

**THE DEVELOPMENT OF VISUO-SPATIAL  
PROCESSING IN CHILDREN WITH AUTISM, DOWN  
SYNDROME AND WILLIAMS SYNDROME**

**Dagmara Annaz**

Thesis submitted for the degree of

**Doctor of Philosophy**

University of London

2006

Birkbeck, University of London, Malet Street, London WC1E 7HX

---

## **ABSTRACT**

This thesis explores visuo-spatial processing in three developmental disorders: autism, Down syndrome (DS), and Williams syndrome (WBS). It is innovative in that it seeks to construct developmental trajectories for visuo-spatial abilities for these children between the ages of 5 and 12. The disorders were compared against a trajectory for typically developing children. Results from a battery of standardised tests were analysed to demonstrate the similarities and differences between the disorders. Study 1 examined individuals' reliance on surrounding face context while recognising individual facial features, taken as a measure of 'holistic' processing, and explored the sensitivity of this skill to rotation. Study 2 examined the ability to perceive featural and configural manipulations to faces when presented in upright or inverted orientations, where sensitivity to configural manipulations in upright faces was taken to be emergence of face recognition expertise. Study 3 examined the ability to recognise a target face in upright and inverted orientations when presented in the child-friendly context of a storybook. Study 4 shifted the focus to construction skills and assessed the ability of individuals to construct a target face from a selection of individual features – in effect, a 'social' version of the Pattern Construction task. The results demonstrated: (1) a lack of emerging face recognition expertise in all the developmental disorders; (2) notable differences between the developmental trajectories of each disorder - they were atypical or delayed in different ways; (3) aversion to eyes in the low-functioning autism group and marked differences between low and high-functioning groups; (4) relatively strong face recognition abilities in WBS influenced on face construction scores; (5) uniform low performance across tasks in the DS group. It is argued that the multiple perspectives offered by cross-syndrome studies of development shed light on the constraints that shape the typical development of face recognition skills.

---

## **ACKNOWLEDGEMENTS**

I am grateful to both my supervisors for their support throughout this project. I am extremely thankful to Dr Michael Thomas who has been a great supervisor, giving me lots of support, guidance and encouragement. I would like to thank Professor Mark Johnson, for giving me the opportunity to work on this project. I am particularly grateful to Professor Annette Karmiloff-Smith for her kind help and for being an inspirational role model for me. It has been a pleasure conducting the research reported here, in no small part due to the excellent company and helpful nature of my colleagues and friends. My thanks go to Dr. Kate Humpreys, Dr. Fiona Richardson, Dr. Gillian Forrester, Roberto Filippi, Dr. Ellen Kotsoni, Steve Newton, Anna Remington, Frank Baughman and Dr. John Swettenham. To my husband Basil, thank you for being so supportive and encouraging over the last 3 years. Finally, I should thank little Amelka, my family and friends for all their support and friendship. I am indebted to all the children who took part in the studies, and their parents and teachers, without whom this thesis would not have been possible. I would also like to thank The Williams Syndrome Foundation, UK. I would like to thank C. Mondloch and N. Brace for providing stimuli for Study 2 and 3. These studies were financed by Birkbeck College.

---

## TABLE OF CONTENTS

Abstract.....	2
Acknowledgements.....	3
Table of contents.....	4
List of figures.....	8
List of tables.....	10
List of abbreviations.....	11
Thesis Overview.....	12
<b>CHAPTER 1: FACE PROCESSING.....</b>	<b>13</b>
1.1 Introduction .....	14
1.2 Are faces special? .....	16
1.2.1 Face Cells.....	16
1.2.2 Prosopagnosia .....	17
1.2.3 Face Recognition versus Object Recognition.....	18
1.2.4 The Face Inversion Effect .....	19
1.2.5 Summary .....	22
1.3 Cognitive theories of face recognition .....	22
1.3.1 Developmental Theories of Face Processing .....	22
1.3.2 The Holistic Hypothesis.....	23
1.3.3 The Configural Hypothesis .....	27
1.3.4 Featural hypothesis .....	34
1.3.5 The Right hemisphere hypothesis .....	35
1.4 Neuro-imaging studies of face processing .....	35
1.5 Summary .....	37
<b>CHAPTER 2: DEVELOPMENTAL DISORDERS.....</b>	<b>39</b>
2.1 Introduction .....	41
2.1.1 Domain specific or general? .....	43
2.1.2 Intact/spared versus impaired .....	44
2.2 Autism Spectrum Disorder (ASD).....	46
2.2.1 Historical Perspective .....	46
2.2.2 Clinical diagnosis.....	46
2.2.3 Prevalence.....	48
2.2.4 Genetics .....	49
2.2.5 Cognitive phenotype.....	50
2.3 Down syndrome (DS).....	62
2.3.1 Historical Perspective .....	62
2.3.2 Genetics .....	62
2.3.3 Clinical diagnosis.....	63
2.3.4 Cognitive Phenotype.....	64
2.4 Williams-Beuren syndrome (WBS) .....	68
2.4.1 Historical Perspective .....	68

---

2.4.2 Genetics .....	68
2.4.3 Clinical diagnosis.....	70
2.4.4 Cognitive phenotype.....	71
2.5 Summary of disorders .....	81
<b>CHAPTER 3: GENERAL METHODOLOGY.....</b>	<b>83</b>
3.1 Introduction .....	84
3.2 General methods adopted in this thesis .....	84
3.2.1 Participants .....	84
3.2.2 Challenges of testing children with developmental disorders .....	90
3.2.3 Stimuli and Apparatus .....	91
3.2.4 General Procedure .....	92
3.3. Data Collection .....	93
3.3.1 Dependent variables.....	93
3.4 Methodological issues and data analysis .....	93
3.4.1 Matching strategies .....	93
3.4.2 Developmental trajectories .....	94
3.4.3 Data analysis .....	95
3.4.4 Floor and ceiling effects.....	96
3.4.5 Definition of delay adopted in the analysis .....	97
<b>CHAPTER 4: STANDARDISED TESTS.....</b>	<b>99</b>
4.1 Introduction .....	100
4.1.1 Questions and Hypotheses.....	100
4.2 British Picture Vocabulary Scale (BPVS) .....	101
4.2.1 Method .....	101
4.2.2 Results.....	102
4.3 Pattern Construction (PC) .....	106
4.3.1 Method .....	107
4.3.2 Results.....	108
4.4 Copying task (CT) .....	111
4.4.1 Method .....	111
4.4.2 Results.....	112
4.5 Benton Facial Recognition task - short version.....	116
4.5.1 Method .....	116
4.5.2 Results.....	118
4.6 Summary of all tests .....	121
<b>CHAPTER 5: DEVELOPMENT OF HOLISTIC FACE RECOGNITION.....</b>	<b>126</b>
5.1 Introduction .....	127
5.2 Study 1: Whole-Part face recognition.....	128
5.2.1 Method .....	130
5.2.1.1 Participants.....	130

---

5.2.1.2 Stimuli.....	130
5.2.1.3 Procedure.....	131
5.2.2 Results.....	133
5.2.1.1 Accuracy.....	133
5.2.1.2 Can the Benton test predict accuracy on the Rotating faces task?.....	145
5.2.1.3 Summary of accuracy results.....	146
5.2.1.3 Response Times.....	148
5.2.3 Discussion .....	152
<b>CHAPTER 6: DEVELOPMENT OF CONFIGURAL &amp; FEATURAL FACE RECOGNITION.....</b>	<b>158</b>
6.1 Introduction .....	159
6.2 Study 2: Jane Faces .....	160
6.2.1 Method .....	162
6.2.1.1. Participants.....	162
6.2.1.2 Stimuli.....	162
6.2.1.3 Procedure.....	163
6.2.2 Results.....	164
6.2.2.1 Identity Recognition .....	165
6.2.2.2 Difference Detection.....	166
6.2.2.3 Summary of difference detection accuracy .....	177
6.2.2.4 Can the Benton test predict accuracy on the Jane task?.....	178
6.2.3 Discussion .....	180
6.3 Study 3: Storybook face recogniton task .....	183
6.3.1 Method .....	184
6.3.1.1 Participants.....	184
6.3.1.2 Stimuli.....	184
6.3.1.3 Procedure.....	185
6.3.2 Results.....	186
6.3.2.1 Summary of results .....	196
6.3.2.2 Can Benton test predict performance on the Storybook task?.....	196
6.3.3 Discussion .....	198
<b>CHAPTER 7: DEVELOPMENT OF FACE CONSTRUCTION ABILTIES.....</b>	<b>200</b>
7.1 Introduction .....	201
7.2 Study 4: Face Constuction task .....	202
7.2.1 Method .....	203
7.2.1.1 Participants.....	203
7.2.1.2 Stimuli.....	204
7.2.1.3 Procedure.....	205
7.2.2 Results.....	206
7.2.3 Benton task and Pattern Construction .....	219
7.2.4 Summary .....	221
7.2.4 Discussion .....	221

---

<b>CHAPTER 8: GENERAL DISCUSSION AND CONCLUSIONS.....</b>	<b>225</b>
8.1 Introduction .....	226
8.1.1 The Development of Holistic Face Recognition.....	227
8.1.2 Development of Configural and Featural face recognition .....	228
8.1.3 Visuo-Construction skills in face processing. ....	230
8.1.4 Featural processing and feature salience .....	231
8.2 Developmental profiles of disorder groups .....	232
8.2.1 Can developmental level explain the results?.....	237
8.2.2 Contribution of developmental trajectories.....	238
8.3 Conclusions .....	239
8.4 Further research .....	242
<b>REFERENCES.....</b>	<b>244</b>
<b>APPENDICES.....</b>	<b>269</b>
Appendix A: Distribution of CARS scores for autism groups.....	270
Appendix B: Trajectories of Whole-Part performance.....	272
Appendix C: Featural-Configural analyses.....	281
Appendix D: Trajectories of Face construction task.....	284

---

## LIST OF FIGURES

Figure 1.1: Example of aligned and misaligned halves of different identities.....	20
Figure 1.2: Example of the ‘Thatcher illusion’ .....	21
Figure 1.3: Example of whole-part stimuli.....	24
Figure 1.4: Graphic representation of processes involved in face recognition.....	39
Figure 2.1: Example of drawings and block design by an adult with DS.....	66
Figure 2.2: Schematic diagram of the small region of chromosome 7 deleted in WBS....	70
Figure 2.3: Illustrates an example of WBS performance on the Navon task.....	79
Figure 3.1: Age distributions of each group.....	88
Figure 3.2: Graphic description of developmental delay.....	98
Figure 4.1: BPVS age equivalent scores for all groups plotted against their CA.....	102
Figure 4.2: LFA group developmental trajectory shown with outliers.....	104
Figure 4.3: Example of PC trial.....	107
Figure 4.4: PC age equivalent scores for all groups plotted against their CA.....	108
Figure 4.5: Copying test age equivalent performance for each group.....	112
Figure 4.6: Example of Benton trial.....	117
Figure 4.7: Developmental trajectories for Benton tests performance.....	118
Figure 4.8: Summary of standardised tests.....	121
Figure 5.1: Example of the whole-part stimuli.....	131
Figure 5.2: TD group trajectories for accuracy scores on the whole-part.....	136
Figure 5.3: TD group trajectories accuracy scores on whole-face with confidence intervals.....	137
Figure 5.4: HFA group trajectories for accuracy scores on the whole-part.....	139
Figure 5.5: LFA group trajectories for accuracy scores on the whole-part.....	141
Figure 5.6: DS group trajectories for accuracy scores on the whole-part.....	142
Figure 5.7: WBS group trajectories for accuracy scores on the whole-part.....	144
Figure 5.8: % accuracy scores on each feature for each group.....	147
Figure 5.9: % accuracy on whole-part recognition trials for each group.....	148
Figure 5.10: TD group trajectories for RT scores on the whole-part.....	149
Figure 5.11: HFA group trajectories for RT scores on the whole-part.....	150
Figure 5.12: LFA group trajectories for RT scores on the whole-part.....	150
Figure 5.13: DS group trajectories for RT scores on the whole-part.....	151
Figure 5.14: WBS group trajectories for RT scores on the whole-part.....	151
Figure 6.1: Examples of ‘Jane’ faces stimuli.....	163
Figure 6.1: TD group trajectory for accuracy scores on the Jane faces.....	167

---

Figure 6.2: Linear regressions with confidence intervals in upright and inverted orientation for the TD group on configural set.....	168
Figure 6.3: HFA group trajectories for accuracy scores on the Jane faces.....	170
Figure 6.4: LFA group trajectories for accuracy scores on the Jane faces.....	172
Figure 6.5: DS group trajectories for accuracy scores on the Jane faces.....	173
Figure 6.6: WBS group trajectories for accuracy scores on the Jane faces.....	175
Figure 6.7: Developmental trajectory for accuracy scores on the Jane faces task against Benton age.....	179
Figure 6.8: Example of the Storybook stimulus used in the study.....	185
Figure 6.9: TD group trajectory for accuracy scores on the Storybook.....	186
Figure 6.10: TD group trajectory for RT on the Storybook task.....	187
Figure 6.11: HFA group trajectory for accuracy scores on the Storybook.....	188
Figure 6.12: HFA group trajectory for RT on the Storybook.....	189
Figure 6.13: LFA group trajectory for accuracy scores on the Storybook.....	190
Figure 6.14: LFA group trajectory for RT on the Storybook.....	191
Figure 6.15: DS group trajectory for accuracy scores on the Storybook.....	192
Figure 6.16: DS group trajectory for RT on the Storybook.....	193
Figure 6.17: WBS group trajectory for accuracy scores on the Storybook.....	194
Figure 6.18: WBS group trajectory for RT on the Storybook.....	195
Figure 7.1: Example of the construction stimuli.....	204
Figure 7.2: Example of a typical testing procedure on construction task.....	205
Figure 7.3: TD group trajectories on accuracy of feature choice.....	208
Figure 7.4: TD group trajectories on accuracy on 1 <sup>st</sup> -order-configuration.....	209
Figure 7.5: HFA group trajectories on accuracy on feature choice.....	210
Figure 7.6: HFA group trajectories on accuracy on 1 <sup>st</sup> -order-configuration.....	211
Figure 7.7: LFA group trajectories on accuracy of feature choice.....	212
Figure 7.8: LFA group trajectories on accuracy of 1 <sup>st</sup> -order-configuration.....	213
Figure 7.9: Example of a face construction.....	213
Figure 7.10: DS group trajectories on accuracy of feature choice.....	215
Figure 7.11: DS group trajectories on accuracy of 1 <sup>st</sup> -order-configuration.....	216
Figure 7.12: WBS group trajectories on accuracy of feature choice.....	217
Figure 7.13: WBS group trajectories on accuracy of 1 <sup>st</sup> -order-configuration.....	218
Figure 7.14: Example of incorrect placement of eye feature.....	218
Figure 8.1: Schematic representation of face recognition in HFA group.....	233
Figure 8.2: Schematic representation of face recognition in LFA group.....	234
Figure 8.3: Schematic representation of face recognition in DS group.....	235

---

Figure 8.4: Schematic representation of face recognition in WBS group.....	236
Figure 8.5: Examples of photos.....	241

## LIST OF TABLES

Table 1.1: Description of holistic, configural and featural terms.....	15
Table 2.1: Summary of visuo-spatial profiles of each group.....	82
Table 3.1: Summary information on participants excluded from the studies.....	87
Table 3.2: Chronological age details for all groups.....	88
Table 3.3: Classification of parental SES (%).....	89
Table 3.4: Details of parental education level (shown in %).....	90
Table 4.1: Examples of copying.....	116
Table 4.2: Summary of standardised results.....	121
Table 5.1: Summary of each group's accuracy levels.....	135
Table 5.2: Summary of each group's performance on whole-part paradigm against Benton test age.....	146
Table 5.3: Summary of each group's performance on whole-part against CA.....	147
Table 5.4: Median RT for all the groups on whole-part paradigm.....	152
Table 6.1: Means and SE for accuracy % in Identity Recognition.....	166
Table 6.2: Means and SE for accuracy % in Difference Detection.....	166
Table 6.3: Summary of each group's performance on Jane task plotted against CA.....	177
Table 6.4: Summary of each group's performance on Jane task plotted against Benton..	178
Table 6.5: Summary of each group's performance plotted against CA.....	196
Table 6.6: Summary of each group's performance on Storybook against Benton age test.....	197
Table 7.1: Summary of % accuracy on individual features.....	207
Table 7.2: Summary of results against Benton test age.....	220
Table 7.3: Summary of results against PC age.....	220
Table 7.4: Summary of results for the Construction task.....	221

---

## **LIST OF ABBREVIATIONS USED IN THIS THESIS**

ASD = Autism Spectrum Disorder

CA = Chronological Age

CARS = Childhood Autism Rating Scale

DS = Down Syndrome

ERP = Event-Related Potential

FFA = Fusiform Face Area

HFA = High Functioning Autism

fMRI = Functional Magnetic Resonance Imaging

LFA = Low Functioning Autism

MA = Mental Age

NVMA = Non-Verbal Mental Age

PC = Pattern Construction

RT = Response Time

TD = Typically Developing

VMA = Verbal Mental Age

WBS = Williams-Beuren Syndrome

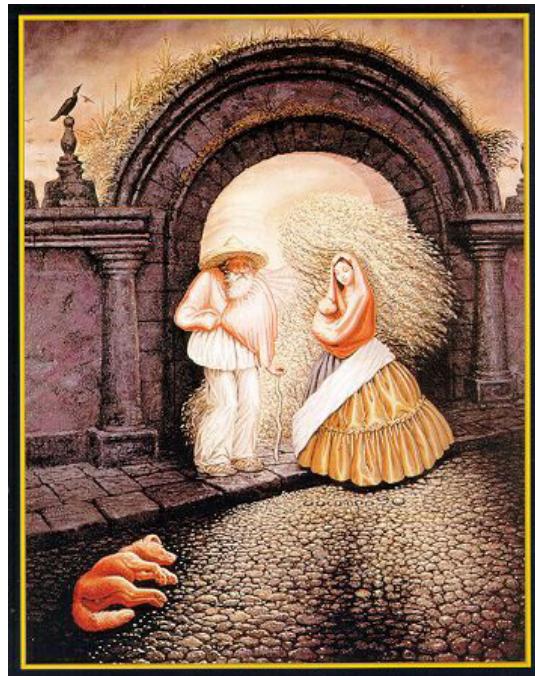
---

## **THESIS OVERVIEW**

The main aim of this thesis was to investigate the development of face processing in children with developmental disorders within configural and holistic framework. This thesis is divided into eight chapters. Chapters 1 and 2 review current theories of face processing in typically developing children and adults, and Chapter 2 discusses these findings in developmental disorders literature, and how it might be applied to development. Chapter 3 presents a brief overview of the methods used in this thesis, including the experimental paradigms, methods used for data collection and stimulus preparation, and the statistical approach employed. Chapter 4 presents developmental data from standardised tests and discusses each group's profile. Chapter 5 investigates whether children with developmental disorders show increased sensitivity to holistic face recognition, and compares their profiles to the TD group. Chapter 6 investigates configural face recognition and how it develops over time in TD group, and then assesses disorder groups. Chapter 7 uses a visuo-constructive task to investigate face processing. Finally in Chapter 8 the experimental results are discussed in the light of current theories of face processing and predictions. Further suggestions are made for future research.

## **CHAPTER 1**

### **FACE PROCESSING**



## **1.1 INTRODUCTION**

Faces are amongst the most important stimuli for human social functioning and have been described as the 'single most important pattern in our environment' (Ellis & Young, 1989, p. 7). Faces encompass a multitude of meaningful information, including age, race, gender, emotions, as well as feature configuration. The information that can be extracted from faces is crucial for human daily lives and social communication.

In the last decade or so, a great deal of research within the areas of psychology and neuropsychology has been devoted to the understanding of our particular face recognition abilities. Much of the interest in face processing arose from the observation of individuals who were unable to recognise faces due to acquired brain damage, a condition known as Prosopagnosia - while retaining other visuo-spatial abilities such as object recognition (e.g., de Renzie, 1986). Conversely, Farah (1991) described patients who had impairments in object and word recognition but normal facial recognition, a condition known as Agnosia. The significance of the double dissociation, combined with the importance of facial stimuli in our lives, created a huge surge to formulate theories of facial recognition and its underlying mechanisms.

Nowadays, it is widely accepted that the visual perception of faces can involve different underlying processes and strategies, which are complex and fine-tuned. These can change depending on the developmental stage of an individual and, most importantly, they can follow atypical developmental pathways in developmental disorders.

The literature on face recognition is vast and covers aspects that are beyond the scope of the current thesis. This chapter will focus on why faces are important to study and how face recognition skills develop. Particular attention will be given to the literature on face processing theories and experimental studies that test the holistic, configural, and featural dimensions of face processing and their development. The ambiguous use of terms such as configural and holistic created much chaos and different opinions as to what is meant by each term. Sometimes they are used interchangeably, sometimes to refer to different behavioural phenomena.

Although distinction of these terms is not simple, definitions used in the current thesis are outlined in Table 1.1.

Table 1.1: Description of the terms.

<u>TERM</u>	<u>DEFINITION</u>	<u>OTHER TERMS USED</u>
<b>FEATURAL</b>	Recognising by the individual face features	local, piecemeal, part-based, componential
<b>CONFIGURAL</b>		
<b><i>First-order</i></b>	Basic arrangements of the parts (i.e. eyes above nose)	
<b><i>Second-order</i></b>	Recognising small differences between individuals in the spacing among the facial features, (e.g., distance between the eyes or between the mouth and chin)	global, holistic
<b>HOLISTIC</b>	face features are integrated into a whole or Gestalt-like representation and stored as an unparsed perceptual whole without	global, coarse, configural template-matching

The current thesis is aimed to provide an in-depth analysis of face processing development in children with developmental disorders: autism, DS and WBS. Since the major theoretical debate concerns whether face recognition develops normally in autism, DS and WBS, it will be our contention that explanations must be couched in terms of developmental trajectories (Karmiloff-Smith, 1998; Karmiloff-Smith, et al., 2004). The developmental trajectory approach to disorders offers more insight with respect to the way in which development may have proceeded over time in a deviant fashion, even though the behavioural proficiency may end up similar to controls.

In order to address the question, the following aims have been set out: i) to establish developmental profile of each group's face recognition performance; and ii) to use a cross-syndrome comparisons on face processing tasks to investigate different constraints that may exist. Comparisons will relate the performance to chronological age in both the control and clinical samples and, in the latter, also between experimental face processing tasks and other background measures discussed in chapter 4 (see Karmiloff-Smith et al., 2004; Thomas et al., 2001, for a similar analytical approach).

## **1.2 ARE FACES SPECIAL?**

### **1.2.1 Face Cells**

One line of evidence for the neuroanatomical specialisation of face recognition comes from single-cells recordings with non-human primates. In their seminal studies, Gross and colleagues (Gross, Bender & Rocha-Miranda 1969; Gross, Rocha-Miranda, Bender, 1972) reported that some cells within the temporal cortex of macaque monkeys responded selectively to complex shapes, such human and monkey faces, providing the first evidence for face selective neurons. These findings were followed by a number of investigations that extended the initial results greatly. For instance, ‘Face neurons’ were found in areas such as Superior Temporal Sulcus (STS) and inferior temporal cortex (Rolls, 2004; Rolls & Tovee, 1994).

It was established that some cells are between two to ten times more responsive to faces than to geometrical stimuli or three-dimensional objects (hence the name ‘face cells’), (Perrett, Oram, Hietanen & Benson, 1994). Some cells in the upper region of STS appeared to be most responsive to facial expression and gaze direction and to be mainly unaffected by face identity.

Furthermore, face cells have been found to be sensitive to the overall configuration of the face (i.e., to the spatial arrangement and separation of the features in the face). They do not respond to scrambled faces (Perrett, Oram & Ashbridge, 1998), so it appears that the features themselves are not enough to trigger responses. In contrast, cells responsive to non-face objects appeared to respond to parts of objects rather than the overall configuration (Farah, 2000).

### **1.2.2 Prosopagnosia**

Another line of evidence for the anatomical specialisation of face recognition abilities was triggered by reports of adult patients suffering from Prosopagnosia, a rare neurological disorder, who lost their ability to recognise familiar faces while maintaining normal object recognition skills (Bodamer, 1947; de Renzie, 1986). Prosopagnosia commonly results from damage to the region of cortex that lies between the temporal and occipital cortex. Recently, two types of prosopagnosia

have been distinguished due to their nature of causality: adulthood and developmental prosopagnosia.

The individuals with acquired prosopagnosia had normal face recognition abilities before they suffered from head trauma, stroke, or degenerative diseases. They show damage to the ventral occipitotemporal and temporal cortices. For example, in a case study, patient LH had damage to an area within the occipitotemporal cortex and experienced impaired face recognition while recognition of objects was described as normal. ERP (Event-related potential) studies of acquired prosopagnosia revealed that these individuals do not display a normal N170 component<sup>(1)</sup>. Eimer and McCarthy (1999) found that the N170 was completely absent in a severely prosopagnosic patient (PHD) in response to upright or inverted faces. It has been suggested that patients with prosopagnosia rely on a feature-based strategy to construct face representations in the way that objects are processed. However, since face recognition is impaired, there must be some properties of faces such that this strategy is insufficient.

In contrast, individuals with developmental prosopagnosia never developed normal face abilities, either due to a genetic condition, brain damage (pre-natally or in early childhood) or unknown causes. It has been reported that some individuals with developmental prosopagnosia have been diagnosed with autism, due to mismanagement of social situations caused by their inability to recognise faces (Duchaine & Nakayama, 2004). A small number of individuals with developmental prosopagnosia appear to be associated with more general visual processing problems (de Haan & Campbell, 1991). Functionally, the deficit appears similar to many patients with the acquired form of the disorder, but structural imaging has shown no gross structural abnormalities in occipitotemporal cortex (Barton, Cherkasova, Press, Intriligator & O'Connor, 2003). On the basis of case studies of patients with prosopagnosia, some researchers have argued for an innately dedicated face processing system (e.g., Kanwisher, Stanley & Harris, 1999). This was further supported by rare cases of patients with agnosia, whose face recognition ability was normal but object recognition was impaired (e.g., Moskovitch, Winocur &

---

<sup>1</sup> The most widely reported is an early face sensitive negative component over the lateral temporal and occipital sites at about 170 ms post-stimulus presentation, which has been termed the 'N170'.

Behrmann, 1997). For instance, patient C.K. appeared to have retained ability to recognise faces but showed a profound visual agnosia for everyday objects. He recognised upright non-scrambled faces in photographs, line drawings, caricatures and cartoons as good as controls but could only recognise about 50% of non-face line drawings (Behrmann, Moskovitch & Winocur, 1994). However Gauthier & Tarr (1997) have argued that even in cases such as that of CK, it is possible that impairments in face processing would be found if variables such as response times or biases were investigated instead of simply the number of correct recognitions.

### **1.2.3 Face Recognition versus Object Recognition**

The findings by Gross and colleagues (Gross et al., 1969; Gross et al., 1972) of ‘face neurons’ was reinforced by the studies on newborns. Johnson and colleagues (Johnson, Dziurawiec, Ellis & Morton, 1991) were the first researchers to propose that neonates have an ‘innate face-like representation’ mechanism. In their study of newborn babies, they found that neonates preferred looking at faces over other patterns (Johnson, et al., 1991), and in 6-month olds, faces were distinguished from other highly familiar objects (de Haan & Nelson, 1999).

Another line of evidence came from several behavioural and neuroimaging studies, which indicated that, as we gain more visual experience with one class of objects, those objects may gain certain visual privileges (e.g., Gauthier, Tarr, Anderson, Skudlarski & Gore, 1999; see Tarr & Cheng, 2003 for discussion), such as speed and potentially, orientation sensitivity.

Gauthier and colleagues (Gauthier & Tarr, 1997) argued that faces are unique, as they are the most commonly encountered case of expert subordinate visual recognition (that is expertise in discriminating between different components in the same category). In one of their studies, participants were trained to become experts at discriminating members of a novel class of stimuli named ‘Greebles’. These stimuli are visual patterns that share common structure and are recognised by name. The authors found that trained participants showed the hallmark of expertise, i.e., faster recognition of individual greebles than novice participants. This advantage disappeared when greebles were presented in inverted orientation, an effect also

found in face recognition (see next section). It is certainly the case that experiences shape our perceptual and neural processing of the visual system, as in the case of recognising words (Shaywitz et al., 2004), non-native faces (Sangrigoli & de Schonen, 2004), and items with which we have unique expertise (e.g., Gauthier & Tarr, 1997; Gauthier et al., 1999).

Face-object discrimination has been studied to a greater extent in adult populations and primates. It is thought that in adult populations, faces gain special entry into the visual processing stream relative to non-face objects (e.g., Allison, Puce, Spencer & McCarthy, 1999). Non-face objects such as rearranged faces, letters, and random dots, have been shown to require longer presentation times to be processed than do faces. For example, Eimer showed that faces are distinguished from other objects early in the visual stream, within the first 200 msec (Eimer, 2000), suggesting a special clearance in the visual system only for faces. Most theories of visual object processing suggest that objects are recognised by feature analysis, which seems to be adequate for distinguishing heterogeneous class. However, recognition of faces, which are largely homogenous stimuli, requires fine within-category individuation, so that feature-by-feature recognition appears insufficient (e.g., Diamond & Carey, 1986; see Tarr & Cheng, 2003, for a review).

#### **1.2.4 The Face Inversion Effect**

This section will draw attention to the three effects that illustrate a specialisation for the recognition of upright faces: i) the face inversion effect, ii) the Thatcher illusion, and iii) the face composite illusion.

In order to investigate whether inversion particularly affects the recognition of faces, Yin (1969) used a forced-choice recognition paradigm with pictures of human faces, airplanes, houses, and stick figures of men in motion as stimuli. In one condition the stimuli were learnt and tested in the upright orientation. In another condition the stimuli were learnt in the upright orientation and then tested in the inverted orientation. Generally, when the stimuli had to be recognised in the inverted position, error rates increased for all stimuli. Interestingly, in the upright orientation, performance was disproportionately higher for faces when compared with the other

objects. However, while faces were recognised best in the upright orientation, performance on inverted faces dropped below the recognition levels of the other object classes. The finding that upside-down faces are disproportionately more difficult to recognise than other inverted objects has been referred to as ***the face inversion effect***. Since then, the inversion effect has been considered as the hallmark of face-specific recognition and is often used as a marker of expertise in face processing.

One paradigm that has been extensively used to investigate the inversion effect, uses face composites made from the top half of one faces and the bottom half of another face (e.g., Hole, 1994; Young, Hellawell & Hay, 1987), shown in Figure 1.1. Young and colleagues (1987) used pictures of famous people and found that adults were slower and less accurate at naming the top half, presumably because the two halves of the face formed a completely new ‘gestalt’ which was hard for participants to break down. However, their performance improved when two halves of the faces were misaligned. Hole (1994) found that the ***composite effect*** was not seen when the faces were aligned yet inverted or when the exposure time for aligned upright faces was longer, as participants were able to use a feature-by-feature comparison (Hole, 1994). The composite face effect paradigm has often been used to test holistic processing, which is described in section 1.3.2 of this chapter.



Figure 1.1: Example of aligned and misaligned halves of different identities.

Another impressive demonstration of orientation sensitive face recognition is the '**Margaret Thatcher illusion**' task (Thompson, 1980). The illusion was created by rotating facial features, such as the eyes and the mouth within the context of the face. The modified picture of the face appeared grotesque, yet when viewed upside-down the face appeared normal. See Figure 1.2.

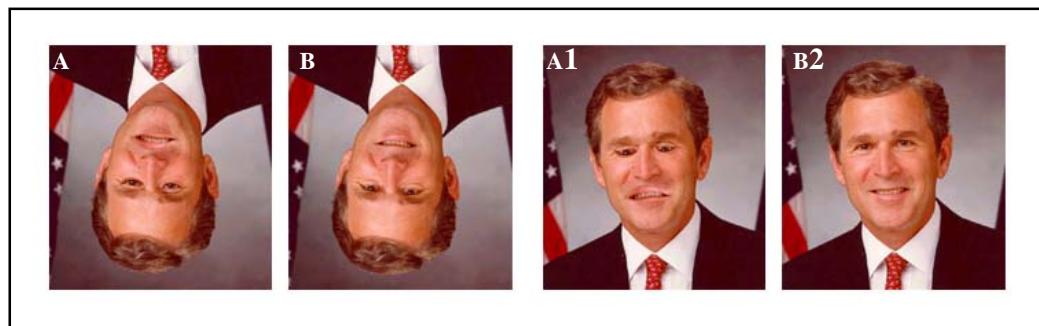


Figure 1.2: Example of the 'Thatcher illusion', here with George W. Bush: Grotesque faces (A & A1), and normal faces (B & B1).

Several studies have found evidence of face inversion effects even in infancy (e.g., de Haan & Nelson, 1998). For example, neonates prefer to track upright rather than inverted face-like configurations (e.g., Johnson, et al., 1991). However, this could be attributed to disruption of the first order relations of the face-like configuration, thus rendering it inoperative as a trigger for the newborns' hypothesised innate tendencies to attend to faces (Johnson et al., 1991), (see next section).

The studies discussed so far demonstrated that faces are orientation sensitive, but why is it? Rock (1988) provided one explanation. In his view (mental rotation hypothesis), recognition of inverted faces overtax a mental rotation mechanism. Rotated faces have to be processed by mentally rotating face-features one after another, which makes it difficult for configural or holistic information to recover (see Diamond & Carey, 1986, for a similar view).

### 1.2.5 Summary

In summary, based on the cases of patients with prosopagnosia and other empirical results such as inversion effect, one can conclude that faces are ‘special’ as a result of a dedicated face-processing module (Farah, 1996; Kanwisher, McDermott & Chun, 1997). Others regard this exceptional human ability as arising through an interaction of innate basic preferences and experience from the environment (Diamond & Carey, 1986; Gauthier & Nelson, 2001; Johnson et al., 1991). Inversion effect has proved to be a crucial tool in investigating face recognition, and will be discussed in many studies throughout this chapter, as well as being used for some of the experimental studies in the current thesis.

## 1.3 COGNITIVE THEORIES OF FACE RECOGNITION

This section reviews the behavioural evidence from adults and children that has been used to construct cognitive theories of face recognition. These revolve around three putative types of processing: holistic, configural and featural. However, a developmental account that draws a distinction between innate and learnt aspects of the system will be discussed first.

### 1.3.1 Developmental Theories of Face Processing

One of the earliest theories on early face recognition was proposed by Morton and Johnson (1991). According to their ***CONSPEC*** & ***COLERN*** theory, early face processing is driven by innate factors, and at later stages the processing is driven by interactions between these innate factors (Conspec) and a learning-based system (Colern). According to the theory, Conspec is mediated by subcortical visuomotor pathways, and is an early primitive mechanism that is triggered once an infant first views faces. Conspec responds exclusively to faces and ensures that they are a frequent input to the Colern mechanism that is cortically mediated. Johnson (1991) suggested that Conspec declines approximately from the age of 2 months and is replaced by Colern upon cortical maturity. Recently, Johnson (2001) updated this theory and proposed the ***Interactive Specialisation*** theory to explain the role of fusiform face area (the putative site of the cortical mechanism). He suggested that early in development, infants have diffuse cortical specialisation for faces, which

becomes more specific with environmental exposure to faces, and the face-processing system becomes more specialised and localised. Similarly, Nelson (2001) in his ‘perceptual narrowing’ approach emphasises the role of environment, suggesting that infants shape their face processing system based on the visual experience they encounter. He proposed that the face recognition system develops from a broadly-tuned non-specific recognition to a fine-tuned to human face-discrimination system.

### 1.3.2 The Holistic Hypothesis

According to one hypothesis face processing relies more on *holistic* representations than does the processing of other stimuli (Diamond & Carey, 1986; Farah, Wilson, Drain & Tanaka, 1998; Farah, et al., 1995; Tanaka & Farah, 1993). This proposal is based on the idea of *Gestalt psychology* that the ‘whole’ is more than the sum of its parts, and such that processing is translated into a template-like or norm-based. According to this position, a face is seen as an organised, meaningful pattern, and is difficult to break down into its parts without harming perception and encoding of a face and its features.

Evidence for the holistic type of processing comes from a number of experimental paradigms. One of the most influential study was designed by Tanaka and Farah (1993), who operationalised the concept of holistic processing by developing a task in which participants were presented with a series of faces and were tested on their recognition of features such as eyes, mouth, or nose. The test criteria consisted of showing the feature in isolation and showing it within the context of the whole face, hence the name ‘Whole-Part Paradigm’. The stimuli are illustrated in Figure 1.3. The trials were shown in upright and inverted orientations. Initially, adult participants ( $n = 20$ ) took part in the learning session (time not specified), to learn names of the faces. During the task proper, they were presented with a whole face or an isolated face feature and were asked whether they had seen the face or feature previously. The results showed that in the upright orientation, individuals were more accurate on the features presented in the whole face (74%) than in isolation (65%). However, when features were presented inverted, the performance on whole-face decreased considerably (65%) whereas accuracy on the part-faces was not influenced by inversion (64%) conditions. Thus an inversion effect was only observed in the

whole-face trials. In addition, Tanaka and Farah (1993) tested participants on scrambled faces and house stimuli. In the house condition, presentation of the stimuli in different orientations had no influence on their performance. In contrast to the whole-part face paradigm, there was little difference between the recognition of the houses in the whole-house and part-house conditions (81% and 79% respectively). The authors proposed that there is a face-specific recognition system that processes holistic information from upright faces (see Farah, 1996; Kanwisher, McDermott & Chun, 1997, for similar argument). On the other hand, objects and inverted faces are processed in a featural manner using a more general-purpose object recognition system.

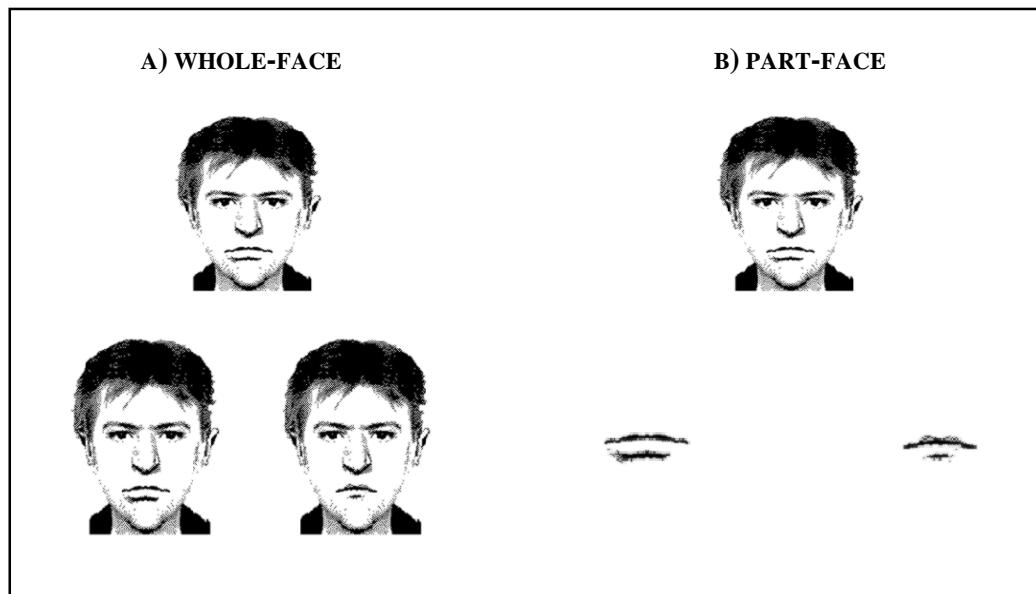


Figure 1.3: Example of whole-part stimuli. Upper stimulus is a target: the participant must select which of the lower two photos matches with the target. Adapted from Tanaka and Farah (1993).

Recently, Lewis and Glenister (2003) have extended the whole-part paradigm to face recognition at 90 degrees orientation, to determine whether there are any holistic encoding changes at the intermediate level of rotation. They tested 21 adults and found that the advantage of recognising a feature in the context of the whole-face decreased gradually with rotation and the inversion effect was found when orientation was past 90 degrees rotation. The effect found in many previous studies

(e.g., Tanaka & Farah, 1993). Moreover, rotation affected recognition of features presented in a context of a whole face and without context. Lewis and Glenister (2003) interpreted this result as configural disruption (configuration within each feature) of isolated features. This finding contradicts the originally reported findings by Tanaka and Farah (1993) and more recent study by Joseph and Tanaka (2003).

Palermo and Rhodes (2002) examined the role of attention in holistic face recognition. They modified the whole-part task by adding an attention condition (experiment 2). In this condition, participants were presented with a target face bounded by two ‘distracter’ faces. Adult participants ( $n=72$ ) were asked whether they had seen the target face previously and if the distracter faces were same or different. Unlike Tanaka and Farah (1993) there was now no accuracy difference between the whole-face and part-face in the attention condition. It was concluded that holistic face recognition is an attentionally demanding mode of processing.

The holistic processing style has also been observed in studies of the perception of emotional expressions. If the top half of a face shows one expression and is fused with the bottom of a face showing a different expression, recognition of the composite parts is slower compared to when the face halves are misaligned (Calder, Young, Keane & Dean, 2000). This suggests that recognition of facial expression is also associated with holistic processing.

### ***1.3.2.1 The Development of Holistic Encoding***

The composite-effect and whole-part paradigms have also been used to study the development of holistic processing in typically developing children and children with developmental disorders (see Chapter 2 for discussion for the latter). Carey and Diamond (1994) used the composite paradigm with two groups of children (mean age in group one: 7:1, and group two: 10:9) and a group of adults (mean age: 28:3). The children were asked to name the upper part of the face while the lower part of the face belonged to a different person. The task was shown in upright and inverted orientations. Carey and Diamond found that the face composite effect was as strong in 6-year-old children as in adults, suggesting early development of holistic processing.

Tanaka, Kay, Grinnell, Stansfield and Szechter (1998) tested children (age groups: 6, 8 and 10 year olds) using the whole-part paradigm. In their task, children were asked to memorise the faces with the names assigned to them. The children were then shown the feature of the memorised face in a whole face context or in isolation. The authors observed a similar pattern of performance as that previously observed in the adult studies i.e., a whole-face advantage across all groups. Similarly, when Mondloch, Le Grand and Maurer (2002) used the contour condition (set of faces created by placing same internal face features within the outer contour of different faces) to test development of holistic encoding, they found adult-like levels of holistic processing in all age groups tested. These findings suggest that holistic face processing reaches adult-like levels relatively early, at least by 6 years of age, the youngest group tested in these studies.

This conclusion was partially supported by Hay and Cox (2002), who asked 6 and 9 year olds to match photographs of either whole-face or part-face features. Children were presented with pictures of varied school-friends and unfamiliar children in different orientations. Face processing showed marked improvement with age. There was an emergence of the face inversion effect in the older group (9 years old), which was not present in the younger children (6 years old). It was also found that the younger group displayed better recognition of the eye regions in isolation compared to the older group.

A developmental trend from featural to holistic processing of faces and an effect of face inversion with increasing age has been demonstrated in another categorisation study carried out by Schwarzer (2000). Children between 6 and 10 years and adults control group were asked to sort upright and inverted line drawings of faces into two categories (1 = adult's face; 2 = child's face). Face categories were based on either similarity of face features (eyes, nose or mouth) or overall similarity of the faces. The results suggested that the 6-to-7-year-olds categorised faces using featural processing (similarity of face features) regardless of the face orientation, whereas the 10-year-olds showed an increase in the use of holistic categorisation (similarity of overall face stimulus) of the upright faces. The adult group used mostly holistic processing for the upright faces and featural for the inverted faces.

Le Grand and others have examined the role of early visual experience in the development of holistic processing using the composite paradigm (Le Grand, Mondloch, Maurer & Brent, 2003). They compared the size of composite-face effect in normal control adults and 12 patients treated for bilateral congenital cataracts (9 to 23 years old). Consistent with the previous studies, a strong composite-face effect was found in the control group and their performance increased by 30% on misaligned trials. In contrast, the patient group did not show the effect, and surprisingly performed better on aligned faces than the control group. Thus it seems that lack of experience with human faces during infancy can cause disturbance in the development of normal holistic processing.

While the above studies have focused entirely on face recognition, some investigators have argued that so-called ‘face holism’ is not exclusive only to faces (see Donnelly & Davidoff, 1999), but can be expanded to many other complex objects and objects of expertise (Gauthier & Logothetis, 2000).

### **1.3.3 The Configural Hypothesis**

An alternative or complement to the holistic face processing hypothesis, emerged in the form of configural face processing hypothesis. According to this proposal face recognition is dependent on both: the features of the face and the way in which they are arranged (their configuration).

Diamond and Carey (1986) suggested that there are two types of configural information (see also Bruce, 1988, and recently Maurer, Le Grand, & Mondloch, 2002). The first one is termed sensitivity to first-order relations and describes seeing faces with their features arranged in relation to each other: eyes above the nose and mouth below the nose. The relations are called first-order because a term such as ‘above’ specifies no distances and therefore incorporates a range of absolute relationships. This arrangement is one to which human infants appear to be sensitive to, from early development (Johnson, et al., 1991). One way of assessing sensitivity to first order relations is to present stimuli such as Mooney faces, which are images of human faces rendered in photographic half tone (Mooney, 1957). This

manipulation preserves the relationship between the main features while eliminating much of the detail.

It is uncontroversial that faces belong to a relatively homogeneous visual category (this does not apply only to human faces). It is therefore argued that recognition of individual faces requires more specific encoding than first-order sensitivity or holistic encoding<sup>(2)</sup> (see Tarr & Cheng, 2003, for a review). Sensitivity to second-order relations, also known as *configural* or *second-order configural* encoding, permits us to individuate faces based on sensitivity to the spatial distances among internal features, for example the distance between the eyes and nose. This type of face encoding is associated with ‘expert’ recognition in adults and maturity of the face recognition system. It is of interest to elucidate, the transitional stage at which children achieve this expert configural encoding.

The principal behavioural evidence that leads to the postulation of second-order configural processing is inversion effect and configural stimuli manipulation. The former (described previously, section 1.2.4) relies on the disruption of configural processing through image inversion. Several studies suggest that individuals are forced to use slower and less efficient featural face encoding when faces are inverted (e.g., Leder & Bruce 2000; Leder, Candrian, Huber & Bruce, 2001; Valentine, 1988). The later technique relies on a more direct measure of sensitivity to configural information by altering the distances between the features of face stimulus (Freire, Lee & Symons, 2000; Leder & Bruce, 1998, 2000; Leder, et al., 2001; Le Grand, Mondloch, Maurer & Brent, 2001; Mondloch et al. 2002). Thus sensitivity to configural information is indeed disrupted by inverting the face stimuli and second-order stimuli manipulation, whereas the effect of local changes, such as blackening teeth, is relatively unaltered (e.g., Leder & Bruce, 1998; Leder & Bruce, 2000; Searcy & Bartlett, 1996).

---

<sup>2</sup> Holistic processing therefore comes with some limits on the acuity with which templates can be discriminated.

### **1.3.3.1 Configural Face Processing in Adults**

Face processing in adults is thought to rely on second-order configural processing and processing of the features of the face has been dismissed as relatively unimportant (e.g., Bartlett & Searcy, 1993; Diamond & Carey, 1986; Rhodes, 1993). For instance, Mondloch et al. (2002) tested adults on featural and configural conditions in upright and inverted conditions. The participants showed a larger inversion effect for the configural condition than they did for the featural one. This study was adapted in the current thesis, thus full details of the study will be discussed in chapter 6 (Jane Faces, study 2).

Recently, in an elegant study developed by Ge and collaborators (Ge, Wang, McCleery, & Lee, *in press*), configural face recognition was investigated by comparing Chinese written symbols to face stimuli. Chinese symbols are learnt from very early age and most Chinese people are highly familiar with them. Also, like face Chinese characters contain featural and configural information and are recognised at the individual level. However, featural manipulation changes the meaning of a character, but configural manipulation does not alter identity of a Chinese character. The authors hypothesised that adult participants ( $n=16$ ) should show an inversion effect when tested on the face condition but should not show an inversion effect when tested on Chinese characters due to the lack of importance of configural information in recognising Chinese characters. They suggested that inversion effect occurs due to the top-down activation of a specialised expertise processes. Half of the participants were asked to make same-different judgment in upright and inverted faces and another half was assigned to recognise Chinese characters. This was followed by a condition that included ambiguous figures which could be perceived as either faces or Chinese characters (configurally manipulated). Consistent with the authors' predictions, an inversion effect was observed in the face and ambiguous figures conditions, but not the Chinese character condition, thus providing clear evidence for configural face processing. Moreover, the authors suggested that the absence of an inversion effect in the Chinese character condition implies that the effect can be traced to specific activation of the face expert system.

A further source of evidence that adults rely more on configural than featural information in faces comes from studies which have filtered face images that contain certain spatial frequencies. Face images filtered to show only the low spatial frequencies will reveal preserved configural information within the face, while the features of the face will not be invisible. By contrast, face images filtered to reveal only the high spatial frequencies will show both the features and their configuration. In a couple of early studies, Harmon and colleagues (e.g., Harmon & Julesz, 1973) pixelated face images to remove the high-spatial-frequencies yet found that the faces could still be identified. The authors concluded that low spatial frequencies are sufficient to allow recognition of familiar faces and that high-spatial-frequency is relatively redundant in face identification. For example, Collishaw and Hole (2000) found that adults could recognise blurred faces with removed high-spatial-frequencies reasonably well, but that they were unable to do this if the faces were inverted, presumably because inversion meant that they were unable to access the remaining configural information in the faces. However other results are not consistent with these conclusions. Fiorentini, Maffei and Sandini (1983) demonstrated that both low- and high-spatial-frequency information alone can be sufficient to identify faces although their data did also reveal that the best results were obtained with a combination of the two spatial frequencies. Sergent (1989) pointed out that these discrepant results may be due to methodological differences (e.g., the use of face matching versus face naming).

### ***1.3.3.2 The development of configural processing***

In general, it is accepted that children's ability to recognise faces improves with age. Freire and Lee (2001) used a learning paradigm to examine the development of face recognition and its vulnerability during encoding. The face stimuli were manipulated either configurally or featurally, and paraphernalia such as hats or glasses were added (see also Leder & Bruce, 2000). Each child (age range: 4 to 11 years olds) was told a story about a person (named Bob) and shown his photo. Subsequently, the child was asked to identify Bob from a set of three other pictures. If the answer was correct then paraphernalia trials were added. It was found that paraphernalia trials disrupted processing of both featural and configural information, suggesting that paraphernalia

items are encoded as very salient and that young children have an unstable system with limited capacity. Two other important findings were highlighted here. First, children as young as 4 years of age were able to use configural information (although more poorly than adults) to discriminate a target face from distracters. Second, children up to around 7 years old were considerably slower to learn to recognise a face in the configural task compared with 11 year olds, suggesting a slow and gradual improvement in sensitivity to configural processing.

Brace et al. (2001) investigated the face inversion effect with children as young as 2 years of age using a total of 153 children. In contrast to Diamond and Carey's study (1977), their findings revealed that children from 6 years of age showed an inversion effect. Furthermore children between the ages 2 - 4 were surprisingly faster at recognising inverted faces than the upright ones, which the authors referred to as 'inverted inversion effect'. Brace et al. (2001) suggested number of possible explanations for the early configural encoding. First, configural encoding might depended on task demands, levels of difficulty and contextual support. Second, children might have both configural and featural information available to them, but their lack of experience to individuate and recognise a large number of people makes them less efficient and slower at using configural encoding. Third, the switch from featural to configural encoding still holds, but rather than occurring at around 10 years of age, it takes place at around 6 years of age. The age discrepancy in the studies may be due to task design. Brace et al. (2001) designed a child-oriented procedure in which a face recognition task was embedded in a story book, thereby giving contextual support. The 'inverted inversion effect' found in the youngest group of children was also found in a patient with prosopagnosia (Farah, Tanaka & Drain, 1995). The authors suggested that the 'inverted inversion effect' can indicate that an individual was able to use object recognition resources to process inverted faces. Thus, one may suggest that face and object systems are not fine-tuned in young children.

The other major piece of evidence that has been used to argue that configural face processing develops over time comes from study by Mondloch and colleagues (2002). The authors demonstrated that second-order configural encoding develops

over a longer time by creating and using a set of faces (called Jane and her sisters) that differed either in the shape of internal features (featural set) or differed in the spacing between the features (second order configural set). Thirty-six children aged 6, 8 and 10 years old and adults were tested on both sets presented sequentially in upright and inverted orientation. The target face appeared for 200 ms, while the second (test) face appeared until the participant responded whether the faces were same or different. Response time and accuracy was measured. The results showed that children of all ages were more accurate on the featural set, but their performance on the configural set was significantly lower, up to around 10 years of age. These data are consistent with previous research showing some evidence of configural processing in young children (Brace et al., 2001; Carey & Diamond, 1994; Freire & Lee, 2001), but lagging behind the development of featural recognition. For example, children in the youngest group studied (6-year-olds) performed almost as accurately as adults on upright featural face recognition.

From the developmental perspective, most findings support the initial view of Diamond and Carey (1977; 1986) that there is an increasing reliance on configural information with age, while younger children employ a less efficient featural strategy. It was also proposed that there is a qualitative ‘switch’ to configural processing at age ten (e.g. Diamond & Carey, 1977). However, there are wide discrepancies in the ages at which configural processing is thought to reach adult-like levels and the suddenness of the transition remains debatable.

It was also established that younger children (below 10 years) were more distracted than older children by added paraphernalia such as hats or glasses (Freire & Lee, 2001). Thus, as suggested by Chung and Thomson (1995), the nature and timings of any changes in face processing development are hard to establish, as the methodologies employed in various studies and the results obtained vary widely.

### ***1.3.3.3 Holistic and Configural processing and expertise***

Holistic and configural processes seem to use information from the whole face. This chapter discussed paradigms used to investigate both types of encoding for example, composite and whole-part paradigms for holistic processing and spatial feature manipulation for configural processing. Both types of processing are sensitive to face rotation. So are they really different? Previous studies have shown that the onset of holistic processing to precede that of configural processing. In addition to the difference in the temporal onset of these processing systems, they have distinct improvement rates, being more rapid in the former, whilst that of the latter is more gradual (e.g., Carey & Diamond, 1994, Mondloch et al., 2002).

Diamond and Carey (1986) proposed that configural encoding increases with expertise within a stimulus category discrimination. They showed that as well as a large face inversion effect, a similar effect could be found for inverting pictures of dogs for dog experts. This has also been found for handwriting experts (Bruyer & Crispeels, 1992). One prediction from Diamond and Carey's theory is that the degree to which face processing relies on configural information in the face should increase with development and experience of faces, and vast number of studies appear to be largely consistent with this.

However, other predictions can be made: as adults are trained to become experts on a class of stimuli, configural processing of these stimuli should increase with training and adults should show less configural processing (and worse performance) with classes of face stimuli with which they are less expert (for example, other race effect).

Gauthier and Tarr (1997) used the artificial Greeble stimuli to test these hypotheses. They attempted to replicate the Tanaka and Farah (1993) whole-face context advantage for part recognition with Greebles. They found that Greeble parts were indeed discriminated better in the context of a whole Greeble than when feature was shown alone, and this was true whether or not the subjects were expert in Greeble recognition, suggesting that holistic processing, unlike configural, may not increase with expertise.

### **1.3.4 Featural hypothesis**

Throughout this chapter, a ‘featural’ face recognition has been used as the alternative recognition to holistic and configural processing. This theory has its origin in object recognition theory (Biederman, 1987). The term ‘features’, when used in face processing, normally refers to the eyes, nose, mouth or face shape. At present, it is not clearly elucidated whether featural information in the context of face processing or any other object is processed sequentially or in parallel. Furthermore, features are sometimes subdivided into internal and external. However, the precise definition of what makes a feature is problematic. The term featural is often used interchangeably with many other terms such as local, componential or piecemeal and there is no consistent definition of what constitutes a facial feature (Rakover, 2002). Features also assume a level of descriptive (e.g., the eye is itself a configuration of smaller scale features such as lids, iris, etc.).

Until recently, there has been little attention to possible developmental differences in facial feature saliency (Cabeza & Kato, 2000). Several studies found that there are differences of relative importance between certain features for the face recognition. Imitation studies have shown that young infants rely more on mouth features than any other features (e.g., Meltzoff & Moore, 1994), however the eye region becomes most salient in children from the age of 6 years old (Hay & Cox, 2000). Leder and Carbon (2003) investigated facial feature saliency in a forced-choice face recognition task. Presentation of eyes alone gave a high recognition rate of 78%, however the accuracy rate dropped dramatically when mouth was presented to 47% and 34% when the nose was presented. From the developmental perspective, most studies have looked at the dichotomy of configural–featural face recognition and little attention has been given to the importance of different features (Brace et al., 2001; Carey & Diamond, 1977; Mondloch et al., 2002). For instance, Rakover and Teucher (1997) suggested that featural information is the most important information for face recognition as it carries highly informative values even more important than configurations. Others suggest a dominant role of features due to the fact that they can be easily recognise regardless of context (‘embedded in the ‘whole’ face or in isolation).

### **1.3.5 Right hemisphere hypothesis**

Some studies suggest that the right hemisphere plays a dominant role in the processing of faces in adults. For example, de Schonen and Mathivet (1989) demonstrated that infants as young as 4 to 10 months showed faster ocular saccades to their mother's face than to a stranger's face when the faces were presented to the right hemisphere. The authors suggest that it could be attributed to the way in which the two hemispheres may be processing the different types of information such as configural information would be processed in the right hemisphere, whereas the left hemisphere could be associated with the featural information.

## **1.4 NEURO-IMAGING STUDIES OF FACE PROCESSING**

In the last decade or so, numerous studies have been carried out using neuroimaging tools such as positron emission tomography (PET) and functional magnetic resonance imaging (fMRI) to investigate brain activation during face recognition tasks. Fusiform gyrus has been identified as a region of increased activation during face recognition tasks. For example, Haxby and colleagues (1994) found areas in occipitotemporal cortex to be activated by faces more than other classes of object, particularly parts of the lateral fusiform gyrus. Several other studies replicated this finding using a wide variety of comparison stimuli in passive viewing tasks (e.g., Gauthier, et al., 2000; Tong, Nakayama, Moscovitch, Weinrib & Kanwisher, 2000) and as a result, the area of fusiform gyrus has been termed the 'fusiform face area' (FFA). However, recently, the interpretation of selective activations in passive viewing tasks has been brought into question (Price & Devlin, 2003). Furthermore, despite the consistent demonstration that the fusiform gyri plays an important role in face recognition, debate on the role of the FFA continues. Several fMRI studies have found that there is no change in the activation of the FFA even when faces are shown in inverted orientation (e.g., Aguirre, Zarahn & D'Esposito, 1999; Haxby et al., 1999). However, inverting Mooney faces (faces composed simply of black and white shadow and light), (Mooney, 1957), which are almost impossible to detect as faces when upside down, did lead to a significant drop in FFA activation in fMRI (Kanwisher, Tong & Nakayama, 1998). On the other hand, the results from other studies suggest that FFA activity may correspond to more than simply face detection.

Gauthier and colleagues (Gauthier, et al., 1999) found that inverting the face stimulus corresponds to a drop in the response of the FFA, despite the fact that a face can still be detected.

One important point to note is that the FFA is not the only area that is activated during face viewing. The story is more complex and activation has been seen in many other brain areas, particularly in the regions of bilateral occipitotemporal cortex, including the inferior occipital gyri and the superior temporal sulcus (Haxby, Hoffman & Gobbini, 2000; Johnson 2005) as well as areas within the left fusiform gyrus, bilaterally in the anterior fusiform gyrus, the left posterior inferior temporal gyrus and the medial occipital lobe (see Gauthier & Logothetis, 2000, for discussion). For example, a region of the inferior occipital gyrus has now become termed the OFA (occipital face area) as it responds more to faces than non-face visual stimuli, particularly in the right hemisphere (e.g., Gauthier, et al., 1999; Haxby et al., 1999). This region may be associated with the perception of facial features (Haxby, et al., 2000) and may provide input to the FFA and other face selective regions.

Use of neuroimaging tools in investigating the development of face processing in infants and young children has been problematic so far. In a study of older children, Passarotti and colleagues (2003) found that although 10-12 year-old children showed activation in the FFA, this was more distributed in both temporal regions. This suggests that even in these older children, the face processing system is still developing from a distributed to a more discretely localised one. The authors replicated this study with older children (13 to 15 years old), and shown that even at these ages, the brain regions associated with processing faces are not yet quite adult-like (Passarotti, Paul, Rudiak-Gould & Stiles, 2001).

The ERP technique has been used extensively with infants and children as it is a non-invasive method (unlike PET). The general pattern from infant ERP studies is that the cortical regions associated with face processing become increasingly specialised and localised over time (see, e.g., Johnson & de Haan, 2001). Taylor and colleagues (2001) investigated the electrophysiological correlates of upright and inverted face perception and isolated eye perception in 4 to 15 year-old children. The authors

supported the finding that the neural response to upright faces continued to develop until adulthood. In addition, they found that the N170 response to isolated eyes matures earlier, by 11 years of age. They interpreted this as indicating that the ‘configural’ processing of the spacing between features in upright faces develops more slowly than the ‘featural’ processing of isolated features (eyes, in this instance).

## **1.5 SUMMARY**

At present it is not clear what relationship is between the holistic and configural mechanisms of face recognition. Identification of precise ages at which these two types of processing become operational and mature remains to be identified. All of the described studies on holistic and configural encoding in adults suggest that orientation is a critical variable, as illustrated in the inversion conditions. Faces are best recognised in the upright orientation and performance on inverted faces was generally lower and slower. Inversion effects have been found in 7-year-olds (Flin, 1985), 6-year-olds (Carey & Diamond, 1994; Tanaka, 1998), and 5-year-old children (Brace et al., 2001) suggesting a specialised encoding of faces at these ages.

However, above all, the role of expertise seems to play a significant role in this phenomenon. Through years of experience, engaging in social communication, the face recognition system becomes more specialised and limited to recognition of faces in the upright position. Concerning the developmental course of the holistic and configural specialisation, studies reviewed in this chapter showed that children achieve adult-like holistic face recognition by 6 years old, whereas configural processing develops more slowly until the age of 10 (Mondloch et al., 2002). This still remains hotly debated.

Feature processing and holistic processing appear to operate at an early age (Carey & Diamond, 1994; Tanaka et al., 1998), but the age at which configural processing emerges is unclear. Some studies indicate that configural processing does not reach adult levels until adolescence, but this has been challenged by findings showing that the inversion effect does not increase after age 8 (Itier & Taylor, 2004). Interestingly, although face recognition continues to improve throughout childhood (Carey & Diamond, 1994; Mondloch et al., 2002), it does not reach adult-levels without visual

input to the right hemisphere during the first six months of life (Le Grand, Mondloch, Maurer & Brent, 2001; Le Grand et al., 2003).

Figure 1.4 represents a schematic outline of the processes involved in face recognition, i.e., holistic, configural and featural summing of the empirical findings to date. The issue of whether first-order configural processing is mediated by subcortical or cortical pathways is still open to question. It is important to note that here that a static presentation of this kind downplays the developmental origins of the structures involved.

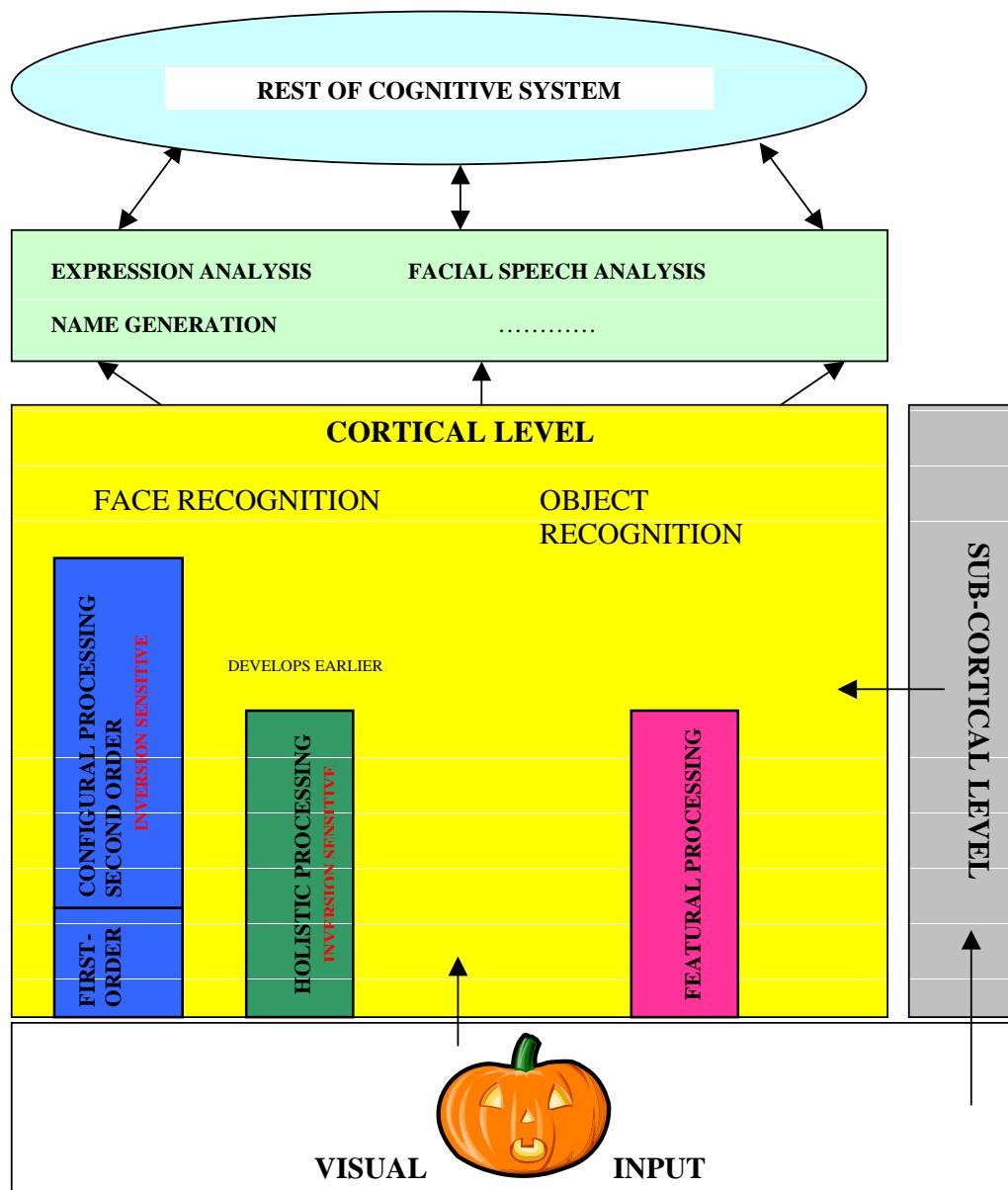


Figure 1.4: Graphic representation of processes involved in face recognition. The top part of the model is based on Bruce and Young's model (1987). The vertical mismatch between configural and holistic pathways is intended to capture the earlier emergence of holistic effects.

The next chapter will focus on the basis and current theories on face processing in three developmental disorders: autism, Down syndrome and Williams-Beuren syndrome. Subsequent chapters will highlight the practical aspects of the current thesis and its findings.

## **CHAPTER 2**

### **DEVELOPMENTAL DISORDERS**



## 2.1 INTRODUCTION

The main aim of this thesis is to explore and evaluate developmental changes that may occur during face recognition performance between clinical groups and typically developing control groups. The analyses will be carried out within the *Neuroconstructivist* theoretical framework. This framework places great prominence on the role of inter-relation between brain development and cognitive development and seeks to account for any developmental deviations/delays from the typical progression in terms of constraints that operate in the developmental processes (Karmiloff-Smith et al., 1998). It suggests that one needs to examine the development of both normal and atypical groups, right from early infancy or even prenatally to have a clearer picture of low-level prerequisites for normal mechanisms involved in the development of the particular cognitive structure.

Historically, a common approach to developmental disorders has been to identify a damaged module at the cognitive level and describe its functioning as “impaired” or “intact/spared”. The Neuroconstructivist framework argues that while this approach may be descriptively adequate, as an explanation it is more appropriate for characterising adult cognitive functioning in the endstate. Neuroconstructivism urges against a simplistic use of double dissociations and adult neuropsychology in accounting for findings from research on developmental disorders.

The adult framework can be informative and trigger some important questions. It enables researchers to map the architecture of cognitive modules and their purported independence by establishing single and double dissociations within and between domains. However, when applied to developmental disorders or even typically developing children, it is empirically dubious as it ignores the dynamics of developmental changes underlying disorders (Karmiloff-Smith, 1997, 1998). Neuroconstructivism argues that the adult modular structure cannot be assumed to be present in the infant but is itself a product of the developmental process. Studies to date that have taken the neuroconstructivist developmental approach to behavioural phenotypes have shown that areas of purported relative strength at one stage of development (middle childhood or adolescence) were not relatively stronger at

earlier stages of development (Paterson, Brown, Gsoedl, Johnson, & Karmiloff-Smith, 1999).

Paterson et al. (1999) showed that infant cognitive profiles in Williams-Beuren syndrome and Down syndrome cannot be predicted from the adult endstate of cognitive functioning. One of the most compelling examples is language in infants with Williams-Beuren syndrome which is very poor and at the same level as DS, but then improves at a far quicker rate and becomes significantly better than the DS comparison group by adulthood.

Another line of evidence illustrating fundamental differences in cognitive constraints comes from brain damaged patients. In adults with acquired damage, the affected systems had previously developed normally. Hence it is possible that damage to a fully developed mature system could lead to selective impairments and use of different cognitive mechanisms. However, in individuals with developmental disorders such as Williams-Beuren syndrome or Down syndrome, the genetic abnormalities constrain the development trajectory of cognitive abilities from the beginning. The implication is that these systems have been impaired under very different conditions. In the case of developmental disorders, it is constrained by atypical gene expression and/or atypical brain development, whereas in acquired damage in adults, the damage occurred after full system development. These crucial differences are often ignored, although they are likely to lead to quite different processing impairments. Besides, when similar levels of behavioural functioning are found, it is frequently inferred, or even *a priori* assumed that equivalent cognitive processes drive the behaviour across different groups. The causative differences of constraints can only be explored by more in-depth analyses of the processes underlying the behavioural performance, which may differ between populations. In other words, equivalent scores with respect to behavioural performance do not necessarily equate to similar cognitive processes (Karmiloff-Smith, 1998).

In line with this reasoning, the sensitivity of standardised tests is open to discussion, raising the risk that scores in the normal range may be achieved by atypical cognitive processes. Therefore, it has been argued that the use of sensitive tasks is crucial to properly assess underlying processes where claims of normality are made on the

basis of standardised test scores alone (Karmiloff-Smith, et al., 1998; Karmiloff-Smith et al., 2004).

### 2.1.1 Domain specific or general?

The notion of domain specificity has been under scrutiny, highlighting aspects such as the role of variations in the early state that could give rise to domain specific differences in endstate. Neuroconstructivism accepts relative modularity as a product of development and as the possible characterisation of the normal adult system, but it rejects it as a starting point for the infant cognitive system based on evidence from developmental cognitive neuroscience. It posits that even a small deficiency or abnormality early on can have cascading but differential effects on later development, making the outcome to appear as domain-specific (or at least domain-uneven) although it may have originated in a domain-general impairment (Annaz & Karmiloff-Smith, 2005; Karmiloff-Smith, 1997, 1998; Karmiloff-Smith et al., 2004). It invokes a strong role for plasticity and for processing capacities that are relevant to domains rather than specific to them. An explanation of developmental deficits consists of identifying how these initial domain relevancies have been altered in the disorder, and then how the subsequent process of emergent modularisation has been perturbed.

Thomas (2005) points out two unanswered problems associated with the Neuroconstructivism. The first asks for a clearer picture of the initial domain-relevancies that pre-date a particular domain, and of the nature of the process that eventually delivers domain-specific functional structures. The second difficulty is related to methodological issues of building developmental trajectories from infancy through to adulthood. Thomas argues that one should not assume that the same task is treated the same way, i.e., using the same mechanisms, at very different ages. The notions of **interactivity** and **compensation** will be key in characterising how atypical development proceeds at the cognitive level. **Interactions** and **compensation** have significant implications on the formation and functioning of mechanisms over developmental time (Karmiloff-Smith & Thomas, 2003; Thomas 2003, 2005). In addition, Morton (2004) proposed that compensation must also be considered at behavioural and biological levels.

### 2.1.2 Intact/spared versus impaired

In the literature on developmental disorders, one frequently encounters terms such as ‘spared’, ‘intact’ and ‘impaired’ when describing atypical development (for example: Hoffman, Landau, & Pagani, 2003; Rouse, Donnelly, Hadwin, & Brown, 2004; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). The notion of a selective deficit implies the ‘impairment’ of a single process or domain and the preservation (i.e., normal functioning across time) of others. When a brain has developed normally, resulting in specialised, localised functions, it is possible that brain injury may produce selective damage(s) with other components still operating normally. Hence, one might consider them to be ‘spared/intact’. However, in a developmental disorder, the appropriate terminology for this hypothesis would have to imply that a function has *developed normally* from infancy, through childhood to adulthood, with no interactions with other developing parts of the brain. This is unlikely, since early on the infant brain is highly interconnected (Neville, *in press*). Only with development and progressive pruning do brain regions become more specialised.

Rather than considering behavioural outcomes as preserved or damaged modules that are wholly ‘intact/spared’ or ‘impaired’ throughout development, Karmiloff-Smith (1998) argues that small changes in the initial state can become magnified throughout development into domains of relative strengths and weaknesses. Early development may be a crucial window of opportunity for intervention, as these small changes have not yet snowballed into impairments in whole domains of processing. Thus, the use of ‘intact/impaired’ terminology in characterising developmental aspects of functioning could hinder rather than help the study of the dynamics of atypical development.

Karmiloff-Smith advocates the importance of investigating not only domains of weaknesses but also domains in which individuals show proficiency. Besides, if changes to domain-relevant properties are initially widespread, and some properties are less relevant to a given domain, then that domain might exhibit lesser and perhaps more subtle impairments (Karmiloff-Smith, 1998; Karmiloff-Smith, Scerif & Ansari, 2004).

Thus, it is indeed crucial to differentiate between ‘normal’ scores at the behavioural level from their underlying cognitive and brain processes. Phenotypical outcomes could stem from very small differences in different parameters such as: developmental timing, gene expression, neuronal formation and their migration and density, and many other genetic and biochemical factors or brain neural network. Also, one must also bear in mind environmental factors when studying developmental disorders (Mareschal et al., 2005). A child must always be considered within the environment he/she lives; if the child’s cognitive system is developing atypically, the child may experience an atypical environment, both physically and socially.

The remainder of this chapter is dedicated to description of current knowledge on the visuo-spatial abilities of the individuals with autism, Down syndrome and Williams-Beuren syndrome, with the summary of each disorder at the end of this chapter.

