The Development of Visual Cognition in Infants with Williams and Down's Syndromes.

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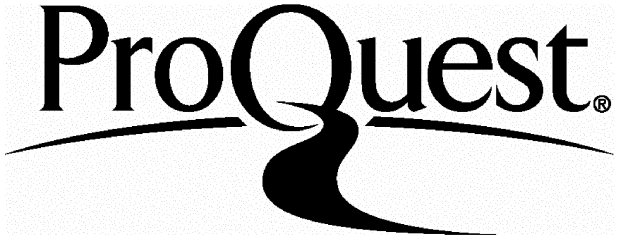

ABSTRACT

Williams Syndrome is a rare genetic syndrome, which exhibits an uneven cognitive profile. In adults with Williams Syndrome, the profile is of impaired visuo-spatial skills, number concepts, and problem solving, with good language and face processing skills. Recent research has shown that language and face processing are not 'preserved' abilities, in that the processes involved differ from those used by normal controls. Down's syndrome is a more common genetic syndrome, with a relatively level cognitive profile. The purpose of this thesis was to examine aspects of visual cognition in infants and young children with Williams and Down's syndromes, to determine whether the cognitive profile found in adults is present from infancy, or whether the end state is a product of the relationship between impaired systems interacting in an aberrant way with environmental factors. Four groups of infants and young children were recruited and tested on a range of measures of visual cognition: children with Williams syndrome, children with Down's syndrome, chronological age matched controls, and mental age matched controls. All children were tested with the Bayley Scales of Infant Development II, in order to obtain a general measure of development for matching purposes. The experimental tasks given to all four groups examined face processing, saccade planning, sustained attention, and temperament. Relationships between measures were also examined. The results indicate that the pattern of impairments found in adults with Williams syndrome are not wholly reflected in infancy. Although precursors of impairment were found for some domains, other areas of difficulty experienced in adulthood were not found in infancy. Findings are discussed in terms of the impact of impairments on the development of other cognitive abilities, and methodological problems in testing atypically developing infants are considered.

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1

WILLIAMS AND DOWN'S SYNDROMES: DEFINING AREAS OF RESEARCH.

1.1 Introduction

The development of abilities in the domain of visual cognition is particularly important in infancy, as in sighted infants these skills guide the infant's selection of information from the environment for further processing. Furthermore, selection of appropriate aspects of the environment to attend to, and processing such stimuli in an appropriate manner, is likely to be influential in learning and developmental outcome. If these processes deviate from the norm in any way, then it would seem to follow that there would be some detrimental impact on the normal path of development.

The study of infants in atypically developing populations, namely Williams syndrome (WS) and Down's syndrome (DS), offers a unique insight into development, from the perspective of comparing systems in infancy with those that are researched and reported in adulthood. This approach is useful in attempting to unravel the developmental trajectory for these populations. Such trajectories culminate in a well-documented cognitive profile in adulthood, but less is known about the developmental journey that takes them there. Thus this approach compares the end state with the starting state, by examining whether strengths and deficits in the adult phenotype are also present in the infant phenotype. If the pattern of strengths and weaknesses found in adult populations is not mirrored by that found in infancy, in the form of predispositions or precursors in domains tested, then this might suggest that the

profile found in adulthood is a product of a possibly different set of impairments in infancy. Thus early differences in these populations may themselves act to magnify the development of impairments.

The focus of this thesis is to compare visual cognition in infants with Williams and Down's syndromes, both with cognitive abilities displayed by their older counterparts, as well as across the infant groups. The purpose of these comparisons is to try to determine what abilities and deficits are present from a very early age, and how specific this pattern of abilities is to each of the syndromes studied. Furthermore, if there are differences between the infant and adult profiles, this research aims to address the ways in which early deficits might impact on other abilities, resulting in the final pattern of performance displayed in adulthood.

Comparison of infant and adult phenotypes in Williams and Down's syndromes also allows for the consideration of two contrasting approaches within the field of cognitive development. The first concerns the flexibility or plasticity of developing cortical functions, and their relationship to cognitive processes. Any differences in cognitive performance between infants and adults in these populations could have resulted from changes at a cortical level, which have occurred at some point between infancy and adulthood. Ascribing validity to this proposal relies on mapping between cognitive behaviours, and what is known about the biological and neuroanatomical structures that support them.

The second approach, which might be considered the converse of the first, is the comparisons that are often made between adult neuropsychological models, and atypical development. This approach tends to view the adult system as intact, with

various deficits within the brain in neuropsychological disorders and patients. When this approach is then applied to infants, the assumption is made that the infant brain can be modelled in a similar way, and that developmental disorders are the product of focal deficits in otherwise intact systems. Through examining the performances of infants with Williams and Down's syndromes, comparing these with adult performance, and with what is known about their neuroanatomy, it is possible to address this issue, and examine whether developmental trajectories are much more complex than have been supposed by such static neuropsychological models.

In this thesis, I aim to provide a summary of the literature on the development of visual cognition, both in typically developing infants, and in infants with Down's and Williams Syndromes, and to report several experimental studies on aspects of visual cognition. This chapter begins with definitions and descriptions of the populations of interest, Williams and Down's syndromes. This will include details relating to the genetic and neuroanatomical backgrounds to these syndromes, as well as a brief overview of cognitive processes, which will be covered in greater detail in the relevant chapters. The issues of cortical plasticity and adult neuropsychological models of development outlined above will also be considered more fully later in this chapter. Finally, research questions addressed in this thesis will be explored in greater depth.

1.2 Williams Syndrome

Williams Syndrome, previously also known as infantile hypercalcemia, is a rare genetic condition characterised by mild to moderate cognitive impairment. It was first recognised as a distinct condition in 1961 (Williams, Barret-Boyes, and Lowe,

1961), and has an estimated incidence of 1 in 20,000 live births. It affects males and females equally, and occurs in all ethnic groups. The most defining physical characteristic of the syndrome is the facial appearance, which is typically described as elfin, consisting of small upturned or retroussé nose, wide mouth, full lips, small chin, and a stellate iris pattern. Characteristic medical features include heart and blood vessel problems, especially supravalvular aortic stenosis, feeding problems and failure to thrive in early infancy, hernias, hyperacusis, and musculoskeletal problems. A small proportion of people with Williams syndrome also present with hypercalcemia.

1.2.1 Cognitive Profile of Williams Syndrome

The cognitive profile of Williams syndrome has attracted the attention of researchers because it presents a distinctive and uneven pattern of abilities and deficits in adulthood. In spite of relatively low IQs, typically in the 50s and 60s, language is relatively spared, while visuo-spatial abilities are poor, with the exception of face processing. Visuo-spatial memory, both long- and short term is impaired, while phonological short term memory is relatively unaffected (Vicari, Brizzolara, Carlesimo, Pezzini, and Volterra, 1996). Motor skills, number, problem solving and planning are also seriously affected (Bellugi, Sabo, and Vaid, 1988; Karmiloff-Smith, Klima, Bellugi, Grant, and Baron-Cohen, 1995). Recent investigation has revealed that what were previously considered 'spared' language abilities are somewhat unusual, in that language typically develops late, spontaneous speech is good, but is marked by unusual words (Bellugi, Bihrle, Neville, Jernigan, and Doherty, 1992), and grammar is somewhat impaired (Karmiloff-Smith, Grant, Berthoud, Davies, Howlin, and Udwin, 1997). Furthermore, examination of language abilities in infants and young children with WS reveal that the advantage for language found in adulthood is

not present early in life, and is just as impaired as language abilities found in matched children with DS (Paterson, Brown, Gsödl, Johnson, and Karmiloff-Smith, 1999).

The overall pattern of relative preservation and impairment of abilities is reflected within the realm of visuo-spatial cognition. Bellugi and colleagues have administered a wide range of visuo-spatial tasks on a group of individuals with Williams syndrome, ranging from 10 to 20 years old (e.g. Bellugi, Bihrle, Jernigan, Trauner, and Doherty, 1990; for a review, see Bellugi, Wang, and Jernigan, 1993). On visuo constructive measures (e.g. block design tasks), they are markedly impaired, demonstrating a selective disability on integration of component parts. Free hand drawing and copying also show an inability to integrate parts. On the Delis Hierarchical Processing Test, individuals with Williams syndrome typically replicate only the local features, without configuring them into the global form. Similarly, on the Block design subtest of the Wechsler Intelligence Scale for Children-Revised (WISC-R), WS subjects fail to reproduce the correct configuration (Bihrle, Bellugi, Delis, and Marks, 1989). Recently, Pani, Mervis and Robinson (1999) have suggested that the core deficit in visuo constructive performance in Williams syndrome is a problem in switching between processing styles once one type of processing has been adopted, rather than a specific deficit in global processing.

Bellugi et al. (1993) have suggested that the pattern of strengths and weaknesses exhibited by WS subjects is comparable with the abilities of right-hemisphere damaged patients. However, visuo-spatial perceptual measures reveal that these similarities are superficial. Unlike right-hemisphere damaged subjects, individuals with Williams syndrome perform well on test of object identification in noncanonical views (Wang, Doherty, Rourke, and Bellugi, 1995).

The pattern of face processing abilities in Williams syndrome further belies the comparison with right-hemisphere damaged subjects. Discrimination of unfamiliar faces, both upright and inverted, does not differ significantly from normal adult performance (Bellugi et al., 1992). Performance on the Mooney Closure Test, which requires identification of degraded silhouettes of faces, is good, even though it is thought that this test taps closure ability rather than face recognition (Bellugi, Sabo, and Vaid, 1988). However, in comparison, performance on tests of closure with non-facial stimuli is poor.

The pattern of good face processing in the presence of poor visuo-spatial skills with non-facial stimuli, in individuals with Williams syndrome, supports the fractionation of face processing as a separate domain. However, it is proposed that face processing as demonstrated by this population may develop differently from normal (Karmiloff-Smith, 1997). Greater attention is paid to facial features, and disturbed processing of inverted faces is less pronounced than in normal subjects. What is of interest is the status of these abilities at birth, and the processes by which they develop to give the unique profile found in older children and adults with Williams syndrome.

1.2.2. Brain Structure of Williams Syndrome

The brain structure of adults with Williams syndrome has been investigated, using both morphometric MRI (e.g. Bellugi, Wang and Jernigan, 1993), and histological examination (Galaburda, Wang, Bellugi, and Rossen, 1994). Total cerebral volume is reduced, specifically parietal, posterior temporal, and occipital regions, although there is a near normal volume relationship of frontal to posterior cortex. Architectonic analysis has revealed that there is also increased cell packing density in several brain regions, exaggerated horizontal organisations of neurons within layers, decreased

myelination, and abnormally clustered and oriented neurons, particularly in the visual cortex.

When compared with matched individuals with Down's syndrome, the cerebellum is significantly larger in WS than DS, the former being normal in size (Jernigan, Bellugi, Sowel, and Hesselink, 1993). Cerebral volume, reduced in WS, is also reduced in DS. However, smaller anterior regions, and greater reductions in temporal limbic structures are reported in DS.

The pattern of brain structure described above has been linked to the behavioural and cognitive profile in Williams syndrome. Galaburda et al. (1994) suggest that reduced dorsal parietal areas, and immature organisation of neurons which are most noticeable in the visual cortex, can be related to the visuo-spatial deficits seen in Williams syndrome. Relatively normal development in limbic, frontal cortical, and neocerebellar structures have been linked to good language and face processing skills (Jernigan et al., 1993). Finally, while it is apparent that brain structure in Williams syndrome differs from normal brain structure in a number of ways, it is important to note that no specific focal lesions are apparent.

1.2.3. Genetic Bases of Williams Syndrome

Recent investigation into the genetic aetiology of Williams syndrome has resulted in the comparison with a vascular system defect thought to be due to deletion on the long arm of chromosome 7. This deletion (locus 7q11.23), is approximately 1.5 megabases, and is believed to encompass the elastin (ELN) gene, as well as LIMK 1, a protein gene expressed in the brain, and other genes contiguous to elastin (Ewart, Morris, Atkinson, et al., 1993; Frangiskakis, Ewart, Morris et al., 1996, Tassabehji,

Metcalf, Fergusson, et al., 1996). This microdeletion may be of maternal or paternal origin (Ewart et al., 1993). This genetic information is now used in diagnosis of the syndrome; the FISH (fluorescent in situ hybridisation) test detects elastin deletion on chromosome 7 in 95-98% of individuals with Williams Syndrome.

The microdeletion reported has led to speculation on the link between deleted genes, neuroanatomy, and behavioural profile in Williams syndrome. Galaburda et al. (1994) suggest that similarities may exist between the elastin gene, and extracellular matrix proteins, some of which are also found on 7q, which contribute in neurodevelopment. The elastin gene has also been implicated in connective tissue abnormalities in WS, including facial dysmorphology and supravalvular aortic stenosis (Ewart et al. 1993). The LIMK 1 gene deletion has been linked to impaired visuo-spatial skills (Frangiskakis et al. 1996).

While such genotype/phenotype links as those described above are exciting to consider, they must be treated with caution. Recent findings suggest that the role of individual genes in phenotype expression may not be as straightforward as has been suggested. A number of individuals have been reported with deletions of the elastin and LIMK 1 genes, who do not necessarily present with typical facial dysmorphology, or deficits in spatial cognition (Tassabehji, Metcalf, Karmiloff-Smith et al. 1999; Karmiloff-Smith, Brown, Grice, and Paterson, submitted). These individuals present with supravalvular aortic stenosis, but despite elastin and LIMK1 gene deletions, present a very different cognitive profile from that typically found in Williams syndrome, suggesting that the relationships between genes and phenotype is more likely to involve a complex interaction between a number of genes.

1.3. Down's Syndrome

Down's syndrome is the most common chromosomal abnormality, and is the leading cause of mental retardation today, with a prevalence of 1 in 800 live births (Dolk, De Wals, Gillerot, et al., 1990). Distinctive physical characteristics include facial dysmorphology, typically upward slanting, almond shaped eyes, wide, flat nose bridge, small mouth, along with flattened back of the head, and shortness of stature. Medical problems typically include congenital heart disease, hearing and vision defects, respiratory tract infections, and poor muscle tone.

1.3.1. Cognitive Profile of Down's Syndrome

Unlike the cognitive profile found in Williams syndrome, the pattern of cognitive abilities in Down's syndrome is typically somewhat more uniform. The delay in cognitive development is generally considered to be at two thirds (Oates, personal communication) to half (Berry, Gunn, and Andrews, 1984) of normal development, and learning difficulties typically increase with age (e.g. Carr, 1985). Language abilities may be somewhat poorer than other cognitive skills (Fowler, 1990), although a few cases of exceptional language abilities have been reported (e.g. Rondal, 1995). In general, spatial skills tend to be better than language skills, although in comparison with Williams syndrome, the cognitive profile of Down's syndrome is more even.

In terms of visuo-spatial processing, subjects with Down's syndrome are impaired on visuo-constructive measures (Bellugi et al., 1990), reproducing global rather than local features in hierarchical processing tests, and on the Block Design subtest of the WISC-R. Picture copying tasks also reveal a tendency to reproduce global configuration, while featural details are omitted (Bihrle et al., 1989). Performance on discrimination of upright and inverted faces is equally poor (Bellugi et al., 1992).

Auditory processing appears to be more problematic for people with Down's syndrome than visual processing (e.g. Rohr and Burr, 1978). This tendency is also reflected in memory, as auditory short-term memory is more impaired than visual short-term memory (Jarrold, Baddeley, and Hewes, 1998).

Traditionally, research in cognitive functioning in children with Down's syndrome has tended to focus on the delay in development, which culminated in the viewpoint that development in Down's syndrome is a slowed down version of normal development. This viewpoint argues that children with DS go through the same stages of development as typically developing children, reaching a plateau in development at around the age of the typically developing six to eight year old. This perspective has recently been challenged by researchers who argue that development in DS is different from that in typically developing infants (e.g. Morss, 1983; Wishart and Duffy, 1990; Wishart, 1993). The challenge is based on findings in several areas of cognition that developmental processes in DS are quite different from those found in typically developing infants. Wishart (1993) reports reversals in competence and task avoidance strategies in children with DS are indicative of a tendency to avoid learning opportunities. Furthermore, when skills are acquired, they are often poorly utilised. Support for the claim that developmental processes in DS differ from those in typical development is also found in results from early intervention programmes, which tend to produce short-term gains, and do not generalise to other areas of cognition (Sloper, Glenn, and Cunningham, 1986). Intervention programmes are largely based on normal development, therefore if development in DS proceeds along a different pathway, it is not surprising that intervention fails to produce significant gains in DS (Wishart, 1991).

1.3.2. Brain Structure of Down's Syndrome

Gross characteristics of Down's syndrome brains include relatively small cerebellum and brainstem, a rounded cerebrum, and reduced cerebral volume. The anterior cortex is small, in relation to the relatively normal sized posterior cortex. Architectonic analysis has also revealed reduced cell packing density, with fewer granular cells reported in visual, auditory, and somatosensory areas (Ross, Galaburda, and Kemper, 1984). Brain stem structures, such as the thalamus, are normal in size (Jernigan, Bellugi, Sowell, et al. 1993).

Studies of brain structure in infancy in Down's syndrome reveal that prenatal brain development appears comparable to that found in typically developing infants (Brooksbank, Walker, Balazs, and Jorgensen, 1989). Brain development seems to be normal until the age of two months, when there may be delayed myelination (Wisniewski, 1991), and by three months, there is a reduction in frontal lobe volume. As with Williams syndrome, no focal lesions are apparent in DS brain structure.

1.3.3. Genetic Bases of Down's Syndrome

The genetic aetiology of Down's syndrome has been shown to result from the presence of extra material on chromosome 21. There are three main types of Down's syndrome, the most common of which is trisomy 21, accounting for 90-94% of cases, in which all the cells have an extra chromosome 21. Translocation, in which extra chromosome 21 material is attached to another chromosome, is found in around 4% of people with Down's syndrome. Mosaicism, in which only some of the cells have an extra chromosome 21 accounts for 2% of incidence of Down's syndrome. To a certain extent, diagnosis can be performed prenatally using the triple test, and amniocentesis.

1.4. Cognitive, Medical and Experiential Equivalence of Down's and Williams Syndromes.

Down's and Williams syndromes may be comparable in several respects, and can provide complementary chronological age and IQ matched groups. Overall, development and intelligence as measured on standardised tests are comparable. Both groups show impairment of Piagetian conservation, and equivalent deficits of concept formation (Bellugi et al., 1992) and problem solving. Both of these syndromes are also associated with delayed language acquisition, and infants from both populations are often encouraged to communicate by Makaton signing at an early age. Therefore both groups are also likely to participate in early intervention programmes, such as the Portage Guide to Early Education (1985), a home-based intervention programme, designed for pre-school children at risk of delayed development. Both groups also tend to display delayed motor development, and can therefore act as controls in terms of tasks that have motor skills components.

While medical conditions differ to some degree between theses two syndromes, there is also a large element of common ground. Both Williams and Down's syndromes tend to present with heart problems, which often necessitate lengthy medical procedures and hospital stays at an early age. Although there has been little research on the effects on development of such procedures at an early age, it can be seen that both populations are comparable in this respect. Finally, although infants with Williams and Down's syndromes tend to have visual problems, which must be taken into consideration in view of the visual nature of the tests administered in this thesis, they tend to present with a similar level of visual impairments. Visual problems in these populations will be discussed in greater depth later in this chapter.

In addition to cognitive and medical similarities, infants with Williams and Down's syndromes are likely to have had relatively similar life events and experiences in their home environment. Parents of infants from these populations will probably have had to come to terms with a diagnosis of the syndrome affecting their child in the first few months or years of the child's life, and may have had to deal with similar effects of diagnosis of medical problems. Therefore in terms of family stress factors, Williams and Down's syndrome infants are comparable. Both groups of infants may have experienced various forms of testing, and thus have both been subjected to different testing environments, and meeting a number of practitioners.

1.5. Selection of Control Groups.

The focus in this thesis is on the cognitive abilities in infants with two developmental disorders, Williams syndrome and Down's syndrome. The equivalence between these groups in terms of general level of experience and developmental delay, described above, means that each group can act as a control for the other on the experimental tasks reported in this thesis. This ensures that differences between these groups cannot be due to any general effects of developmental delay. However, for purposes of ease of exposition, the main focus will be the WS group. In view of the uneven profile reported in adults with WS, it is of interest to compare across domains and across age groups, to try to decode the underpinnings of the syndrome. Therefore to some degree the Down's syndrome population will act as a control group for WS. However, it is also of interest to investigate and report the performance of infants with Down's syndrome in the studies reported in this thesis, as several of the tasks have not previously been administered to this population.

Two groups of typically developing infants/young children were also selected for inclusion in the studies reported here; mental age-matched controls (MA), and chronological age-matched controls (CA). MA infants are included to provide a control for general level of development, while CA children provide a control for length of experience and exposure to environmental stimuli.

The selection of control groups in studies of developmental disorders has attracted criticism, as once again the question is raised of what is being matched. Matching criteria are typically based on the scores from standardised tests, depending on the domain of interest. However, Wishart and Duffy (1990) suggest that the use of control groups matched on the basis of mental age infers that the developmental processes in the two groups are the same. This is not the assumption here. Indeed, it is the processes underlying development in Down's and Williams syndrome which are of interest in this thesis. Specific problems concerning matching also arise in relation to the measure used in the studies reported here, the Bayley Scales of Infant Development II (Bayley, 1993), but these will be covered in detail in chapter 2.

1.6 The Status of the Visual System in Infancy and Early Childhood.

In considering the realm of visual cognition, it is important to establish abilities in visual perception, to ensure that differences between groups are not due to perceptual problems. While it may often be difficult to establish where perception ends and cognition begins, it is useful to review the visual capabilities of the populations studied.

1.6.1. The New-born's Visual System.

The infants visual capabilities at birth support a certain amount of visual processing, but there is a great deal of further development during the first year of life. At birth, the optic nerve is incompletely myelinated, and the eye diameter and the pupillary opening are both smaller than in adults (Reese and Lipsitt, 1970). Evidence from animal studies suggests that the concentration of cones in the fovea is lower than in adults, only reaching adult levels after migration of rods toward the periphery, and cones toward the centre (Hendrickson and Kupfer, 1976). Visual acuity, as measured by preferential looking, is about one-thirtieth of adult levels at birth, but improves rapidly over the first few months. At birth, contrast sensitivity is also around one-thirtieth to one-fortieth of that found in adults (Atkinson and Braddick, 1981). Binocular vision is not thought to develop until the end of the fourth month (Held, 1985).

The new-born has a focal distance of approximately 19 cm (Haynes, White and Held, 1965), can follow a moving stimulus (Dayton, Jones, Steele and Rose, 1964), and indeed shows a preference for moving stimuli (Slater, Morison, Town, and Rose, 1985), although smooth tracking of moving stimuli is not generally found until 2 months of age (Aslin, 1981). The time taken to plan and execute a saccade is substantially longer than in adults, and the visual target is typically reached through a number of eye movements, each of which undershoot in the direction of the target (Aslin and Salapatek, 1975). In summary, it can be seen that although the new-born infant has a certain degree of control over eye movements, there are many respects in which infant vision and the visual system differ from those of adults. However, the development of infant vision occurs rapidly, in certain areas reaching adult levels in a mere few months. Therefore the visual system of the infant is capable of supporting a

large amount of visual processing, as demonstrated in the studies reviewed here, but for each measure used, it was important to establish that the particular processes being tested were not beyond the infant's basic visual abilities.

1.6.2. Visual Processes in Williams and Down's Syndromes.

Individuals with Williams and Down's syndromes both tend to experience visual problems, and in infancy this is often in the form of strabismus. A larger proportion of children with Williams syndrome are strabismic than is generally found in the normal population, which leads to deficits in stereoscopic vision (Anker, 2000; Anker and Atkinson, 1997). Visual acuity is also affected, but appears to improve with age. However, when tested on a battery of visuo-perceptual tasks, no differences were found between children with Williams and Down's syndrome (Govers, Borghgraef, Descheemaeker et al. 1997).

Stimuli in the tasks reported here are both two and three dimensional. It must be acknowledged that infants Williams and Down's syndromes may be somewhat disadvantaged on tasks using three dimensional stimuli, as stereoscopic deficits in these groups may imply problems with depth perception. However, while unlikely, it is possible that these groups compensate for a deficit in stereoscopic vision through a greater dependence on other cues used in depth perception. Although there may be some doubt as to perceptual equivalence between the atypical and typically developing groups, it is reasonable to assume that perceptual abilities are not dissimilar in Williams and Down's syndromes. Differences between these groups on tests of aspects of visual cognition are therefore unlikely to be due to low-level perceptual deficits.

1.7. Approaches to Studying Developmental Disorders: Addressing Fundamental Issues.

As stated in the introduction, the study of developmental disorders, and more specifically the comparison of infant and adult phenotypes, can be used to address two contrasting approaches to the study of the functional architecture of the cognitive system. The first is essentially a preformist view, from a neuropsychological perspective, that cognitive systems are largely preformed, or innate. It is further claimed that such innate cognitive systems are modular, and double dissociations in adult neuropsychological patients are often offered to support this claim. This preformist view has been applied to developmental disorders, where parallels have been drawn between acquired and developmental disorders. An example of this approach can be seen in the literature on developmental dyslexia. Temple and Marshall (1983) report on HM, a developmental dyslexic child, who resembled adult phonological dyslexics. These latter tend to have problems reading non-words. HM is then contrasted with another dyslexic child, CD, who has problems reading irregular words, and thus resembles adult surface dyslexics (Coltheart, Masterson, Byng, Prior, and Riddoch, 1983). This type of developmental double dissociation is then taken as evidence that reading systems are preformed, and that the atypically developing brain is just like a normal brain, in which some parts (modules) are impaired. Although most proponents of this approach agree that there is some impact on development of interactions between genes and environment, the role of the environment is to a great extent reduced to that of triggering innately specified systems.

The contrasting approach to the preformist view comes from developmental cognitive neuroscience, which is much more concerned with the dynamics of development. It is acknowledged that innate constraints are present, but rather than specifying modular

systems, these constraints are initially merely domain relevant, and only become domain specific through the process of development. In relation to developmental disorders, adult phenotypes are viewed as the endpoint of this process, where impairments in cognitive skills are the product of differences in the infant phenotype interacting throughout development both with themselves, and with environmental factors. Thus very small differences in infancy may result in atypical interactions, which in turn cause the developmental trajectory to veer further from the normal path of development. This approach highlights how comparisons between acquired and developmental disorders makes assumptions about the developing brain and cognitive systems, that may result in the actual processes which culminate in the adult phenotype being overlooked. This approach can be found in the recent literature on the development of Down's syndrome. As stated in section 1.3.1, several researchers have questioned the classification of development in Down's syndrome as simply a slowed-down version of normal development (Morss, 1983; Wishart and Duffy, 1990; Wishart, 1993). Instead, it is claimed that the developmental pathway in Down's syndrome is different, and not just delayed.

Both of these contrasting approaches have been applied in the literature regarding Williams syndrome. The preformist approach has taken the documented uneven cognitive profile in Williams syndrome as support for innately specified cognitive modules. Thus WS is represented as having intact language and face processing modules, but an impaired visuo-spatial processing module (Pinker 1994; 1999). However, these claims are based on performance in adults and older children with Williams syndrome, and make the assumption that the adult phenotype will mirror the infant phenotype. Furthermore, such claims are based on an oversimplification of studies of Williams syndrome, which increasingly report that abilities such as

language and face processing are not 'intact', in that the cognitive processes involved in successful behaviour differ from those seen in the normal population.

The constructivist developmental cognitive neuroscience approach views the pattern of cognitive abilities in Williams syndrome in more relative terms. Thus language and face processing are spared relative to visuo-spatial processing, in that people with Williams syndrome are more proficient in the first two areas, compared to more severe problems in the latter. It is proposed that the cognitive profile in adulthood culminates from a potentially different profile in infancy. The infant with Williams syndrome begins with small genetic differences, which may influence processing styles and abilities, and which results in atypical specialisation of cognitive functions as development progresses. Thus over- or under-specialisation for cognitive functions may occur, resulting in the pattern of relative strengths and weaknesses (Karmiloff-Smith, 1998), while still differing in many respects from normal cognitive functioning. This approach has been applied in a study of language and number skills in young children with Williams and Down's syndromes (Paterson et al. 1999). Infants with Williams syndrome performed well on a test of numerosity judgement, even though this is poor in adulthood. Conversely, the proficiency in vocabulary found in adulthood in Williams syndrome was not found in infants.

Examination of cognitive functioning in infants with Williams and Down's syndromes, and comparisons with findings from adult populations, will facilitate an evaluation of the two approaches outlined above. Comparing the infant and adult phenotypes will help determine whether the pattern of abilities found in adulthood is already present from an early age, which would tend to support the preformist argument. Alternatively, if infant performance is different from that found in adults,

this would indicate the presence of dynamic developmental influences, as proposed by the neuroconstructivist developmental cognitive neuroscience approach. Investigation in a number of areas within the domain of visual cognition, which represent areas of strengths and weaknesses in Williams and Down's syndromes will further facilitate this comparison. In the next section, specific areas in which these comparisons will be made are discussed.

1.8. Defining Areas of Research.

The realm of visual cognition is extensive, therefore certain limitations must be imposed on the areas within this field which can be considered. To a certain degree, these limitations are dictated by pre-existing knowledge about the particular abilities of the populations being considered. In addition, the areas of interest are also selected in part because of their interdependence, as well as the manner in which they relate to each other. The areas of visual cognition that will be addressed are face processing, sustained attention, and spatial frames of reference used in saccade planning. These will also be compared to more general measures, including performance on a standardised test of development, and temperament. A review of the literature on each of the areas outlined above will be presented in the relevant chapters. This will include literature on both typical and atypical development in the areas concerned. Finally, relationships between all such areas will be examined, in an attempt to unravel the interaction between impaired and relatively intact abilities at an early stage in Williams syndrome.

1.8.1. Matching in Atypical Populations.

In chapter 2, the issues concerning control groups and matching in studies with atypical populations are discussed in detail. This chapter also contains details about all the participants, who formed the basis of the samples for all other tasks reported in this thesis. Results from administering The Bayley Scales of Infant Development II, and establishing the control groups are also reported.

1.8.2. Spatial Frames of Reference.

In chapter 3, aspects of visuo-spatial skills will be examined. As stated previously (sections 1.2.1. and 1.3.1.), visuo-spatial skills are impaired in both Williams and Down's syndromes. However, different classes of impairment are generally found; while both groups tend to have problems on visuo-constructive measures, in Williams syndrome this is seen as a problem recreating global aspects of figures, while for Down's syndrome the problem appears to be with local features (Bihrle, Bellugi, Delis, and Marks, 1989; Bellugi et al. 1990). While these results appear to present a pleasing dichotomy, what is pertinent here is how infant performance compares to that of the adult.

Spatial frames of reference are a type of mental representation that utilises a co-ordinate system used to process spatial information, and have been demonstrated to become more sophisticated during the course of normal development. The investigation of spatial frames of reference in infants with Williams and Down's syndromes might indicate whether differences here can account for differences exhibited in later childhood and adulthood, and whether these can be related to neural mechanisms, such as those proposed to account for spatial deficits in Williams syndrome. This will be examined in chapter 3.

1.8.3. Sustained Attention.

Although little empirical work has been reported on attention in Williams syndrome, attentional problems are often remarked upon by parents and teachers. Attention has been reported as being poor in Down's syndrome children (e.g. Fisher, 1970), and adults (Tomporowski, 1990), although it has also been suggested that poor attention is not a uniform trait across all individuals with Down's syndrome (Green, Dennis and Bennets, 1989).

Sustained attention is generally held to be the phase of attention after orienting, in which effortful processing takes place. Differences in sustained attention that might be revealed between atypically and normally developing infants, may highlight early factors which compound later developing deficits. The key issues here are whether infants with Williams and Down's syndromes exhibit deficits of sustained attention, whether any deficits are the same in both populations, and the contribution these might make to developmental outcome. A study of sustained attention in Williams and Down's syndromes will be reported in chapter 4.

1.8.4. Temperament.

In chapter 5, the role of temperament in development is addressed. Of particular interest are dimensions within measures of temperament which relate to attention, in order to further explore any differences between groups from the study of sustained attention. While temperament in Down's syndrome has been widely researched, little work has been done on temperament in Williams syndrome, although anecdotal evidence suggests that several personality problems may be common in adults with Williams syndrome. The focus here is primarily on attentional aspects of temperament, but it is also of interest to determine whether personality aspects

claimed to be a factor in adults with Williams and Down's syndromes are also present from an early age.

1.8.5. Face Processing.

Face processing presents one aspect in which adults with Williams and Down's syndromes exhibit very different abilities, therefore it would be of interest to determine whether these differences exist from an early age, or develop later as a product of different deficits. The exact nature of face processing in infants with Williams syndrome is also worthy of investigation, as this is reported to be an area of proficiency in adulthood, although recent investigations have begun to demonstrate that the processes involved are somewhat atypical. If face processing is abnormal in older children and adults with Williams syndrome, but more representative of normal processing in infants, or exhibiting different abnormalities, then this would indicate that some degree of environmental input is involved, and also that plasticity within the cortex may be implicated. In chapter 6, a study of face processing is presented which looks at the creation of prototypical faces in Williams and Down's syndromes, and control groups.

1.8.6. Relationships Between Measures: an Examination of Group Coherence.

In chapter 7, the relationship between the measures reported in this thesis will be examined. This is an attempt to understand the impact of deficits in one area, on other areas of cognition. It may also reveal whether or not some areas of impairment are key deficits in the populations studied. While it is acknowledged that the complexity of such an interaction would demand a great deal more work to establish the underlying pattern, this type of exercise will hopefully give some indication of the processes involved.

An investigation of the coherence and structure of the groups studied will also be reported. Variability within groups will be examined, in order to determine the uniformity, or diversity, of groups. This is particularly important in the case of Williams syndrome, because research already suggests that sub-groups exist within the Down's syndrome population (e.g. Green, Dennis, and Bennets, 1989). If this is the case, then the same may be true in Williams syndrome. This may be of relevance from an educational point of view, in terms of determining appropriate intervention and training strategies, as well as informing other researchers in this area.

1.9. Experimental Design and Procedural Considerations

The areas of research outlined above will be tested in a cross-sectional design, looking at the performance on tasks of WS, DS, MA, and CA groups. At the outset, it was also the intention that a longitudinal component would be included in these set of studies, in order to chart development within individuals from the different groups. However, due to attrition rates, the number of infants tested more than once was deemed too small to allow comparisons across groups.

Testing atypically developing infants presents a challenge in terms of task design. It is important to present tasks which are engaging enough to keep the child's attention, but which also do not overtax the child's abilities, while also tapping into the measure of interest. Several of the tasks reported in this thesis are based on measures normally used with younger age-groups. Modifications were therefore made to the original tasks so that they were more appropriate for the age groups tested here. However, although every effort was made to ensure that task design fulfilled the above criteria,

through extensive piloting, it must be acknowledged that designing tasks for atypically developing infants can always be improved.

Each infant was seen twice; one test session to administer the Bayley, and a second in which all other experimental studies were run. A 15-20 minute warm-up session took place before each session. Most of the studies were short (5-10 minutes), and varied in terms of equipment used, in order to maintain infants' interest. If the infant became upset, or parents or experimenters felt that a break was necessary for other reasons, testing would stop for a short period. However, some infants did not complete all the tasks, and participant details will be given for each experimental chapter, as there are slight differences for each study. On a few occasions, infants who were tested twice completed tasks on the second visit that they had refused on the first, and on these occasions, data from the second session are included, and noted in the appropriate section on participants.

1.10. Summary

The tasks and abilities reviewed in this thesis, and the cognitive structures that support them, represent only a fraction of the literature on the developmental cognition. However, the findings reported give an indication of the current debate on the nature of infant knowledge within the realm of visual cognition. The key issues, relating to cortical plasticity and comparisons of developmental disorders with adult neuropsychological models, will be considered in light of the studies reported here, and ways in which the study of infants with Williams and Down's syndromes may go some way toward aiding the resolution of this conflict will be presented.

2

STANDARDISED ASSESSMENT OF COGNITIVE DEVELOPMENT IN ATYPICALLY DEVELOPING POPULATIONS.