## **2.2 AUTISM SPECTRUM DISORDER (ASD)**

### **2.2.1 Historical Perspective**

In his comprehensive clinical accounts, Leo Kanner described 11 children with profound problems in communication, language, lack of response to other individuals and resistance to change, giving a description of “autistic disturbances of affective contact” (Kanner, 1943). Similarly, Hans Asperger published a paper about a group of children with behaviours similar to those described by Kanner (Asperger, 1944). They both borrowed the term “autism” from Eugene Bleuler’s description of schizophrenia to characterise the “withdrawal from reality” which can be seen in both conditions. This link with schizophrenia led to the theoretical position that “infantile autism” was in fact a very early form of that disorder. Accordingly, in North America, the term “infantile autism” was replaced by terms such as “childhood schizophrenia” and “childhood psychosis.” It was Rutter (1972) who pointed out differences in symptomology between the children with early-onset psychosis in childhood from those with a later onset. Also, Lorna Wing demonstrated a link, not between autism and schizophrenia, but rather, between autism and mental retardation<sup>(1)</sup>. Wing clearly formulated the notion of a triad of impairments in socialization, social communication, and social play (Wing, 1993; Wing & Gould, 1979).

### **2.2.2 Clinical diagnosis**

Autism is defined broadly by the presence of deficits in three core domains of functioning: social reciprocity and engagement; communication and language skills; and stereotyped repetitive behaviours with restrictive interests, and sensory exaggeration (Diagnostics and Statistical Manual of Mental Disorders-IV, American Psychiatric Association, 1994). Gradual progress was made towards earlier identification of the disorder, and currently diagnosis occurs at around 30-38 months, and is characterised by many behavioural and medical problems.

---

<sup>1</sup>The term mental retardation will be replaced by learning difficulties hereafter.

In the early years, children with ASD fail to follow eye gaze, dislike being picked up or touched, and are less curious about their environment than typical children (DSM-IV; American Psychiatric Association, 1994). With increasing age, the number of abnormal behaviours increases, the most striking feature of which is their inability to play reciprocally with others. Individuals with ASD show major problems in understanding non-verbal communication (gestures, social imitation), as well as in speech production. Other characteristics include: hyper- or hypoactivity, abnormal eating behaviours and sleep patterns, self-injurious behaviours, and aggression and ticks (DSM-IV; American Psychiatric Association, 1994).

Individuals diagnosed with autism can display a variety of symptoms ranging from an inability to speak to those with superior intelligence (Frith, 2003). Many clinicians, psychologists and teachers adopt the use of labels such as low/severe-, middle- and high-functioning autism and/or Asperger syndrome, but these all fall together under umbrella of “Autism Spectrum Disorder” (ASD). Approximately 75% of individuals with autism have learning difficulties and the other 25% test in the borderline intellectual range. Severity of the disorder will be considered carefully in the current studies, and the performance of children with low-functioning autism (LFA) will be separately assessed from the children with high-functioning autism (HFA). Children in HFA and LFA groups were distinguished using tests whose details can be found in chapter 3 section 3.2.1.

The definition of Asperger syndrome has so far been problematic (for further discussion see Frith, 2004). Asperger syndrome was included in the DSM in 1994 and taken together with autism and Pervasive Developmental Disorder Non-Otherwise Specific (PDD-NOS) under umbrella of Autism Spectrum Disorder. The phenotype of autism is therefore highly heterogeneous. Thus using “Asperger’s syndrome” and “high-functioning autism” terms interchangeably can cause confusion and problems when comparing studies. Thus, some researchers strongly advocate the need to distinguish these two terms based on differences in language skills, as those with Asperger disorder do not exhibit language problems (Ozonoff, 2003).

### **2.2.2.1 Personality**

Individuals with autism are reported to prefer consistency, maintaining the same routines, and have a tendency to become attached to objects. The slightest change in routine may cause serious difficulty for the families of children with autism. Change can produce behaviours such as screaming and crying lasting for hours and leading to severe tantrums (Norton & Drew, 1994). Many behaviors occur repetitively for long periods of time or diminish for an unspecific period of time and reoccur again. Other common behavioral characteristics that are found in many other developmental disorders can vary widely across individuals with autism. However, some of the most prevalent behaviours in individuals with autism are: obsessive behaviors such as retracing exact steps to locations that are familiar, as well as repetitive flapping of the arms and hands. Also, poor toilet skills, refusing food, aimless wandering with no fear of getting lost, climbing on dangerous and inappropriate objects such as kitchen counters, roofs and railings, and little or no communication. Bizarre body movements and stiffening of the body, biting, pinching, and hitting, self-injuries, usually accompanied by tantrums, are often reported (Gillberg & Coleman, 2000).

### **2.2.3 Prevalence**

Epidemiological studies of autism reported rates of two to five cases per 10,000 children (e.g., Volkmar & Pauls, 2003). A huge increase of 556% was reported between 1991-1997, making autism cases higher than the prevalence of cancer or Down syndrome (Muhle, Trentacoste & Rapin, 2004). This was supported by subsequent studies that reported an increase in the prevalence of autism up to 30 per 10,000 in children under 8 years old and approximately 60 per 10,000 overall (Fombonne, 2003). Fombonne (2003) accounted for this increase by calling on to a number of factors such as changing diagnostic methods, heightened public and professional awareness, changes in definition criteria and small sample studies. Finally, autism is three to four times more common in males than females. The reason for this remains unclear, although could be due to genetic differences on the X chromosome.

#### **2.2.4 Genetics**

Studies of twins have demonstrated a high concordance rate between monozygotic twins, of around 60 %, and rising to 92% in the UK twin study (Muhle, Trentacoste, & Rapin, 2004), but only 10 % in dizygotic twins. This suggests that there is a large genetic component to autism susceptibility (Medical Research Council Review of Autism Research, 2001).

Several studies indicate that parents of children with autism, themselves display many autistic characteristics, albeit often more mildly. For instance, they show impairments in pragmatic language (e.g., Folstein et al., 1999) and executive function (Hughes, Leboyer, & Bouvard, 1997), and describe themselves as preferring routines and as having difficulties with changes in their environments. They perform above normal on the Embedded Figures Test and below norm at emotion recognition tasks (Baron-Cohen & Hammer, 1997), a pattern similar to their autistic offspring. Also, Dawson and colleagues found that parents of individuals with autism exhibited a significant decrement in their face recognition abilities relative to their verbal and visuo-spatial skills (Dawson et al., 2005).

Despite the evidence from twin and family studies, the identity and number of genes involved are hard to pinpoint. The current consensus is that a number of genes may be responsible for an individual's susceptibility to ASD (Korvatska, Van de Water, Anders, & Gershwin, 2002; Muhle et al., 2004). Evidence indicates that at least 10 genes may be involved in the causation of autism (Hutcheson et al., 2003). Thus far, abnormalities have been identified at chromosome 15(q11-q13), and some candidate genes include: FOXP2, and RAY1/ST7. Also, IMMP2L and RELN at chromosome 7(q22-q33), and close to the serotonin transporter gene (5-HTT) on chromosome 17(q11-q12) have been identified.

In his recent review Baron-Cohen (2005) pointed out that chromosome 17(5-HTT) is of particular interest, as serotonin innervates the limbic system which plays a role in emotion recognition. Skuse and colleagues (2003) in their study on Turner syndrome suggested that genes on X chromosome (Xp 11.3) can play a significant role in impaired social cognition. Also, Jamain and colleagues associated autism with the X-linked neuroligin genes that could potentially help to explain the 4:1 ratio of males to

females (Jamain, et al., 2003). By contrast, Stone and colleagues (2004) identified a male-specific linkage peak at chromosome 17(q11). These results suggested that sexual dichotomy is an important factor in the genetics of autism, and surprisingly extending to the macroscopic structures of the brain.

It has been suggested that certain structures such as limbic system and cerebellum volume have reduced cell growth, excessive cell loss, or reduced dendritic arborisation (Akshoomoff, 2005). Some postmortem and MRI studies have shown that overall brain size in autism is increased by 2-10%. However, most studies have failed to replicate, which can be attributed to the large variability within the autistic population and the techniques used.

#### ***2.2.4.1 Associated medical conditions***

Some of the medical problems associated with ASD are epilepsy, depression, vision and hearing impairments. The prevalence of epilepsy among all children is estimated at 2 to 3%, compared with some 30% in autism. Approximately 15 to 37% of cases of autism have co-morbid medical conditions and most common associations include fragile X syndrome, tuberous sclerosis and untreated phenylketonuria. Also, language disorders, Attention-Deficit/Hyperactivity Disorder, motor disorientation disorder, and dyslexia neurofibromatosis have been reported (Gillberg & Coleman, 2000).

#### **2.2.5 Cognitive phenotype**

The cognitive profile of children with ASD is highly variable, ranging from nonverbal with low IQ and severe autistic symptoms to high-functioning individuals with an above average IQ with milder autistic symptoms (for example “unsociable”). Eighty percent of individuals with autism have a full-scale IQ below 70 (Deb, 1997). Moreover, they are often described as having areas of significant cognitive deficits and areas of relative strengths as described below.

### **2.2.5.1 Face processing**

The aim of this section is to explore configural and holistic processing of face recognition. Therefore, components such as gender, emotion and age recognition of faces will be only considered briefly. Also, it should be noted that of the disorders that are considered in this thesis, face recognition abilities in autism have been studied the most extensively, in part due to the social deficits found in individuals with this disorder.

From the observational studies of home videotapes of first birthday parties, and numerous parental reports, it was demonstrated that the failure to attend to others' faces was the best discriminator between children with autism versus typically developing ones (Osterling & Dawson, 1994).

Anecdotal reports such as: "*He did not recognise me when I changed the colour of my hair*" (personal communication, 2002); "*She would not look at me or make baby noises; she ignored me completely. And although she happily held objects, she would not hold my finger ...*" (Stenhli, 1996), coupled with substantial evidence from the research studies, strongly suggest that individuals with autism process and remember faces in an abnormal way (Ellis, Ellis, Fraser, & Deb, 1994; Klin et al., 1999; Langdell, 1978). Perceptual problems also affect the perception of emotional expressions of faces (Hobson, 1986; Hobson, Outson & Lee, 1988), the perception of direction of gaze (Jolliffe & Baron-Cohen, 1997) and sometimes even the perception of gender (Hobson, 1987; Njikiktjen et al., 2001). Interestingly, however, some studies have reported normal face recognition in individuals with autism (e.g., Boucher, & Lewis, 1992; Davies, Bishop, Manstead, & Tantam, 1994; Rouse, et. al., 2004; Teunnise, & de Gelder, 2003).

Several studies that aimed to explain the mechanisms by which those abilities emerged or failed to emerge, reported that individuals with autism do not recognise faces holistically and display a reduced inversion effect, which indicates that they may have less expertise in face processing, and therefore rely on featural recognition (Hobson et al., 1988; Langdell, 1978; Tantam, Monaghan, Nicholson & Stirling,

1989; see Joseph & Tanaka, 2003; López, Donnelly, Hadwin, & Leekam, 2004 for contrary results).

The pioneering study by Langdell (1978) tested the ability of children with autism to recognise the faces of their peers from partial cues. The participants were shown features of the faces and were asked to recognise them either from the eye, nose or mouth region of the face. It was found that children with autism were significantly better than typical children at recognising faces on the basis of an isolated view of the mouth region, but they performed significantly worse than comparison groups matched on overall mental age (MA) and chronological age (CA) in using eye cues to identify faces. Also, children with autism failed to show the “face inversion effect” in comparison to the control groups. No significant correlation was found between performance IQ and the task. These results suggested that children with autism do not use configural processing and instead rely on feature-based processing (see Elgar & Campbell, 2001 for a similar proposal).

In accord with the results reported by Langdell (1978), Hobson and colleagues (1988) found that older children with autism recognised inverted faces significantly better than did MA-matched comparison participants. The authors suggested that the possible mechanisms of processing face recognition must be different either in encoding strategies or in efficiency from those used by control individuals. So, individuals with autism could use similar strategies for recognition of upright but different encoding for inverted faces which might be “abstract or meaningless” for them.

In another study, the largest carried out to date, Klin et al. (1999) tested 102 young children with autism (mean CA: 7.3), PDDNOS and non-PDD disorders (mental retardation and language disorders) matched on CA and nonverbal IQ. The Face Recognition task subtest of the Kaufman Assessment Battery for Children (K-ABC), and two comparison tests - Gestalt Closure and Spatial Memory were employed. The results showed that children in the autism group were significantly worse than any other group on the face recognition test when matched on verbal-MA and non-verbal MA. On the Gestalt Closure test they performed slightly worse when matched on non-verbal MA, but did not show a difference in performance when matched on

verbal-MA. The authors concluded that the children with autism exhibited deficits on face recognition that were not attributable to overall level of cognitive functioning.

Several studies reported that individuals with autism show poorer memory for faces compared to controls. For example, de Gelder and colleagues (de Gelder, Vroomen, & van der Heide, 1991) examined 17 children with autism (mean age: 10:11 years) and 17 typically developing children (mean age: 8:6 years) matched on the Peabody Picture Vocabulary test (PPVT), (Dunn & Dunn, 1997). The task consisted of two parts. In the first part of study, children were shown a set of 16 photos, one photo after another for 5 seconds each. In the second part, children were shown two photos, one from the first part and a second distractor photo. Children were asked whether they had seen either of these people before. The results demonstrated that children in the autism group were significantly worse than the TD group on the face recognition. This was further supported by another group of researchers who examined memory for faces and social scenes in adults with autism (Williams, Goldstein, & Minshew, 2005).

However, these results are not without challenge. Some recent studies have shown that people with autism are able to attend to and process contextual information. For example, Joseph and Tanaka (2003) examined the hypothesis that holistic face processing is impaired in children with autism. Twenty-two high functioning children with autism (age range: 8:0-14:4 years old), and 20 typically developing children (age range: 8:0-14:4 years old) participated in the study. The children were presented with pictures of children employing a whole-part paradigm. The results revealed that children with autism were markedly deficient when face identification depended on the eyes and showed no emergence of inversion effect on either of the conditions (whole- or part-face). However, a significant improvement was observed when face identification depended on mouth region. In addition, children in the autism group exhibited a whole-over-part advantage and an inversion effect, both of which are associated with maturation of holistic processing. The authors concluded that individuals with autism show holistic processing but with specific sensitivity to the lower part of the face in particular mouth.

Rouse et al. (2004) aimed to further explore the mechanisms of face recognition in individuals with autism. They presented 11 children with high-functioning autism and 30 children in control groups with two experimental paradigms based on the Thatcher illusion paradigm. Children with autism were matched for MA and CA. In study 1, all individuals were shown faces and buildings in upright and inverted orientations, and were asked to point to a picture that looked “strange or funny”. The results showed that children in autism group were similarly susceptible to the Thatcher illusion as both control groups. However, children in the CA control group reached ceiling scores in the upright face condition. The authors concluded that their data and the results from Joseph and Tanaka (2003) study (who explored holistic processing) suggested that the children with autism show normal configural face processing. This is somewhat confusing and presumptive. First, it seems that the authors suggest both that the Thatcher illusion task and whole-part task tap the same mode of processing. Second, configural and holistic processing are two different types of processing that develop at different rates and are affected differently by stimuli manipulations (reviewed in chapter 1; see also Maurer et al., 2002). Although, this was acknowledged in the paper, both terms nonetheless were used interchangeably.

López and colleagues re-examined the whole-part paradigm. Seventeen adolescents with autism and 17 typically developing children matched for chronological age participated in their study. Participants were asked to match a target face either to a whole-face or to a part-face feature. The study included a condition that cued participants to the relevant face feature for matching. The results showed that in the control group, the cue did not moderate the whole-face matching advantage, but in the autism group it did generate a whole-face advantage, while in the uncued condition no difference between whole face and feature matching was evident. The authors suggested that individuals with autism are able to use holistic processing in face processing under suitable conditions (López et al., 2004).

Other behavioural studies have explored configural and featural processing by altering spatial frequencies in their studies (see chapter 1 section 1.3.3 for detailed information). Deruelle et al. (Deruelle, Rondan, Gepner & Tardif, 2004) examined 11 high-functioning children with autism or Asperger syndrome and two groups of TD children matched on Verbal Mental Age (VMA) and on CA. The children had to match faces on either high-spatial frequency (which is useful for discriminating fine details such as facial features) or low-spatial frequency information (which provides information regarding configuration). The results suggested that the autism group performed better on HSF faces than LSF faces which was in contrast to the control groups (experiment 2). Curby and colleagues found similar results with autism group showing a greater reliance on HSF information compared to LSF information. Thus, these results demonstrated a consistent pattern of bias towards featural processing by the individuals with autism (Curby, Schyns, Gosselin, & Gauthier 2003).

The reported problems in face processing are consistent with a series of recent functional imaging studies demonstrating atypical or weak activation of the fusiform gyrus (Critchley et al., 2000; Grelotti et al., 2005; Pierce, Muller, Ambroses, Allen & Courchesne, 2001, Schultz et al., 2000). For instance, Schultz et al. (2000) observed that children with ASD displayed greater activity in the right inferior temporal gyri and less activation of the right fusiform gyrus during face discrimination. The authors suggested that individuals with ASD process faces in the same way as objects, using a feature-based strategy.

Contrasting results were reported by several other research laboratories that showed significant fusiform activation in high-functioning autism groups, especially in the right hemisphere, and with greater activation in response to familiar than unfamiliar faces (Dalton et al., 2005; Hadjikani et al., 2004; Pierce et al., 2004). Hadjikani and colleagues suggested that the discrepancy between their study and others is due to factors such as use of stronger field magnet and differences in the experimental procedure. The authors suggested that although individuals with autism activated the FFA and other brain areas normally, other more complex components or interconnections involved in social perception and cognition might be atypical in this clinical population.

Emerging evidence from brain studies suggests that abnormal development of the amygdala can give subsequent deficits in the areas of recognition of face identity and facial expression perception. Landmark fMRI studies have shown the amygdala in individuals with autism to be hypoactive during perceptual tasks (Baron-Cohen et al., 1999; Critchely et al., 2000; Pierce et al., 2001). Critchely et al. (2000) found that individuals with autism did not activate a cortical ‘face area’ when judging emotional expressions and also failed to activate the amygdala. Dysfunction of the amygdala may be one of causal factors underlying face recognition difficulties in individuals with autism (Baron-Cohen et al., 2000; Howard et al., 2000).

Dawson and colleagues used the ERP technique with individuals with autism to assess brain activity in response to upright versus inverted faces. All of their studies showed abnormalities in the different aspects face processing of people with autism (Dawson, et al., 2002; McPartland, Dawson, Webbs, Panagiotides & Carver 2004). In the McPartland study, 9 individuals with autism (14 to 42 years old) and 14 TD individuals (16 to 37 years old) were tested. The participants were shown four categories of stimuli: upright and inverted faces and upright and inverted furniture. The results from this study showed that autistic group had a minimal difference in N170 latencies to upright versus inverted faces, in contrast to the robust difference in latency to upright vs. inverted faces in the control group. This implies that the neural system related to face processing is less efficient and lacks sensitivity to configural alterations of the stimuli, which are indicators of atypical cortical specialisation of face processing. However, replication of this study with narrower age range in both groups and with larger number of participants would be useful to ascertain the role of age in the development of the neural system.

Another line of research found that children with autism display atypical gaze control in relation to social stimuli such as faces (Dawson, Meltzoff, Osterling & Rinaldi, 1998, Pelphrey et al., 2002; Swettenham et al., 1998; see van der Geest, Kemner, Verbaeten & van Engeland, 2002, for contrasting results). Swettenham and colleagues (1998) observed the spontaneous looking behaviour of typically developing 20 month-old children and of children diagnosed with autism, during playing sessions. They found that children with autism spent significantly less time looking at people

and more time looking at the objects when compared to controls. Pelphrey et al. (2002) tested 5 HFA adults (IQ in normal range) and 5 TD controls on visual scanning of photographs of human faces. They found that individuals in the autism group, unlike TD controls, spent significantly longer time viewing non-feature areas and less time on examining internal features of the faces, in particular the eyes region. The authors also described the scan-paths of the autistic participants as “erratic, undirected, and disorganized, often reflecting the processing of only one or two relatively unimportant features of the face (e.g., an ear, the chin, or region of the hair line)”.

Consistent with these findings, Klin, Jones, Schultz, Volkmar and Cohen (2002) measured visual paths and percentage of viewing time of 15 individuals with autism (mean age: 15:4) and 15 control individuals matched on age and IQ. Participants watched clips from *Who's afraid of Virginia Wolf* dominated by close-ups of faces. The results showed that individuals in ASD group focused more time on the mouth region of the faces than any other regions of the faces, with the eyes being of least interest. These findings are consistent with a study by Langdell (1978) and more recently by Joseph and Tanaka (2003). The authors suggested that bias to a single feature is a result of impairment in holistic recognition of faces, a different conclusion to that suggested by Joseph and Tanaka (2003).

Various theoretical proposals have been put forward to account for abnormal face recognition in autism. Deruelle et al., (2004) suggested that the deficits found in autism probably have their roots in a failure of early developmental processes. Hobson (1988) proposed that children with autism who score in normal range on a face recognition test may be treating faces as abstract patterns during the matching procedure without any emotional relations. Also, most of the experimental results correlate with hypothesis of Weak Central Coherence (Frith, 1989; Happé, 2000). Weak central coherence proposes that fundamental problem in autism is the difficulty in integrating individual pieces of information to establish meaningful configuration, thus reliance or bias towards featural information rather than on the overall context.

Dawson et al., (2001) hypothesised that abnormal social attention in autism is related to their inability to understand and attend to social reward. More specifically, the reward mechanisms that draw typically developing individuals' attention to the eye region are dysfunctional in autism. Social reward is unpredictable (smile, kiss or hug) in contrast to non-social rewards (a sound in response to a button). These social deficits have been explained in the 'Theory of Mind' account (Baron-Cohen, Tagger-Flusberg, & Cohen, 1993). Baron-Cohen et al. (1993) suggested that autism causes a child to be "mind-blind" by not understanding that other people think differently than themselves. Individuals with autism may have difficulties in understanding and conveying thoughts and emotions even within their closest social environment (their parents or peers).

Recently, Baron-Cohen (2002) formulated a theory called 'Systemising and Empathising' which aims to explain the nature of social problems that individuals with autism can have. The main notion of the 'Empathising' part of this theory is that individuals with autism have a profound impairment in the development of empathy. For example, inferring other's emotional states and imagination of others' minds is hypothesised to be impaired, leading to serious difficulties in development of social communication. On the other hand, the 'Systemising' part of the theory focuses on males' greater abilities in visuo-spatial skills and their response to more structured environments. This theory also claims to account for the higher male ratio in the autism disorder.

Many of the studies described in the current review suggested that individuals with autism are poor at face recognition because they use featural information during face encoding. However, current evidence is insufficient (experimentally and theoretically) to conclude that individuals with autism are less sensitive to configural processing than typically developing children. Reduced sensitivity to inversion provides only indirect evidence for configural processing, and it could be evidence for immature holistic processing. For example, stimulus manipulations such as the Thatcher illusion are not clearly defined regarding which aspects of face recognition could be facilitated.

Some individuals with autism have been shown to be able to use holistic processing. Converging evidence comes from studies regarding the importance of the mouth region. Another point to note is that none of the studies looked at the relationship with the severity of the disorder. Mostly high-functioning individuals with autism (IQ range between 70-100) participated in the studies. Such differences have been taken into consideration in the current thesis, with children with both high- and low-functioning autism recruited and compared.

### ***2.2.5.2 Visuospatial Cognition***

Many studies have shown that individuals with autism demonstrate superior performance on pitch processing and memory (e.g., Heaton, Hermelin & Pring, 1998), pattern discrimination (Plaisted, O'Riordan & Baron-Cohen, 1998), pattern construction/block design subtests of the WAIS/BAS (Shah & Frith, 1993), on detecting embedded figures, and on memory for objects (Mortton, Belleville, & Menard, 1999).

The notion of good visuo-spatial performance in this clinical population is often supported by observational reports: “*- By three, he knew most of the countries of the world by glancing at their shape on a map or puzzle, and he could identify puzzle pieces of the countries and states whether they were upside down ...*” (Stenlli, 1996). The teacher says: “*Be careful, do not take out your credit cards in front of these two boys as they memorise the numbers in a second and try to enter them on the internet shopping*”. This statement referred to two 10- years-old boys with autism whose IQ is below 70. (From anecdotal reports gathered during the testing sessions).

Shah and Frith (1993) demonstrated that individuals with autism were superior to a TD group in performance on the lock design task. The task was to reproduce abstract patterns using patterned blocks. Participants with autism completed the task more quickly and with fewer mistakes than individuals in the TD group. While TD children benefited from seeing the target design pre-segmented, those with autism performed well whether the design was pre-segmented or un-segmented. According to Shah and Frith (1993) pre-segmentation did not improve performance any further

in participants with autism, because they could spontaneously apprehend the elements of the stimulus even when presented un-segmented. Again, as in the face recognition section, these findings have been explained in terms of Weak Central Coherence (WCC) theory. It is argued that good or superior functioning of individuals with autism on tasks such as block design and visual illusions can be attributed to their distinctive cognitive style that leads to peaks and valleys in performance.

Recently, in a battery of experiments Behrmann et al. (2005) investigated the processing styles of individuals with autism with respect to faces and objects. They tested 14 individuals with high-functioning autism (19 to 53 years of age), and 27 typical control participants, individually matched for gender, age and education level. In their second study the authors used a well studied Navon figure paradigm (Navon, 1977). The Navon test is a hierarchical figure composed of two layers of information, a holistic level (a whole letter image), and a featural level (smaller elements which make up the whole image). The autistic group was not only slower in their response, but also produced a very different pattern in comparison to the control group, i.e., their response on the featural level was faster than on the holistic level of letter identification. In sum, the majority of the studies that have examined visuo-spatial abilities have pointed to an atypical advantage for featural processing in autism population. However, it is noteworthy that none of the studies examined whether there is a developmental shift in the attention bias to featural or holistic processing across normal development.

#### ***2.2.5.3 Language***

Language and communication problems are some of the key symptoms characterising autism. Some of the initial problems that parents of children with autism report are their lack of language, or loss of language which had begun to develop. One of the most often described features of autistic language is echolalia. They can repeat words, sentences, and sometimes whole songs and movies with the exact words and intonation. Instrumental use of language rather than social use is another striking feature of autistic language. Individuals with autism show poor

pragmatic abilities regardless of the severity of the condition (Tager-Flusberg, 1996). By contrast, grammatical and semantic impairments are often attributed to the severity of the disorder. For example, individuals with High Functioning Autism (HFA) have mild to moderate problems with grammar, in contrast individuals with Low Functioning Autism (LFA) who have severe difficulties with grammar and communicate via single words or signs, and rote-learnt phrases.

Some difficulties with linguistic meaning are very specific to this disorder: for instance, the use of words and phrases in a narrow, context-bound way, difficulties with abstract terms and lack of acquisition and/or use of terms that refers to states of mind or emotions (e.g., Hobson & Lee, 1989).

In one of their studies Tager-Flusberg and Joseph (2003) subdivided 47 children with autism (aged 6-13 years old) on the basis of their verbal and non-verbal IQ to differentiate autistic phenotypes. Of a particular note is that the authors found low verbal IQ to be linked to lack of communication and reciprocal social interaction. It was found that those children scored low on verbal IQ and relatively high on non-verbal tasks, they showed severe symptoms of autism.

## **2.3 DOWN SYNDROME (DS)**

### **2.3.1 Historical Perspective**

The name ‘Down’ comes from John Langdon Down, an English doctor who in 1866 described the syndrome with the characteristics accounted for below. DS has received a great deal of research interest in comparison to other genetic developmental disorders. Surprisingly, most studies concentrated on investigating development in infancy and toddlerhood, which is in contrast with other disorders such as Williams-Beuren syndrome or Prader-Willi. Although, individuals with DS have been often used as a comparison group, there have not been many studies focusing cognitive domains, in particular face recognition, in older children and adults with DS.

### **2.3.2 Genetics**

Down syndrome (DS) is associated with the presence of three copies of chromosome 21 (trisomy 21), which is caused by non-disjunction of chromosome 21 during cell division. The exact relationship between trisomy 21 and DS is not fully established since some studies suggest a link between the syndrome and mitochondrial dysfunction at the cellular level (Roizen & Patterson, 2003). There are three types of the syndrome, with approximately 94 % of all people with DS falling into trisomy 21 type (in which all cells have an extra chromosome 21). The other two types include translocation trisomy 21, which arises from the permanent attachment of chromosome 21 to another chromosome (mainly chromosome 14, hence [t(14;21)]). This is also known as Robertsonian translocation (named after the Australian Chromosome expert who described this type of translocation), and it is prevalent in about 3-4% of DS cases ([www.nas.com/downsyn/benke.html](http://www.nas.com/downsyn/benke.html)). The third and final type of DS is mosaic DS, which is prevalent in about 2-4% of DS cases. In this the individual has a mixed population of cells (the trisomy 21 population and a normal cell population). The physical features in these individuals vary depending on the extent of the trisomy population, However, they tend to be milder compared to trisomy 21 ([www.nas.com/downsyn/benke.html](http://www.nas.com/downsyn/benke.html)).

### Prevalence

The prevalence of the syndrome in the U.K. is around 1 in 600 to 800 live births and the functions of the relevant genes causing the phenotypic outcomes of the syndrome are yet to be discovered (Bellugi, Lichtenberger, Jones, Lai & St. George, 2000; Roizen & Patterson, 2003).

### **2.3.3 Clinical diagnosis**

Diagnosis of Down syndrome is usually made at or soon after the birth and is primarily based on physical characteristics. The phenotypic anomalies that are associated with the syndrome include hypotonia, flat facial profile, small nose, a big space between the first and second toe, a below average weight and length at birth (Korenberg, 1991). Other characteristics include: loss of hearing, poor auditory short-term memory, language impairments and speech production difficulties (Pary, 1989).

However, in recent years, a few other techniques have been used. They include screening of pregnant mothers for trisomy of chromosome 21, measuring biochemicals in the foetal amniotic fluid (Beta HCG, Alfa Feto Protein, & Estriol). A more definitive way is the use fluorescent in-situ hybridisation (FISH) test to detect the extra copies of chromosomes. Also, there are a number of ways of other prenatal testing to screen for DS, such as the use of amniocentesis (carried out at 14-16 weeks), [www.nas.com/downsyn/benke.html](http://www.nas.com/downsyn/benke.html).

#### **2.3.3.1 Personality**

Individuals with DS are often described as friendly, and as having charming personalities (Wishart & Johnston, 1990). One study reported that older children and adults with DS are predictable in their behaviour, less active and persistent and more distractible than other children (Gunn & Cuskelly, 1991). Carr (1995) collated parental reports which showed that over 50% of 11-year-old children with DS were described as “lovable”, “affectionate” and “getting on well with other people”.

### **2.3.4 Cognitive Phenotype**

DS is the most common cause of learning difficulties. The pattern of cognitive abilities is often reported to be rather uniform, displaying gross delays or deficits across three main domains: language, face processing and visuo-spatial abilities (Wang, 1996; Weeks, 2002, but see Jarrold & Baddeley, 1997 for contrasting evidence). Children with DS have an average IQ falling in the range between 36-107, but significantly declining with increasing age to between 40 and 70 (Wang, 1996).

Recent research on the cognitive phenotype in individuals with DS has mainly focused on deficits in verbal working memory (Jarrold, Baddeley, & Phillips, 2002; Laws, 1998). Individuals with DS have often been characterised with relative strengths in visuo-spatial processing and poor verbal processing skills (Jarrold, Baddeley & Hewes, 1999; Klein & Mervis, 1999; Wang & Bellugi, 1994).

#### ***2.3.4.1 Face processing***

Only a handful of studies have examined face processing in DS. Wishart and Pitcairn (2000) carried out two studies to investigate face processing skills in children with DS. In their first study, 16 children aged 8 to 14 years old were tested on two tasks (identity-matching, expression-matching to a story). They were compared to typically developing children matched on overall mental age. The results revealed that although children with DS were slower at identity-matching tasks, their performance was not significantly different from TD. However, their performance was significantly poorer on the expression-matching task. Children had particular difficulties in decoding emotions such as surprise and fear. The second study also focused on identity and expression recognition; however faces were presented in different orientations (at 0°, 90° and 180°). Children were presented with familiar faces (their peers) and unfamiliar ones, and asked to choose the face they had seen before. Again, the results indicated that children with DS were less accurate and slower in response than TD. Furthermore, unlike the TD group the accuracy and response time of the children in DS group were not sensitive to orientation of the faces. These studies provided little information about the currently debatable featural

and configural face processing, as the measures used in these studies were not designed to tap configural or featural encoding.

Most recently, in a preliminary study, Kaiser and colleagues examined face recognition in adults with DS. They used a same/different matching task in which the configuration and size of eyes and mouth was manipulated. Individuals with DS performed significantly worse when the configuration and size of the eyes were altered than the mouth feature. It was concluded that individuals with DS are impaired on processing information from the eye region of the face, which might have a cascading effect on social/emotional functioning (Kaiser, Virji-Babul, Iarocci, McLaughlin, & Tanaka, 2005). However, this study is preliminary with limited details available, so more studies will be necessary to explore these issues.

#### ***2.3.4.2 Visuo-spatial Cognition***

Current evidence gives rise to speculations that some aspects of visuo-spatial processing are stronger than others in people with DS. In particular, visual memory, visual-motor integration, and visual imitation seem to be areas of relative strength within visuospatial processing, whereas spatial memory and visuo-constructive abilities seem to be areas of relative weakness (Fidler, 2005).

Evidence of strengths in visual processing during early development in DS can be found in studies of visual recognition memory, where infants with DS show similar event-related brain potential morphology, visual attention, and visual fixation to TD infants (Karrer, Karrer, Bloom, Chaney, & Davis, 1998). By contrast, in visual exploration in play situations, 6-months-old infants with DS were delayed in comparison to TD infants (Gunn, Berry, & Andrews, 1982). Other studies reported impaired visual attention on habituation tasks, and delays in various aspects of eye contact including functional use of eye contact to explore the environment in a parent-child interactive setting (Berger & Cunningham, 1983; Brown et al., 2003).

Another line of research has focused on the investigation of visuo-spatial short term memory using Corsi blocks, in which the experimenter taps a number of wooden blocks in a pre-specified sequence and each participant is required to repeat the same

sequence by tapping the blocks (equivalent in space of the verbal digit span). Most of the studies have reported that, although individuals with DS were slower, their performance was not significantly different from control groups. Evidence suggests that visuo-spatial short term memory is atypically affected on verbal tasks only (Jarrold & Baddeley, 1997).

Some indirect evidence from the standardised tests suggests that children with DS use a holistic strategy (Bellugi, Lichtenberger, Mills, Galaburda & Korenberg, 1999). In their comparative study of Williams-Beuren syndrome and Down syndrome individuals, Bellugi and colleagues reported that in a drawing task, people with DS produced a good holistic pattern but failed to reproduce the features correctly (Figure 2.1A). Moreover, on block-design tasks, their performance was very poor and they made errors of internal detail (Figure 2.1B). Difficulties with integration of simple shapes have also been emerged on the Navon task where individuals with DS tended to produce only the global forms of the letters (Figure 2.1C).

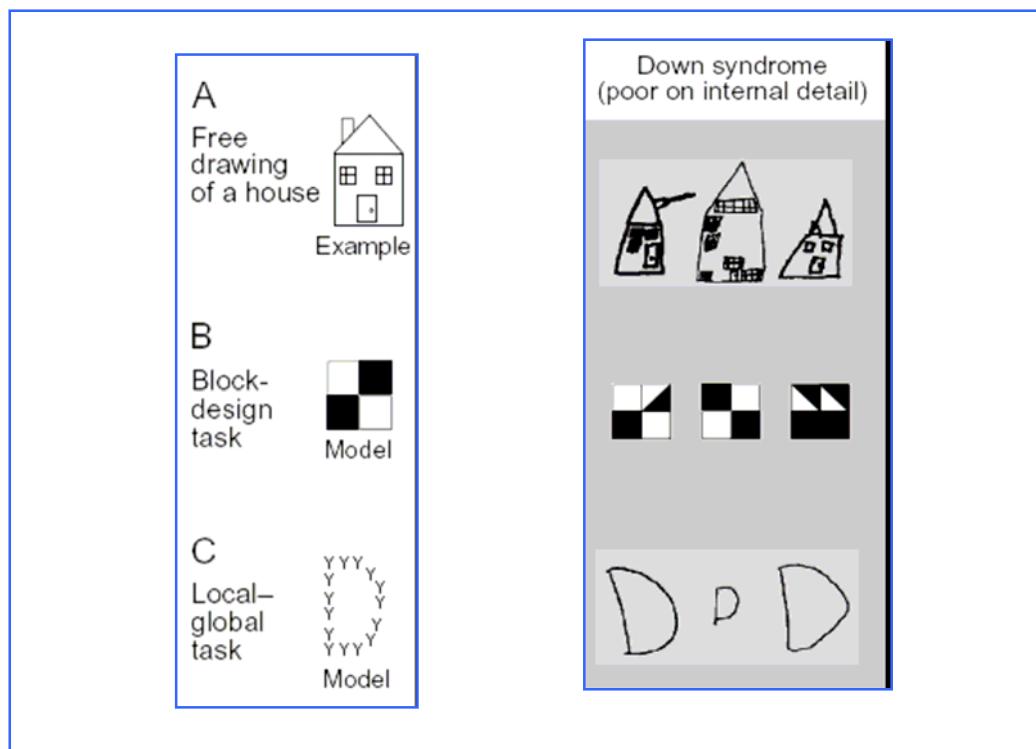


Figure 2.1: Adapted from Bellugi et al., (1999). A) Example of drawings by an adult with DS; B) Example of a block design; C) Navon task trial.

#### ***2.3.4.3 Language***

Studies of language development in children with DS indicate that their expressive language is particularly delayed in comparison to typically developing children and matched on non-verbal MA (Miller, 1999). Although, early language milestones emerge at typical times in cognitive development, with increasing age children with DS diverge from the norms. They show slower rates of development in grammatical morphology and expressive language (Beeghly & Cichetti, 1997).

Problems in DS language have often been associated with poor verbal short-term memory (e.g., Jarrold, Baddeley, & Phillips, 1999). In a longitudinal study of language comprehension by Laws and Gunn (2004), 33 individuals with DS (30 in a follow-up study) were tested on a battery of language tests including: the British Picture Vocabulary Scale, the Test for the Reception of Grammar (TROG) and on the non-word repetition test. The results suggested that there was no progress with age on the TROG test and non-words repetition test. The individuals reached a plateau and some of them declined with age. However, their performance on the BPVS, although poor, continued to increase at a slow rate. The authors concluded that factors such as hearing deterioration, decline in memory, and moving to residential homes could be considered as risk factors for the language plateau.

## 2.4 WILLIAMS-BEUREN SYNDROME (WBS)

### 2.4.1 Historical Perspective

Williams-Beuren syndrome was first identified by J.C.R Williams, a cardiologist from New Zealand who noted that several of his patients shared similar characteristics that included a heart defect (supravalvular aortic stenosis), learning difficulties and some facial characteristics (including a turned-up nose and a small chin). Shortly thereafter, in Germany, M. Beuren identified individuals with similar characteristics that expanded on the phenotype to include overfriendliness and dental anomalies. Thus, the syndrome is often referred to as Williams-Beuren syndrome (henceforth, WBS) or Williams syndrome (WS). Since then, significant advances have been made in the areas of genetics, biochemistry, cognitive and behavioural features providing a clearer understanding and characterisation of the WBS phenotype.

### 2.4.2 Genetics

The genetic basis of WBS was first established in 1993 (Ewart et al., 1993), and was shown to arise from the deletion of a number of genes on one copy of chromosome 7(q11.23), now known to be at least 28 genes (Tassabehji, 2003). The authors tested a group of 9 individuals with WBS using fluorescent in-situ hybridisation test (FISH), and discovered a link between the elastin gene on the long arm of chromosome 7 and autosomal dominant supravalvular aortic stenosis (SVAS). Given this link and the association between WBS and SVAS, the authors proposed that WBS is a contiguous gene disorder in which one copy of the elastin gene is deleted leading to the vascular abnormalities. All individuals with WBS showed the clinical features typically associated with the syndrome and it was put forward that the elastin hemizygosity (deletion of one copy) is involved in the pathogenesis of WBS. However, the hemizygosity of the elastin gene alone does not explain all the neurobehavioral aspects of WBS. Other genes absent from the WBS-associated deletion at the chromosomal subunit 7(q11.23) adjacent to elastin are believed to contribute to the pathogenesis of WBS (Robinson et al., 1996; Tassabehji et al., 1996).

Another gene deletion that was identified is the Lim Kinase-1 (LIMK1), which encodes the protein tyrosine kinase and is expressed in the developing brain (Proschel, Blouin, Gutowski, Ludwig, & Noble, 1995), and may therefore have an effect of axonal guidance during brain development (Tassabehji et al., 1996). Several research groups (e.g., Frangiskakis et al., 1996; Mervis, Morris, Bertrand & Robinson, 1999; Monaco, 1996) have proposed that the spatial deficit in WBS may be associated with the deletion of the LIMK1 (Figure 2.2). However, Tassebehji et al. (1999) showed that although LIMK1 deletion could have a contributory role in visuospatial deficits it does not necessarily play a causal role in the deficit. They tested three individuals diagnosed with SVAS who were identified with LIMK1 deletion, but the individuals did not display any visuo-spatial problems (for further findings see also, Donnai & Karmiloff-Smith, 2000; Gray, Karmiloff-Smith, Funnell, & Tassabehji, in press). Three further genes typically deleted in WBS, Syntaxin1A, WBSCR9 and RFC2 are also thought to be involved in brain development (Donnai & Karmiloff-Smith, 2000). Recently, GTF2I gene was identified and it is suggested to be involved in neuronal maturation (Morris, et al., 2003).

It therefore seems likely that the relationship between genotype and cognitive phenotype in WBS is more complicated than at first assumed, with the uneven cognitive profile resulting from a complex interaction between multiple genes in the deleted region (Donnai & Karmiloff-Smith, 2000; Karmiloff-Smith, Scerif, & Thomas, 2002). Thus, caution should be exercised when trying to attribute a dysfunction to a specific genetic mutation.

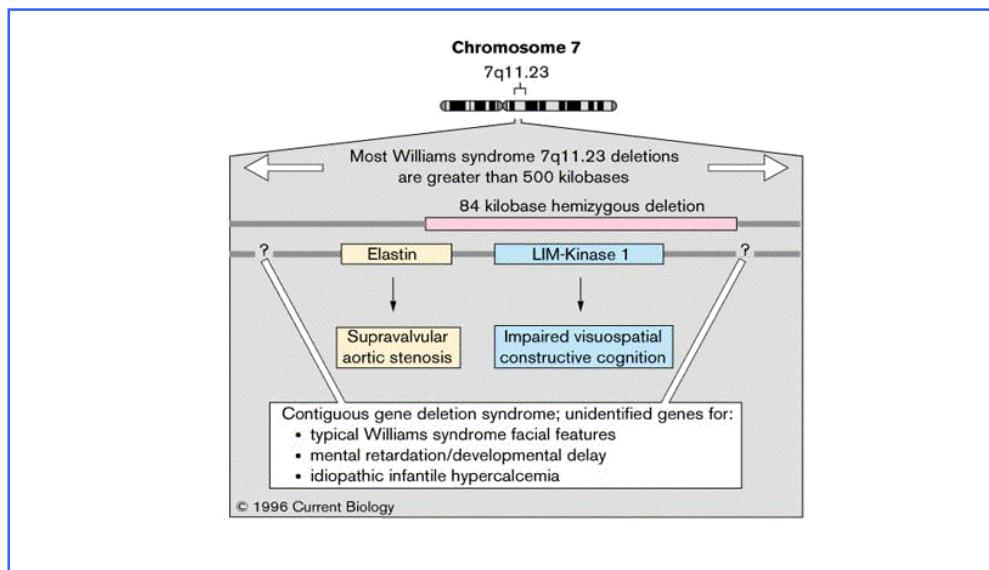


Figure 2.2: Schematic diagram of the small region of chromosome 7 deleted in WBS. Adapted from Monaco (1996).

### 2.4.3 Clinical diagnosis

The incidence of WBS is approximately 1 in 20,000 (Morris, Demsey, Leonard, Dilts & Blackburn, 1988), although recently it was estimated at 1/7500 (Stromme, Bjornstad, & Ramstad, 2002). Individuals with WBS are characterised by a number of distinctive features such as cardiac and dental anomalies, hypercalcemia<sup>(2)</sup>, facial dysmorphology, small stature, premature ageing of skin, hoarse voice, hyperacusis<sup>(3)</sup>, musculoskeletal and renal abnormalities (Korenberg et al., 2000). Around fifty percent of individuals with WBS have a strabismus<sup>(4)</sup>, mainly in a form of esotropia<sup>(5)</sup> with refractive errors being common (Donnai & Karmiloff-Smith, 2000).

<sup>2</sup> This is an abnormally high level of calcium in the blood, which causes loss of appetite, nausea, vomiting, constipation, abdominal pain, and kidney and bowel problems.

<sup>3</sup> Intolerance to certain sounds

<sup>4</sup> A visual defect in which one eye cannot focus with the other on an object due to imbalance of the eye muscles, often called a squint.

<sup>5</sup> A form of strabismus in which one or both of the eyes deviate inward.

### **2.4.3.1 Personality**

Individuals with WBS are often described as overfriendly, gregarious and typically unafraid of strangers (Einfeld, Tonge, & Rees, 2001). From parental reports, it is known that children with WBS are very empathic and responsive to the emotional states of others (Bellugi et al., 1999). Some studies indicate that individuals with WBS show a higher rate of emotional and behavioural problems in comparison to other disorders. They tend to exhibit overactivity, poor concentration and excessive anxiety to places and unfamiliar settings. Similarly to individuals with autism, they often display repetitive behaviours and preoccupation with certain objects (such as insects, cars, or postcards), particular topics (travelling, illness, future events such as birthdays, etc.) or certain people (a teacher, a television star or neighbour), (Korenberg et al., 2000).

### **2.4.4 Cognitive phenotype**

WBS attained great interest when Bellugi and her colleagues (Bellugi, Marks, Bahrle, & Sabo, 1988) proposed that individuals with WBS have a distinct cognitive profile showing peaks and valleys in their cognitive skills, with language, face recognition and social interaction abilities claimed to be “intact” while spatial and numerical skills were seriously impaired (e.g., Bellugi et al., 1988, Bellugi, Bahrle, Trauner, Jernigan & Doherty, 1990). Overall IQ levels range from 40 to 90 (Korenberg et al., 2000), with majority scoring between 55-69 (Searcy et al., 2004).

#### **2.4.4.1 Face processing**

Landmark studies have argued that face recognition develops normally in WBS, such that individuals with WBS achieve scores in the normal range on some face processing tasks (Bellugi et al., 1988; Udwin & Yule, 1991). Bellugi and colleagues (2000) reported that a group of 16 individuals with WBS who had performed at the “severely deficient” level on the Benton Judgment of Line Orientation Test (discussed in following section) simultaneously performed at nearly normal adult levels in the face-matching subtask of the same test battery (Benton Hamsher, Varney & Spreen, 1983b). Similarly good performance was seen on other face

recognition tasks such as the Warrington Face Memory Test (Warrington, 1984) and the Mooney Closure Task (Mooney, 1957).

To date, there are no experimental reports of face recognition in infants and very young children with WBS. Some observational studies that used tasks to indirectly examine face processing, revealed that infants with WBS spend significantly more time focused on faces than on objects (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Laing, Grant, Thomas & Karmiloff-Smith, 2005; Mervis & Bertrand, 1997). This led many researchers to believe that this inordinate attention to faces is a precursor to good face recognition skills in individuals with WBS.

Tager-Flusberg et al. (2003) argued for normally developing face recognition in WBS. They investigated holistic face recognition using the part-whole paradigm adapted from Tanaka and Farah (1993) and the Benton Facial Recognition test (described in detail in chapter 4). The authors tested a large group of 47 individuals with WBS (age range from 12-36 years old) and 36 CA-matched control participants. The participants with WBS showed a whole-face processing advantage in the upright condition, similar to that shown by typical participants. Furthermore, the authors reported that both groups (WBS and typical group) showed the same pattern of recognising faces by individual features, performing best on recognition of the eyes. However, closer examination of the data reveals that participants with WBS were at floor level on some of the conditions, rendering the interpretation of results difficult. Also, as already discussed in the face recognition chapter, the whole-part paradigm does not tap configural processing, a fact acknowledged by the authors. It is simply not sufficient to choose the whole-part task to investigating face processing abilities in WBS. Furthermore, the Benton test used in the study can be resolved just by using featural processing (Duchaine & Nakayama, 2004).

While some argue for normally developing face recognition skills in WBS, a large number of studies have shown that people with WBS may attain normal range scores in face recognition tests by using different routes of processing to achieve good performance. It was proposed that individuals with WBS rely more on featural encoding than configural encoding (Deruelle, Mancini, Livet, Cassé-Perrot & de

Schonen, 1999; Karmiloff-Smith, 1997; Karmiloff-Smith et al., 2004; Mills et al., 2000).

Karmiloff-Smith (1997) set out to ascertain whether individuals with WBS recognise faces using configural information. The author used several face processing tasks, some of which required configural processing, while others required featural processing. The results indicated that performance in the WBS group was poor on the configural face recognition task. However, the results were preliminary and configural and featural face information was not directly manipulated.

Deruelle and collaborators (1999) tested 12 children and adults with WBS (age ranged between 7 and 23 years old) matched on chronological age and mental age. Participants were shown a series of pictures of faces and of houses (experiment 2) presented in upright and inverted conditions in a same-different judgement task. In the face conditions, individuals with WBS did not show inversion effect, in contrast to CA and MA-matched control groups, whereas, in the house condition, there was no difference between the groups. The authors explained these results by arguing that the WBS group showed greater reliance on featural information in both the upright and inverted conditions, whereas the controls used predominantly featural processing for the inverted faces and configural processing for the upright faces. This led the authors to speculate that WBS face processing is not delayed but follows a different developmental pathway, confirming Karmiloff-Smith's previous study (1997). In another study, Deruelle et al. (1999) investigated the processing of configurally- and featurally-modified schematic faces and geometric shapes (experiment 3). Yet again, the CA- and MA-matched participants produced significantly fewer errors than the WBS group on configural items, but there were no group differences with respect to the featural ones. The Deruelle study showed that individuals with WBS are biased to process featural over configural information, regardless of the type of stimuli.

Recently, in line with Derulle's claims, Karmiloff-Smith et al. (2004) demonstrated in a series of face recognition studies that individuals in WBS group lack normal sensitivity to configural properties of face stimuli. They tested configural processing through the use of inversion and the manipulation of configural or featural

information in face stimuli. In one of their studies, fourteen individuals with WBS (age range: 14-51 years old) and TD group individually matched on CA were tested on their sensitivity to configural face recognition using Jane faces task (described in chapter 1 section 1.3.3). The results showed that individuals in the WBS group did not display the normal emergence of the inversion effect and were also significantly less sensitive on the configural face condition in comparison to the control group. In the second study, 14 individuals with WBS (age range: 12-54:10 years old) and 111 typically developing children (age range 2:8-11:5 years old)<sup>(6)</sup> were tested on recognising faces shown in upright and inverted orientations. Taking advantage of the wide age range the authors used a novel approach of building cross-sectional developmental trajectories (an approach adapted in the current thesis) to compare TD and WBS developmental trajectories. While the WBS group displayed similar performance in terms of accuracy and response time, their developmental trajectory did not show the emergence of the inversion effect. The Benton test was also used to assess whether it could predict participants' performance on the experimental tasks. The performance scores on the Benton tests improved with increased age. The same improvements were apparent for tests where configural processing was required, suggesting that the Benton can be solved via a featural strategy (see Duchaine, 2004, 2005, for similar evidence with prosopagnosics and TD adults). The authors construe that the WBS face recognition follows an atypical developmental processing encompassing a featural bias.

This is further supported by the findings from several ERP studies. For instance, Grice et al. (2001) investigated the neural basis of featural face processing in people with WBS and ASD by examining binding- related gamma activity in response to the upright and inverted faces. They found that individuals with WBS and ASD did not show any difference in gamma activity when faces were presented in upright or inverted orientations. However, there was a qualitatively different gamma burst for each group. It was speculated that a failure to integrate isolated features to compose a whole object might be related to binding<sup>(7)</sup> processes in the brain. The authors concluded that although WBS and ASD groups display similarities at a behavioural

---

<sup>6</sup> Data for the control group were provided by Brace and colleagues (Brace et al., 2001).

<sup>7</sup> According to neuronal binding, neurons within different neuronal assemblies fire in synchrony to unite different features of neuronal representations.

level (i.e. featural face processing) they differ at neurological level, and may use different strategies for featural face processing.

Mills and colleagues reported that 18 adults with WBS performed similarly to the typical control adults in their behavioral responses to inversion of face stimuli. Both groups showed a 10% drop in same/different judgments for face stimulus pairs as well as a 50 msec slower reaction time for inverted faces compared to upright faces (Mills et al., 2000). Also, during the behavioural study ERPs were recorded, and the results revealed both developmental delays and abnormal patterns. Early components of the ERP waveform, taken to reflect structural aspects of face perception, showed abnormal patterns in all individuals with WBS. By contrast, late components of the ERP waveform (during the same/different judgment task) taken to reflect face recognition, were quite similar among WBS adults to the pattern seen in typically developing 13-year-old children. Unlike typical control adults who showed distinct ERP patterns to mismatched faces when the stimuli were upright versus inverted, the ERP patterns for upright and inverted faces were not distinguishable in the control 13-year-olds and WBS adults. The authors reported that this pattern was not observed in normal adults, children, and infants at any age, nor in any clinical populations that they tested (including Down syndrome, language impaired children, and children with early left- or right-hemisphere brain injury). These data not only suggest that face processing mechanism is not normally developing but also that individuals with WBS may use a general object processor to recognise faces rather than a face-specialised mechanism.

In one of very few neuroimaging studies investigating cognitive abilities in a WBS group, Mobbs et al. (2004) aimed to elucidate the neural systems that underlie face processing skills in WBS. The authors compared performance accuracy of 11 individuals with WBS to 11 typically developing children (age and gender matched) on faces seen at different orientations with both direct and averted gaze. An unusual pattern of increased frontal activation in the right prefrontal cortex was observed in the WBS group. It was suggested that this observation could underpin empathetic behaviour rather than general face identity abilities. However, the authors also suggested that the results were preliminary and caution should be taken, as other

possible explanations (such as level of experimental difficulty) could not be ruled out.

Briefly, there is no doubt that face-processing is a relative strength in WBS, but it would be wrong to maintain earlier claims of normally developing face recognition which were based on behavioural scores of older children and adults with WBS, and frequently in comparison Down syndrome. Moreover, several behavioural and electrophysiological studies point towards atypical development of face processing in WBS compared to controls.

#### ***2.4.4.2 Visuo-spatial Cognition***

Individuals with WBS consistently display poor performance on visuo-spatial tasks (e.g. Bellugi et al., 1988, Bellugi, Wang & Jernigan, 1994; Bahrle, Bellugi, Delis & Marks, 1989; Rondan, Mancini, Liver & Deruelle, 2003; Wang, Doherty, Rourke & Bellugi, 1995). They tend to show severe difficulties on standardised tasks such as block design task <sup>(8)</sup>, Raven's Progressive Matrices, Benton Line Orientation and drawing task <sup>(9)</sup>, and freehand drawings.

They typically score worse than their full IQ scores would predict, and worse than overall mental age comparison control groups (e.g., Bellugi et al., 1988; Bellugi, Bahrle, Neville, Doherty & Jernigan, 1992; Mervis et al., 1999). It is worth mentioning that no relationship has been found between early visual problems and visuo-spatial functioning (Atkinson, Anker, Braddick, Nokes, Manson et al., 2001). Findings from the block design task (known as pattern construction in British Ability Scale) have been particularly influential, thus considered in more detail below.

The block design task requires the participant to assemble a number of blocks into a coherent whole pattern that resembles a model image. There is no memory constrain, since the model remains visible throughout. Studies report that individuals with WBS fail to reproduce the trials even at a very simple level, as they tend to focus on

---

<sup>8</sup> A subtest of Weschler test that is equivalent to Pattern Construction subtest of the British Ability Scale subtest.

<sup>9</sup> This test forms part of the Boston Diagnostic Aphasia.

individual blocks. Bellugi and colleagues found that during a block construction task the individuals with WBS selected the appropriate blocks but failed to produce the whole configuration of the patterns (termed here as “holistic”). They proposed that the deficits arise from processing biases (i.e., preference to attend to featural information), (Bellugi et al., 1994; Bellugi, Korenberg, & Klima, 2001; Bellugi et al., 1988; Bellugi, et al., 1992; Dall’oglio & Milani, 1995; Frangiskakis et al., 1996; Howlin, Davies & Udwin, 1998; Udwin, Yule & Martin 1987).

Farran et al. (2001) showed that people with WBS have normal facilitation of block design performance when the target design is segmented (Farran, Jarrold, & Gathercole, 2001; Mervis et al., 1999). Farran and colleagues suggested that poor performance on the block construction might be related to inability to use mental imagery rather than a featural processing bias. Twenty-one individuals with WBS and 21 TD control group matched on Ravens Coloured Progressive Matrices (RCPM) participated in the study. The authors employed a novel two-dimensional block construction task that was manipulated to investigate processing preference and the ability to use mental imagery. The results demonstrated that in the WBS group response accuracy decreased dramatically when rotation of the pattern was more than 60 degrees. They argued that individuals with WBS can perceive information at both the local and global levels, but that they have difficulty using this information in visuo-spatial construction tasks at the global level (Farran & Jarrold, 2002). However, Farran has subsequently argued that holistic processing was atypical in the WBS, based on their performance on perceptual grouping tasks (Farran, 2005).

Hoffman, Landau and Pagani (2003) attempted to deconstruct WBS performance on the block construction task and argued that the task involves many capacities including perception, spatial working memory and executive planning processes. The authors used two kinds of block construction tasks with different levels of difficulty. A simple puzzle contained block of the same colour and complex puzzles contained an arrangement of two colours. The results showed that individuals with WBS performed at normal levels for simple puzzles, but were impaired on complex

puzzles task. This finding suggests that individuals with WBS can successfully form perceptual groupings of small sizes.

However, Pani, Mervis, and Robinson (1999) found in a visual search task that the performance of individuals with WBS was highly similar to controls matched on CA and gender. The authors reported that individuals with WBS struggled to disengage from global processing if the task required local processing for success. This suggests that they demonstrate global precedence in visual search. Pani et al. (1999) have suggested that visuo-spatial constructive deficits result from a general weakness in planning and organising information in working memory. However, it is noteworthy that success on a holistic visual search task is not comparable with failure on a block construction task. The integration of the featural information, the spatial relations between the features and the holistic information that is required in a block design task are far more demanding and thus comparison across tasks is difficult.

Evidence of a featural bias also comes from drawing studies. In a freehand drawing task of common objects such as bicycles, individuals with WBS tended to draw only the features of the objects and failed to produce the overall configuration of the object (Stiles, Sabbadini, Capirci & Volterra, 2000). Similar patterns were found when individuals with WBS were tested on Navon-type stimuli. This task involves the reproduction of drawings of alphabetical letters, for example Ys arranged in the shape of a letter D, thus representing individual features and holistic levels respectively. As illustrated in Figure 2.3, numerous studies have reported that individuals with WBS (unlike control groups) attended to individual letters (features) at the expense of the holistic form of the image (Bihrlle et al., 1989).

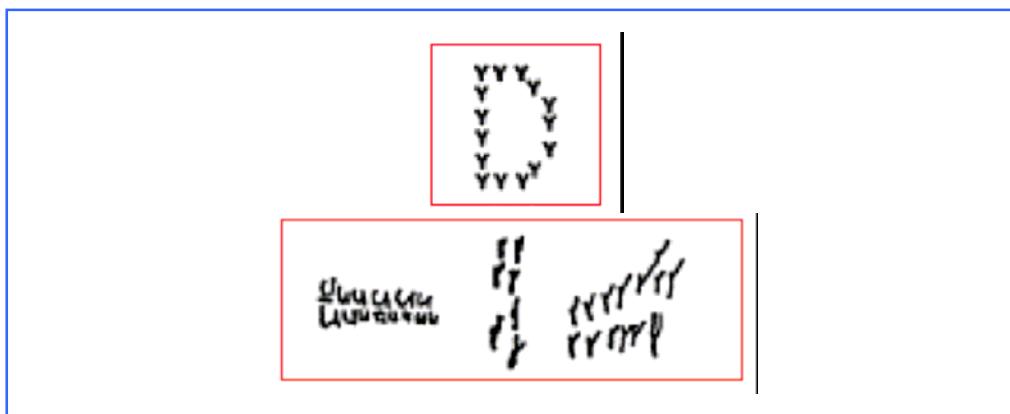


Figure 2.3: Illustrates an example of WBS performance on the Navon task. Adapted from Bihrl et al. (1989).

Farran, Jarrold, and Gathercole (2003) employed perceptual and drawing versions of the Navon task. They confirmed that in construction tasks, individuals in WBS group were attending only to local elements of the Navon letters. However, on the perceptual version of the task, individuals in WBS group attended to both local and global levels of the stimuli, demonstrating a similar pattern of processing as TD control group (matched on NVMA or on CA). The authors concluded that individuals with WBS are able to attend to the holistic level, but in the case of drawings where output is required, featural elements become more salient than holistic ones. These conclusions were further examined by Abreu, French, Annaz, Thomas and de Schonen (2005) who tested 13 children with WBS (mean age: 8:8) and explored their sensitivity to featural versus holistic levels in a perception task that incorporated static and moving versions of computerised Navon-type stimuli. Their preliminary findings showed that children in the WBS group exhibited perceptually equal sensitivity to both levels of processing compared with children in the TD group (matched on CA), supporting Farran's claims.

Neuroimaging studies have looked for causal evidence of the poor spatial abilities in WBS. For example, Atkinson and colleagues have suggested that there may be a deficit in frontal function linked to the selection and control of spatial behaviour. Atkinson and colleagues have also proposed that spatial deficits in WBS are associated with deficits at the higher levels in the dorsal stream (MT/V5). This view

is supported by evidence of abnormal motion coherence, a test of dorsal stream function, but normal form coherence that involves the ventral stream (Atkinson, King et al., 1997; Atkinson, Braddick et al., 2005).

The evidence from the studies outlined above points to differences in visuo-perceptual and construction abilities in individuals with WBS. Although, individuals with WBS seem to follow an atypical processing pathway, they show worse performance on visuo-constructive tasks than on visuo-perceptual ones (see also Deruelle, Rondan, Mancini, & Livet, 2005, for discussion).

#### ***2.4.4.3 Language***

Language is one of the domains of ability that has often been characterised as ‘intact’ in individuals with WBS. Bellugi et al (1988) claimed that expressive language in WBS is “complex in terms of morphological and syntactic structure, including full passives, embedded relative clauses, a range of conditionals and multiple embeddings”. Clahsen and Almazan suggested that grammar develops normally, despite memory for vocabulary being impaired in individuals with WBS (Clahsen & Almazan, 1998).

In contrast, other groups have reported that individuals with WBS develop language significantly later and differently from their peers. Karmiloff-Smith and colleagues (1997) suggested that it is unlikely that any aspect of language can be truly unimpaired. On closer inspection, they have found in several studies that not only is language development delayed in WBS, but also that its trajectory of development differs from that of controls (e.g., Laing, Hulme, Grant, & Karmiloff-Smith, 2001; Mervis, et al., 1999; Thomas et al., 2001; Zukowski, 2001).

Further support for the atypical functioning and/or organisation of language in individuals with WBS was demonstrated using electrophysiological studies. In their ERP study, Neville, Mills, and Bellugi, (1994) investigated brain activation to grammatical information and open-class words which convey meaning. For the MA-matched control group, open-class words elicit an N400, which is larger over the posterior right hemisphere, whereas for the grammatical words they elicit an N400

which is more anterior and left lateralised. In contrast, people with WBS showed no difference between the classes of the words, and all the N400 were lateralised to the left. The results suggest an atypical organisation of language functions in individuals with WBS.

## **2.5 SUMMARY OF DISORDERS**

In the light of the above evidence, it is plausible to assume that individuals with autism attend to featural stimuli in an atypical manner (in line with WCC theory). In general, individuals with autism perform poorly on social stimuli such as faces, but their constructive skills fall within the normal range on behavioural tasks. In the majority of studies, children with high-functioning autism and Asperger syndrome were recruited. Thus, role of severity of the disorder has not been addressed and correlated with any of the experimental outcomes.

The WBS profile seems to be somewhat different from the individuals with autism, because those with WBS are fascinated by faces. People with WBS perform well on face recognition tasks in contrast to individuals with autism, but it has been suggested that both groups display a configural deficit. Another contrasting aspect between these groups is their difference in performance on constructive skills, with WBS performing very poorly and individuals with autism very well.

Lastly, it is difficult to characterise visuo-spatial profile of individuals with Down syndrome. Some studies suggest that they have a ‘global style’ of processing, but how this relates to configural or holistic encoding remains to be addressed. Table 2.1 illustrates summary of visuo-spatial profiles of each disorder group.

Table 2.1: Summary of visuo-spatial profiles of each group (relative to overall MA).

PROFILE	HOLISTIC	CONFIGURAL	FEATURAL	CONSTRUCTION	PERCEPTUAL
<b>AUTISM</b>	UNKNOWN	UNKNOWN	GOOD	GOOD	POOR
<b>DS</b>	GOOD	UNKNOWN	UNKNOWN	POOR	GOOD
<b>WBS</b>	UNKNOWN	UNKNOWN	GOOD	POOR	GOOD

Finally, in order to use the theoretical perspective summarised in the introductory section of this chapter, it is crucial to adapt the methodological design. There a number of methodological issues such as the comparison of patient-control groups which are carefully examined and discussed throughout this thesis. The methodological issues will be reviewed and discussed in the next chapter.

A number of studies suggest differences between visuo-spatial perceptual and construction tasks. In the current thesis an attempt was made to build developmental trajectories of each disorder group on each task, to compare them directly to typical developmental trajectories. The main question whether children in clinical groups develop sensitivity to holistic processing (Study 1) and configural processing (Study 2 and Study 3). Constructive abilities during the face processing task were also tested (Study 4) and compared to the perceptual performances of the all groups. More specific research questions are introduced in the relevant chapters.

---

## CHAPTER 3

### GENERAL METHODOLOGY



*Do you see a young woman or an old woman?*

### **3.1 INTRODUCTION**

Typically developing children and children with developmental disorders took part in this research project. In the following chapter, methodological issues surrounding matching strategies are evaluated, the participants' characteristics, general data collection methods and analyses will be described.

### **3.2 GENERAL METHODS ADOPTED IN THIS THESIS**

#### **3.2.1 Participants**

##### ***3.2.1.1 Recruitment and Diagnosis***

All studies were initially approved by Birkbeck College Ethics Committee, prior to recruitment of participants. In the case of participants with WBS, a formal research proposal was submitted and accepted by the WBS Professional Advisory Panel. In addition, the experimenter gave a presentation about the studies and its objectives to the schools involved in the project.

Each parent was sent a consent letter and detailed information about the studies. Parents then returned a slip indicating whether or not they would like their children to take part in the studies. Some parents did not wish their children to be videoed during the testing sessions, thus we defined full consent as parents giving consent to video their children. In partial consent video recording was not allowed during the session.

Participants in all groups had normal or corrected to normal colour vision on the basis of parental, teachers' reports or self-reports. Handedness was ascertained in the same way.

##### **Typically developing control group**

Children in the TD group were recruited from mainstream schools and a nursery in north London. The children were randomly selected from the required age group.

*Children with developmental disorders*

Children with autism and DS were recruited from the London area schools and via charities such as Parents for Down syndrome (London Borough of Barnet). Children with WBS were recruited through The Williams Syndrome Foundation, and came from throughout the UK.

All of the children in the autism group were already clinically diagnosed with autism according to DSM-IV criterion. Children in the DS group are known to have tested positively for trisomy of chromosome 21. Children with WBS had been diagnosed clinically as well as by means of the fluorescence in situ hybridisation (FISH) genetic test for microdeletion in specific gene markers.

The heterogeneity within the autism population is substantial. For instance, some children may avoid any eye contact and show no interest in others, whereas other children may attempt to interact with people in various ways. Also, IQ levels range from profound learning difficulties to well above average across children with autism.

The majority of the current studies recruit high functioning children with autism and/or Asperger syndrome. The aim of this study was to recruit children with different functioning levels. The rationale for splitting the children with autism into two groups, high and low functioning groups, (HFA and LFA) was to i) enable us to compare our HFA group performance with the previous literature and ii) investigate whether children in the LFA group produce a different developmental profile.

Children from autism group were further divided into HFA and LFA according to Childhood Autism Rating Scale (CARS) (Schopler, Reichler, & Rochen- Renner, 1993). In this test, children are rated on 15 categories using a 7-point scale. The categories include: imitation, emotional response, body and object use, visual and listening response, verbal and nonverbal communication, adaptation to change and activity level. After the child had been rated the scores from all categories were pooled. Children with a score above 30 points were categorised as autistic<sup>(1)</sup>.

---

<sup>1</sup> All of the children in our study scored above 30 points (autistic category)

Those falling within the autistic range were divided into two categories 1) mild-to-moderate autism (30 - 36.5 points) or 2) severe autism (37 - 60 points). In the present thesis, children from the former were assigned to HFA group, and from the later to LFA group. The categorisation system is based on a comparison of CARS scores with the corresponding expert clinical assessments of over 1,500 children. The distributions of CARS scores for these participants can be found in Appendix A.

Rellini, Tortolani, Trillo, Carbone, and Montecchi (2004) recently examined the reliability of the CARS. They assessed 65 children aged 18 months to 11 years on CARS and Autism Behaviour Checklist (ABC) in correspondence to DSM-IV.

The CARS identified 100% of cases of autism, whereas the ABC identified only 54%. Also, Saemundsen, Magnússon, Smári, and Sigurdardóttir (2003) examined sensitivity of the CARS and the Autism Diagnostic Interview-Revised (ADI-R) in diagnosing autism and found the CARS significantly better than ADI-R in classifying young children with autism.

### ***3.2.1.2 Response rates***

#### ***Typically developing control group***

Thirty parents were asked for their consent to test the children; also each child was verbally asked whether he or she wished to participate in the studies. Twenty-seven parents gave consent and two children declined to participate in the studies. Although most parents gave full consent, current internal rules in the mainstream schools did not permit the use of video equipment during testing sessions.

#### ***Children with developmental disorders***

**Autism groups:** 42 parents were approached for their children to participate in the study. 39 parents gave consent for their children to take part in the study, 3 parents declined due to their emotional problems related to the diagnosis of their children. 27 full consents and twelve partial consents were granted. 6 children were later excluded from the testing for a number of reasons (see section 3.2.1.3.). Thus, the final sample comprised 33 children.

**DS group:** 23 parents were approached to allow their children to participate in the study. 17 parents gave consent for their children to take part in the testing. 8 full consents and 9 partial consents were given. 2 children were excluded from the final sample. The final sample comprised of 15 children.

**WBS group:** 19 parents were contacted and 18 gave full consent for their children to take part in the studies. 1 family declined for their child to participate in the project.

### ***3.2.1.3 Exclusion criteria***

The initial aim of the thesis was to test each participant on each study, however this was not possible in practice. Some individuals moved schools, or were away from the country for some time. Also, some children were unable to take part in the studies due to their behavioural or emotional problems. These factors contributed to slight variations in participant number in each study.

In line with many studies of children with developmental disorders, not all the children tested were included in the final sample. Children were excluded for failure to complete the study due to tantrums, attentional problems or poor language comprehension. Some children were excluded before any testing had started, due to other medical problems. Other reasons for excluding children were specific to the experimental paradigm being used. Children described in Table 3.1 were not able to participate in all of the studies. Although care was taken to test all the children, some individuals were able to participate in only some studies.

Table 3.1: Summary information on participants excluded from the studies.

NO.	GROUP	CA	LANGUAGE	BEHAVIOUR
1	LFA	8:2	Non-verbal	Doesn't recognise parents' photos, tantrums
2	LFA	6:5	Non-verbal	Doesn't recognise parents' photos, tantrums
3	LFA	9:4	Non-verbal	Poor language comprehension
4	LFA	7:4	Non-verbal	Severe tantrums
5	HFA	6:1	Verbal	Colour-blind, tantrums
6	LFA	10:8	Verbal	Severe tantrums
7	DS	9:7	Verbal	Poor language comprehension
8	DS	7:6	Verbal	Poor language comprehension

### 3.2.1.4 Ages chosen for studies

TD children participating in the studies ranged from 3 to 12 years old, and children in clinical groups ranged from over 5 years of age to 12 years old (see details in Table 3.2). The gender bias for the autistic groups is characteristic of this disorder. The age distribution for all participants is shown in Figure 3.1.

Table 3.2: Chronological age details for all groups

GROUP	NO.	MEAN (yrs in months)	SD	AGE RANGE	GENDER	
					GIRLS	BOYS
TD	25	7:2	2:8	2:9-12:5	13	12
HFA	16	8:5	1:8	5:4-11:2	3	13
LFA	17	8:6	1:8	5:3-11:4	2	15
DS	15	9:0	1:9	6:1-12:5	5	10
WBS	18	8:6	2:0	5:8-12:1	8	10

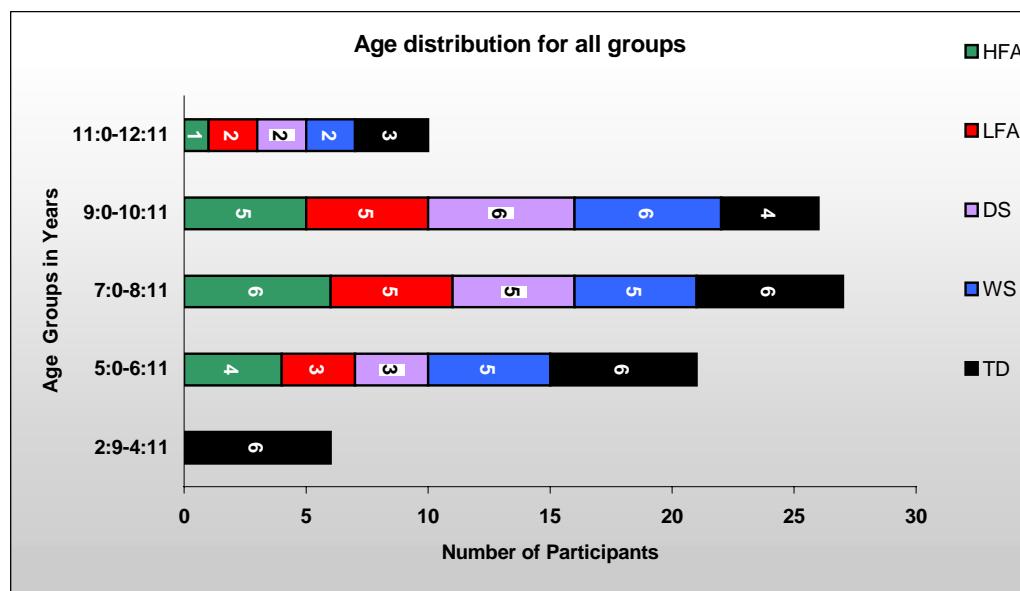


Figure 3.1: Age distributions of each group.

### **3.2.1.5 Parental educational level and socio-economic-status classification (SES)**

All parents from clinical groups were given a questionnaire regarding their SES (See Table 3.3) and education level (see Table 3.4) Occupations were classified according to the Standardised Occupational Classification 2000 (Office of National Statistics, 2004). Note that children with autism are split here into HFA and LFA groups.