2.1. Introduction

This purpose of this chapter is to review a standardised assessment of overall cognitive development in atypically developing populations, and to establish the validity of using such a test as a matching tool. This will then be used to determine participants to be included in the experimental and control samples for the experimental studies which follow. To obtain a standardised measure of development, the Bayley Scales of Infant Development II (BSID II, Bayley; 1993) were administered to each infant. The aim of the BSID II is to assess the current developmental functioning of infants and children over the first 42 months of life. Scores from this test were used to establish global equivalence in the WS, DS, and MA groups, and to ascertain that children in the CA and MA groups were functioning within normal limits. Data will be presented from this assessment as to how infants in each of these groups were matched.

The use of standardised assessment tests with atypically developing groups has attracted some criticism. The focus of this debate concerns stability of performance, and consequently questions whether or not performances on developmental assessments by atypically developing populations are a true demonstration of their

abilities, or merely a measure of performance on one particular occasion. Although addressing the issue of performance stability is not one of the aims of this chapter, this issue will be reviewed in this section, in order to justify the use of standardised tests for the purposes described.

2.1.1 The Role of Standardised Tests of Cognitive Development

The development of standardised tests of cognitive development in infancy resulted from the growing realisation that rapidly advancing abilities that emerge during infancy required specific tests to explore developmental milestones at different ages. Thus the purpose of such tests is to establish whether or not a specific ability has been acquired. The development of standardised assessment tests was also motivated by the need for early identification of children who deviate from normal development. This usually means children exhibiting developmental delay, although such tests are also used to assess children with accelerated development. Once a child has been identified as exhibiting developmental delay, an intervention programme can, in principle, be designed and implemented, in an attempt to help the child reach his or her full potential. Further testing is then recommended in order to chart the child's progress, and to modify intervention programmes.

Over the last 30 years, a number of standardised tests of infant intelligence has been developed and used in both clinical and research settings. However, it has been demonstrated that the relationship between scores on infant development tests and later IQ scores is not a strong one, before the age of 24 months (Lewis and Brooks-Gunn, 1981), when emerging language allows for a different type of task to be administered. Several critics have asked whether such infant tests, as administered to

infants under the age of two, are not simply measures of perceptual and motor skills, rather than of cognitive processes. Bornstein, Slater, Brown, et al. (1997) point out that in the Mental scale of the BSID II, items for infants under the age of two years largely measure perceptual-motor development, such as 'eyes follow rod', and 'fingers hole in pegboard'. By the age of two, there are fewer perceptual-motor tasks; items are more cognitive, relating to abilities such as verbal comprehension and comparison of mass.

Several researchers have embarked upon studies designed to establish alternative tests of infant intelligence. Fagan (1984, 1985) suggests that the abilities measured by existing standardised tests of infant development do not relate to intelligence in later childhood, and that novelty preference and habituation in infancy are much better predictors of later intelligence. Similarly, Messer, McCarthy, McQuiston, et al. (1986) argue that mastery motivation in infancy is a better predictor of later cognitive performance than standardised infant development tests. Mastery motivation is typically defined as persistent task-directed behaviour, which is independent of the task being carried out. Bornstein et al. (1987) present a review of measures of information processing and causal understanding as predictors in infancy of later intelligence. While this area of research is promising in terms of the development of new tests of infant intelligence, until psychometric problems such as poor predictive validity and reliability can be resolved, existing standardised tests of infant development must remain the dominant form of assessment.

2.1.2 The Bayley Scales of Infant Development II

The Bayley Scales of Infant Development-Second Edition (BSID II) was revised and developed from the Bayley Scales of Infant Development (BSID; Bayley, 1969). It is one of the most widely used tests of infant development. It was standardised on a sample of 1700 children in the United States of America, 100 at each of the 17 age groups identified in the test, and is reported as having good reliability and validity (Bayley, 1993). The scale consists of three subtests; the Mental Scale, the Motor scale, and the Behavior Rating Scale, and the age range covered is from 1 to 42 months. Although both the Mental Scale and the Behavior Rating Scale were administered during testing in this thesis, in the current study the focus will be mainly on the Mental Scale.

The Mental Scale consists of items designed to tap a number of emerging abilities in infancy and early childhood, such as problem solving, number concept, language development, and sensory integration. Each age group has a specific set of items to be administered, with a cue sheet suggesting the order of administration. Consecutive item sets overlap to a degree. Items increase in difficulty through the set, based on the performance of children from the standardisation sample, ranging from 90% of the children of a given age passing an item located early in the item set, to 15% of the same aged children passing an item towards the end of the set. The item set to be administered is determined by the child's chronological age, although the Bayley manual suggests that premature children under two years may be tested at their corrected age.

For a pass (or credit, as it is termed in the Bayley manual) to be scored, the child has to complete a task under the criteria laid out in the manual. There is often a time limit for task completion, or a fixed number of trials to be administered. Other scoring options for each item include refusal to perform the task, omitting the task, and parental report that although the child has not performed the task during the test session, it is a task that parents have seen the child perform. However, anything other than a pass is not entered into the total score.

After administering a complete item set, basal and ceiling rules determine whether testing should stop, continue to the next item set, or go back to next lower set. Because of the overlap between item sets, administering an additional item set involves testing with fewer items than the original set. Some items may be scored from performance on the original item set. After administering the test, a raw score is calculated, which consists of the number of items passed within an item set, added to the total number of items before that set. The raw score is then converted into a Mental Development Index (MDI) using the child's chronological age. The MDI can then be used to classify the infant as exhibiting normal, accelerated, or delayed development. A developmental age can also be derived from the raw score.

2.1.3 Specific Aspects of BSID II Mental Scale Scores

The primary objective in administering BSID II to all the infants in this study was to aid in matching between groups. However, it was also important to demonstrate reliable administration of the test, both in terms of different experimenters administering the test, as well as the place of testing, which was either at the infant's home or in the lab.

In addition to a global score of development, the Bayley II also yields facet scores, in which individual test items are grouped to give scores for specific areas of development, i.e. language, social, motor and cognitive. Facet scores are achieved by finding the developmental age level at which a child passes a predominant number of items. However, over the developmental age of 13 months, many age groups have only one or two items per facet, and predominance at a specific age level is harder to determine.

By extending the idea of facet scores further, it is possible to examine groups of items of particular interest in the populations being tested. The Bayley manual suggests:

> 'you may also examine the child's performance on an item or set of items to gain more insight into that performance. Interpretation at the item or subset level should be undertaken with considerable caution, given the unreliability (i.e. lack of replicability) of any single item. However, one can look at a child's performance on an item or subset of items to generate hypotheses for further evaluation.' (pp. 229).

One application of this might be to look at spatial items within the Bayley, as it is well documented that adults with Williams syndrome are spatially impaired. For example, people with Williams syndrome show serious visuo-constructive deficits in drawing tasks. One way to examine whether this tendency exists in infants and young children with Williams syndrome would be to look at their performance on a subset of visuo-constructive tasks within the Bayley, such as block construction items.

2.1.4 Applying BSID II to Atypical Populations

In order to establish clinical validity of the BSID II, data for children from several high-incidence clinical groups, including premature infants, HIV+ infants, and infants with Down's syndrome, were collected by the test developers. The Down's syndrome group consisted of 60 children with chronological ages ranging from 3 to 42 months, and mental ages (not established on the BSID II) ranging from 2 to 30 months. When tested on the Bayley, this group performed below all the other clinical groups, and had a mean Mental Development Index (MDI) score greater than three standard deviations below the mean of the standardisation sample. From this clinical sampling, the Bayley is claimed to be appropriate for testing children at risk for developmental delay, although the manual suggests that repeated test sessions are the best way to establish level of functioning in children, particularly those with one or more risk factors which might result in development that deviates from the norm. The manual urges caution in the interpretation of individual results in relation to the data presented from the clinical sample, and concludes by stressing that further research is required to better determine the effectiveness of the BSID II as used with children who differ from the standardised sample.

Several problems arise when using standardised assessment tests on groups with atypical development, only some of which are addressed by the Bayley manual. The first of these concerns the possible instability of the performance of DS infants on standardised cognitive tasks. Stability of performance in typically developing children has been investigated during standardisation of the Bayley, and it is reported as having good test-retest stability, with a correlation coefficient of .87 for the Mental Scale for all ages (Bayley, 1993). However, several studies have shown that infants with DS fail to consistently pass the same items when tested repeatedly. Wishart and

Duffy (1990) gave a series of object search tasks to 18 DS children, three at each of the following ages; 6, 12, 18, 24, 36, and 48 months. These tasks were re-administered one to two weeks later to check stability of performance. Performance varied on the two sessions within each of the age groups on at least one task level, all task levels varied overall, and performance varied in over 50% of subjects. Wishart and Duffy also tested the same children over the same intervals with the first version of the Bayley, the Bayley Scales of Infant Development. Although raw scores did not differ between first and second tests, analysis of individual item to item agreement showed that there was a high degree of item instability. Similarly, Morss (1983) found that DS infants were significantly less likely to repeat successful task performance in an object concept task than normally developing infants. These findings may indicate that successful task performance in infants with Down's Syndrome may not necessarily represent a stable acquisition. Moreover, they also indicate that single test sessions may not reveal the developmentally delayed child's competence, but simply reflect performance on that occasion.

Wishart and Duffy propose that factors such as task engagement and motivation may contribute to performance instability in DS children. They found that task failure was often due to DS children's refusal to participate, thereby failing the task by default. DS children, they argue, will often engage in elaborate avoidance behaviours. However, several studies claim to find no difference between typically developing infants and DS infants in terms of task persistence (e.g. MacTurk, Vietze, McCarthy, et al., 1995; Ruskin, Kasari, Mundy, and Sigman, 1994). In addition, Hasan and Messer (1997) found no indication of failure to engage in six children with DS, when tested on Uzgiris and Hunt object concept and means-ends scales. Furthermore, Hasan and

Messer claim to find stable performance with these DS children when tested repeatedly at monthly intervals.

The second issue in relation to administration of standardised assessment tools with atypically developing populations concerns the level at which to start testing. As previously stated, the starting point for typically developing infants is determined by their chronological age. The selection of starting points when testing atypically developing infants is far less easy to determine. The Bayley manual advises selecting the item set closest to the child's current level of functioning, using any available information, such as caregiver report or other test results. This approach allows for a large amount of variability in the item set selected, which in turn can have a dramatic impact on the score achieved. Because all the items below the item set administered are credited as a pass, extra points can be achieved through the selection of one item set rather than another. For example, by administering the 17-19 month set, rather than the 14-16 month set, the child would automatically be credited with 10 more points, before the addition of the score from that item set.

When testing a child with an item set outside his or her chronological age, the Bayley manual advises that adhering to the basal and ceiling rules will ensure that the score obtained is a valid one. However, the length of the test sessions may indicate that a child may fall within the criteria for the item set by failing latter items because of lack of interest or fatigue. Testing sessions for children over 15 months can be as long as an hour, and although the manual advises that it is possible to take a break and restart later, the child's motivation and energy levels may result in failing the necessary amount of items within an item set. Although this approach is certainly to be recommended when using the scales in a clinical assessment setting, in order to

establish the child's true level of functioning, it lends itself less to the research setting where it is often used as a tool to establish matched control groups.

The issue of ambiguous starting points has been addressed in the literature concerning administration of the Bayley with premature infants. Ross and Lawson (1997) tested premature infants at 7 months and 12 months (corrected age) on either the chronological age item set, or corrected age item set. Although mean differences between the groups tested on chronological or corrected age was small, considerable differences were evident for individual children. In reply, Matula, Gyurke, and Aylward (1997) state that the item set selected should be as close as possible to the chronological age, as they acknowledge that the item set selected will have an impact on a child's score. However, the case of infants with Down's syndrome may require the use of item sets that are considerably further removed from their chronological age than the difference between chronological age and corrected age in premature infants. As previously mentioned, the delay in cognitive development in Down's Syndrome infants is generally considered to be at half (Berry, Gunn, and Andrews, 1984) to two-thirds (Oates, personal communication) of normal development, and may decelerate over time (Niccols, Latchman, and Williamson, 1999). Therefore the selection of item sets to administer with this group can be ambiguous and problematic.

Although it can be seen that there are several problems associated with using BSID II as a matching tool for atypically developing populations, if used with caution it can provide a basic level on which to match groups. Thus with the samples tested in the present studies, the approach was to provide a level playing field, by administering the same item set within age groups. This was done to provide a level from which to

begin exploring the processes underlying development in the different groups involved, without assuming that the processes are the same in delayed and non-delayed infants. Indeed, the emphasis for the remaining experimental studies is the possible difference between processes employed by typically and atypically developing infants.

In summary, the aims of this section are to establish validity for using BSID II as a matching tool across groups, ensuring reliability of administration, and to explore in particular visuo-spatial performance within the test.

2.2 Method

2.2.1 Design

A between subjects design was used, with four groups: WS, DS, CA, and MA.

2.2.2 Participants

The original sample consisted of 14 WS infants, 21 DS infants, 18 CA-matched infants, and 21 MA-matched infants. These infants were seen as part of a larger study at the MRC Cognitive Development Unit, and are now being seen at the Neurocognitive Development Unit, Institute of Child Health. WS infants were recruited through the Williams Syndrome Foundation, and all were positive on the FISH test for elastin deletion on chromosome 7. DS infants were recruited through the Down's Syndrome Association, as well as through normal recruitment procedures employed at the Cognitive Development Unit. All infants with Down's syndrome were full trisomy-21. CA and MA infants were recruited through the Cognitive Development Unit.

From the original sample of infants tested, a number were excluded for different reasons. 2 DS infants were excluded. One presented with nystagmus, or wobbly eye syndrome, and one had severe epilepsy. One of the CA group was excluded due to accelerated development on BSID II. Five infants were excluded from the MA group, 3 due to delayed development, and 2 due to accelerated development. This left a sample of 14 WS, 19 DS, 17 CA and 16 MA. Mean ages and age ranges for all groups are presented in Table 2.1. All participants were reported as having normal, or corrected for normal vision. Those who normally wore glasses did so during testing. Infants in the WS, DS and CA groups were tested as close as possible to 24, 30 and 36 months. MA infants were matched to the WS and DS infants on the basis of developmental ages obtained from the BSID II.

Table 2.1 Chronological age in months of the main sample.

Group	Mean	Minimum	Maximum
WS (n=14)	29 months	23 months 16 days	37 months 7 days
DS (n=19)	29 months 3 days	23 months 25 days	36 months 25 days
CA (n=17)	29 months 28 days	23 months 13 days	36 months 28 days
MA (n=16)	15 months 6 days	11 months 24 days	20 months 15 days

2.2.3 Procedure

The children were tested in the presence of the caregiver, either at home, or at the Cognitive Development Unit (CDU). Preferred location for testing was the CDU, and all infants tested there would visit on a separate occasion from other tests, to ensure that they were not overtired. For families with WS and DS children who had travelled a long distance, this often meant two sessions on consecutive days, staying in a hotel overnight. However, this was not always feasible, and for those families who were unable to attend the CDU on two occasions, a home visit was arranged as close as possible to the other test session, to administer the Bayley.

Before testing began, a warm-up session of 15 to 20 minutes allowed the child to become more familiar with the experimenter, and the environment, for those tested in the lab. Testing in the lab took place in a quiet room, with all visual distractions removed. When tested in the home, parents were encouraged to remove siblings to another room. Children were tested seated in an infant seat, at a table opposite the experimenter, with the parent or carer seated beside the child. Children who would not settle in the infant seat were tested on the parent's lap. All sessions were videotaped.

The point at which testing was started within the Bayley, for the infants with Williams and Down's syndromes, was determined from piloting, and the same item set was administered to infants in these groups within age sets. Thus all WS and DS 24-month-olds were tested with the 13-month item set, 30-month-olds with the 14-16 month item set, and 36-month-olds with the 17-19 month item set. The CA and MA groups were tested using item sets according to their chronological age.

The starting point for matching was the WS infants. DS infants were matched to each WS infant on the basis of chronological age and gender, and as closely as possible on raw scores obtained from the BSID II. CA infants were matched to the experimental groups on chronological age, and MA infants on developmental age, achieved by transforming raw scores obtained by the clinical groups. Both CA and MA groups were also matched to WS and DS groups on gender.

As previously stated, in both the CA and MA groups, infants were dropped from further analysis if they demonstrated delayed or accelerated development. MA infants who failed to match WS and DS infants on developmental age were also excluded.

Visuo-spatial subset

Three experimenters, experienced in administering the Bayley, independently compiled a list of visuo-spatial items from the items within the Bayley which were administered to the WS, DS, and MA groups. Data from the CA group are not included, as item sets administered to this group were different to those administered to the WS, DS, and MA groups, and consisted of fewer visuo-spatial items, because of the difference in age. Lists were cross referenced and only items which appeared in all three lists were then identified as a subset of visuo-spatial items for further analysis. Items identified for the subset are as follows;

79 Fingers Holes in Pegboard
86 Puts Three Cubes in Cup
87 Places One Peg Repeatedly in 70 seconds
89 Puts Six Beads in Box
90 Places One Piece (Blue Board) in 150 seconds
92 Closes Round Container
93 Places Circle Piece (Pink Board) in 180 seconds
95 Puts Nine Cubes in Cup
97 Builds Tower of Two Cubes
98 Places Pegs in 70 seconds
104 Uses Rod to Attain Toy
112 Places Four Pieces (Blue Board) in 150 seconds
115 Completes Pink Board in 180 seconds
119 Places Pegs in 25 seconds
120 Completes Reversed Pink Board
123 Builds Tower of Six Cubes
130 Completes Blue Board in 75 seconds
132 Places Beads in Tube in 120 seconds

2.3 Results

The majority of the WS and DS groups failed to obtain an MDI score, and were categorised as MDI < 50. Raw scores will therefore be reported throughout.

2.3.1 Group Raw Scores

Average raw scores on BSID II for all groups are shown in Table 2.2. (Individual raw scores are presented in the Appendix). To ascertain that the MA, DS, and WS groups were matched as to developmental age, a one-way Anova was carried out on the data. There was a significant difference between groups ($F(3,62) = 89.56$; $p < 0.0001$), the CA group scoring significantly higher than all the other groups (Tukey's HSD, $p < 0.05$). Therefore the MA, DS, and WS groups can be seen to be equivalent in terms of developmental age.

Table 2.2 Mean BSID II Raw Scores.

Group	Raw Score	
	Mean	SD
WS (n=14)	101.43	10.00
DS (n=19)	100.89	9.96
CA (n=17)	146.76	9.30
MA (n=16)	99.44	10.81

2.3.2 Reliability of Administration

Four experimenters were involved in testing, although 2 of them (JHB and SJP) administered most of the tests. Rates of testing for experimenters are: JHB: 62%, SJP: 27%, SH: 5%, MG: 6%. Overall percentages for location of testing are: CDU: 62%, Home: 38%. Raw scores for each group by a) different testers, and b) different location of testing are presented in Table 2.3.

Table 2.3 Bayley Raw Scores by Location and Tester.

		TESTER								PLACE			
Age	Grp	JHB		SJP		SH		MG		CDU		HOME	
(mo)		Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
24	WS	90.50	6.25	97.50	2.12	/	/	/	/	89.50	9.19	94.50	4.80
	DS	93.86	3.81	88.00	*	/	/	/	/	91.50	4.95	93.67	4.13
	CA	138.25	1.71	139.00	*	135.00	*	/	/	137.83	1.94	/	/
30	WS	104.50	3.70	105.00	2.83	/	/	/	/	106.50	3.54	103.75	2.99
	DS	99.00	2.28	117.00	*	/	/	/	/	102.00	*	101.50	7.79
	CA	143.00	*	147.00	4.00	145.00	*	/	/	145.40	3.58	149.00	*
36	WS	117.50	6.36	/	/	/	/	/	/	/	/	117.50	6.36
	DS	116.33	4.51	112.00	*	/	/	/	/	118.50	3.54	112.00	0.00
	CA	153.00	5.66	/	/	/	/	162.00	2.64	158.40	5.98	/	/
	MA	99.50	10.93	101.17	12.19	101.00	*	87.00	*	99.44	10.81	/	/

* denotes no SD available, as experimenter tested only 1 child in this category.
/ denotes no children in this category tested by experimenter.

To check whether there was any difference between scores achieved by different testers, an ANCOVA was carried out, with tester as the factor of interest, and co-varying for group, age, and place of testing. No significant differences were found in relation to tester ($F(3, 65) = .567$; n.s.). The same procedure was applied to the place of testing data, co-varying for group, age, and tester. The effect of place of testing was not significant ($F(1,65) = 3.51$; n.s.). Therefore it has been established that factors such as different testers, and different locations for testing did not have an effect on scores.

2.3.3 Visuo-spatial subset

The scores in Table 2.4 represent visuo-spatial items passed as a percentage of visuo-spatial items attempted. Examination of the means shows all groups scored similarly.

A one-way Anova was performed on the data. No differences were found between groups ($F(2,46) = 0.13$; $p >$ n.s.).

Table 2.4 Mean percentage of visuo-spatial items passed.

	Mean	SD
WS (n = 14)	64.79	17.00
DS (n = 19)	63.33	16.48
MA (n = 16)	61.59	17.34

2.4 Discussion

The purpose of this chapter was to establish validity for using the BSID II as a matching tool across groups. The results show that the WS, DS, and MA groups are matched in terms of developmental age, and that extraneous factors such as different testers or location of testing did not affect scores. Administration of the Bayley also allowed further refinement of the sample, by identifying and then excluding infants in the two typically developing groups who were performing outside normal limits, exhibiting either accelerated or delayed development.

A sub-set of spatial items were selected by three experienced Bayley testers, and scores on these items for WS, DS, and MA infants were analysed. This analysis was performed in order to determine whether the visuo-spatial deficits found in adults with Williams syndrome, and visuo-spatial abilities in adults with Down's syndrome, are present from an early age. However, no difference was found between groups. One possible interpretation of this result concerns the type of item administered. All the items administered were from sets aimed at under 2 year olds. As mentioned in the introduction, these items typically measure perceptual-motor development. Therefore, failure to find a deficit on visuo-spatial items at the ages tested here could imply that

the visuo-spatial impairments found in adults with Williams syndrome is indeed a cognitive deficit, and is not influenced by perceptual-motor impairments. Although this is a tempting perspective, the nature of the analysis reported here cannot really support it. As stated in the Bayley manual, interpretation at the sub-set level should be done with extreme caution. The item set selected for this analysis has not been standardised, and therefore this result cannot be viewed as reliable.

Ideally, when using standardised tests with atypically developing infants, tests which have not established test-retest stability outside the normally developing population should be administered twice. In addition, the practice of administering the same set of items to each child within age groups results in scores that may not be an accurate representation of each child's abilities. The issue of stability of performance in the WS and DS groups, as mentioned in the introduction, has not been addressed here, as only a small number of WS and DS infants provided data from two test sessions, and repeat data from typically developing infants were not collected. When dealing with atypically developing populations, particularly those such as Williams syndrome which are rare, repeated test sessions are not always practical, and even less so when the age groups to be tested are young infants and toddlers, often travelling long distances for testing. Finally, while administering one item set to each age group does not give a truly clinical score for each child, it does have the advantage of ensuring that the same conditions apply within groups, and is easier and quicker to run. This obviates problems of overly long test sessions with infants of this age.

BSID II is by no means ideal as a matching tool when dealing with atypically developing groups. However, if used with caution, and provided results are

interpreted with some restraint, then it can act as a starting point of global equivalence for examination of other specific abilities.

3

SPATIAL FRAMES OF REFERENCE

3.1 Introduction

One of the earliest ways in which the developing infant can begin to investigate his world is through visual exploration. Before motor control has developed sufficiently to allow the infant to explore the environment by touching or grasping, visual exploration allows the infant to interact with his world, and to begin to exert some control over his responses to it. In addition, the selection of aspects of the environment for further processing by the infant also shapes learning. Therefore it can be seen that the development of mental representations used to plan saccades, in order to select appropriate aspects of the environment to attend to, is of major importance in infancy. Furthermore, the measurement of eye movements is often used in studies of infant cognition, therefore it is important to establish the infant's capabilities, in order to understand what such movements might signify.

Successful spatial orientation relies upon accurate perception of the physical self in relation to the environment, and adaptation to the changes both within that environment and of the body. Spatial knowledge may be represented by frames of reference, a co-ordinate system used to code positions in space which can then be used to monitor stimuli and plan eye movements. The particular type of mental representation used will dictate how efficiently different aspects of visual stimuli can be processed and integrated.

3.1.1 The Development of Spatial Knowledge

As infants develop, increasingly complex representations of spatial information are used throughout infancy and then childhood, before adult representations are formed. Some authors claim that at around the age of six months, children use egocentric representations, i.e. they use body- or head-centred coordinates (e.g. Bremner, 1978a, 1978b). Between six and 16 months, there is an increasing tendency to use geographic, sometimes labelled non-egocentric, representations (Acredolo, 1978). This has been related to increased mobility in the infant, which allows greater exploration and interaction with spatial aspects of the environment (Gustafson, 1984). At this stage the infant also needs to be able to update spatial information due to self movement. The transfer to using geographic frames of reference can also be accelerated, as demonstrated in studies which have increased the salience of geographic objects (Acredolo and Evans, 1980), and have also increased familiarity of the environment (Acredolo, 1979). Acredolo (1976) further distinguishes between different forms of geographic representations, which progress from proximal to distal, and towards decreasing mobility of reference objects. Bremner (1993) cautions that there may be a tendency to oversimplify the coding systems used to represent spatial information, as infants may code using multiple frames of reference at the same time. Finally, adult representations are typically allocentric, which although sometimes equated with geographic representations, may be considered as a particular form of non-egocentric representation (Pick, 1988), and perhaps the most sophisticated.

Recent research has demonstrated that the egocentric spatial frames of reference found in 6-month-olds are not present from birth. Using a double-step saccade paradigm, Gilmore and Johnson (1997) have demonstrated that 3-month-old infants are unable to combine retinal information and information about eye movements, and

are therefore dependent on retinocentric representations. However, when 7-month old infants are tested on the same task, their responses are predominantly egocentric, i.e. they can combine retinal and non-retinal (or extra-retinal) information. The task required infants to look at an array of stimuli which was presented briefly on a monitor, such that two saccades were required in rapid succession in order to locate both targets. The correct sequence of saccades required integration of retinal information concerning the position of the first and second target, and information concerning the eye position, which altered after the initial saccade to the first target. The ability to integrate retinal and non-retinal information requires the infant to update spatial information, similar to the updating which takes place in older, locomotive infants who have progressed to using geographic frames of reference.

3.1.2 Neural Bases of Spatial Knowledge

Gilmore and Johnson relate the difference between 3- and 7-month-olds use of spatial frames of reference, to the shift from subcortical to cortical control over eye movements during the first few months after birth (Johnson, 1990). Before the age of two months, it is proposed that visual processing is dominated by the subcortical pathway from the retina to the superior colliculus. This serves to influence the slower developing cortical system by orienting the infant to salient environmental stimuli, which is necessary for the development of spatial processing circuits within the parietal cortex. This would suggest that the specialisation of such circuits is not wholly predetermined, but is to some extent shaped by the quality and quantity of experiential input.

The shift from subcortical to cortical control of saccade planning may be revealed by performance on a recently developed marker task. Johnson, Gilmore, Tucker, and Minister (1996) propose that vector summation, in which saccades are made to the average position between two targets, is a key characteristic of visual processing in the subcortical pathway from the retina to the superior colliculus. Johnson et al. (1996) found that vector summation, as measured by end-points of saccades made in response to two simultaneously presented stimuli, progressively declined in 2-, 4-, and 6- month-old infants, which they claimed was indicative of the gradual shift from subcortical to cortical control of eye movements.

The transfer to cortically controlled eye movements, as indicated by egocentric spatial frames of reference and decline in vector summation, occurs in typically developing infants by the age of six to seven months. The effect of delayed maturation of the cortical mechanism is not known, but it may have an impact on the development of visuo-spatial abilities.

3.1.3 Two Cortical Visual Pathways

Ungerleider and Mishkin (1982) first proposed that there are two cortical visual systems in the adult brain. The 'what' pathway, which follows a ventral occipito-temporal route, allows for object identification. The 'where' pathway, located in the dorsal system from occipital to parietal lobes, supports the processing of spatial locations. Goodale and Milner (1992) also propose a model of two cortical visual pathways, but argue that the distinction between pathways concerns the output from (rather than input to) visual processing. As in the Ungerleider and Mishkin model, there is a 'what' pathway, which is used in the perception and recognition of objects,

but rather than a 'where' pathway, they suggest a 'how' pathway, which is concerned with the perceptual control and guidance of actions. The parietal cortex, one of the areas within the dorsal visual pathway, has been shown to be involved in head or body centred representations for saccade planning. Chen and Nakamura (1998) present a review of lesion and neuropsychological studies, which indicate that the rat posterior parietal cortex is largely implicated in coding for head- or body-centred space. This evidence would seem to support the further specification of the dorsal pathway as an action planning route. Goodale and Milner further suggest that the model can be divided into multiple streams within the two pathways, with interconnections between various systems.

Recent research suggests that the dorsal and ventral pathways in the two cortical visual pathways model do not emerge simultaneously in infancy, with the ventral pathway appearing before the dorsal (Atkinson, 1998). In developmental terms this would appear to be reasonable, as initially infants can rely on the subcortical orienting mechanism in order to locate and attend to important environmental stimuli, such as faces.

3.1.4 Visuo-spatial Processing in Infants with Down's and Williams Syndromes

Both Down's syndrome and Williams syndrome are associated with deficits of visuo-spatial processing in childhood and adulthood. In Williams syndrome, these take the form of absence of integration on visuo-constructive measures (Bellugi, Bihrle, Jernigan, Trauner, and Doherty, 1990), and impaired long- and short-term visuo-spatial memory (Vicari, Brizzolara, Carlesimo, Pezzini, and Volterra, 1996), although visuo-perceptual abilities appear to be less affected (Wang, Doherty, Rourke, and

Bellugi, 1995). In a comprehensive study of the cognitive abilities of three adolescents with Williams syndrome, Bellugi and colleagues found deficits in tasks relating to spatial knowledge (Bellugi, Sabo, and Vaid, 1988). These children exhibited severely impaired performances on the Benton Line orientation test (Benton, Varney, and Hamsher, 1978), which involves matching the spatial orientation of lines to an array. However, they performed normally in a test for unilateral visual neglect, thus demonstrating that spatial attention is evenly distributed in both hemispheres, and across the environment.

While people with Down's Syndrome also exhibit visuo-constructive deficits, they differ from Williams Syndrome in that they tend to focus on global features, while in Williams Syndrome the focus is more on local features (Bellugi et al., 1990). However, children with Down's Syndrome who have been identified as demonstrating impaired performance on block construction tests, are able to identify and recognise block displays, as well as 2-dimensional representations of such displays (Stratford, 1980). However, these results are based on the performance of older children with Down's Syndrome, and comparatively little is known about spatial abilities at a very early age, particularly regarding the nature of frames of reference adopted to guide visual saccades in infancy.

While relatively little is known about the manifestation or origins of spatial deficits in early infancy in Williams syndrome, a recent study shows impairments on a visuo-spatial task with WS children, some of whom were as young as 4 years, and also offers an account of the impairment in neurological terms. Atkinson, King, Braddick, et al. (1997) propose that the visuo-spatial deficits found in Williams syndrome may result from impairments in the dorsal cortical stream of visual processing. As

previously mentioned, this system is proposed to be involved in processing spatial location, and visual guidance of action. Atkinson and co-workers tested fifteen children with WS, aged 4-14 years, and a control group, on three tasks; two dorsal and one ventral. They found that the WS children were impaired relative to controls on the two dorsal tasks; a preferential looking measure of motion coherence thresholds, and a visuo-manual card posting task. Although Atkinson et al. argue that these results support the claim that there is a dorsal stream deficit in Williams syndrome, they caution that the wide variability in performance exhibited by the WS group is unlikely to be explained fully by a dorsal stream deficit, and that contributions from other neural bases are likely to be involved.

One of the factors suggested to contribute to the typically child's developing sense of spatial perception is self-locomotion, which is believed to impact on the child's ability to use geographic frames of reference (Acredolo, 1978). Infants with Williams and Down's syndromes tend to exhibit delays in achieving motor milestones, although there is a wide variation in motor skills within each of these groups (Kataria, Golstein and Kushnick, 1984; Cobo-Lewis, Oller, Lynch, and Levine, 1996). If mobility is important in the development of more sophisticated spatial frames of reference, then it might be argued that delayed mobility could have a detrimental effect on this area of development.

The aim of this study is to test infants with Williams and Down's syndromes using the double-step saccade paradigm employed by Gilmore and Johnson, to determine whether later developing visuo-spatial deficits are marked by early eccentricities in terms of spatial frames of reference used to guide visual action. The task requires spatial location processing. Therefore it is proposed that infants with Williams

syndrome will be impaired on this task, relative to typically developing controls. This would support the Atkinson et al. account of the impaired dorsal pathway in Williams syndrome. It is also proposed that infants with Williams syndrome and Down's syndrome may differ from typically developing infants in terms of vector summation, which would indicate delayed maturation of the cortical control mechanism.

3.2 Method

3.2.1 Design

Four groups were tested in a between-subjects design; Williams Syndrome (WS), Down's Syndrome (DS), chronological age-matched controls (CA), and mental age-matched controls (MA).

3.2.2 Participants

13 WS infants, 19 DS infants, 17 CA infants and 15 MA infants were tested from the main group reported in chapter 2. Mean ages and age ranges for all groups are presented in Table 3.1.

Table 3.1 Mean and range of ages for each group.

	Mean	Minimum	Maximum
WS (n=13)	28 months 27 days	23 months 16 days	37 months 7 days
DS (n=19)	29 months 3 days	23 months 25 days	36 months 25 days
CA (n=17)	29 months 28 days	23 months 13 days	36 months 28 days
MA (n=15)	15 months 9 days	11 months 24 days	20 months 15 days

3.2.3 Procedure

The infants were placed in a raised car seat 50 cm from the centre of a 51 cm colour monitor, and the room lights were dimmed. In the case of infants who would not settle when their carer left the immediate area, carers took up position directly behind the car seat. Those infants who would not settle at all in the car seat were tested on the carer's lap. The sessions were recorded from a video camera mounted above the monitor, zoomed in to obtain a close-up view of the infant's head and eyes. A time code generator was used to allow for subsequent frame-by-frame coding.

Each session consisted of distracter stimuli which were started before the infant was in place, followed by 18 calibration trials, 48 experimental trials, and 12 break trials. Breaks were presented after each block of four experimental trials. Eight combinations of fixation-target sequences were presented in pseudo-random order in each pair of blocks of experimental trials. Fixation and target stimuli were high contrast designs, 4 degrees in width, accompanied by random auditory tones.

Each experimental trial began with a fixation display appearing in one of 4 positions, centre right, centre left, centre top, or centre bottom. The experimenter monitored the infants reactions on video, and when it was determined that the infant was looking at the fixation display, a computer key press would terminate the fixation display, and the target stimuli would appear. Target durations were determined from pilot studies, and were 70 ms for target 1, and 100 ms for target 2. Target 1 was always in one of the two positions diagonally adjacent to the fixation point, with target 2 always positioned opposite target 1, as in Figure 3.1A. Targets were 17 degrees from the middle of the fixation point.

A 1.5 second response interval was allowed before the start of the next trial. On trials where the infant was not fixating, attempts were made to attract his attention to the correct area of the screen, by calling his name and encouraging him to look at the picture, or by shaking bells in the correct position behind the monitor. Continued failure to fixate resulted in the trial being sacrificed in order to maintain the infants' interest.