Table 3.3: Classification of parental SES (shown in %).

SES	HFA		LFA		DS		WBS	
	M	F	M	F	M	F	M	F
Managers and senior officials	6	13	-	6	-	13	-	11
Professional occupations	38	50	29	41	27	40	28	45
Associate professional & technical occupations	19	25	12	18	33	21	11	17
Administrative and secretarial occupations	31	-	35	-	20	-	44	-
Skilled trades occupations	-	6	-	12	-	13	-	11
Personal service occupations	6	6	6		13	-	17	-
Sales & customer service occupations	-	-	6	6	-	-	-	-
Process, plant and machine operatives	-	-	-	-	-	-	-	-
Unemployed (not classified)	-	-	12	-	7	-	-	-

Note: M= Mother, F= Father. In some cases fathers' details were not provided.

Table 3.4: Details of parental education level (shown in %).

EDUCATION	HFA		LFA		DS		WBS	
	M	F	M	F	M	F	M	F
None	-	-	-	-	-	-	-	-
GCSE/O-Level	6	6	12	6	13	7	11	6
Secretarial/Technical	18	13	17	12	20	20	17	6
A-Level	13	13	24	18	27	13	17	11
Professional	6	18	12	6	7	7	11	11
University degree	57	50	35	41	33	40	44	50

Note: M= Mother, F= Father In some cases fathers' details were not provided, and so these columns do not total to 100%.

### 3.2.2 Challenges of testing children with developmental disorders

As many as 50% of children with autism lack verbal communication (Volkmar & Klin, 2000), thus most children in the LFA group use Picture Exchange Communication System cards (PECS) and Makaton sign language to communicate with others. These tools were used in the current project to familiarise the children with the experimenter as well as the testing procedure. PECS cards are one of the most effective tools of non-verbal communication with LFA children (Ganz, & Simpson, 2004; Magiati, & Howlin, 2003). Also, this method of communication proved to be useful with some of the children with DS, who had poor verbal communication.

Given the cognitive and attention difficulties in children with developmental disorders, it is important to make the tasks entertaining, attention grabbing and short. In the interests of collecting meaningful data, often it is necessary to have several testing sessions. Thus, studies in this thesis were designed or divided into sections that did not last longer than 15 minutes. This was particularly useful for children with DS and LFA group. It is frequently necessary to use rewards such as stickers, and praise to motivate the child to elicit an appropriate response.

Moreover, the testing environment can vary substantially between participants, which may affect their performance. All efforts were made to test children at school and keep the testing sessions short (depending on the child's abilities). Testing sessions were scheduled in the mornings from Monday to Thursday, as children were tired and non-compliant in the afternoons and on Fridays. There was a maximum of two-week interval between consecutive testing sessions.

The final issue concerned group sizes. Given the rarity of WBS, and difficulty in gaining access to children with DS or autism, it is often challenging to recruit large groups of children with these disorders, especially if the study is focusing on a relatively narrow age range. However, it is important to employ sufficient participant numbers to disentangle systematic variation between groups from random noise, particularly if the tests involved produce only a small range of scores.

While case studies and studies of small groups provide useful starting points for further investigation, caution should be exercised when generalising such results to the syndrome/disorder as a whole (Grelotti et al., 2005; Happé, Malhi, & Checkley, 2001; Stiles, Sabbadini, Capirci, & Volterra, 2000; Temple, 2003). Conversely, it is also important to realise that the average profile may not be representative of all individuals. Hence, choice of appropriate testing tools, analysis and the level of task difficulty needs to be carefully considered (Ansari, Donlan, Thomas, Ewing, Peen & Karmiloff-Smith, 2003; Farran, Jarrold, & Gathercole, 2003; Jarrold & Brock, 2004; Mervis & Klein-Tasman, 2000).

### **3.2.3 Stimuli and Apparatus**

#### ***3.2.3.1 Stimuli***

All studies in this thesis used face photographs. Colour photographs were used where possible to retain the maximum amount of information in the faces. However, in cases where stimuli were supplied by other research groups, it was sometimes necessary to use black and white pictures (e.g., Study 2 - Jane faces) in order to allow comparison to previous studies. All faces were of young to middle-aged Caucasian adults (with the exception of Study 3 where photos of children's faces were used).

Some of the face photographs were manipulated using specialist computer software such as Faces 3.0 developed by InterQuest (Study 1 - rotating faces) and Morphed software (Study 4 - construction). Details of the manipulations are given in the relevant chapters. All face stimuli were presented full frontal with neutral facial expressions.

### **3.2.3.2 Apparatus**

Studies 1, 2 and 3 were run on a Dell laptop and presented on a 17" touch-screen monitor. Stimulus presentation was controlled using the SuperLab Pro v.2.0 software package. The equipment used in Studies 4 is detailed in the relevant chapters.

### **3.2.4 General Procedure**

#### *Typically developing children*

Each participant was taken to a quiet experimental room on two different days. Younger children (under 5 years old) were visited 3 times depending on the individuals' needs, again with a maximum interval of two-week period.

#### *Children with developmental disorders*

The experimenter visited each child at least once prior to the testing sessions. Each child was taken into the experimental room only when the experimenter was satisfied that he/she was not distressed or unwell. All participants were tested individually. Experimental rooms used for children with autism were very plain with no equipment to minimise distractions. Equipment other than that used in the study was hidden from the child's view.

Each child was taken to a quiet experimental room with the investigator and sometimes with a learning support teacher. Learning support teachers were requested to refrain from any form of communication with the children during the testing sessions, unless it was absolutely necessary. The presence of the learning support teacher was not necessary on most occasions (92%).

### ***3.2.4.1 Standardised measures***

Participants completed four standardised tests. These were two subtests of the British Ability Scales II: pattern construction and copying task (Elliott, Smith, McCulloch, 1996), British Picture Vocabulary Scale (Dunn, Whetton, & Burley, 1997) and the Benton Facial Recognition Test (Benton, Sivan, Hamsher, Varney, & Spreen, 1983). See chapter 4 for detailed description and results of these tests.

## **3.3. DATA COLLECTION**

### **3.3.1 Dependent variables**

One of the dependent variables for Studies 1, 2 and 3 was the number of correct responses. Another dependent variable was response time (RT) measurement (Study 1 and 3). In order to improve data quality and decrease the noise, cues such as fixation crosses were used to warn the participants about the appearance of the stimuli. RTs were analysed only for correct responses and used in study 1 and 3. In RTs analyses, median RTs were calculated instead of using means, since the latter approach required excluding outliers. For Study 4 (Construction Task), numbers of correct responses was analysed.

## **3.4 METHODOLOGICAL ISSUES AND DATA ANALYSIS**

### **3.4.1 Matching strategies**

#### ***3.4.1.1 General Issues***

Many research studies on developmental disorders such as WBS and autism employ partial or general measures of IQ for matching with a control group (e.g., Klin et al., 1999; Pezzini, Vicari, Volterra, Milani, & Ossella, 1999; Plesa-Skwerer, Sullivan, Joffre, & Tager-Flusberg, 2006; Temple, Almazan, & Sherwood, 2002; Teunisse, & de Gelder, 2003; Tager-Flusberg, & O'Sullivan, 2000; Tager-Flusberg et al., 2003). Also, individuals with WBS or autism have often been matched to individuals with DS or other clinical groups based on their cognitive abilities in this manner (e.g., Bellugi et al., 1988, 1990, 1994, Bahrle, Neville, Doherty, & Jernigan, 1992; Plesa-

Skwerer et al., 2006). The results obtained using this approach may be ambiguous, since individuals with WBS perform better on language tasks than individuals with DS.

Similarly, if we match people with DS or WBS with autism group on visuo-spatial abilities, the cognitive profile of the people with autism would be overestimated. Comparisons with younger TD individuals are also common (e.g., Lakusta, & Landau, 2005; Pléh, Lukács, & Racsmány, 2003). In a recent study participants were matched on an area of their special interest (Grelotti et al., 2004).

The problem which investigators face is that experimental and control groups need to be compared or matched on some basis. One obvious criterion for matching is on chronological age, perhaps also matching for gender. However, to assess if a skill is in line with an overall pattern of delay in some cognitive domain, it is desirable to match a control group based on “mental age” (MA), for instance in language ability or visuo-spatial skills. In recent years the technique of matching individuals for mental age has come under scrutiny (Jarrold & Brock, 2004). For example, Karmiloff-Smith, et al., (2004) used a developmental trajectories approach in their analysis, instead of individual matching on MA or any standardised test.

### **3.4.2 Developmental trajectories**

To gain a fuller understanding of whether and how the development of cognitive competencies in clinical populations differs from typical development, one may build ***developmental trajectories*** (theoretical background discussed in chapter 2). This approach seeks to build a task-specific typical developmental trajectory by measuring performance across a range of ages in the normal population. Firstly, given an individual with a disorder, one can then establish whether his/her performance fits *anywhere* on the typical trajectory. This comparison is theory neutral. Secondly, one can assess whether the individual fits on the trajectory at the position predicted by their CA. Finally, by utilising one (or more) tests of “mental age”, one can assess whether the individual fits on the normal trajectory according to their general level of performance in this (or other) domain(s).

Given a group of individuals with a disorder who span an age range, it becomes possible to construct an atypical developmental trajectory for this particular disorder and contrast this against the TD group. This approach offers a more direct way of addressing the question “Does the target behaviour develop normally or atypically in the disorder?” Later, it will become more apparent that studying disorders can lead to a reconsideration of the notion of delay, in the question that is sometimes asked of disorders: “Is the target behaviour in this disorder atypical or simply delayed?”

The best and most informative way of gaining an insight into how developmental changes occur in clinical groups or typically developing individuals is to conduct longitudinal studies. However, these studies are highly time-consuming and may put parents, children and teachers under a lot of pressure. An alternative to the longitudinal method and one that is used in this thesis is to build *developmental trajectories* by means of a cross-sectional design. This approach has been successfully used in recent studies (Karmiloff-Smith et. al., 2004; Thomas, et al. 2001). Thomas et al. (2001) compared individuals with WBS with four groups of TD controls (6, 8, 10 year olds and adults) on 2 past-tense elicitation tasks. This method allowed the authors to establish which level of typical development best fitted the performance of the WBS group.

### 3.4.3 Data analysis

A focus on developmental trajectories led to the use of the General Linear Model (GLM) in the current thesis. Linear methods were used to derive a relationship between age and performance (accuracy and RT), and where necessary, variables were transformed to ensure that the relationship between age and performance was approximately linear.

A linear regression line has an equation of the form  $Y = a + bX$ , where  $X$  is the explanatory variable and  $Y$  is the dependent variable. In the current analyses the  $Y$  is accuracy or RT,  $X$  is the test age or raw score (in case of Benton test). A positive gradient means that performance is increasing with age. Conversely, a negative value of  $b$  means that performance decreases with age.

A regression line may be plotted to any data, so it is essential that the line reflects a true relationship between the variables. Therefore it is important that the regression line accounts for a certain portion of the variability in the data. If  $R^2 = 1$ , the regression line is a perfect fit between the dependent and independent variables. Outliers may have a major impact on regression line therefore the distribution of the data was checked for significant deviance from normality by visual inspection of the frequency histograms for the data in each condition. Also, Cook's distance and Levene's distance were inspected to determine whether a particular data point alone affects regression estimates. If a particular data point was deviant, the regression line was considered without it to check whether the point influenced the significance of regression results.

#### **3.4.4 Floor and ceiling effects**

Ideally, all groups should fall in the sensitive range of the test used. However, when comparing disorders and control groups, this is not always possible. One potential problem in assessing trajectories is that of floor and ceiling effects. These can mask group differences or produce artifacts in trajectories. For example, individuals with HFA and TD controls may both be at ceiling but this does not mean that there would be no group differences if the test were more difficult (see chapter 4). Despite this caveat, it is often the case that ceiling performance in clinical groups such as WBS is taken as indicating normal development (see e.g., Clasen, & Almazan, 1998). In a similar way, floor effects, where the individuals are scoring at the lowest level, are also problematic (see, for example, Tager-Flusberg et. al., 2003).

If individuals arrive at ceiling level right from the youngest age tested, or do not improve with time (floor level), then two options should be considered: i) an in-depth data analysis that explores individual cases or, ii) the choice of task should be reconsidered to include a more sensitive measure of performance.

In the current thesis, there were instances of floor and ceiling scores in some groups that led to a potentially misleading intercept or gradient values for the developmental trajectories. We dealt with these problems in the following manner: if there was a floor effect but subsequently i) the scores did increase, then the intercept value was

described; ii) if there was a floor effect but the scores steadily improved above certain age, then we analysed just the section of the trajectory where performance was above the floor level. For the ceiling effects, a similar approach was taken.

### 3.4.5 Definition of delay adopted in the analysis

There is a myriad of terms used to describe individuals whose behavioural characteristics do not fall within the “normal” range. Researchers often describe participants who score in normal level on standardised scores as developing normally (“intact”, “spared” and “preserved”).

Can behavioural scores tell us that underlying cognitive processes have developed normally? Karmiloff-Smith, and Thomas (2003) argued that achievement of normal behavioural scores can be supported by different cognitive processes, compared to TD individuals. For example, individuals with WBS have good face recognition but they use different processes (see chapter 2 for discussion).

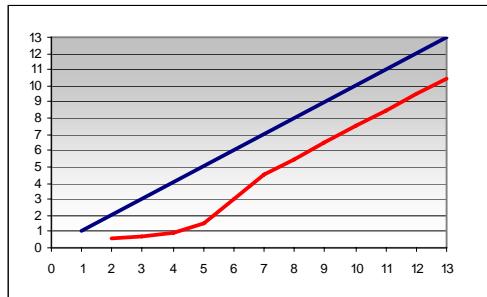
Another problem in using those terms is to classify the developmental trajectories using a linear regression analysis. Therefore, henceforth we use an analytic approach based on linear regression models. The term *delay* will be used with additional descriptors of the trajectories (for instance chapter 4). The terms relating to delay will be descriptive of behaviour and no claims about mechanism are made here. The typical developmental trajectory (normal onset & rate) is contrasted with other developmental trajectories that may be described as follows:

- a) *delayed onset* trajectory implies normal rate (not statistically different from normal trajectory, but different intercept),
- b) *delayed rate* of developmental trajectory implies normal onset but slower rate of increase in performance,
- c) *delayed rate & onset* includes both points a & b,
- d) *zero rate* (gradient not significantly different from 0) of development describes trajectory that does not increase with age.

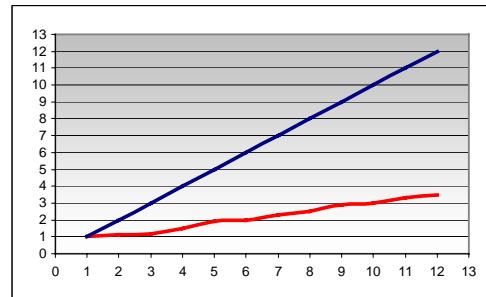
Graphic demonstration of these developmental trajectories, are shown in Figure 3.2. For simplicity of the analysis, only linear regression analyses were carried out in this thesis. However, if needed the data points were transformed to improve linearity.

Another example of non-linear trajectories that were present in the current analysis is called here a ***premature plateau*** (Figure 3.2-e) where development ceases to improve beyond certain age. This constitutes a marked reduction in rate of performance and is independent of onset (normal onset).

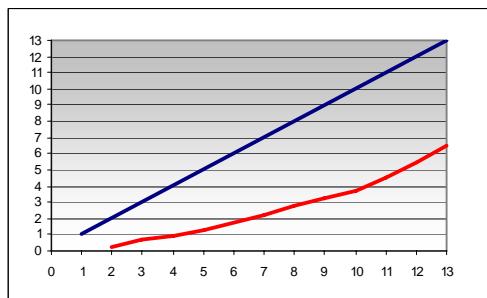
a) Delayed onset



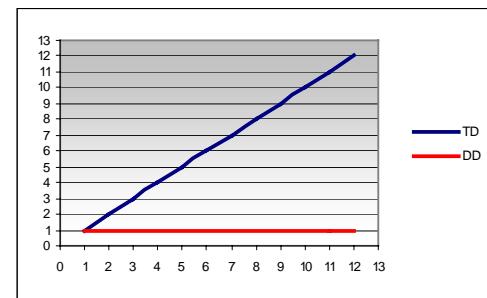
b) Delayed rate



c) Delayed onset & rate



d) Zero rate



Graphic representation of non-linear regression that occurred in the current studies

e) Premature plateau

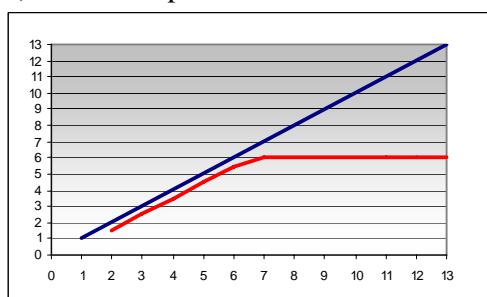


Figure 3.2: Graphic description of developmental delay. Blue line shows a normal development (TD), red line shows developmental disorder trajectory (DD). X-axis represents age, y-axis represents performance scores.

---

## CHAPTER 4

### STANDARDISED TESTS



*Can you see a face or a man playing a saxophone?*

## **4.1 INTRODUCTION**

In order to explore the verbal and non-verbal contribution to the development of face recognition in both TD children and those with developmental disorders, measures of both verbal and visuo-spatial competence were obtained from participants in all groups.

This thesis is one of the first to focus in detail on the developmental trajectories generated by these developmental groups across the age range studied (developmental trajectories are discussed earlier in chapter 3.4.2). The BPVS and two subtests of British Ability Scale (BAS) were employed: pattern construction (PC) and copying task (CT). Also, the Benton Facial Recognition Test (Benton test) was utilised. Although this test is widely used to compare disorder groups it has not been strictly standardised.

### **4.1.1 Questions and Hypotheses**

The literature reviewed in chapter 1 and 2 raised a number of general questions and suggestions which have motivated the experimental studies presented within this chapter:

1. Does performance on the standardised visuo-spatial tests and the Benton test increase in line with CA in children with developmental disorders as observed in the TD control group?
2. Based on the previous research literature it is predicted that children with WBS would perform at normal/near normal scores at BPVS and Benton test and below normal range on copying and PC test.
3. Does performance on the standardised tests and the Benton test differ between HFA and LFA groups, given that the groups were divided on the CARS, which is not specifically cognitive?

In the following sections, the results from the standardised tests will be summarised for (1) the TD children, (2) the children with autism split into HFA, and (3) the LFA, (4) the children with DS, and (5) the children with WBS. For each test, the developmental trajectory for the TD control group is considered. For the standardised tests, one would expect this group to demonstrate a one year increase in test age for each year of increase in CA. The trajectories for each clinical group will be compared to the TD trajectory.

Finally, the reliability of any apparent differences between disorder groups will be clarified (where the reliability of these differences is potentially invalidated through multiple post-hoc comparisons, this will be noted).

## **4.2 BRITISH PICTURE VOCABULARY SCALE (BPVS)**

The BPVS is a receptive vocabulary test. The test is used from age three to fifteen. Thus, the minimum floor score is 28 months and ceiling score is 204 months. Details of the test can be found in BPVS II Manual (Dunn, et al., 1996).

### **4.2.1 Method**

#### ***4.2.1.1. Participants***

In the current test, one boy in WBS group refused to participate in the test, so that the WBS sample was N = 17, mean age = 8:04, SD = ±2:10, age range = 5:08-12:01. Details of participants are in chapter 3, section 3.2.1.

#### ***4.2.1.2 Procedure***

The BPVS test requires the experimenter to assess accuracy only. There is no time limit and children are given practice trials. During the test the child is asked to choose a picture that illustrates the meaning of a word presented orally by the experimenter. There are four possible pictures. General procedure of the methodology is outlined in general methods chapter 3 section 3.4.

### 4.2.2 Results

Figure 4.1 depicts the performance of each group in terms of test age [TA] plotted against increasing chronological age (CA). The solid lines indicate a best-fit regression through each group's data, and the dashed line indicates floor performance on this test. If the TD group is similar to the sample on which the BPVS was standardised, there should be a tight relationship between TA and CA, and a linear trajectory with a gradient of 1. For each year a child gets older, his or her test age should increase by one year.

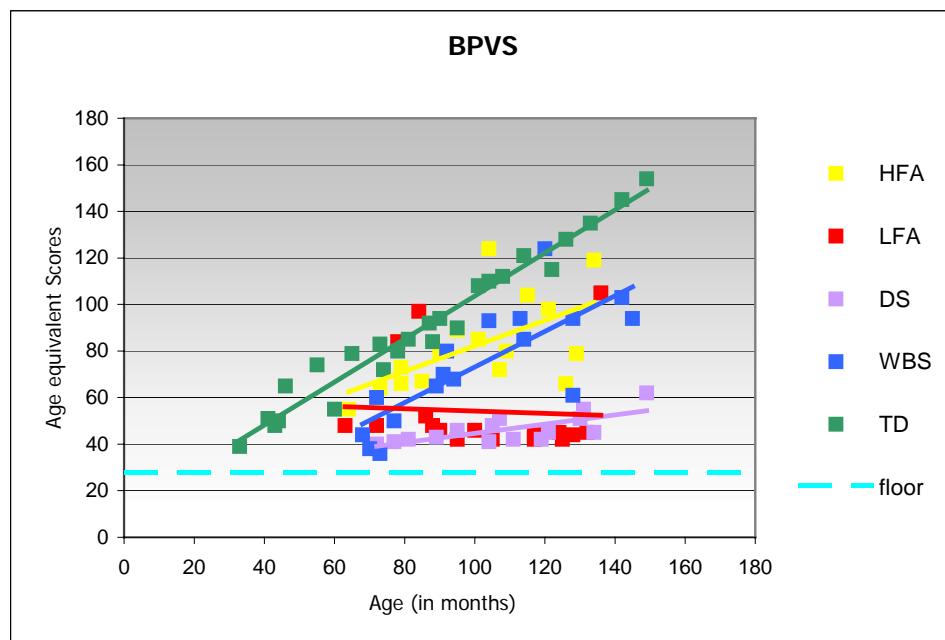


Figure 4.1: BPVS age equivalent scores for all groups plotted against their CA.

#### **4.2.2.1 TD control group**

The trajectory generated by TD group accounted for almost all variance in the group ( $R^2 = .96$ ,  $F(1, 23) = 591.33$ ,  $p < .001$ ). The trajectory indicated that our sample had a BPVS test age improving sharply (gradient:  $.92$ , [.85, 1.00]<sup>(1)</sup>), and had a slightly earlier onset of development on this test (intercept:  $11.23$  [3.10; 18.51]). The control sample therefore generated a valid trajectory of receptive vocabulary development.

#### **4.2.2.2 HFA group**

The HFA group demonstrated more variability in its trajectory than the TD group. Nevertheless, the trajectory still accounted for a third of the variance ( $R^2 = .34$ ,  $F(1,14) = 7.28$ ,  $p = .017$ ). There was a significant increase in BPVS performance with age, with test age showing an increase of over 5 months for each year of CA, across the range measured (gradient =  $.55$ , [.27, .83]).

##### Comparison to TD trajectory

The onset for this trajectory was not significantly different from the TD group (intercept:  $27.39$ , [-17.38, 72.16]). While the HFA trajectory runs below the TD trajectory, the greater variability of the HFA scores meant that there was no overall group difference ( $F(1,37) = 1.07$ ,  $p = .307$ ). However, BPVS performance increased significantly more slowly than in the TD group (interaction of group and age:  $F(1,37) = 5.89$ ,  $p = .020$ ). In sum, the HFA group exhibited a *delayed rate* in vocabulary development.

#### **4.2.2.3 LFA group**

Unlike the HFA group, the performance generated by the children with LFA was close to floor level, scoring well below the TD, HFA and WS groups, as well as showing greater variability in its trajectory ( $R^2 = .003$ ;  $F(1, 15) = .05$ ,  $p = .830$ ). The performance scores of LFA group did not produce a valid developmental trajectory.

---

<sup>1</sup> 95% confidence intervals = [x, y]. Henceforth, data will be reported in the form of regression coefficient [5% confidence interval, 95% confidence interval]

The regression analysis generated a high intercept value but this was an artefact of floor scores (intercept: 59.19; [7.78, 110.59])<sup>(2)</sup>. The performance of LFA group did not increase with age but instead slightly decreased (gradient: -.05, [-.54, .44]).

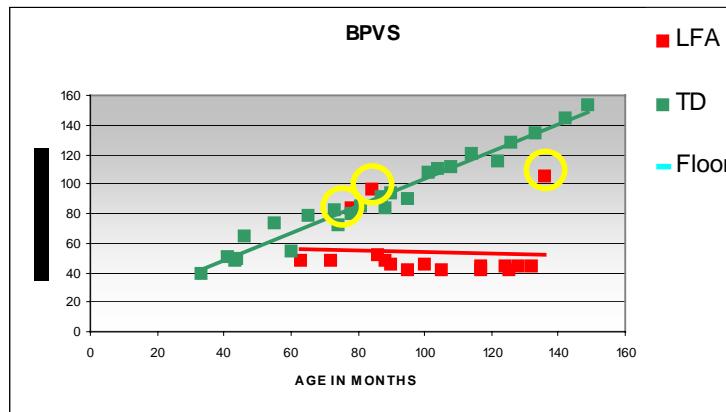


Figure 4.2: The LFA group developmental trajectory shown with outliers.

It is evident from Figure 4.2 that there were three children who performed at the same level as TD children on this test. Interestingly, these children were identified with the following: special interest in reading dictionaries, high parental education level and low score on CARS.

It was decided to reanalyse the data by removing those scores and the new trajectory showed to account for almost 40% of the variance ( $R^2 = .39$ ,  $F(1, 12) = 7.63$ ,  $p = .017$ ) However, the data showed the same pattern as in the full trajectory of *zero rate* in language development (gradient: -.09; [-.15, -.02]), where the performance was slightly decreasing with age.

#### Comparison to TD trajectory

The LFA group performed at an overall lower level compared to the TD trajectory (main effect of group:  $F(1,37) = 7.13$ ,  $p = .011$ ), also the LFA group revealed a significantly slower rate of development (interaction of group and age:  $F(1,38) = 30.46$ ,  $p < .000$ ). Figures 4.1 and 4.2 show that LFA trajectory consistently runs

<sup>2</sup>Henceforth, the intercept value that is an artefact of floor scores will not be reported.

closely to the floor line and does not improve with age, indicating a *zero rate* of performance.

#### **4.2.2.4 DS group**

The trajectory generated by DS group accounted for half of the variance ( $R^2 = .52$ ,  $F(1,13) = 14.32$ ,  $p < .001$ ). Their performance was close to the floor level and increased at a slow rate of 2 months for each CA year (gradient:  $.20 [.09, .31]$ ,  $F(3,36) = 437.99$ ,  $p < .000$ ).

##### Comparison to TD trajectory

The development of children with DS was much slower than TD children (interaction of group and age:  $F(1,37) = 94.67$ ,  $p < .001$ ). However, there was not an overall significant difference between the groups ( $F(1,36) = 2.93$ ,  $p = .095$ ). The DS trajectory runs closely to floor level, indicating receptive vocabulary development at a far slower rate than our TD control group. We define it here as *delayed rate* and possibly *delayed onset*<sup>(3)</sup> of developmental performance.

#### **4.2.2.5 WBS group**

Similarly to HFA and TD group, WBS group generated a trajectory that accounted for over half of the variance ( $R^2 = .62$ ,  $F(1,15) = 24.54$ ,  $p < .001$ ). Although the performance of the younger children was slightly delayed (intercept:  $-3.28 [-37.53, 30.98]$ ), the overall test performance rose sharply showing an increase of around 8 months for each year of CA (gradient:  $.76 [.44, 1.09]$ ).

##### Comparison to TD trajectory

Despite the fact that the WBS trajectory runs consistently below the TD trajectory, there was no reliable group difference ( $F(1,38) = 1.28$ ,  $p = .265$ ). Additionally, there was no difference in increase in performance with age (interaction of group and age:

---

<sup>3</sup> This could not be verified due to high level of intercept, an artefact of floor scores.

( $F(1,38) = 1.57$ ,  $p = .217$ ). In sum, the WBS group exhibited a normal pattern of development on the BPVS test.

#### **4.2.2.6 Comparison of different groups**

Lastly, we clarify potential differences between (i) HFA vs. WS (ii) LFA vs. DS groups.

##### Comparison of HFA and WBS trajectories

Comparison of HFA and WBS groups revealed that these two groups performed similarly on the test and there was no overall group difference (main effect:  $F(1,29) = 1.37$ ,  $p = .251$ ). Test performance increased in both groups at the same rate (interaction of group and age:  $F(1,29) = .73$ ,  $p = .404$ )<sup>(4)</sup>.

##### Comparison of LFA and DS developmental trajectories

Children with LFA performed worse than children with DS (main effect of participant group:  $F(1,27) = 8.55$ ,  $p = .001$ ). Further, their performance did not increase with age as in DS group (interaction of group and age:  $F(1,27) = 7.68$ ,  $p = .001$ ). Figure 4.1 shows that the LFA group performance was more heterogeneous compared to DS group.

### **4.3 PATTERN CONSTRUCTION (PC)**

PC is a non-verbal subtest of BAS II (Elliot, et al., 1987). This test measures visuo-spatial abilities of children aged 3:0 to 17:11, thus the minimum floor level age equivalent score is 34 months and ceiling level is 215 months. The test requires the child to make two-dimensional patterns from blocks that are two or three-dimensional (in later trials) when given a target pattern (see Figure 4.3). The test is considered to possess high overall reliability ( $r = .91$ ), with reliability coefficients ranging from .80 to .93 across all of the age groups. It has a moderate correlation with the GCA ( $r = .77$ ).

---

<sup>4</sup> Critical value of Alpha was reduced to .025 for all the comparison tests.

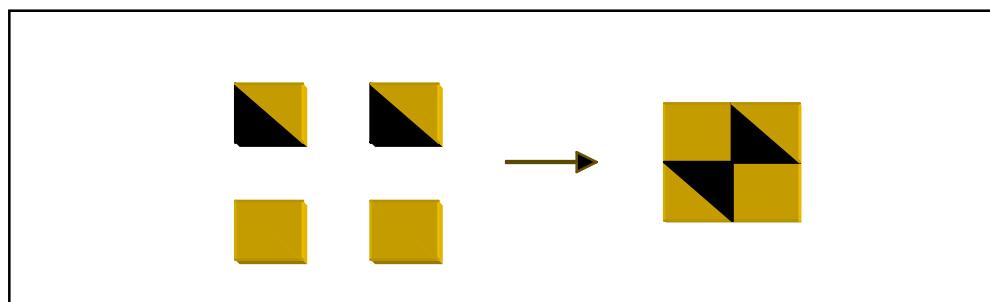


Figure 4.3: Example of PC trial.

### 4.3.1 Method

#### 4.3.1.1 Participants

All participants completed this test. See chapter 3.2.1 for full details of each group.

#### 4.3.1.2 Procedure

Firstly, each child was given few blocks to play with, in order to familiarise themselves with the texture, shape and/or smell of the items. Then the administrator demonstrated the first item in the test. The test was ended when the child accurately finished a total of 15 items or was unable to construct 4 items in 5 consecutive trials. All the trials were timed. Some children in DS and WBS groups tried to complete the patterns by building their designs directly on top of the model picture. This strategy may be helpful but only on the early items where the picture is much smaller than the blocks themselves. In the later trials the use of this strategy results in the covering up of the pictures that the participants are trying to copy. However, children in all groups were asked to follow a standard procedure.

### 4.3.2 Results

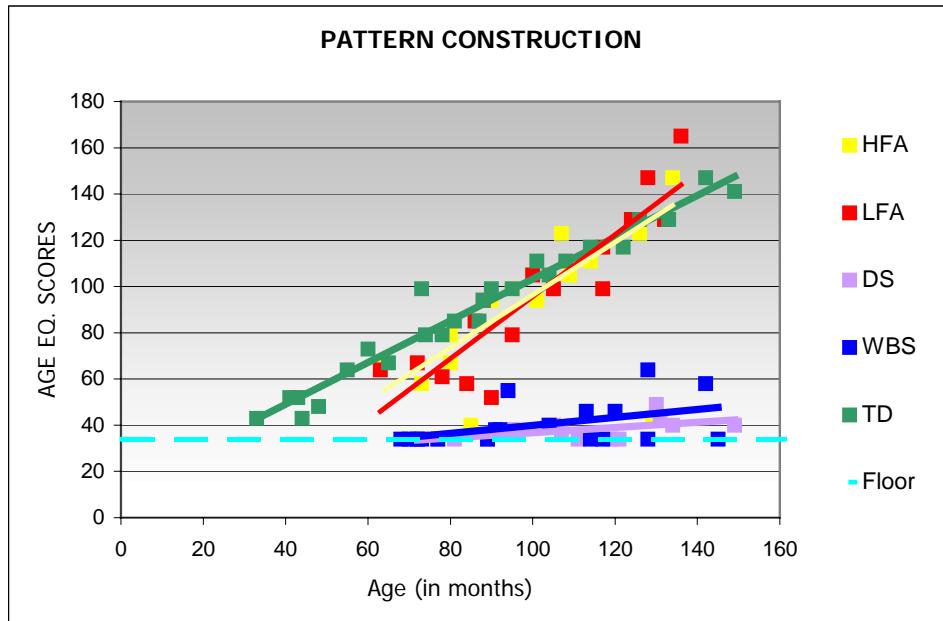


Figure 4.4: PC age equivalent scores for all groups plotted against their CA.

#### 4.3.2.1 TD control group

The trajectory generated by TD group accounted for almost all variance ( $R^2 = .96$ ;  $F(1,23) = 567.30$ ,  $p < .000$ ). The rate of performance on PC was in advance of their CA (gradient: .90, [.82, .98]). The children had a slightly earlier onset of development on the test (intercept: 13.09, [5.88, 20.30]). The control sample therefore generates a valid and slightly advanced trajectory of visuo-spatial development (Figure 4.4).

#### 4.3.3.2.2 HFA group

HFA group trajectory accounted for over the third of the variance ( $R^2 = .34$ ,  $(F(1,14) = 7.10$ ,  $p = .020$ ). Converse to the TD group, children in the HFA group had a slightly later onset of development on this test (intercept: -14.87; [-106.98, 77.24]). However, the rate of performance was increasing sharply with over one year in relation to their CA (gradient: 1.11; [.22, 2.01]).

#### Comparison to TD trajectory

There was no overall group difference ( $F(1,37) = .88$ ,  $p = 0.353$ ), and both groups performance increased with age at the similar rate (interaction of group and age:  $F(1,37) = .51$ ,  $p = .478$ ). Thus, statistically, the HFA group performance demonstrated a normal pattern of development, but with greater variability at both ends of development (Figure 4.4).

#### **4.3.3.2.3 LFA group**

The trajectory generated by the children with LFA explained over 80% of the variance ( $R^2 = .82$ ,  $F(1,15) = 115.55$ ,  $p < .000$ ). Again, as in HFA group performance rate for this trajectory was very high (gradient: 1.34; [1.00, 1.69]) suggesting a sharp increase in performance with age. However, the negative value of intercept suggests a delayed onset of the performance (intercept: -38.69; [-74.65, -2.73]).

#### Comparison to TD trajectory

Overall, performance of the groups was significantly different ( $F(1,38) = 15.11$ ,  $p < .001$ ). Also, the rate of increase with age was significantly different, this could be due to the delayed performance in the younger children with in LFA group (interaction of group and age:  $F(1,38) = 11.38$ ,  $p = .002$ ). In sum, the LFA group had a *delayed onset* on this task, but their rate of performance was significantly faster than TD group. Figure 4.4 illustrated these results.

#### **4.3.3.2.4 DS group**

Performance of the children with DS generated a trajectory that explained over a third of the variance ( $R^2 = .34$ ,  $F(1,13) = 6.8$ ,  $p = .020$ ). Their performance increased only slightly with age, with a rate equivalent to one month for each year of CA (gradient: .11; [.02, .21]).

### Comparison to TD trajectory

Although, there was no overall group difference ( $F(1,36) = 2.55$ ,  $p = .119$ ), children with DS showed to perform significantly poorer compared to TD group (interaction of group and age:  $F(1,36) = 119.82$ ,  $p < .001$ ). The DS trajectory runs closely to floor level, with a very slow increase in performance *delayed rate* of test performance and possibly *delayed onset*<sup>(5)</sup>.

### **4.3.3.2.5 WBS group**

As shown in Figure 4.4, the trajectory generated by the children with WBS explained 20% of variance ( $R^2 = .20$ ,  $F(1,16) = 4.11$ ,  $p = .060$ ). Again the rate of performance was just over one month increase for each year of CA (gradient: .17 [-.01, .36] indicating that across the age range sampled performance did not increase significantly.

### Comparison to TD trajectory

As with the DS group, there was no significant difference between the groups (main effect of group:  $F(1,38) = 1.02$ ,  $p = .276$ ), but a strong interaction of group and age (interaction of group and age:  $F(1,38) = 74.01$ ,  $p < .001$ ), indicating that performance did not increase with age in the WBS group. It is evident from Figure 4.4 that the WBS trajectory consistently runs closely to floor level suggesting a *delayed rate* of performance and *delayed onset*<sup>(6)</sup>.

### **4.3.3.2.6 Intra- and inter-group comparison**

#### Comparison of HFA and LFA trajectories

A comparison of HFA and LFA groups revealed that these two groups performed similarly on the test ( $F(1,29) = .29$ ,  $p = .599$ ), and both groups displayed similar rate of increase in their performance on PC task (interaction of group and age:  $F(1,29) = .29$ ,  $p = .596$ ).

---

<sup>5</sup> This could not be verified due to high level of intercept which was an artefact of floor scores

<sup>6</sup> Critical value of Alpha was reduced to .025 for all the comparison tests.

### Comparison of DS and WBS trajectories

Both groups had a very slow rate of increase in performance with age (interaction of group x age:  $F(1,29) = .31$ ,  $p = .583$ ), also there was no overall group difference (main effect of group:  $F(1,29) = .06$ ,  $p = .811$ ).

## **4.4 COPYING TASK (CT)**

The coping task is a non-verbal subtest of the BAS. The test measures individual's visuo-spatial analysis for children aged 3:6 to 7:11. Thus, the floor level is 40 months and the ceiling level is 95 months. Copying is considered to possess medium overall reliability ( $r = .86$ ), with reliability coefficients ranging from .82 to .88 across all of the nine age groups. It has a medium correlation with the GCA ( $r = .65$ ).

### **4.4.1 Method**

#### ***4.4.1.1 Participants***

See chapter 3 section 3.2.1 for details. One child in WBS group completed the test 5 months later than the other children in the WBS group.

#### ***4.4.1.2 Procedure***

Before attempting the task, children were allowed sometime for free drawings, use of pencil grippers was allowed. Items in the proper task start very simple (straight line) and progress to more complex geometric figures. The participants were able to view the design the entire time while drawing, and no time limit was assigned for completion of the task. See BAS manual for detailed description of procedure (Elliott et al., 1997).

#### 4.4.2 Results

As shown in Figure 4.5, many children in TD and HFA group performed at ceiling level (red dotted line in the graph), so each data analysis was carried out with and without scores at the ceiling scores. Where the ceiling scores altered the quantitative pattern of effects in the data, this will be reported.

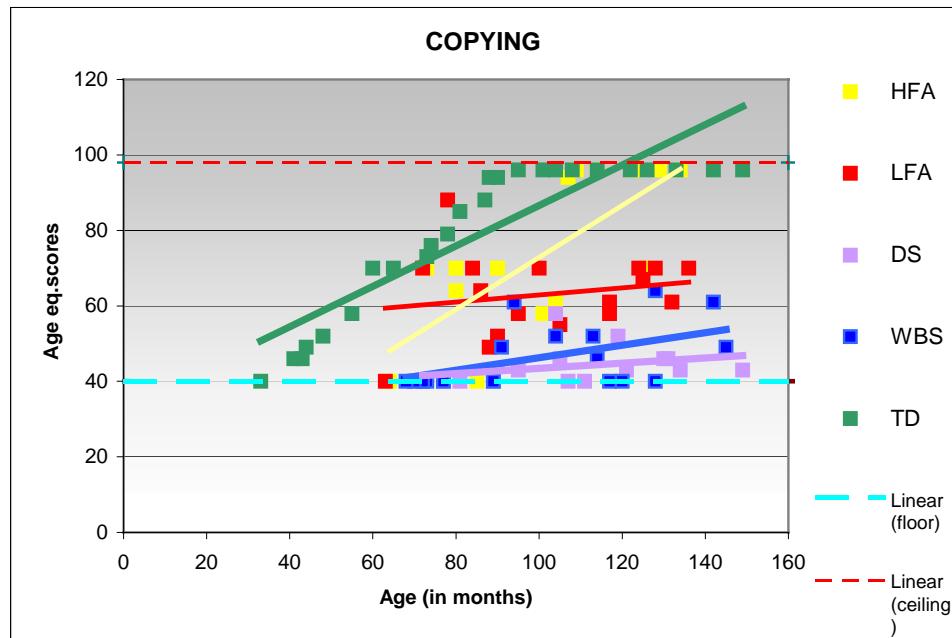


Figure 4.5: Copying test age equivalent performance for each group.

##### 4.4.3.1 TD control group

As illustrated in Figure 4.5, the trajectory generated by the TD children produced ceiling scores at around 8 years old. Thus, it was decided to treat the analysis in two ways. First, the whole trajectory was analysed, including the ceiling scores. Secondly, an additional trajectory was analysed for those children below 8 years of age. In this case the full trajectory accounted for over 80% of the variance ( $R^2 = .81$ ,  $F(1,23) = 96.28$ ,  $p < .001$ ), and revealed a significant increase in copying performance with age (gradient = .53, [.42, .65]). The trajectory below 8 years accounted for a much higher amount of the variance ( $R^2 = .98$ ;  $F(1,13) = 255.09$ ,  $p <$

.001) and the rate of performance with age showed an increase of over 9 months for each year of CA (gradient: .95, [87, 1.02]). The onset of the performance was in the normal range (intercept: 7.62; [2.67, 12.57]). The TD trajectory produced a marginally advanced developmental trajectory of CT.

#### **4.4.3.2 HFA group**

There was some indication of a ceiling effect in the HFA group, with 5 individuals above the age of 9 years (110 months old) exhibiting ceiling scores. However, one individual aged 10:8 was not at ceiling. It was therefore decided to treat these data in the same way as described in TD group. The full trajectory accounted for around half the variance ( $R^2 = .53$ ,  $F(1,14) = 15.63$ ,  $p < .001$ ), and revealed a significant increase in copying performance with age (gradient = .68 [.31, 1.06]). The trajectory below 10 years did not account for a significant amount of the variance and so did not produce a valid trajectory ( $R^2 = .19$ ,  $F(1,9) = 2.16$ ,  $p = .180$ ). This latter analysis is compromised by small number of participants, but it is worth mentioning that this truncated trajectory produced a slower increase of performance with age (gradient: .37 [-.20, -95]). Therefore, there is some indication that the relationship between age and performance was not linear in the HFA group, with a slow increase until around 8:5 years, then a sudden jump in performance. Note, that this pattern was not observed in the BPVS and PC tasks with the same children.

#### Comparison to TD trajectory

Comparison of the full HFA trajectory to the TD suggested an overall group difference in the development of copying in the HFA group, but this only emerged as a trend (main effect of group:  $F(1,37) = 3.46$ ,  $p = .071$ ). On the other hand, there was no reliable difference in the rate of increase of performance with age in the two groups (interaction of group x age:  $F(1,37) = .96$ ,  $p = .334$ ). In contrast, comparison of the truncated trajectory revealed a significantly slower increase in performance with age in the HFA group, but no overall group difference (main effect of group:  $F(1,22) = 1.48$ ,  $p = .240$ ); interaction of group x age:  $F(1,22) = 7.37$ ,  $p = .013$ ). To some extent, interpretation of the HFA group depends on how ceiling scores are

treated. There seem to be two distinct patterns of performance i) a scattered and delayed performance (up to 100 months) and, ii) a sudden improvement in performance running closely to the same age TD controls.

#### **4.4.3.3 LFA group**

The LFA group showed a large variability in their trajectory in comparison to other groups. The trajectory generated by the LFA group did not account for a significant amount of variance thus did not produce a valid developmental trajectory ( $R^2 = 0.04$ ,  $F(1,15) = .60$ ,  $p = .445$ ). The trajectory that was generated did not indicate a significant improvement of performance with age (gradient: .09; [-.17, .36]).

##### Comparison to TD trajectory

Comparison of both trajectories revealed that there was an overall group difference (main effect<sup>7</sup>:  $F(1,28) = 14.05$ ,  $p = .001$ ), and children in LFA group produced a significantly slower increase with age (interaction of group and age:  $F(1,38) = 12.62$ ,  $p < .001$ ). Although, the interpretation of the scores is difficult without having a valid developmental trajectory, it should be noted that most children scored higher than DS and WS groups. Only one child scored at floor level and the rest of the group scored in the range of 50-88 points. Thus, one could speculate that their performance rate was at *zero rate*.

#### **4.4.3.4 DS group**

Similar to LFA group, the trajectory generated by the DS group did not account for a significant amount of variance and thus did not produce a valid developmental trajectory ( $R^2 = .09$ ,  $F(1,13) = 1.31$ ,  $p = .276$ ). This group had the lowest rate of performance out of all clinical groups (gradient: .07; [-.06, .20]).

---

<sup>7</sup> The ceiling scores in TD group were excluded from the analysis

#### Comparison to TD trajectory

The DS group demonstrated a significant difference in the rate of performance with age in comparison to TD group (interaction of group and age:  $F(1,38) = 20.57$ ,  $p < .001$ ). Also, there was an overall group difference when the ceiling scores were removed in TD group (main effect:  $F(1, 26) = 20.49$ ,  $p < .001$ ). In sum, the performance of DS group is mostly at the floor level across range sampled suggesting a *zero rate* of performance on the test.

#### **4.4.3.5 WBS group**

The trajectory generated by the children with WBS accounted for almost 25% of variability in the group ( $R^2 = .24$ ;  $F(1,15) = 4.83$ ,  $p = .044$ ). There was a very little increase in performance rate with age (gradient:  $.17$ ; [.005, .33]).

#### Comparison to TD trajectory

Children with WBS had a slower rate of performance than TD group (interaction of group and age:  $F(1,38) = 14.39$ ,  $p = .001$ ). There was an overall group difference (main effect<sup>(8)</sup>:  $F(1, 28) = 7.36$ ,  $p = .011$ ). Performance of children with WBS showed a *delayed rate* and possibly a *delayed onset* as well.

#### **4.4.3.6 Comparison of different groups**

##### Comparison of DS and WBS trajectories

Lastly, we compared the performance of children from the WBS and DS groups to clarify potential differences. Analyses of the data revealed that these two groups perform similarly on the test and there was no overall group difference (main effect:  $F(1,28) = 17.08$ ,  $p = .389$ ), nor an interaction between group and age ( $F(1,28) = 40.22$ ,  $p = .538$ ). Many children in both groups scored at floor level. Examples of copying can be found in Table 4.1.

---

<sup>8</sup> The ceiling scores in TD group were excluded from the analysis.

Table 4.1: Examples of copying for each disorder group.

AGE	HFA	LFA	DS	WBS
6				
8				
10				
12				

## 4.5 BENTON FACIAL RECOGNITION TASK - SHORT VERSION

The Benton test is a commercially available test to assess individuals' face recognition abilities. The short- form of the Benton test has 13 items with 27 possible points while the long form has 22 items with 54 possible points.

### 4.5.1 Method

#### 4.5.1.1 Participants

Participant details are outlined in Chapter 3.

#### 4.5.1.2 Procedure

The short version of the Benton test was administered in our study. On each item, subjects were presented with a target photo and were asked to choose the target individual from six faces presented simultaneously with the target photo. The short version of the test is graded and consists of two parts: (1) matching a frontal view of the target with an identical photograph, (2) matching a frontal view of the target individual with three photos of the target taken from different angles. No time limits were placed, and scores were classified as normal when 21 or above was achieved. The photos used in the test consist of unfamiliar male and female faces with their hair and clothing shaded out so that subjects must rely on the face. The faces are centered within a black background, and the entire image is approximately 6.5 cm by 6.5 cm (see Figure 4.6).

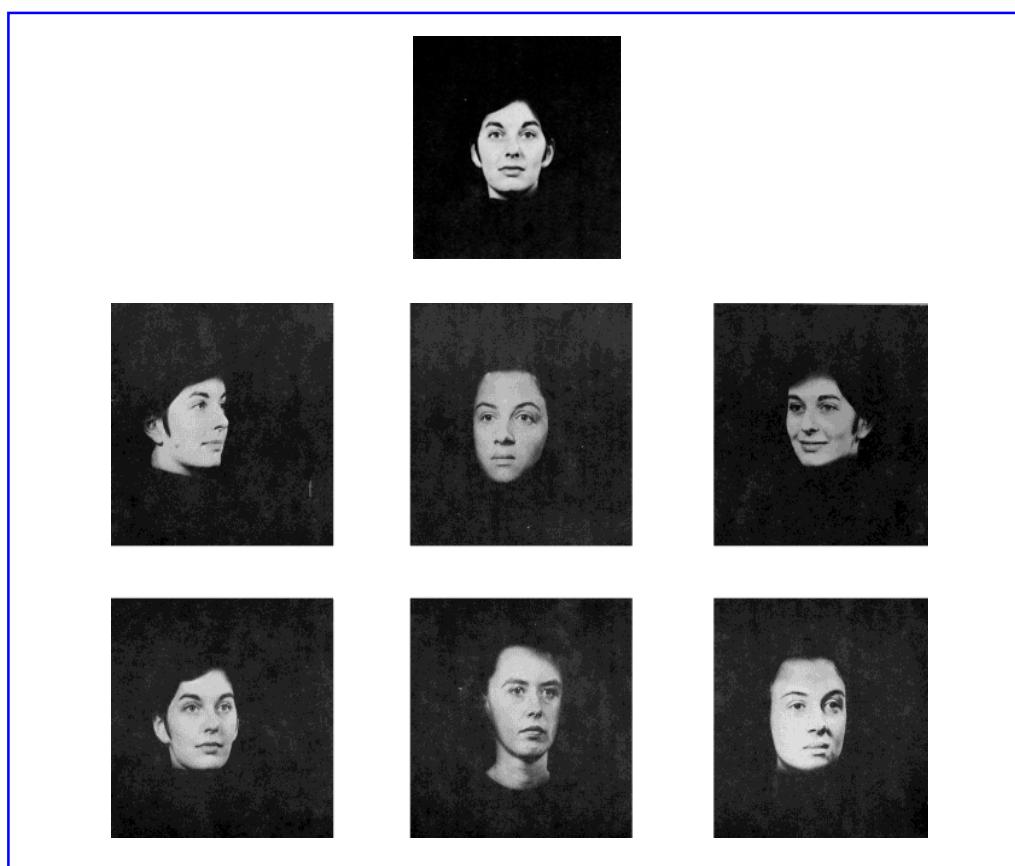


Figure 4.6: Example of Benton test trial. Participants are asked to find three photos of the target face shown at the top.

### 4.5.2 Results

At the time of writing, no standardisation of Benton short form had taken place. Therefore, raw scores rather than test ages were compared in the following analysis. These still permit comparisons to TD trajectory. Figure 4.7 shows raw scores on short version performance. Participants scoring below 11 points were at chance level, whereas scores of 20 points and above were at normal adult-like face recognition. The intercept value will not be reported in this section since raw scores values are used in the analysis.

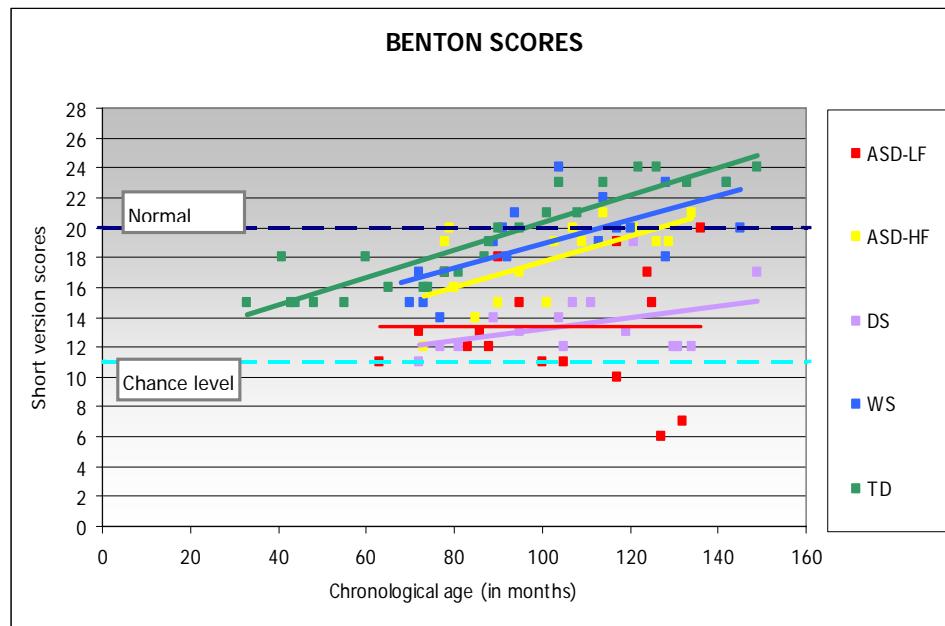


Figure 4.7: Developmental trajectories for Benton tests performance.

#### 4.5.2.1 TD control group

All children above 8 years of age (95 months old) performed within normal adult-like face recognition (20 points and above). None of the children scored lower than 15 points. The trajectory generated by the TD children accounted for over 80% of variance ( $R^2 = .84$ ;  $F(1,23) = 122.65$ ,  $p < .001$ ), and the performance raised significantly with age (gradient:  $.09$ , [.08, .11]). Scores from this group will be used to derive test ages for the disorder groups. This will be derived from formula ( $y = .92x + 11.11$ ) from their linear regression.

#### **4.5.2.2 HFA group**

Performance of children with HFA generated a trajectory that accounted for almost half of the variance ( $R^2 = .49$ ,  $F(1,14) = 13.41$ ,  $p = .003$ ). Of the disorder groups, the HFA group produced the steepest rate of increase with age (gradient:  $.09$ , [.04, .14]).

##### Comparison to TD trajectory

The TD and HFA groups performed similarly (main effect of group:  $F(1,37) = 1.01$ ,  $p = .321$ , interaction of group and age:  $F(1,37) = .013$ ,  $p = .909$ ). A number of individuals with HFA group appeared to fall below the TD trajectory (Figure 4.7); however the HFA group demonstrated greater variability, thus preventing reliable differences.

#### **4.5.2.3 LFA group**

The LFA group's performance did not generate a reliable developmental trajectory ( $R^2 = .001$ ,  $F(1,15) = .001$ ,  $p = .981$ ). Six children scored in the range of chance level and only one child scored in normal range. For the trajectory generated, the gradient was not significantly different from zero, indicating no improvement of performance with age (gradient:  $-.001$ , [-.02, .98]).

##### Comparison to TD trajectory

The main effect of participant group was not significant ( $F(1,38) = .42$ ,  $p = .522$ ), however, children in LFA group were significantly delayed in their development compared to TD (interaction of group and age:  $F(1,38) = 6.89$ ,  $p = .012$ ). In sum, the children in LFA group performed at “zero rate”.

#### **4.5.2.4 DS group**

Children in this group did not generate a valid developmental trajectory ( $R^2 = .16$ ,  $F(1,13) = 2.54$ ,  $p = .163$ ). Further, their performance was not increasing with age (gradient:  $.04$ , [-.01, .09]), suggesting a zero *rate* of performance.

### Comparison to TD trajectory

There was no overall group difference between TD and DS groups (main effect:  $F(1,36) = .62$ ,  $p = .438$ ) , however, there was a significant difference in their performance rate (interaction of group and age:  $F(1,36) = 5.73$ ,  $p = .022$ ).

#### **4.5.2.5 WBS group**

The trajectory generated by the children with WBS explained almost 40% of variance ( $R^2 = .39$ ,  $F(1,16) = 9.67$ ,  $p = .007$ ). This group's performance increased significantly with age (gradient:  $.07$ , [.02, .12]).

### Comparison to TD trajectory

There was no overall group difference between TD and WBS children (main effect:  $F(1,39) = .28$ ,  $p = .600$ ), nor a reliable difference in the rate of increase of performance with age (interaction of group and age:  $F(1,39) = 1.17$ ,  $p = .287$ ). Figure 4.5 suggests that WS trajectory closely follows the TD trajectory although their performance was more heterogeneous. In sum, for face recognition, the WBS group followed a normal trajectory of development.

#### **4.5.2.6 Comparison of different groups**

##### Comparison of HFA and WBS trajectories

Comparison of HFA and WBS groups revealed that these two groups performed similarly on the test and there was no overall group difference (main effect:  $F(1,30) = .92$ ,  $p = .344$ ), nor a difference in rate of performance with age (interaction of group and age  $F(1,30) = .35$ ,  $p = .561$ ).

##### Comparison of LFA and DS trajectories

Comparison of LFA and DS groups showed that both groups performed similarly (no main effect of participant group:  $F(1,27) = .52$ ,  $p = .479$ ), and the rate of performance with age was indistinguishable (no interaction of group and age:  $F(1,28)$

= .55, p = .462). It should be noted that although there are no statistical difference in these groups, due to variability. Only one child in DS group scored at chance level in comparison to six children in LFA.

#### 4.6 SUMMARY OF ALL TESTS

A large number of comparisons was carried out, hence we summarise them in two ways. Figures 4.8 shows mean age equivalent scores. Note that age range is not strictly comparable, since TD group was younger than all clinical groups. Second, Table 4.2 summarises trajectories in terms of types of development we identified in chapter 3.

Table 4.2: Summary of results for each group.

TEST	TD	HFA	LFA	WBS	DS
<b>BPVS</b>	NORMAL	DELAYED RATE	ZERO RATE	NORMAL	DELAYED ONSET & RATE
<b>PC</b>	NORMAL	NORMAL	DELAYED ONSET	DELAYED ONSET & RATE	DELAYED ONSET & RATE
<b>COPYING</b>	NORMAL	UP TO 8 YRS DELAYED RATE; THEN NORMAL	ZERO RATE	DELAYED ONSET & RATE	ZERO RATE
<b>BENTON</b>	NORMAL	NORMAL	ZERO RATE	NORMAL	ZERO RATE

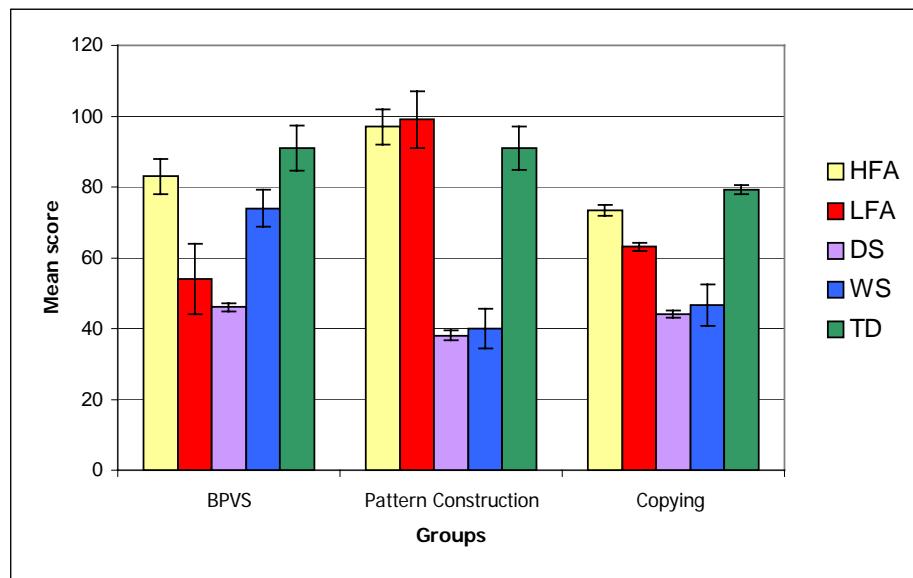


Figure 4.8: Summary of standardised tests. Note that ceiling scores for 10 individuals in the TD group are included; the error bars display the standard error of the mean. Also, the age range differs for each group.

### BPVS

Children with HFA scored showed a delayed rate of language acquisition in comparison to the TD group, whereas children in the LFA group did not improve with age and instead they got worse with age on the task thus showing an atypical pattern of development. Results obtained from autism groups were surprising as children with autism were not divided on language abilities, but the severity of behavioural symptoms (CARS). In line with study carried out by Lawson and Gunn (2004), children with DS showed a delayed onset and continued to improve at a slow rate on the test. Language is one of the domains that have been described as normal and indeed in the current study, WBS group displayed a normal receptive vocabulary performance. Although, the results show that WBS group did not significantly differ from control group, their developmental trajectory lags behind TD group at all times (see Figure 4.2). Moreover, it should be noted that BPVS represents only receptive vocabulary abilities of language thus any inference regarding language abilities cannot be based on group outcome on this test.

### **Pattern Construction**

Children in both autism groups performed as well as TD group on the pattern construction. The current data offers no support to previous reports that individuals with autism demonstrate superior performance on tasks such as pattern construction (Shah & Frith, 1993). Both autism groups performed comparably to the TD group. As predicted, children in the DS and WBS groups displayed poor performance on the test, a similar profile shown by many previous studies (e.g., Bellugi et al., 1988, 1994). Anecdotal reports in the current study on the task completion by some children with DS (around 70%) and WBS (around 60%) that the children could score higher on the task if other method was employed. Instead of putting cubes on the desk children often used a target example to put the cubes on top of the pictures. We could speculate that using this method individuals' performance would have improved (see Farran, 2003, for discussion on task difficulty).

### **Benton test**

Children with HFA and WBS scored in the normal range on Benton test thus demonstrating normal face perception. However, these results should be taken with caution because alternative ways to normal scores on Benton faces have been reported. Duchaine and Weidenfeld (2003) showed that Benton task can be successfully completed by using feature-based recognition skills. The authors modified the test by deleting the internal features except for the eyebrows and hair. The results showed that the presence of only few features were sufficient to recognise faces. This finding is further supported by another study (Duchaine & Nakayama, 2004) where individuals with prosopagnosia were able to score within the normal scale on the test. Surprisingly, children in the LFA group showed a very poor performance on the test and contrasting developmental profile to the HFA group. Why did the LFA group not use a featural strategy on the Benton test? It seems that social impairment and lack of expertise on face recognition overrode use of featural encoding. Children in the DS group produced similar scores on the test.

### **Copying task**

This task produced ceiling scores very early on in the TD group, thus differences between the disorder groups such as HFA can potentially be masked. Current findings suggest that children in the HFA and LFA groups display different developmental profiles on the copying task. Children in the HFA group showed a delayed performance up to the age of 8 and then their scores were in normal range of performance, whereas, children in the LFA group did not improve with age and exhibited atypical developmental profile. A possible explanation of HFA group performance is that the children are mainstreamed at slightly later age than TD individuals or other abilities had compensatory role in the performance. It is well documented that children with WBS and DS are poor on visuo-spatial construction tasks and children participating in the current study showed similar performance.

### **Overall Summary**

In summary the results from the battery of standardised tests confirmed that each clinical group displayed characteristic cognitive profile of their disorder. For instance children in the autism groups showed good performance on the pattern construction, the WBS group showed commonly discussed uneven cognitive profile (good language scores and poor PC scores) and DS group showed progressively slower rate of development with age.

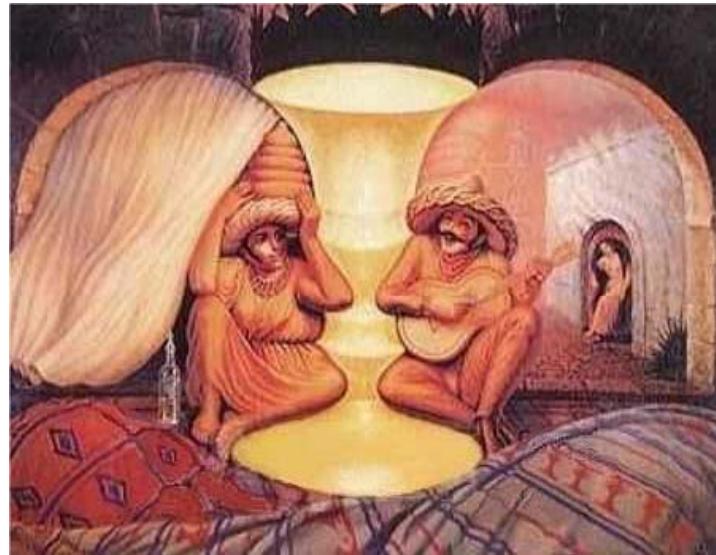
In order to investigate possible relationships between face processing accuracy and other cognitive abilities or age we computed analysis between standardised and experimental measures. These included the use of scores on the Benton test (chapters 5, 6 and 7) and pattern construction (chapter 7). These analyses were also used to directly compare current results with previous studies (Karmiloff-Smith et al., 2004; Tager-Flusberg et al., 2003). Current experimental studies investigate visuo-spatial skills, and hence standardised measures such as BPVS will not be used.

The developmental changes in face processing abilities between the TD groups will be compared with the developmental trajectory of clinical groups. In such analyses, main effects provide an indication of differences between control and clinical groups

in the absolute magnitude of developmental changes in the dependent variables. An interaction between group and development would suggest differences in the quality of the developmental differences between TD and clinical groups. In other words, an interaction of group and development indicates that the slope of developmental change differs between groups. In addition, each empirical chapter will conclude with characteristics according to the definition of delay described in chapter 3 (see p.98 for details).

## **CHAPTER 5**

### **DEVELOPMENT OF HOLISTIC FACE RECOGNITION**



*Is this a portrait of an old couple, or is it that of two young musicians sitting on the ground?*

## **5.1 INTRODUCTION**

The roles of parts and wholes in perception have been studied for a long time, starting with a debate between structuralists who championed the role of parts and Gestalt psychologists who emphasised the role of wholes. Some early behavioural studies argued that adults generally perceive faces organised as wholes, whereas young children can also identify faces as organised as wholes but they might be less skilful when doing so (Carey & Diamond, 1994; Pellicano & Rhodes, 2003). Moreover, it was proposed that a strategy that is most often used by young children is the recognition of faces based on a single feature such as mouth. However, as described in earlier chapters (chapter 1 and 2), this featural encoding does not convey sufficient information for accurate and rapid face discrimination, thus more advanced encoding of face recognition has been proposed within the framework of holistic and configural information.

Recently, the holistic approach has enjoyed its renaissance with a large number of investigations. However, little research has equated the role of developmental changes with holistic face recognition in children with developmental disorders. As stated in chapter 2, the aim of this thesis is to gain an insight into the development of face recognition competences by examining holistic and configural aspects of face recognition. The current study sought to address when typically developing children develop sensitivity to holistic face encoding and how children in the clinical groups compare to them.

The experimental method employed in the current study was based on the whole-part paradigm by Tanaka and Farah (1993), recently improved upon by Joseph and Tanaka (2002) and extended by Lewis and Glenister (2003). Briefly, the whole-part paradigm (described in detail in Chapter 1) was developed to differentiate between featural and holistic face processing (e.g., Tanaka & Farah, 1993; Tanaka & Senco, 1997; Lewis & Glenister, 2003). In the original whole-part paradigm, children were presented with a target face that was identified with a name (for example: ‘This is Tom’) for 5 seconds. Then they were presented with a target face and a distracter-face, which differed by one feature, and were asked to show ‘Which is Tom?’

In the part-face condition, children were presented with a target feature and a distracter feature and were asked to show, for example, ‘Which is Tom’s nose?’ Using this approach, it has been shown that children from around 6 years of age and adults recognise facial features better when they are embedded in the whole face rather than when presented in isolation when viewed upright. Furthermore, any holistic processing advantage would not be operative when a stimulus is presented in an inverted orientation. It has been shown that inversion disrupts both: holistic and configural processing of a whole face. Hence whole-part face paradigm and face inversion have been associated with maturity of holistic face recognition, they are both used in the current study.

The eye-feature has been shown to elucidate the most accurate performance in the TD individuals (Tanaka & Farah, 1993) and individuals with Williams syndrome (Tager-Flusberg et al., 2003). In contrast, children with autism were most accurate when face recognition depended on the mouth (Joseph & Tanaka, 2003). Details of the whole-part procedure used to test holistic processing in autism and Williams syndrome groups are outlined in chapter 2. It was suggested that individuals in autism and WBS groups showed normal holistic face processing. In light of the evidence provided by those authors, and the lack of developmental approach to holistic face abilities in the clinical groups, the general aim of this study was to investigate how developmental changes of holistic processing occur in typically developing children and disorder groups.

## **5.2 STUDY 1: WHOLE-PART FACE RECOGNITION**

In the whole-part version of the task used here, participants were presented with a target face/feature simultaneously in different orientations. The specific investigations and predictions are described below:

1) Do children show an increasing advantage of feature recognition when presented in the context of a whole-face rather than in isolation (referred to as ‘part-face’)?

**Predictions:**

- i) Children in TD (Tanaka & Farrah, 1993) and DS groups will demonstrate an increasing advantage of recognising features presented in the context of faces rather than in isolation for the upright faces;
- ii) Children in the autism groups (e.g., Joseph & Tanaka, 2003) and WBS group will perform better on the part-face condition (e.g., Elgar & Campbell 2001; Karmiloff-Smith et al., 2004);
- iii) All the disorder groups will perform significantly poorer on the whole-face trials on the upright condition compared to the TD group.

2) What is the onset/preference of the holistic face recognition?

**Predictions:**

- i) Children in the TD group as young as 6 years old will show holistic face recognition marked by increased performance on whole-face in upright orientation and decreased performance on inverted trials;
- ii) Children in the clinical groups will be significantly less sensitive to holistic processing. However, children in the DS group will show similar performance to the TD group.

3) Do inversion effects emerge on whole- and/or part-face conditions?

**Predictions:**

- i) Children in the TD and DS groups will become increasingly more influenced by rotation of the stimuli and their accuracy will decline while their response time will increase with age. Based on the previous findings, it is expected that this effect will start to emerge by 6 years of age;
- ii) Children in the disorder groups, except for DS group, will not be affected by the rotation of the face stimuli.

4) What is the most salient feature during the face recognition?

**Prediction:** Children in TD, DS and WBS groups will exhibit the normative pattern of better performance on the eye-feature than nose or mouth, while children with autism, will focus predominantly on the mouth feature.

5) Can performance on the Benton Face recognition test predict level of performance in the whole-part task?

**Prediction:** Previous findings suggested that Benton test could be resolved by the use of featural information. Thus, it was predicted that the Benton test will not be a good predictor of holistic recognition.

### **5.2.1 Method**

#### ***5.2.1.1 Participants***

Three children in the WBS group did not complete this task (age range: 5:8 – 12:8). Details of other groups can be found in chapter 3, section 3.2.1.

#### ***5.2.1.2 Stimuli***

Three high quality faces were generated using Faces 3.0 software (published by IQ Biometrix, Inc.). For each prototype face, two types of eyes, nose and mouth were generated. The same face feature was never used across other prototypes. Use of this high performance face-reconstruction software meant that methods such as cropping and positioning of the face features were eliminated as potential confounds. Use of a narrow choice of features of similar shapes and matched for eye-colour also reduced the possibility of confounds. Figure 5.1 shows a sample of whole- and part-face stimuli.

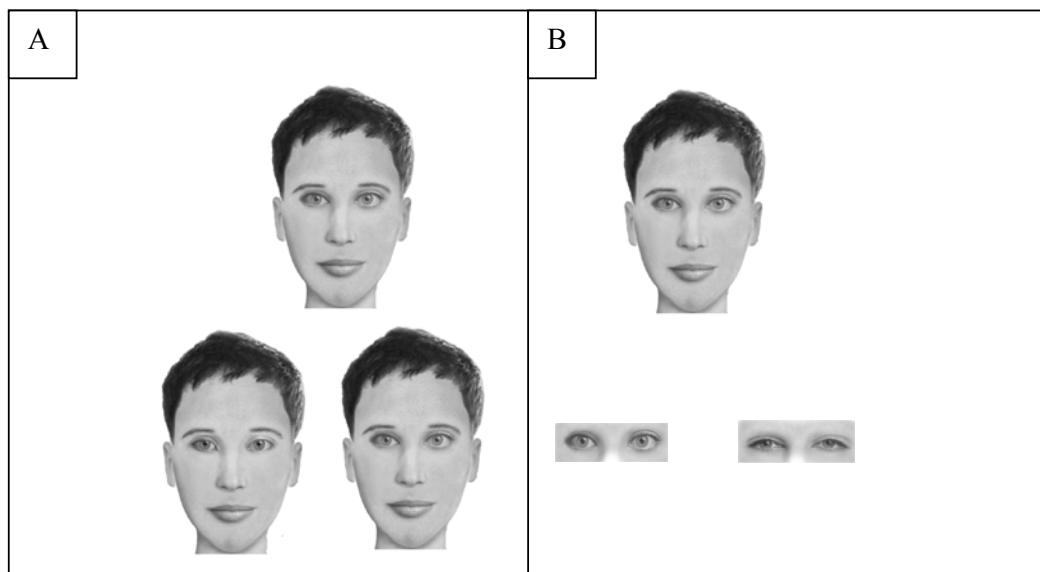


Figure 5.1: Example of the whole-part stimuli. A) whole-face condition; B) part-face condition (stimuli not used in the study due to large differences in shape of the eyes). The upper face is the target. The participants had to decide which of the lower alternatives matched the target face.

### 5.2.1.3 Procedure

All participants were tested on 36 upright trials followed by 36 trials in 90° (clockwise) orientation and 36 inverted trials. The trial order was randomised within orientation in blocks. Stimuli were presented simultaneously on a 17-inch touch-screen computer monitor using SuperLab Pro 2.0 software. Children were seated facing the computer monitor at a viewing distance of approximately 30 cm and with their eye level at the centre of the screen. All children first took part in a practice task consisting of 6 trials (3 whole-faces and 3 part-faces) where feedback was given, to familiarise them with the testing procedure and the touch-screen equipment. Participants were informed that identification of the face by a single feature was necessary in some of the trials. The experimenter initiated the task by saying: “*Now we are going to play a game. Look at this face (experimenter points to the target faces) can you touch the face that you think looks the same? Sometimes you will see the face and features such as eyes, nose or mouth. Can you show me which feature is the same as in the face. Are you ready?... Try to answer as fast as possible*”.

Following the practice trials, participants were presented with an original face, a target face and a distracter face in the whole-face condition. The distracter face was a face that differed by only one feature from the target face. The distracter face was a face that differed by only one feature from the target faces (Figure 5.1A). Alternatively, in the part-face condition participants were presented with the original face, a target feature and distracter feature (Figure 5.1B). They were asked to touch the face or part-feature that was the ‘same’ as the original face. The side of correct face/feature was counterbalanced. After each response, a fixation cross appeared which gave the experimenter an opportunity to check whether the child was getting tired or distracted by external noise. In order to initiate the next trial, the experimenter or the child pressed a space-bar. The only feedback given during the experiment was non-specific praise. To encourage children to complete the task, stickers were provided at various points. Between each condition all children were given longer breaks, no shorter than 30 minutes and no longer than 2 hours.

The dependent variables were the percentage of correct choices for the upright, 90° and inverted trials in the whole and part conditions. Accuracy for individual features was also analysed. Response times were also measured and analysed, however due to level of noise in these data, the RT will be considered only briefly.

### **5.2.2 Results**

Accuracy and RT data were taken from all participants in the studies.

1. Trajectories were analysed using fully factorial analysis of co-variance (ANCOVA) for each group, where the within-participants factors were orientation (upright, 90° and inverted), presentation context of each face-feature (whole-face and part-face) and identification of the face by individual feature (eyes, nose or mouth), and age was the co-variant.
2. A direct comparison of each clinical group to TD group was carried out using an ANCOVA (3x3x2) with Group (TD group compared to 4 clinical groups) as between-participants factor with age as co-variant, the within-group factors of face orientation and stimuli presentation (whole- or part-) with age as co-variant.
3. As in the previous results sections (chapter 4), the TD group will be first described in detail. Full analyses were performed for each group. However due to a large number of effects and interactions only ones that are specific to current predictions will be discussed in detail. Each disorder group will be characterised on its own and then against to TD group. Each disorder group will be described and then compared to the TD group. Performance scores will be plotted against Benton test (see chapter 4 for details) to explore whether this test is a good predictor of expert face recognition.

#### **5.2.1.1 Accuracy**

Table 5.1 displays the mean number and percentage of trials correct for each group for each condition and collapsed across all conditions for individual groups. Whereas, Figures 5.2-5.6 depict the performance of each group in terms of correct percentage accuracy scores plotted against increasing chronological age (CA) in months. The solid lines indicate a best-fit regression through each group's data <sup>(1)</sup>.

---

<sup>1</sup> Note that individual points are not displayed in the graphs due to a large number of conditions and individuals; instead  $R^2$  is shown for each regression line, reflecting the percentage of the variance captured by each best-fit trajectory.

One again, regressions were checked for violation of linearity and outliers. Due to larger variability in the disorder groups, comparison statistics will include a measure of effect size (partial eta-squared).

Table 5.1: Percentage correct accuracy on individual features for each condition.  
 \* Mean number of correct responses for each group in each condition

GROUP	WHOLE					PART								
	Eyes%	M(SD)*	Nose%	M(SD)*	Mouth%	All%	Eyes%	M(SD)*	Nose%	M(SD)*	Mouth%	M(SD)*	Mouth%	All%
<b>UPRIGHT</b>														
TD	90	5.4 (0.8)	55	3.3 (1.2)	75	4.5 (1.4)	73	93	5.6 (0.6)	77	4.6 (1.0)	85	5.1 (0.7)	85
HFA	75	4.5 (1.0)	54	3.3 (1.4)	70	4.2 (1.2)	67	84	5.1 (1.3)	61	3.7 (1.3)	88	5.3 (0.9)	80
LFA	32	1.9 (0.9)	35	2.1 (1.1)	53	3.2 (1.0)	40	44	2.8 (0.9)	57	3.4 (0.7)	77	4.6 (1.1)	60
DS	92	5.5 (1.1)	52	3.1 (1.1)	68	4.1 (0.7)	71	62	3.7 (0.9)	38	2.3 (0.8)	47	2.8 (1.1)	49
WBS	81	4.9 (1.5)	66	3.9 (1.0)	51	3.1 (1.0)	61	71	4.3 (1.1)	78	4.7 (1.3)	69	4.1 (1.1)	81
														90
<b>INVERTED</b>														
TD	75	4.5 (1.0)	59	3.6 (1.2)	73	4.4 (1.3)	69	85	5.1 (1.0)	73	4.4 (1.1)	88	5.3 (0.9)	83
HFA	77	4.6 (1.0)	55	3.3 (1.1)	68	4.4 (1.1)	66	83	5.0 (1.4)	67	4.0 (1.0)	85	5.1 (0.1)	78
LFA	30	1.8 (0.7)	36	2.2 (0.8)	58	3.5 (0.7)	42	51	3.1 (1.7)	58	3.5 (0.6)	76	4.6 (0.9)	62
DS	63	3.8 (0.7)	53	3.2 (1.0)	62	3.7 (0.8)	60	67	4.0 (0.8)	41	2.5 (1.1)	47	2.8 (1.1)	51
WBS	77	4.6 (1.1)	60	3.6 (0.9)	60	3.6 (1.3)	63	73	4.4 (1.0)	70	4.2 (1.0)	76	4.5 (1.0)	73

### 5.2.1.1.1 TD control group

Trajectories for TD group split by whole-part and orientation conditions are shown in Figure 5.2.

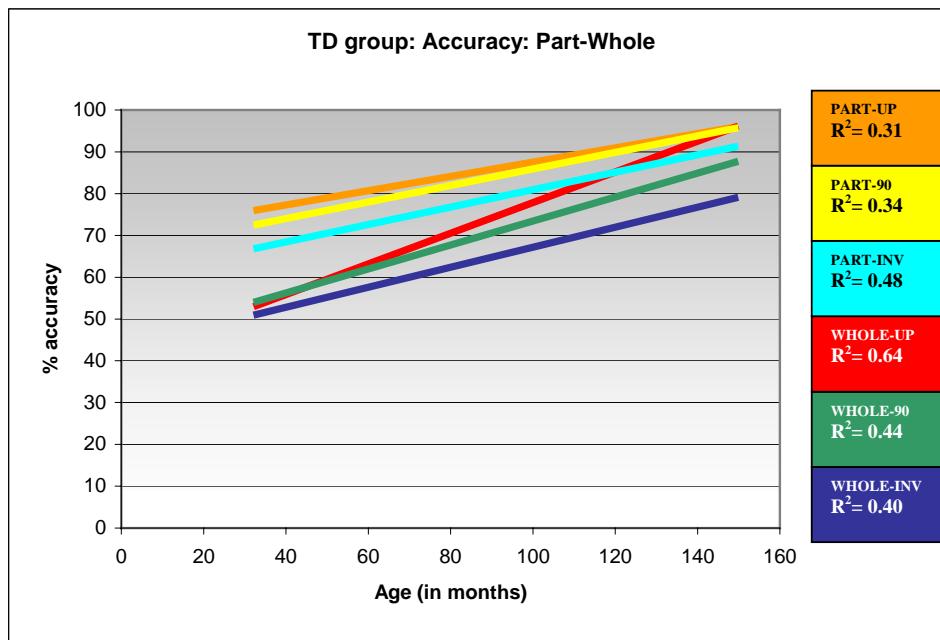


Figure 5.2: TD group developmental trajectories for accuracy scores on the whole-part.

The proportion of correct accuracy scores of whole-face or part-face features was calculated for each child in each of the six conditions (three orientations: upright, 90°, inverted) by either whole- or part-face features (eyes, nose or mouth). Firstly, the average performance scores over the three orientations (collapsed across stimulus type) showed a gradual but not significant decrease as the face was rotated: upright faces: = 79%; 90° faces = 76% and inverted faces = 71%. As shown in Figure 5.2 overall performance accuracy was better in recognition of part-faces (average accuracy = 82%) than whole-faces (average accuracy = 69%), (main effect of whole-part face:  $F(1, 23) = 78.32, p < .001, \eta_p^2 = .378$ ). Overall, performance accuracy improved with increasing age (effect of age:  $F(1, 23) = 48.32, p < .001, \eta_p^2 = .924$ ). Of main interest was the presence of a significant 3-way interaction between orientation, stimulus type and performance across ages ( $F(1,23) = 32.32, p = .041, \eta_p^2 = .170$ ) Analysis of this interaction indicated that rotation of stimuli had increasingly strong effect on the whole-face condition with age but remained the

same size for the part-face condition. Also, further analysis of the whole- and part-face recognition in the upright orientation revealed that the part-face advantage was only present until around 8:09 years of age (Appendix B, Figure 1). A significant 3-way interaction indicated that rotation effect becomes increasingly strong with age (orientation x whole-part x age:  $F(1,23) = 32.32$ ,  $p = .041$ ,  $\eta_p^2 = .170$ ). However, the inversion effect was only found in the whole-face conditions ( $F(1, 23) = 14.42$ ,  $p < .001$ ,  $\eta_p^2 = .329$ ). As illustrated in Figure 5.3, the emergence of the inversion effect was evident as early as 6 years old children, which is in line with the results reported by Tanaka and Farah (1993).

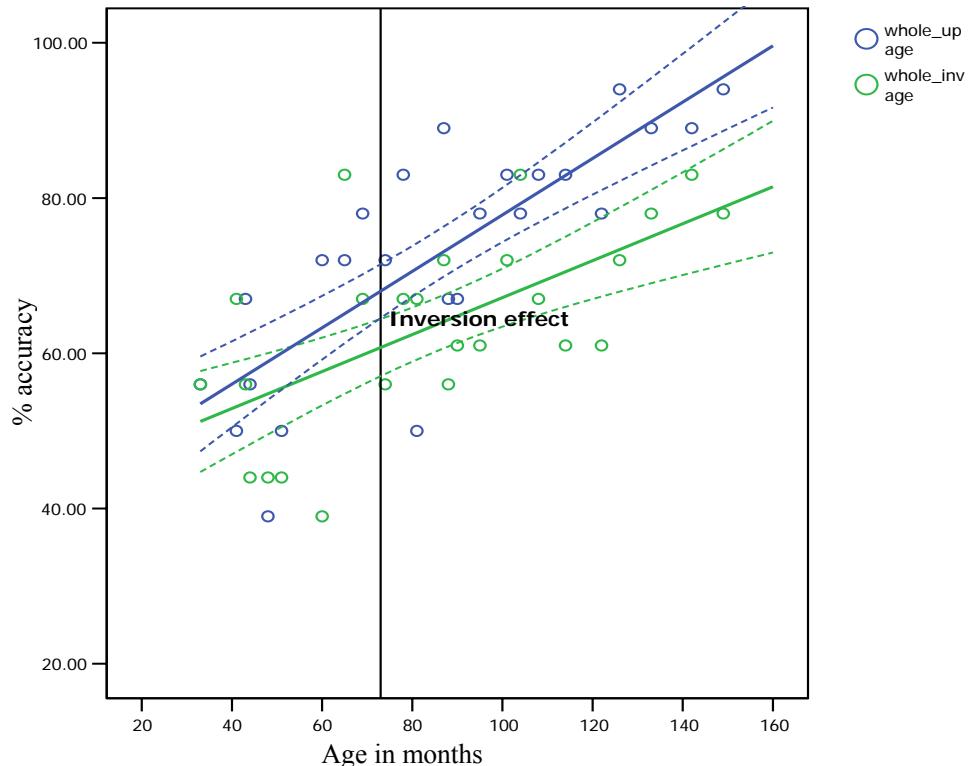


Figure 5.3: TD group developmental trajectories accuracy scores on whole-face condition with 5% and 95% confidence intervals.

### Feature salience

The main effect of features was not significant ( $F(1, 23) = .73$ ,  $p = .789$ ,  $\eta_p^2 = .008$ ). However, individual features (eyes, nose or mouth) were considered separately for whole- and part-face conditions to identify which individual feature generated the highest accuracy. In the whole-face condition, children performed significantly better in the upright orientation when recognition depended on the eyes, achieving ceiling scores from the age of 9 years old (main effect of feature:  $F(1,23) = 5.09$ ,  $p = .034$ ,  $\eta_p^2 = .181$ ), as depicted in Figure 2a in Appendix B and Table 5.1. A robust inversion effect was also found when recognition depended on eyes (interaction of eyes x orientation:  $F(1,23) = 11.71$ ,  $p = .002$ ,  $\eta_p^2 = .337$ ). Also, the rate of development varied for each feature at a marginal level of significance (interaction of feature x age:  $F(1,23) = 4.07$ ,  $p = .053$ ,  $\eta_p^2 = .150$ ), possibly due to different starting accuracy levels. In the part-face there was no main effect of features ( $F(1,23) = .074$ ,  $p = .789$ ,  $\eta_p^2 = .003$ ) nor significant interactions.

In summary, children in the TD group showed a typically observed pattern of holistic encoding with a gradual increase in inversion effect with age. Eyes were a privileged feature in upright whole faces.

#### *5.2.1.1.2 HFA group*

As depicted in Figure 5.4, the HFA group did not show any difference in their average accuracy scores when the face was rotated: upright faces = 73%; 90° faces = 72% and inverted faces = 71%; (effect of orientation:  $F(1,14) = .901$ ,  $p = .359$ ,  $\eta_p^2 = .060$ ). Children performed marginally better on recognition of part -face stimuli (average accuracy = 78%) than whole-faces (average accuracy = 66%), (effect of whole-part face:  $F(1,14) = 28.33$ ,  $p < .058$ ,  $\eta_p^2 = .305$ ). Overall, performance accuracy improved with increasing age (effect of age:  $F(1,14) = 30.45$ ,  $p < .001$ ,  $\eta_p^2 = .685$ ). The 3-way interaction was not significant as the performance was not influenced by orientation and stimulus type (interaction of orientation x whole-part x age:  $F(1,14) = .609$ ,  $p = .610$ ,  $\eta_p^2 = .090$ ).

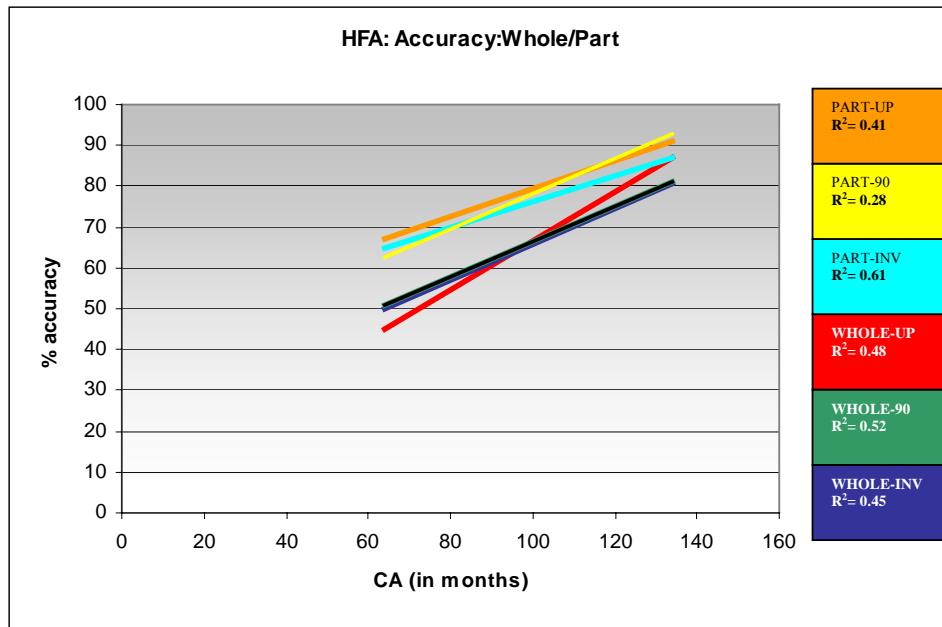


Figure 5.4: HFA group developmental trajectories for accuracy scores on the whole-part.

### Feature salience

Overall, face recognition did not depend on any individual features (effect of features:  $F(1,14) = .410$ ,  $p = .535$ ,  $\eta_p^2 = .028$ ). Separate analyses for whole- and part-feature conditions were carried out. Table 5.1 and Figure 2b in Appendix B depict that in the part-face condition, children performed better when recognition was depending on the eyes (87%) and mouth (87%) but not nose (73%), (eyes vs. nose:  $t(15) = 3.6$ ,  $p = .003$ ; mouth vs. nose:  $t(15) = 3.2$ ;  $p = .005$ ) and achieving ceiling scores on mouth feature at around 9 years of age. This was not reliable for the whole-face condition as there were no differences between the features.

### Comparison to TD group

Overall, the HFA group was less accurate than TD group (effect of group:  $F(1,37) = 8.84$ ,  $p = .005$ ,  $\eta_p^2 = .193$ ) with a slower rate of development than the TD group (effect of age x group:  $F(1,37) = 17.21$ ,  $p < .001$ ,  $\eta_p^2 = .522$ ). In the whole-face condition, the HFA group did not exhibit an inversion effect which was in contrast to the TD group (orientation x group x age:  $F(1,37) = 8.84$ ,  $p = .005$ ,  $\eta_p^2 = .193$ ). Lack

of an inversion effect indicates a reliance on feature information during face recognition and immaturity of holistic processing. There were no other significant main effects and interactions. Overall, both groups showed similar developmental profiles on face recognition and performed better on part-face recognition, continuously improving with age (4-way interaction of orientation x whole-part x age x group:  $F(1,37) = .01$ ,  $p = .906$ ,  $\eta_p^2 = .022$ ).

#### *5.2.1.1.3 LFA group*

Firstly, inspection of the  $R^2$  values suggested a larger variability in this group than the other clinical groups (Figure 5.5). Children with LFA were not influenced by rotation of the stimuli: upright faces = 51%; 90° faces = 52% and inverted faces = 56%; (effect of orientation:  $F(1, 15) = .077$ ,  $p = .785$ ,  $\eta_p^2 = .005$ ). Again, as in the groups discussed above, the average performance accuracy was better in part- face recognition (part average accuracy = 61%) than on whole-face (average accuracy = 41%), (main effect of whole-part face:  $F(1, 15) = 32.31$ ,  $p < .050$ ,  $\eta_p^2 = .378$ ). Overall performance accuracy increased with age (effect of age:  $F(1, 15) = 4.74$ ,  $p = .050$ ,  $\eta_p^2 = .255$ ) and there was no influence of orientation nor stimulus type on accuracy scores with increased age (interaction of orientation x whole-part x age:  $F(1, 15) = .04$ ,  $p = .852$ ,  $\eta_p^2 = .002$ ). A strange pattern was observed in the whole-face condition, whereby children performed significantly more accurately on the inverted trials than upright and 90° with increased age (interaction of orientation x age:  $F(1,15) = 14.88$ ,  $p = .049$ ,  $\eta_p^2 = .437$ ). Figure 5.5 clearly demonstrates the unusual treatment of inverted whole-faces in this group.

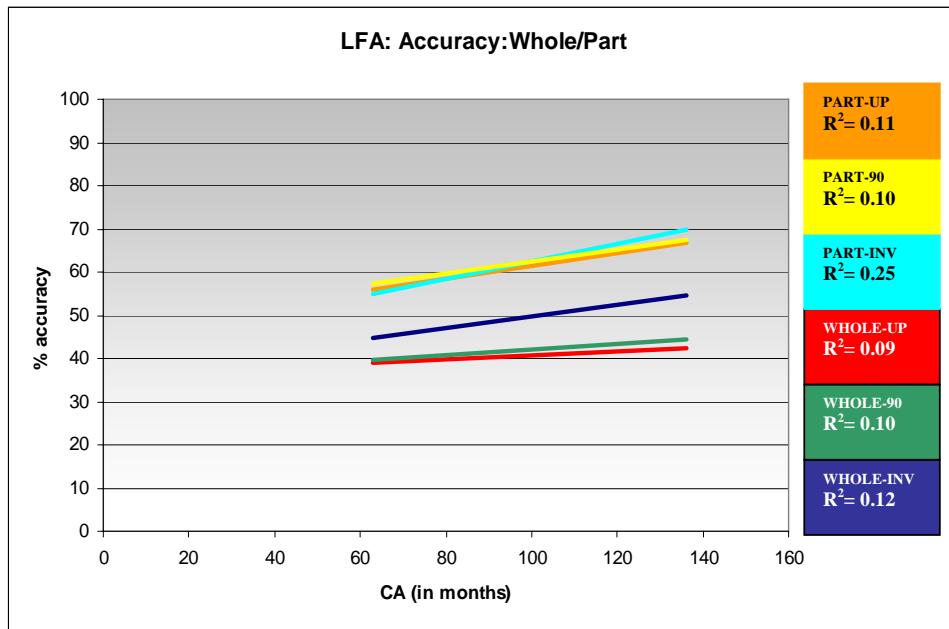


Figure 5.5: LFA group developmental trajectories for accuracy scores on the whole-part.

### Feature salience

Inspection of Table 5.1 indicates poorer overall accuracy on eyes (48%), whereas mouth (66%) modulated the highest accuracy regardless of condition (eyes vs. mouth:  $t(16) = -7.1$ ,  $p < .001$ ; mouth vs. nose:  $t(16) = -5.8$ ;  $p < .001$ ). Figure 2c in Appendix B illustrates this finding.

### Comparison to TD group

Overall, the LFA group was less accurate in comparison to TD group (effect of group:  $F(1,38) = 26.89$ ,  $p < .001$ ,  $\eta_p^2 = .414$ ). In the whole-face condition the LFA group performed not only significantly worse (interaction of orientation x age x group:  $F(1, 38) = 4.21$ ,  $p = .045$ ,  $\eta_p^2 = .100$ ), but also showed opposite pattern of recognition, performing better on inverted trials. In sum, the LFA group showed a different developmental profile in comparison to the TD group (4-way interaction of orientation x whole-part x age x group:  $F(1,38) = 3.85$ ,  $p = .035$ ,  $\eta_p^2 = .111$ ), thus did not show the emergence of holistic processing on their developmental trajectory.

#### 5.2.1.1.4 DS group

As depicted in Figure 5.6, average accuracy declined with rotation of the stimuli in the DS group: upright = 60%; 90° = 54% and inverted = 52%; (effect of orientation:  $F(1,13) = 5.86, p = .031, \eta_p^2 = .311$ ). Unlike other groups, children in the DS group did not show part- over whole-face an advantage on their developmental trajectories (part average accuracy = 60%, whole average accuracy = 50%), although the difference was in this direction (effect of whole-part:  $F(1,13) = .71, p = .795, \eta_p^2 = .071$ ). In general, performance accuracy improved with increased age (effect of age:  $F(1, 13) = 5.14, p = .049, \eta_p^2 = .255$ ). The hallmark of normality was present as the 3-way interaction was significant (interaction of orientation x part-whole x age:  $F(1, 13) = 4.44, p = .050, \eta_p^2 = .254$ ). Children became more sensitive to orientation with increased age in both conditions (whole-face: interaction of orientation x age:  $F(1, 13) = 9.95, p = .008, \eta_p^2 = .431$ ; part-face:  $F(1, 13) = 20.80, p < .001, \eta_p^2 = .988$ ) .

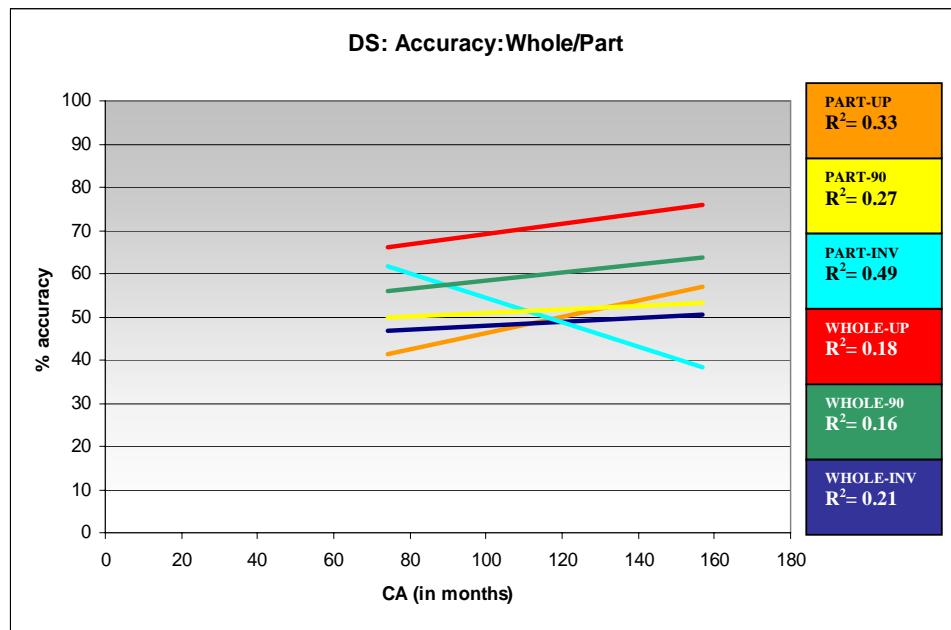


Figure 5.6: DS group developmental trajectories for accuracy scores on the whole-part.

### Feature salience

Although overall performance was not feature dependent (main effect of feature:  $F(1,13) = .986$ ,  $p = .383$ ,  $\eta_p^2 = .059$ ), once conditions were analysed separately children scored at ceiling level when recognition of the face was based on eyes in the upright orientation whole-face condition (see Figure 2d in Appendix B). Similarly to the TD group, in the upright condition, an inversion effect on the eyes and mouth features was found albeit of marginal significance on the former (eyes:  $F(1,13) = 4.08$ ,  $p = .056$ ,  $\eta_p^2 = .247$ ; mouth:  $F(1,13) = 15.28$ ,  $p = .002$ ,  $\eta_p^2 = .435$ ).

### Comparison to TD group

There was no group difference between DS and TD groups (effect of group:  $F(1,36) = .21$ ,  $p = .651$ ,  $\eta_p^2 = .006$ ), but performance in DS group was significantly worse than TD group with delayed rate of development (effect of age x group:  $F(1, 36) = 14.64$ ,  $p < .001$ ,  $\eta_p^2 = .289$ ). Both groups demonstrated better face recognition when it depended on eyes and mouth features in whole-face (interaction of feature x age x group:  $F(1, 36) = 1.91$ ,  $p = .176$ ,  $\eta_p^2 = .049$ ), and were both affected by the stimulus orientation (orientation x age x group:  $F(1,36) = .56$ ,  $p = .459$ ,  $\eta_p^2 = .015$ ). However, the DS group's profile of development was atypical in comparison to TD in number of ways: DS group was less accurate, showed inversion effect on both conditions and earlier advantage of whole-vs.-part recognition than TD group (4-way interaction of orientation x part-whole x age x group:  $F(1, 36) = 8.03$ ,  $p = .007$ ,  $\eta_p^2 = .182$ ).

#### *5.2.1.1.5 WBS group*

For the WBS group, the average accuracy was approximately equivalent across the conditions: upright = 71%,  $90^\circ$  = 68% and inverted = 64%; (effect of orientation:  $F(1,13) = .168$ ,  $p = .691$ ,  $\eta_p^2 = .013$ ), with no indication that the inversion effect altered across chronological age (interaction of orientation x age:  $F(1,13) = .064$ ,  $p = .804$ ,  $\eta_p^2 = .044$ ). As shown in Figure 5.7, the average performance accuracy was higher in part-face recognition (part average accuracy = 74%) than on whole-face (average accuracy = 62%) with marginal significance (main effect of whole-part face:  $F(1,13) = 13.31$ ,  $p < .051$ ,  $\eta_p^2 = .378$ ). Surprisingly, overall performance

accuracy did not increase with age (effect of age:  $F(1,13) = .589$ ,  $p = .456$ ,  $\eta_p^2 = .043$ ) but when conditions were analysed separately, it appeared that children improved marginally with age in part-face trials (effect of age:  $F(1,13) = 11.21$ ,  $p = .056$ ,  $\eta_p^2 = .184$ ), driven most strongly in the upright condition. Another interesting pattern emerged in the whole-face condition whereby children performed better in the  $90^\circ$  and inverted condition than upright with increased age (interaction of orientation x age:  $F(1, 13) = 14.88$ ,  $p = .049$ ,  $\eta_p^2 = .437$ ).

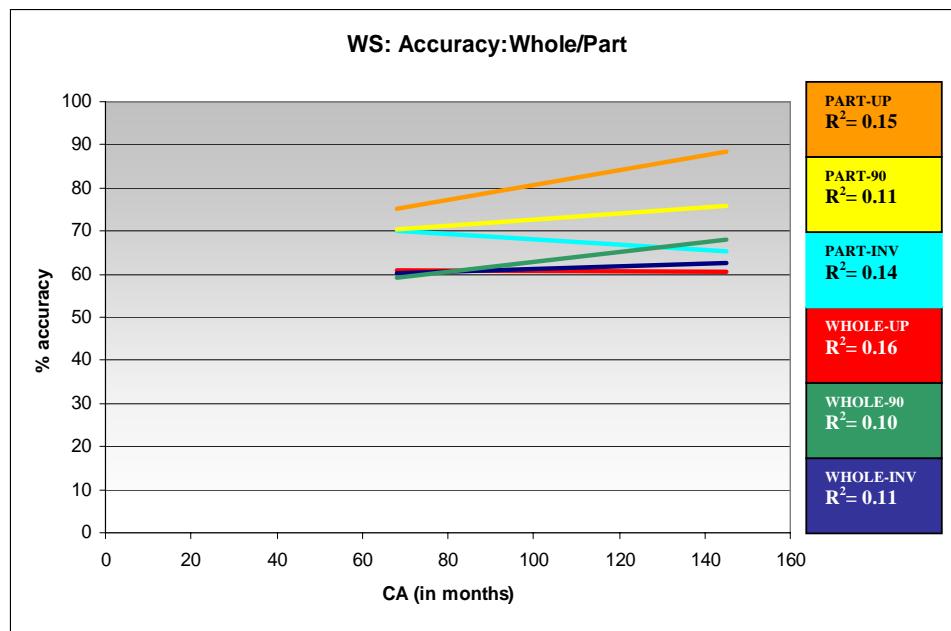


Figure 5.7: WBS group developmental trajectories for accuracy scores on the whole-part.

### Feature salience

Although there was no significant difference in accuracy between features (effect of features:  $F(1, 13) = 1.46$ ,  $p = .246$ ,  $\eta_p^2 = .089$ ), in the whole-face condition children were most accurate when face recognition depended on eyes (eyes vs. mouth:  $t(14) = 3.67$ ,  $p = .003$ ; eyes vs. nose:  $t(14) = 3.29$ ;  $p = .005$ ). Figure 2e in Appendix B illustrates this finding.

### Comparison to TD group

Overall, the WBS group's accuracy was significantly poorer and improved at a slower rate than TD group (main effect of group:  $F(1,36) = 29.69$ ,  $p < .001$ ,  $\eta_p^2 = .452$ ; interaction of group x age:  $F(1,36) = 16.02$ ,  $p < .001$ ,  $\eta_p^2 = .308$ ). Separate analysis for whole- and part-face conditions revealed that WBS group exhibited similar profile of performance as TD group in part-face condition (interaction of feature x orientation x age x group:  $F(1,36) = 2.65$ ,  $p = .110$ ,  $\eta_p^2 = .066$ ). However, taken all together, children in WBS group showed atypical pattern of face recognition in comparison to TD group (4-way interaction of orientation x part-whole x age x group:  $F(1, 36) = 4.52$ ,  $p = .040$ ,  $\eta_p^2 = .112$ ), consistent with the idea that individuals with WBS use featural encoding in their face recognition.

#### **5.2.1.2 Can the Benton test predict accuracy on the Rotating faces task?**

The next step was to explore whether the Benton test could predict performance on whole-part paradigm (performance for each group on this test was described in chapter 4). The main question using this approach is: Are the patterns of performance exhibited by each disorder group in line with their face recognition abilities? The CA ages (on X axis) were replaced with Benton age equivalent scores, derived from our TD sample (see section 4.5.2.1). For brevity, the results are discussed and presented in Table 5.2 and Appendix B (Table 3).

Benton predicted overall accuracy scores only in HFA group (main effect of test age:  $F(1, 14) = 20.73$ ,  $p < .001$ ,  $\eta_p^2 = .597$ ). There was no main effect of test age in other disorder groups: (LFA group:  $F(1, 15) = 2.56$ ,  $p = .131$ ,  $\eta_p^2 = .146$ ; DS group:  $F(1, 13) = .92$ ,  $p = .356$ ,  $\eta_p^2 = .066$ ; WBS group:  $F(1, 13) = .01$ ,  $p = .953$ ,  $\eta_p^2 = .001$ ). Close inspection of summary Table 5.2 indicates that even when chronological age was replaced with test age, it did not normalise groups' developmental profile on the task. For example, one can see that none of the groups, except for DS, showed inversion effect on the whole-face condition.

Table 5.2: Summary of each group's performance on whole-part paradigm against Benton test age. \* Normal profile is based on no-significant 4-way interaction of orientation x whole-part x age x group (compared to TD group).

GROUP	BETTER ACCURACY	INVERSION	INVERSION	NORMAL PROFILE*
		WHOLE	PART	
HFA	PART	NO	NO	YES (but no inversion effect)
LFA	PART	NO	NO	NO
DS	NS DIFFERENCE	YES	YES	NO
WBS	PART	NO	NO	NO

### 5.2.1.3 Summary of accuracy results

Table 5.2 depicts summary of results on whole-part paradigm task. To summarise the relevant accuracy for different features and on whole-part conditions, collapsed group averages are shown in Figures 5.8 and 5.9. Note that chronological age differences between the groups were not equated in these graphs.

Table 5.3: Summary of each groups' performance on whole-part paradigm against CA.

GROUP	BETTER ACCURACY	INVERSION	INVERSION	FEATURE SALIENCE *	NORMAL PROFILE **
		WHOLE	PART		
TD	PART up to 8:9 yrs old	YES from 6 yrs old	NO	WHOLE: EYES & MOUTH	-----
				PART: NS DIFFERENCE	
HFA	PART	NO	NO	WHOLE: NS DIFFERENCE	YES (but no inversion effect)
				PART: EYES & MOUTH	
LFA	PART	NO	NO	WHOLE: MOUTH	NO
				PART: MOUTH	
DS	NS DIFFERENCE	YES	YES	WHOLE: EYES & MOUTH	NO
				PART: NS DIFFERENCE	
WBS	PART	NO	NO	WHOLE: EYES	NO
				PART: NS DIFFERENCE	

\*best accuracy on individual features; \*\* normal profile = ns 4-way interaction

(orientation x whole-part x age x group).

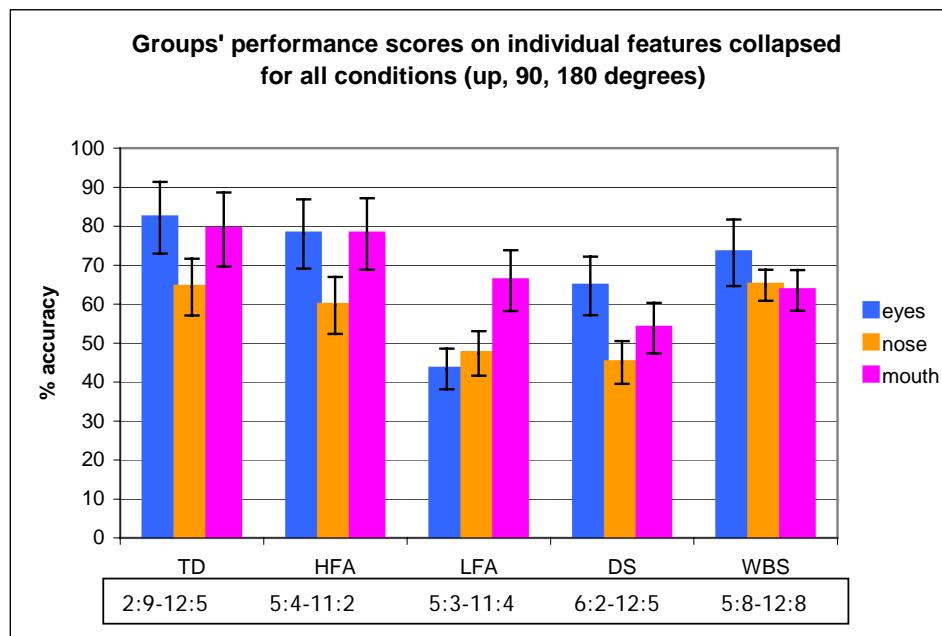


Figure 5.8: Percentage of correct accuracy scores on each feature for each group. CA range of each group is shown.

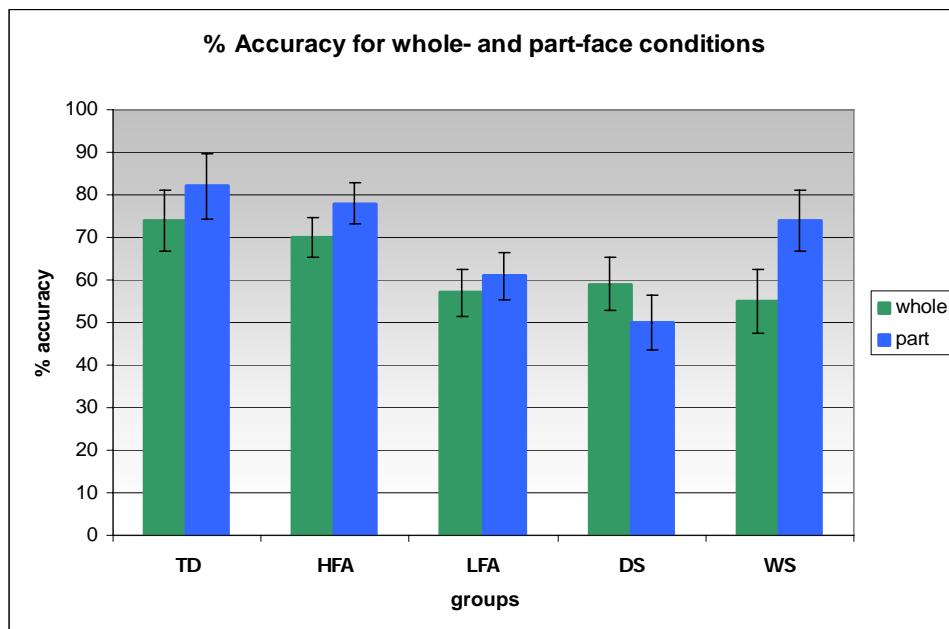


Figure 5.9: Percentage of correct responses on whole and part recognition trials for each group.

### **5.2.1.3 Response Times**

Median reaction times for whole- and part-face over the three orientations were compared to chronological age. Trajectories were constructed linking log (RT) to CA, as this transformation was found to best linearise the data.

Control typically developing children exhibited a significant reduction in reaction time with age ( $F(1,23) = 25.11$ ,  $p < .001$ ,  $\eta_p^2 = .522$ ). However, a significant time cost of recognising inverted faces in the whole-face condition emerged ( $F(1,23) = 28.13$ ,  $p < .001$ ,  $\eta_p^2 = .631$ ). The TD trajectories are shown in Figure 5.10 and trajectories for each clinical group are shown in Figures 5.11 - 5.14.

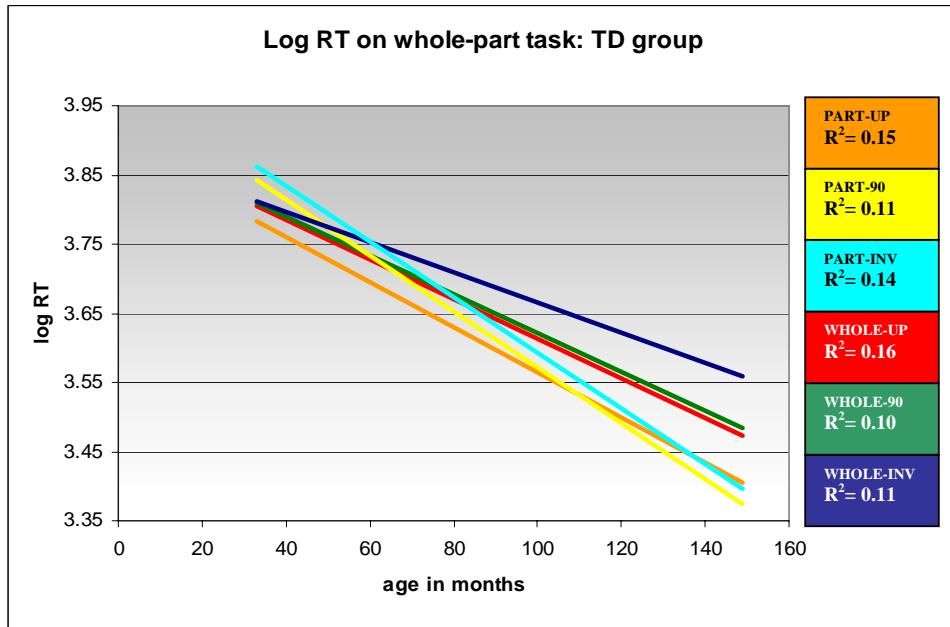


Figure 5.10: TD group developmental trajectories for RT scores on the whole-part faces.

A main effect of age was not significant in any of the disorder groups (HFA:  $F(1,14) = 3.30$ ,  $p = .091$ ,  $\eta_p^2 = .191$ ; LFA:  $F(1,15) = 1.87$ ,  $p = .192$ ,  $\eta_p^2 = .111$ ; DS:  $F(1,13) = .96$ ,  $p = .344$ ,  $\eta_p^2 = .069$ ; WBS:  $F(1,13) = .21$ ,  $p = .653$ ,  $\eta_p^2 = .016$ ). Also, unlike the TD group, none of the clinical groups exhibited time cost on the inverted trials in the whole-face condition (main effect of orientation for HFA group:  $F(1,14) = .15$ ,  $p = .705$ ,  $\eta_p^2 = .011$ ; LFA group:  $F(1,15) = .12$ ,  $p = .107$ ,  $\eta_p^2 = .106$ ; DS group:  $F(1,13) = .27$ ,  $p = .616$ ,  $\eta_p^2 = .020$ ; WBS group:  $F(1,13) = .38$ ,  $p = .551$ ,  $\eta_p^2 = .028$ ). An interesting pattern emerged in the LFA group where responses became slower on the whole-face condition with increased age but in contrast they became faster on the part-face trials with increased age (interaction of orientation x age:  $F(1,15) = 8.64$ ,  $p = .010$ ,  $\eta_p^2 = .365$ ). This finding is illustrated in Figure 5.12. There were no other main effects and interactions for separate clinical groups.

Direct comparison of the TD group with the clinical groups revealed that there were no overall differences in the response time between the TD and HFA group ( $F(1,37) = 3.07$ ,  $p = .088$ ,  $\eta_p^2 = .077$ ) and the TD and DS group ( $F(1,16) = 2.21$ ,  $p = .146$ ,  $\eta_p^2 = .058$ ). However, children in the LFA and WBS groups became significantly slower with age in comparison to the TD group (LFA group:  $F(1,38) = 6.04$ ,  $p = .019$ ,  $\eta_p^2$

= .137; WBS group:  $F(1,36) = 7.32$ ,  $p = .010$ ,  $\eta_p^2 = .169$ ). There were no other significant main effects and interactions.

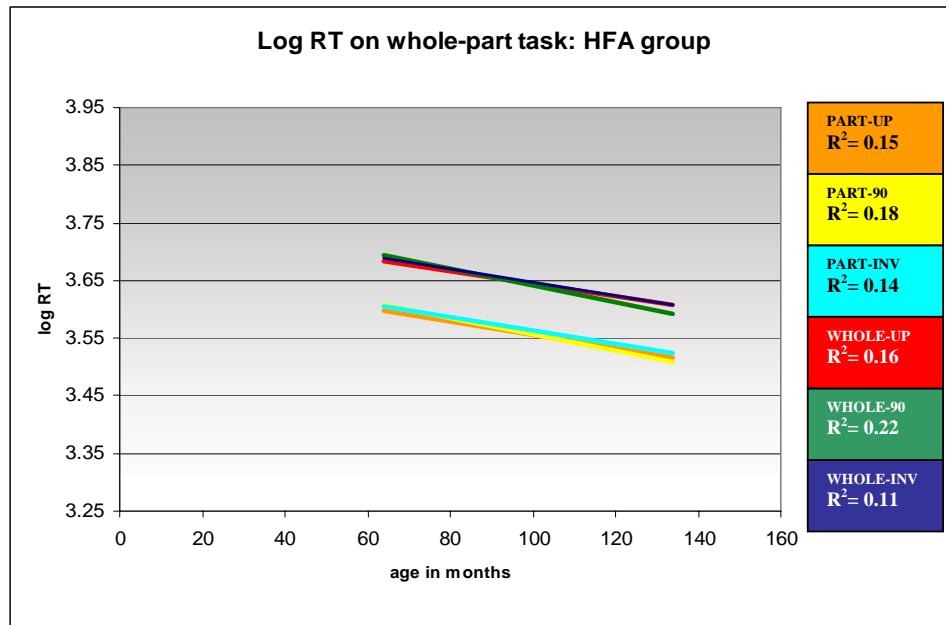


Figure 5.11: HFA group developmental trajectories for RT scores on the whole-part faces.

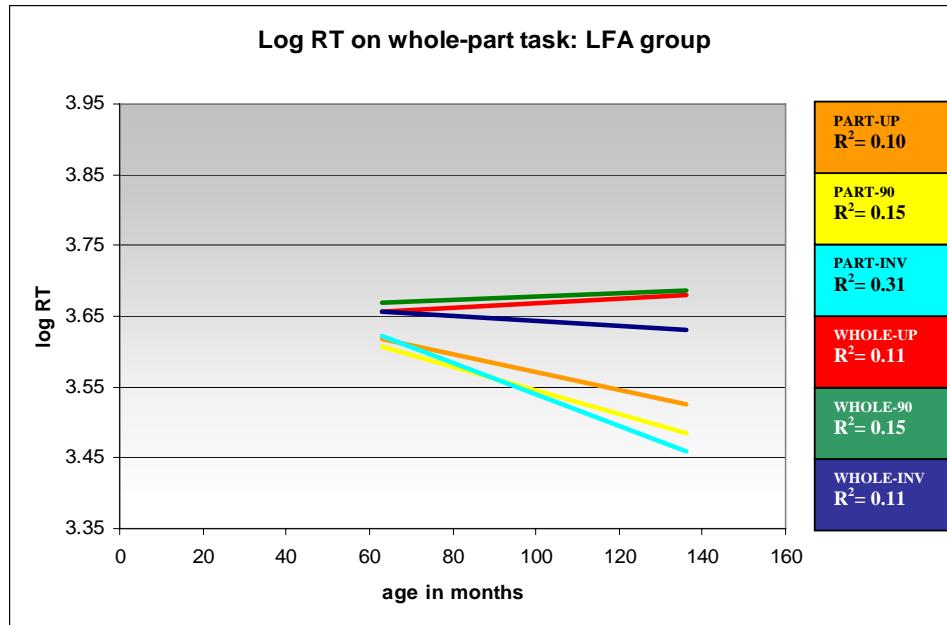


Figure 5.12: LFA group developmental trajectories for RT scores on the whole-part faces.

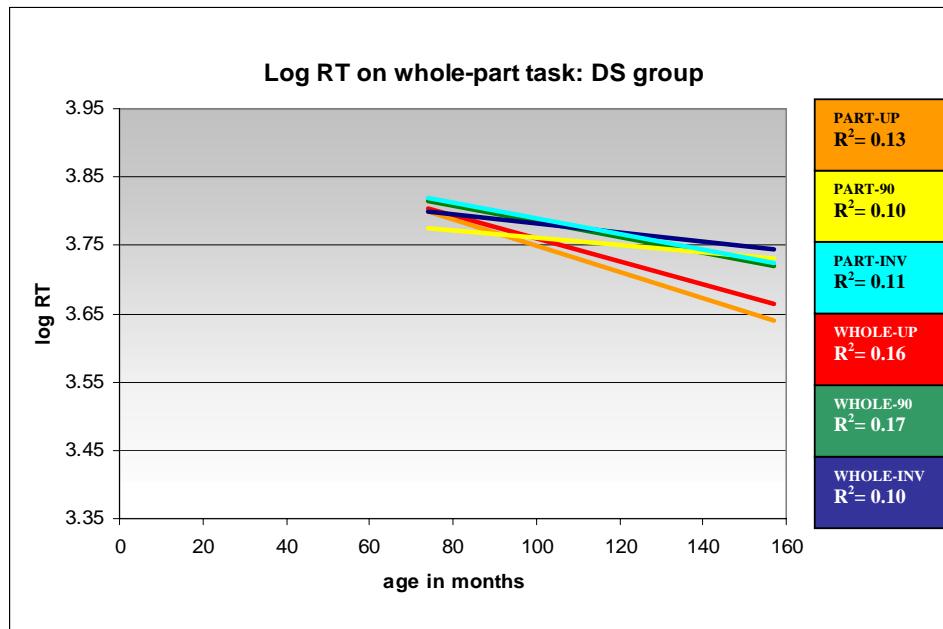


Figure 5.13: DS group developmental trajectories for RT scores on the whole-part faces.

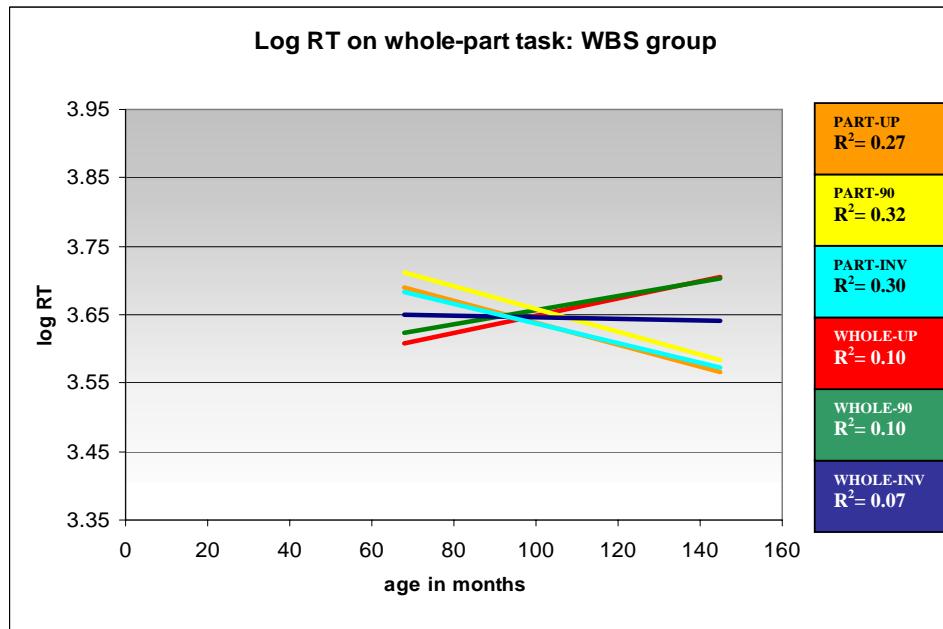


Figure 5.14: WBS group developmental trajectories for RT scores on the whole-part faces.

Table 5.4: Median RT (in seconds) with SD for all the groups on whole-part paradigm. Note that the TD group mean age is younger than the clinical groups.

GROUP	WHOLE-UP	PART-UP	WHOLE-90	PART-90	WHOLE-INVERTED	PART-INVERTED
TD	4.8 (2.1)	4.3 (1.8)	4.8 (1.8)	4.6 (1.7)	5.1 (2.0)	4.8 (1.5)
HFA	4.4 (1.3)	3.6 (0.7)	4.4 (0.9)	3.6 (0.8)	4.5 (0.9)	3.7 (0.7)
LFA	4.7 (1.1)	3.8 (1.0)	4.7 (0.8)	3.5 (0.7)	4.4 (1.1)	3.5 (0.7)
DS	5.5 (1.8)	5.5 (1.6)	6.0 (1.9)	5.8 (2.1)	6.0 (2.2)	6.0 (2.0)
WBS	4.6 (1.8)	4.3 (1.9)	4.7 (2.1)	4.5 (1.7)	4.5 (2.1)	4.3 (1.8)

Overall, RT data turned out to be less sensitivity for identifying developmental changes on this task; consequently no other details will be reported in this section.

### 5.2.3 Discussion

The purpose of this study was to establish whether children in the disorder groups develop sensitivity to holistic processing and to determine how their developmental trajectories compare to the typically developing control group. In the case of the typically developing group the data showed that their performance on face recognition developed linearly. Furthermore contrary to the prediction, children in the TD group showed advantage of feature recognition in the part-face. However, this effect was significant until the age of 8:9 years old, when children became equally good on recognition of both whole- and part-faces.

The rotation of the face from the upright to inverted orientation showed a gradual decrease in performance but only in the whole-face condition. Consistent with the prediction, the emergence of adult-like inversion effect was observed as young as 6 years old. This result has been demonstrated consistently in previous studies (e.g., Tanaka & Farah, 1993; Yin, 1969) and it is the sign of an emerging specialisation for holistic and configural processing. Another consistent pattern established previously (e.g., Tanaka and Farah, 1993) is that overall performance on part-face condition was

not influenced by orientation. However, when recognition was dependent on eyes, children showed an inversion effect in the part-face condition. The eye-feature inversion was observed in several studies, and it was suggested that individual features such as eyes are highly detailed (as discussed in chapter 1). It is possible that configural encoding of individual features as well as holistic processing of the whole face can occur simultaneously (e.g., Lewis & Glenister, 2003; Rhodes et al., 1993).

Some of the differences in the results obtained in the current study and previous ones could be attributed to methodological variations between the current study and the previous ones. One main issue is that Tanaka and Farah (1993) included an additional component of memorising names of the faces, which would require more complex cognitive skills as proposed by Bruce and Young model (1986). Moreover, the whole-face advantage was observed in 9-year-olds, which was the youngest group studied in a more recent study by Joseph and Tanaka (2003). Thus direct comparison is not possible due to differences in ages and exclusion of the age variable in their statistical analysis. In the current study, age was used as continuous variable whereas in most of the previous studies, age was used as a categorical variable.

With respect to the responses exhibited by children in disorder groups, they showed different developmental patterns in comparison to typically developing children. As predicted, children in the autism and WBS groups performed better on the part-face condition. Conversely, children in DS group demonstrated an early developmental advantage of recognising features presented in the context of faces. Also, all the disorder groups showed significantly poorer performance than the TD group on whole-face recognition. In line with the prediction, none of the disorder groups showed a sign of the inversion effect, which suggests that none of the groups have developed normal specialised face recognition expertise.

Furthermore, the clinical groups diverged in other important ways from the TD group. Interestingly, each disorder group showed different profiles of performance from the TD group and from each other ('inter-group') and within different subsets of the same group ('intra-group'), which will be discussed here in turn.

***Inter- and intra-group comparisons: Autism***

In the case of autism groups, overall there was a massive difference in their performance, with HFA group performing significantly better than LFA on the task. Children in the HFA group exhibited increased sensitivity to holistic face processing in the upright condition, which was mainly dependent on recognition by eyes and mouth. This is in part consistent with the findings of López and colleagues (2004), who also showed the presence of holistic processing when participants were cued to face features during recognition (see also Joseph & Tanaka, 2003; Mottron, Burack, Iarocci, Belleville & Enns, 2003). In contrast, children in the LFA group were poor at eye recognition and relatively good at mouth recognition across all of the conditions. This pattern of reverse recognition was found in previous studies (Joseph & Tanaka, 2003; Langdell, 1978; Klin, et al., 2002). Together, these findings suggest an unusual preference for the mouth region in face processing by individuals with severe autism in the current study. This finding is broadly consistent with studies in autism that reported a variety of abnormalities in processing information such as the direction of other people's eye-gaze which is related to impairment in social and cognitive functioning (Baron-Cohen, Campbell, Karmiloff-Smith, Grant, & Walker, 1995; Leekam, Baron-Cohen, Perrett, Milders, & Brown, 1997; Leekam, Hunnissett, & Moore, 1998; Swettenham et al., 1998). Leekam and colleagues found differences between children with high and low VMA based on BPVS on a gaze following study. The authors reported that children of low VMA showed difficulties with following another person's head turn. On this basis, it was proposed that development of language and attention may account for atypical development of these skills (Leekam, Hunnissett, & Moore, 1998).

In summary, the LFA group's performance was the poorest out of all groups, and did not show holistic face recognition, consistent with previous studies (e.g., Plastead, Swettenham, & Rees, 1999). This suggests that children who show holistic processing are also able to use eyes as the most salient feature in the face recognition. In addition, the results showed that both autism groups were less accurate than typically developing children in the part-face condition. This finding presents a contradictory position to the weak central coherence theory in this population (discussed in chapter 2). Strong differences within the profiles of the

autism population raise the possibility that the ability to respond to holistic tasks may be a marker for severity of perceptual atypically.

### ***Inter-group comparisons***

As predicted, children with DS showed an earlier advantage of whole-face condition in comparison to the TD group. Also, it was the only group that showed an inversion effect in whole- and part-face conditions. These findings could provide a valuable clue that individuals with DS have an early reliance on holistic face encoding compared to typically developing children. It also prompts a possible link to previous studies by Bahrle et al. (1990) who described the performance of individuals with DS as exhibiting a ‘global style of processing’ in drawings and on constructive tasks. Under definition in the current thesis, this would index holistic processing. Another result that is in line with the prediction was that children in DS exhibited the normative pattern of better eye than other features face recognition. There are clear indications that developmental profile exhibited by the DS group could be characterised as delayed rather than atypical. Inspection of Benton test age results also show that the DS group has similar characteristics to the TD group but at much younger age (see Figure 4 in Appendix B). One might ask whether the DS results are stemming from low abilities levels, however, the current investigation suggests that the DS children recruited, are characteristic of their group, displaying the abilities that are representative of their group.

As predicted, children in the WBS group did not show a whole-face advantage and did not demonstrate an inversion effect with increased age. This finding supports the hypothesis that face processing develops atypically in WBS (e.g., Karmiloff-Smith, et al., 2004; Grice et al., 2001). Karmiloff-Smith (1997) reported that although people with WBS enjoy looking at faces and therefore receive frequent face input, but it may be that the quality of the underlying processing is atypical. A study by Brown and colleagues (2003) offers a potential precursor that the visual scanning of the whole face may be limited by ‘sticky fixation’, thus narrowing the focus to one small area such as the eye region (Brown et al., 2003). These data point to early problems in the general domain of visual processing that would have cascading effects on the developing system over time (Karmiloff-Smith et al., 1998).

The current data offer no support to the view that the face processing develops normally in the WBS population. It contradicts earlier whole-part study that individuals with WBS process faces holistically (Tager-Flusberg et al., 2003). The authors examined developmental change across wider age range than considered here (12:01-36:02). They found no correlation between age and overall accuracy performance in WBS or CA groups. Contrasting accuracy performance on whole- and part-faces in the upright condition emerged from their study (71.3% vs. 61.5%, respectively) and in the current study (61% vs. 81%, respectively). However, the interpretation of the data from Tager-Flusberg et al. study is problematic because performance on the potentially more informative inversion condition was at floor, and thus their results are un-interpretable in this regard.

In summary, the results of the study reported here show that participants in the clinical groups display not only delay of various types but atypical development in holistic processing in relation to their CA. Importantly, this also holds with respect to their level of face recognition performance on the Benton test, which suggests atypical underlying processing, except for DS group. Even when Benton scores fell within the normal range, our trajectories showed that the performance of disorder groups was nevertheless lower than the TD group, implying that even ‘scores-in-the-normal-range’ on this task do not imply typical development. Benton scores for face processing in the disorder groups did not predict success levels on holistic processing in whole-part face-recognition tasks. Another important factor of presenting stimuli simultaneously to reduce memory load did not have significant positive impact on face recognition in the clinical groups, in particular LFA group which showed the most peculiar profile on this task.

Furthermore, it is possible that preference for the use of featural information exists until holistic processing is fully matured and a template-like face matching strategy is available to the visual system. This speculation is also consistent with some low-level vision studies that suggest that maturity of primary (cortical) visual system is slow and extends up to late childhood (Kovacs, 1999). Whether this can be used as a direct support for the current study still remains to be investigated.

Results from the studies described above provided a more direct insight into the changes that occur during face recognition development. It also showed that scores in normal range could be achieved via atypical route thus providing direct evidence to theoretical assertion by Karmiloff-Smith (1997).

## **CHAPTER 6**

### **DEVELOPMENT OF CONFIGURAL AND FEATURAL FACE RECOGNITION**



*Can you find the 13 hidden faces?*

## **6.1 INTRODUCTION**

In this chapter, configural and featural face recognition will be examined using two behavioural studies. Configural and featural face recognition require people to detect somewhat subtle differences between the face features and their spatial positioning respectively. Adults can recognise faces under different conditions such as poor lighting or from different angles of face presentation. It has been suggested that configural encoding alone is sufficient for recognising an individual face, however featural recognition can also be sufficient to recognise an individual face as shown by studies using photos of blurred faces where the shape of the features is lost, but is not as efficient as configural information in terms of speed and accuracy (Collishaw & Hole, 2000). These qualitative and quantitative differences between configural and featural encoding have important implications in the real world, and have been a hotly debated issue even in some very early studies of visual perception. For example, Galton (1879), who carried out some of the first psychometric experiments, argued that face recognition is based on higher-order variables than individual features, and sensitivity to configural differences between faces is essential for accurate and fast face recognition. Many current studies suggest that sensitivity to configural information can be attributed to experience gained from exposure to a large set of faces in the environment (e.g., Kanwisher et al., 2004, but see Le Grant et al., 2001, for contrasting argument). The current thesis investigates the development of sensitivity to configural and featural role of configural encoding but it will not address the role of configural and featural recognition in relation to face-specificity, nor will it delve into its role in other homogeneous objects recognition, as it is beyond the scope of this thesis.

From a developmental perspective, it is well established that featural processing is a skill used early in life and precedes the use of configural strategies (Diamond & Carey, 1977; Mondloch et al., 2002). However, given that there are multiple aspects to face processing, reaching adult-levels of performance may mean different things for different processes. The literature is mixed as to when holistic processing and configural strategies emerge. Depending on the paradigm used, the configural information skills have been suggested to emerge by 10 years (Carey & Diamond, 1977; Diamond & Carey, 1977; Mondloch, Geldart, Maurer, & Le Grand, 2003), by 5 years (Brace et al.,

2001), and even suggested to be available by 4 years (Freire & Lee, 2001; Pellicano & Rhodes, 2003).

As described in earlier literature review chapters, there are some hints that configural, featural and holistic aspects of face recognition are dissociable cognitive processes. The previous chapter investigated the development of holistic processing in typically developing children and children with developmental disorders. Studies in the current chapter investigated the developmental course of configural face recognition in comparison with featural processing. Note that only second-order configural processing (perceiving distances between the internal face-features) is examined in this chapter.

Configural face processing was assessed through two different experimental studies and this chapter is divided into two sections entitled Study 2: Jane Faces and Study 3: Story-Book. In Study 2, face recognition was investigated with systematically constructed faces, in which configural or featural properties were manipulated and faces were presented in either upright or inverted orientations. In Study 3, sensitivity to inversion was investigated to tap configural encoding. A task that embeds the recognition of upright and inverted faces in the naturalistic context of a story-book was employed. An important difference between the Studies 2 and 3 is use of memory component in the Study 3. Also, Study 3 was designed to be child-friendly with heavy use of contextual support. The specific investigations and predictions are described in the relevant studies.

The vast majority of research concerning face recognition abilities in the clinical groups has been confined to group comparisons. Current studies investigate sensitivity to configurally manipulated faces in cross-syndrome studies. The relationship between the age and face development in these clinical groups has not been studied so far. This is also the first study to investigate face recognition abilities in children with DS.

## **6.2 STUDY 2: JANE FACES**

The main aim of this study was to investigate when developmental changes of configural processing occur in typically developing children and in children with developmental disorders. The specific investigations and predictions are as follows:

1) When do children show emergence of configural face recognition?

**Predictions:**

- i) Children in the TD group will show an increasing sensitivity to configurally altered faces (e.g., Brace et al., 2001; Diamond & Carey, 1986; Mondloch et al., 2002);
- ii) Children in the clinical groups will be significantly less sensitive to configural processing in comparison to the TD group, since configural processing is associated with face recognition expertise (e.g., Deruelle et al., 2004; Hobson et al., 1988; Kaiser et al., 2005; Karmiloff-Smith et al., 2004).

2) Do children show an inversion effect in the configural condition?

**Predictions:**

- i) TD group will show increased sensitivity to inversion (since this disrupts configural processing). Based on the previous findings it is expected that this effect will be evident by 6 years of age (e.g., Mondloch et al., 2002);
- ii) Children in the disorder groups will not be as affected by inversion of the configurally altered face stimuli (e.g., Hobson et al., 1988; Karmiloff-Smith et al., 2004)

3) What are the developmental trajectories of performance on featurally altered faces?

**Predictions:**

- i) Children in the autism, TD and WBS groups will develop similar sensitivity to featurally altered faces based on previous studies (e.g., Deruelle et al., 1999, 2004);
- ii) Children in the DS group will be less accurate than other groups on the featural condition as suggested by previous researchers (e.g., Bellugi, 1989).

4) Can performance on the Benton test predict level of performance on the ‘Jane’ task?

**Prediction:** Previous findings suggest that the Benton test can be resolved by the use of featural information. Thus, it was predicted that the Benton test will not be a good predictor of configural recognition but will be a good predictor of featural face recognition (Karmiloff-Smith, 1997; Duchaine & Nakayama, 2004).

### 6.2.1 Method

#### 6.2.1.1. Participants

All participants completed the task. See chapter 3 for full details.

#### 6.2.1.2 Stimuli

The stimuli were kindly provided by Catherine Mondloch and Daphne Maurer at the McMaster University, Canada. The stimulus manipulation was based on the original technique used by Freire and Lee (2001). Stimuli were derived from a black and white photo of a woman (called ‘Jane’ in this study) to create new versions of the face. The featural version was created by replacing the eyes or the mouth features of Jane’s face with features of other people. In the configural version (referred to as the ‘spacing set’ by Mondloch et al., 2002) features such as the eyes were moved in either direction (horizontally or vertically) of the inner face, for example, the eyes were moved closer together by 4mm relative to the original. Several research groups have used these stimuli and demonstrated that children and adults are sensitive to the featural and configural manipulations of the stimuli sets (Mondloch et al., 2002; Karmiloff-Smith et al., 2004, de Schonen et al., 2005). All stimuli were 10.2 cm wide and 15.2 cm high. More detailed information about the stimuli and details of other versions such as external contour can be found in Mondloch et al., (2002). Figure 6.1 illustrates a sample of featurally alerted faces (see panel A) and configurally alerted faces (see panel B) in upright and inverted orientations.

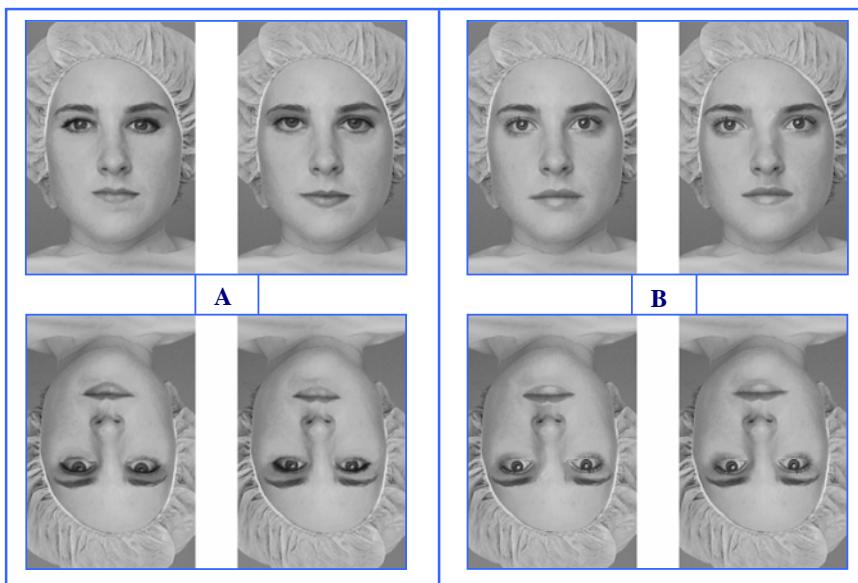


Figure 6.1 Examples of ‘Jane’ faces changed featurally (panel A), and configurally (panel B). Pairs of faces were shown simultaneously and participants had to decide whether faces were same or different. Stimuli provided by Mondloch et al. (2002).

### **6.2.1.3 Procedure**

The procedure employed a well-tested paradigm for differentiating between featural and configural processing of real faces (Mondloch et al., 2002). Participants were presented with two faces simultaneously<sup>(1)</sup> and were asked to determine whether the two faces were the same or different. Trials were blocked into featurally or configurally altered sets and were shown in the upright and inverted orientations. Trials were blocked to encourage the participants to adopt specific face-processing strategies (Mondloch et al., 2002). Stimuli were presented simultaneously on a 17-inch computer monitor using SuperLab Pro 2.0 software.

The testing session began with a game and practice trials, to ensure that all participants understood the instructions and meaning of the words “same” and “different”. The experimenter played a short game with each child, which involved placing objects which were the “same” on one side and “different” separately. Once the experimenter was satisfied that the child understood the rules of the game, the practice trials began.

---

<sup>1</sup> Mondloch et al. (2002) presented the stimuli sequentially.

Small number of practice trials (3 upright and 3 inverted for each condition) was also used due to a short attention span of children with developmental disorders. The experimenter initiated the task by saying: “*Now we are going to play another game, like the game you have just played. Look. This is Jane and these are her sisters* (original model with other modified versions of Jane were presented on the screen). *Some sisters look the same because they are twins. Do you know any twins?... Some sisters look different and they are not twins. Now we are going to play a game where sometimes you will see twin sisters, sometimes not. When you see two faces, that you think look the same press this button* (experimenter shows the relevant button) *and when you think that the faces look different press this button* (experimenter shows the relevant button). *Are you ready?... Try to answer as fast as possible.*

During each trial, two target faces were presented simultaneously, to which the participant had to respond with the “same” or “different” keys on the keyboard. The stimulus was displayed until the response button was pressed. Only two keys on the keyboard were visible, so the child was not able to press wrong key. There were two cards placed under the relevant key, one had same colours (representing ‘same’ response - S key) and the other card had two different colours (representing ‘different’ response - L key). Each participant was presented with 30 trials from the featural and configural sets respectively. For each participant, the upright block was always presented before the inverted block and the order of configural and featural blocks within these was counterbalanced. Each block consisted of 15 ‘same’ (identity trials) and 15 ‘different’ (transformed trials) randomised trials.

It was noticed that most children were very proficient at using computers regardless of their cognitive abilities. As in the previous study, children were provided with stickers as rewards during the breaks. The dependent variable was accuracy of correct choices for the upright and inverted trials in the featural and configural conditions.

### 6.2.2 Results

The task comprised two components: ***difference detection*** (where the difference was due either to a configural or featural transformation) and ***identity recognition*** (for all items where no change had been made between model and target). The transformation

trials and identity trials for featural and configural blocks were analysed separately (see Karmiloff-Smith et al., 2004 for similar strategy). This split was made because the configural and featural factor only applies to transformations. All trials were shown in upright and inverted orientation. Accuracy levels were taken from all participants in the studies. Accuracy results on difference detection trials will be described in detail as they were the most relevant to our study.

1. As in the previous chapter, trajectories were analysed using an analysis of covariance ANCOVA for each group, where the within-participant factors were orientation (upright and inverted) and transformation of the task (configural and featural) with age as co-variant.
2. A direct comparison of each clinical group to TD group was carried out using an ANCOVA with Group (TD group compared to 4 clinical groups) as between-participants factor, within-participant factors of face orientation and transformation, and age as co-variant. As in the previous results sections, the TD group will be described in detail first. Each disorder group will be characterised on its own and then compared to the TD group. Results from the featural and configural sets will then be described separately for each group. In addition, some disorder groups will be compared to each other to explore detailed similarities and differences between them. Lastly, performance scores will be plotted against Benton test (see chapter 4 for details) to explore whether this test is a good predictor of expert face recognition. A summary of each group's performance can be found in Table 6.3.

#### **6.2.2.1 Identity Recognition**

Whether identity recognition trials were presented in configural or featural blocks had no effect on performance in any of the groups. There was no significant interaction of block type on orientation or any other variables, suggesting that trial blocking of featural versus configural condition did not trigger specific face-recognition strategies to affect identify recognition (see Table 6.1). A comparison of accuracy levels in identity recognition revealed no significant difference between TD group and HFA group (main effect of group:  $F(1,37) = 2.29$ ,  $p = .139$ ,  $\eta_p^2 = .058$ ), and DS group (main effect of

group:  $F(1,36) = .09$ ,  $p = .773$ ,  $\eta_p^2 = .002$ ). Children with WBS performed equally well as TD group on featural set, but were far better than TD group on configural trials (main effect of group:  $F(1,37) = 7.94$ ,  $p = .008$ ,  $\eta_p^2 = .177$ ). In contrast, the LFA group performed worse than the TD group on both sets (main effect of group:  $F(1,38) = 7.51$ ,  $p = .009$ ,  $\eta_p^2 = .165$ ).

Table 6.1 Means and standard errors (SE) for accuracy % in Identity Recognition

GROUP	TRIAL BLOCK							
	FEATURAL				CONFIGURAL			
	UPRIGHT		INVERTED		UPRIGHT		INVERTED	
	Mean %	(SE)%	Mean %	(SE)%	Mean %	(SE)%	Mean %	(SE)%
TD	73	3.7	75	3.0	69	3.4	70	2.7
HFA	72	3.4	65	4.1	67	4.0	57	4.4
LFA	58	3.0	61	3.4	51	4.7	51	4.5
DS	63	4.0	57	2.8	61	5.0	56	2.8
WBS	71	2.4	68	3.5	86	3.6	82	5.2

### 6.2.2.2 Difference Detection

The mean accuracy levels for each group are provided in Table 6.2. Figures 6.1- 6.6 depict the performance of each group in terms of correct percentage accuracy scores plotted against increasing chronological age (CA). The solid lines indicate a best-fit regression through each group's data. Due to a large number of main effects and interactions only comparisons directly relevant to current predictions will be reported and p-values for the remaining comparisons can be found in Appendix C (Table 1).

Table 6.2: Means and standard errors (SE) for accuracy % in Difference Detection

GROUP	TRIAL BLOCK							
	FEATURAL				CONFIGURAL			
	UPRIGHT		INVERTED		UPRIGHT		INVERTED	
	Mean %	(SE)%	Mean %	(SE)%	Mean %	(SE)%	Mean %	(SE)%
TD	81	2.8	79	2.5	62	4.8	23	2.3
HFA	79	3.8	80	2.7	42	5.4	47	5.9
LFA	60	4.0	63	3.1	22	3.5	17	3.3
DS	64	4.1	59	3.6	39	6.0	24	3.9
WBS	79	2.7	71	2.1	28	4.4	12	4.1

### 6.2.2.1.1 TD control group

The trajectories generated by TD group accounted for between 62% and 78% of the variance (featural-upright:  $R^2 = .73$ ,  $F(1,23) = 60.52$ ,  $p < .001$ ; featural-inverted:  $R^2 = .62$ ,  $F(1,23) = 37.53$ ,  $p < .001$ ; configural-upright:  $R^2 = .79$ ,  $F(1,23) = 87.41$ ,  $p < .001$ ), except for the configural inverted trajectory which appeared to be more variable ( $R^2 = .23$ ,  $F(1,23) = 6.82$ ,  $p = .016$ ). The control group therefore generated valid cross-sectional developmental trajectories on this task.

Overall, performance accuracy improved with age (main effect of age:  $F(1,23) = 59.61$ ,  $p < .001$ ,  $\eta_p^2 = .722$ ). As shown in Figure 6.1, the TD group exhibited a characteristic pattern in difference detection, whereby recognition of configurally-transformed faces was harder to detect than featurally-transformed faces (main effect of transformation:  $F(1,23) = 39.19$ ,  $p < .001$ ,  $\eta_p^2 = .630$ ). Also, presentation of the faces in different

orientations had an influence on the performance (main effect of orientation:  $F(1,23) = 12.32$ ,  $p = .002$ ,  $\eta_p^2 = .349$ ), but inverting the face added to the difficulty only for configurally changed faces (interaction of transformation x orientation:  $F(1,23) = 7.68$ ,  $p = .011$ ,  $\eta_p^2 = .250$ ).