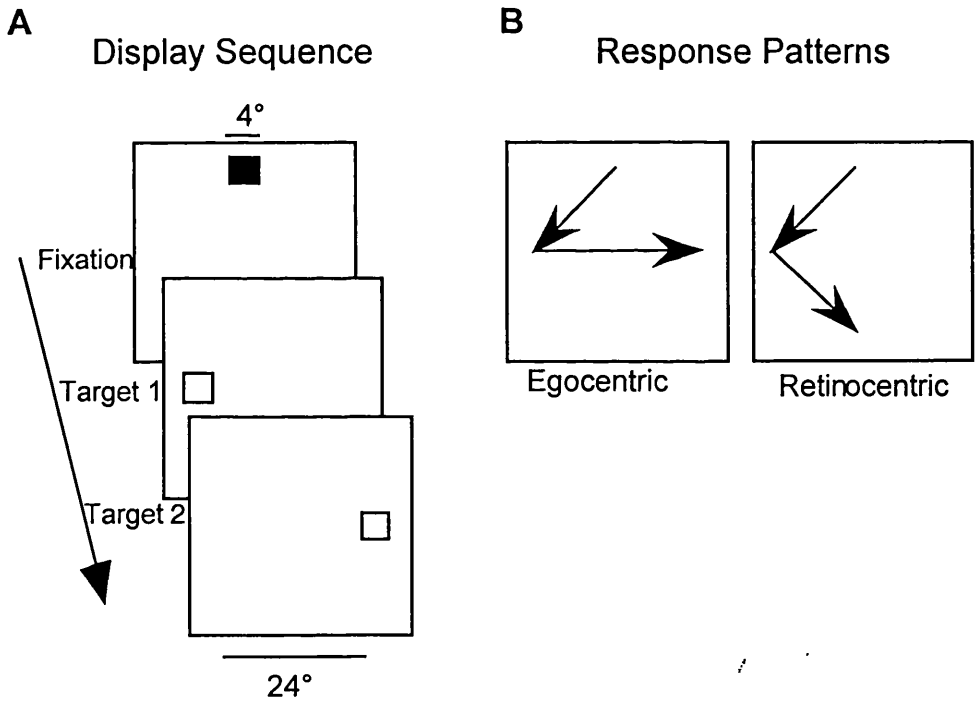

Figure 3.1 (A) Presentation of a fixation stimulus was followed by two identical targets that flashed sequentially. Fixation positions varied in pseudo-random order, between four possible locations: centre top (as shown), centre bottom, centre left, and centre right. (B) Response types: a body-centred sequence consisting of two saccades, one to each target location; and a retinocentric sequence consisting of two saccades, each equivalent to the positions of the targets at the time of presentation to the fovea.

3.2.4 Coding Eye Movements

Eye movements were subsequently analysed independently by 2 trained coders, one of whom was the author, and the other was completely naïve to the experimental hypothesis. Both coders were blind as to trial type. Any disagreement between coders resulted in that trial being coded independently by a third coder, or judge. Onset times and end locations of first and second saccades were coded from PAL format videotapes, i.e. 50 half frames per second. Location was coded as one of nine positions on an imaginary 3x3 grid. Each coder would spend approximately two hours coding each participant. Trials were only coded as valid if the following criteria were met:

1. Infants made a valid first saccade, i.e.;
   Infants were fixating to the correct location at the onset of the first target.
   First saccades were made to valid locations, i.e. not away from the monitor.
   Infants made at least one saccade after fixation.
2. Trials were not included when the first saccade was made before the offset of the second target, in order to ensure that retinal and non-retinal information was separate.
3. The starting times of saccades as determined by coders corresponded to within one frame either way, and locations of the endpoint of a saccade was the same for both coders. Any discrepancy between coders was judged by a third coder, who would code the trial independently, and reject one or both decisions.

Any trials which failed to meet any of these criteria were excluded from the main analyses. Videotape measures of this type have been shown to be comparable in terms of accuracy with electro-oculogram (EOG) data (Tucker et al., 1996). After coding

was completed, trials were then categorised as egocentric, retinocentric, or other (see Figure 3.1B).

3.3 Results

3.3.1 Reliability

Reliability between two of the three coders was calculated based on the number of trials where two of the three coders agreed on start times and endpoints of both saccades, as a proportion of trials completed, and are as follows;

All = 92%, WS = 90%, DS = 91%, CA = 92%, MA = 95%.

Table 3.2 Trials Completed and Eliminations (% of trials completed) for all Groups.

	Trials Completed (out of 48 possible trials)		% Valid Trials (valid trials as a % of trials completed)		Elimination 1 (no fixation, first look, or valid 1st)		Elimination 2 (first look before 2nd target offset)		Elimination 3 (disagreement between coders)	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
WS (n=13)	45.77	5.29	51.15	15.56	36.05	19.25	3.05	3.96	9.76	7.92
DS (n=19)	47.74	1.15	59.37	12.92	23.96	13.06	7.47	9.00	9.25	7.92
CA (n=17)	42.94	10.05	58.12	15.75	22.45	11.54	10.83	13.60	8.46	7.33
MA (n=15)	45.6	4.44	65.33	13.56	24.94	10.40	4.84	4.88	5.08	5.35

3.3.2 Valid Trials and Elimination Rates

Table 3.2 shows the trials contributed by each of the groups, and the pattern of elimination of trials. Univariate one-way Anovas were carried out on the percentage of valid trials contributed by each of the groups, and on each of the elimination measures. There was no significant difference between groups on percentage of valid trials ($F(3,60) = 2.243$, n.s). For the elimination measures, there was a significant difference between groups on Elimination 1 ($F(3,60) = 2.903$, $p < 0.05$), WS infants

scoring higher on this measure than CA infants (Tukey's HSD, $p < 0.05$). There were no group differences on Elimination 2 ($F(3,60) = 2.126$, n.s.), or Elimination 3 ($F(3,60) = 1.275$, n.s.). Thus WS infants had significantly more eliminations due to no valid first look than the CA infants.

3.3.3 First Saccade Responses

A summary of first saccade types (as a percentage of all saccades) is presented in Table 3.3. The types of saccade presented are:

Target 1: Looks to the position of the first target

Target 2: Looks to the position of the second target

Centre: Looks to the central (vector averaged) position

Other: Looks to any other position on the 3x3 grid

Table 3.3 First Saccade Types.

	Target 1		Target 2		Centre		Other	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD
WS (n=13)	26.36	9.33	34.87	16.82	26.58	13.37	12.18	5.12
DS (n=19)	49.49	12.99	33.98	12.93	10.47	10.27	6.06	4.41
CA (n=17)	48.51	20.92	38.51	19.45	6.60	5.91	6.38	4.86
MA (n=15)	43.63	12.77	48.51	14.41	4.32	5.55	3.59	4.88

Univariate one-way Anovas were carried out on each of the first saccade response types. There was a significant difference between groups on saccades to Target 1 ($F(3,60) = 7.359$, $p < 0.001$), saccades to the Centre ($F(3,60) = 16.517$, $p < 0.0001$), and saccades to Other positions on the 3x3 grid ($F(3,60) = 7.911$, $p < 0.001$). Differences on all these measures were found to be between the WS group and all other groups (Tukey's HSD, $p < 0.05$), with the WS group scoring lower than other groups on looks to Target 1, and higher then other groups on looks to the Centre, and

Other positions. There were no significant group differences on saccades to Target 2 ($F(3,60) = 2.680$, n.s.).

3.3.4 Second Saccade Responses

Response types made in the second saccade are presented in Table 4. Responses presented are those made after a saccade to the first target, and response types reported are:

Egocentric: Successful looks to the egocentric position

Retinocentric: Looks to the retinocentric position

Fixation: Looks back to the position of the fixation point

Other: Looks to any other valid position on the 3x3 grid

No Look: No second look made

Table 3.4 Second Saccade Types

	Egocentric		Retinocentric		Fixation		Other		No Look	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
WS (n=13)	19.14	18.53	10.35	15.64	9.61	15.94	8.40	11.79	52.48	30.78
DS (n=19)	47.72	13.85	11.14	10.83	4.93	5.04	13.27	12.77	22.94	14.60
CA (n=17)	53.46	21.95	14.33	23.70	6.64	6.72	4.36	6.65	21.22	15.79
MA (n=15)	51.04	10.52	8.58	9.34	8.64	7.87	6.61	8.13	25.13	12.87

One-way Anovas were performed on each of the second saccade measures. Significant differences were found on Egocentric looks ($F(3,60) = 12.55$; $p < 0.001$), and No Second Looks ($F(3,60) = 8.48$; $p < 0.001$). Differences on these measures were found between the WS group and all the other groups (Tukey's HSD, $p < 0.05$). Differences at the level of Retinocentric looks, Looks back to Fixation, and Other looks were not significant.

3.3.5 Ratio of Egocentric Responses

Two ratios of egocentric responses were then calculated. The first was the ratio of egocentric to egocentric plus retinocentric responses, and the second was the ratio of egocentric to all second responses. These are presented as they represent a more accurate depiction of the pattern of response types than the simple measure of proportion of valid trials which resulted in egocentric responses. This calculation eliminates the trials on which there was no second saccade, which represent a large number for the WS group. Mean scores of both measures for all groups are shown in Table 3.5.

Table 3.5 Ratio of Egocentric Responses

	Ego: Ego+Retino		Ego:All Second Looks	
	Mean	SD	Mean	SD
WS (n=11, n=12)	64.61	44.24	45.13	40.90
DS (n=19)	82.31	16.51	63.56	19.42
CA (n=17)	81.21	25.05	67.58	23.72
MA (n=15)	86.16	14.43	68.87	14.45

Two WS infants did not contribute any egocentric or retinocentric looks, and therefore a meaningful ratio of egocentric to egocentric plus retinocentric responses cannot be calculated for these infants. Similarly, one WS infant failed to provide a meaningful ratio of egocentric to all second looks. These infants have therefore been excluded from this analysis.

Due to non-homogeneity of variance in both data sets, attempts were made to transform the data in order to normalise the variance. This was successful for the Ego:Ego+Retino ratio, using a cubed transformation (Box and Cox, 1964). However, a suitable transformation was not revealed for the Ego:All Second Looks data, when a

Box and Cox diagnostic plot (1964) was applied, therefore untransformed data were entered into the analysis. A one-way Anova was performed on the transformed Ego:Ego+Retino data, but there were no significant differences between groups ($F(3,58) = 0.1589$, n.s.). A Kruskal-Wallis one-way Anova was performed on the Ego:All Second Looks data, but although scores for the WS group were lower than for all other groups, there was no significant difference between groups (chi-square (3) = 4.0229; n.s).

3.3.6 Exploring Individual Differences in Williams Syndrome

Although there were no significant differences between groups for either measure of ratio of egocentric responses, standard deviations for the WS group were much larger than for other groups. This would seem to indicate that there was a large variation in the response patterns of infants with WS, and the raw data seemed to support this, as can be seen from a representation of the Ego:All second looks ratio (Figure 3.2).

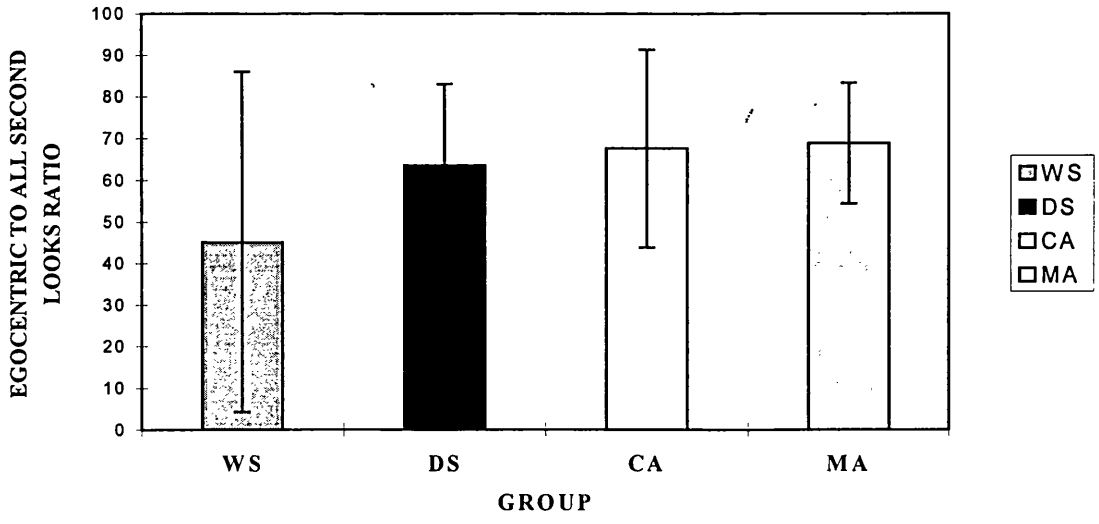

Figure 3.2 Ratio of Egocentric:All second saccades for all groups (Mean +/- SD).

This raised the possibility that there may be subgroups within the WS population, which at a basic level might be described as one group which could perform the task, and one group which could not. In order to explore this possibility further, each of the raw ratio scores for the WS group were divided into two groups using a median split. The Ego: Ego + Retino data had a median of 85.71, and as there were 11 infants in this group, the infant with the median score was excluded from this analysis. The Ego: All Second Looks data had a median of 41.67. Thus for each ratio score there were two WS groups: one group with a high ratio score, and another with a low ratio score.

Having established the proposed WS subgroups on the basis of ratio of egocentric responses, the results of the response types for first and second saccades were calculated for these new groups, and are presented in Table 3.6. The results for the median split of each of the ratio measures were compared with independent samples t-tests, in order to ascertain that using the median split had resulted in discrete groups. There were significant differences between the high scoring and low scoring groups on both ratio measures (Ego:Ego + Retino; $t(8) = -4.74$, $p < 0.05$; Ego:All Second Looks; $t(10) = -5.56$, $p < 0.001$). Therefore using the median split had resulted in groups which differed significantly from each other in terms of both measures of egocentric ratio score. These groups were also compared on their raw scores from the Bayley Scales of Development II, in order to determine whether this grouping would be reflected in terms of mental age. However, there was no difference between groups on Bayley raw scores (Ego:Ego + Retino; $t(8) = 0.20$, n.s; Ego:All Second Looks; $t(10) = -0.39$, n.s).

Table 3.6 Results of Median Split of Egocentric Ratios

	Ego: Ego+Retino				Ego:All Second Looks			
	WS Low (n=5)		WS High (n=5)		WS Low (n=6)		WS High (n=6)	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD
Median split	25.00	35.36	100.00	0.00	11.10	17.20	79.17	24.58
Bayley	99.2	11.15	98.00	7.65	99.67	9.99	102.17	11.99
%Valid Trials	45.42	14.91	53.12	16.63	50.70	7.98	50.52	22.63
Target 1*	26.10	11.15	29.16	9.07	27.40	12.47	27.25	4.94
Target 2*	33.26	25.20	40.84	11.09	36.87	16.89	34.37	19.31
Centre*	29.54	18.27	19.30	5.72	26.87	14.82	24.55	13.50
Other*	11.10	5.71	10.69	3.29	8.86	2.72	13.83	4.34
No Look†	34.67	32.02	61.33	21.71	40.00	33.62	57.05	21.91

* = measures from the first saccade, † = measure from the second saccade

Comparisons were also made between high and low ratio groups on the other measures which were recorded during the task, in order to establish whether or not the proposed subgroups in Williams Syndrome differed throughout the task. If these groups also differed on measures such as ability to locate the target on the first saccade, this might indicate that differences between WS infants on the egocentric ratio measures would be due to a more general deficit, rather than a specific problem using egocentric frames of reference. The only measure which will be reported from the second saccade is the 'No Looks' measure, as all other measures contributed to the median split data.

The mean results from the two groups obtained from the median split of the Ego: Ego + Retino ratio appear to reveal some interesting differences (see Table 3.6). However, although the low ratio group scored higher than the high ratio group on looks to the centre for the first saccade, and failure to make a second look, on the second saccade, these differences were not found to be significant when independent samples t-tests were applied to the data (Centre; t (4.78) = 1.20, n.s., t-value for unequal variances

reported, as Levene's test for equality of variance was significant, No Look; t (8) = -1.54, n.s.). In terms of the nature of the proposed subgroups, this result indicates that the groups did not differ on measures which did not directly contribute to the Ego: Ego + Retino ratio.

The mean results obtained when the Ego: All Second Looks ratio data was split to form two groups were also examined (see Table 3.6). There do not appear to be any large differences between groups on any of the measures from the first saccade, but the high ratio group appear to be making fewer second looks than the low ratio group. However, this difference was not significant when analysed using an independent samples t-test (t (10) = -1.04, n.s.). The only measure from the first saccade on which there were significant differences between groups was looks to other positions, in which the high egocentric ratio group scored higher than the low ratio group (t (10) = -2.38, $p < 0.05$).

In summary, the results of analysis of the groups formed from a median split of the Ego: Ego + Retino ratio indicate that these groups differed only on the egocentric ratio. This would appear to suggest that the deficit in the low ratio group is a specific problem using egocentric spatial frames of reference. Results of analysis of groups formed from a median split of the Ego: All Second Looks ratio demonstrated that the high ratio group looked more to other positions on the first saccade than the low ratio group. This prompted comparison with the other groups, as a difference on this measure might indicate that although the high ratio group was performing well on the task, they may not have been processing information in the same way as the control groups. When a one-way Anova was performed on looks to other positions on the first saccade, including data from DS, CA, MA, and the two new WS groups, there was a

significant difference between the high ratio WS group, and all the control groups ($F(4,58) = 5.912$, $p < 0.001$; Tukey's HSD, $p < 0.05$).

3.4 Discussion

The aim of this study was to test the hypothesis that infants with Williams syndrome would be impaired on the task using the double-step saccade paradigm, relative to normally developing chronological and mental age matched controls, and relative to the mental age matched Down's syndrome group. More specifically, it was proposed that the ratio of saccades indicating egocentric frames of reference would be smaller for the WS group than other groups, indicating a dorsal route deficit in the WS group. The hypothesis that WS infants would also show more vector summation than control groups, as exhibited by looks to the central location, was also tested.

3.4.1 Results of the Dorsal Stream Impairment and Vector Summation Hypotheses.

The results show that overall the children with Williams syndrome performed very differently from children with Down's syndrome, chronological age matched controls, and mental age matched controls. The difference between the WS group and all other groups on the ratio of egocentric looks failed to reach significance, and therefore the hypothesis that WS infants as a group are impaired on the task, is not supported. There is, however, some indication that some of the WS infants may be impaired in terms of using egocentric spatial frames of reference. Finally, the finding that WS infants exhibited significantly more looks to the centre location than other groups supports the hypothesis that WS infants are more reliant on sub-cortical mechanisms of saccade planning than other groups. However, the pattern of results exhibited by the WS group revealed some unexpected findings, and overall present an intriguing

package, which needs to be unravelled to determine the nature of WS performance on this task.

3.4.2 Eliminations and their Implications

Trials were eliminated on the basis of 1) failure to contribute a valid first saccade, 2) saccades made before the offset of the second target, and 3) disagreement between coders in terms of time or endpoints of saccades. Analysis of the response rates and elimination of trials for each group showed that the WS group had significantly more trials eliminated on the basis of failure to contribute a valid first saccade, than the CA group. Because different responses were grouped together within this category of eliminations (lack of fixation, looks away, and no first look), the cause of the difference between these groups cannot be established. Therefore factors such as obligatory attention or other differences in attention between the groups cannot be eliminated as contributing to these results at this stage. Obligatory attention is the tendency to display prolonged orientation to stimuli (Stechlar and Latz, 1966), and is generally found in one to three month olds. It could be argued that if WS infants had this type of attention disengagement problem, this would result in fewer looks being made after fixation by this group. More general differences in attention could also contribute to fewer looks being made, as this could result in trials being terminated in an attempt to maintain the infant's interest. The nature of attention in the groups studied here will be addressed in Chapter 4, and the relationship between the results of this study and those of the attention study reported in Chapter 4, will be examined in Chapter 7.

3.4.3 First Saccade Responses

For the first saccade, the measures reported were proportion of looks to the first target, the second target, the central position, and all other looks to any valid positions on the grid. Looks to the first target indicate the ability to locate a single target, and on this measure, the proportion of looks made by WS infants was lower than the other groups, including the Down's syndrome group. WS infants also differed from all the other groups on looks to the centre location. This was the vector summation measure, indicating reliance on sub-cortical mechanisms for saccade planning. Although WS infants scored a higher proportion of looks in this category than other groups, it should be noted that this was not the predominant response type for this group. In fact the greatest proportion of looks made by the WS group was to the Second target, although they did not differ from controls on this measure. Finally, WS infants also made a higher proportion of looks than all other groups to other areas on the grid.

This pattern of results suggests that the WS infants were impaired relative to controls on target location, and rely more than controls on sub-cortical orienting, which results in making responses to the vector averaged location. However, these results do not indicate whether there is any causal relationship, or what the direction of such a relationship might be. Thus the WS infants may make fewer saccades to the position of the first target because of reliance on the sub-cortical mechanism, which directs them to the centre location. Alternatively, they may have to resort to using a sub-cortical mechanism because cortical control of saccade planning is absent or impaired. The latter would appear to be consistent with the impaired dorsal stream hypothesis, although a third interpretation, that these results represent a more general impairment of saccade planning, should also be considered.

3.4.4 Second Saccade Responses

Second saccade responses also showed differences between the WS infants and the other groups. The proportion of egocentric looks made by the WS group was significantly lower than all of the other groups, indicating they are unable to combine extra-retinal information with retinal information, to the same extent as the other groups. The WS group also differed from all the other groups on the proportion of trials in which there was no second look, WS infants contributing more trials on which no second saccade was made. There was no difference between groups on proportion of retinocentric responses, looks back to the fixation point, or looks to other positions.

In terms of the overall pattern of second saccades made by groups, the predominate response type for the WS group was failure to make a second saccade, while for all other groups the predominant response was looks to the egocentric position. As with the elimination data, failure to make a second saccade could also be due to obligatory attention, or to more general differences in attention levels.

3.4.5 Ratio of Egocentric Responses

The ratio of egocentric responses to other measures is a more accurate representation of responses over the task, and is therefore a more stringent test for egocentric frames of reference than the simple measure of proportion of egocentric responses. Analysis of both ratios of egocentric responses, the ratio of egocentric to egocentric plus retinocentric, and the ratio of egocentric to all second looks, revealed that the WS infants did not differ from control groups on these measures. However, this analysis did reveal large variations in the WS group, which prompted further exploration of the data from this group, and suggested the possibility that there may be subgroups within

the WS population. The data would seem to indicate that there may be some infants with WS who exhibit impaired use of egocentric frames of reference, while some infants appear to be able to perform the task, although their performance still differs from controls. This pattern of results might be interpreted as a specific deficit in using egocentric frames of reference in the low ratio WS group, while the high ratio WS group have a more general target locating deficit. This suggestion concerning sub-groups in Williams syndrome will be further explored in Chapter 7, and it will also be of interest to determine whether differences occur within the WS population on other tasks.

One of the problems in interpreting the pattern of behaviours exhibited by the WS group concerns the complexity of the task. In order to succeed on the task, a child has to attend to the stimulus array, correctly locate the position of the first target, update spatial information due to the eye movement made, and finally integrate this information with information retained about the position of the second target. The performance of the infants with Williams syndrome might be interpreted as demonstrating impairments on many, if not on all these components. An alternative, more general interpretation of task failure, as exhibited by the WS infants, could be related to information processing. Bremner (1993) suggests that spatial tasks should be analysed in terms of their information processing requirements, and it could be argued that the pattern of results for the WS infants demonstrates impaired information processing abilities. However, both the mentally aged matched control group, i.e. younger typically developing infants, and the infants with Down's syndrome, were able to perform the task, therefore an information processing impairment in Williams syndrome would seem to be unlikely. It would be of interest to conduct further studies in order to obtain separate measures of each of the task

related components, as well as a reliable measure of information processing, in order to better determine the problem areas.

Further research into the relationship between the development of spatial representations and other cognitive abilities may also be beneficial in the examination of visual cognition in Williams syndrome. Acredolo (1990) proposes that there may be a link between the development of symbolic function and the child's ability to use landmarks in specifying location. It may be of interest to examine symbolic function in Williams syndrome, possibly through language measures, and compare this with the role of landmarks in spatial representation in this group. Acredolo also suggests a link between measures of temperament, such as activity level and attentiveness, to the development of spatial skills. Relationships between such measures will be examined and discussed in chapter 7.

While the underlying cause of the impaired performance demonstrated by the infants with Williams syndrome is as yet unclear, there is little doubt that these infants are experiencing significant problems processing this type of information. This disadvantage may result in problems processing a wide range of stimuli, such as environmental and educational stimuli, which in turn may impact on development in other areas of cognition.

The results of the study reported in this chapter raise the issue of the role of attention, and whether deficits in attention in the WS group may have contributed to this pattern of results. In order to examine this possibility further, the next chapter will focus on aspects of attention in typical and atypical development.

4

SUSTAINED ATTENTION

4.1 Introduction

Attentional problems are frequently associated with atypically developing infants and children, and several studies have focused on attention in specific clinical groups, such as Down's syndrome. Little work has been done on attention in Williams Syndrome, although anecdotal claims of attention problems and hyperactivity are often reported. The purpose of this chapter is first to review the literature on attention, and three proposed phases of attention, and more specifically the development of these phases, in both normally and atypically developing infants. Data will then be presented from a structured observational study of sustained attention in infants with Williams and Down's syndromes, and chronological and mental age-matched controls.

Decades of research into attention as a cognitive process have led away from the assumption that attention is a unitary concept, to the finding that attention is multifaceted. One basic definition of attention in adults may be thought of as the processes whereby certain aspects within the environment are selected for further, or enhanced cognitive processing. This selection may be driven by bottom-up, or top-down processes, or a combination of both. Attention may also be overt, in that the behaviours indicating attention are visible, such as eye or head movements, or it may be covert, internal in nature (Posner, 1980). Of greater interest here is the nature of

attention in infants, how it develops, and the impact of variations in attention on development.

It is generally considered that attention plays a major role in the development of visual cognition in infancy, as has been demonstrated in the cases of hyperactive children (Palisin 1986), and premature and full-term infants (Ruff, Lawson, Parrinello, and Weissberg, 1990). The results of such longitudinal studies have led to the claim that the effect of low attention in infancy is both detrimental and cumulative (Lawson and Ruff, unpublished manuscript). Thus this section will review the proposed link between attention and the development of visual cognition, in both normally developing, and atypical populations.

4.1.1 Phases of Attention.

Within the area of attention in infancy, several investigators have differentiated between the phases involved. Ruff and Lawson (1991) distinguish between reactive attention, which involves general arousal and orienting toward novel stimuli, and sustained attention, which is the period during which the infant is effortfully engaged with the stimuli. These phases can also be defined in terms of overt or covert attention, in that overt attention generally refers to orienting, while sustained attention, although normally measured by behaviour, is thought to be a covert process. Ruff and Lawson state that normal functioning also requires the ability to shift attention; without this, the infant will be unable to move on to other relevant aspects within the environment. Similarly, Lansink and Richards (1997), describe three phases of attention; stimulus orienting, sustained attention, and attention termination. The focus of this section will be on sustained attention, as it has been proposed that this phase reflects enhanced information processing (Casey and Richards, 1988), but stimulus orienting and attention termination will also be considered, as these phases delineate the onset and offset of periods of sustained attention.

4.1.1.1 Stimulus Orienting

Stimulus orienting refers to the process whereby an individual will first detect and then locate a stimulus, before actively engaging in effortful processing. The development of orienting is thought to progress from subcortical control in new-borns, to more cortical mechanisms by the age of two to three months. New-born infants orient more readily towards stimuli in the temporal visual field, which is thought to input to the subcortical visual pathway from the eye to the superior colliculus (Johnson, 1990). By the age of two months, infants are more able to orient towards stimuli in the nasal visual field, indicative of cortical control of orienting. Speed of orienting also changes over the first few months; one month olds will take longer to orient to peripheral stimuli than three month olds, and will also demonstrate more directional errors (Atkinson, Hood, Wattam-Bell, and Braddick, 1992).

4.1.1.2 Sustained Attention

Sustained attention, sometimes referred to as focused attention, equates to the period after orienting, when relatively intense processing or concentration is involved. However, it should be noted that sustained attention does not necessarily follow from orienting, as infants may be looking at a stimulus without necessarily engaging in the enhanced processing which defines sustained attention. This process of looking at stimuli without deep processing is sometimes referred to as casual attention. The traditional approach toward the measurement of sustained attention has been the observation of children's interactions with toys during free play, or within the more structured confines of laboratory based observations. In both types of situation, such observations are typically videotaped, and subsequently coded by trained coders. The actual behaviours which are coded differ between studies. For example, coding has been done on the basis of a global measure of attention (Ruff and Lawson, 1990), a combination of physical alterations such as facial expression and motor movement (Ruff and Lawson, 1990), or the duration of time spent on/off task (Krakow and Kopp, 1982).

The reliability of these measures of sustained attention is claimed to be good; Ruff and Lawson (1991) give inter-observer correlations of >.90 for infants of 6 months and older, and .87 for 5 month-old infants. Ruff and Lawson also claim good validity for this type of measure of attention. Based on the hypothesis that infants will be less distractible during periods of sustained attention, than during periods of casual attention, these investigators have shown that when presented with a novel object, sustained attention has priority over other behaviours in both 7 and 12 month-old infants. Sustained attention has been shown to decline with exposure, and increase on presentation of another, novel object. Ruff and Lawson claim that such findings support the validity of the measure as one of 'cognitively effortful engagement'. Support for this also comes from evidence that the amount of information to be processed, varied by presenting complex or simple objects, is related to the amount of examining carried out, as found in a study with 7 and 10 month-old infants (Oakes and Tellinghausen, 1994).

Several studies have applied physiological measures in the investigation of sustained attention. Lansink and Richards (1997) have looked at heart rate during attention, and have used this measure to support their tripartite model of attention. During stimulus orienting, there is an initial deceleration in heart rate, which is maintained during sustained attention, before heart rate returns to pre-stimulus level, signalling the termination of the attention phase. This pattern of heart rate has been explained in terms of the role played by the vagus nerve which, it is proposed, results in increased blood outflow during periods of sustained attention, which is then inhibited during attention termination (Richards and Casey, 1991).

Lansink and Richards (1997) have also examined the relationship between heart rate and behavioural measures of sustained attention, and concluded that together these measures offer a greater degree of accuracy than either on its own. Although the majority of the results were the same when either measure was considered, some

differences occurred. For example, longer latencies to looking at a distracter were found when heart rate was used as a measure, for 6 and 10-month-olds, while this was only found in the 9-month-olds when behavioural measures were used.

The general trend demonstrated in the development of sustained attention is one of greater sustained attention with increasing age. Ruff and Lawson (1991) claim that this is due initially to improved motor control from 5 to 7 months, and subsequently through increased sophistication about what is to be learned, from 7 to 11 months. They do not, however, elaborate on the form such increased sophistication might take. There is also a general tendency for latency to enter sustained attention to decrease with age, as the maturing nervous system facilitates quicker and more efficient organisation of processing.

It may also be the case that as infants develop, in addition to increased sustained attention, they also demonstrate more periods 'off task', particularly if attention is examined during problem solving. This increase in the number of periods off task may represent the need to disengage from the stimuli in order to process, or consolidate what has been learned, or may equate to more rapid habituation with age.

Richards (1985) has charted the development of sustained attention, as measured by heart rate, from 2 to 6 months, and has noted that while heart rate changes occur in periods of sustained attention over this time, other phases of attention are not marked by such changes. This change in sustained attention may be linked to the development of the arousal system, and its increasing influence on the developing cortex (Richards, 1995). Richards compares the development of these systems and the development of the neural system underlying attention, eye movement and fixation, and notes that the changes in each follow a parallel development. However, it is not clear whether there is a mutual influence between systems, or whether one is dependent on the other.

The significance of sustained attention in development is clear, if periods of sustained attention are indeed phases which reflect learning. If these events are few, or short, the infant will learn less about the world than an infant whose sustained attention is considered to be 'appropriate'. This can be seen in the proposed link between sustained attention and impulse control. Ruff, Lawson, Parrinello, and Weissberg (1990) found associations between two measures of impulse control (time delay in a delayed response task, and errors of commission in a reaction time task), and measures of sustained attention in a longitudinal study of normal and premature infants. Although relationships between these measures were not strong, Ruff et al. believe that such measures can be useful in defining children who have deficits of sustained attention, and in differentiating between these and children who have deficits of reactive attention, or orienting. Using data from the same study, Ruff et al. claim that measures of attention and inattention are remarkably stable over the first few years. It is also interesting to note that Ruff found greater stability and predictive validity for a number of their measures of attention and inattention, than for either of the standardised tests of development used in the study.

4.1.1.3 Attention Termination

Attention termination refers to the process whereby án individual's attention is disengaged from a stimulus, normally prior to orienting to another target. This is a necessary component within development, as having completed processing the current object of attention, it allows infants to move on to, and learn about, other environmental stimuli (Ruff and Lawson, 1991). From studies with lesion patients, Posner, Rafal, Choate, and Vaughan (1985) claim that mechanisms controlling attention termination, or disengagement, are located in the parietal lobe, while Guitton, Buchtel, and Douglas (1985) found that patients with damage in frontal eye field areas are unable to inhibit saccades to salient peripheral visual targets.

The development of attention termination in infancy encompasses the phenomenon of obligatory attention, first described by Stechlar and Latz (1966), which is characterised by prolonged fixation to a stimulus, often concluding only when the infant becomes distressed. This pattern of behaviour is not believed to be present at birth, but may be found most strongly in one to three month-olds. Johnson (1990) suggests that obligatory attention may be due to increased inhibition of the colliculus, resulting in stimuli in the peripheral visual field failing to trigger orienting.

Problems in attention termination have also been shown in older children, when presented with distracting stimuli or invalid cues (Enns and Brodeur, 1989). Deficits of attention disengagement have also been demonstrated in several clinical groups, including children with ADHD (Wood, Maruff, Levy, Farrow, and Hay; 1999), children with developmental coordination disorder (Wilson, Maruff and McKenzie; 1997), and Alzheimers patients (Filoteo, Delis, Roman, Demadura, et al.; 1996). Abnormally rapid disengagement of attention has been found in Parkinson's disease patients (Fileto, Delis, Demadura, Salmon, et al.; 1994). Several studies have also claimed to find impairments of attention in autism (see Courchesne, 1987, for a review). Courchesne, Townsend, Akshoomof, et al., (1994) claim that people with autism demonstrate impaired attention shifting, although not impaired disengagement, and link this to Purkinge neuron loss in the cerebellum.

4.1.2 Sustained Attention in Williams and Down's syndromes

If sustained attention plays a major role in the development of normal infants, then the contribution it makes within atypically developing populations is worthy of investigation. Demonstrations of impaired sustained attention in these groups may be linked to their individual developmental outcomes, such that early identification of this problem may be beneficial in the implementation of intervention schedules. But perhaps of greater interest are the causes underlying deficits of sustained attention, and their interaction with the development of other cognitive abilities and

impairments associated with individual syndromes. Thus charting the development of sustained attention over time, in infants with Down's syndrome, Williams syndrome, and normally developing infants, is of particular interest.

There has been a general trend within the research literature towards the classification of the Down's syndrome population as a unitary, stereotypical group. Thus Down's syndrome children are typically described as exhibiting poorer general attention than normal children, while they do not differ from other mentally delayed groups matched on IQ (e.g. Fisher, 1970). Similar results have also been demonstrated in adults with Down's syndrome. Tomporowski (1990) claimed to find impaired sustained attention in a group of 16 mentally retarded adults, 3 of whom were diagnosed as having Down's syndrome, when tested on vigilance tasks. However, there are also studies which indicate a degree of variability of attention within the DS population. Green, Dennis and Bennets (1989) claim that within the small sample of 2-4 year-olds with Down's syndrome they tested, there was a subgroup (4 out of 13 subjects) who exhibited a pattern of attention which they compared to attention deficit disorder. The subjects in this group demonstrated both qualitative impairments, in terms of reduced total attention time, and quantitative impairments, in that these subjects had a higher proportion of shorter attention spans, with more intervals between periods of attention. Green et. al. argue that this represents a discrete grouping, and not merely the extreme end of a continuous variable. They propose that this distinction may be found in the Down's syndrome population as a whole.

Using a different coding system, Krakow and Kopp (1982) found qualitative rather than quantitative differences between children with Down's syndrome and developmental age matched controls. Both 29 month-olds and 3-4 year-olds were similar to the control groups in terms of duration of object orientation, but spent less time than the control groups in social orientation, e.g. giving a toy to the mother, and more time than controls was spent completely unoccupied. In addition, almost two

thirds of the older group of Down's syndrome engaged in repetitive or stereotypic play, which was not demonstrated by any of the control subjects. This perseveration of behaviour can be related to the issue of impulse control. Krakow and Kopp suggest that this pattern of behaviour reflects an inability to spontaneously shift activities, which in turn reduces learning about the environment. Failure to shift activities may also be related to a deficit of attention disengagement which, as stated previously, has been found in a number of clinical groups.