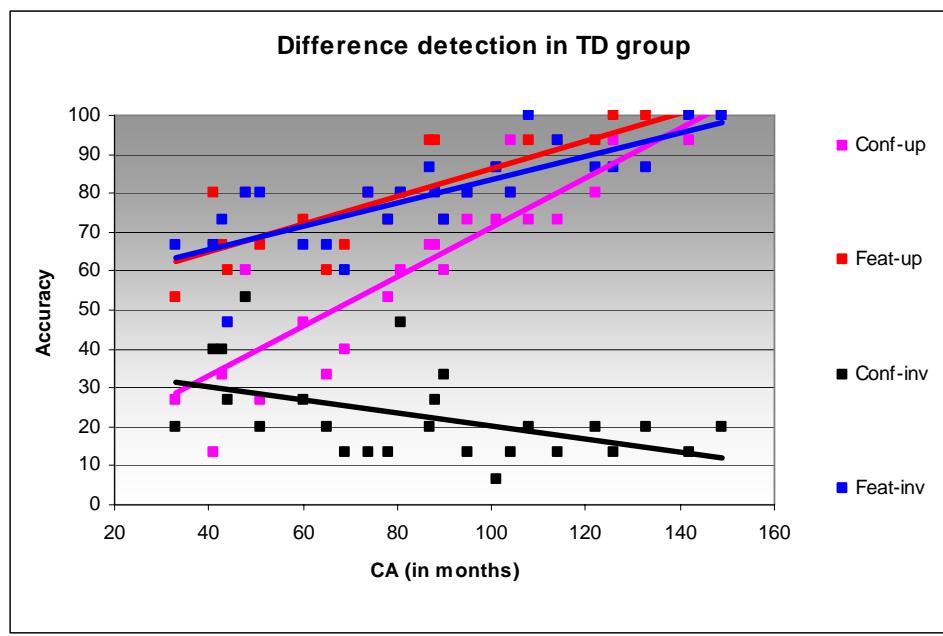


Figure 6.1: TD group developmental trajectory for accuracy scores on the Jane faces.

### Featural trials

In featural trials, performance increased with age (main effect of age:  $F(1,23) = 63.76$ ,  $p < .001$ ,  $\eta_p^2 = .735$ ), and orientation of the stimuli had no influence on the accuracy level (main effect of orientation:  $F(1,23) = .40$ ,  $p = .533$ ,  $\eta_p^2 = .017$ ). Performance accuracy increased with age regardless of orientation the faces were presented (interaction of age x orientation:  $F(1,23) = 1.29$ ,  $p = .268$ ,  $\eta_p^2 = .053$ ).

### Configural trials

Figure 6.1 depicts a steady increase in accuracy scores on upright configurally transformed faces with age (main effect of age:  $F(1,23) = 23.94$ ,  $p < .001$ ,  $\eta_p^2 = .510$ ). However, recognition of faces in inverted orientation had a detrimental influence on performance (main effect of orientation:  $F(1,23) = 12.46$ ,  $p = .002$ ,  $\eta_p^2 = .351$ ), showing a decrease in accuracy with age (interaction of age x orientation:  $F(1,23) = 78.00$ ,  $p < .001$ ,  $\eta_p^2 = .772$ ). Figure 6.2 below depicts this effect.

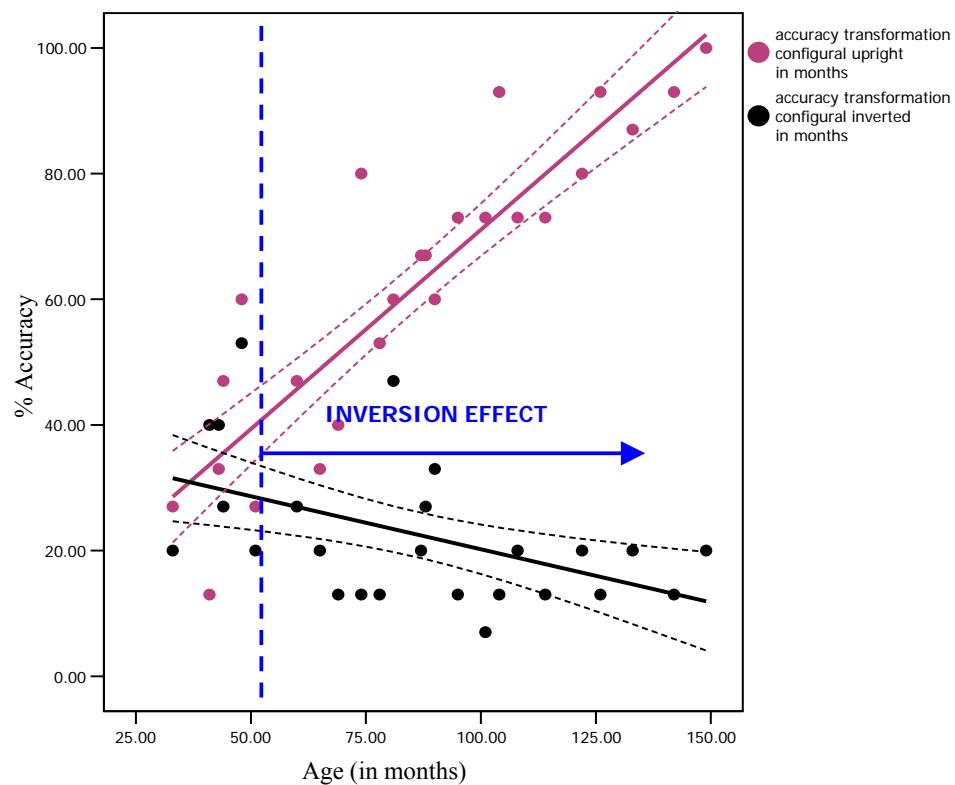


Figure 6.2 shows linear regressions with confidence intervals for configural sets in both orientations for the TD group. Dashed lines show 95% confidence intervals, continuous lines are linear regressions for each condition. Dashed blue line indicates significant differences between those conditions.

In summary, performance of the TD group is consistent with previous studies showing that the development of featural processing is in advance of the development of configural processing (see also Table 6.2). These results are supported by two effects: i) improved accuracy on upright configural trials, which was evident in children as young as 6-year-olds who scored above chance level on (Figure 6.1), and ii) emergence of a classic inversion effect which was evident from the age of 5.8 years old (Figure 6.2). The different pattern for featural and configural transformation across development produced a significant 3-way interaction (orientation x transformation x age:  $F(1,23) = 50.89$ ,  $p < .001$ ,  $\eta_p^2 = .689$ ). This is the hallmark of normal development. In the following sections, developmental profiles of the clinical groups will be assessed whether this hallmark emerged in the trajectories of each disorder group<sup>(2)</sup>.

#### *6.2.2.1.2 HFA group*

As shown in Figure 6.3, recognition of configurally-transformed faces was harder than featurally-transformed faces for children in the HFA group (main effect of transformation:  $F(1,14) = 20.21$ ,  $p < .001$ ,  $\eta_p^2 = .591$ ). Orientation of the faces did not have any influence on accuracy levels (main effect of orientation:  $F(1,14) = .36$ ,  $p = .561$ ,  $\eta_p^2 = .025$ ), and performance improved with age (main effect of age:  $F(1, 14) = 15.29$ ,  $p = .002$ ,  $\eta_p^2 = .522$ ).

---

<sup>2</sup> We will be looking for a 4-way interaction (orientation x transformation x age x group).

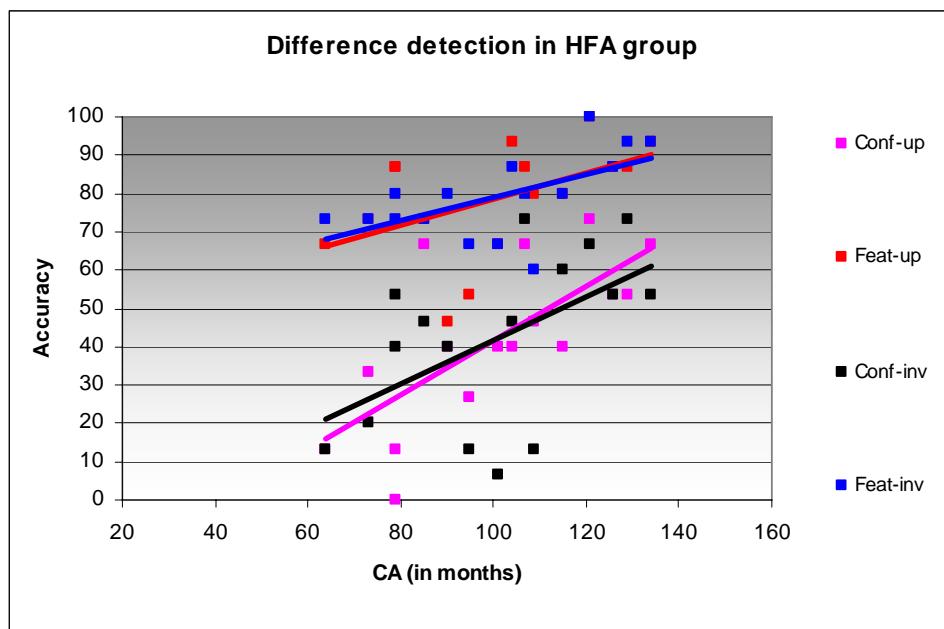


Figure 6.3: HFA group developmental trajectories for accuracy scores on the Jane faces.

### Featural trials

The HFA group performance on featural trials increased with age (effect of age:  $F(1,14) = 7.42$ ,  $p = .016$ ,  $\eta_p^2 = .346$ ), and orientation of the stimuli had no influence on the accuracy level (effect of orientation:  $F(1,14) = .09$ ,  $p = .764$ ,  $\eta_p^2 = .007$ ). Performance accuracy increased with age regardless of the orientation in which the faces were presented (interaction of age x orientation:  $F(1,14) = .08$ ,  $p = .780$ ,  $\eta_p^2 = .006$ ).

### Configural trials

As shown in Figure 6.3, there was more variability in the performance on configural trials. Orientation had no influence on accuracy (main effect of orientation:  $F(1,14) = .24$ ,  $p = .632$ ,  $\eta_p^2 = .017$ ) and performance significantly improved with age (main effect of age:  $F(1,14) = 14.84$ ,  $p = .002$ ,  $\eta_p^2 = .515$ ) regardless of orientation (interaction: age x orientation:  $F(1,14) = .25$ ,  $p = .625$ ,  $\eta_p^2 = .017$ ).

### Comparison to TD trajectory

Overall, the HFA group was less accurate (delayed onset) in comparison to the TD group (effect of group:  $F(1,37) = 5.41$ ,  $p = .026$ ,  $\eta_p^2 = .128$ ). HFA group was the only group exhibiting a significant group difference in comparison to the TD group, for brevity of this section it will not be reported for the remaining TD and disorder group comparisons in the following section. The HFA and TD groups had a similar rate of improvement with age (effect of age x group:  $F(1,37) = 3.48$ ,  $p = .070$ ,  $\eta_p^2 = .086$ ), but note that it was at marginal level. In general, both groups were differentially influenced by face orientation with age (interaction of orientation x age x group:  $F(1,37) = 6.18$ ,  $p = .018$ ,  $\eta_p^2 = .143$ ). Separate analysis on featural trials demonstrated that both groups exhibited similar profile of performance by improving with age regardless of orientation (interaction of orientation x age x group:  $F(1,37) = .008$ ,  $p = .930$ ,  $\eta_p^2 = .187$ ). However, a significant difference between the groups was observed on configural condition where the HFA group performed significantly worse on upright configural trials and did not show emergence of a classic inversion effect (interaction of orientation x age x group:  $F(1,37) = 7.20$ ,  $p = .011$ ,  $\eta_p^2 = .163$ ). Lack of an inversion effect indicates a reliance on featural encoding and immaturity of cognitive encoding. Overall, children in the HFA demonstrated an atypical profile of face recognition in comparison to the TD group as they did not exhibit a hallmark of normal face recognition as shown by significant 4-way interaction (interaction between: task x orientation x age x group:  $(F(1,37) = 5.12$ ,  $p = .030$ ,  $\eta_p^2 = .122$ ).

#### *6.2.2.1.3 LFA group*

The LFA group displayed the most variability in the performance of the disorder groups. As shown in Figure 6.4, recognition of configurally-transformed faces was harder than featurally-transformed faces (effect of transformation:  $F(1,15) = 8.01$ ,  $p = .013$ ,  $\eta_p^2 = .348$ ). Orientation of the faces had a marginal influence on accuracy levels (effect of orientation:  $F(1,15) = 3.99$ ,  $p = .064$ ,  $\eta_p^2 = .210$ ), although this main effect masks several interactions. More importantly, overall performance did not improve with age (effect of age:  $F(1, 15) = 3.07$ ,  $p = .100$ ,  $\eta_p^2 = .170$ ).

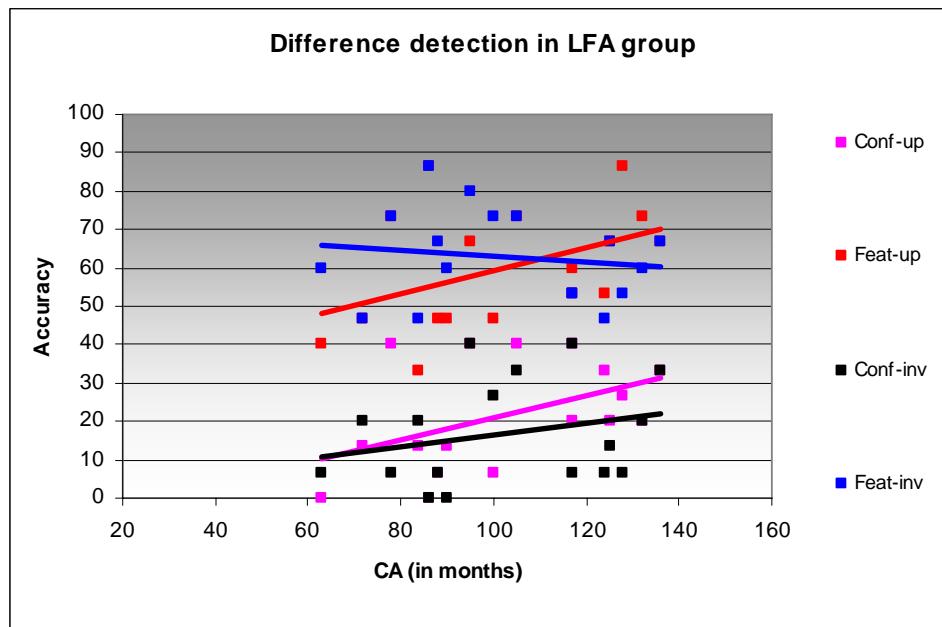


Figure 6.4: LFA group developmental trajectory for accuracy scores on the Jane faces.

### Featural sets

The LFA group performance on featural trials did not improve with age (effect of age:  $F(1,15) = .73$ ,  $p = .408$ ,  $\eta_p^2 = .046$ ). The LFA group revealed a significant accuracy decrease when recognising inverted faces (effect of orientation:  $F(1,15) = 9.45$ ,  $p = .008$ ,  $\eta_p^2 = .387$ ), and was significantly correlated with increasing chronological age (interaction of age x orientation:  $F(1,15) = 8.64$ ,  $p = .010$ ,  $\eta_p^2 = .365$ ). Decrease in accuracy on inverted trials emerged at about 9 years olds (up to this age individuals performed better on inverted trials than upright ones). As shown in Table 6.2, overall accuracy on the inverted trials was higher (63%) than on the upright ones (60%).

### Configural sets

Again, as in the featural trials, performance did not reliably increase with age, although the effect size is suggestive of underlying effect (effect of age:  $F(1,15) = 3.02$ ,  $p = .103$ ,  $\eta_p^2 = .167$ ). There was no sign of an inversion effect (effect of orientation:  $F(1,15) = .36$ ,  $p = .560$ ,  $\eta_p^2 = .023$ ) and orientation had no influence on performance change with age (interaction of age x orientation:  $F(1,15) = .85$ ,  $p = .371$ ,  $\eta_p^2 = .054$ ).

### Comparison to TD trajectory

Direct comparison against the normal developmental trajectory revealed that the LFA group was differentially affected by orientation with increased age (3-way interaction of orientation x age x group:  $F(1,38) = 33.32, p < .001, \eta_p^2 = .467$ ). Separate analysis of each condition revealed that there was significant difference between the groups on featural trials (3-way interaction of orientation x age x group:  $F(1,38) = 7.13, p = .010, \eta_p^2 = .165$ ), which was also observed on the configurally altered condition (3-way interaction of orientation x age x group:  $F(1,38) = 14.97, p < .001, \eta_p^2 = .294$ ).

In summary, the LFA group showed an atypical profile of performance in comparison to the TD group: i) they did not improve with increased age on both configurally and featurally transformed faces and ii) exhibited an “inverted inversion effect” on featural trials, and younger children were better on featural inverted faces but this performance declined with age. In sum, there was no hallmark of normally developing face recognition skills (task x orientation x age x group:  $(F(1,38) = 26.37, p < .001, \eta_p^2 = .410)$ .

#### *6.2.2.1.4 DS group*

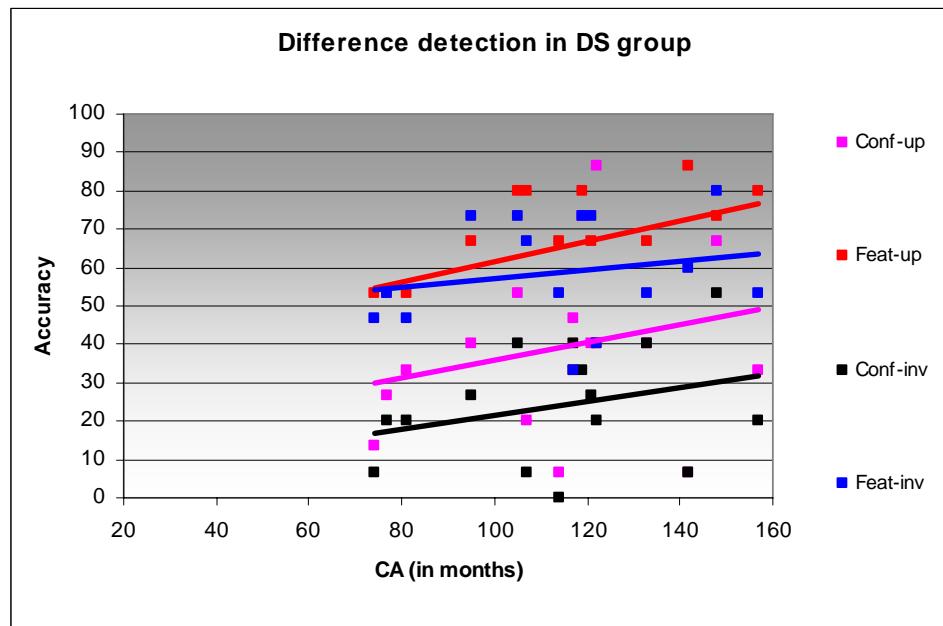


Figure 6.5: DS group developmental trajectory for accuracy scores on the Jane faces.

As depicted in Figure 6.5, the DS group showed a familiar pattern of better accuracy performance on featural over configural trials on their developmental trajectories (effect of transformation:  $F(1,13) = 13.34$ ,  $p = .004$ ,  $\eta_p^2 = .572$ ). Overall, presentation of the faces in different orientations had no influence on accuracy levels (effect of orientation:  $F(1,13) = .01$ ,  $p = .946$ ,  $\eta_p^2 = .001$ ). Also, performance did not improve with increasing age (effect of age:  $F(1, 13) = 3.0$ ,  $p = .106$ ,  $\eta_p^2 = .188$ ).

#### Featural sets

There was no significant effect of the stimulus orientation (effect of orientation:  $F(1,13) = .91$ ,  $p = .400$ ,  $\eta_p^2 = .055$ ). The performance did not increase with increasing age (effect of age:  $F(1,13) = 1.72$ ,  $p = .212$ ,  $\eta_p^2 = .117$ ) and was not modulated by orientation of the faces (interaction of age x orientation:  $F(1,13) = 2.02$ ,  $p = .179$ ,  $\eta_p^2 = .135$ ).

#### Configural sets

Orientation of the stimulus did not affect accuracy levels in configurally transformed faces (effect of orientation:  $F(1,13) = .20$ ,  $p = .666$ ,  $\eta_p^2 = .015$ ). Again, as in the featural trials, the performance did not increase with age (effect of age:  $F(1,13) = 1.21$ ,  $p = .294$ ,  $\eta_p^2 = .084$ ), and children with DS did not show an emergence of inversion with increased age (interaction of age x orientation:  $F(1,13) = .07$ ,  $p = .802$ ,  $\eta_p^2 = .105$ ).

#### Comparison to TD trajectory

Direct comparison against the TD group, revealed that the DS group exhibited a significantly worse performance (illustrated in Figure 6.5) and were less affected by stimuli orientation regardless of age (interaction of orientation x group:  $F(1,36) = 10.11$ ,  $p = .003$ ,  $\eta_p^2 = .219$ ). Separate analysis of configurally transformed trials revealed that there was a 3-way interaction of orientation x age x group ( $F(1,36) = 14.97$ ,  $p < .001$ ,  $\eta_p^2 = .294$ ), but similar performance between the TD and DS groups on featurally transformed faces was observed (interaction of orientation x age x group:  $F(1,36) = .90$ ,  $p = .349$ ,  $\eta_p^2 = .024$ ). The groups had different rates of performance with increasing age with DS group being significantly worse (effect of age:  $F(1,36) = 18.62$ ,  $p < .001$ ,  $\eta_p^2 = .341$ ). In sum, children in DS group performed significantly less accurate than the TD

group. Furthermore, large variability in DS group was apparent, and could have had masking effect of a slow emergence of inversion effect in configural or/and featural trials (task x orientation x age x group:  $F(1,36) = 12.87$ ,  $p < .001$ ,  $\eta_p^2 = .263$ ).

#### 6.2.2.1.4 WBS group

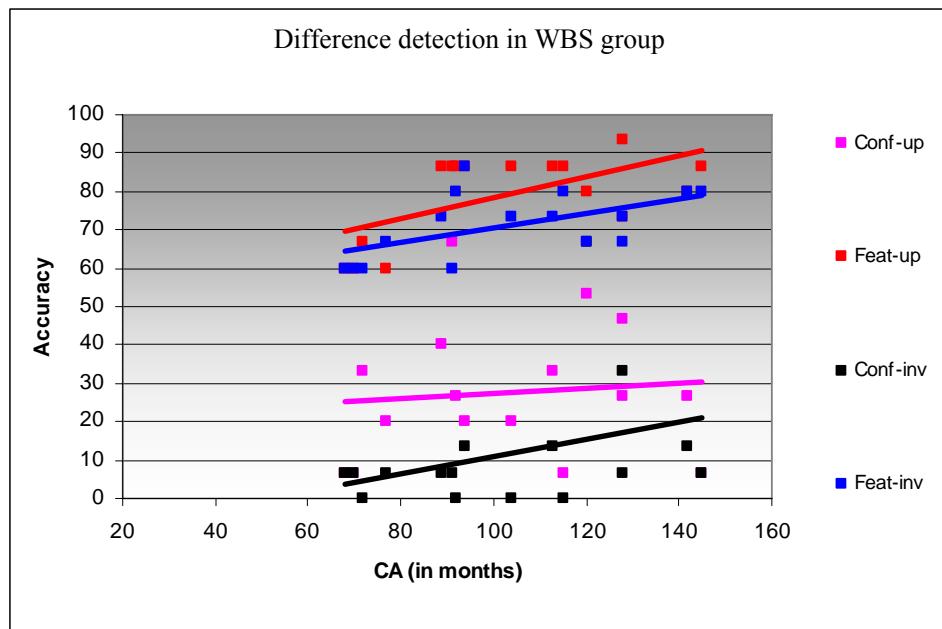


Figure 6.6: WBS group developmental trajectory for accuracy scores on the Jane faces.

As seen from Figure 6.6, transformation of the faces had a significant effect on the accuracy level in the WBS group (effect of transformation:  $F(1,14) = 5.68$ ,  $p = .032$ ,  $\eta_p^2 = .289$ ), however inverting faces did not have a significant influence on the performance levels (effect of orientation:  $F(1,14) = 1.46$ ,  $p = .247$ ,  $\eta_p^2 = .095$ ). Overall, performance improved with age (effect of age:  $F(1, 14) = 6.07$ ,  $p = .027$ ,  $\eta_p^2 = .302$ ) regardless of orientation and transformation (interaction of transformation x orientation x age:  $F(1,14) = 2.94$ ,  $p = .108$ ,  $\eta_p^2 = .174$ ).

#### Featural sets

Performance on featural trials was not influenced by orientation (effect of orientation:  $F(1,14) = .04$ ,  $p = .953$ ,  $\eta_p^2 = .001$ ), and increased with age (effect of age:  $F(1,14) = 9.23$ ,  $p = .009$ ,  $\eta_p^2 = .397$ ), but there was no interaction of age x orientation: ( $F(1,14) = .89$ ,  $p = .362$ ,  $\eta_p^2 = .060$ ).

### Configural sets

As in the featural trials, orientation had no influence on the accuracy level (effect of orientation:  $F(1,14) = 2.83$ ,  $p = .104$ ,  $\eta_p^2 = .169$ ), however the performance was poor and did not improve with increased age (effect of age:  $F(1,14) = .92$ ,  $p = .354$ ,  $\eta_p^2 = .062$ ).

### Comparison to TD trajectory

Direct comparison against the normal developmental trajectory revealed that children in the WBS group were not affected by seeing faces in different orientations as the TD group was (3-way interaction of orientation x age x group  $F(1,37) = 15.92$ ,  $p < .001$ ,  $\eta_p^2 = .301$ ). Separate analysis of conditions showed that configurally transformed faces were more difficult to recognise for the WBS group showing no improvement with age and no sign of an inversion effect (3-way interaction of orientation x age x group:  $F(1,37) = 25.55$ ,  $p < .001$ ,  $\eta_p^2 = .409$ ). On the featural trials, WBS group showed similar treatment of faces with increased age (interaction of orientation x age x group  $F(1,37) = .09$ ,  $p = .769$ ,  $\eta_p^2 = .002$ ). In summary, the WBS group exhibited an atypical profile of performance in comparison to the TD group, whereby lack of inversion effect and poor performance on upright configural trials was evident (task x orientation x age x group:  $(F(1,37) = 27.06$ ,  $p < .001$ ,  $\eta_p^2 = .422$ ).

#### *6.2.2.1.5 Intra- and inter-disorder comparisons*

##### Intra-disorder comparison: autism

Overall, there was no main effect of group ( $F(1,29) = .46$ ,  $p = .504$ ,  $\eta_p^2 = .016$ ), however children in the LFA group appeared to be significantly less accurate than the HFA group with increased age (interaction of age x group:  $F(1,29) = 4.92$ ,  $p = .035$ ,  $\eta_p^2 = .145$ ). The groups exhibited a differential effect of inversion on featurally transformed faces (interaction of orientation x group x age:  $F(1,29) = 4.67$ ,  $p = .039$ ,  $\eta_p^2 = .139$ ), with the LFA group showing an inversion effect.

### Inter-disorder comparison: HFA and WBS

Overall, there was no group difference (main effect of group:  $F(1,28) = 1.16$ ,  $p = .292$ ,  $\eta_p^2 = .040$ ), but the WBS group showed a slower rate of improvement with age (effect of age x group:  $F(1,28) = 427$ ,  $p = .048$ ,  $\eta_p^2 = .132$ ). Separate analysis of conditions revealed similar pattern of development on featural sets ( $F(1,28) = .06$ ,  $p = .813$ ,  $\eta_p^2 = .002$ ), however the HFA group showed faster rate of development on configural trials (interaction of age x group:  $F(1,28) = 4.71$ ,  $p = .039$ ,  $\eta_p^2 = .144$ ).

#### **6.2.2.3 Summary of difference detection accuracy**

Notably in terms of delay, in the configural condition only HFA group showed delayed onset <sup>(3)</sup> and borderline delayed rate <sup>(4)</sup> ( $p = .070$ ) and none of the disorder groups showed either type of delay. Why is this? Due to the nature of the task, children in the TD group get worse with age on inverted configural trials (see Figure 6.1 & 6.2). If disorder groups are not doing well on a task for other reasons, it may cause overlap of data points and wash out group differences. Table 6.3 depicts summary of each group profile on the difference detection conditions of Jane faces task.

Table 6.3: Summary of each group's performance on Jane task plotted against CA.

GROUP	NORMAL PROFILE *	BETTER ACCURACY	INVERSION	
			FEATURAL	CONFIGURAL
TD	YES	FEATURAL	NO	YES FROM 5:08
HFA	NO	FEATURAL	NO	NO
LFA	NO	FEATURAL	YES (REVERSE AT YOUNG AGE)	NO
DS	NO	FEATURAL	NO	NO
WBS	NO	FEATURAL	NO	NO

\*Normality of the performance for the TD is marked by a 3-way interaction of transformation x orientation x age for the TD group. For the disorder groups, normality is therefore a NS 4-way interaction of orientation x transformation x age x group in comparison to the TD group.

<sup>3</sup> Significant main effect of group comparison between TD and HFA

<sup>4</sup> Significant effect of age x group comparison between TD and HFA

#### **6.2.2.4 Can the Benton test predict accuracy on the Jane task?**

In order to establish whether Benton test could predict accuracy on difference detection in the Jane faces task, the CA scores were replaced with Benton age equivalent scores (see section 4.5.2.1), (equivalent to MA matches). If controlling for test age normalises performance one would expect main effects of group and interactions including group to become non-significant when the disorder group is compared to the TD group.

Overall, none of the disorder groups showed the normal pattern of performance when their abilities level on the Benton task was controlled for (see Table 3, in Appendix C). Similarly, when featural and configural conditions were analysed separately, an atypical pattern of performance on configural trials was observed in all disorder groups. This finding is illustrated in Figure 6.7. A summary of the results can be found in Appendix C (Table 2-4).

Table 6.4: Summary of each groups' performance on Jane task plotted against Benton test.

GROUP	NORMAL PROFILE *	BETTER ACCURACY	INVERSION	
			FEATURAL	CONFIGURAL
TD	----	----	----	----
HFA	NO	FEATURAL	NO	NO
LFA	NO	FEATURAL	YES	NO
DS	NO	FEATURAL	NO	NO
WBS	NO	FEATURAL	NO	NO

\*Normality of the performance for the disorder groups is expressed as a NS 4-way interaction of orientation x transformation x age x group in comparison to the TD group. Note that Benton scores were derived from the TD group hence it is not included in results.

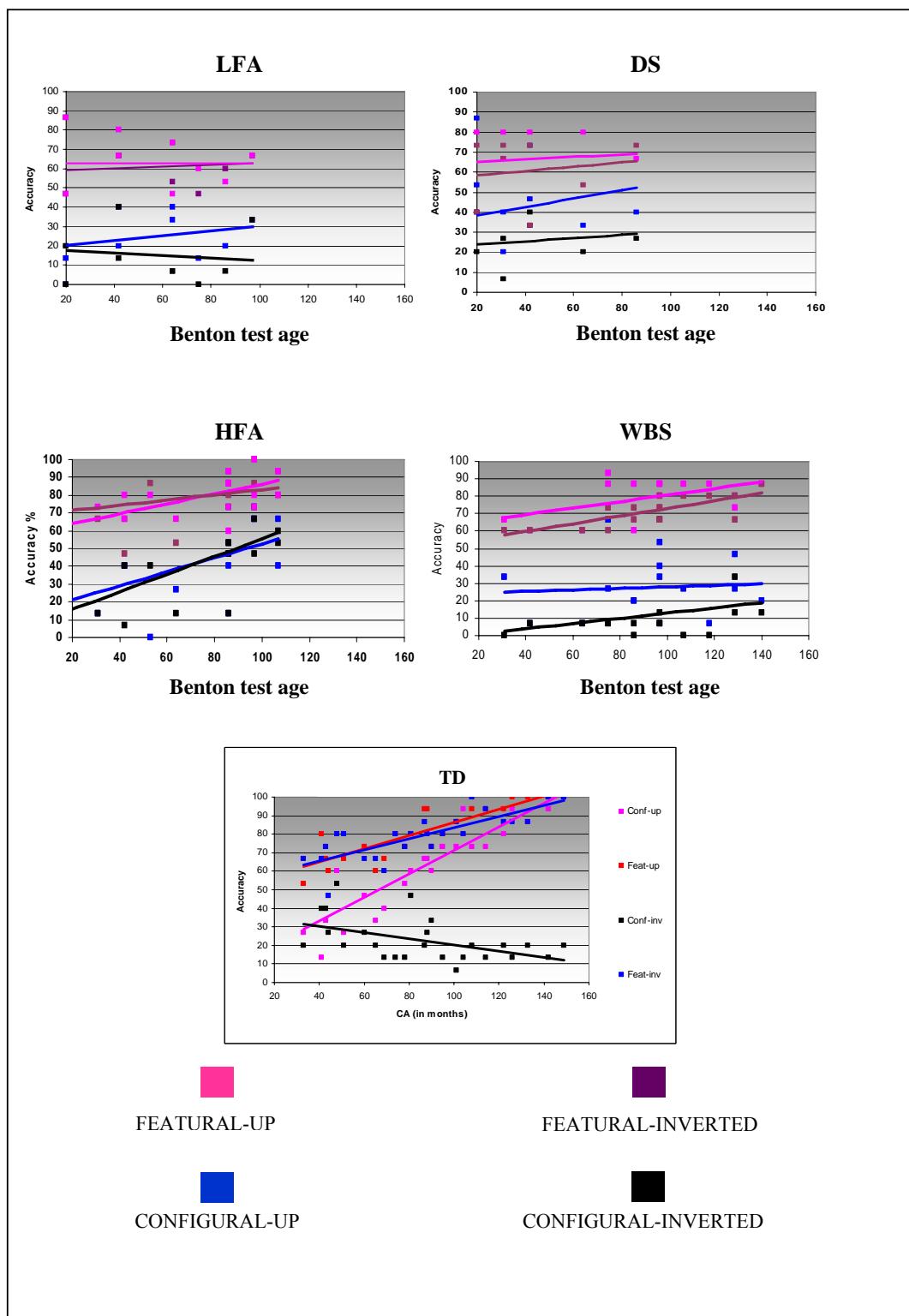


Figure 6.7: The relationship between Benton face recognition test age and performance. Note that TD was not included as Benton test scores were standardised on this group. No group demonstrated the normal profile.

### 6.2.3 Discussion

The main aim of this study was to establish whether children in the disorder groups develop sensitivity to configural changes at a comparable level to the typically developing children. Separate results were obtained for identity and transformation block. In the identity trials, children in the TD group and disorder groups did not show any sign of inversion, suggesting the use of a matching strategy rather than face-specific strategies.

In the case of typically developing group, the findings from this study are consistent with previous data (e.g., Brace et al., 2001; Freire & Lee, 2001; Mondloch et al., 2002) showing that face processing abilities improve with chronological age and inversion has a negative effect on recognition of configurally transformed faces. In line with current predictions, recognition of featurally altered faces improved very quickly with age, and was also constantly better than configurally altered faces (81% vs. 62%, respectively), showing that the development of configural encoding lags behind the development of featural encoding.

As predicted, sensitivity to configural information increased with age and reached an adult-like level at about 12-year-olds, which is consistent with Mondloch's data and with the idea that slow development of configural processing in relation to featural processing is largely due to a longer development of 'expertise' in face processing (Mondloch et al., 2002). However, this expertise can be also acquired in other homogenous objects as shown by Gauthier et al. (1999) with 'greebles' or bird experts (see chapter 1 for further details).

Consistent with current predictions, children showed an early inversion effect on the configural condition at 5:8 years of age, thus demonstrating that upright configurally changed faces are resolved by a configural strategy under the assumption that inversion differentially disrupts configural over featural information. Similarly to the previous studies (Freire et al., 2000; Leder & Bruce, 2000), an inversion effect was not found on featurally changed faces. This, however, is in contrast to Mondloch et al. (2002), who reported a small inversion effect on the featural condition. This discrepancy is possibly

due to procedural differences such as use of simultaneous presentation in the current study along with Freire et al. (2000) and Leder and Bruce (2000) and sequential by Mondloch et al. (2002). A further difference between the studies is that the identity trials were analysed separately to reflect the experimental design.

Similarly to the study on development of holistic face recognition described in the previous chapter, the disorder groups diverged from each other and typically developing controls in many important ways, and are discussed in turn below. As predicted, children with HFA were less sensitive to configurally changed faces, and were not affected by orientation of the stimulus showing lack of inversion effect (e.g., McPartland, et al., 2004). However, they showed similar developmental trajectories to the TD group when using featural information in face recognition. There were large intra-disorder differences in the autism population, with the HFA group improving significantly more accurate with increased age than the LFA group. Perhaps the most surprising result, giving good performance on pattern construction (see chapter 4 for results) is that children in the LFA group performed poorly not only on configural condition (average accuracy 20%) but also on featural condition (62%) in comparison to the HFA and TD group. An atypical performance was observed on the inverted trials where younger children showed better performance on inverted trials then decreases at around 9 years of age and accuracy on upright trials increase, similar performance was found previously by Hobson and colleagues (1988). It is plausible that upright faces give an aversive response to children with LFA but with age, as the frequency of seeing faces increases (children become enrolled full-time at school, play-schemes, clubs), they become more able to deal with face stimuli, albeit not proficiently. Overall, performance of both autism groups was atypical in different ways, even when their performance was matched on their Benton face recognition level.

In line with current predictions, children with Down syndrome showed poorer performance on the task. Also, large group variability could have had a masking effect on their configural and featural performance. Benton test did not predict the normal pattern of performance, however Figure 6.7 suggests the slow emergence of an inversion effect. This finding, along with previous reports, indicates overall poor face recognition abilities (Kaiser et al., 2005; Wishart & Pitcairn, 2000).

As predicted, children in the WBS group showed poor sensitivity to configural information in upright orientations and did not demonstrate an inversion effect with increased age. These findings are consistent with earlier reports in the literature that people with WBS process faces abnormally, relying on face-features rather than face-configuration to recognise faces (Deruelle et al., 1999; Karmiloff-Smith, 1997; Karmiloff-Smith et al., 2004).

Current results are also in line with other visuo-spatial tasks. For instance, Bellugi and colleagues argued that instead of attending to both configural and featural aspects of a spatial display, people with WBS attend exclusively to the individual features, and ignore the overall configuration (e.g., Bellugi et al., 1992), echoing the proposals made about atypical face processing (e.g., Karmiloff-Smith, 1997; Karmiloff-Smith et al., 2004). However, this study contradicts Tager-Flusber and colleagues' (2003) findings. These authors claim that face recognition skills in individuals with WBS develop normally, though this study did not use a direct configural manipulation. It is noted here that these authors did not provide any direct evidence on developmental aspects of face recognition skills.

Current findings show that participants in the clinical groups recognise faces by using different strategies compared the typically developing (except for the DS group). Moreover, these groups show different profiles from each other, suggesting different developmental constraints. Importantly, this also holds with respect to their level of face recognition performance on the Benton test, which results suggest involvement of atypical processes on Jane task. Even when Benton scores fell within the normal range (e.g., for the WBS group), our trajectories showed that performance of disorder groups was nevertheless atypical, meaning that even ‘scores-in-the-normal-range’ do not imply typical development. Due to the fact that several studies reported poor memory for faces or other objects in developmental disorders (e.g., de Gelder, et al., 1991) stimuli were presented simultaneously to reduce memory load and task difficulty level (see Farran, et al., 2003, for discussion on task difficulty) and so performance in the current study should be at their highest level of abilities. In summary, the clinical groups showed atypical development of face recognition abilities. They were less accurate than the TD controls on configurally transformed faces and showed lack of inversion effect, thus

lack of development of face expertise skills. In the next study, memory component is included to further investigate its role on configural face recognition.

### **6.3 STUDY 3: STORY-SUPPORTED FACE RECOGNITION TASK**

The purpose of this study was to examine the face recognition abilities of children in our disorder groups using a child friendly procedure. Contextual support and a small number of trials enabled Brace and colleagues (2001) to successfully test children as young as 2:8 years old and explore the age at which an inversion effect first emerges. Recently, Karmiloff-Smith et al. (2004) also used this task with individuals with WBS. The specific investigations and predictions are described below:

- 1) Do children with developmental disorders show progressive developmental emergence of the inversion effect?

**Predictions:**

- i) Children in the TD group will show increased sensitivity to inversion (e.g., Brace et al., 2001; Mondloch et al., 2002);
- ii) Children in the disorder groups will not be affected by the inversion of the face stimuli (e.g., Karmiloff-Smith et al., 2004; McPartland et al., 2004).

- 2) Does the performance of the disorder groups improve with age?

**Predictions:**

- i) Children in the TD group will show increased accuracy and faster response with age for upright faces (e.g., Brace et al., 2001; Mondloch et al., 2002);
- ii) Children in the disorder groups will be significantly less accurate and slower than the TD group on the upright faces (e.g., Karmiloff-Smith et al., 2004).

- 3) Can performance on the Benton Face recognition test predict level of performance on the story-supported face recognition task?

**Prediction:** If the face processing is atypical then it may not be predicted by developmental age on the Benton test.

### 6.3.1 Method

#### 6.3.1.1 Participants

Two children with WBS and two children with LFA failed to correctly identify either a single upright face or a single inverted face and were excluded from subsequent analyses. The 16 remaining participants with WBS had a mean CA of 8:6 (SD: 2:7; range: 5:7 - 12:8). The 15 remaining children in the LFA group had a mean CA of 8:8 (SD: 1:8; range: 6:0 - 11:3). One child in DS group initially refused to participate in the study, thus the testing session was rescheduled for another day.

The control data from a previous study by Brace et al. (2001) were made available for the current study. The TD group consisted of 111 children with a mean CA of 8:1 (SD: 2:7, range: 2:8 - 11:5). There were 12 children between 2:8 - 4:4, 20 children between 5;2-6;11, 26 children between 7:2 - 8:8; 28 children between 9:2 - 10:2, and 25 children between 11:1 - 11:5. This age range enabled us to build a large trajectory of typical developmental changes on this task. Details of other groups' participants are detailed in chapter 3, section 3.2.1.

#### 6.3.1.2 Stimuli

The stimuli were kindly provided by Nicola Brace at the Open University, UK and were modified for use with a touch-screen monitor using Superlab 2.0. The stimuli consisted of two parts: a Storybook and a computer game. The Storybook was a hand-painted story about two boys, called Jamie and Tom. One of the boys is kidnapped by a witch and taken to her castle. The witch turns the boy into a variety of objects (such as a bear) that retain only the boy's face, and she then hides him in amongst 8 other boys that she has kidnapped. The only way for the other boy to rescue his friend is to play a game of hide and seek in order to spot his friend in amongst the other boys/objects, which are either the correct way up or hung upside-down. In the first two pages of the book, pictures of Jamie and Tom are present, whereas in the next five pages the story continues without any pictures of the boys, to ensure that subsequent recognition of the faces is delayed by about three minutes. The hide and seek computer game includes upright and inverted pictures of one of the two target faces (Jamie and Tom) among 8

distracter faces. Two versions of the task were run with different target faces. Each participant saw one of the two versions (for further details, see Brace et al., 2001). Figure 6.8 illustrates a sample of a stimulus used in the study.

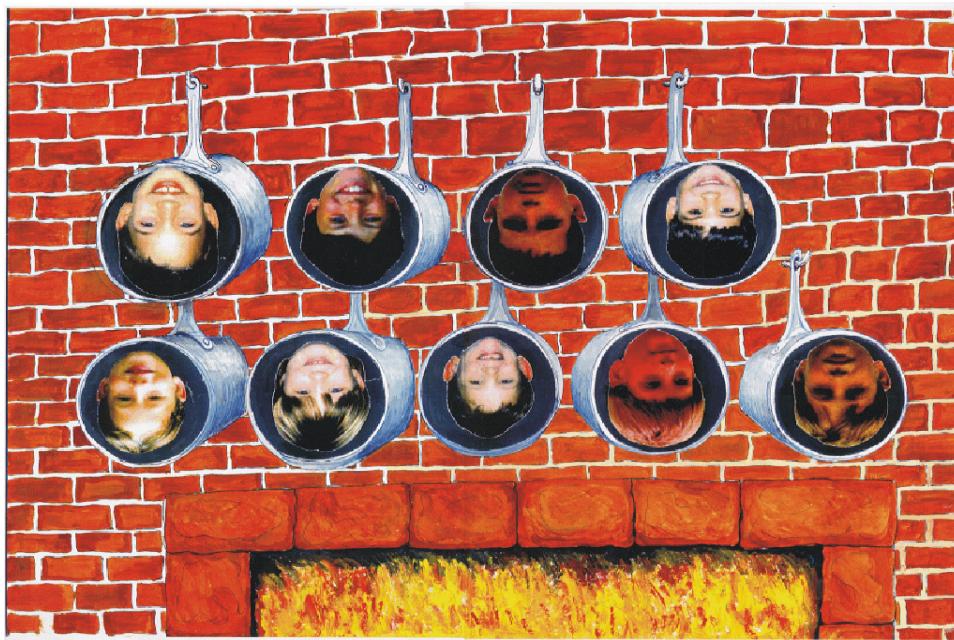


Figure 6.8: Example of the stimulus used in the study. Children were asked to find Jamie/Tom and touch the screen as quick as possible. Stimuli provided by Brace et al. (2001).

### 6.3.1.3 Procedure

In the first part of the study the experimenter (or the participant if he/she read easily) read the story aloud, during which the participant was asked to point to the pictures of the two boys called Jamie and Tom, and to repeat their names after the experimenter. On completion of the first part of the story, the participant was asked if he/she would like to play a computer game of finding the lost boy (Jamie and Tom). Eight trials were run, including two practice trials. For each trial, a picture was presented on a touch-screen with the target face hidden amongst 8 distracter faces of varying similarity to the target face. The position and orientation of the target face within the array of 9 faces was systematically varied. Once the detection game was completed, the story reading was continued to achieve a happy ending.

### 6.3.2 Results

Trajectories were analysed using an analysis of co-variance (ANCOVA). This test requires the relationship between performance and age to be roughly linear. The accuracy rates and median RT on upright and inverted faces (each out of 3) were compared to chronological age<sup>(5)</sup>. Since there were 8 distractors per trial, chance performance in this face identification task was 11%.

#### 6.3.1.4.1 TD control group

##### Accuracy

As depicted in Figure 6.9, the TD group exhibited a significant improvement in accuracy with age ( $F(1,109) = 12.40$ ,  $p < .001$ ,  $\eta_p^2 = .102$ ) and orientation had no influence on accuracy performance (main effect of orientation:  $F(1,109) = 1.19$ ,  $p = .278$ ,  $\eta_p^2 = .011$ ). The performance accuracy on inverted trials did not alter significantly across the developmental profile (interaction of age x orientation:  $F(1,109) = .01$ ,  $p = .915$ ,  $\eta_p^2 = .001$ ). A low number of trials possibly prevented to demonstrate differences on accuracy in different orientations.

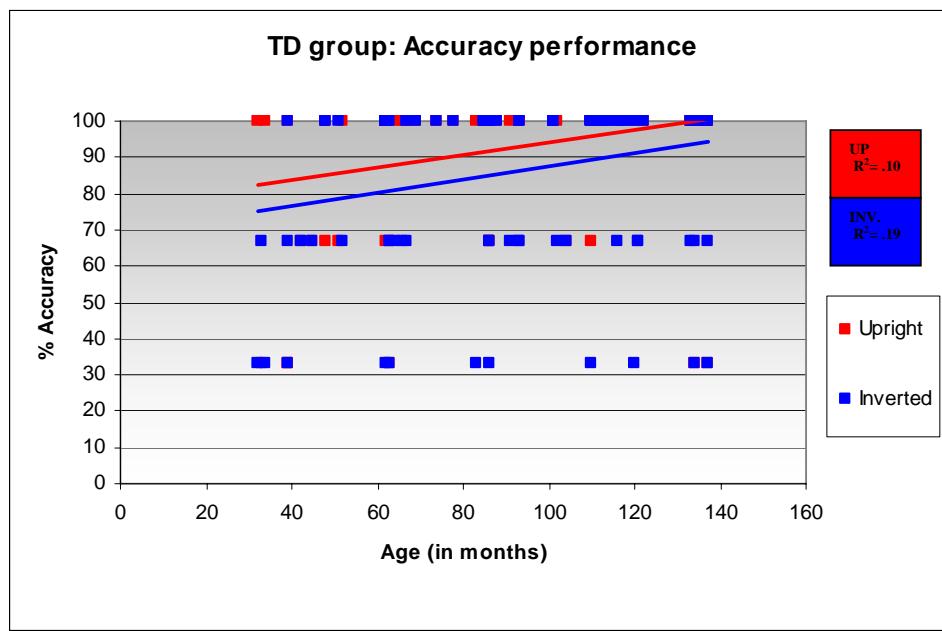


Figure 6.9: TD group developmental trajectory for accuracy scores on the Storybook. Data provided by Brace et al. (2001).

<sup>5</sup> In general, all groups displayed larger variability in accuracy than RT data due to the lower sensitivity of the accuracy measure.

### Reaction Time

Typically developing children exhibited a significant reduction in reaction time with age (main effect of age:  $F(1,109) = 38.50$ ,  $p < .001$ ,  $\eta_p^2 = .026$ ) and a significant time cost of recognising inverted faces (main effect of orientation:  $(F(1,109)=4.50$ ,  $p=.036$ ,  $\eta_p^2 = .040$ ). Moreover, the cost of recognising inverted faces significantly increased with age, consistent with the emergence of configural faces processing expertise (interaction of orientation x age:  $F(1,109) = 5.93$ ,  $p = .016$ ,  $\eta_p^2 = .005$ ). This finding is illustrated in Figure 6.10.

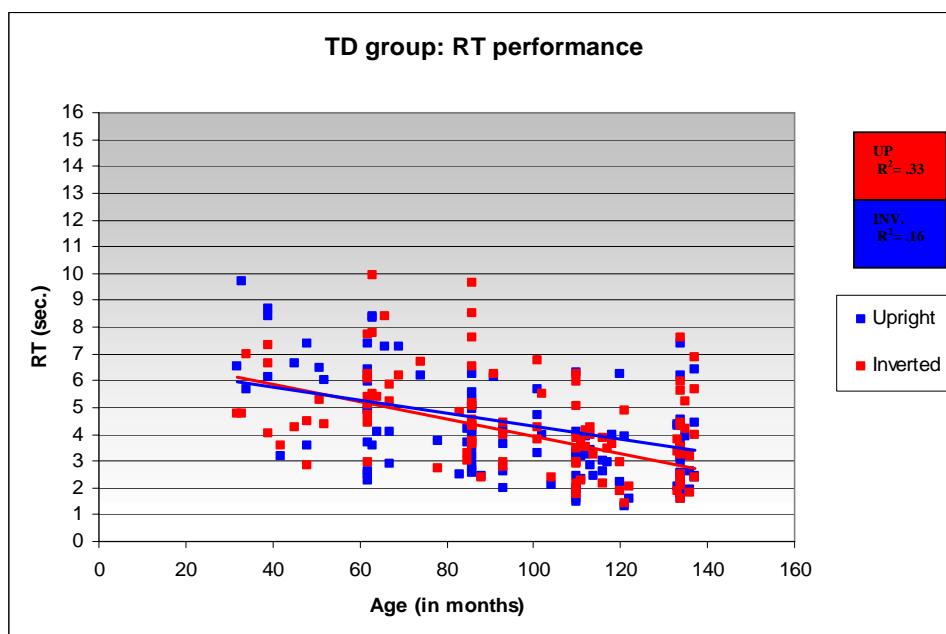


Figure 6.10: TD group developmental trajectory for RT on the Storybook task.

#### *6.3.1.4.2 HFA group*

### Accuracy

Overall accuracy scores did not significantly improve with age (main effect of age:  $F(1,14) = 2.84$ ,  $p = .114$ ,  $\eta_p^2 = .017$ ), but when analysed separately it was shown that performance accuracy increased with age on the inverted trials ( $F(1, 14) = 15.30$ ,  $p = .002$ ,  $\eta_p^2 = .022$ ). Performance accuracy was not negatively affected by the presentation

of inverted faces (main effect of orientation:  $F(1,14) = 1.09$ ,  $p = .314$ ,  $\eta_p^2 = .07$ ). Moreover, this effect did not change with increased age, indicating lack of emergence of configural faces processing expertise (interaction of orientation x age:  $F(1,14) = .75$ ,  $p = .402$ ,  $\eta_p^2 = .051$ ). This finding is illustrated in Figure 6.11.

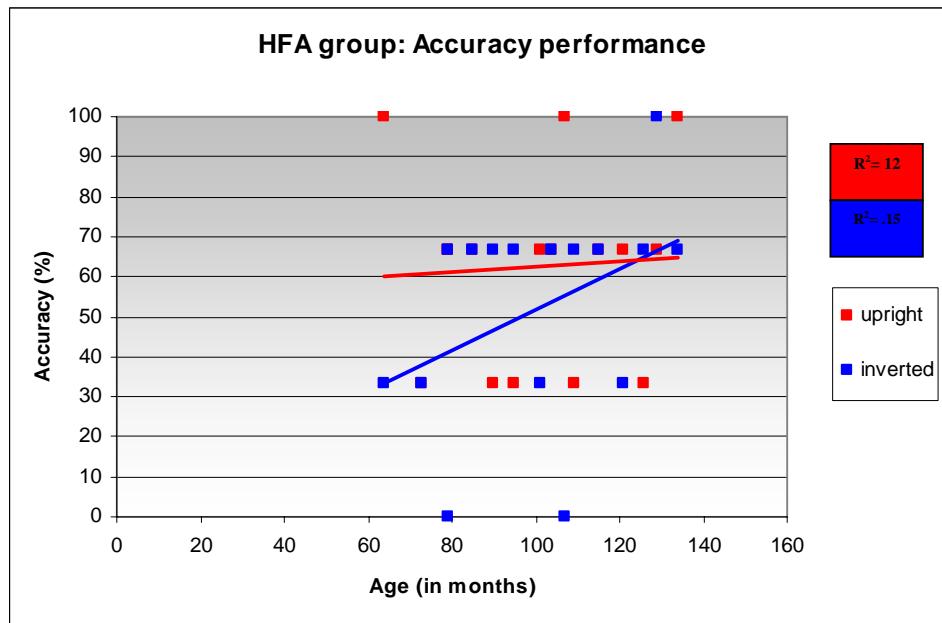


Figure 6.11: HFA group developmental trajectory for accuracy scores on the Storybook.

#### Reaction Time

As shown in Figure 6.12, the HFA group, response time did not improve with increased age ( $F(1,14) = .01$ ,  $p = .948$ ,  $\eta_p^2 = .001$ ). There was no indication of an inversion cost ( $F(1,14) = 1.59$ ,  $p = .232$ ,  $\eta_p^2 = .117$ ) and orientation of the faces did not have any influence across age (interaction of orientation x age:  $F(1,14) = 1.45$ ,  $p = .252$ ,  $\eta_p^2 = .108$ ).

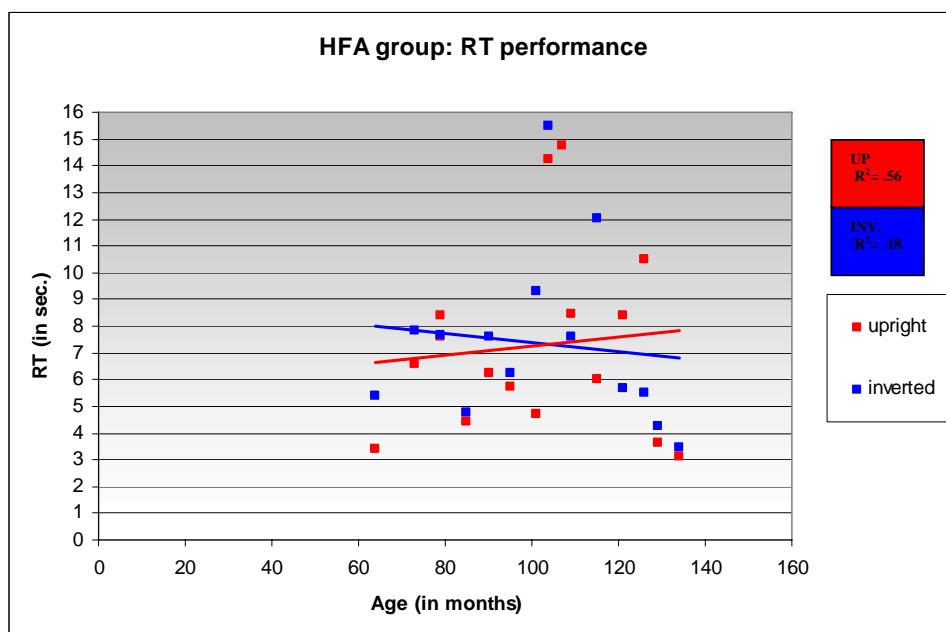


Figure 6.12: HFA group developmental trajectory for RT on the Storybook task.

#### Comparison to TD trajectory

A direct comparison revealed an overall significant group difference in accuracy (delayed onset),  $F(1,123) = 4.80$ ,  $p = .030$ ,  $\eta_p^2 = .038$  but not in response time ( $F(1,123) = .01$ ,  $p = .994$ ,  $\eta_p^2 = .001$ ). Both groups showed a similar rate of development in accuracy (interaction of group x age:  $F(1,123) = .32$ ,  $p = .572$ ,  $\eta_p^2 = .003$ ) and in RT (interaction of group x age:  $F(1,123) = 1.74$ ,  $p = .190$ ,  $\eta_p^2 = .014$ ). Overall, both groups displayed a similar profile of performance on accuracy data (3-way interaction of orientation x age x group:  $F(1,123) = 2.03$ ,  $p = .157$ ,  $\eta_p^2 = .016$ ). However, a significant difference between the groups was revealed in the RT data (interaction of orientation x group x age:  $F(1,123) = 4.17$ ,  $p = .043$ ,  $\eta_p^2 = .033$ ). As depicted in Figure 6.12, the HFA group showed an atypical developmental profile as they did not show an inversion effect on accuracy and RT trials and showed a delayed onset in accuracy performance.

### 6.3.1.4.3 LFA group

#### Accuracy

As illustrated in Figure 6.13, accuracy scores of children in the LFA group did not increase with age (main effect of age:  $F(1,13) = .71$ ,  $p = .421$ ,  $\eta_p^2 = .067$ ). While the LFA group performance appeared to be better on the inverted trials (inverted: 54% and upright: 36%) there was no main effect of orientation ( $F(1,13) = 2.57$ ,  $p = .140$ ,  $\eta_p^2 = .204$ ) nor interaction between orientation x age: ( $F(1,13) = 1.04$ ,  $p = .333$ ,  $\eta_p^2 = .094$ ). As previously mentioned, the small number of trials and larger variability in the disorder groups possibly masked some of the effects and interactions. Response time data indicate a similar effect of faster recognition on inverted trials. This suggests that differences were not due to the sample size, task sensitivity and level of variability in performance.

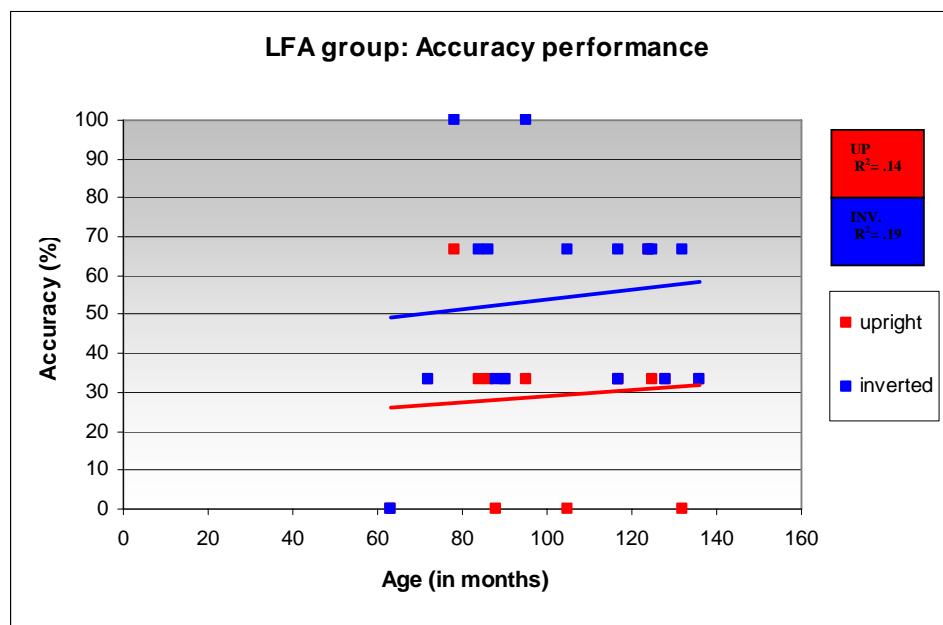


Figure 6.13: LFA group developmental trajectory for accuracy scores on the Storybook.

#### Response Time

Overall, there was no significant reduction in response time with increased age (main effect of age: ( $F(1,13) = 2.19$ ,  $p = .169$ ,  $\eta_p^2 = .180$ ). A surprising pattern of faster face recognition in inverted orientation was evident (main effect of orientation: ( $F(1,13) =$

$4.83$ ,  $p = .053$ ,  $\eta_p^2 = .325$ ). Moreover, the speed of recognising inverted faces significantly decreased with age, which is inconsistent with the emergence of configural faces processing expertise (interaction of orientation x age:  $F(1,13) = 5.41$ ,  $p = .042$ ,  $\eta_p^2 = .351$ ). Figure 6.14 illustrates these findings.

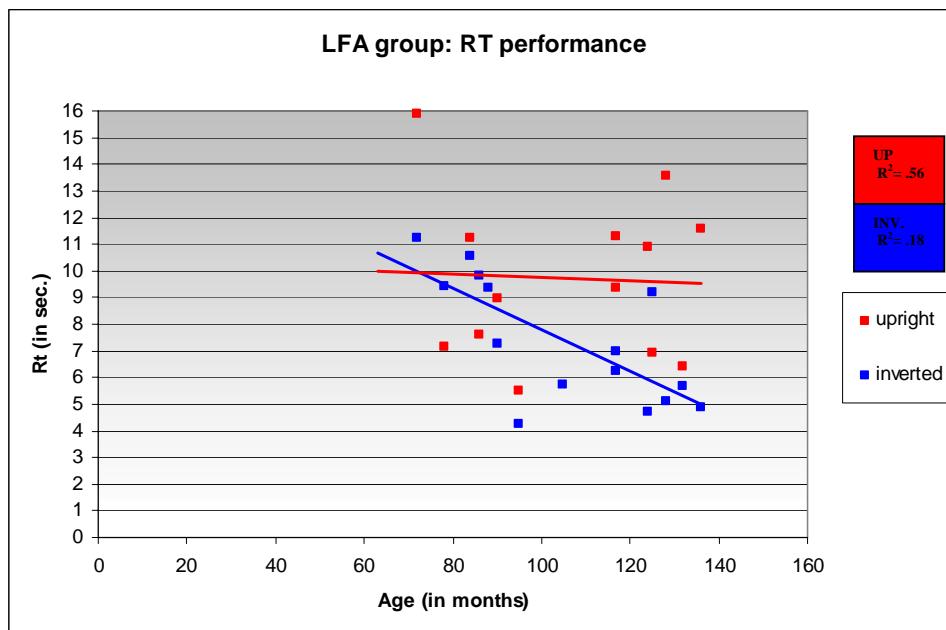


Figure 6.14: LFA group developmental trajectory for RT on the Storybook task.

#### Comparison to TD trajectory

Direct comparison between LFA and TD revealed no overall significant group difference in accuracy ( $F(1,121) = .04$ ,  $p = .852$ ,  $\eta_p^2 = .001$ ), despite overall lower level of accuracy in the LFA group (TD group: 91%; LFA group: 44%) but a significant difference in reaction time was evident ( $F(1,121) = 5.64$ ,  $p = .019$ ,  $\eta_p^2 = .045$ ). Both groups showed a similar rate of development in accuracy (interaction of group x age:  $F(1,121) = 2.72$ ,  $p = .102$ ,  $\eta_p^2 = .022$ ) and in RT (interaction of group x age:  $F(1,121) = .06$ ,  $p = .808$ ,  $\eta_p^2 = .001$ ). Children in the LFA group displayed a similar profile of performance when accuracy performance was considered (3-way interaction of orientation x age x group:  $F(1,121) = 1.15$ ,  $p = .285$ ,  $\eta_p^2 = .010$ ), but not on the RT data (interaction of orientation x group x age:  $F(1,121) = 17.74$ ,  $p < .001$ ,  $\eta_p^2 = .130$ ). Results from the current analysis point out that RT data are more sensitive measure of face recognition abilities. In summary, the LFA group showed atypical profile in a

number of ways, i) faster recognition of inverted faces, ii) increased accuracy with age on inverted faces, and iii) an overall slower RT.

#### 6.3.1.4.4 DS group

##### Accuracy

As illustrated in Figure 6.15, participants in the DS group displayed a significant increase in accuracy with age ( $F(1,13) = 4.71$ ,  $p = .049$ ,  $\eta_p^2 = .266$ ). However, orientation of the stimuli had no influence on their performance ( $F(1,13) = 2.39$ ,  $p = .146$ ,  $\eta_p^2 = .055$ ), and did not change with increased age ( $F(1,13) = .89$ ,  $p = .364$ ,  $\eta_p^2 = .064$ ).

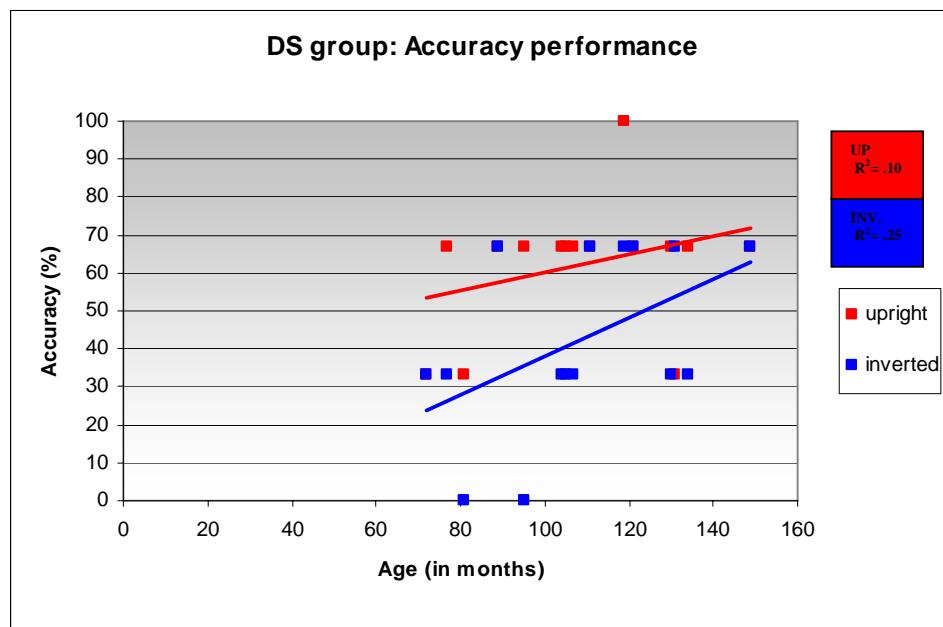


Figure 6.15: DS group developmental trajectory for accuracy scores on the Storybook.

##### Response time

Children in the DS group exhibited a significant reduction in response time with age (main effect of age:  $F(1,13) = 8.15$ ,  $p = .014$ ,  $\eta_p^2 = .385$ ), but there was no sign of time cost of recognising inverted faces (main effect of orientation:  $F(1,13) = .14$ ,  $p = .717$ ,  $\eta_p^2 = .010$ ). Moreover, there was no indication of RT cost in recognising inverted faces

with increased age, hence the lack of emergence of configural faces processing expertise (interaction of orientation x age:  $F(1,13) = .10$ ,  $p = .753$ ,  $\eta_p^2 = .008$ ). This finding is illustrated in Figure 6.16.

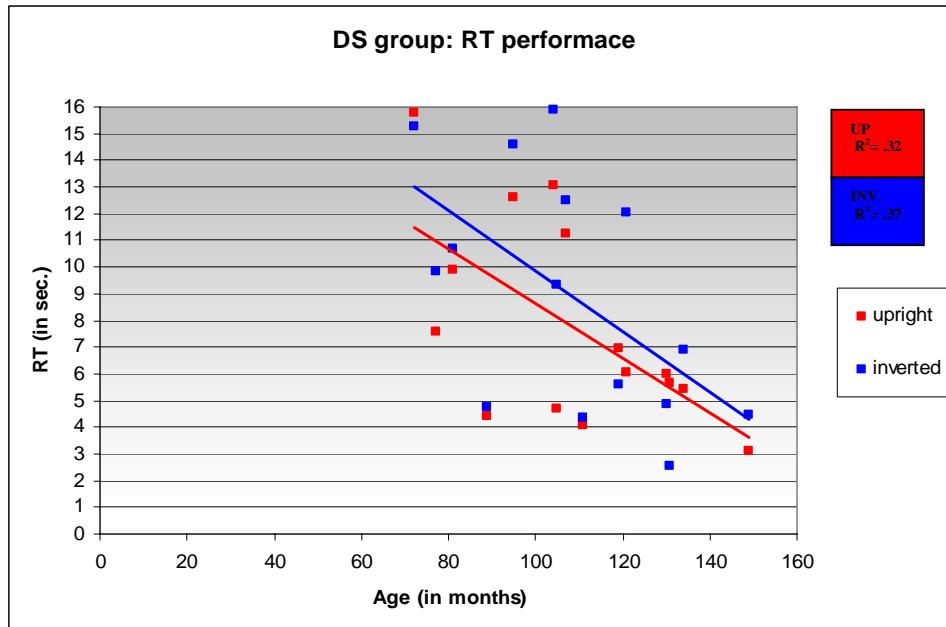


Figure 6.16: DS group developmental trajectory for RT on the Storybook task.

#### Comparison to TD trajectory

There were significant differences between the groups on accuracy trials ( $F(1,122) = 8.37$ ,  $p = .005$ ,  $\eta_p^2 = .004$ ) and in response time ( $F(1,122) = 30.99$ ,  $p < .001$ ,  $\eta_p^2 = .203$ ), suggesting a later developmental onset of performance in the DS group. The groups showed a similar rate of development in accuracy (interaction of group x age:  $F(1,122) = 1.08$ ,  $p = .301$ ,  $\eta_p^2 = .029$ ) but not in RT (interaction of group x age:  $F(1,122) = 13.76$ ,  $p < .001$ ,  $\eta_p^2 = .101$ ). In summary, the DS group showed a similar developmental profile in comparison to the TD group on their accuracy (3-way interaction of orientation x age x group:  $F(1,122) = 1.05$ ,  $p = .307$ ,  $\eta_p^2 = .010$ ), and the RT data (interaction of orientation x group x age:  $F(1,122) = 1.04$ ,  $p = .310$ ,  $\eta_p^2 = .008$ ), although their developmental onset on accuracy and RT was delayed. Furthermore, there was no emergence of inversion effect.

### 6.3.1.4.5 WBS group

#### Accuracy

Children in WBS group exhibited increased accuracy scores with age (main effect of age:  $F(1,13) = 19.04$ ,  $p < .001$ ,  $\eta_p^2 = .594$ ). However, there was no indication that orientation had influenced the performance accuracy (main effect of orientation:  $F(1,13) = .88$ ,  $p = .365$ ,  $\eta_p^2 = .064$ ), and no sign that the inversion effect emerged across chronological age (interaction of orientation x age:  $F(1,13) = .55$ ,  $p = .471$ ,  $\eta_p^2 = .041$ ). This finding is illustrated in Figure 6.17.

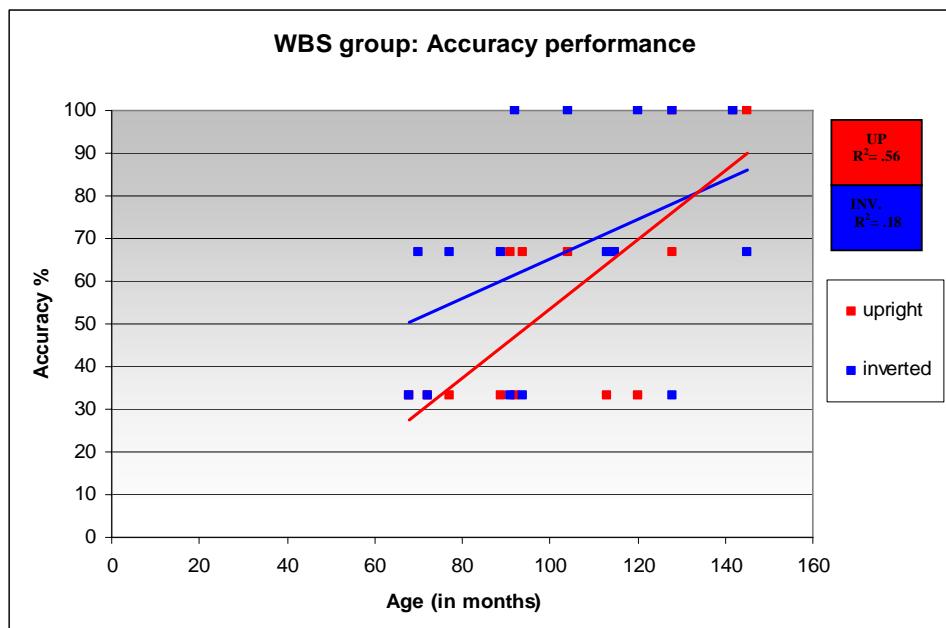


Figure 6.17: WBS group developmental trajectory for accuracy scores on the Storybook.

#### Response Time

As shown in Figure 6.18, the WBS group revealed a significant decrease in response time with age ( $F(1,13) = 28.44$ ,  $p < .001$ ,  $\eta_p^2 = .686$ ). Also, inversion had no effect on response time performance ( $F(1,13) = .01$ ,  $p = .921$ ,  $\eta_p^2 = .001$ ). Moreover, it did not change across development (interaction of orientation x age:  $F(1,13) = .01$ ,  $p = .993$ ,  $\eta_p^2 = .101$ ).