The development of attention in Down's syndrome can be examined in a study by Vietze, McCarthy, McQuiston, MacTurk, and Yarrow (1983), who presented 6, 8, and 12 month old infants with Down's syndrome with objects in three conditions. As with normal controls, manipulation of objects increased with age, and visual exploration as a behaviour on its own decreased. However, DS infants in all age groups looked more, and explored less, than normally developing 6 month old infants.

A small number of studies have been carried out on attention in Williams syndrome, although the majority of these focus on the prevalence of Attention Deficit Hyperactivity Disorder (ADHD) in children and adolescents with WS. The Williams Syndrome Association list distractibility as a common problem, and state that ADHD is often diagnosed in school age children with Williams syndrome. In a comprehensive study of the medical and psychological characteristics of a group of people with Williams syndrome, Morris, Demsey, Leonard, Dilts and Blackburn (1988) report that ADHD was characteristic of 84% of 4-16 year old participants. Finegan, Sitarenios, Smith and Meschino (1994) claim that children with WS are four times more likely to have ADHD than children in the general population. Udwin, Yule and Martin (1987) reported that seventy-two percent of a sample of 6-15 year old children with WS, displayed hyperactivity in school, at home, or both, as rated by parents and teachers on the Rutter Questionnaires (Rutter, 1967). This is higher than rates for age matched normal or mentally handicapped children (Rutter et al., 1970).

Treatment of children with Williams syndrome with methylphenidate, a stimulant medication used in the treatment of ADHD, has resulted in some improvement in attention, and less impulsivity, irritability and frustration, in two case studies (Power, Blum, Jones, and Kaplan, 1997; Bawden, MacDonald and Shea; 1997).

Although there is a paucity of more empirical research on attention and Williams syndrome, the prevalence of ADHD in children with Williams syndrome indicates that children and adolescents do tend to present with problems of attention. The reported incidence of ADHD in people with Down's syndrome vary, but tend to be much lower than those reported for Williams syndrome (Green, Dennis and Bennets, 1989; Cocchi and Favuto, 1997). If the incidence of ADHD in Williams syndrome is as high as has been reported, then it might be logical to expect that deficits of attention disengagement, which are claimed to be present in ADHD (Wood, Maruff, Levy, Farrow, and Hay; 1999), would also be present in people with Williams syndrome.

The comparison of sustained attention abilities in infants with Williams syndrome and Down's syndrome may reveal something concerning the development of this process. Two issues are of interest here; the first concerns whether the patterns of attention claimed to be present in adults and older children with Williams and Down's syndromes are present from infancy, or if these problems develop later. Secondly, if infants with Williams syndrome do show deficits in this area, are they qualitatively similar to those exhibited by infants with Down's syndrome? Thus an empirical study will be reported in which sustained attention is compared across WS, DS, and control groups. The method used is the structured observation and coding system developed by Ruff (e.g. Ruff, 1990; Ruff and Lawson, 1990, 1991). This was chosen in preference to heart rate measures of attention (e.g. as used by Richards), due to irregular heart function in many children with WS and DS. It is hypothesised that both WS and DS infants will be impaired relative to control groups, both on measures of

duration and number of periods of sustained attention, which would indicate that the attention problems found in adulthood are present from an early age.

4.2 Method

4.2.1 Design

4 groups were tested in a between-subjects design; Williams Syndrome (WS), Down's Syndrome (DS), chronological age-matched controls (CA), and mental age-matched controls (MA).

4.2.2 Participants

14 WS infants, 19 DS infant, 17 CA infants and 16 MA infants were tested from the main sample described in chapter 2. Mean ages and age ranges for each group are presented in Table 4.1.

Table 4.1 Mean and range of ages for each group.

	Mean	Minimum	Maximum
WS (n=14)	29 months 29 days	23 months 16 days	36 months 15 days
DS (n=19)	29 months 3 days	23 months 25 days	36 months 25 days
CA (n=17)	29 months 28 days	23 months 13 days	36 months 28 days
MA (n=16)	15 months 6 days	11 months 24 days	20 months 15 days

4.2.3 Procedure

Infants were placed on a child's booster seat which was attached to a normal chair, and seated on the right of the long side of a table. The parent or carer sat on the child's left, and the experimenter sat at an angle, at the corner opposite the child. A box containing the toys was to the left of the experimenter, out of the child's view. A video camera was placed directly facing the child at head level, and all sessions were recorded for subsequent coding. A representation of the apparatus and set-up can be seen in Figure 4.1.

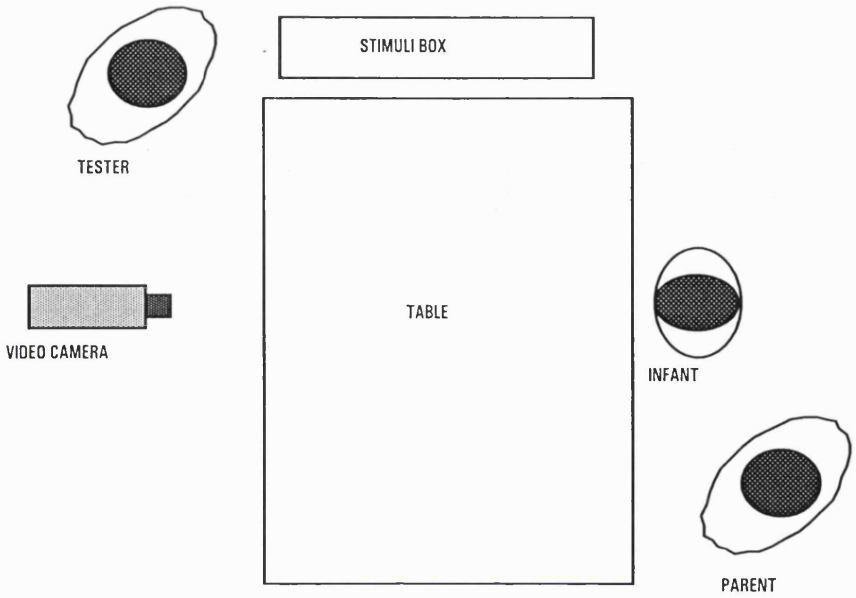

Figure 4.1 Position of apparatus, participant, and experimenter, in the sustained attention study.

The test session consisted of three parts; the warm-up, the experimental trials, and a debriefing for the parents. The warm-up consisted of two trials, in which toys were placed on the table in front of the child for 45 seconds each. Order of presentation of the toys was varied across participants. Parents were advised before the session that they should not talk to the child, and also that the experimenter would not talk to, or make eye contact with the child. At the end of 45 seconds, the toy was removed, and the next toy introduced. In cases where the child would not give up the toy, the next toy was introduced as a distracter, enabling the experimenter to remove the first toy. The purpose of the warm-up session was to allow the child to become familiar with the setting, and to minimise any disturbance the child might have experienced due to the fact that the adults were not speaking. The experimental trials were exactly the same as in the warm-up, but different toys were used. During the debriefing, parents were asked whether the child was familiar with any of the experimental toys, and their responses recorded.

Six toys were used during the experimental trials, but three were later eliminated from further coding; one was a two-part toy and was therefore judged to be more difficult to code than the other toys, as attention could be focused on either of two component parts. Another toy was not age appropriate for many of the older infants, and a third was judged to be gender specific. The three toys which were coded were:

1. A suction toy, which was 9 inches high, with a suction cap at the bottom. The upper part of the toy was a corkscrew shape, with an outer part which could be screwed round the corkscrew. Attached to this outer part were three revolving balls.
2. A farmyard pop-up, 12 inches wide, which consisted of four different types of coloured buttons, which would reveal farm animals when pressed.
3. A wooden rod, ball and elastic toy, 6 inches in diameter. Small wooden balls could slide along the rods and elastic, and the whole object could be compressed and expanded.

4.2.4 Coding

Time codes were added to the videotapes after experimental sessions, to allow frame-by-frame coding (50 half frames per second). Subsequent coding from videotape was carried out by two trained coders. One coder, who was blind as to the experimental hypothesis, coded all sessions. A second coder coded 25% of sessions, selected at random, in order to obtain inter-rater reliability scores. The first coder also coded 10% of sessions a second time, in order to obtain intra-rater reliability scores.

Coding was based on that of Ruff and Lawson (1991). Dependent variables obtained were duration of attention, number of periods of attention, and a global rating of attention. Each coding session would begin with the coder watching a complete session (one period of 45 seconds with one toy) in real time, to get an overall view of events during that session. The coder would then rate the session for the global

features of attention, on a three point scale; 3 was indicative of poor attention, while 1 indicated good sustained attention. The dimensions rated were:

A. Steadiness of Gaze

B. Facial Expression

C. Position of Toys

D. Self-Consciousness

E. Amount of Extraneous Movement

F. Speed of Movement

G. Talking/Vocalizing

The coder would then code each session for specific periods of sustained attention, noting start and end times in half frames. The two coders trained on coding periods of sustained attention using videotapes and training material provided by Holly Ruff (see Appendix), until reliability within and between coders was at least 90%. After coding, any periods of sustained attention which were less than 2 seconds in duration were removed before analysis, as Ruff (1991) claims that these are so short that they are not really indicative of sustained attention.

4.3 Results

Having ascertained that there were no differences between groups in familiarity with the toys presented, responses from all three toys were pooled, and combined results will be reported throughout.

4.3.1 Reliability

To establish reliability both within and between coders, Pearson product-moment correlations were calculated for the duration of sustained attention, as coded by Coder 1 on two separate occasions for the intra-rater reliability, and as coded independently by Coder 1 and Coder 2 for the inter-rater reliability. Coder 1 intra-rater reliability =

.98 (n = 9), intra-rater reliability by group: WS = .99 (n = 2), DS = .99 (n = 2), CA = .96 (n = 2), MA = .98 (n = 3), inter-rater reliability (Coder 1 and Coder 2) = .99 (n = 8). These figures are comparable with the inter-observer correlations of >.90 reported by Ruff and Lawson (1991).

4.3.2 Duration of Sustained Attention

Total duration of periods of sustained attention with all 3 toys was calculated, and average total duration for all 4 groups is presented in Table 2. The WS group have the longest total duration of sustained attention, and the DS group the shortest.

Table 4.2 Average duration of sustained attention for all 3 toys, in seconds.

	Mean	SD
WS (n=14)	63.75	33.72
DS (n=19)	33.18	23.15
CA (n=17)	54.86	19.16
MA (n=16)	54.65	28.08

A one-way Anova was performed, which revealed a significant difference between groups ($F(3,62) = 4.29$; $p < 0.05$). A significant difference was found between the WS and the DS groups (Tukey's HSD, $p < 0.05$), the WS group having significantly greater total duration of periods of sustained attention than the DS.

4.3.3 Number of Periods of Sustained Attention

The number of periods of attention demonstrated by each infant was calculated for all toys. Average number of periods of sustained attention for each group is presented in Table 4.3.

Table 4.3 Average number of periods of sustained attention

	Mean	SD
WS (n = 14)	4.50	1.70
DS (n = 19)	3.32	1.29
CA (n = 17)	4.59	1.70
MA (n = 16)	5.06	2.41

A one-way Anova was performed on the data, which revealed a significant difference between groups ($F(3,62) = 3.05$. $p < 0.05$). This was found to be between the lowest number of periods of sustained attention for the DS group, and the highest for the MA group (Tukey's HSD, $p < 0.05$). There was no significant difference between the DS and WS groups.

Table 4.4 Means and Standard Deviations of Ratings for each Global Dimension for all Groups. Standard Deviations in parenthesis.

	WS (n = 14)	DS (n = 19)	CA (n = 17)	MA (n = 16)
Gaze Steadiness	2.41 (0.53)	2.67 (0.46)	2.52 (0.39)	2.58 (0.41)
Facial Expression	2.41 (0.37)	2.64 (0.34)	2.50 (0.46)	2.38 (0.49)
Position of Toys	2.17 (0.52)	2.40 (0.36)	2.06 (0.32)	2.25 (0.46)
Self-Consciousness	2.36 (0.51)	2.68 (0.41)	2.56 (0.47)	2.52 (0.42)
Movement Amount	2.02 (0.58)	2.61 (0.39)	2.27 (0.50)	2.25 (0.59)
Speed of Movement	2.17 (0.34)	2.33 (0.48)	1.99 (0.33)	1.73 (0.39)
Talking/Vocalizing	2.14 (0.58)	1.81 (0.30)	2.42 (0.56)	1.81 (0.42)

4.3.4 Global Ratings

Mean ratings for each of the seven global measures of sustained attention were calculated, and are presented in Table 4.4. Possible scores ranged from 1 to 3, with a lower score indicative of sustained attention. Analysis with a repeated measures Anova revealed that the main effect of group was not significant ($F(3,62) = 0.156$; n.s.). The main effect of global measure was significant ($F(6,372) = 28.56$; $p < 0.001$), and the interaction of group by global measure was also significant ($F(18,372) = 5.37$; $p < 0.001$). Analysis of the interaction revealed that significant differences were only

found on three of the measures. On Amount of Extraneous Movement, the DS scored higher than the WS ($F(3,65) = 3.804$; $p < 0.05$; Tukey's HSD, $p < 0.05$), indicating better sustained attention in the WS group. On Speed of Movement, both the WS and the DS groups scored higher than the MA group ($F(3,65) = 7.283$; $p < 0.01$; Tukey's HSD, $p < 0.05$), which implies that the MA group were showing better sustained attention on this measure. On Talking and Vocalizing, the CA group scored higher than the DS and MA groups ($F(3,65) = 6.774$; $p < 0.01$; Tukey's HSD, $p < 0.05$). This could be indicative of lower attention in the CA group, as according to the scoring system, sustained attention is not normally accompanied by other activities such as speech. However, this result might also be an effect of different language abilities between groups, in that the CA group were more advanced on language measures (Paterson, 2000), and were therefore more able to vocalize.

4.3.5 No periods of Sustained Attention

In order to further explore the impaired DS performance on the measures of number of periods of sustained attention, the data were examined for failure to exhibit periods of sustained attention on any of the three trials. Thus each infant was coded as demonstrating at least one period of sustained attention with each toy, or failing to demonstrate sustained attention with at least one toy. Frequencies of group membership are presented in Table 4.5.

Table 4.5 Contingencies of infants showing Sustained Attention with all 3 toys or not. Percentages of group size by category are shown in parenthesis.

	Minimum of one failure to exhibit SA	SA with all 3 toys
WS (n=14)	4 (13.79)	10 (27.03)
DS (n=19)	15 (51.72)	4 (10.81)
CA (n=17)	4 (13.79)	13 (35.14)
MA (n=16)	6 (20.69)	10 (27.03)

For infants categorised as failing to demonstrate periods of SA, the DS group represents over 50%, while the proportion of DS infants who showed at least one period of SA with each of the three toys was smaller than in other groups (Chi-Square = 13.94; df = 3; $p < 0.05$; Cramer's V = 0.46; $p < 0.05$)

4.3.6 Categorisation of SA periods as Short, Medium, or Long.

Periods of sustained attention could vary in length from 2 to 45 seconds, therefore a more detailed analysis of the lengths of SA periods was desirable. This analysis was performed in order to determine whether groups were demonstrating tendencies to sustain attention for short or long periods. This would allow further exploration of the difference between the WS and DS groups on the measure of average duration of sustained attention. The 25th and 75th percentiles were determined from the raw data from all the groups, and these were used to categorise periods of SA as short, medium, or long. Mean durations for each group in each of these categories is reported in Table 4.6.

Table 4.6 Mean duration of periods of SA (in seconds) categorised as short, medium, or long.

	SHORT		MEDIUM		LONG	
	Mean	SD	Mean	SD	Mean	SD
WS	2.54	2.15	17.93	12.98	43.37	30.72
DS	3.90	4.20	13.41	11.46	15.88	25.88
CA	3.39	3.37	23.03	15.99	28.44	24.57
MA	3.79	3.45	23.23	14.09	27.64	26.71

The most noticeable difference appears to be in the long periods of SA, where the mean duration for WS infants is greater than that of all other groups, and particularly that of the DS infants. When analysed with a repeated measures Anova, the data showed a significant effect of group ($F(3,62) = 4,3$; $p < .05$), and a significant effect of time ($F(2,124) = 31.09$; $p < 0.01$). The interaction of group by time was not significant ($F(6,124) = 2.1$; n.s). The main effect of time was to be expected, and was

due to the differences between categories. Of more interest is the main effect of group, which was found to be due to differences between WS and DS infants (Tukey's HSD, $p < 0.05$).

4.4 Discussion

The aim of this chapter was to examine sustained attention in Williams and Down's syndromes, compared to chronological and mental age-matched controls. It was predicted that both WS and DS infants would be impaired relative to controls on measures of sustained attention such as total duration of periods of sustained attention, and number of periods of sustained attention. This would indicate that detriments of attention reported in adults with Williams and Down's syndromes are present from an early stage. However, although differences were found for the experimental groups, they are not in line with predictions, and therefore the results do not fully support the hypothesis.

On the proposed measures of duration and number of periods of sustained attention, infants with Down's syndrome were shown to perform at a lower level than all other groups. Specifically, DS infants had significantly shorter average total duration of sustained attention than infants with WS, although CA and MA infants also scored higher than DS infants on this measure. The results from analysis of the number of periods of sustained attention indicate that DS infants have fewer periods of sustained attention; although this difference was only significant when compared with the MA infants, there is also a trend towards more periods of sustained attention in the WS and the CA groups.

The global ratings of specific dimensions of sustained attention are less detailed, in that they are judged over the whole session with a toy, and will therefore reflect changes in levels of attention, as infants move between periods of sustained attention. However, it is interesting to note that the measures where differences occur are either

ratings of movement (amount of extraneous movement, or speed of movement), or talking/vocalizing. The DS group score was the poorest for both extraneous movement and speed of movement, while the WS group was poor on speed of movement only. Given the reported prevalence of ADHD in older children and adolescents with Williams syndrome, they might have been expected to perform worse on both these measures. The poor score by the CA group on the Talking/Vocalizing measure may simply reflect better language skills in this group. As this group scored higher on tests of vocabulary and other language measures (Paterson, 2000), they were more able to vocalize than other groups, and this may have been reflected in the results reported here.

Further examination of the data was attempted in order to try to unravel the differences which occurred. Categorisation of periods of attention into short, medium and long revealed that there was a difference between the WS and DS groups. However, differences were not revealed between these groups in relation to any specific category.