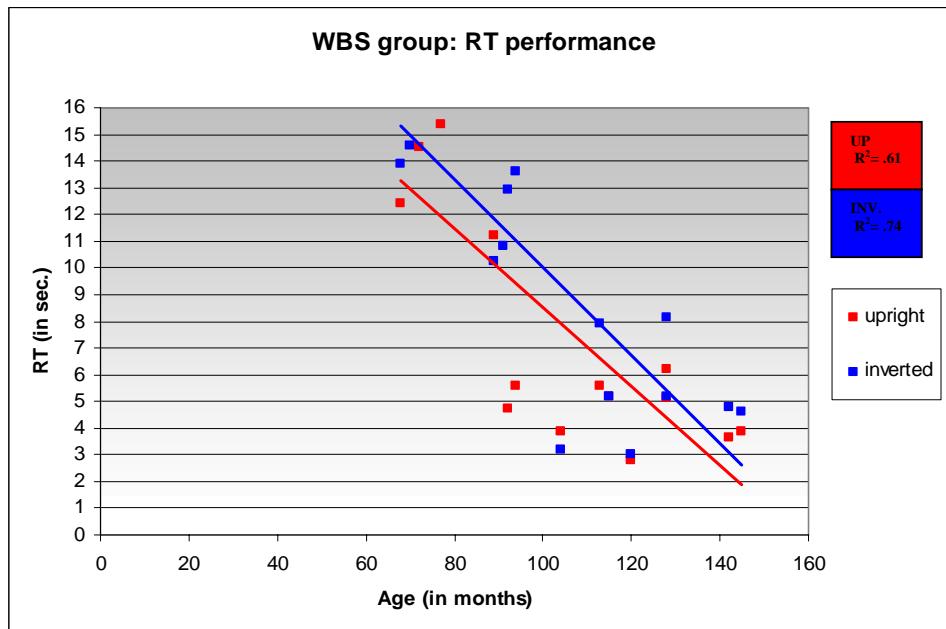


Figure 6.18: WBS group developmental trajectory for RT on the Storybook task.

#### Comparison to TD trajectory

Direct comparison between groups revealed an overall significant difference in both accuracy and reaction time: (effect of group on accuracy:  $F(1,122) = 17.88$ ,  $p < .001$ ,  $\eta_p^2 = .128$ ; RT:  $F(1,122) = 85.62$ ,  $p < .001$ ,  $\eta_p^2 = .412$ ). Furthermore, both WBS accuracy scores and RTs improved at a slower rate than the TD group (accuracy:  $F(1,122) = 7.62$ ,  $p = .007$ ,  $\eta_p^2 = .059$ ; RT:  $F(1,122) = 50.03$ ,  $p < .001$ ,  $\eta_p^2 = .291$ ). However, there was no significant difference in 3-way interactions (accuracy:  $F(1,122) = 1.92$ ,  $p = .168$ ,  $\eta_p^2 = .015$ ; RT:  $F(1,122) = 1.26$ ,  $p = .265$ ,  $\eta_p^2 = .010$ ). In summary, WBS group showed a delayed onset and rate of performance on the task when compared to the TD group. Finally there was a lack of inversion in accuracy and RT conditions in the WBS group.

### 6.3.2.1 Summary of results

Summary of each group results obtained in the current study can be seen in the Table 6.5.

Table 6.5: Summary of each group's performance plotted against CA

GROUP	DEVELOPMENTAL PROFILE*		INVERSION	
	ACCURACY	RT	ACCURACY	RT
TD	YES	YES	NO	YES
HFA	DELAYED ONSET	DELAYED ONSET & RATE <b>ATYPICAL</b>	NO	NO
LFA	NS	DELAYED ONSET <b>ATYPICAL</b>	NO	YES (REVERSE)
DS	DELAYED ONSET	DELAYED ONSET & RATE	NO	NO
WBS	DELAYED ONSET & RATE	DELAYED ONSET & RATE	NO	NO

Normality of the performance is expressed as a 2-way interaction of orientation x age for the TD group and for the disorder groups, as a NS 3-way interaction (normal profile = ns 3-way interaction of orientation x age x group in comparison to the TD group).

### 6.3.2.2 Can Benton test predict performance on the Storybook task?

As in the previous studies, performance scores on the Benton test were translated into test ages via our TD group and performance plotted against this instead of CA. It was found that the Benton test age scores did not predict a normal face recognition profile when groups were controlled for face recognition abilities. These findings are summarised in Table 6.6.

For the HFA group, performance on the Benton did not predict accuracy ( $F(1,14) = 1.62$ ,  $p = .447$ ,  $\eta_p^2 = .098$ ) nor did it predict reaction time on the Storybook task ( $F(1,14) = 1.26$ ,  $p = .498$ ,  $\eta_p^2 = .102$ ); in this analysis the inversion effect was non-significant:

$F(1,14) = 3.23, p = .100, \eta_p^2 = .099$ ). In the LFA group, scores on the Benton did not predict their performance on the Storybook task (accuracy:  $F(1,13) = 1.06, p = .337, \eta_p^2 = .094$ ; RT:  $F(1,13) = 2.55, p = .198, \eta_p^2 = .182$ ); inversion effect non-significant:  $F(1,13) = 2.23, p = .141, \eta_p^2 = .209$ ). In the DS group, performance on the Benton test did not predict accuracy ( $F(1,13) = 2.62, p = .147, \eta_p^2 = .156$ ) or RT on the Storybook task ( $F(1,13) = .16, p = .798, \eta_p^2 = .088$ ); inversion effect non-significant:  $F(1,13) = .23, p = .712, \eta_p^2 = .079$ ). Lastly, in the WBS, performance on the Benton did not predict accuracy ( $F(1,13) = .82, p = .347, \eta_p^2 = .067$ ) nor did predict response time on the Storybook task ( $F(1,13) = .26, p = .498, \eta_p^2 = .122$ ); inversion effect non-significant:  $F(1,13) = .53, p = .451, \eta_p^2 = .049$ ).

Table 6.6: Summary of each groups' performance on Storybook task plotted against Benton age test.

GROUP	DEVELOPMENTAL PROFILE*		INVERSION	
	ACCURACY	RT	ACCURACY	RT
TD	----	----	----	----
HFA	NO	NO	NO	NO
LFA	NO	NO	NO	YES
DS	NO	NO	NO	NO
WBS	NO	NO	NO	NO

\*Normality of the performance for the disorder groups is expressed as a NS 3-way interaction of orientation x age x group in comparison to the TD group. Note that Benton scores were derived from the TD group hence it is not included in results.

### 6.3.3 Discussion

The main aim of this study was to establish whether children in the disorder groups show progressive face recognition abilities in a child-friendly task. In this study, reaction times were the more sensitive measure given the small range of accuracy performance available on the six trials. Response time data explained more variability than the accuracy data for all groups (see  $R^2$  values). Overall, the developmental trajectories of disorder groups failed to show the progressive emergence of a face inversion effect. In the case of typically developing group, the findings from this study are consistent with current predictions and previous data (e.g., Diamond & Carey, 1986; Freire et al., 2000), showing that face-processing abilities improve with chronological age and that inversion has a negative effect on recognition of faces. In accordance with previous studies described in this thesis, the disorder groups diverged from the TD group in many ways, and are discussed in turn below.

The performance of children in the autism groups on the RT was atypical, performing worse on Storybook task with no sign of inversion effect. Surprisingly, both groups showed better performance on the inverted trials. This finding suggests that children with autism encode faces using a featural encoding mechanism. Previous studies suggest that individuals with autism perform poorly in recognition memory for faces (e.g., Hobson, 1988) and other stimuli such as cats, motorbikes and horses (Blair, Frith, Abell, & Cipolotti, 2001).

In the case of the DS group, they showed similar developmental profile in comparison to the TD group but had delayed onset on accuracy and delayed onset and rate on RT. The DS group did not show an inversion effect, but whether lack of this effect is due to atypically in face recognition or a generally slower cognitive system is hard to establish from these data. More importantly, basing the trajectories on Benton test did not serve to normalise children's performance on the Storybook task.

The WBS group showed a delayed onset and rate on the Storybook task. As predicted, individuals with WBS were not affected by the inversion of the face stimuli. This finding confirmed previous results that individuals with WBS use a different system to

recognise faces employing featural strategy (Karmiloff-Smith, 1997; Karmiloff-Smith, et al., 2004).

In terms of relating disorder groups' performance on the Storybook task to the Benton task, current findings showed no correlation between these tasks. The current study concurs with previous studies that WBS behavioural proficiency on Benton face processing tasks seems to stem from an atypical developmental trajectory rather than from a normally developing face-processing system. This is also supported by our previous study on a population with this disorder (Karmiloff-Smith et al., 2004).

## CHAPTER 7

### CONSTRUCTION ABILITIES OF FACES



## **7.1 INTRODUCTION**

This chapter represents a digression from perception to construction tasks to examine face recognition abilities in the TD and clinical groups. This is a more exploratory approach to the face processing abilities. Some of the most intriguing questions we aim to examine are i) whether children in autism group are able to use their good construction skills to compensate for their poor face abilities and ii) can children with WBS use their relatively good face recognition abilities in a construction task?

Most of the visuo-spatial studies investigating face processing abilities use perceptual recognition tasks. However, development is dynamic and highly complex, where the maturation of certain cognitive processes may have an impact upon the development of other processes at different developmental stages (e.g., Johnson, 2005; Karmiloff-Smith, 1998). Hence, it is essential to examine visuo-spatial processing of face recognition through different tasks, each of which may recruit a different subset of developing mechanisms. In this chapter, face processing abilities are considered from the perspective of a construction rather than a perceptual task. The present study employed a visuo-construction task in which children were required to reconstruct the image of a target face by placing face features within a blank outline of a face.

Construction tasks differ from perceptual tasks, as they require the partitioning of a whole figure into parts. Correct image reproduction necessitates correct spatial rearrangement of the parts in relation to each other to achieve the correct configural formation. This is particularly true in face construction, where the integration of parts is an essential factor for successful completion of the task (Farah, Wilson, Drain, & Tanaka, 1995; Leder & Bruce, 2000; Maurer, Le Grand, & Mondloch, 2002; Tanaka & Farah, 1993). In addition, the construction task requires a sequence of actions, and therefore that a visuo-spatial representation (e.g., the difference between the target and goal state) must feed into action planning and selection.

Using a wide variety of paradigms, investigations have revealed that individuals with autism show enhanced detection of part-feature targets in visual search tasks (e.g., Plaisted, Swettenham, & Rees, 1999). They perform well on tasks such as block

design and object assembly that require a local focus (Minshew, Goldstein, & Siegel, 1997) and exhibit superior performance in detecting embedded figures (Happé, 1999; Shah & Frith, 1983). The results of our standardised test sets revealed a similar high level of performance for both HFA and LFA groups (see chapter 4 for scores on pattern construction).

Individuals with DS and WBS display a contrasting visuo-spatial profile to that found in the autism population, showing a particularly weak ability on construction tasks such as Navon or block construction (e.g., Bellugi et al., 2001). This profile is also evident in performance on pattern construction and copying task seen in chapter 4. Children with DS are often reported to have a ‘global’ style of processing (see chapter 2 for details), so it is interesting what form of preference they will take on this task. Individuals with WBS are impaired on visuo-construction tasks but are good at recognising faces. How, then, will they perform when constructing faces?

## **7.2 STUDY 4: FACE CONSTRUCTION TASK**

The face construction task developed in the current thesis initially set out to assess face-feature accuracy and two stages of face configural integration: first-order configural integration (eyes above nose and mouth below nose) and second-order configural integration (spatial relationship between the features). Although second-order configural accuracy is recoverable from the data, feature selection accuracy and first-order configural accuracy will be the focus of current analysis.

The specific investigations and predictions for disorder groups on this task were as follows:

1) Do children in the disorder groups show an increasing performance with age on the visuo-construction abilities in a face task?

**Predictions:**

- i) Children in TD and autism groups will show normal progressive performance accuracy, based on their good construction skills;
- ii) The performance of children in DS and WBS groups will be significantly poorer on the task (Bellugi et al., 1999; Fidler, 2005).

2) Do children show increased accuracy performance on the first-order configural processing?

**Prediction:** Several studies showed that even newborn infants focus on first-order configuration, thus it was predicted that children in all groups will develop a normal sensitivity to first-order configural processing (e.g., Johnson, et al., 1991).

3) What was the most salient feature during the face construction?

**Predictions:**

- i) Children in TD, DS and WBS groups will exhibit the normative pattern of better accuracy on eyes than other features (Hay & Cox, 2000);
- ii) Children in HFA and LFA groups would be more accurate in selecting the correct mouth feature (e.g., Hobson et al., 1988; Joseph & Tanaka, 2003).

4) Can performance on Pattern Construction or Benton recognition tests predict normal face construction?

**Predictions:**

- i) Benton will not predict normal constructive skills for all disorder groups since a good face representation may feed into an atypical construction process;
- ii) PC will not predict normal constructive skills for the autism groups as faces are social stimuli (an area of their weakness), but it will predict performance for the DS and WBS (their low constructional abilities may be the limiting factor on performance).

## **7.2.1 Method**

### ***7.2.1.1 Participants***

All participants completed the task. Details of all groups' participants can be found in chapter 3, section 3.2.1.

### **7.2. 1.2 Stimuli**

Six pictures of adults (3 females and 3 males) were taken with a digital camera (Nikon CoolPix 3500). Each face feature (eyes, nose and mouth) was distorted to achieve another two featurally changed new features using Deformer v 2.0. Use of this software allowed all textural information to be preserved. The photos were printed out on a high-quality paper and were 21 cm wide and 25 cm high. Photos and individual features were laminated and magnetic strips were attached to each of them. All the features were grouped by type (e.g., eyes, noses and mouths) and were placed on a magnetic board (see Figure 7.1).

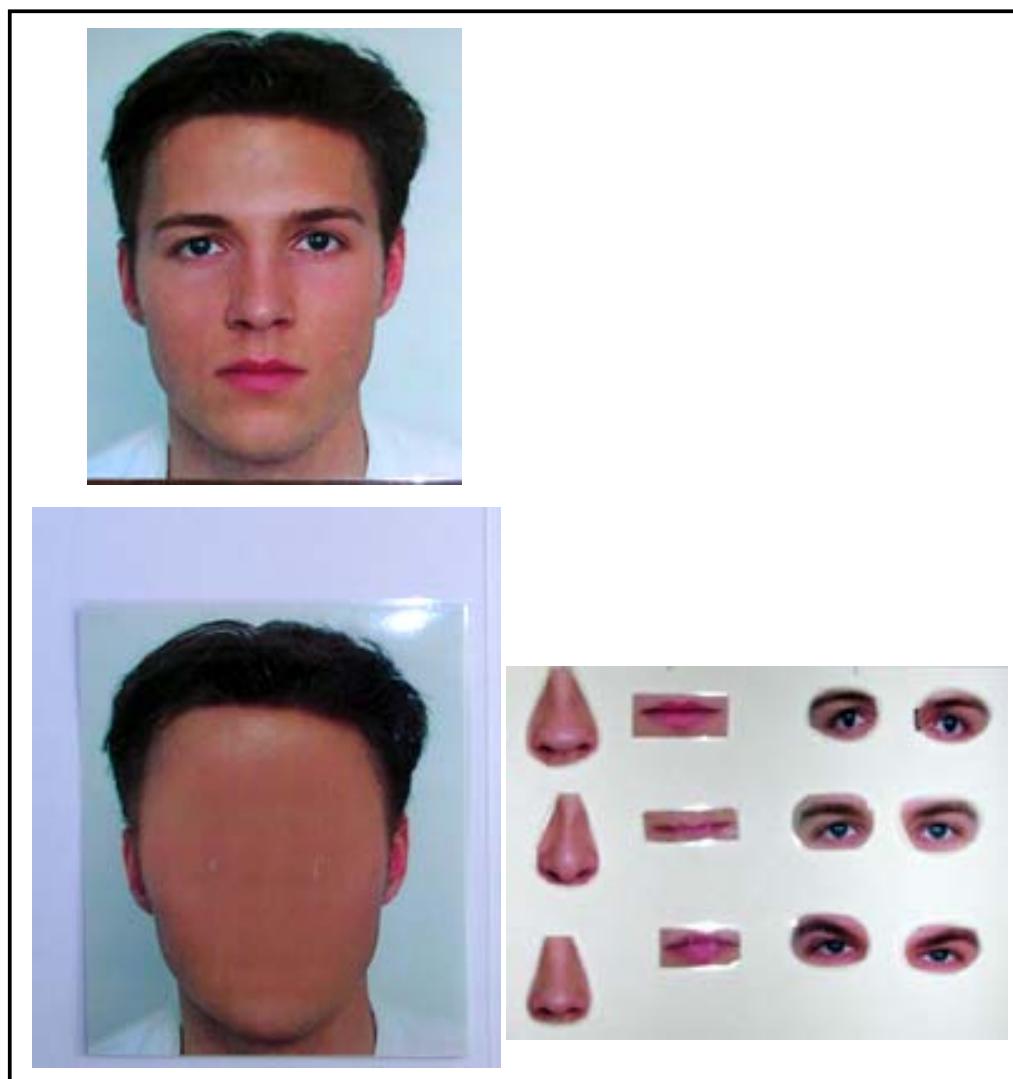


Figure 7.1: Example of the face construction stimuli.

### **7.2.1.3 Procedure**

Each child was shown a target face and a blank face outline (with no features) on a magnetic board. Another board with a choice of features including one target and two distracter features (3 x pairs of eyes, 3 x noses, and 3 x mouths) were placed next to the child. Eyes were placed as pairs. Placement of features on the board was randomised. The child was asked to play a game to make a face. Each child was given one practice trial to ensure that he/she understood the task procedure. If the child needed to be prompted, the experimenter asked the child: “What would you like to start with?” Each participant was asked to confirm that he/she finished the completion of making a face. The session lasted approximately 10 minutes.



Figure 7.2: Example of a typical testing procedure. Photo printed with parental consent.

There were two conditions: featural and first-order configural placement. In the featural condition, if a child chose a correct feature 1 point was given. There was a maximum score of 3 points per trial (one point per feature). Zero points were given if the child (i) chose the wrong feature or/and (ii) the child mixed the pairs of eyes, i.e., chose one eye from one pair and another eye from another pair. In the first-order configural condition, participants received 1 point per correctly placed feature (1

point for the eyes at the top of the face, 1 point for the nose in the middle of the face, and 1 point for the mouth at bottom of the face). There was a maximum of 3 points per trial. Zero points were given if: i) feature was placed outside the face area or/and ii) eyes were placed in a wrong orientation i.e. mouth upside down or eyes placed in a reverse position (right eye on left side of the face).

### **7.2.2 Results**

Accuracy data were taken from all participants in the study.

1. Trajectories were analysed using fully factorial analysis of co-variance (ANCOVA) for each group, where the within-participants factors were: accuracy scores on each feature (eyes, nose or mouth) and first-order configuration, and age was the co-variate.
2. A direct comparison of each clinical group to TD group was carried out using an ANCOVA (3x3) with Group (TD group compared to 4 clinical groups) as between-participants factor.
3. As in the previous results sections, the TD group is first described in detail. The proportion of correct accuracy scores on each feature (eyes, nose and mouth) and accuracy on the first-order configurational orientation was calculated for each individual. Accuracy results are presented first and summarised in tables (see chapter 4 for similar approach). This format will subsequently be used to summarise the results of predicting performance in the disorder groups according to their test ages on Benton and PC. Table 7.1 displays percentage accuracy scores on individual features by each group.

Table 7.1: Summary of % accuracy and standard error (SE) on selecting individual features.

GROUP	EYES		NOSE		MOUTH	
	%	SE	%	SE	%	SE
TD	77	25	75	23	93	10
HFA	65	21	64	16	81	15
LFA	23	17	38	20	57	20
DS	53	12	28	15	60	10
WBS	50	17	52	10	58	10

#### 7.2.2.1. TD group

##### Featural accuracy

As illustrated in Figure 7.3, children exhibited a significant improvement in accuracy with age (effect of age:  $F(1,23) = 89.63$ ,  $p < .001$ ,  $\eta_p^2 = .796$ ). The accuracy scores were significantly influenced by features (main effect of feature:  $F(1,23) = 57.81$ ,  $p <.001$ ,  $\eta_p^2 = .715$ ) performing at a ceiling level on the mouth feature above 8 years of age. A significant interaction between features and age (interaction of feature x age:  $F(1,23) = 34.32$ ,  $p <.001$ ,  $\eta_p^2 = .599$ ) revealed that performance accuracy increased with age at different rates modulated by feature. As shown in Table 7.1, children in the TD group showed significantly better accuracy scores on the mouth feature (mouth vs. eyes:  $t(15) = -3.81$ ;  $p = .002$ ; vs. nose:  $t(15) = -5.46$ ;  $p < .001$ ).

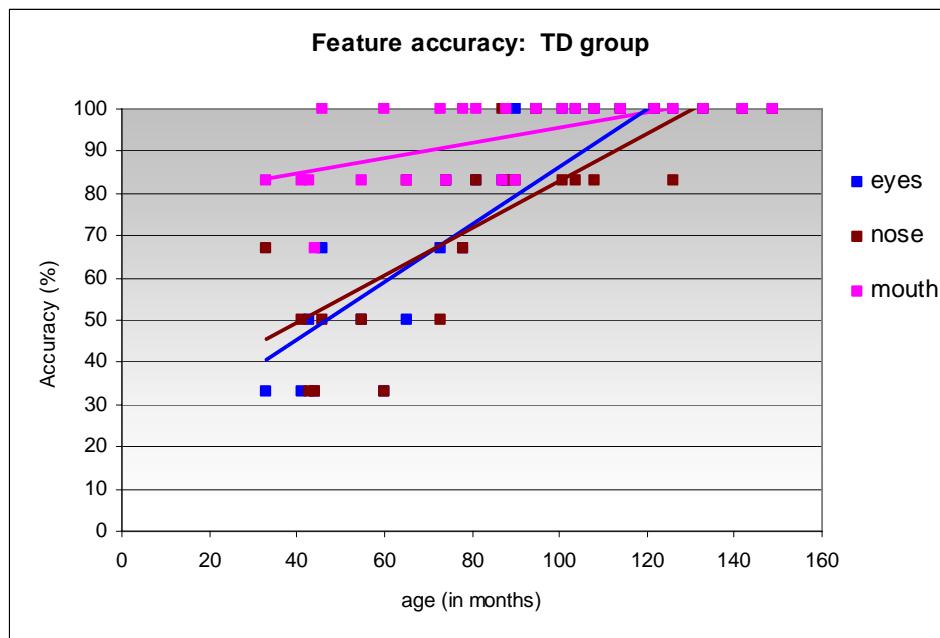


Figure 7.3: TD group developmental trajectories on accuracy of feature choice.

#### First-order configural accuracy

The study's other aim was to examine performance on configural placement of the features. Children's performance improved rapidly with age (effect of age:  $F(1,23) = 18.46$ ,  $p < .001$ ,  $\eta_p^2 = .445$ ). There were significant differences in performance scores on features (effect of features:  $F(1,23) = 7.90$ ,  $p = .010$ ,  $\eta_p^2 = .256$ ). Once again, the mouth feature was the least difficult in a configural placement, achieving ceiling scores at a very young age (see Figure 7.4). An interaction between features and age revealed that increased accuracy was feature dependent (interaction of feature x age:  $F(1,23) = 5.37$ ,  $p = .030$ ,  $\eta_p^2 = .189$ ). There were some concerns of ceiling effects in this condition, but as will be seen in later sections, the condition proved to be reasonably discriminating across the disorder groups.

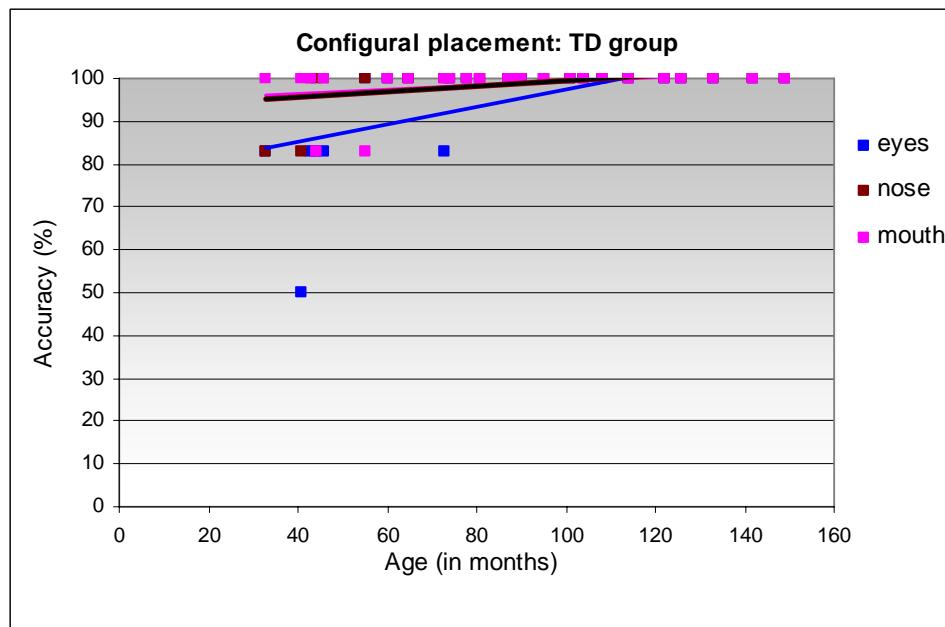


Figure 7.4: TD group developmental trajectories on accuracy of first-order-configural feature placement.

### 7.2.2.2 HFA group

#### Featural accuracy

Figure 7.5 shows that overall performance accuracy improved with age (main effect of age:  $F(1,14) = 24.91$ ,  $p < .001$ ,  $\eta_p^2 = .640$ ) but was not dependent on any particular feature (main effect of feature:  $F(1,14) = 2.50$ ,  $p = .136$ ,  $\eta_p^2 = .152$ ), although there is some indication that children performed better on mouth feature as illustrated in Figure 7.5 and Table 7.1. There was no interaction between features and age (interaction of feature x age:  $F(1,14) = .715$ ,  $p = .412$ ,  $\eta_p^2 = .049$ ) suggesting that different features had no influence on accuracy performance regardless of age.

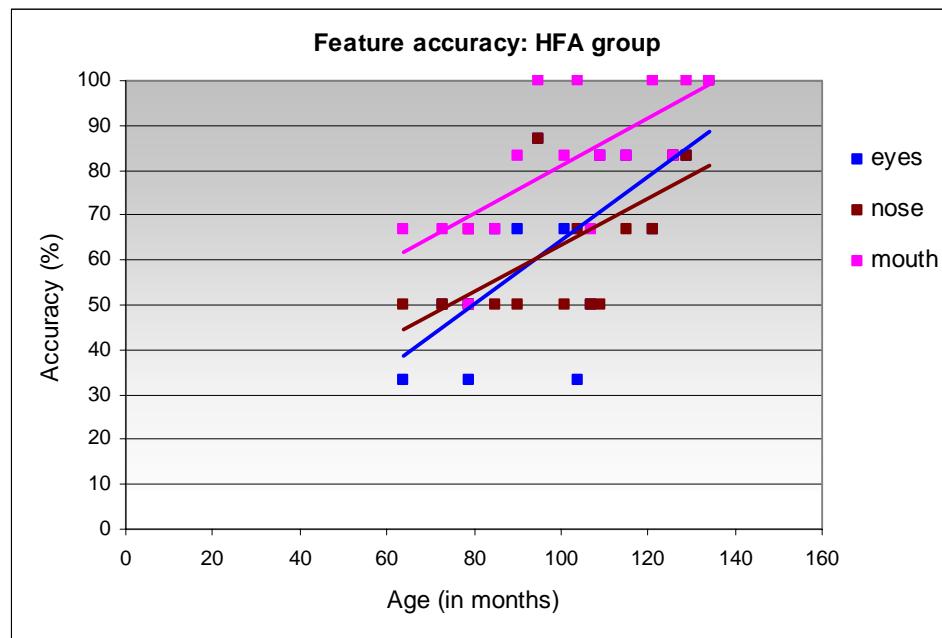


Figure 7.5: HFA group developmental trajectories of accuracy on feature choice.

#### First-order configural accuracy

Overall, performance accuracy increased with age (main effect of age:  $F(1,14) = 20.49$ ,  $p < .001$ ,  $\eta_p^2 = .594$ ). There was a significant main effect of feature ( $F(1,14) = 9.79$ ,  $p = .007$ ,  $\eta_p^2 = .411$ ), suggesting a differential effect of features on performance. Figure 7.6 demonstrates that children found placing the eyes feature more difficult than mouth or nose, similar to the TD group. A ceiling effect on mouth and nose but not eyes generated a significant interaction between features and age ( $F(1,14) = 6.53$ ,  $p = .023$ ,  $\eta_p^2 = .318$ ).

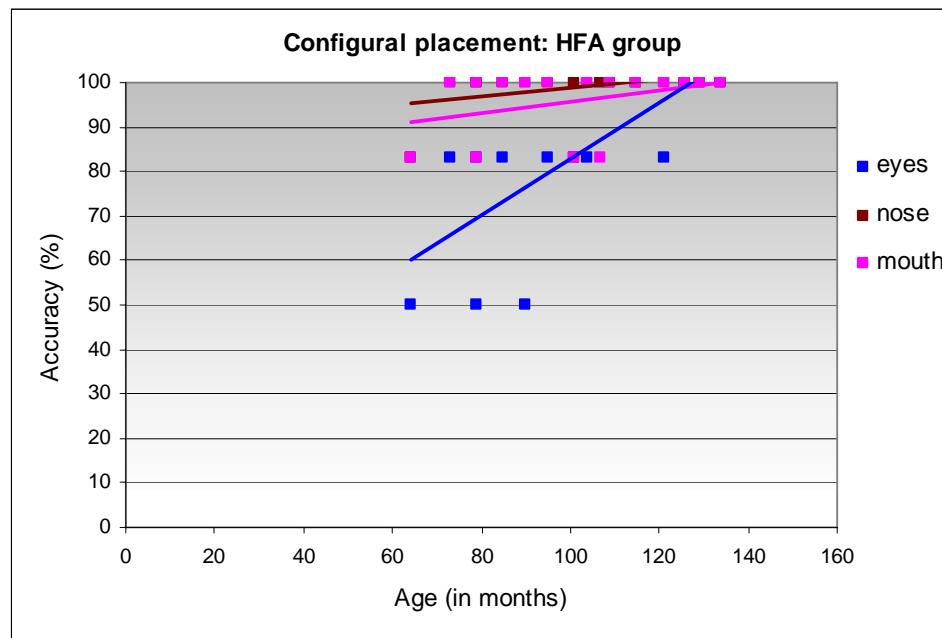


Figure 7.6: HFA group developmental trajectories on accuracy on feature choice.

#### Comparison to the TD trajectory

In the featural condition, the HFA group was less accurate than the TD group (main effect of group:  $F(1,37) = 6.56$ ,  $p = .015$ ,  $\eta_p^2 = .151$ ), but showed a similar rate of accuracy improvement with age (effect of group x age:  $F(1,37) = .99$ ,  $p = .326$ ,  $\eta_p^2 = .026$ ). There was a significant 3-way interaction suggests atypical development in the HFA group (features x age x groups:  $F(1,37) = 2.30$ ,  $p = .138$ ,  $\eta_p^2 = .058$ ). In the configural placement condition, the HFA group had a delayed onset and rate of performance in comparison to the TD group (main effect of group:  $F(1,37) = 22.31$ ,  $p < .001$ ,  $\eta_p^2 = .376$ ; effect age x group:  $F(1,37) = 12.83$ ,  $p < .001$ ,  $\eta_p^2 = .258$ ). There was a trend towards a significant interaction (interaction of feature x age x group:  $F(1,37) = 3.61$ ,  $p = .065$ ,  $\eta_p^2 = .089$ ), driven by the HFA group's greater difficulty in configural placement of their eyes choice (Figure 7.6). Separate analysis of accuracy improvement with age on eyes alone showed a significant difference between the HFA and TD ( $F(1,37) = 20.01$ ,  $p < .001$ ,  $\eta_p^2 = .346$ ).

### 7.2.2.3 LFA group

#### Featural accuracy

As shown in Figure 7.7, the overall performance of the LFA group was poor and children in the LFA group were the only group that did not significantly improve with age (main effect of age:  $F(1,15) = 3.77$ ,  $p = .071$ ,  $\eta_p^2 = .201$ ), although there was a trend suggesting improvement. Again, as in the previous studies, variability in performance was large. Children found the mouth feature to be the most important for constructing faces (see Figure 7.7 and Table 7.1 for overall scores) but this did not seem to have a large effect or was masked by a large variability (main effect of feature:  $F(1,15) = .18$ ,  $p = .679$ ,  $\eta_p^2 = .012$ ). A non-significant interaction between features and age revealed that performance accuracy was not affected by different features ( $F(1,15) = .860$ ,  $p = .140$ ,  $\eta_p^2 = .049$ ).

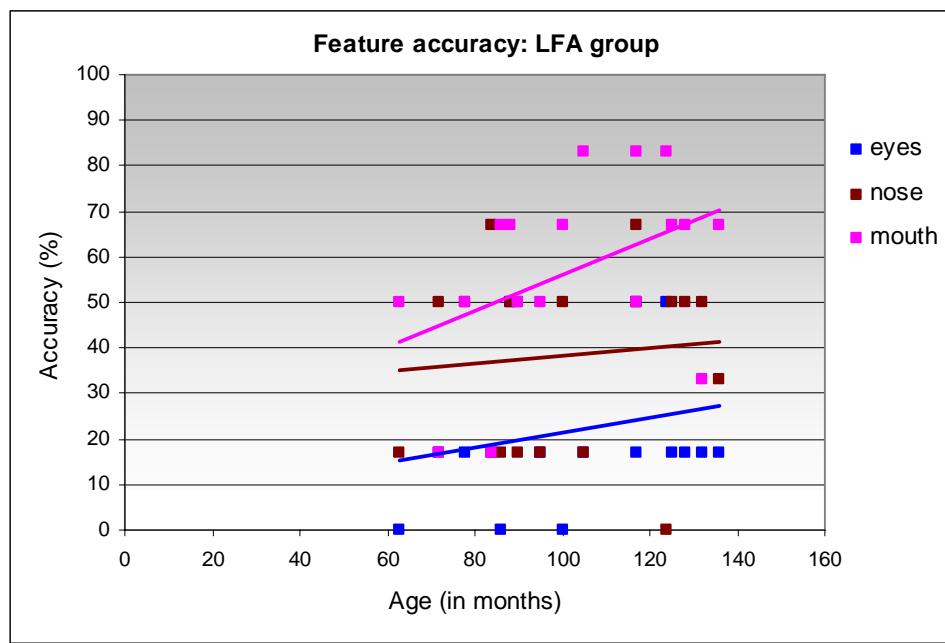


Figure 7.7: LFA group developmental trajectories on accuracy of feature choice.

#### First-order configural accuracy

As illustrated in Figure 7.8, majority of children were unable to place features in correct first-order positions regardless of feature sets and children's ages (main effect

of feature:  $F(1,15) = .03$ ,  $p = .862$ ,  $\eta_p^2 = .002$ ; main effect of age:  $F(1,15) = .36$ ,  $p = .557$ ,  $\eta_p^2 = .023$ ; interaction of feature x age:  $F(1,15) = .03$ ,  $p = .954$ ,  $\eta_p^2 = .003$ ). Most children placed the mouth feature in the middle part of the target face (see Figure 7.9) possibly highlighting its salience for them. This low performance is remarkable given the levels that the same group achieved on the highly similar pattern construction task (see Figure 4.4. in chapter 4).

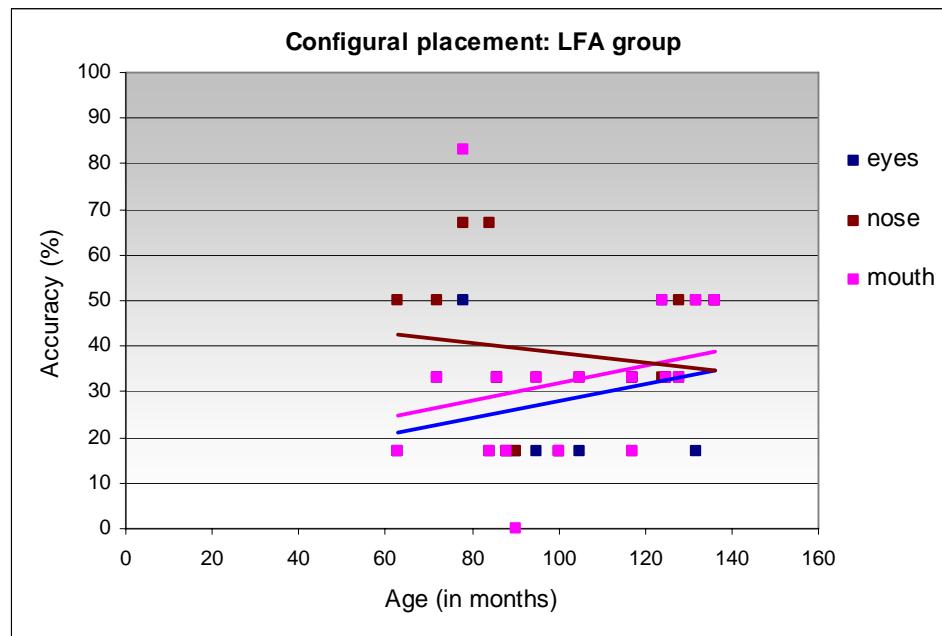


Figure 7.8: LFA group developmental trajectories on accuracy of first-order-configural feature placement.

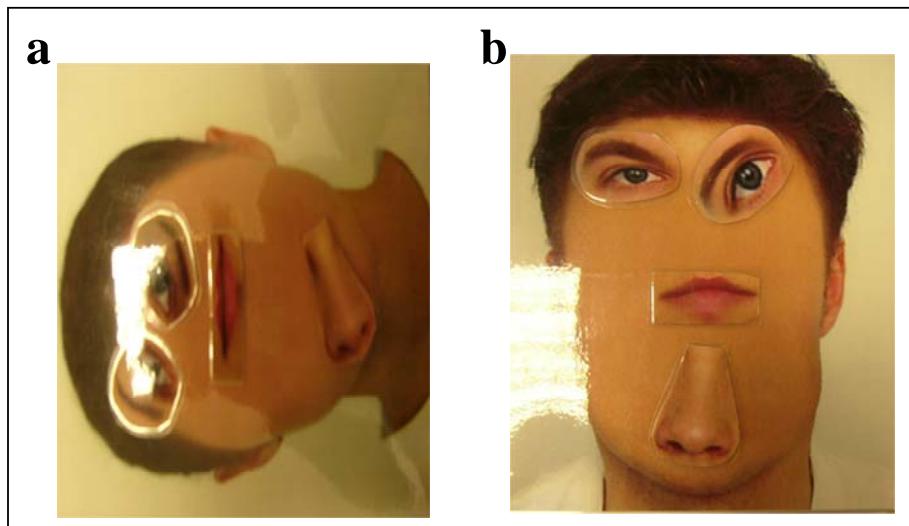


Figure 7.9: Representative example of faces constructed by a child in the LFA group.

### Comparison to the TD trajectory

In the featural condition, the LFA group was less accurate than TD group (main effect of group:  $F(1,38) = 4.44$ ,  $p = .042$ ,  $\eta_p^2 = .105$ ), and exhibited a delayed rate of development (effect of group x age:  $F(1,38) = 5.36$ ,  $p = .126$ ,  $\eta_p^2 = .124$ ). A significant 3-way interaction indicates that children in the LFA group expressed an atypical development of face construction skills on this task, mostly obvious in the much lower performance on eyes selection (features x age x group:  $F(1,38) = 10.38$ ,  $p = .003$ ,  $\eta_p^2 = .214$ ). On the configural placement condition, children in the LFA group showed delayed onset of performance accuracy (main effect of group:  $F(1,38) = 33.93$ ,  $p < .001$ ,  $\eta_p^2 = .472$ ) but a similar rate of improvement with age compared to the TD group (group x age:  $F(1,38) = .01$ ,  $p = .966$ ,  $\eta_p^2 = .001$ ). The lack of difference here is slightly misleading. The TD group were not improving much because they were at ceiling. The LFA group were not improving much with age, at around 40%. A 3-way interaction was not significant (interaction of feature x age x group:  $F(1,38) = 1.14$ ,  $p = .293$ ,  $\eta_p^2 = .029$ ) possibly to large group variability masked the differences between the groups. In summary, children in the LFA group showed atypical developmental profile on the feature condition and displayed an atypical treatment of the mouth feature on the configural placement task (see Figure 7.9).

#### *7.2.2.4 DS group*

##### Featural accuracy

Children exhibited a significant improvement in accuracy with age (main effect of age:  $F(1,13) = 20.65$ ,  $p < .001$ ,  $\eta_p^2 = .614$ ), depicted in Figure 7.10. Performance accuracy was not significantly modulated by different features (main effect of feature:  $F(1,13) = 2.63$ ,  $p = .129$ ,  $\eta_p^2 = .168$ ), which did not change across development trajectory (interaction of feature x age:  $F(1,13) = 1.28$ ,  $p = .278$ ,  $\eta_p^2 = .090$ ). This effect size suggests that the role of features may have been masked by the small sample and indeed children with DS showed a significantly lower accuracy on nose feature when simple t-tests were used (nose x eyes:  $t(15) = 8.04$ ,  $p < .001$ ; nose

x mouth:  $t(15) = -7.93$ ,  $p < .001$ ). This contrasts with other groups. Table 7.1 illustrates this finding.

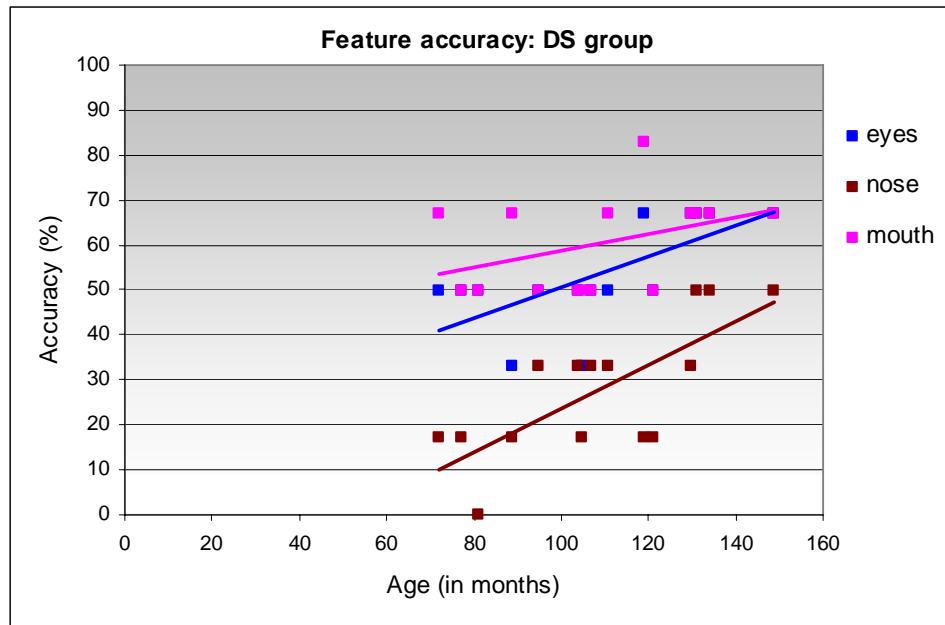


Figure 7.10: DS group developmental trajectories on accuracy on feature choice.

#### First-order configural accuracy

Overall accuracy increased with age (main effect of age:  $F(1,13) = 17.81$ ,  $p < .001$ ,  $\eta_p^2 = .578$ ). There was no difference in performance on different features (main effect of feature:  $F(1,13) = 1.04$ ,  $p = .327$ ,  $\eta_p^2 = .074$ ), and accuracy was not influenced by different features sets with increased age (interaction of feature x age:  $F(1,13) = .15$ ,  $p = .709$ ,  $\eta_p^2 = .011$ ). This finding is illustrated in Figure 7.11.

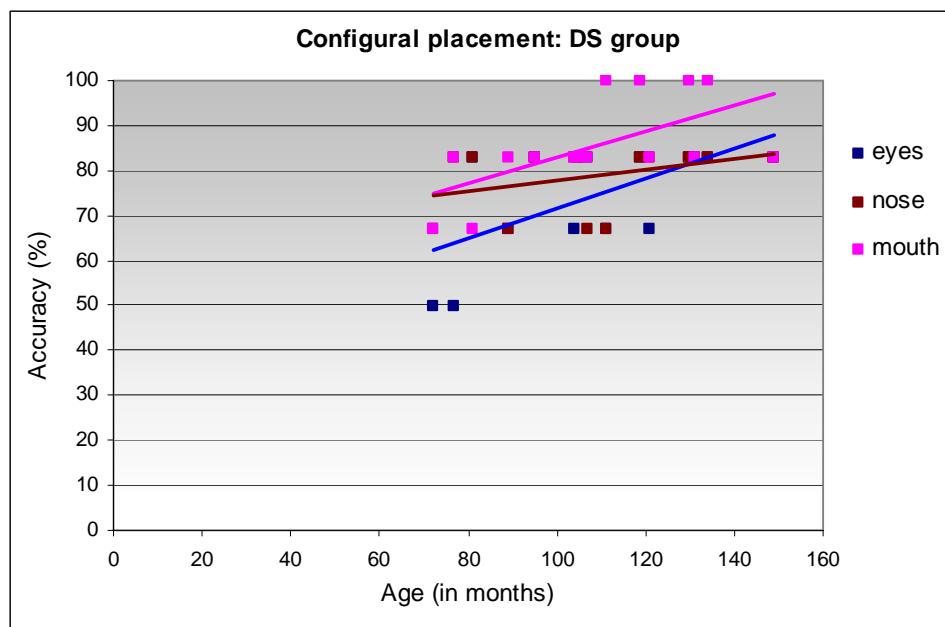


Figure 7.11: DS group developmental trajectories on accuracy of first-order.

#### Comparison to the TD trajectory

For the feature condition, children in the DS group showed delayed onset but not rate in comparison to the TD group (main effect of group:  $F(1,36) = 8.21$ ,  $p = .007$ ,  $\eta_p^2 = .186$ ; group x age:  $F(1,36) = 3.13$ ,  $p = .085$ ,  $\eta_p^2 = .080$ ). Furthermore, a significant 3-way interaction (feature x age x group:  $F(1,36) = 5.55$ ,  $p = .024$ ,  $\eta_p^2 = .134$ ) suggests that children in DS group showed a different profile of performance on the task. However, this result should be treated with caution as children in the TD group were at ceiling for the mouth feature. Nevertheless, while eyes and nose accuracy were equivalent in the TD group, nose was lower than eyes in the DS group. Therefore, a comparison of performance on the nose feature was carried out to see if it better discriminates the groups. In this case the DS group had delayed onset but not rate (main effect of group:  $F(1,36) = 10.05$ ,  $p = .003$ ,  $\eta_p^2 = .218$ ; group x age:  $F(1,36) = .38$ ,  $p = .541$ ,  $\eta_p^2 = .010$ ). In the first-order configural placement condition, children in the DS group had delayed onset and rate of accuracy performance in comparison to the TD group (main effect of group:  $F(1,36) = 45.45$ ,  $p < .001$ ,  $\eta_p^2 = .558$ ; group x age:  $F(1,36) = 7.96$ ,  $p = .008$ ,  $\eta_p^2 = .181$ ). However, a 3-way interaction was not significant (features x age x group:  $F(1,36) = .462$ ,  $p = .501$ ,  $\eta_p^2 = .013$ ), suggesting a normal but delayed developmental profile of performance in the DS group.

### 7.2.2.5 WBS group

#### Featural accuracy

As shown in Figure 7.12, performance accuracy in the WBS group improved significantly with increasing age (main effect of age:  $F(1,15) = 10.26$ ,  $p = .006$ ,  $\eta_p^2 = .406$ ) and was not modulated by any individual feature (main effect of feature:  $F(1,15) = .18$ ,  $p = .116$ ,  $\eta_p^2 = .156$ ). Also, the performance scores did not depend on any individual feature with increasing age (interaction of feature x age:  $F(1,15) = .860$ ,  $p = .237$ ,  $\eta_p^2 = .092$ ).

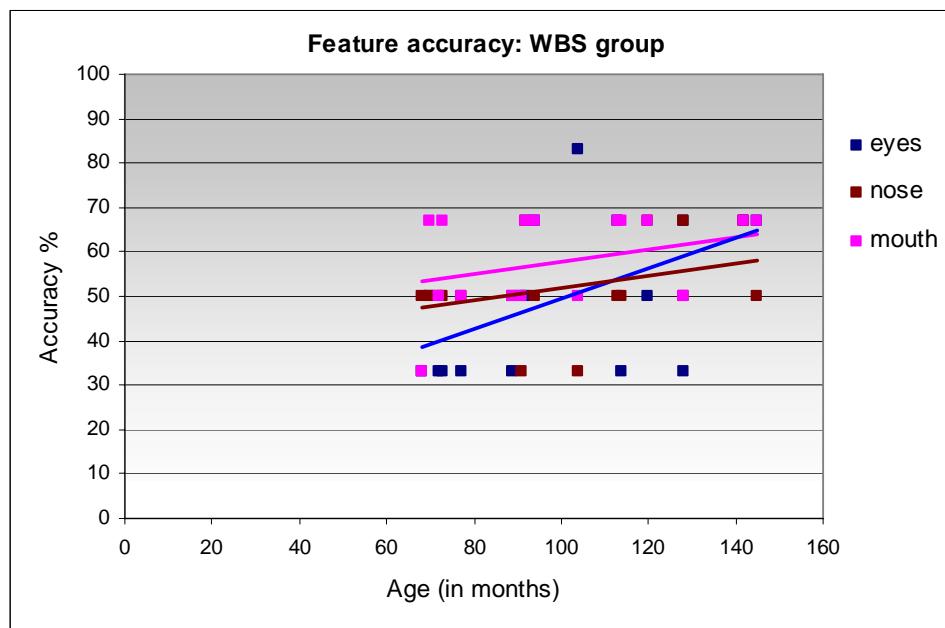


Figure 7.12: WBS group developmental trajectories on accuracy of feature choice.

#### First-order configural accuracy

In general, children with WBS found it difficult to place features in the correct positions. In particular, some children were placing eyes features incorrectly by reversing their sides. This was observed in 25% of children. None of the other groups showed this effect (Figure 7.13). Performance accuracy increased with age (main effect of age:  $F(1,15) = 6.72$ ,  $p = .020$ ,  $\eta_p^2 = .309$ ). Additionally, a significant difference in feature performance was found, driven by poor placement of the eyes

(main effect of feature:  $F(1,15) = 7.35$ ,  $p = .016$ ,  $\eta_p^2 = .329$ ). However, the feature sets had no influence on accuracy scores with increased age (feature x age:  $F(1,15) = 1.29$ ,  $p = .273$ ,  $\eta_p^2 = .079$ ). Figure 7.14 shows examples of the inability of children in the WBS group to place the eye features correctly, reversing left and right eyes.

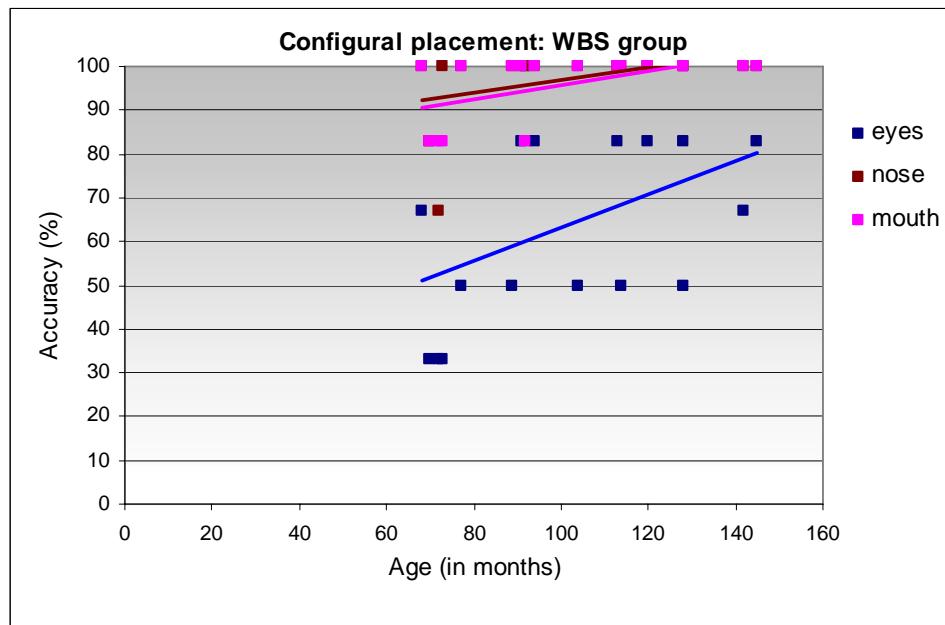


Figure 7.13: WBS group developmental trajectories on accuracy on first-order-configural feature placement

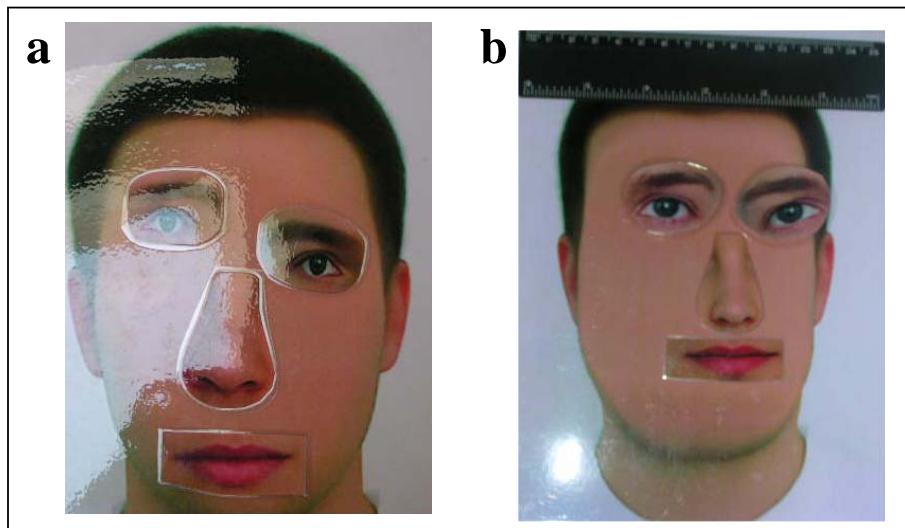


Figure 7.14: Examples of incorrect placement of the eyes features by a) KH, age 10; b) CM, age 8.

Comparison to the TD trajectory

In the featural accuracy, comparison of children with WBS to the TD revealed similar onset of accuracy on the task (main effect of group:  $F(1,38) = 1.02$ ,  $p = .318$ ,  $\eta_p^2 = .026$ ) but WBS group had a slower rate of improvement with increased age (effect of group x age:  $F(1,38) = 9.23$ ,  $p = .004$ ,  $\eta_p^2 = .195$ ). A 3-way interaction was not significant (features x age x groups:  $F(1,38) = 2.88$ ,  $p = .098$ ,  $\eta_p^2 = .070$ ). In the first-order configural accuracy condition, WBS group performance had delayed onset of accuracy compared to the TD group (main effect of group:  $F(1,38) = 15.07$ ,  $p < .001$ ,  $\eta_p^2 = .284$ ) and delayed rate at marginal level (effect of group x age:  $F(1,38) = 3.99$ ,  $p = .053$ ,  $\eta_p^2 = .095$ ). Again, 3-way interaction was not significant suggesting similar developmental profile of performance in both groups (interaction of feature x age x group:  $F(1,38) = .13$ ,  $p = .724$ ,  $\eta_p^2 = .103$ ). In sum, children in WBS group showed a delayed rate of development on both conditions in comparison to the TD group.

### **7.2.3 Benton task and Pattern Construction**

Performance on the Benton test age (generated from current TD group, see chapter 4.5.2.1 for individual scores) predicted accuracy level on the feature accuracy condition only for the WBS group ( $F(1,15) = 6.86$ ,  $p = .019$ ,  $\eta_p^2 = .314$ ). For the first-order configural condition, it predicted performance for the HFA group ( $F(1,14) = 9.72$ ,  $p = .008$ ,  $\eta_p^2 = .412$ ) and the WBS group ( $F(1,15) = 10.49$ ,  $p = .006$ ,  $\eta_p^2 = .410$ ). There was no significant interaction between feature type and Benton test age in any group.

Performance test age on PC predicted accuracy scores for feature placement for the TD group ( $F(1,23) = 67.50$ ,  $p < .001$ ,  $\eta_p^2 = .212$ ) and the DS group at marginal level ( $F(1,15) = 4.33$ ,  $p = .058$ ,  $\eta_p^2 = .498$ ). A significant interaction of feature x PC age was observed only in the TD group ( $F(1,24) = 26.23$   $p < .001$ ,  $\eta_p^2 = .533$ ). Also, PC test age successfully predicted accuracy on first-order configural placement for the TD group ( $F(1,24) = 15.44$ ,  $p < .001$ ,  $\eta_p^2 = .402$ ) and DS group ( $F(1,13) = 6.52$ ,  $p = .024$ ,  $\eta_p^2 = .334$ ).

Tables 7.2 and 7.3 show a comparison of the disorder groups on Benton and PC test age with the TD group. Recall, a significant 3-way interaction suggests an atypical developmental profile of the group. Trajectories for each group plotted against Benton test age and PC age respectively can be found in Appendix D (Figure 1 - 2).

Table 7.2: Summary of disorder group's comparisons to the TD group on feature and configural placement accuracy, when controlled for their abilities on Benton.

GROUP	BENTON TEST AGE					
	FEATURES			FIRST-ORDER CONFIGURATION		
	main effect of group	2-way*	3-way**	main effect of group	2-way	3-way
<b>HFA</b>	NS	NS	NS	NS	NS	NS
<b>LFA</b>	p=.069	p=.001	p=.030	p<.001	NS	NS
<b>DS</b>	p=.035	p=.005	p=.056	NS	NS	NS
<b>WBS</b>	p=.036	p=.010	p=.028	p<.001	p=.012	NS

\* Disorder groups compared to TD (group x Benton test age), \*\* group x age x condition (feature or configural). NS = no difference between the disorder group and TD.

Table 7.3: Summary of disorder group's comparisons to the TD group on feature and configural placement accuracy when controlled for their abilities on PC.

GROUP	PATTERN CONSTRUCTION TEST AGE					
	FEATURES			FIRST-ORDER CONFIGURATION		
	main effect of group	2-way*	3-way**	main effect of group	2-way	3-way
<b>HFA</b>	NS	p=.003	p<.001	NS	NS	NS
<b>LFA</b>	NS	p<.001	p<.001	p<.001	NS	NS
<b>DS</b>	NS	NS	NS	p<.001	p=.004	NS
<b>WBS</b>	NS	NS	NS	p=.010	p=.029	p=.047

\* Disorder groups compared to TD (group x PC age equivalent), \*\* group x age x condition (feature or configural). NS = no difference between the disorder group and TD.

### 7.2.4 Summary

Table 7.4 summarizes each group's performance on the build-a-face task in comparison to the TD group.

Table 7.4: Summary of results for construction task for all groups.

GROUP	DEVELOPMENTAL PROFILE	
	FEATURES	FIRST-ORDER CONFIGURAL
TD*	NORMAL	NORMAL
HFA	DELAYED ONSET	DELAYED ONSET & RATE
LFA	DELAYED ONSET & RATE <b>ATYPICAL</b>	DELAYED ONSET <b>ATYPICAL</b>
DS	DELAYED ONSET <b>ATYPICAL</b>	DELAYED ONSET & RATE
WBS	DELAYED RATE	DELAYED RATE

\*Normality of the performance for the disorder groups is expressed as a NS 3-way interaction (task x age x group) in comparison to the TD group. Delayed onset = sig. main effect of group; delayed rate = sig. effect of group x age.

### 7.2.4 Discussion

The purpose of this study was to establish whether children in the TD group and disorder groups develop sensitivity to faces, viewed through the lens of a construction task. In the case of the TD group, their performance rapidly improved with age achieving ceiling scores very early in their development, particularly in first-order configural placement. This supports previous claims that sensitivity to first-order information is available from early infancy (Johnson, 1991). Contrary to initial predictions in favour of the eyes children in the TD group showed better accuracy for mouth feature selection in the construction task, perhaps because of its high physical contrast and social importance. On the other hand, eyes provide us with even more powerful social information, such as another person's gaze, intentions and emotional states. Perhaps the effect emerges because eye recognition requires us to identify two components (right and left eye), which are harder to identify in isolation. Another possibility is that correct identification of eyes requires configural

processing on a finer scale. This skill develops steadily with increased age as seen in Jane faces and Storybook studies.

In the feature accuracy condition, children in the disorder groups displayed delayed (HFA and WBS groups) or atypical (LFA and DS) developmental patterns in comparison to TD children. In contrast, in the configural placement condition, only children in LFA group showed an atypical profile while the other groups' performance was delayed either in onset, rate, or both.

Contrary to initial predictions children in the HFA group showed a delayed (onset) performance on featural accuracy and were less sensitive to first-order configural processing (delayed onset and rate). This result is also in contrast to another very similar study where the same group of children with HFA showed normal performance on object (house) construction task (Thomson, Annaz & Thomas, 2005). An atypical developmental profile emerged from the LFA children who were poor on feature accuracy and found the mouth feature to be of prime importance in an atypical way as reported earlier by Hobson (1988) and Joseph and Tanaka (2003). Children in the LFA group were also the only group that violated first-order configural face processing by placing the mouth in the middle of the face. These findings suggest that children with LFA group are unable to use their good construction skills in a face task. It also contradicts the weak central coherence theory that suggests good feature processing in autism population (discussed in chapter 2). Moreover, lack of sensitivity to first-order configural face processing is inconsistent with research studies that show that even young infants are sensitive to the first-order configural face processing (Johnson et al., 1991). These data represent a novel finding with respect to face stimuli. Clearly, the social status of the stimuli affected the performance of individuals with autism. However, it remains unclear whether children thought they were accurately reproducing the target face, or they deliberately did not reproduce it to end up with a less aversive pattern for them.

Children with DS revealed an atypical profile in the feature accuracy condition. However, this was possibly triggered by their greater variability in performance. In general, children with DS showed a delayed development on the task. They demonstrated a significantly poorer accuracy level on the nose feature than other

groups. The reason for this performance may be due to slower development of general visuo-spatial processing and noses neither operate, as an important feature for social communication, nor possess distinctive regions and contrast spectrum. The current data concur with earlier studies that reported poor spatial memory and visuo-constructive abilities and a peculiar lack in the accuracy on feature processing in DS (Fidler, 2005). Some indirect evidence from the standardised tests suggests that individuals with DS use a holistic strategy in drawing tasks and fail to reproduce the features correctly (Bellugi, et al., 1999).

In general, individuals with WBS revealed relatively good performance on the task, given their low ability on object construction tasks (Farran & Jarrold, 2003; Pani et al., 1999). The majority of individuals scored above 50%, which suggests a high success level considering that usual scores on pattern construction are at floor (Bellugi et al., 1999). Moreover, good performance on featural accuracy may be due to the featural preference reported previously by Bellugi et al. (1994) and Farran and Jarrold (2003). Nonetheless, the WBS group showed a delayed rate of performance on this task. Some peculiar results were obtained on first-order configural condition, where a number of children placed eyes in the reverse order (right eye on left side and visa versa). One could speculate that the strabismus, which affects many individuals with WBS, may have played a role. Interestingly, the pattern of data on the configural dimension almost mirrors that of a similar construction task using houses. Morrison, Annaz and Thomas (2005) showed that children with WBS found configural placement of the windows difficult, displaying a similarly delayed trajectory as for the placement of eyes (see Appendix D, Figure 3).

Hoffman et al. (2003) reported that individuals with WBS are able to complete pattern construction tasks at simple levels. This appears to be the case for the current task, which makes limited demands. However, it does not explain why children with DS and LFA performed poorly on this task, given that individuals with LFA usually perform well on any object visuo-construction task (e.g., Happé, 2000). The social nature of the stimulus appears to have a strong influence on each group's performance. Individuals with WBS are hypersociable, with a tendency to look at faces in preference to other visual stimuli from a very young age (e.g., Karmiloff-

Smith, Bellugi, Grant, & Baron-Cohen, 1995). Thus, it is possible that more socially relevant stimulus such as faces or even houses are more favourable, resulting in the enhancement of the accurate face construction. Current findings also highlight the necessity for a more rigorous approach in future studies, in which distinctive “task-dependency” plays an important role in data analysis.

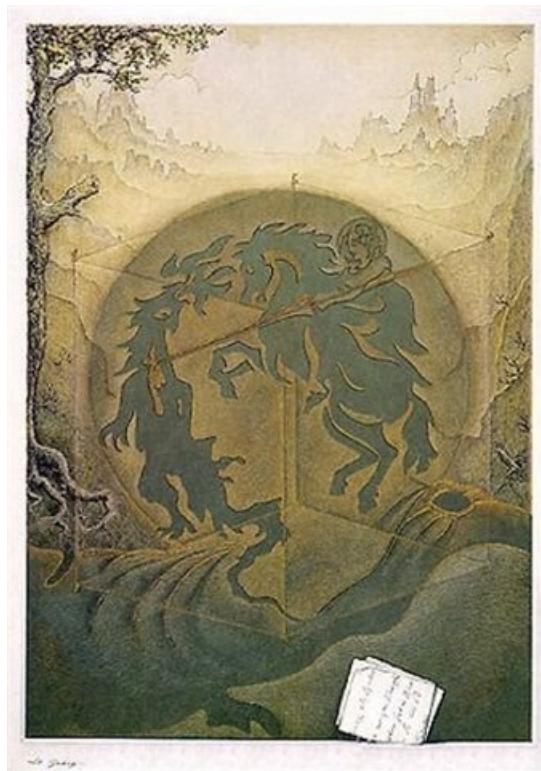
Lastly, current results demonstrate that Benton scores predicted normal level of performance on the feature accuracy condition only for HFA group in comparison to the TD group. However, the Benton test did predict normal performance of all disorder groups on the first-order placement task. As predicted, when children were matched according to PC, the DS and WBS groups showed similar performance to the TD group on feature accuracy condition. However, in the first-order configural placement, PC predicted normal performance for all groups except for WBS.

Although it was not viable to include an inverted condition in the current battery of tests, given previous findings, the following predictions might be made: Children in the TD group will show no difference between the conditions (features are easy to pick up and first-order effect is robust). Children in HFA and LFA groups will improve, but the LFA group will show larger improvement (inverted faces are not as aversive). There will be no change in performance for DS and WBS.

This completes set of current studies. We will now turn to a consideration of the patterns that emerged across the studies between and within the groups, and what these patterns tell us about the way that atypical constraints can shape visuo-spatial development of developmental disorders.

## CHAPTER 8

### GENERAL DISCUSSION AND CONCLUSIONS



## **8.1 INTRODUCTION**

Face processing like many other abilities develops over many years (e.g., Johnson, 2005; Mondloch et al., 2002) and is interconnected with other processes such as auditory perception during speech recognition (Neville, *in press*). Developmental changes that occur in face recognition are complex and as Morton (2004) pointed out, atypical development is nearly always caused by multiple factors that may exist at different levels including biological, cognitive and behavioural.

The main aim of this thesis was to investigate whether children with autism, Down syndrome and Williams syndrome develop normal face recognition abilities at the cognitive and/or behavioural levels. This question was addressed by assessing their developing sensitivity to holistic, configural and featural aspects of face processing. This thesis represents one of the first attempts to use trajectory based cross-syndrome studies to investigate the development of face recognition abilities. Tracing developmental trajectories using linear regression analysis was utilized to explore each groups' abilities in depth and to distinguish different ways in which development could be 'delayed' or atypical.

In this chapter, we consider the experimental findings in relation to the theories introduced earlier in the face processing and developmental disorders chapters. The first part of this chapter will discuss the main findings in relation to the development of holistic, configural and featural face recognition. In the second part, we will seek to identify the different constraints that may shape development in each disorder. Following Morton (2004) we shall endeavor to distinguish between behavioural and cognitive levels, and thus between re-description and explanations. Finally, methodological issues encountered in the current thesis and future research will be discussed.

### **8.1.1 The Development of Holistic Face Recognition**

The whole-part study revealed that none of the disorder groups followed the normal developmental trajectory on holistic face performance. Beginning with the autism groups, children in the HFA group showed delayed onset in sensitivity to holistic processing (based on their scores on upright whole-face recognition). However, this reasonably good performance of the HFA groups might be delivered by atypical focusing on individual face features. Some supporting evidence for feature-based strategy comes from HFA group's performance on part-face condition and pattern construction, which seemed to follow the normal developmental trajectory. It is in line with the lack of inversion effect shown by HFA group, also reported in previous studies (e.g., Deruelle et al., 2004; Hobson et al., 1988; Langdell, 1978; Schultz et al., 2000).

A contrasting profile was shown by children in the LFA group, who displayed poor performance on the task, and followed an atypical developmental trajectory by performing better on inverted faces (in whole-face condition). It seems that the featural skills that helped HFA group to achieve a relatively good performance on the face task, was not utilized by the LFA group in this instance. Factors such as aversive response to faces may have affected the performance outcome of individuals with LFA. If our marker of holistic processing is taken to be an inversion effect in the whole-faces condition, then children in the autism population do not seem to be developing it normally. Several possible explanations are proposed here. First, previous studies reported that individuals with autism, even as young toddlers, spend less time looking at faces (e.g., Osterling & Dawson, 1994) and show atypical eye gaze (Jolliffe & Baron-Cohen, 1997). It is possible that their visual system receives less input in early life, which is necessary for normal face development (Le Grand et al., 2003). There is also a hint from an ERP study carried out by Grice, Halit, Farroni, Baron-Cohen, Bolton and Johnson (2005) that young children with autism show early problems in face recognition due to atypical subcortical involvement, which in turn, may lead to atypical specialisation and organisation of representations at cortical level. But how can we explain the differences between the autism groups? Based on the findings discussed

above, the differences between our two groups of autism might be attributed to qualitative and quantitative factors in early development. Qualitative factors could include gaze aversion, orienting preference, sensitivity to nonverbal cues such as gesture, facial expression and body language; quantitative might encompass degree of featural processing. Also, appropriate social play and conversational language at young age may contribute to quantity of face input; all may be necessary for normal face recognition skills, albeit at different levels of contribution to the system.

Children in the DS group appeared to use holistic processing but their progression was at a delayed rate in comparison to the TD group. At present, this finding can only be compared to results from standardised test (Bellugi et al., 1999), consistent with the idea that individuals with DS use holistic information on the block design and Navon tasks.

Many studies reported that individuals with WBS, unlike those with autism, appear to be fascinated by faces and spend much of their time looking at them (e.g., Bellugi et al., 1988; Udwin & Yule, 1991). However, in the current study, children in the WBS group did not show behavioural markers of normal face development i.e. emergence of inversion effect and a difference between whole-part conditions. This suggests that the WBS group did not develop sensitivity to holistic face information. As in the HFA group, it is possible that the WBS group, successfully recognised faces by applying their good featural processing style to progress on the recognition of face-features. Furthermore, their performance on the Benton test (which is in the normal range) did not predict their level of holistic face recognition. This also supports the claim that the Benton test can be resolved just by using featural processing (Duchaine & Nakayama, 2004).

### **8.1.2 Development of Configural and Featural face recognition**

The Jane faces and Storybook studies were designed to test development of configural and featural face recognition. These studies differed in their design in a number of ways. In particular, the storybook study had a memory component and sacrificed a degree of

test sensitivity in favor of ecological validity. However, both studies revealed converging results. In line with previous reports, children in the TD group showed gradually increasing sensitivity to configurally changed Jane faces and the emergence of an inversion effect with increased age in both studies (Brace et al., 2001; Carey & Diamond, 1994; Freire & Lee, 2001; Mondloch et al., 2002). Similar to the whole-part study, none of the disorder groups followed normal developmental trajectories on this task, showing the lack of an inversion effect and reduced sensitivity to configurally manipulated faces in the Jane faces study.

Once again, an unusually good performance by LFA group was evident in the inverted faces condition in both studies. Children in the HFA group were the only disorder group to improve on configural trials, thus once more it is proposed that good behavioural scores on configural condition could be achieved via a well established featural system in the HFA group. The question of how can one use featural information to resolve configural differences remains to be examined<sup>(1)</sup>. These findings are in line with claims that children with autism and WBS show an atypical face recognition skills (Ellis, et al., 1994; Klin et al., 1999; Langdell, 1978; Karmiloff-Smith et al., 2004), and children in the DS group have severely delayed face recognition ability (Kaiser et al., 2005). Poor specialisation for face in the DS group could stem from problems in visual exploration in play situations (Gunn, Berry, & Andrews, 1982) and eye contact (Berger & Cunningham, 1983, Brown et al., 2003) that have been reported in early childhood.

Also, several neuro-imaging studies have demonstrated that poor specialisation for faces that fails to activate the FFA could stem from lack of expertise with faces (Passarotti, et al., 2001). Furthermore, performance on the Benton test did not predict normal configural processing in any of the disorder groups.

---

<sup>1</sup> One possibility is that second-order configural information creates features, e.g. the distances between the eyes might be a light coloured area. When eye separation is changed, this feature changes too!

### **8.1.3 Visuo-Construction skills in face processing.**

The current literature on face processing does not specify the relationship of perception and construction. For example, in the WBS literature, a distinction is made between perception and construction, with the former occurring before construction. Pani and colleagues (1999) have argued that perception (the early stage) develops normally in WBS and construction (the later stage) is impaired. However, the results obtained from the current study do not support Pani's argument. Instead, the current data suggest that atypical performance on one domain can have cascading effects on another domain due to continuous dynamics during development (e.g., Karmiloff-Smith, 1998). Moreover, as advocated by Thomas (2005), processes such as interactivity and compensation can play an important role in perception versus construction tasks. For instance, children in the HFA group showed a delayed onset of development during our construction task, but performance of the LFA group was poor and followed an atypical trajectory; thus the social stimuli used in Study 4 hindered the performance of the autism groups. In contrast, the social nature of construction task helped children with WBS to acquire better performance than that usually observed on pattern construction in this and in previous studies (e.g., Bellugi et al., 1988). Although many of the studies provide strong support for impaired visuo-construction skills in WBS, two longitudinal case studies showed progress that passed through normal stages of development. The authors suggested that the effects of training could have positive impact on performance (Stiles et al., 2000).

Poor performance by children in the DS group continued across all studies carried out in this thesis. Children with DS showed a delayed onset in their developmental performance on the construction task, which re-affirm earlier findings in previous studies on visuo-spatial construction (Fidler, 2005).

Lastly, the results from the face construction study provide further evidence that face recognition does not have to be computationally special (e.g., Diamond & Carey, 1986; Morton & Johnson, 1989). For instance, children in the HFA group were able to achieve

relatively high scores via featural processing on the face tasks although they did not seem to develop normal ‘face expert’ processing. That is, they could recognise faces as if they were objects.

There seems to be an array of complex inter-dependency between different cognitive abilities that play an important role in normal face recognition (i.e., language, non-verbal social communication).

#### **8.1.4 Featural processing and feature salience**

The Jane faces study examined developmental trajectories of featural face recognition, while whole-part and face construction studies investigated facial feature saliency. In several cases, disorder groups followed normal but delayed developmental trajectories on featural face recognition. Several previous studies suggested that faces are recognised sequentially in a top-down ordering from eyes to nose and mouth, thus eyes seem to play a very important role for the recognition process. Although this might seem probable, it does not necessarily mean that children are encoding eyes better than other features. There is no doubt that the informative and privileged nature of eyes that comes from their high contrast, symmetrical positioning (e.g., in emotion recognition) has an immense influence on participants’ performance. However, current data showed that children in the TD group sometimes were equally good on eyes and mouth but significantly worse on the nose feature. Similar profiles were observed in the disorder groups, with the exception of the LFA group who showed atypical performance on the mouth region, which was considerably better than the other features. This finding was reported in earlier studies of autism population (Hobson, 1988; Joseph & Tanaka, 2003), as well as in typically developing infants that appeared to rely on a single feature such as mouth, which is a particularly important feature for them (Schwarzer & Massaro, 2001). However, results from the face construction task showed an overwhelmingly atypical performance in which children in the LFA group placed the mouth feature in the middle of face, presumably because mouth is considered to be most informative feature for them as it is the least aversive or just as equally important, because of its combined

vocal and lip movement. Another interesting pattern that emerged in all studies, was the poor accuracy on eyes until around 8 years olds in the LFA group, followed by an improvement, possibly because these children overcame their aversive response to eyes or were able to use their featural processing in a more advance way.

## **8.2 DEVELOPMENTAL PROFILES OF DISORDER GROUPS**

Based on findings from the four sets of studies, possible atypical constrains in the development of in each group will be shown in the form of a schematic diagram based on the normal model introduced in Chapter 1. Findings using behavioural measures of face recognition suggest that children in the HFA group do not develop holistic and configural (second-order) face processing and instead use a general featural mode of processing in face recognition. Current data show a novel finding in that the featural processing in the HFA group follows a normal developmental trajectory and is sufficiently powerful to play a compensatory role for other domains of weaker development i.e. configural and holistic processing of faces (Figure 8.1). Note, that use of a red cross does not imply a focal lesion but an atypically developing system with sub-optimal performance.

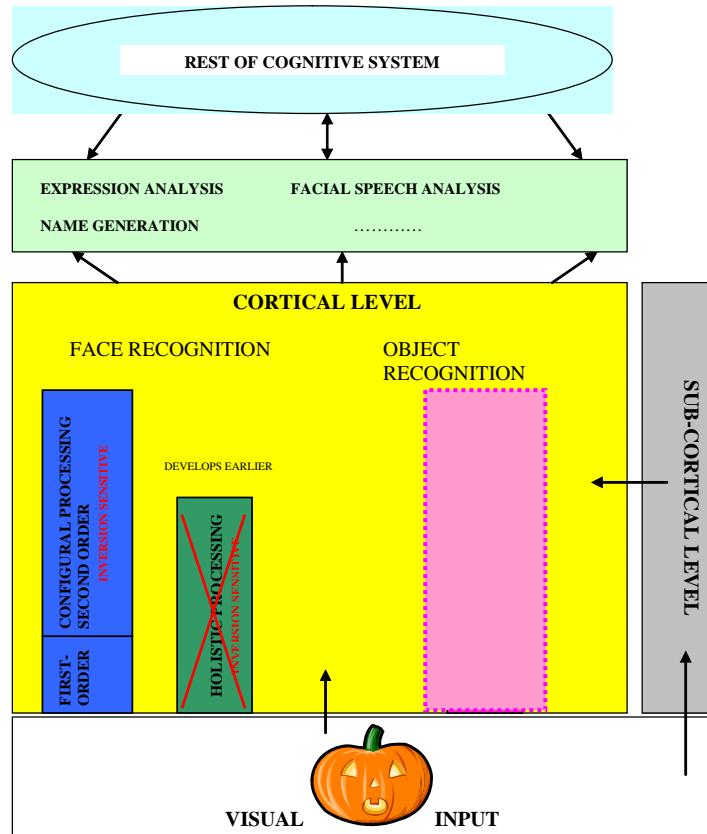


Figure 8.1: Schematic representation of face recognition in HFA group.

Children in the LFA were the only children to demonstrate impairment on the first-order configural processing in the face construction study as well as second-order configural processing. This impairment might be associated with atypical subcortical level due to the lack of orienting to people, which then causes atypical development of face processing at the cortical level (Critchely et al., 2000). Often minimal use of language and lack of social communication could have an impact on normal development of face recognition abilities (Figure 8.2).

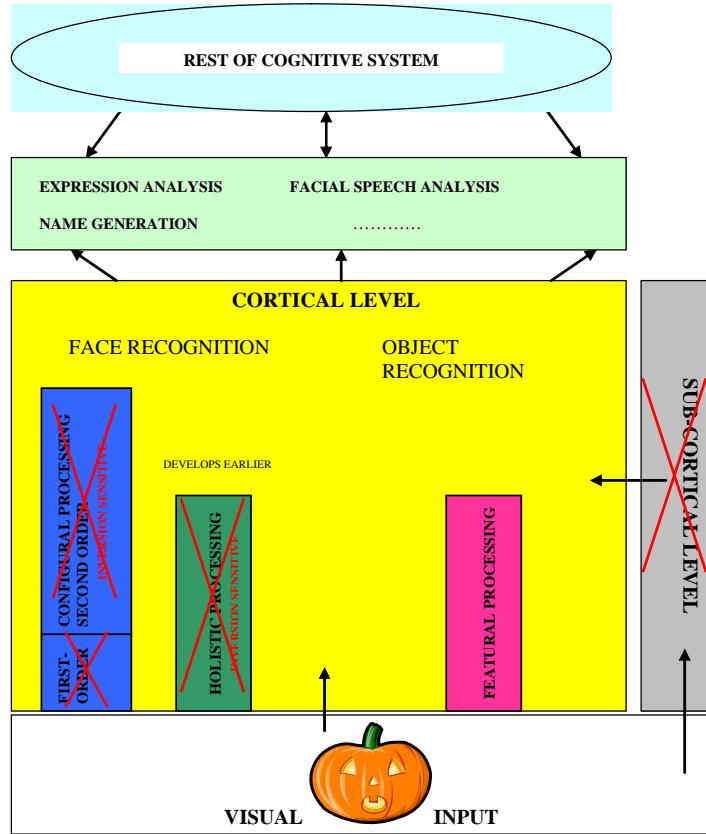


Figure 8.2: Schematic representation of face recognition in LFA group.

In children with DS, the developmental of face recognition exhibited either a delayed onset or rate, or sometimes both, depending on the task. This could be attributed to the generally lower abilities on most of the cognitive skills of this group (Beeghly & Cichetti, 1997; Jarrold & Baddeley, 1997; Laws & Gunn, 2004). However, their basic social orienting does appear to appropriately influence their face development. This is depicted in Figure 8.3.

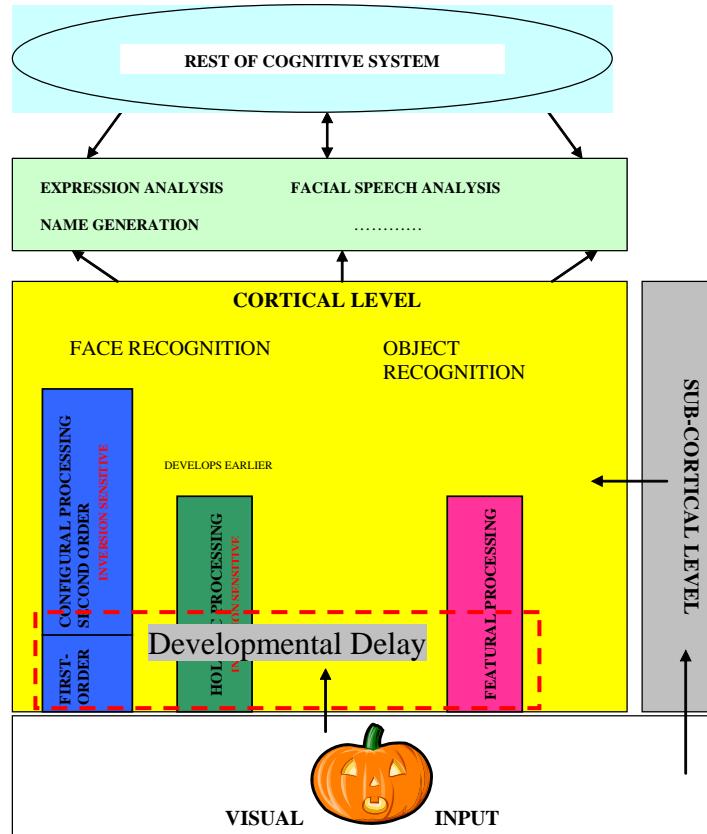


Figure 8.3: Schematic representation of face recognition in DS group. Dashed red lines indicate delay on all the components of visual recognition.

Children in the WBS group demonstrated good performance on face recognition via the featural processing route. Comparable to the HFA group, children with WBS did not show an inversion effect, but unlike the HFA group, they could not use featural processing as a substitute to configural processing for face recognition. However, on the face construction task, children utilised their “good face abilities” to their advantage, so as to compensate for their poor construction skills. This suggests top-down processing in the construction task (see Figure 8.4).

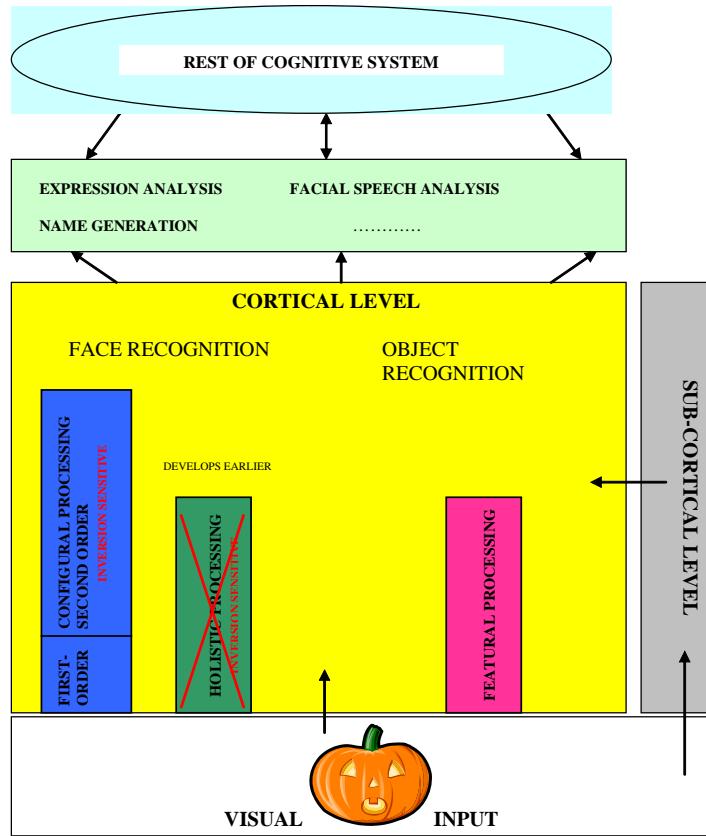


Figure 8.4: Schematic representation of face recognition in WBS group.

In contrast to typically developing individuals who build face representations using a variety of details associated with faces such as names, clothing, emotions, places and relations, children with developmental disorders seem to have impoverished access and processing of information. Children in the autism groups do not develop a stable face representation due to factors such as the lack of sufficient exposure to faces from early infancy. In the case of LFA group it is clear that their social engagement with people is very limited. Moreover, their impairment in emotion recognition has negative effects on development of face recognition. On the other hand, children in the HFA group, may have lesser problems in social communication and larger exposure to faces, and are thus able to use their good feature processing system to recognise faces, but this system is inefficient at recognizing fine differences between people. Thus, it is simply erroneous to assign the differences between the two groups to biologically- and/or cognitively-based attributes. Such differences are multi-factorial and complex in nature,

and environmental contributions must be taken into consideration, such as school environment. Semantic knowledge about faces may also play a role in recognition (Dawson et al., 2005). This lack of preference for faces in autism group might have a cascading effect on shaping category learning by directing attention, and attending to other objects at the expense of faces. In contrast, the WBS group's lack of expertise in faces seems to be of a different nature. Although individuals with WBS typically receive high face input, the quality of such repeated experiences might be inadequate. For example, 'sticky fixation' discussed in chapter 2 may limit the visual scanning pathway of the whole face and narrow the focus to a small area such as the eyes. This notion is in line with the Interactive Specialisation theory that suggested the face-processing system becomes more specialised and localised with environmental exposure to faces (Johnson, 2001, 2005).

### **8.2.1 Can developmental level explain the results?**

The specialization of face processing and its progressive separation from object processing appears to be a product of development, with the face recognition system emerging as a gradual specialisation of an initially more general-purpose system. As discussed in the literature review, maturity of face recognition abilities is directly linked with chronological age development. Several things are worth noting about these age-related data. First, and not unexpectedly, the correlation between CA and face expertise (configural and holistic performance) in the TD group are substantial and have been found in the majority of previous studies of typically developing children (e.g., Brace et al., 2002; Mondloch et al., 2003). Second, in all the current studies with our clinical groups' configural and holistic face recognition did not correlate with CA. Could other background matrices explain our findings? This question requires further exploration of developmental metrics such as BPVS which could aid our theoretical understanding of what elements constitute precursors to face recognition. However, most previous studies have found no significant correlations between face recognition abilities with IQ (Nowicki & Duke, 1994).

In terms of low general intelligence in the DS group it remains to be seen whether face recognition skills are impacted upon domain-general pattern. One way of assessing it would be to recruit children with higher IQ levels and compare their face processing abilities with our DS group.

### **8.2.2 Contribution of developmental trajectories**

The studies examined in the current thesis showed that using the developmental trajectories approach could reveal many important clues about different stages or levels of performance. We illustrated that comparisons of linear regression slopes and intercepts to interpret levels of delay give detailed information about group's developmental changes. These findings indicate that use of term *delay* without additional description is somewhat problematic, as course of development is not static and can change with age. This thesis advocates Karmiloff-Smith approach, which states that in order to understand development we must explore developmental changes right from infancy to adulthood. Let's take the example of WBS group we examined. The early claims about an intact face processing module in WBS are now challenged, not only with respect to the behavioural data themselves, but targeting the underlying cognitive and brain processes involved.

Current investigations of face processing demonstrate that face performance is not at normal levels in any of the clinical groups. It shows that a developmental delay is a very complex process and should not be described as simply *delayed*. Tracing full-developmental trajectories, unlike traditional group comparisons, provide clues to the processes that occur during the development of the face recognition. Also we are able to look at group variability, individual points and make comparisons of certain components of trajectories.

### **8.3 CONCLUSION**

The results presented in this thesis shed light on the similarities and differences between the typically developing children and children with developmental disorders. It also raises important issues regarding the time at which face expertise is acquired. Current findings indicate that children with developmental disorders follow atypical developmental trajectories during face recognition tasks, which depending on the disorder are expressed in different ways. Despite continuous reports of featural processing ‘style’ in individuals with autism and WBS, behavioural data demonstrated that individuals with autism are different from those with WBS (Inter-disorder differences). Additionally, differences within autism can be identified depending on and modulated by the severity of the condition (Intra-disorder differences). This is important for two reasons. First, similarities found on one task at behavioural, genetic or neural levels, do not necessarily correlate with abilities at the other levels. Second, it is essential to consider the role of compensation, interaction and changes over time when describing the behavioural phenotype. In order to reject the assumption that different types of developmental brain damage can result in similar cognitive profiles, a combination of studies at neural, computational and behavioural levels need to be carried out. In summary, findings presented in this thesis are inconsistent with the claim that face perception develops normally in any of these disorders, as shown by behavioural scores.

The current thesis demonstrated that each disorder group showed a different developmental profile during face recognition tasks and these profiles were atypical in different ways. However, only it became obvious from the comparisons of the developmental trajectories of disorder groups to the TD group and by between disorder comparisons. In this regard, the tasks used in the current thesis proved to be sensitive enough to tap the differences between the groups. It was shown that delay of performance should not be simply ignored as unimportant or described as ‘simple’ delay. Studies in this thesis showed that delay itself is dynamic. Sometimes children can

be delayed at first on the task and then improve with age, thus other descriptions are needed to accurately define developmental changes of a group. Data from Studies 1 to 3 pointed to a lack of emergence of inversion in all disorder groups (except for DS in whole-part study). This was shown with respect to the CA as well as on other developmental criteria such as the Benton test and pattern construction task.

While a longitudinal approach would be ideal, use of cross-sectional studies to address developmental trajectories is a good starting point. These enable us to examine patterns and rates of development of particular cognitive abilities at several time-points during development. Currently data are being collected from the same groups of participants on standardised tests to gather longitudinal trajectories.

In summary, many different factors can be implicated in face recognition, some of which will be considered. Current findings are consistent with the notion that experience plays an important role in development of configural and holistic processing (marked by the inversion effect and sensitivity to configurally manipulated faces). Several previous studies reported that individuals with autism and DS have poorer face recognition

abilities than the typically developing children and adults, suggesting that their lack of interest in faces from a very young age may be contributing to their lack of face expertise. In contrast, individuals with WBS seem to experience lots of facial input due to their sociable personalities. It is unclear how these two conflicting dissociations can result in a similar outcome, which culminates in the lack of facial expertise. I am of the opinion that a plausible explanation for the disparities between these disorders is governed by the quantity of visual input as well as its quality.

Relatively little research has been conducted into the role of attention in face perception. One intriguing finding from several studies using simple stimuli indicates that while attention is not required to encode features such as colour or size, it is necessary to combine the features into a percept (e.g. Treisman, 1993). Also, in one of the studies on letter and word perception, it was found that letters were encoded effortlessly, whereas

attention was required to join the letters into words (Treisman & Souther, 1986). Based on these findings, it was proposed that facial feature recognition requires little attention, whereas encoding configural or holistic aspects of faces is attentionally demanding (Palermo and Rhodez, 2002). Although it is believed here that children who participated in the studies were attending to the stimuli presented to them, the possibility of inattention still remains. The only way to exclude this possibility would be if saccadic eye movements are examined.

At the time of writing, none of the behavioural studies examining developmental disorders have controlled for luminance differences within the face category. It may be argued that the potential contrast difference between individual faces may have also contributed to the difference between the groups (Farran, 2005).

One interesting report from real life further supports current findings for the autism population. Several children with autism who took part in this project are currently tested by another research laboratory, in which the Research Assistant is often mistaken for a school teacher (see Figure 8.5), despite the fact that the two only share a common hair outline. Even when children are corrected that this is not Miss J.N., they still continue to mistake these identities. This anecdotal report is consistent with infant data and feature salient hypothesis. Infants are known to use the contour of the face for recognising their mothers (Schwarzer & Massaro, 2001). This lends support to the conclusion that children with LFA have an immature face recognition processing.

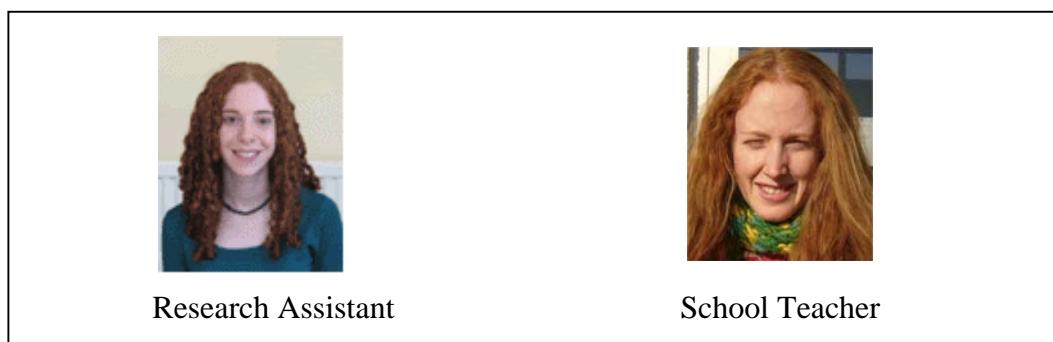


Figure 8.5: Photos of RA and a school teacher. Reproduced with permission.

## **8.4 FURTHER RESEARCH**

There are a number of analyses that were beyond the scope of the current thesis, while others were not investigated due to time constraints and access to participants. Thus, several directions for future research shall be proposed. These would include a repetition of the current studies with a contextual support such as cueing participants to the feature or configuration of the face. It has been suggested that cueing can improve overall performance and enhance holistic processing (Donelly & Davidoff, 1999; Lopez et al., 2004). If children with developmental disorders improve on the tasks when additional help is given it could be incorporated to their school or home programme curriculums.

It is clear that a deeper understanding of the perceptual and construction abilities of face recognition awaits further empirical investigation. This is particularly important given the widespread recommendation of programmes to learn face recognition and emotion. Although the current project highlights problems of individual variability within the clinical groups, and divided the autism population based on their severity of the disorder, based on the current findings in autism groups, there are clearly more things that can be

done in future to improve our understanding of the disorders and to be able to compare findings from different research groups. Such items would include more detailed demographics of individuals and the use of questionnaires such as SVQ, ADOS.

Longitudinal studies of face recognition in developmental disorders are non-existent and it would be important to replicate current studies by testing children at various developmental time points. Ideally, these would obtain a more accurate picture of developmental changes. Examine both behavioral responses, using measures with sensitive tasks, and physiological responses in the same study.

Additional areas for exploration would include determining whether children with autism develop configural processing in recognition of objects other than faces remains

open for verification. One way to examine this process is to use letter configurations, as in the study by Ge et al. (2006).

Current findings do not support the mental rotation theory proposed by Rock (1988). If recognition of inverted faces overtax a mental rotation mechanism, and faces have to be processed by mentally rotating face features one after another, does this mean that children in our disorder groups have a good mental rotation mechanism? If so, how can we explain some of the low performance scores?

The studies and methodological approaches used in this thesis provide compelling evidence advocating for the use of the Neuroconstructivist approach in guiding and interpreting developmental behavioural data. The Neuroconstructivist approach implies that fine examination of developmental changes of supposedly normally developing areas of behavioural functioning may uncover atypical or delayed course of development. Central to this approach is the use of developmental trajectories to focus on changes that occur with chronological age and cross-syndrome comparisons to give multiple perspectives on the constraints that shape normal development and may differ in developmental disorders.

- Abreu, A. M., French, R. M., Annaz, D., Thomas M., & de Schonen, S. (2005). A "visual conflict" hypothesis for global-local visual deficits in Williams syndrome: simulations and data. *Proceedings of the 27th Annual Meeting of the Cognitive Science Society*, Stresa, Italy, 21 July 2005.
- Aguirre, G. K., Zarahn, E., & D'Esposito, M. (1998). An area within human ventral cortex sensitive to "building" stimuli: evidence and implications. *Neuron*, 21(2), 373-383.
- Akshoomoff, N. (2005). The neuropsychology of autistic spectrum disorders. Special issue: Autistic Spectrum Disorders. *Developmental Neuropsychology*, 27(3), 307-310.
- Allison, T., Puce, A., Spencer, D. D., & McCarthy, G. (1999). Electrophysiological studies of human face perception. I: Potentials generated in occipitotemporal cortex by face and non-face stimuli. *Cerebral Cortex*, 9(5), 415-430.
- American Psychology Association (1994). *Diagnostic and statistical manual of mental disorders (4<sup>th</sup> ed.)*. Washington, DC: American Psychiatric Association; 1994.
- Annaz, D. & Karmiloff-Smith, (2005). Cross-syndrome, cross-domain comparisons of developmental trajectories. Commentary on Milne, E., Swettenham, J. & Campbell, R. Motion perception in autism: a review. *Current Psychology of Cognition*, 23(1), 75-83.
- Ansari, D., Donlan, C., Thomas, M., Ewing, S., Peen, T., & Karmiloff-Smith, A. (2003). What makes counting count? Verbal and visuo-spatial contributions to typical and atypical number development. *Journal of Experimental Child Psychology*, 85 (1), 50-62.
- Asperger, H. (1944) Die "Autistischen Psychopathen" im Kindersalter. *Arch. Psychiat. NervKrankh.* 117, 76-136. (English translation in Frith, 1991).
- Atkinson, J., Anker, S., Braddick, O., Nokes, L., Mason, A., & Braddick, F. (2001). Visual and visuo-spatial development in young children with Williams syndrome. *Developmental Medicine and Child Neurology*, 43, 330-337.
- Atkinson, J., King, J., Braddick, O., Nokes, L., Anker, S., & Braddick, F. (1997). A specific deficit of dorsal stream function in Williams' syndrome. *NeuroReport*, 8, 1919-1922.
- Atkinson, J., Braddick, O., Rose, F. E., Searcy, Y. M., Wattam-Bell, J., & Bellugi, U. (in press, 2005). Dorsal-stream motion processing deficits persist into adulthood in Williams syndrome. *Neuropsychologia*.
- Baron-Cohen, S. (2002). The extreme male brain theory of autism. *Trends in Cognitive Science*, 6(6), 248-254.

- Baron-Cohen, S. (2005). Autism and the origins of social neuroscience. In A. Easton and N. Emery (Eds.) *The Cognitive Neuroscience of Social Behaviour*. Psychology Press.
- Baron-Cohen, S., & Hammer, J. (1997). Parents of children with Asperger syndrome: What is the cognitive phenotype? *Journal of Cognitive Neuroscience*, 9, 548 -554.
- Baron-Cohen, S., Ring, H. A., Bullmore, E. T., Wheelwright, S., Ashwin, C., & Williams, S. C. (2000). The amygdala theory of autism. *Neuroscience and Biobehavioral Reviews*, 24, 355–364.
- Baron-Cohen, S., Ring, H. A., Wheelwright, S., Bull-more, E. T., Brammer, M. J., Simmons, A., et al., (1999). Social intelligence in the normal and autistic brain: An fMRI study. *European Journal of Neuroscience*, 11, 1891–1898.
- Baron-Cohen, S., Tager-Flusberg, H. and Cohen, D. (1993). *Understanding other minds: perspectives from autism*. Oxford University Press.
- Barton, J. J., Cherkasova, M. V., Press, D. Z., Intriligator, J. M., & O'Connor, M. (2003). Developmental prosopagnosia: a study of three patients. *Brain and Cognition*, 51(1), 12-30.
- Beeghly, M., & Cichetti, D. (1997). Talking about self and others: Emergence of an internal state lexicon in young children with Down syndrome. *Development and Psychopathology*, 9, 729-748.
- Behrmann, M., Avidan, G., Leonard L. G., Kimchi, R., Luna, B., Humphreys, K., et al. (2005). Configural processing in autism and its relationship to face processing. *Neuropsychological*, 44(1), 110-129.
- Behrmann, M., Moskovitch, M., & Winocur, G. (1994). Intact visual imagery and impaired visual perception in a patient with visual agnosia. *Journal of Experimental Psychology, Human Perception and Performance*, 20(5), 1068-87.
- Bellugi, U., Bahrle, A., Neville, H., Doherty, S., & Jernigan, T. L. (1992). Language, cognition, and brain organization in a neurodevelopmental disorder. In M. Gunnar & C. Nelson (Eds.), *Developmental behavioral neuroscience: The Minnesota Symposia on Child Psychology*. Hillsdale NJ: LEA.
- Bellugi, U., Bahrle, A., Trauner, D., Jernigan, T., & Doherty, S. (1990). Neuropsychological, neurological, and neuroanatomical profile of Williams syndrome children. *American Journal of Medical Genetics (Supplement)*, 6, 115–125.
- Bellugi U., Korenberg, J. R., & Klima, E. S. (2001). Williams syndrome: An exploration of neurocognitive and genetic features. In M. Posner (Ed.) *Journal of Clinical Neurosciences Research, Special Issue 1*, 217-229.

- Bellugi, U., Lichtenberger, L., Jones W., Lai, Z., & St. George, M. (2000). The neurocognitive profile of Williams Syndrome: a complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*, 12(Suppl 1), 7-29.
- Bellugi, U., Lichtenberger, L., Mills, D., Galaburda, A. & Korenberg, J. (1999). Bridging cognition, brain, and molecular genetics: Evidence from Williams syndrome. *Trends in Neurosciences*, 22, 197-207.
- Bellugi, U., Marks, S., Bahrle, A., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams Syndrome. In D. Bishop & K. Mogford (Eds.). *Language development in exceptional circumstances* (pp. 177-189). Hillsdale, NJ: Lawrence Erlbaum.
- Bellugi, U., Wang, P., & Jernigan, T. L. (1994). Williams syndrome: an unusual neuropsychological profile. In S. Broman & J. Grafman (Eds.). *Atypical cognitive deficits in developmental disorders: Implications for brain function* (pp. 23-56). London: Erlbaum.
- Benton, A., Hamsher, K. d. S., Varney, N., & Spreen, O. (1983a). *Benton judgment of line orientation, form H*. NY: Oxford University Press.
- Benton, A., Hamsher, K., Varney, N. R., & Spreen, O. (1983b). *Benton test of facial recognition*. NY: Oxford University Press.
- Benton A. L., Sivan A. B., Hamsher K., Varney N. R., & Spreen, O. (1983). Contribution to neuropsychological assessment. NY: Oxford University Press.
- Berger, J., & Cunningham, C. C. (1983). Development of early vocal behaviours and interactions in Down's syndrome and nonhandicapped infant-mother pairs. *Developmental Psychology*, 14, 44-50.
- Biederman, I. (1987). Recognition by components: A theory of human image understanding. *Psychological Review*, 94, 115-147.
- Bahrle, A. M., Bellugi, U., Delis, D., & Marks, S. (1989). Seeing either the forest or the trees: Dissociation in visuospatial processing. *Brain and Cognition*, 11, 37-49.
- Blair, R.J., Frith, U., Abell, F., & Cipolotti, L. (2002). Fractionation of visual memory: agency detection and its impairment in autism. *Neuropsychologia*, 40(1), 108-118.
- Bodamer, J. (1947). Die Prosopagnosie. *Archives of Psychiatry Nervenkrank*, 179, 6-53.
- Boucher, J. & Lewis, V. (1992). Unfamiliar face recognition in relatively able autistic children. *Journal of Child Psychology and Psychiatry*, 33, 843-859.

- Brace, N. A., Hole, G. J., Kemp, R. I., Pike, G. E., Van Duuren, M., & Norgate, L. (2001). Developmental changes in the effect of inversion: using a picture book to investigate face recognition. *Perception*, 30(1), 85-94.
- Brown, J., Johnson, M. H., Paterson, S., Gilmore, R., Gsödl, M., Longhi, E. et al. (2003) Spatial representation and attention in toddlers with Williams syndrome and Down syndrome. *Neuropsychologia*, 41, 1037-1046.
- Bruce, V. (1988). *Recognising faces*. Hillsdale, NJ: Erlbaum.
- Bruyer, R., & Crispeels, G. (1992). Expertise in person recognition. *Bulletin of the Psychonomic Society*, 30(6), 501-504.
- Cabeza, R. E., & Kato, T. (2000). Features are also important: contributions of featural and configural processing to face recognition. *Psychological Science*, 11, 429-433.
- Calder, A. J., Young, A. W., Keane, J., & Dean, M. (2000). Configurational information in facial expression perception. *Journal of Experimental Psychology, Human Perception and Performance*, 26, 527-551.
- Carey, S., & Diamond, R. (1977). From piecemeal to configurational representation of faces. *Science*, 195(4275), 312-314.
- Carey, S., & Diamond, R. (1994). Are faces perceived as configurations more by adults than by children? *Visual Cognition*, 1(2-3), 253-274.
- Carr, J. (1995). *Down's syndrome: Children growing up*. Cambridge, UK: Cambridge University Press.
- Chung, M. S., & Thomson, D. M. (1995). Development of face recognition. *British Journal of Psychology*, 86(1), 55-87.
- Clahsen, H., & Almazan, M. (1998). Syntax and morphology in Williams syndrome. *Cognition*, 68, 167-198.
- Collishaw, S. M., & Hole, G. J. (2000). Featural and configurational processes in the recognition of faces of different familiarity. *Perception*, 29(8), 893-909.
- Critchley, H. D., Daly, E. M., Bullmore, E. T., Williams, S.C., Van Amelsvoort, T. V., et al., (2000). The functional neuroanatomy of social behaviour : Changes in cerebral blood flow when people with autistic disorder process facial expressions. *Brain*, 123, 2203-2212.
- Curby, K. M., Schyns, P. G., Gosselin, F., & Gauthier, I. (2003). *Face-selective fusiform activation in Asperger's syndrome: A matter of tuning to the right (spatial) frequency*. Poster presented at Cognitive Neuroscience, New York.

- Dall’Oglio, A. M., Milani, L. (1995). Analysis of the cognitive development in Italian children with Williams syndrome. *Genetic Counselling*, 6, 175–176.
- Dalton, K. M, Nacewicz, B. M, Johstone, T., Schaefer, H. S, Grensbacher M. A., Goldsmith H. H., et al., (2005). Gaze fixation and the neural circuitry of face processing in autism. *Nature Neuroscience* 4(8), 519-526.
- Davies, S., Bishop, D., Manstead, A. & Tantam, D. (1994). Face perception in children with autism and Asperger’s syndrome. *Journal of Child Psychology and Psychiatry*, 35, 1033-1057.
- Dawson, G., Carver, L., Meltzoff, A. N., Panagiotides, H., McPartland, J., & Webb, S. J. (2002). Neural correlates of face and object recognition in young children with autism spectrum disorder, developmental delay, and typical development. *Child Development*, 73 (3), 700–717.
- Dawson, G., Meltzoff, A. N., Osterling J., & Rinaldi, J. (1998). Neuropsychological correlates of early symptoms of autism. *Child Development*, 69(5), 1276-85.
- Dawson, G., Osterling, J., Rinaldi, R. Carver, L., & McPartland, J. (2001). Recognition memory and stimulus-reward associations: Indirect support for the role of the ventro-medial prefrontal dysfunction in autism. *Journal of Autism and Developmental Disorders*, 31, 337-341.
- Dawson, G., Webb, S. J., Wijsman, E., Schellenberg, G., Estes, A., Munson, J. et al. (2005). Understanding the Nature of Face Processing Impairment in Autism: Insights from Behavioral and Electrophysiological Studies. *Developmental Neuropsychology*, 27, (3), 403-424.
- Deb, S. (1997) Structural neuroimaging in learning disability. *British Journal of Psychiatry*, 171, 417-9.
- de Gelder, B., Vroomen, J. & van der Heide, L. (1991). Face recognition and lipreading in autism. *European Journal of Cognitive Psychology*, 3, 69-86.
- de Haan, E. H., & Campbell, R. (1991). A fifteen year follow-up of a case of developmental prosopagnosia. *Cortex*, 27(4), 489-509.
- de Haan, M., & Nelson, C. A. (1999). Brain activity differentiates face and object processing in 6-month-old infants. *Developmental Psychology*, 35(4), 1113-1121.
- de Renzi, E. (1986). Current issues in prosopagnosia. In H. D. Ellis & M. A. Jeeves & F. Newcombe & A. W. Young (Eds.), *Aspects of face processing*. Dordrecht: Martinis Nijhoff.
- Deruelle, C., Mancini, J., Livet, M., Cassé-Perrot, C., & de Schonen, S. (1999). Configural and local processing of faces in children with Williams syndrome. *Brain and Cognition*, 41, 276-298.

- Deruelle, C., Rondan, C., Gepner, B., & Tardif.C. (2004). Spatial frequency and face processing in children with autism and Asperger syndrome. *Journal of Autism and Developmental Disorders*, 34(2), 199-210.
- Deruelle, C., Rondan, C., Mancini, J., & Livet, M. O. (in press). Do children with Williams syndrome fail to process visual configural information? *Research in Developmental Disabilities*.
- de Schonen, S., & Mathivet, E. (1989). First come first served. A scenario about development of hemispheric specialisation in face recognition during infancy. *European Bulletin of Cognitive Psychology*, 9, 3-44.
- de Schonen, S., Mancini, J., Camps, R., Maes, E., Laurnet, A. (2005). Early brain lesions and face-processing development. *Developmental Psychobiology*, 46(3), 184-208.
- Diamond, R., & Carey, S. (1977). Developmental changes in the representation of faces. *Journal of Experimental Child Psychology*, 23(1), 1-22.
- Diamond, R., & Carey, S. (1986). Why faces are and are not special: an effect of expertise. *Journal of Experimental Psychology General*, 115(2), 107-117.
- Donnai, D., & Karmiloff-Smith, A. (2000). Williams syndrome: From genotype through to the cognitive phenotype. *American Journal of Medical Genetics: Seminars in Medical Genetics*, 97 (2), 164-171.
- Donnelly, N., & Davidoff, J. (1999). The mental representations of faces and houses: Issues concerning parts and wholes. *Visual Cognition*, 6(3-4), 319-343.
- Duchaine, B. & Nakayama, K. (2004). Developmental prosopagnosia and the Benton Facial Recognition Test. *Neurology*, 62, 1219-1220.
- Dunn L. M., Whetton, C., & Pintilie, D. (1997). *British Picture Vocabulary Scale*. Windsor, UK: NFER-Nelson.
- Dunn, L. M., & Dunn, L. M. (1981). *Peabody Picture Vocabulary Test—Revised*. Circle Pines, MN: American Guidance Services.
- Eimer, M. (2000). Effects of face inversion on the structural encoding and recognition of faces. Evidence from event-related brain potentials. *Brain Research: Cognitive Brain Research*, 10(1-2), 145-158.
- Eimer, M., & McCarthy, R. A. (1999). Prosopagnosia and structural encoding of faces: evidence from event-related potentials. *Neuroreport*, 10(2), 255-259.
- Einfeld, S. L., Tonge, B. J. & Rees, V. W. (2001). Longitudinal course of behavioral and emotional problems in Williams syndrome. *American Journal on Mental Retardation*, 106(1), 73-81.

- Elgar, K., & Campbell, R., (2001). Annotation: the cognitive neuroscience of face recognition: implications for developmental disorders. *Journal of Child Psychology and Psychiatry*, 42(6), 705-17.
- Ellis, H. D., Ellis, D. M., Fraser, W., & Deb, S. (1994). A preliminary study of right hemisphere cognitive deficits and impaired social judgments among young people with Asperger syndrome. *European Child and Adolescent Psychiatry*, 3, 255–266.
- Ellis, H. D. & Young, A. W. (1989). Are faces special? In A. W. Young & H. D. Ellis (Eds.) *Handbook of research on face processing*. Amsterdam, NL: North-Holland.
- Elliot, C.D., Smith, P. and McCulloch, K., (1996). British Ability Scales. NFER-Nelson, Windsor, UK.
- Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., Spallone, P., et al. (1993). Hemizygosity at the elastin locus in a developmental disorder, Williams syndrome. *Nature Genetics*, 5, 11-16.
- Farah, M. J. (1991). Patterns of co-occurrence among the associative agnosia: Implications for visual object representation. *Cognitive Neuropsychology*, 8, 1-19.
- Farah, M. J. (1996). Is face recognition 'special'? Evidence from neuropsychology. *Behavioural Brain Research*, 76(1-2), 181-189.
- Farah, M. J. (2000). *The Cognitive Neuroscience of Vision*. Malden, MA, US: Blackwells.
- Farah, M. J., Tanaka, J. W., & Drain, H. M. (1995). What causes the face inversion effect? *Journal of Experimental Psychology: Human Perception and Performance*, 21(3), 628-634.
- Farah, M. J., Wilson, K. D., Drain, M., & Tanaka, J. N. (1998). What is 'special' about face perception? *Psychological Review*, 105(3), 482-498.
- Farran, E.K. (in press). Perceptual grouping ability in Williams syndrome: Evidence for deviant patterns of performance. *Neuropsychologia*.
- Farran, E. K., & Jarrold, C. (2002). Visuo-spatial cognition in Williams syndrome: Reviewing and accounting for the strengths and weaknesses in performance. *Developmental Neuropsychology*, 23, 175-202.
- Farran, E. K., Jarrold, C., & Gathercole, S. E. (2001). Block design performance in the Williams syndrome phenotype: a problem with mental imagery. *Journal of Child Psychology and Psychiatry*, 42, 719–728.

- Farran, E. K., Jarrold, C., & Gathercole, S. E. (2003). Divided attention, selective attention and drawing: processing preferences in Williams syndrome are dependent on the task administered. *Neuropsychologia*, 41, 676-687.
- Fidler, D. J. (2005). The emerging Down syndrome behavioural phenotype in early childhood. *Infants & Young Children*, 18(2), 86–103.
- Fiorentini, A., Maffei, L., & Sandini, G. (1983). The role of high spatial frequencies in face perception. *Perception*, 12, 195-201.
- Flin, R. H. (1985). Development of face recognition: An encoding switch? *British Journal of Psychology*, 76, 123–134.
- Folstein, S. E., Santangelo, S. L., Gilam, S. E., Piven, J., Landa, R., Lainhart, J., et al. (1999). Predictors of cognitive test patterns in autism families. *Journal of Child Psychology and Psychiatry*, 40(7), 1117-1128.
- Fombonne, E. (2003). The epidemiological surveys of autism and other pervasive developmental disorders: an update. *Journal of Autism and Developmental Disorders*, 33 (4), 395-402.
- Frangiskakis, J. M., Ewart, A., Morris, C. A., Mervis, C. B., Bertrand, J., Robinson, et al., (1996). LIM-kinase1 hemizygosity implicated in impaired visuospatial constructive cognition. *Cell*, 86, 59-69.
- Freire, A., & Lee, K. (2001). Face recognition in 4- to 7-year-olds: processing of configural, featural, and paraphernalia information. *Journal of Experimental Child Psychology*, 80(4), 347-371.
- Freire, A., Lee, K., & Symons, L. A. (2000). The face-inversion effect as a deficit in the encoding of configural information: direct evidence. *Perception*, 29(2), 159-170.
- Frith, U. (1989). *Autism: Explaining the enigma*. Oxford: Basil Blackwell.
- Frith, U. (2003). *Autism: Explaining the enigma*. UK: Blackwell Publishers.
- Frith, U. (2004). Emanuel Miller lecture: confusions and controversies about Asperger syndrome. *Journal of Child Psychology and Psychiatry*, 45, 672-686.
- Ganz, J. & Simpson, R. (2004). Effects on communicative requesting and speech development of the Picture Exchange Communication System in children with characteristics of autism. *Journal of Autism and Developmental Disabilities*, 34, 395-409.
- Gauthier, I., & Logothetis, N. K. (2000). Is face recognition not so unique after all? *Cognitive Neuropsychology*, 17(1-3), 125-142.
- Gauthier, I., & Nelson, C. A. (2001). The development of face expertise. *Current Opinion in Neurobiology*, 11(2), 219-224.

- Gauthier, I., & Tarr, M. J. (1997). Becoming a 'Greeble' expert: exploring mechanisms for face recognition. *Vision Research*, 37(12), 1673-1682.
- Gauthier, I., Tarr, M. J., Anderson, A. W., Skudlarski, P., & Gore, J. C. (1999). Activation of the middle fusiform 'face area' increases with expertise in recognizing novel objects. *Nature Neuroscience*, 2(6), 568-573.
- Gauthier, I., Tarr, M. J., Moylan, J., Skudlarski, P., Gore, J. C., & Anderson, A. W. (2000). The fusiform 'face area' is part of a network that processes faces at the individual level. *Journal of Cognitive Neuroscience*, 12(3), 495-504.
- Ge, L., Wang, Z., McCleery, J. P., & Lee, K. (2006, in press). The effect of face expertise activation on the inversion effect. *Psychological Science*.
- Gillberg, C., & Coleman, M., (2000). *The Biology of the Autistic Syndromes*. Mac Keith Press.
- Gray, V., Karmiloff-Smith, A., Funnell, E. & Tassabehji, M. (in press, 2005). In-depth Analysis of Spatial Cognition in Williams Syndrome: A Critical Assessment of the Role of the LIMK1 gene. *Neuropsychologia*.
- Grelotti, D. J., Klin, A. J., Gauthier, I., Skudlarski, P., Cohen, D. J., Gore, J. C., et al. (2005). fMRI activation of the fusiform gyrus and amygdala to cartoon characters but not to faces in a boy with autism. *Neuropsychologia*, 43 (3), 373–385.
- Grice, S. J., Spratling, M. W., Karmiloff-Smith, A., Halit, H., Csibra, G., de Haan, M., et al. (2001). Disordered visual processing and oscillatory brain activity in autism and Williams syndrome. *Neuroreport*, 12(12), 2697-2700.
- Grice, S. J., Halit, H., Farroni, T., Baron-Cohen, S., Bolton, P., & Johnson, M. H. (2005). Neural correlates of eye-gaze detection in young children with autism. *Cortex*, 41(3), 342-53.
- Gross, C. G., Bender, D. B., & Rocha-Miranda, C. E. (1969). Visual receptive fields of neurons in inferotemporal cortex of the monkey. *Science*, 166(910), 1303-1306.
- Gross, C. G., Rocha-Miranda, C. E., & Bender, D. B. (1972). Visual properties of neurons in inferotemporal cortex of the Macaque. *Journal of Neurophysiology*, 35(1), 96-111.
- Gunn, P., & Cuskelly, M. (1991). Down syndrome temperament: The stereotype at middle childhood and adolescence. *International Journal of Disability, Development, and Education*, 38, 59–70.
- Gunn, P., Berry, P., & Andrews, R. J. (1982). Looking behaviour of Down syndrome infants. *American Journal of Mental Deficiency*, 87, 344–347.

- Hadjikani, N., Chabris, C. F., Joseph, R. M., Clark, J., McGrath, L., Aharon, I., et al. (2004). Early visual cortex organization in autism - An fMRI study. *Neuroreport*, 15 (2), 267–270.
- Happé, F. (2000). Parts and wholes, meaning and minds : Central coherence and its relation to theory of mind. In S. Baron-Cohen, H. Tager-Flusberg, & D. J. Cohen (Eds.), *Understanding Other Minds: Perspective from Autism* (2<sup>nd</sup> edition). Oxford: Oxford University Press.
- Happé, F., Malhi, G., & Checkley, S. (2001). Acquired mind-blindness following frontal lobe surgery? A single case study of impaired ‘theory of mind’ in a patient treated with stereotactic anterior capsulotomy. *Neuropsychologia*, 39, 83–90.
- Harmon, L. D., & Julesz, B. (1973). Masking in visual recognition: effects of two dimensional filtered noise. *Science*, 180, 1194.
- Haxby, J. V., Hoffman, E. A., & Gobbini, M. I. (2000). The distributed human neural system for face perception. *Trends in Cognitive Sciences*, 4(6), 223-233.
- Haxby, J. V., Horwitz, B., Ungerleider, L. G., Maisog, J. M., Pietrini, P., & Grady, C. L. (1994). The functional organization of human extrastriate cortex: a PET-rCBF study of selective attention to faces and locations. *Journal of Neuroscience*, 14(11), 6336-6353.
- Haxby, J. V., Ungerleider, L. G., Clark, V. P., Schouten, J. L., Hoffman, E. A., & Martin, A. (1999). The effect of face inversion on activity in human neural systems for face and object perception. *Neuron*, 22(1), 189-199.
- Hay, D. C., & Cox, R. (2000). Developmental changes in the recognition of faces and facial features. *Infant and Child Development*, 9, 199-212.
- Heaton, P., Hermelin, B., & Pring, L. (1998). Can children with autistic spectrum disorders perceive affect in music? An experimental investigation. *Psychological Medicine*, 29(6), 1405-10.
- Hole, G. J. (1994). Configurational factors in the perception of unfamiliar faces. *Perception*, 23(1), 65-74.
- Hobson, R. P. (1986). The autistic child’s appraisal of expressions of emotion: A further study. *Journal of Child Psychology and Psychiatry*, 27(5), 671–680.
- Hobson, R. P. (1987). The autistic child’s recognition of age- and sex- related characteristics of people. *Journal of Autism and Developmental Disorder*, 17 (1), 63-79.

- Hobson, P., & Lee, A. (1989). Emotion-related and abstract concepts in autistic people: evidence from the British Picture Vocabulary Scale. *Journal of Autism and Developmental Disorders*, 19(4), 601-23.
- Hobson, P., Ouston, J. & Lee, A. (1988). What's in a face? The case of autism. *British Journal of Psychology*, 79, 441-453.
- Hoffman, J. E., Landau, B., & Pagani, B. (2003). Spatial breakdown in spatial construction: Evidence from eye fixations in children with Williams syndrome. *Cognitive Psychology*, 46, 260–301.
- Howard, M. A., Cowell, P. E., Boucher, J., Broks, P., Mayes, A., & Farrant, A. (2000). Converging neuroanatomical and behavioural evidence of an amygdale hypothesis of autism. *NeuroReport*, 11, 2931-2935.
- Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry and Allied Disciplines*, 39, 183-189.
- Hughes, C., Leboyer, M. & Bouvard, M. (1997). Executive function in parents of children with autism. *Psychological Medicine*, 27, 209–220.
- Hutcheson, H., Bradford , S. E., Folstein, M. B., Gardiner, S. L., Santangelo, J. S., Sutcliffe , J. L. et al. (2003). Defining the autism minimum candidate gene region on chromosome 7. *American Journal of Medical Genetics Part B: Neuropsychiatric Genetics*, 117B (1), 90 – 96.
- Jamain, S., Quach, H., Betancur, C., Rastam, M., Colineaux, C., Gillberg, I. C. et al. (2003). Mutations of the X-linked genes encoding neuroligins NLGN3 and NLGN4 are associated with autism. *Nature Genetics*, 34(1), 27-29.
- Jarrold, C., & Baddeley, A. D. (1997). Short-term memory for verbal and visuospatial information in Down's syndrome. *Cognitive Neuropsychiatry*, 2, 101–122.
- Jarrold, C., Baddeley, A. D., & Hewes, A. K. (1999). Genetically dissociated components of working memory: Evidence from Down's and Williams syndrome. *Neuropsychologia*, 37, 637–651.
- Jarrold, C., Baddeley, A. D., & Phillips, C. E. (2002). Verbal short-term memory in Down syndrome: A problem of memory, audition, or speech? *Journal of Speech, Language, and Hearing Research*, 45, 531–544.
- Jarrold, C., & Brock, J. (2004). To match or not to match? Methodological issues in autism-related research. *Journal of Autism and Developmental Disorders*, 34, 81-86.
- Johnson, M. H. (2001). Functional brain development in humans. *Nature Reviews Neuroscience*, 2, 475-483.

- Johnson, M. H. (2005). *Developmental cognitive neuroscience (2<sup>nd</sup> Edition)*. Oxford : Blackwell.
- Johnson, M., H., & de Haan, M. (2001). Developing cortical specialization for visual-cognitive function: The case of face recognition. In J. L. McClelland & R. S. Siegler (Eds.), *Mechanisms of cognitive development: Behavioral and neural perspectives. Carnegie Mellon symposia on cognition* (pp. 253-270). Mahwah, NJ, US: Lawrence Erlbaum Associates, Inc.
- Johnson, M. H., Dziurawiec, S., Ellis, H., & Morton, J. (1991). Newborns' preferential tracking of face-like stimuli and its subsequent decline. *Cognition*, 40(1-2), 1-19.
- Jolliffe, T. & Baron-Cohen, S. (1997). Are people with autism and Asperger syndrome faster than normal on the Embedded Figures Test? *Journal of Child Psychology and Psychiatry*, 38, 527-534.
- Joseph, R. & Tanaka, J. (2003). Holistic and part-based face recognition in children with autism. *Journal of Child Psychology and Psychiatry*, 44 (4), 529-542.
- Kaiser, A. P., Virji-Babul, Iarocci, McLaughlin, P., & Tanaka, J. (2005). *Selective strategies in face processing in Down syndrome*. Poster presented at Cognitive Neuroscience Symposium. New York.
- Kanner, L. (1943). Autistic disturbance of affective contact. *Nervous Child*, 2, 217 - 240.
- Kanwisher, N., McDermott, J., & Chun, M. M. (1997). The fusiform face area: a module in human extrastriate cortex specialized for face perception. *Journal of Neuroscience*, 17(11), 4302-4311.
- Kanwisher, N., Tong, F., & Nakayama, K. (1998). The effect of face inversion on the human fusiform face area. *Cognition*, 68(1), 1-11.
- Karmiloff-Smith, A. (1997). Crucial differences between developmental cognitive neuroscience and adult neuropsychology. *Developmental Neuropsychology*, 13 (4), 513-524.
- Karmiloff-Smith, A. (1998). Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences*, 2 (10), 389-398.
- Karmiloff-Smith, A., Scerif, G., & Ansari, D. (2003). Double dissociations in developmental disorders? Theoretically misconceived, empirically dubious. *Cortex*, 39, 161-163.
- Karmiloff-Smith, A., Scerif, G., & Thomas, M. S. C. (2002). Different approaches to relating genotype to phenotype in developmental disorders. *Developmental Psychobiology*, 40, 311-322.

- Karmiloff-Smith, A., Smith, A., Chin, E., Annaz, D., Elsabbagh, M., Gilchrist, I. D., et al., (submitted). Of mice and men. Genetic contributions to spatial cognition.
- Karmiloff-Smith, A. & Thomas, M. S. C. (2003). Developmental disorders. In M. A. Arbib (Ed.) *The Handbook of brain theory and neural networks, 2nd Edition*. Cambridge, MA: MIT Press.
- Karmiloff-Smith, A. & Thomas, M. (2003). What can developmental disorders tell us about the neurocomputational constraints that shape development? The case of Williams syndrome. *Development and Psychopathology*, 15, 969-990.
- Karmiloff-Smith, A., Thomas, M., Annaz, D., Humphreys, K., Ewing, S., Brace, N., et al. (2004). Exploring the Williams Syndrome Face Processing Debate: The importance of building developmental trajectories. *Journal of Child Psychology and Psychiatry*, 45(7), 1258-1274.
- Karmiloff-Smith, A., Tyler, L. K., Voice, K., Sims, K., Udwin, O., Howlin, P., et al. (1998). Linguistic dissociations in Williams syndrome: evaluating receptive syntax in on-line and off-line tasks. *Neuropsychologia*, 36, 343-351.
- Karrer, J. H., Karrer, R., Bloom, D., Chaney, L., & Davis, R. (1998). Event-related brain potentials during an extended visual recognition memory task depict delayed development of cerebral inhibitory processes among 6-month-old infants with Down syndrome. *International Journal of Psychophysiology*, 29, 167-200.
- Klein, B. P., & Mervis, C. B. (1999). Contrasting patterns of cognitive abilities of 9- and 10-year-olds with Williams syndrome or Down syndrome. *Developmental Neuropsychology*, 16, 177-196.
- Klin, A., Jones, W., Schultz, R., Volkmar, F., & Cohen, D. (2002). Visual fixation patterns during viewing of naturalistic social situations as predictors of social competence in individuals with autism. *Archives Gen Psychiatry*, 59(9), 809-816.
- Klin, A., Sparrow, S., de Bildt, A., Cichetti, D. V., Cohen, D. J., & Volkmar, F. R. (1999). A normed study of face recognition in autism and related disorders. *Journal of Autism Developmental Disorders* 29(6), 499-508.
- Korenberg, J. R. (1991). Down syndrome phenotypic mapping. *Progress in Clinical and Biological Research*, 373, 43-52.
- Korenberg, J. R., Chen, X. N., Hirota, H., Lai, Z., Bellugi, U., Burian, D., et al. (2000). Genome structure and cognitive map of Williams syndrome. *Journal of Cognitive Neuroscience*, 12(Suppl), 89-105.
- Korvatska, E., Van de Water, J., Anders, T. F., & Gershwin, M. E. (2002). Genetic and immunologic considerations in autism. *Neurobiology of Disease*. 9(2), 107-25.

- Kovács, I., Kozma, P., Feher, A., & Benedek, G. (1999). Late maturation of visual spatial integration in humans. *Proceedings of the National Academy of Science USA*, 96(21), 12204-12209.
- Laing, E., Grant, J., Thomas, M. S . C. & Karmiloff-Smith, A. (2005). Love is . . . an abstract word: The influence of phonological and semantic factors on verbal short-term memory in Williams syndrome. *Cortex*, 41 (2), 169-179.
- Laing, E., Hulme, C., Grant, J., & Karmiloff-Smith, A. (2001). Learning to read in Williams syndrome: Looking beneath the surface of atypical reading development. *Journal of Child Psychology and Psychiatry and Allied Disciplines*, 42, 729-739.
- Lakusta, L., & Landau, L. (2005). Starting at the end: the importance of goals in spatial language. *Cognition*, 96(1), 1-33.
- Langdell, T. (1978). Recognition of faces: An approach to the study of autism. *Journal of Child Psychology and Psychiatry* 19, 255-268.
- Laws, G. (1998). The use of nonword repetition as a test of phonological memory in children with Down syndrome. *Journal of Child Psychology and Psychiatry and Allied Disciplines*, 39, 1119-1130.
- Laws, G. & Gunn, D. (2004). Phonological memory as a predictor of language comprehension in Down syndrome: a five-year follow-up study. *Journal of Child Psychology and Psychiatry*, 45(2), 326-37.
- Le Grand, R., Mondloch, C. J., Maurer, D., & Brent, H. P. (2003). Early face processing requires visual input to the right hemispheres during infancy. *Nature Neuroscience*, 6(10), 1108-1112.
- Le Grand, R., Mondloch, C. J., Maurer, D., & Brent, H. P. (2001). Early visual experience and face processing. *Nature*, 410(6831), 890.
- Leder, H. & Bruce, V. (1998). Local and relational aspects of face distinctiveness. *Quarterly Journal of Experimental Psychology Part A*, 51(3), 449-73.
- Leder, H., & Bruce, V. (2000). When inverted faces are recognized: the role of configural information in face recognition. *Quarterly Journal of Experimental Psychology: A*, 53(2), 513-536.
- Leder, H., Candrian, G., Huber, O., & Bruce, V. (2001). Configural features in the context of upright and inverted faces. *Perception*, 30(1), 73-83.
- Leekam S., Baron-Cohen, S Perrett, D. & Milders, M. (1997). Eye-direction detection: A dissociation between geometric and joint attention skills in autism. *British Journal of Developmental Psychology*, 15, 77-95.

- Leekam, S. R., Hunnissett, E., & Moore, D. (1998). Targets and cues: gaze-following in children with autism. *Journal of Child Psychology and Psychiatry*, 39(7), 951-62.
- Lewis, M. B. & Glenister, T. E. (2003). A sideways look at configural encoding: Two different effects of face rotation. *Perception*, 32, 7-14.
- López, B., Donnelly, N., Hadwin, J. A., & Leekam, S. R. (2004). Face processing in high-functioning adolescents with autism: Evidence for weak central coherence. *Visual Cognition*, 11(6), 676-688.
- Magiati, I., & Howlin, P. (2003). A pilot evaluation study of the Picture Exchange Communication System (PECS) for children with autistic spectrum disorders. *The International Journal of Autism*, 7(3), 297-320.
- Mareschal, D., Johnson, M. H., Sirios, S., Spratling, M., Thomas, M. S. C., & Westermann, G. (forthcoming). *Neuroconstructivism: How the brain constructs cognition*. Oxford: Oxford University Press.
- Maurer, D., Le Grand, R., & Mondloch, C. J. (2002). The many faces of configural processing. *Trends in Cognitive Sciences*, 6(6), 255-260.
- McPartland, J., Dawson, G., Webbs, S. J., Panagiotides, H., & Carver, L. J. (2004). Event-related brain potentials reveal anomalies in temporal processing of faces in autism spectrum disorder. *Journal of Child Psychology and Psychiatry*, 45(7), 1235-45.
- Medical Research Council. (2001). Review of autism research: Epidemiology and causes.
- Mervis, C. B., & Bertrand, J. (1997). Developmental relations between cognition and language: Evidence from Williams syndrome. In L. B. Adamson & M. A. Romski (Eds.), *Research on communication and language disorders: Contributions to theories of language development* (pp.75-106). New York: Brookes.
- Mervis, C. B., & Klein-Tasman, B. P. (2000). Williams syndrome: cognition, personality, and adaptive behavior. *Mental Retardation and Developmental Disabilities Research Reviews* 6(2), 148-58.
- Mervis, C. B., Morris, C. A., Bertrand, J., & Robinson, B. F. (1999). Williams Syndrome: Findings from an integrated program of research. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders: Contributions to a new framework from the cognitive neurosciences*. Cambridge, MA: MIT Press.
- Miller, J. F. (1999). Profiles of language development in children with Down syndrome. In J. Miller, M. Leddy, & L. A. Leavitt (Eds.), *Improving the communication of people with Down syndrome* (pp. 11-40). Baltimore: Brookes Publishing.

- Mills, D., Alvarez, T., St. George, M., Appelbaum, L., Bellugi, U., & Neville, H. (2000). Electrophysiological studies of face processing in Williams syndrome. *Journal of Cognitive Neuroscience, 12: Supplement*, 47-64.
- Minshew, N. J., Goldstein, G., & Siegel, D. J. (1997). Neuropsychological functioning in autism: profile of a complex information processing disorder. *Journal of International Neuropsychological Society, 3(4)*, 303-316.
- Mobbs, D., Garrett, A., Menon, V., Rose, F., Bellugi, U. & Reiss, A. (2004). Anomalous brain activation during face and gaze processing in Williams syndrome. *Neurology, 62*, 2070-2076.
- Mondloch, C. J., Le Grand, R., & Maurer, D. (2002). Configural face processing develops more slowly than featural face processing. *Perception, 31(5)*, 553-566.
- Mooney, C. M. (1957). Closure as affected by configural clarity and contextual consistency. *Canadian Journal of Psychology, 11*, 80-88.
- Morris, C. A., Demsey, S. A., Leonard, C. O., Dilts, C. & Blackburn, B. L. (1988). The natural history of Williams syndrome: physical characteristics. *Journal of Pediatrics 113*, 318-326.
- Morris, C. A., Mervis, C. B., Hobart, H. H., Gregg, R. G., Bertrand, J., Ensing, G. J. et al. (2003). GTF2I hemizygosity implicated in mental retardation in Williams syndrome: genotype-phenotype analysis of five families with deletions in the Williams syndrome region. *American Journal of Medical Genetics Part A, 123(1)*, 45-59.
- Morrison, A., Annaz, D., & Thomas, M. (2005). The development of visuo-spatial skills in children with Williams syndrome. Poster presented at Birkbeck, University of London.
- Morton, J. (2004). Understanding Developmental Disorders: *A Causal Modelling Approach*. Blackwell Publishing.
- Morton, J., & Johnson, M. H. (1991). Conspec and conlearn: A two process theory of infant face recognition. *Psychological Review, 98*, 164–181.
- Moscovitch, M., Winocur, G. & Behrmann, M. (1997). What is special about face recognition? Nineteen experiments on a person with visual object agnosia and dyslexia but normal face recognition. *Journal of Cognitive Neuroscience, 9*, 555-604.
- Mottron, L., Belleville, S., & Menard, E., (1999). Local bias in autistic subjects as evidenced by graphic tasks: perceptual hierarchization or working memory deficit? *Journal of Child Psychology and Psychiatry, 40(5)*, 743-55.

- Mottron, L., Burack, J., Iarocci, G., Belleville, S., Enns, J. (2003). Locally oriented perception with intact global processing among adolescents with high functioning autism: evidence from multiple paradigms. *Journal of Child Psychology and Psychiatry*, 44(6), 906-913.
- Muhle, R., Trentacoste S. V., & Rapin, I., (2004). The genetics of autism. *Pediatrics* 113, 472-486.
- Navon, D. (1977). Forest before trees: The precedence of global features in visual perception. *Cognitive Psychology*, 9, 353-383.
- Nelson, C.A. (2001). The development and neural bases of face recognition. *Infant and Child Development*, 10, 3-18.
- Neville, H. J. (in press, 2005). Development and plasticity of human cognition. In U. Mayr, E. Awh & S. Keele (Eds.). *Developing individuality in the human brain: A tribute to Mike Posner*. Washington DC: APA Books.
- Neville, H. J., Mills, D. L., & Bellugi, U. (1994). Effects of altered auditory sensitivity and age of language acquisition on the development of language-relevant neural systems: Preliminary studies of Williams Syndrome. In S. Broman (Ed.). *Cognitive Deficits in Developmental Disorders: Implications for Brain Function* (pp. 67-83). Hillsdale, NJ: Lawrence Erlbaum Associates.
- Njiokiktjien, C., Verschoor, A., de Sonneville, L., Huyser, C., Op het Veld, V., & Toorenar, N. (2001). Disordered recognition of facial identity and emotions in three asperger type autists. *European Child and Adolescent Psychiatry*, 10(1), 79-90.
- Norton, P. & Drew, C. (1994). Autism and potential family stressors. *The American Journal of Family Therapy*, 22, 67-76.
- Osterling, J., & Dawson, G. (1994). Early recognition of children with autism: a study of first birthday home videotapes. *Journal of Autism and Developmental Disorders*, 24(3), 247-257.
- Palermo, R. & Rhodes, G. (2002). The influence of divided attention on holistic face perception. *Cognition*, 82(3), 225-257.
- Pani, J. R., Mervis, C. B., & Robinson, B. F. (1999). Global spatial organization by individuals with Williams syndrome. *Psychological Science*, 10, 453-8.
- Pary, R. J. (1989). Pretreatment systolic orthostatic blood pressure depression in Down's syndrome. *Journal of Clinical Psychopharmacology*, 9(2), 146-7.
- Passarotti, A. M., Paul, B. M., Bussiere, J. R., Buxton, R. B., Wong, E. C., & Stiles, J. (2003). The development of face and location processing: An fMRI study. *Developmental Science*, 6(1), 100-117.

- Passarotti, A., Paul, B., Rudiak-Gould, P. & Stiles, J. (2001). Development affects the ventral and dorsal processing streams differently: a fMRI study on face and location processing in children, teenagers and adults. *Neuroimage*, 13(6), S345.
- Paterson, S. J., Brown, J. H., Gsodl, M. K., Johnson, M. H., & Karmiloff-Smith, A. (1999). Cognitive modularity and genetic disorders. *Science*, 286(5448), 2355-2358.
- Pellicano, L., & Rhodes, G. (2003). Holistic processing of faces in preschool children and adults. *Psychological Science*, 14, 618-622.
- Pelphrey, K. A., Sasson, N. J., Reznick, J. S., Paul, G., Goldman, B. D & Piven, J., (2002). Visual scanning of faces in autism. *Journal of Autism and Developmental Disorders*, 32(4), 249-61.
- Perrett, D. I., Oram, M. W., & Ashbridge, E. (1998). Evidence accumulation in cell populations responsive to faces: an account of generalisation of recognition without mental transformations. *Cognition*, 67(1-2), 111-145.
- Perrett, D. I., Oram, M. W., Hietanen, J. K., & Benson, P. J. (1994). Issues of representation in object vision. In M. J. Farah & G. Ratcliff (Eds.). *The neuropsychology of high-level vision: Collected tutorial essays. Carnegie Mellon symposia on cognition*. (pp. 33-61). Hillsdale, NJ: Lawrence Erlbaum Associates, Inc.
- Pezzini, G., Vicari, S., Volterra, V., Milani, L., & Ossella, M. T. (1999). Children with Williams Syndrome: Is there a single neuropsychological profile? *Developmental Neuropsychology*, 15(1), 141-155.
- Pierce, K., Muller, R. A., Ambrose, J., Allen, G., & Courchesne, E. (2001). Face processing occurs outside the fusiform 'face area' in autism: evidence from functional MRI. *Brain*, 124(10), 2059-2073.
- Plaisted, K., O'Riordan, M., & Baron-Cohen, S. (1998). Enhanced discrimination of novel, highly similar stimuli by adults with autism during a perceptual learning task. *Journal of Child Psychology and Psychiatry*, 39(5), 765-75.
- Plaisted, K., Swettenham, J., Rees, L. (1999). Children with autism show local precedence in a divided attention task and global precedence in a selective attention task. *Journal of Child Psychology and Psychiatry*, 40(5), 733-742.
- Pléh, C., Lukács, Á., & Racsmány, M. (2003). Morphological patterns in Hungarian children with Williams syndrome and the rule debate. *Brain and Language*, 86(3), 377-83.
- Plesa-Skwerer, D., Sullivan, K., Joffre, K., & Tager-Flusberg, H. (2006). Self Concept in People with Williams Syndrome and Prader-Willi Syndrome. *American Journal of Mental Retardation*, 111(1), 15-26.

- Proschel, C., Blouin, M.-J., Gutowski, N. J., Ludwig, R., & Noble, M. (1995). LIMK1 is predominantly expressed in neural tissues and phosphorylates serine, threonine and tyrosine residues in vitro. *Oncogene*, 11, 1271-1281.
- Rakover, S. S. & Teucher, B. (1997). Facial inversion effects: parts and whole relationship. *Perception & Psychophysics*, 59(5), 752-61.
- Rakover, S. S. (2002). Featural vs. configurational information in faces: A conceptual and empirical analysis. *British Journal of Psychology*, 93(1), 1-30.
- Rellini, E., Tortolani, D., Trillo, S., Carbone, S., & Montecchi, F. (2004). Childhood Autism Rating Scale (CARS) and Autism Behavior Checklist (ABC) correspondence and conflicts with DSM-IV criteria in diagnosis of autism. *Journal of Autism and Developmental Disorders*, 34 (6), 703-708.
- Robinson, W. P., Waslynka, J., Bernasconi, F., Wang, M., Clark, S., Kotzot, D., et al. (1996). Delineation of 7q11.2 deletions associated with Williams-Beuren syndrome and mapping of a repetitive sequence to within and to either side of the common deletion. *Genomics*, 34, 17-23.
- Roizen N. J. & Patterson, D. (2003). Down's syndrome. *The Lancet* 36, 1281-1289.
- Rhodes, G. (1993). Configural coding, expertise, and the right hemisphere advantage for face recognition. *Brain and Cognition*, 22, 19-41.
- Rock, I. (1988). On Thompson's inverted-face phenomenon (Research note). *Perception*, 17, 815-817.
- Rolls, E. T. (2004). Convergence of sensory systems in the orbitofrontal cortex in primates and brain design for emotion. Anatomical Records (Part A) *Discoveries in Molecular, Cellular and Evolution Biology*, 281(1), 1212-25.
- Rolls, E. T., & Tovee, M. J. (1994). Processing speed in the cerebral cortex and the neurophysiology of visual masking. *Proceedings in Biological Science*, 257(1348), 9-15.
- Rondan, C., Mancini, J., Livet, M. O., & Deruelle, C. (2003). Perceptual and visuo constructive performance in children with Williams syndrome. *Cognition, Brain, Behavior*, 7, 149–157.
- Rouse, H., Donnelly, N., Hadwin, J. A., & Brown, T. (2004). Do children with autism perceive second-order relational features? The case of the Thatcher illusion. *Journal of Child Psychology and Psychiatry*, 45(7), 1246–1257.
- Rutter, M. (1972). Childhood schizophrenia reconsidered. *Journal of Autism and Childhood Schizophrenia*, 2, 315–38.

- Saemundsen, E., Magnusson, P., Smari, J., & Sigurdardottir, S. (2003) Autism Diagnostic Interview-Revised and the Childhood Autism Rating Scale: Convergence and Discrepancy in Diagnosing Autism. *Journal of Autism and Developmental Disorders*, 33 (3), 319-329.
- Sangrioli, S., & de Schonen, S. (2002). *Visual experience in face processing development in children from 11 months of age to 6 years*. Paper presented at the ESF conference, Maratea.
- Schopler, E., Reichler, R., & Rochen, B. (1993). *The Childhood Autism Rating Scale*. Western Psychological Services, Los Angeles.
- Schultz, R., Gauthier, I., Klin, A., Fulbright, R., Anderson, A., Volkmar, F., et al., (2000). Abnormal ventral temporal cortical activity during face discrimination among individuals with autism and Asperger syndrome. *Archives of General Psychiatry*, 57, 331-340.
- Schwarzer, G. (2000). Development of face processing: the effect of face inversion. *Child Development*, 71(2), 391-401.
- Schwarzer, G., & Massaro, D. W. (2001). Modeling face identification processing in children and adults. *Journal of Experimental Child Psychology*, 79(2), 139-161.
- Searcy et al., (2004). The relationship between age and IQ in adults with Williams syndrome. *American Journal on Mental Retardation*, 109(3), 231-236.
- Searcy, J. H., & Bartlett, J. C. (1996). Inversion and processing of component and spatial-relational information in faces. *Journal of Experimental Psychology: Human Perception and Performance*, 22(4), 904-915.
- Sergent, J. (1989). Ontogeny and microgenesis of face perception. *Current Psychology of Cognition*, 9(1), 123-128.
- Shah, A. & Frith, U. (1993). Why do autistic individuals show superior performance on the block design task? *Journal of Child Psychology and Psychiatry*, 8, 1351-1364.
- Shaywitz, B. A., Shaywitz, K. M., Blachman, B. A., Pugh, K. R., Fulbright, R. T. Skudlarski, P., et al., (2004). Development of left occipitotemporal systems for skilled reading in children after a phonologically-based intervention. *Biological Psychiatry*, 55(9), 926-33.
- Skuse D. (2003). X-linked genes and the neural basis of social cognition. *Novartis Foundation Symposium*, 251, 84-98.
- Stiles, J, Sabbadini, L, Capirci, O., & Volterra, V. (2000). Drawing abilities in Williams syndrome: a case study. *Developmental Neuropsychology*, 18 (2), 213-35.
- Stehli, A. (1996). *The sound of a miracle: Child's triumph over autism*. Fourth Estate.

- Stone, J. L., Merriman, B., Cantor, R. M., Yonan, A. L., Gilliam, T. C., Geschwind, D., et al. (2004). Evidence for sex-specific risk alleles in autism spectrum disorder. *American Journal of Human Genetics*, 75, 1117-1123,
- Stromme, P., Bjornstad, P. G., & Ramstad, K. (2002). Prevalence estimation of Williams syndrome. *Journal of Child Neurology*, 17(4), 269-71.
- Swettenham, J., Baron-Cohen, S., Charman, T., Cox, A., Baird, G., Drew, A., et al. (1998). The frequency and distribution of spontaneous attention shifts between social and nonsocial stimuli in autistic, typically developing, and nonautistic developmentally delayed infants. *Journal of Child Psychology and Psychiatry*, 39(5), 747-53.
- Tager-Flusberg, H., (1996). Brief report: current theory and research on language and communication in autism. *Journal of Autism and Developmental Disorders*, 26(2), 169-72.
- Tager-Flusberg, H., & Joseph R.M., (2003). Identifying neurocognitive phenotypes in autism. *Philos Trans R Soc Lond B Biol Sci.* 358(1430), 303-14.
- Tager-Flusberg, H., Plesa-Skwerer, D., Faja, S., & Robert M. Joseph, R. M. (2003). People with Williams Syndrome Process Faces Holistically. *Cognition*, 89, 11-24.
- Tager-Flusberg, H., & Sullivan, K. (2000). A componential view of theory of mind: evidence from Williams syndrome. *Cognition*, 76, 59-89.
- Tanaka, J. W., & Farah, M. J. (1993). Parts and wholes in face recognition. *Quarterly Journal of Experimental Psychology [A]*, 46(2), 225-245.
- Tanaka, J. W., & Sengco, J. A. (1997). Features and their configuration in face recognition. *Memory and Cognition*, 25(5), 583-592.
- Tanaka, J. W., Kay, J. B., Grinnell, E., Stansfield, B., & Szechter, L. (1998). Face recognition in young children: When the whole is greater than the sum of its parts. *Visual Cognition*, 479-496.
- Tantam, D., Monaghan, L., Nicholson, H. & Stirling, J. (1989). Autistic children's ability to interpret faces: a research note. *Journal of Child Psychology and Psychiatry*, 30, 623-630.
- Tarr, M. J. & Cheng, Y.D. (2003). Learning to see faces and objects. *Trends in Cognitive Science*, 7(1), 23-30.
- Tassabehji, M. (2003). Williams-Beuren syndrome: a challenge for genotype-phenotype correlations. *Human Molecular Genetics*, 12(2), 229-237.
- Tassabehji, M., Metcalfe, K., Fergusson, W.D., Carette, M. J. A., Dore, J. K., Donnai. D., et al. (1996). LIM-kinase deleted in Williams syndrome. *Nature Genetics*, 13, 272-3.

- Tassabehji, M., Metcalfe, K., Karmiloff-Smith, A., Carette, J., Grant, J., Dennis, N., et al. (1999). Williams syndrome: Use of chromosomal microdeletions as a tool to dissect cognitive and physical phenotypes. *Journal of Human Genetics*, 64, 118-125.
- Temple, C. M., Almazan, M., & Sherwood, S. (2002). Lexical skills in Williams Syndrome: a cognitive neuropsychological analysis. *Journal of Neurolinguistics*, 15, 463-495.
- Temple, C. (2003). Deep dyslexia in Williams syndrome. *Journal of Neurolinguistics*, 16 (6), 457-488.
- Teunnise, J. P., & de Gelder, B. (2003). Face processing in adolescents with autistic disorder: the inversion and composite effects. *Brain and Cognition*, 52(3), 285-94.
- Thomas, M. S. C. (2003). Multiple causality in developmental disorders: Methodological implications from computational modelling. *Developmental Science*, 6 (5), 537-556.
- Thomas, M. S. C. (2005). Characterising compensation. *Cortex*, 41(3), 434-442.
- Thomas, M. S. C., Grant, J., Barham, Z., Gsödl, M., Laing, E., Lakusta, L., et al. (2001). Past tense formation in Williams syndrome. *Language and Cognitive Processes*, 16 (2/3), 143-176.
- Thompson, P. (1980). Margaret Thatcher: A new illusion. *Perception*, 9, 483-484.
- Thomson, E., Annaz, D., & Thomas, M. (2006). Visuo-spatial processing in autism. Poster to be presented at British Psychological Society.
- Tong, F., Nakayama, K., Moscovitch, M., Weinrib, O., & Kanwisher, N. (2000). Response properties of the human fusiform face area. *Cognitive Neuropsychology*, 27(1-3), 257-279.
- Treisman, A. (1993). The perception of features and objects. In A. Baddeley & L. Weiskrantz (Eds.), *Attention: selection, awareness, and control* (pp. 5-35). Oxford: Clarendon Press.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, 13(2), 232-244.
- Udwin, O., Yule, W., & Martin, N. D. T. (1987). Cognitive abilities and behavioural characteristics of children with idiopathic infantile hypercalcaemia. *Journal of Child Psychology and Psychiatry*, 28, 297-309.
- Valentine, T. (1988). Upside-down faces: a review of the effect of inversion upon face recognition. *British Journal of Psychology*, 79(4), 471-91.

- Van der Geest, J. N., Kemner, C., Verbaeten M. N., & van Engeland, H. (2002). Gaze behavior of children with pervasive developmental disorder toward human faces: a fixation time study. *Journal of Child Psychology and Psychiatry*, 43(5), 669-78.
- Volkmar, F. R., Klin, A., Schultz, R. T., Rubin, E., & Bronen, R. (2000). Asperger's disorder. *American Journal of Psychiatry*, 157(2), 262-267.
- Volkmar, F., & Pauls, D., (2003). Autism. *The Lancet*, 362, 1133-1141.
- Wang, P. P. (1996). A neuropsychological profile of Down syndrome: Cognitive skills and brain morphology. *Mental Retardation and Developmental Research Reviews*, 2, 102-108.
- Wang, P. P., & Bellugi, U. (1994). Evidence from two genetic syndromes for a dissociation between verbal and visual-spatial short-term memory. *Journal of Clinical and Experimental Neuropsychology*, 16(2), 317-322.
- Wang, P. P., Doherty, S., Rourke, S. B., & Bellugi, U. (1995). Unique profile of visuo-perceptual skills in a genetic syndrome. *Brain and Cognition*, 29, 54-65.
- Warrington, E. K. (1984). *Recognition Memory Test*. Windsor, England: NFER-Nelson.
- Weeks, D., Chua, R. & Elliott, D. (2002). *Perceptual-Motor Development in Down Syndrome*. Champaign, Illinois : Human Kinetics.
- Williams, D. L, Goldstein, G., & Minshew, N. J., (2005). Impaired memory for faces and social scenes in autism: clinical implications of memory dysfunction. *Archives of Clinical Neuropsychology*, 20(1), 1-15.
- Wing, L. & Gould, J. (1979). Severe impairments of social interaction and associated abnormalities in children. Epidemiology and classification. *Journal of Autism and Developmental Disorders*, 9, 11-29.
- Wing, L. (1993). The Definition and Prevalence of Autism: A Review. *European Child and Adolescent Psychiatry*, 2(2), 61-74.
- Wishart, J. G., & Johnston, F. H. (1990). The effects of experience on attribution of a stereotyped personality to children with Down's syndrome. *Journal of Mental Deficiency Research*, 34, 409-420.
- Wishart, J. G., & Pitcairn, T. K. (2000) The recognition of identity and expression in faces by children with Down syndrome. *American Journal on Mental Retardation*, 105, 466-479.
- Yin, R. K. (1969). Looking at upside-down faces. *Journal of Experimental Psychology*, 81, 141-145.

Young, A. W., Hellawell, D., & Hay, D. C. (1987). Configurational information in face perception. *Perception*, 16(6), 747-759.

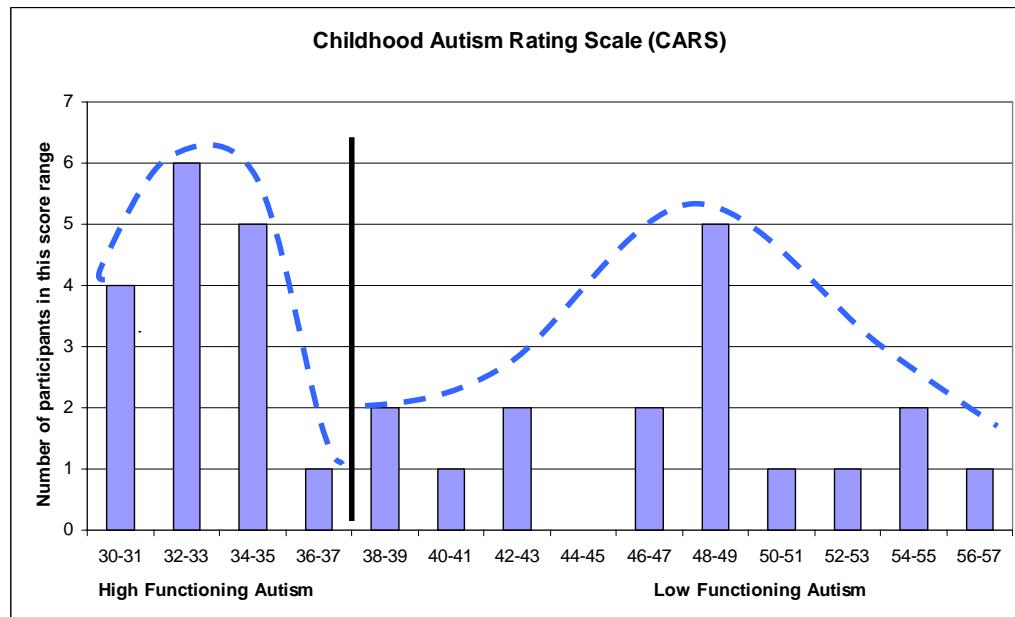
Zukowski, A. (2001). *Uncovering grammatical competence in children with Williams syndrome*. Unpublished doctoral dissertation, Boston University, Boston.

## **APPENDICES**



**Distribution of Childhood Autism Rating Scale scores for children in autism groups.**

Figure 1: Individuals scores on CARS for children with autism. Dotted lines suggest two distributions of the scores.



No	CARS score	
1	32	<b>High-Functioning group</b>
2	31	
3	30	
4	30	
5	31	
6	34	
7	32	
8	32	
9	33	
10	35	
11	35	
12	36	
13	34	
14	35	
15	32	
16	33	
1	38	<b>Low-Functioning group</b>
2	39	
3	55	
4	47	
5	56	
6	48	
7	49	
8	52	
9	48	
10	43	
11	51	
12	49	
13	43	
14	49	
15	41	
16	55	
17	47	

**Developmental trajectories of the Whole-Part paradigm.**

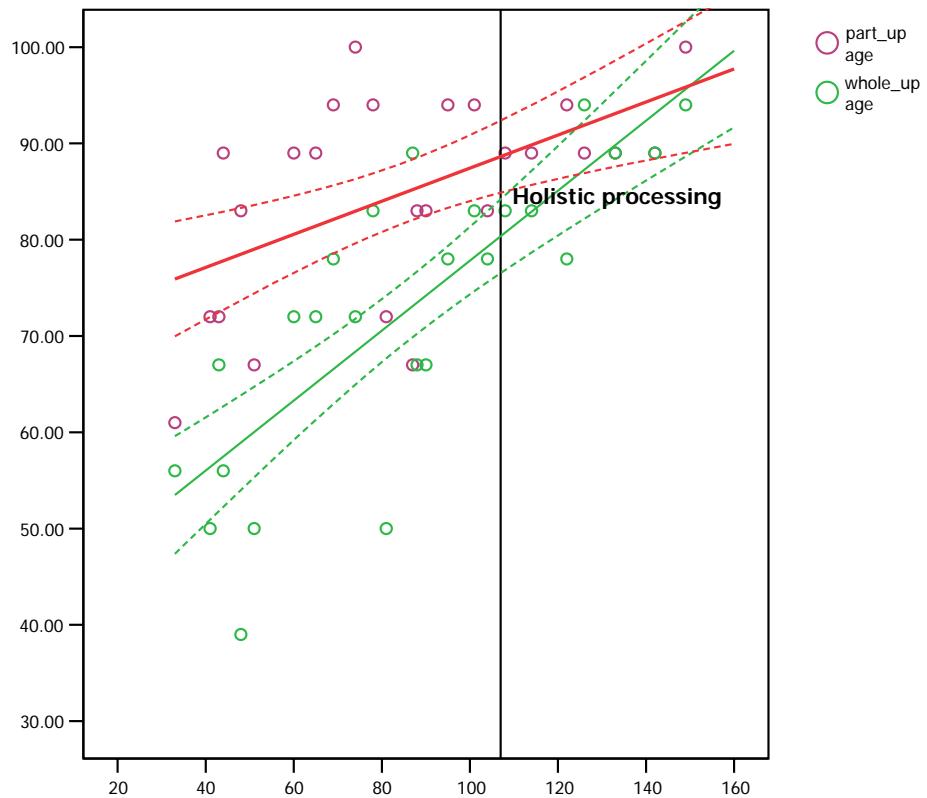
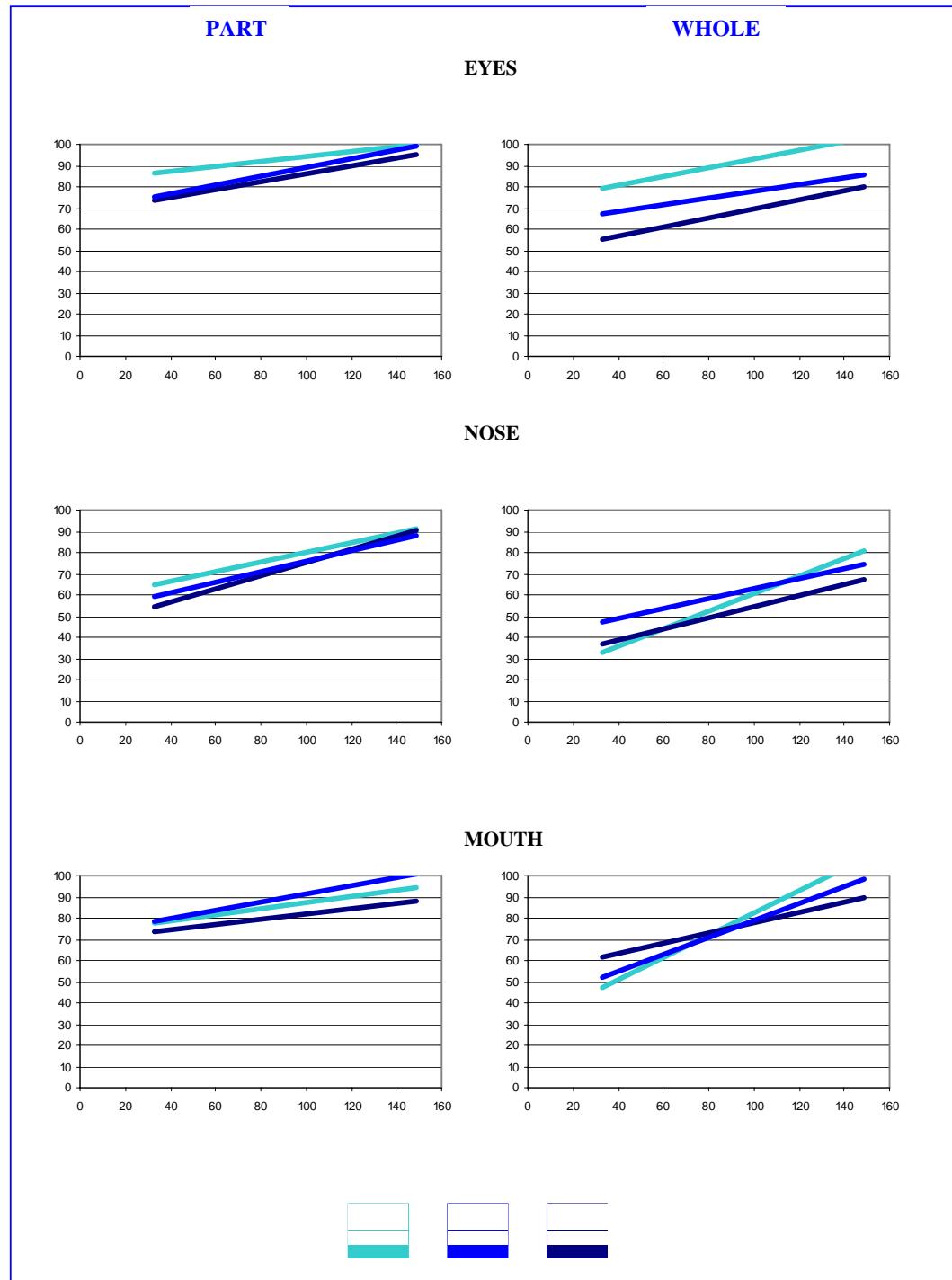


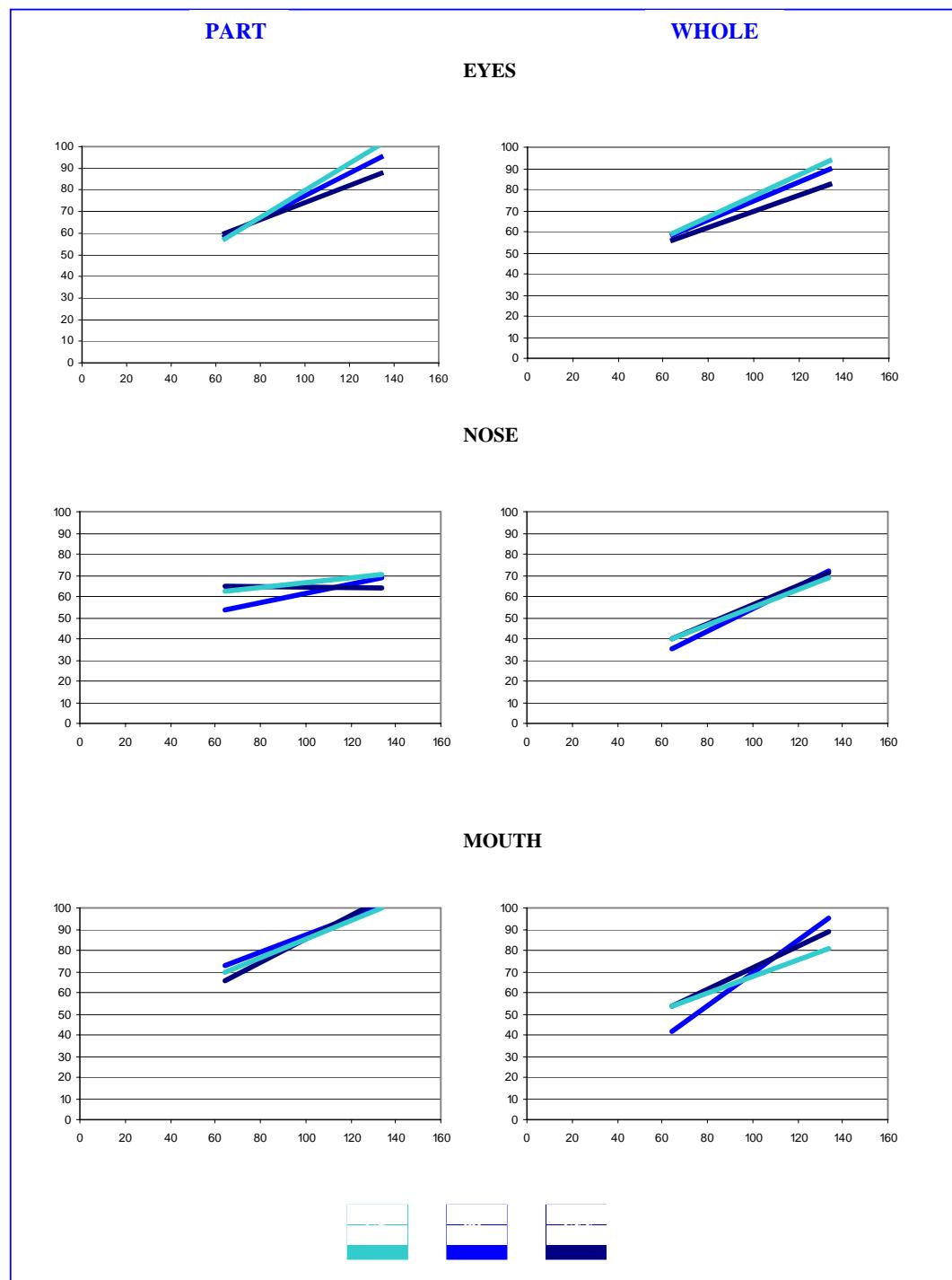
Figure 1: Developmental trajectory of TD group on whole- and part-face condition in upright orientation. X-axis shows age in months and Y-axis shows % accuracy.

Figure 2 a-e: Illustrates developmental trajectories on each feature. X-axis shows age in months and Y-axis shows % accuracy.

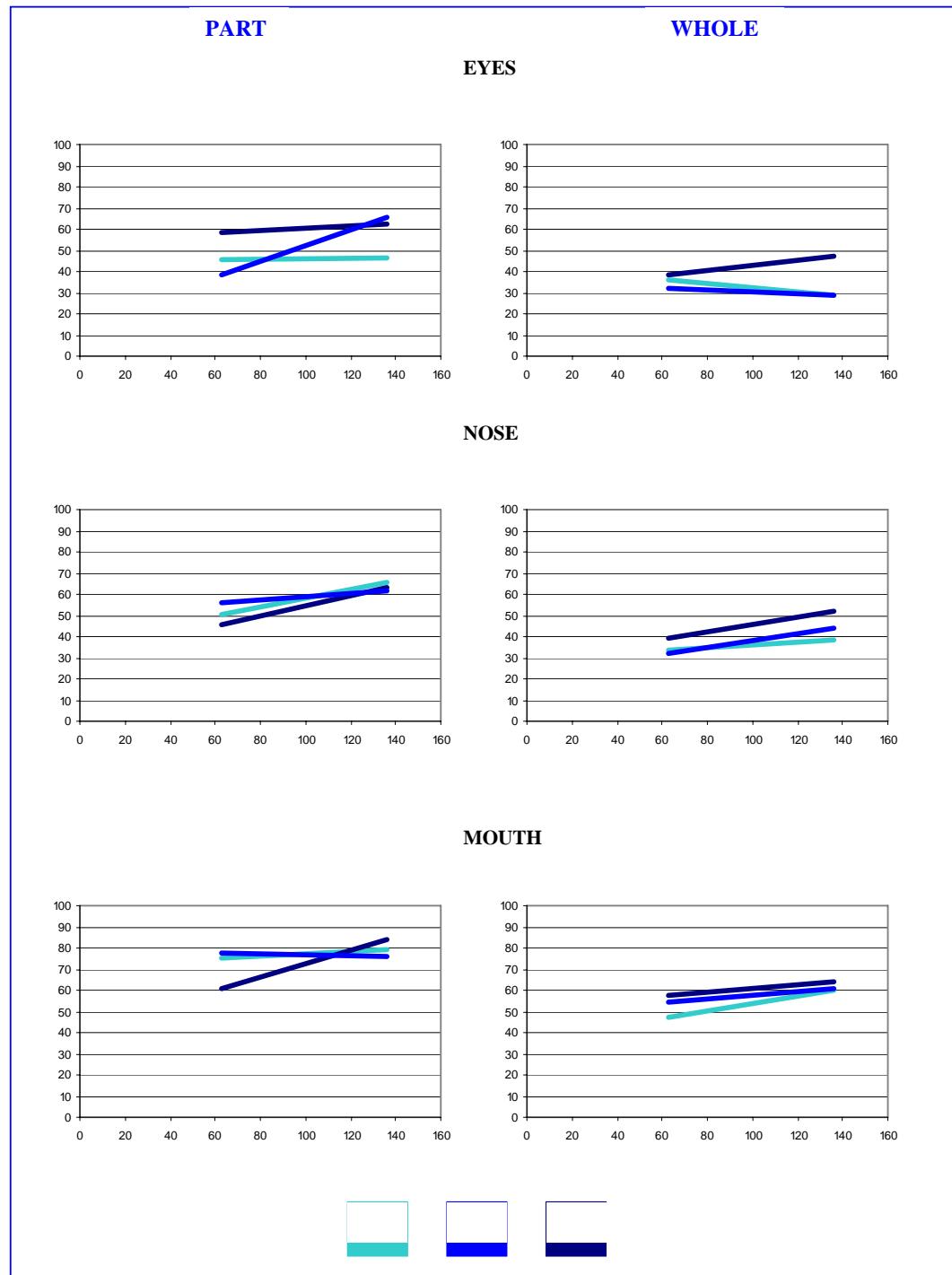
a) Feature recognition in TD group.



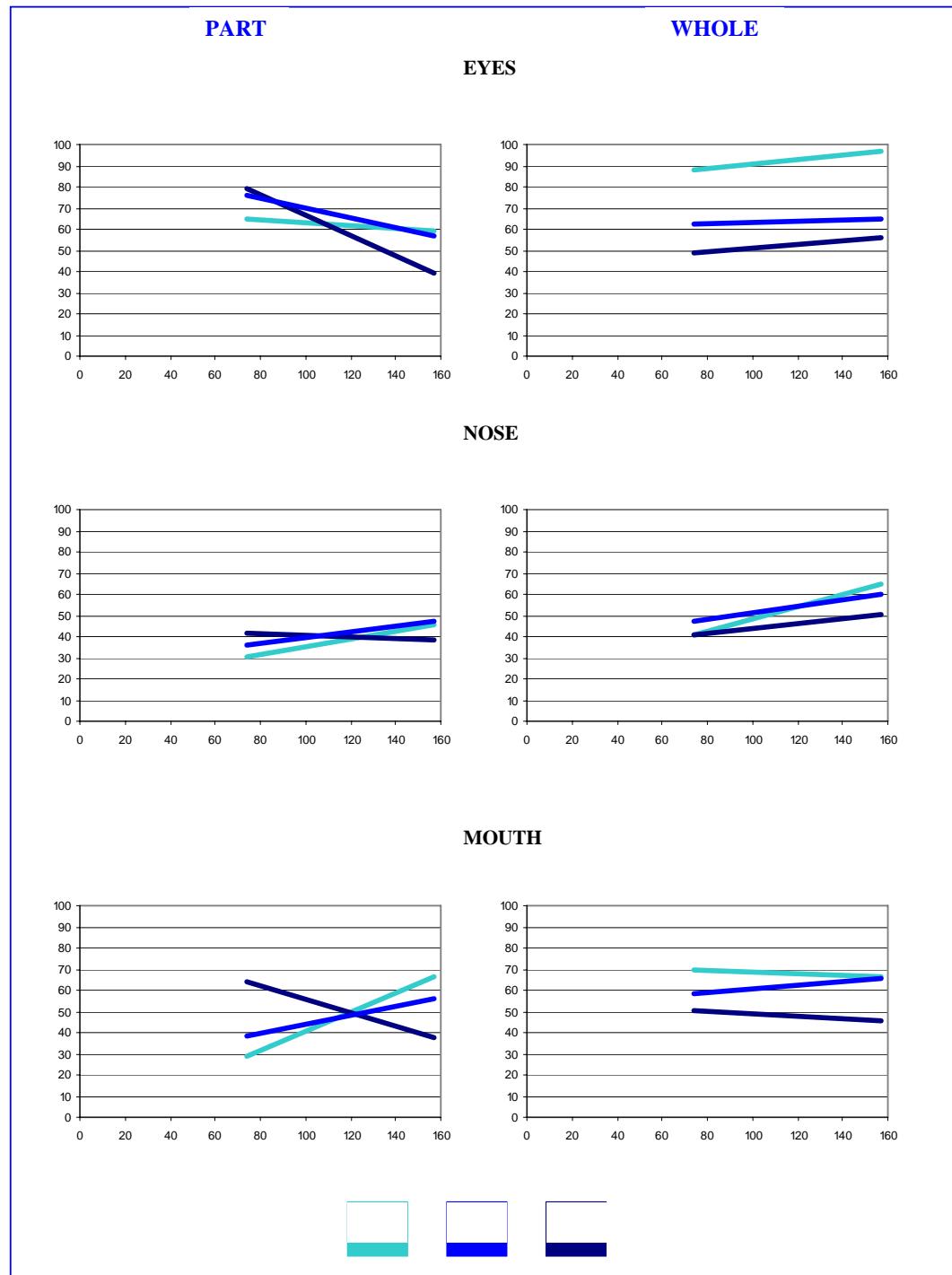
b) Feature recognition in HFA group.



c) Feature recognition in LFA.



d) Feature recognition in DS



#### e) Feature recognition in WBS group

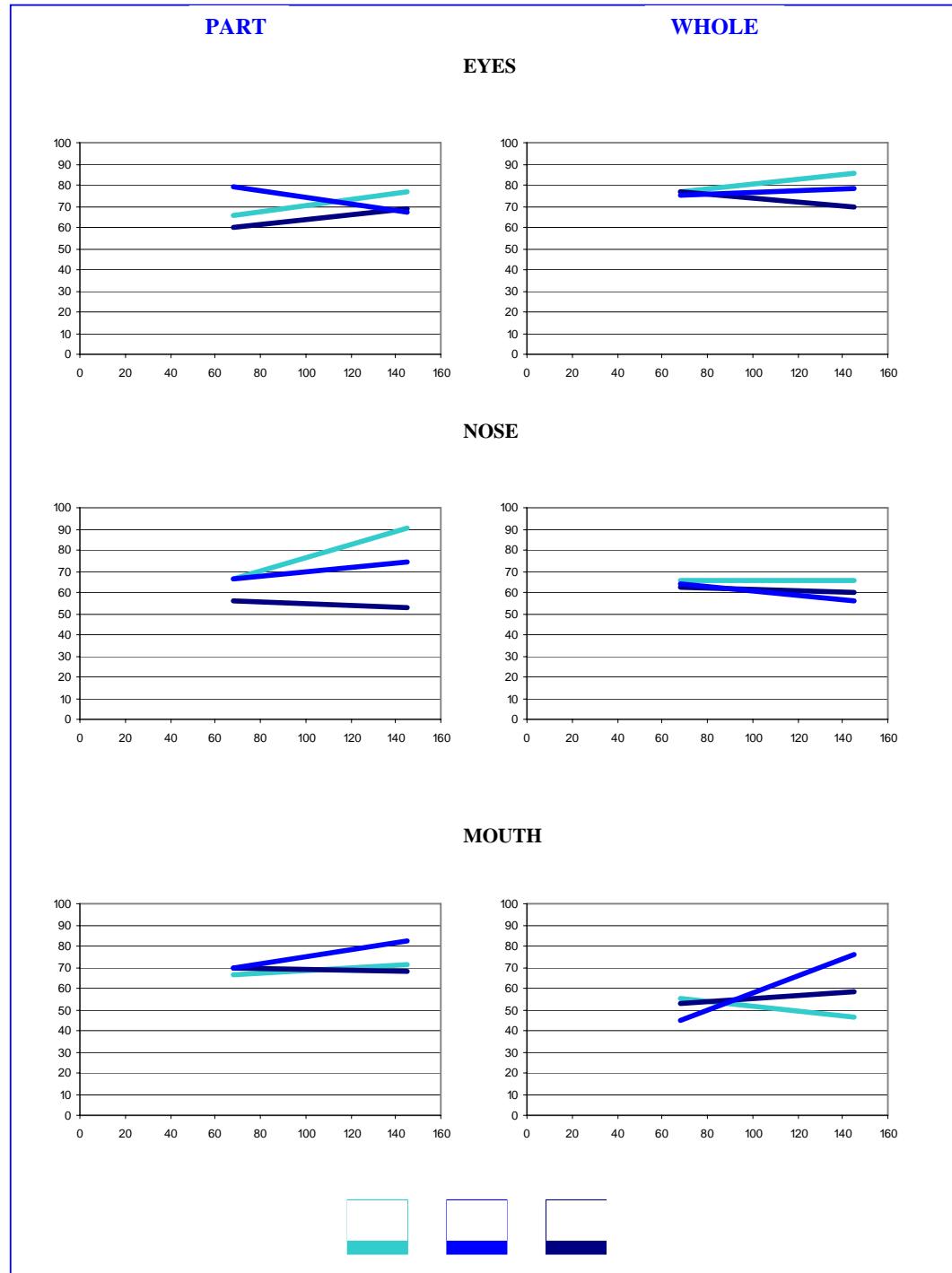
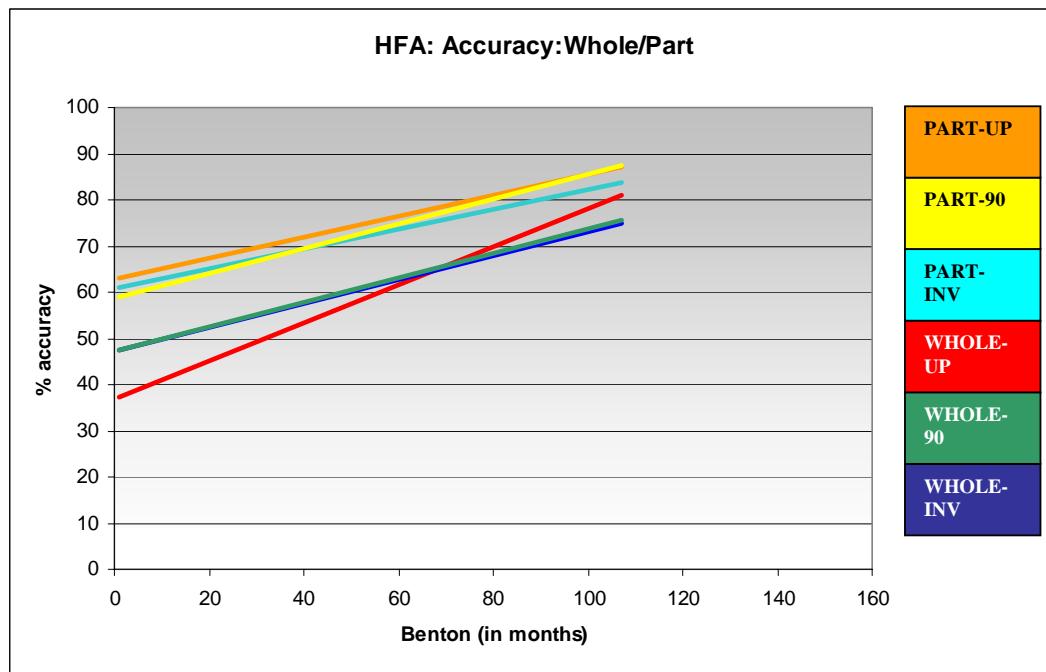
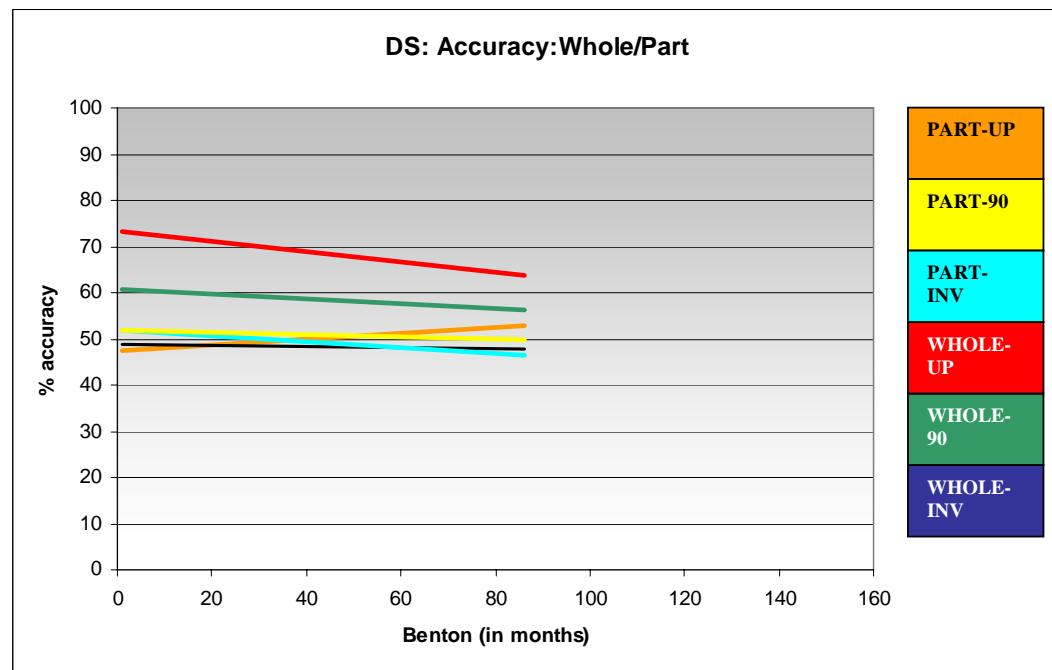
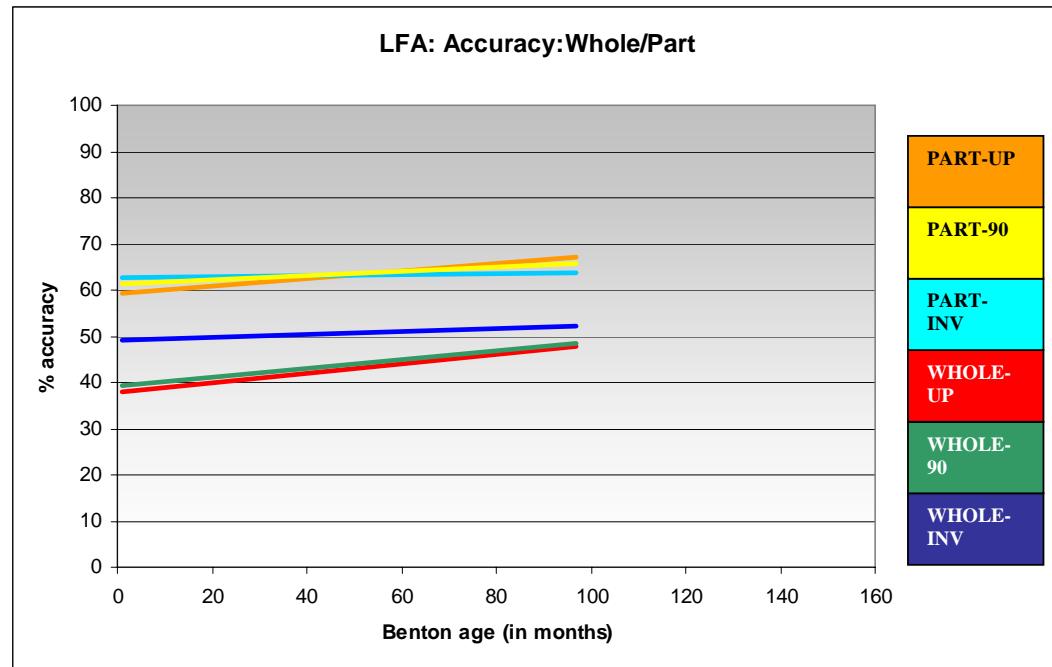


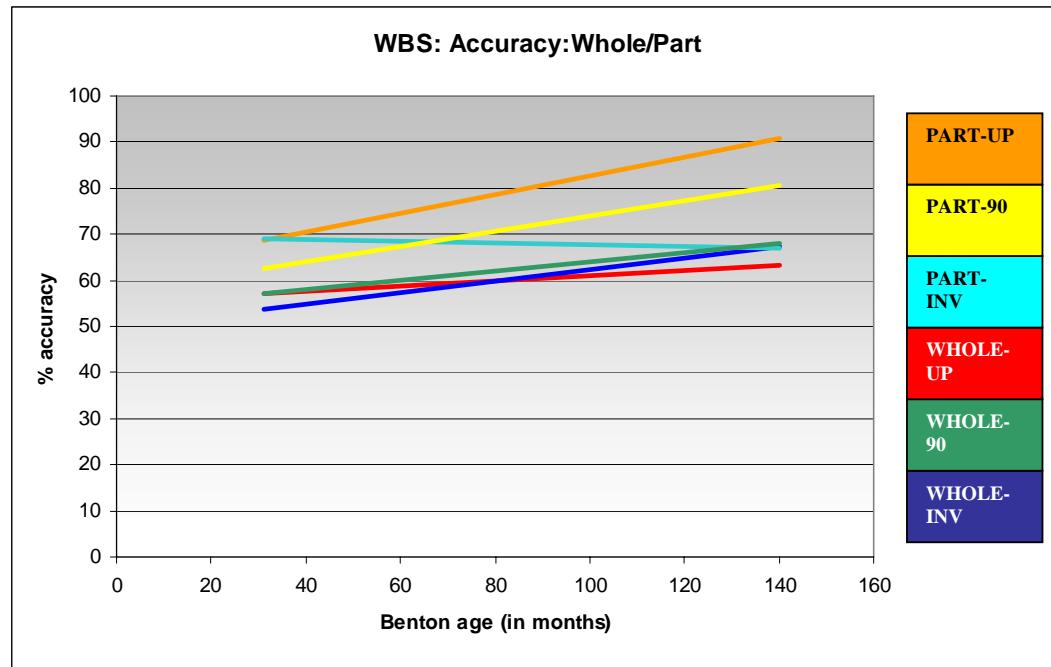
Figure 3: P-values for Jane faces against Benton test age.

Interactions	HFA	LFA	DS	WBS
Task x orientation	P=.390	P=.057	P<.006	P=.921
Task x age	P=.404	P=.658	P=.890	P=.264
Task x group	P=.283	P=.114	P=.906	P=.300
Orientation x group	P<.001	P<.001	P<.001	P<.001
Orientation x age	P=.107	P=.144	P=.135	P=.084

Figure 4: Performance accuracy on whole-part face recognition against Benton age equivalent.







### Featural-Configural analyses on Jane faces task.

Table 1: Summary of each group accuracy results plotted according to their CA. P-values for main effects and interactions from fully-factorial ANCOVAs. Red colour is used for significant p-values, pink for p-values at marginal significance.

A)

GROUPS	MAIN EFFECTS			2-WAY INTERACTIONS			3-WAY T*O*A
	T	O	A	T*O	T*A	O*A	
TD	p=.001	p=.002	p=.001	p=.011	p=.082	p=.002	p=.001
HFA	p=.001	p=.561	p=.002	p=.076	p=.050	p=.562	p=.772
LFA	p=.013	p=.064	p=.100	p=.032	p=.561	p=.051	p=.081
WS	p=.032	p=.247	p=.027	p=.045	p=.663	p=.767	p=.108
DS	p=.282	p=.946	p=.106	p=.479	p=.944	p=.291	p=.665

B)

GROUPS	FEATURAL			CONFIGURAL			T*O*A
	O	A	O*A	O	A	O*A	
TD	p=.533	p=.001	p=.268	p=.002	p=.001	p=.001	
HFA	p=.764	p=.002	p=.780	p=.632	p=.002	p=.625	
LFA	p=.008	p=.408	p=.010	p=.560	p=.103	p=.371	
WBS	p=.953	p=.009	p=.362	p=.114	p=.354	p=.398	
DS	p=.400	p=.212	p=.176	p=.666	p=.294	p=.802	

Table 2: Summary of each disorder group accuracy results plotted according to the Benton age equivalent.

A)

GROUPS	MAIN EFFECTS			2-WAY INTERACTIONS			3-WAY T*O*A
	T	O	A	T*O	T*A	O*A	
HFA	p=.001	p=.884	p=.002	p=.309	p=.036	p=.896	p=.284
LFA	p=.001	p=.514	p=.700	p=.478	p=.962	p=.191	p=.111
WBS	p=.006	p=.210	p=.149	p=.104	p=.663	p=.906	p=.282
DS	p=.002	p=.007	p=.341	p=.578	p=.804	p=.673	p=.458

B)

GROUPS	FEATURAL			CONFIGURAL			T*O*A
	O	A	O*A	O	A	O*A	
HFA	p=.202	p=.021	p=.187	p=.616	p=.001	p=.588	
LFA	p=.413	p=.794	p=.690	p=.770	p=.107	p=.040	
WBS	p=.704	p=.067	p=.459	p=.135	p=.583	p=.603	
DS	p=.062	p=.565	p=.673	p=.061	p=.446	p=.480	

Table 3: Summary of disorder group comparisons to the TD group on accuracy plotted according to their CA

CA	Main effects		2-way		3-way		4-way	
	T	0	A	G	T*G	T*A	T*G*A	T*G*G
HFA vs TD	p=.001	p=.072	p=.001	p=.026	p=.007	p=.001	p=.070	p=.005
LFA vs TD	p=.001	p=.001	p=.001	p=.136	p=.007	p=.142	p=.001	p=.005
WS vs TD	p=.001	p=.970	p=.001	p=.266	p=.771	p=.263	p=.011	p=.217
DS vs TD	p=.006	p=.148	p=.001	p=.243	p=.823	p=.693	p=.001	p=.001
HFA vs LFA	p=.002	p=.001	p=.001	p=.458	p=.501	p=.081	p=.421	p=.082
WS vs DS	p=.397	p=.028	p=.008	p=.796	p=.095	p=.679	p=.742	p=.834
DS vs LFA	p=.133	p=.017	p=.020	p=.952	p=.089	p=.026	p=.156	p=.686
WS vs HFA	p=.001	p=.753	p=.001	p=.292	p=.510	p=.344	p=.398	p=.787
FEATUREAL								
CA	Main effects		2-way		3-way		Main effects	
	0	A	G	0*G	A*G	0*A*G	0	A
HFA vs TD	p=.579	p=.001	p=.646	p=.471	p=.886	p=.927	p=.930	p=.091
LFA vs TD	p=.001	p=.846	p=.001	p=.003	p=.076	p=.010	p=.042	p=.018
WS vs TD	p=.738	p=.001	p=.966	p=.165	p=.825	p=.242	p=.769	p=.902
DS vs TD	p=.253	p=.001	p=.390	p=.059	p=.488	p=.259	p=.349	p=.366
HFA vs LFA	p=.030	p=.024	p=.886	p=.039	p=.078	p=.042	p=.094	p=.468
WS vs DS	p=.458	p=.014	p=.534	p=.099	p=.502	p=.002	p=.601	p=.162
DS vs LFA	p=.009	p=.078	p=.674	p=.004	p=.113	p=.707	p=.193	p=.987
WS vs HFA	p=.766	p=.001	p=.743	p=.463	p=.815	p=.518	p=.813	p=.594
CONFIGURAL								
CA	Main effects		2-way		3-way		2-way	
	0	A	G	0*G	A*G	0*A*G	0	A
HFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
LFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
HFA vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs DS	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs HFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
3-way								
CA	Main effects		2-way		3-way		Main effects	
	0	A	G	0*G	A*G	0*A*G	0	A
HFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
LFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
HFA vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs DS	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs HFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
4-way								
CA	Main effects		2-way		3-way		Main effects	
	0	A	G	0*G	A*G	0*A*G	0	A
HFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
LFA vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs TD	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
HFA vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs DS	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
DS vs LFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001
WS vs HFA	p=.001	p=.001	p=.001	p=.003	p=.076	p=.010	p=.004	p=.001

Table 4: Summary of disorder groups comparisons to the TD group on accuracy plotted according to their Benton age equivalent.

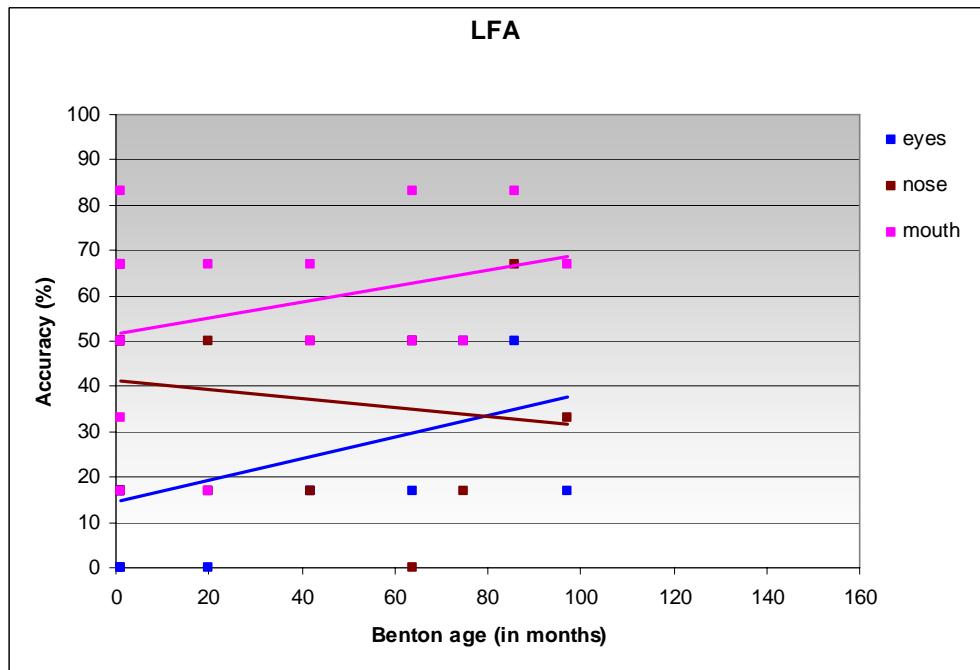
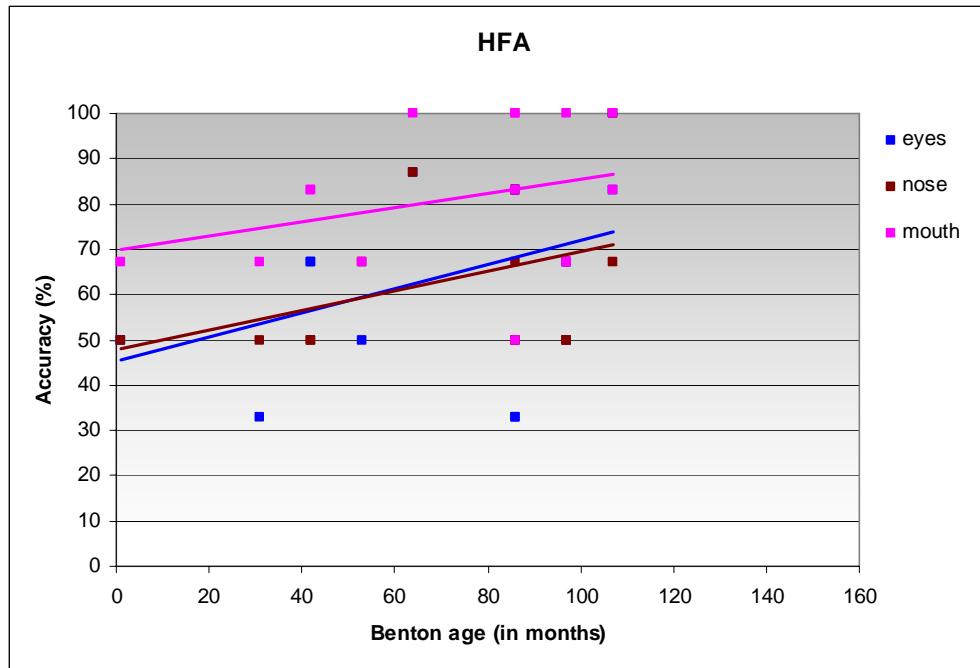
GROUPS	Main effects			2-ways				3-ways		4-ways		
	T	0	A	6	T*0	T*A	T*6	0*A	0*6	A*G	T*0*A	T*0*G
HFA vs TD	p=.001	p=.117	p=.001	p=.500	p=.930	p=.090	<b>p=.019</b>	<b>p=.001</b>	p=.194	p=.262	<b>p=.059</b>	<b>p=.048</b>
LFA vs TD	p=.001	<b>p=.018</b>	p=.002	p=.704	p=.114	p=.664	p=.228	p=.001	<b>p=.011</b>	<b>p=.001</b>	<b>p=.035</b>	<b>p=.002</b>
WBS vs TD	p=.001	p=.979	p=.001	p=.605	p=.947	p=.362	p=.215	<b>p=.001</b>	<b>p=.013</b>	p=.132	<b>p=.005</b>	<b>p=.023</b>
DS vs TD	p=.001	p=.622	p=.002	p=.639	p=.249	p=.983	p=.905	<b>p=.001</b>	<b>p=.001</b>	p=.278	<b>p=.001</b>	<b>p=.069</b>
HFA vs LFA	p=.001	p=.673	<b>p=.003</b>	p=.686	p=.167	p=.145	p=.285	p=.338	p=.880	<b>p=.009</b>	p=.688	p=.316
WBS vs DS	p=.001	<b>p=.023</b>	p=.102	p=.460	p=.149	p=.970	p=.357	p=.850	p=.754	p=.961	p=.883	p=.348
DS vs LFA	p=.001	p=.157	p=.285	p=.356	p=.393	p=.794	p=.275	p=.245	<b>p=.019</b>	p=.505	p=.154	p=.856
WBS vs HFA	p=.001	p=.372	<b>p=.001</b>	p=.924	p=.091	p=.382	p=.776	p=.991	p=.282	p=.055	p=.141	p=.902

GROUPS	Main effects			2-ways				3-ways		Main effects		CONFIGURAL		
	0	A	G	0*A	0'G	A'G	0*A'G	0	A	G	0*A	0'G	A*G	0'A'G
HFA vs TD	p=.101	<b>p=.001</b>	p=.480	<b>p=.050</b>	p=.373	p=.624	p=.424	p=.361	<b>p=.001</b>	<b>p=.093</b>	<b>p=.003</b>	<b>p=.068</b>	<b>p=.029</b>	<b>p=.001</b>
LFA vs TD	p=.303	<b>p=.006</b>	p=.678	p=.327	p=.883	<b>p=.019</b>	p=.891	<b>p=.014</b>	<b>p=.018</b>	p=.300	<b>p=.001</b>	<b>p=.025</b>	p=.073	<b>p=.001</b>
WBS vs TD	p=.970	<b>p=.001</b>	p=.628	p=.178	p=.544	p=.206	p=.990	p=.960	<b>p=.024</b>	p=.287	<b>p=.001</b>	<b>p=.009</b>	p=.277	<b>p=.001</b>
DS vs TD	p=.413	<b>p=.012</b>	p=.764	p=.929	<b>p=.065</b>	p=.188	p=.320	p=.948	<b>p=.025</b>	p=.684	<b>p=.001</b>	<b>p=.002</b>	p=.694	<b>p=.004</b>
HFA vs LFA	p=.157	<b>p=.068</b>	<b>p=.732</b>	p=.239	p=.540	p=.142	p=.545	p=.625	<b>p=.003</b>	p=.370	p=.661	p=.509	<b>p=.007</b>	p=.143
WBS vs DS	p=.497	<b>p=.003</b>	p=.885	p=.130	p=.299	p=.532	<b>p=.032</b>	p=.334	p=.313	p=.846	p=.487	p=.751	p=.379	
DS vs LFA	p=.559	p=.515	p=.927	p=.978	<b>p=.169</b>	p=.715	p=.580	p=.123	p=.372	p=.187	p=.089	<b>p=.065</b>	p=.555	p=.718
WBS vs HFA	p=.243	p=.149	<b>p=.845</b>	p=.949	p=.575	p=.709	p=.431	p=.161	<b>p=.005</b>	p=.830	p=.451	p=.473	<b>p=.039</b>	p=.922

### Developmental trajectories of Face construction task.

Figure 1: Performance accuracy on feature condition against Benton age equivalent.



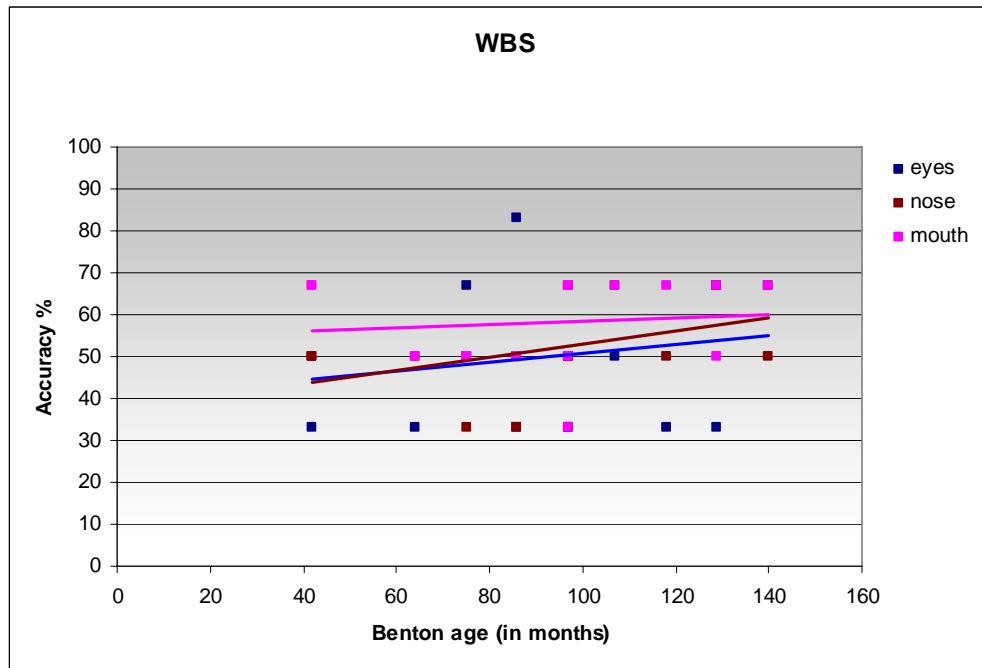
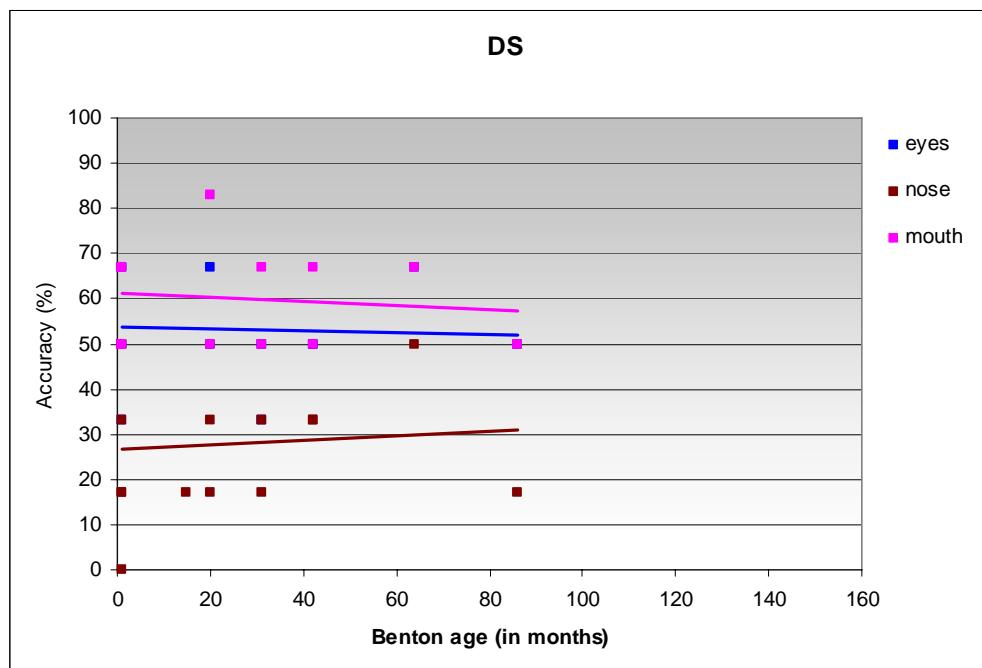
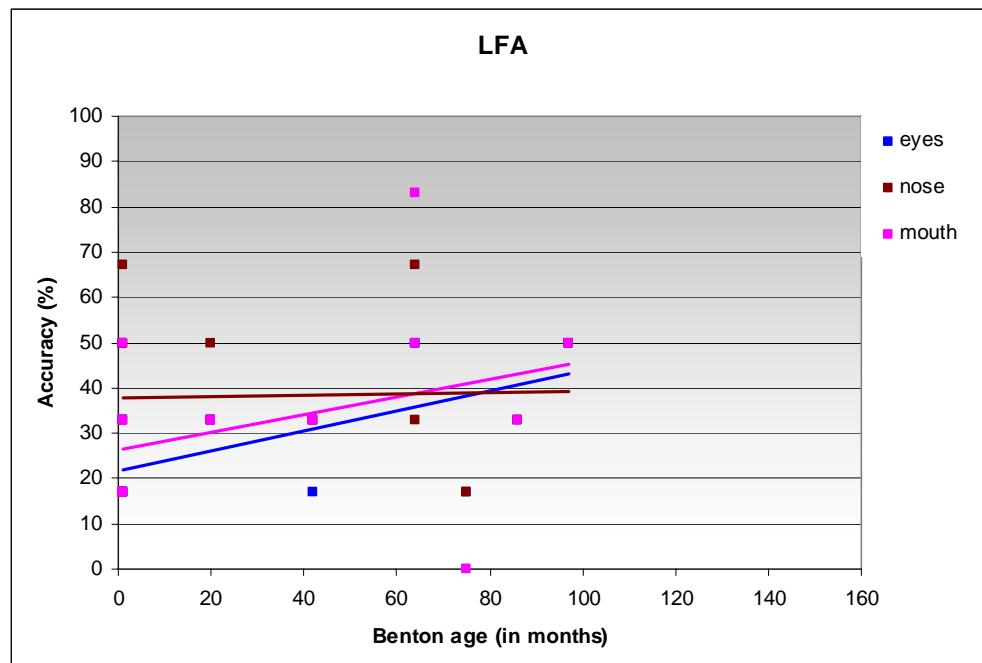
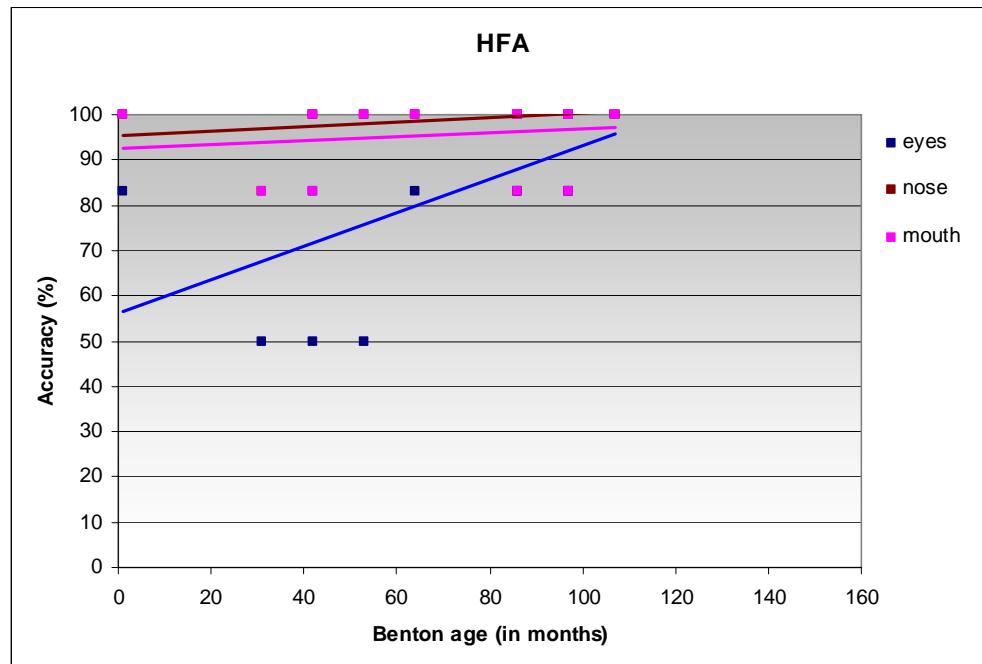


Figure 2: Performance accuracy on first-order configural placement condition against Benton age equivalent.



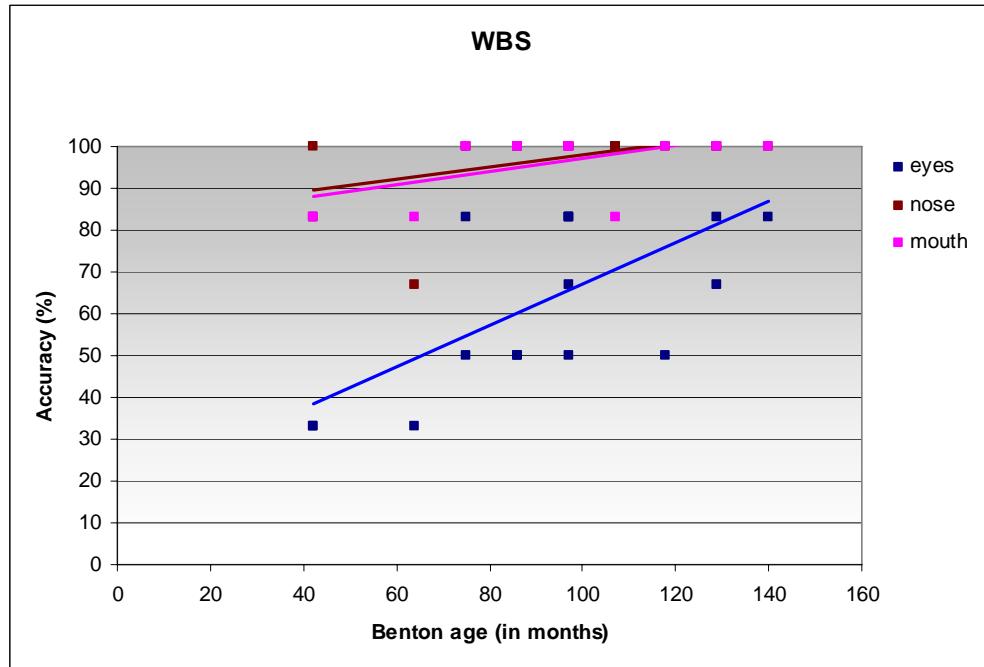
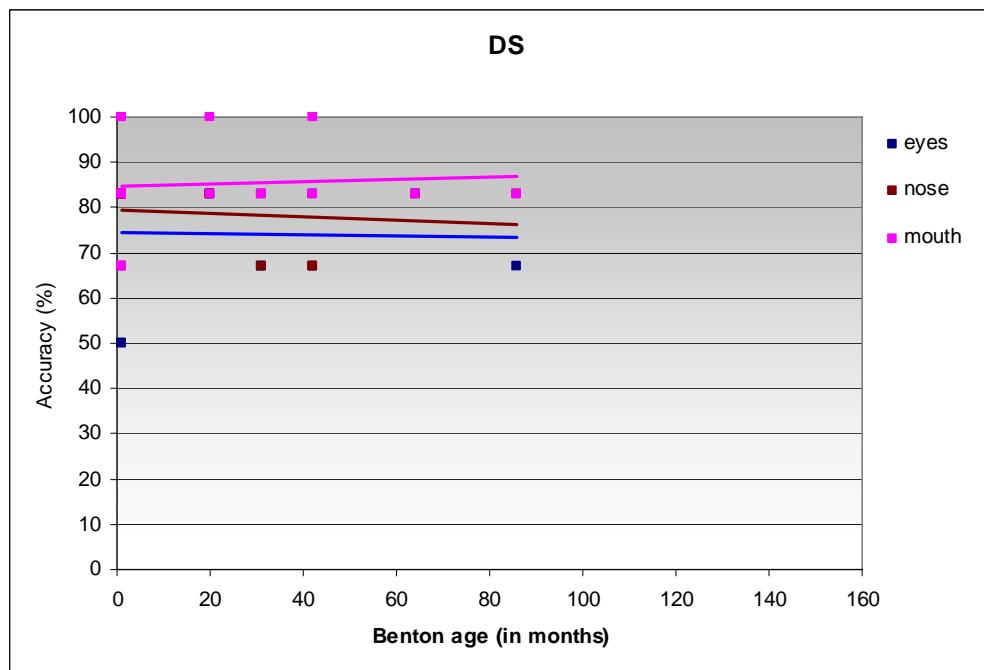
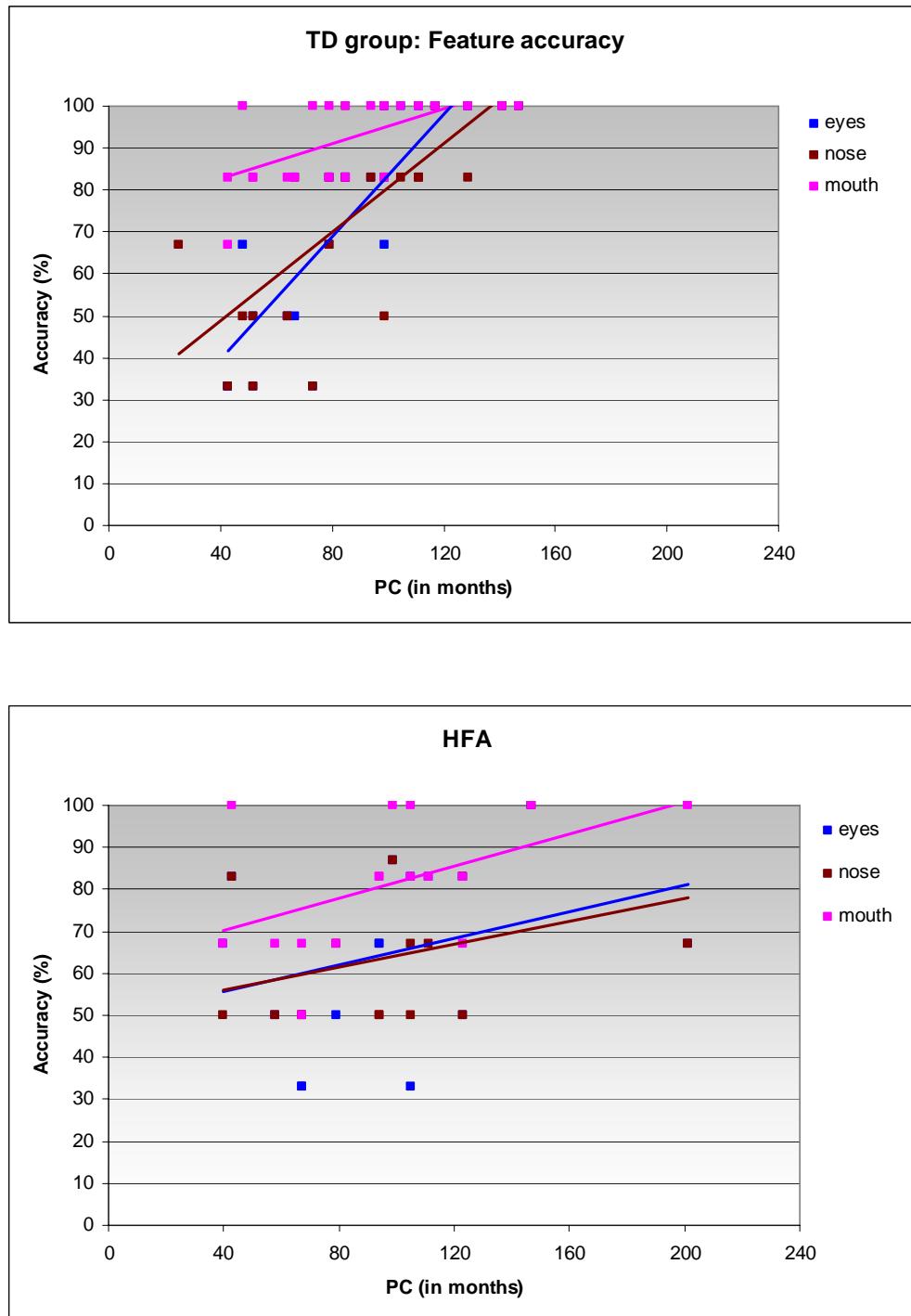
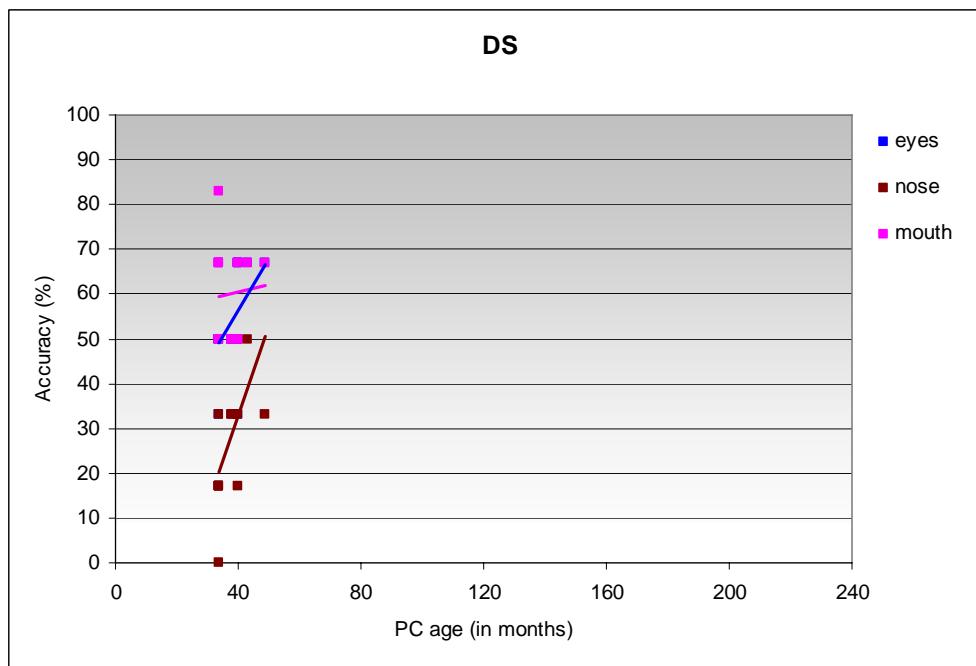
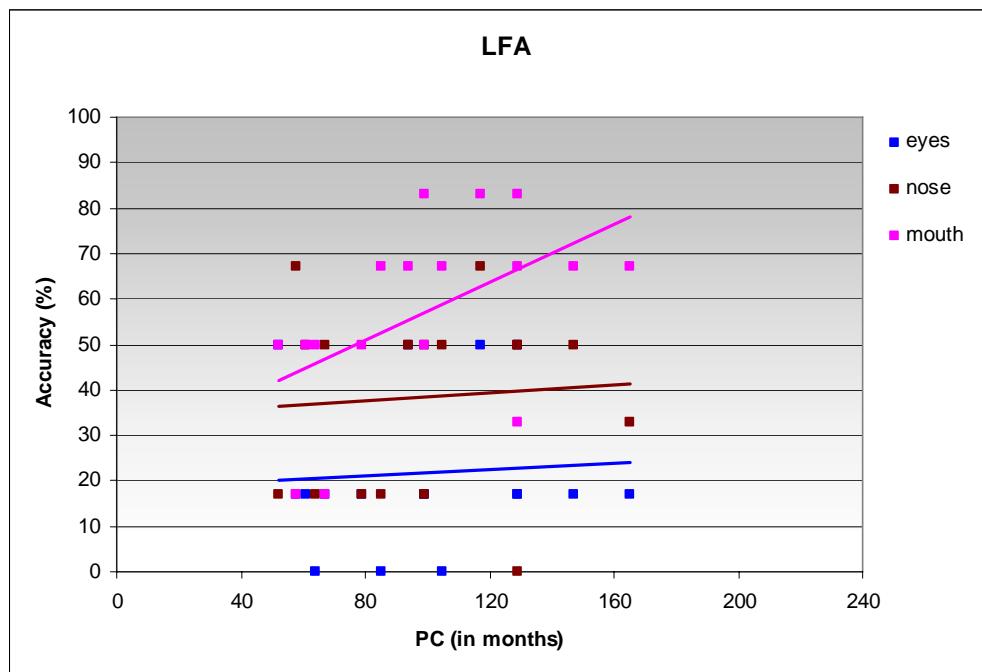


Figure 3: Performance accuracy on feature condition against PC age equivalent.





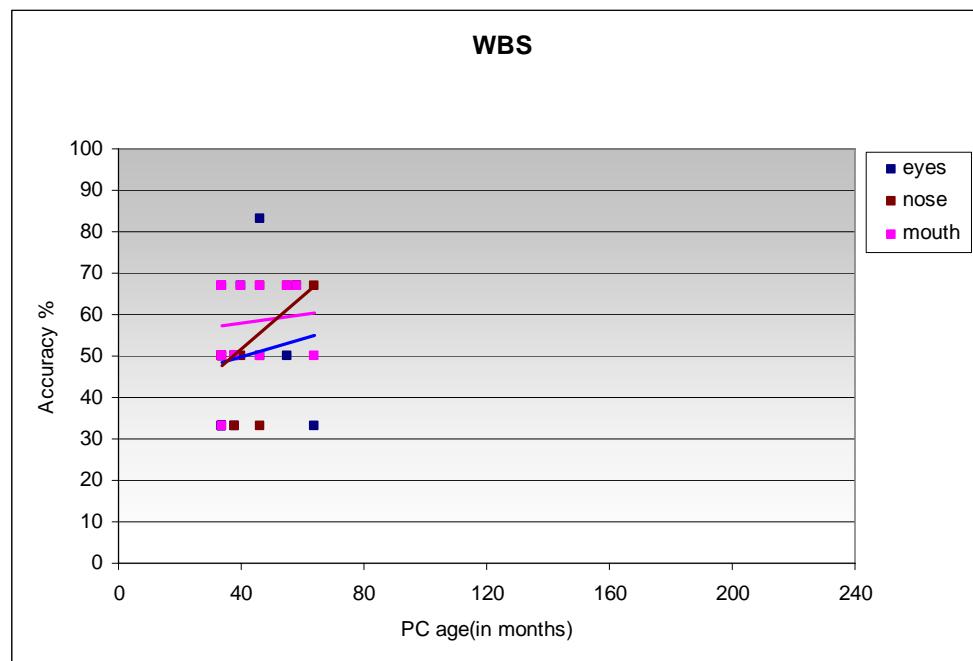


Figure 4: Performance accuracy on first-order configural placement condition against PC age equivalent.