Finally, categorisation of each infant as failing to exhibit sustained attention to at least one of the toys, or successfully exhibiting at least one period of attention for each toy, revealed that DS infants were more likely to fail to display any periods of sustained attention within a session.

Taken as a whole, the results indicate that infants with Down's syndrome perform worse than the other groups, indicating that they are impaired on measures of sustained attention. These results would seem to be in line with many of the studies which claim to find attention deficits in children and adults with Down's syndrome (e.g. Fisher, 1970; Tomporowski, 1990), and are indicative of a stable deficit from infancy through to adulthood within this syndrome.

The surprising result is that infants with Williams syndrome perform as well as typically developing controls, with the exception of faster movements on the global rating measures. As rates of ADHD are reportedly high in WS, it was predicted that infants with WS would also perform poorly on measures of attention. Failure to find this deficit may have several interpretations. The fact that infants with WS do not show the same deficits of attention as found in older children and adults with Williams syndrome suggests that this deficit does not develop until later. This might emerge as children get older, due to interactions between other cognitive deficits, and the way in which such deficits might impact on learning.

It may, however, be the case that the WS group are exhibiting attention deficits which are not being tapped by this task. Infants with WS exhibited greater average duration of periods of attention compared with the DS group. Although these infants did not differ from the CA or the MA groups, it may be that this result indicates a tendency to remain fixated on a stimulus, or a possible attention disengagement deficit. This interpretation might also offer an explanation for the finding of high rates of ADHD in Williams syndrome, as children with ADHD also have attention disengagement deficits.

The results from the typically developing control groups are somewhat problematic, in that as there are no differences between the older and younger typically developing control groups, they do not demonstrate the suggested developmental progression of greater sustained attention with age (Ruff and Lawson, 1991). However, that finding is based on differences between much younger children (5-7 months and 7-11 months). By contrast, the differences between the age groups examined here may be more discrete, and therefore not revealed by this task. In addition, the age range involved (11 months to 30 months) was much broader than that in the Ruff and Lawson study. Although obvious age inappropriate items were excluded, it is

nonetheless difficult to find stimuli which will evoke the same level of interest across these ages.

Another factor which may have contributed to failure to find differences between age groups of the typically developing infants was the decision to maintain silence and avoid eye contact with participants throughout the task. Although the warm up session was designed to allow children to become used to this artificial situation, many of them remained somewhat perturbed by this, and asked questions such as 'what's the lady doing mummy?', and 'why are you quiet, mummy?'. This situation could be seen to interrupt natural attention levels for this group. This may also account for increased talking and vocalizing by this group on the global rating measure.

Given the prevalence of ADHD in Williams syndrome, it would be of interest to explore this more specifically in infants with Williams syndrome. The possible deficit of attention disengagement suggested in this study may be one way to approach this. This would also further test the hypothesis that infants with Williams syndrome may not exhibit the problems of attention seen in later childhood and adulthood. Ruff, Lawson, Parrinello, and Weissberg (1990) suggest that measures of inattention may be more revealing than measures of attention, an area which would be of interest for future study.

In summary, the results from this study indicate that children with Williams syndrome are not impaired on measures of sustained attention, while children with Down's syndrome do exhibit a sustained attention deficit. Infants with WS may have a deficit of attention disengagement mechanisms, but further work is required to investigate this more fully. The results from this study will also be examined in light of the results from the next chapter, which looks at infant temperament, as some dimensions of temperament are proposed to correlate with attention.

5

THE ROLE OF TEMPERAMENT IN ATYPICAL DEVELOPMENT

5.1 Introduction

The purpose of this section is to examine aspects of temperament within infants with Williams and Down's syndrome. The main area of interest is those aspects of temperament which relate to attention, although other features will also be examined.

Temperament has attracted a great deal of research over the last 20 years, driven by a number of theoretical standpoints which differ in their approach and definitions of temperament. However, there are also areas of consensus between these approaches to the study of temperament. A brief review of questionnaire studies of temperament will be presented, followed by a more detailed examination of the measure of temperament used in the current study, the Infant Behavior Questionnaire (Rothbart, 1981). The literature on temperament in Down's and Williams syndromes will also be reviewed, before presentation of the results from a study using the Infant Behavior Questionnaire with the Williams syndrome, Down's syndrome and control samples.

5.1.1 Temperament in Infancy

Several methodologies have been adopted in the study of temperament, including arousal measurement, home observations and laboratory observations, but the most predominant method employed has been parental rating questionnaires. Seminal work in questionnaire studies of temperament in infancy was the research of Thomas and Chess (1977), who conducted the New York Longitudinal Study (NYLS), a parent report assessment of 141 children during the first 2 years of life. Thomas and Chess identified nine dimensions of temperament: activity level, rhythmicity, approach, adaptability, intensity of reaction, threshold of responsiveness, quality of mood,

distractibility, and attention span-persistence. Although this was a pivotal study in temperament research, it has received some criticism. Rothbart (1981) claims that there are problems with lack of homogeneity within the individual dimensions, the sample on which the study was conducted was very restricted in terms of socio-economic status and race, and sampling procedures meant that several different ages were grouped together, which may have confounded individual differences. Thomas and Chess have also been criticised for their approach which tends to downplay the role of genetic or biological factors in temperament (Plomin and DeFries, 1985).

Since the NYLS, several other parent report questionnaires have been devised. The Infant Temperament Questionnaire (Carey, 1970) and the Revised Infant Temperament Questionnaire (Carey and McDevitt, 1978), primarily developed for clinical screening, were based on the Thomas and Chess study, and suffer the same criticisms in relation to nonindependence of scales, in that there was a large degree of overlap between the dimensions identified in these scales. Bates, Freeland, and Lounsbury (1979) developed the Infant Characteristics Questionnaire, to measure parents' perception of infant difficulty, but as such this does not span the total domain of temperament.

Parental reports have been examined for reliability and validity, and appear to stand up to scrutiny reasonably well. Field and Greenberg (1982) found significant relationships between the ratings of mothers and fathers, and between parents and teachers. Wilson and Matheny (1983) report convergent validity between laboratory ratings and parent report on a study of temperament in twins. Worobey (1997) has also shown convergent validity between two measures of infant temperament, the IBQ (Rothbart, 1981), and the Early Infancy Temperament Questionnaire (Medoff-Cooper, Carey and McDevitt, 1994). However, several researchers have found that parental reports reflect not only characteristics of the child but also the mother, and perhaps more importantly, the interaction between mother and child (Bates and

Bayles, 1984; Crockenberg and Acredolo, 1983). Rothbart (1984) cautions that parental reports are not an independent measure of the child's functioning, but are an assessment of behaviour within an interactive system which is contributed to by caregivers, siblings, and other sources of stimulation. Therefore it is not surprising that caregiver characteristics, and the way in which the child functions within the environment provided by the caregiver, should also be tapped by this method.

5.1.2 The Infant Behavior Questionnaire.

The Infant Behavior Questionnaire, or IBQ (Rothbart, 1981) was developed to counter the constraints found in previously developed measures of infant temperament. Thus it is claimed to eliminate the conceptual overlap of other scales, such as Thomas and Chess (1977). In addition, Rothbart (1981) defines temperament as individual differences in reactivity and self-regulation, therefore dimensions were also selected which reflected aspects of reactivity and self-regulation omitted from previous studies. Questionnaires were completed for 463 infants, at 3-, 6-, 9-, and 12-months. Conceptual and item analysis resulted in 6 scales with adequate psychometric and conceptual properties: activity level, soothability, fear/distress and latency to approach sudden or novel stimuli, distress to limitations, smiling and laughter, and duration of orienting. Descriptions of these dimensions are given below:

Activity Level: Child's gross motor activity, including movement of arms and legs, squirming and locomotor activity.

DTNS (Distress and Latency to Approach Sudden or Novel Stimuli): The child's distress to sudden changes in stimulation and the child's distress and latency of movement toward a novel social or physical object.

Duration of Orienting: The child's vocalization, looking at and/or interaction with a single object for extended periods of time when there has been no sudden change in stimulation.

DTL (Distress to Limitations): Child's fussing, crying or showing distress while a) waiting for food, b) refusing a food, c) being in a confined place or position, d) being dressed or undressed, e) being prevented access to an object toward which the child is directing her/his attention.

Smiling and Laughter: Smiling or laughter from the child in any situation.

Soothability: Child's reduction of fussing, crying, or distress when soothing techniques are used by the caretaker of child.

Inter-rater reliability was measured in a sub-sample of 22, in which a second adult (father or babysitter) completed the questionnaire in addition to the infants' mothers. Although the possibility that mothers and second raters would discuss items within the questionnaire could not be eliminated, mothers were asked not to do so. Significant correlations were found for the matched pairs of questionnaires on all dimensions. Stability over time was also assessed; a subsample of 36 mothers filled out questionnaires when their infants were 3-, 6-, 9-, and 12-months old, mothers of thirty-four 6 month olds completed the questionnaire again when infants were 9- and 12-months of age, and thirty-six 9 month olds were retested at 12 months. Overall, stability between ages was adequate, although 3-month scores were not predictive of later scores on distress to limitations, and distress and latency to approach sudden or novel stimuli. Finally, moderate convergent validity and relative temporal stability of the IBQ were found in a longitudinal study of both the IBQ and home observations of infants.

Although the IBQ was designed to eliminate conceptual overlap, positive correlations were found between distress to limitations and fear, distress to limitations and activity level, and smiling/laughter and soothability. Negative correlations were found between smiling/laughter and fear, and between smiling/laughter and distress to limitations. Rothbart (1981) suggests that these intercorrelations, which she interprets as a positive relationship between measures of distress, and a negative relationship

between smiling/laughter and the distress measures, may be indicative of general mood differences. More recently, Rothbart (1999) has proposed a tripartite model of temperament, on to which most of the original dimensions in the IBQ load. Thus activity level and smiling/laughter load on to extraversion/surgency, duration of orienting and soothability load onto regulation, and distress to limitations and attentional shifting load on to frustration/irritability. Rothbart (1999) has further related dimensions on the frustration factor of infancy scales to later measures of attention. When the original sample of the 1981 study were followed up at age 7, it was found that infant frustration predicted low attentional focusing.

The IBQ has recently been extended (IBQ-R; Jones, Garstein, Rothbart and Chasman, 1999) for a more fine grained assessment of temperament. This has resulted in a total of 16 scales and 250 items. In addition to the original scales (activity level, distress to limitations, distress and latency to approach sudden or novel stimuli, duration of orienting, smiling and laughter, and soothability), ten new scales (social fear, high intensity pleasure, low intensity pleasure, falling reactivity/rate of recovery from distress, cuddliness, attentional shifting, perceptual sensitivity, sadness, approach, and vocal reactivity) were found to have internal consistency, with the exception of attentional shifting. However, this new scale was not available at time of testing.

Finally, Rothbart (1984) cautions that the possibility of response bias within parental reports of infant temperament should be not be disregarded, although the use of concrete behavioural items within the IBQ should eliminate this possibility to a certain extent. This issue may be of particular relevance in consideration of the perceptions and possible biases of parents of atypically developing infants.

5.1.3 Temperament in Williams Syndrome and Down's Syndrome

Temperament in infants and children with Down's syndrome has been widely researched (for a comprehensive review, see Ganiban, Wagner, and Cicchetti, 1990).

Many researchers have approached the study of temperament from the standpoint of relating temperament to difficult behaviours, or the impact of problem behaviours on the child's environment and parent-child interactions (Thomas and Chess, 1980). It has also been suggested that difficult temperament can affect cognitive development (Wachs and Gandour, 1983). Gibson (1978) reviewed temperament and personality studies in DS, and states that despite the commonly held stereotype that individuals with DS are obstinate but affectionate, in many studies subgroups have been found which do not fit this stereotype. Furthermore, many parents of children with DS describe their children as having a wide range of personality features (Rogers, 1987). Thus, as in other atypically developing groups, and in the normal population, it should not be assumed that temperament is uniform across individuals.

Rothbart and Hanson (1983) used the IBQ in a longitudinal study of infants with DS. Fifteen parents completed questionnaires when their infants were 3-, 6-, 9-, and 12-months old, and responses were compared to those of parents of typically developing infants at 6-, 9-, and 12-months. The DS group scored lower than controls on the dimension of smiling and laughter, which is consistent with previous research which found delayed onset of laughter (Cicchetti and Sroufe, 1976), and damping of positive affect intensity (Emde, Katz, and Thorpe, 1978). At 6 and 9 months, DS infants were more fearful, and scored higher on the measure of duration of orienting than non-handicapped infants. The latter finding had also been reported previously. Miranda and Fantz (1973) found that 8-month olds with Down's syndrome responded to stimuli longer than typically developing 8-month olds. However, as Rothbart (1984) states, interpretation of specific behaviours is problematic, and this result may not mean that DS infants are processing more information. Rather, Miranda and Fantz argue that this pattern of behaviours reflects a delay in the ability to process visual information.

The finding that 6- to 9-month old infants with DS scored higher on measures of duration of orienting (Rothbart and Hanson, 1983; Miranda and Fantz, 1973) would seem to contradict the findings reported in the previous chapter. In the study of sustained attention reported in chapter 4, the 24- to 36-month old children with DS showed less duration of attention than the CA and MA typically developing control groups, and the WS group of the same age. One interpretation of this apparent contradiction would be that by 24 months, infants with DS may not need longer to process information, as suggested by Miranda and Fantz. However, it would seem unlikely that the DS group would be processing information faster than the typically developing control groups. An alternative explanation is that the two measures, duration of orienting from the IBQ and duration of sustained attention, are not tapping the same abilities. While duration of orienting is largely a measure of looking, the sustained attention task measures more enhanced processing.

The problem of response bias in parental reports, as mentioned previously, has also been addressed in relation to parents of infants with Down's syndrome. The impact of the birth of an infant with Down's syndrome can be stressful, and interact with parental expectations. Parents may have problems coming to terms with the reality of having a baby with Down's syndrome, and maintain inappropriate expectations (Emde, Katz, and Thorpe, 1978). Several studies have found poor correlations between ratings of mothers of DS infants when using questionnaires such as the Carey ITQ, as well as a more global measure of the mothers' impressions of their childrens' temperament (Bridges and Cicchetti, 1982; Gunn, Berry, and Andrews, 1981). Bridges and Cicchetti argue that while the global measure yields a relatively accurate assessment by the mother of the child's temperament, the ITQ as a measure may reflect mother-infant interactions, as was also argued in the study of temperament in typically developing infants.

The stereotypical approach to defining personality in DS persists in studies of adults with DS. Typical descriptions of people with DS as cheerful, friendly and affectionate are somewhat contradicted by studies which find higher levels of depression and other affective disorders in DS, compared to prevalence rates in the general population of people with mental retardation (Collacott, Cooper, and McGrother, 1992).

In contrast to the number of studies on temperament in DS, to date little work has been carried out on temperament in Williams syndrome, particularly in infancy and early childhood. A few studies have looked at stress and dysfunction in families with a child with Williams syndrome. Einfeld (1998) found no substantial dysfunction in families of children with Williams or Down's syndromes, when compared with families of children with Autism and Prader-Willi syndrome. It could be argued that dysfunction may be related to difficulties in temperament, but equally many other problems associated with individual syndromes may account for this finding.

A number of studies of personality in adolescents and adults with Williams syndrome have focused on vulnerabilities to specific psychopathologies. Dykens and Rosner (in press; cited in Dykens, 1999a) found high levels of global anxiety, worry, and perseverative thinking in WS, which appear to be associated with high levels of specific fears and phobias (Dykens, 1999b, cited in Dykens, 1999a). Depression, and low self-esteem has also been reported in a number of adolescents with WS (Pober and Dykens, 1996).

The Williams Syndrome Association of the U.S.A. list overly sociable personalities as a trait of the syndrome, adding that people with Williams syndrome are often less fearful of strangers than the normal population. Anecdotal reports also claim that

adults with Williams syndrome often have overly anxious personalities, and many display obsessive behaviours. Empirical work is needed in this area in order to establish the validity of such accounts. In addition, the proposed impact of difficult temperament on cognitive development should also be explored with relation to Williams syndrome.

The aim of the study presented here was to examine temperament in WS, DS and chronological and mental age matched controls, using the IBQ, in an attempt to further explore differences in attention between groups. The IBQ was selected in preference to scales for toddlers (such as the Toddler Behavior Assessment Questionnaire; Goldsmith, 1996), because of the mental ages yielded by DS and WS groups on the Bayley Scales of Development II. Use of this scale also allowed for direct comparison with the mental age-matched controls, although possibly at the cost of reliable comparison with CA controls, as many items on the scale may not be appropriate for older children. Selection of the IBQ also allows the comparison of results reported in younger infants with DS (Rothbart and Hanson, 1983) with the older group studied here. Furthermore, examination of the attentional aspects of the IBQ may also help resolve the possible contradiction between the results from the sustained attention study reported in chapter 4, and those reported by Rothbart and Hanson.

5.2 Method

The Infant Behaviour Questionnaire was given to the parents of all the infants tested, full details of whom can be found in chapter 2. These were completed at home and returned by freepost mail. Age ranges and mean age for each group are presented in Table 5.1. Response rates were somewhat higher for the WS (86%) and MA (88%) groups, than DS (58%), and CA (53%), and response rate overall was 70%.

Table 5.1 Chronological Age in months of IBQ respondents

	Mean	Minimum	Maximum
WS (n=12)	29 months 7 days	23 months 16 days	37 months 7 days
DS (n=11)	27 months 22 days	23 months 25 days	36 months 15 days
CA (n=9)	30 months 4 days	23 months 20 days	36 months 11 days
MA (n=14)	14 months 29 days	11 months 24 days	20 months 15 days

5.3 Results

Table 5.2 shows the mean scores for each of the groups on the 6 dimensions in the IBQ. It should be noted that parents of three of the CA infants felt that all questions relating to soothability were not applicable. Therefore there are 3 fewer scores on this measure.

Table 5.2 Mean scores on IBQ dimensions for each group.

	Activity		DTNS		Orientation		DTL		Smiling		Soothability	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
WS (n=12)	3.11	0.86	2.54	0.84	3.94	1.38	3.84	0.79	5.28	1.08	5.03	1.35
DS (n=11)	4.46	0.54	3.45	0.69	4.50	1.36	3.51	0.90	5.19	0.52	5.21	0.99
CA (n=9, 6)	4.12	1.42	3.18	0.95	5.06	1.16	3.34	0.61	5.07	0.60	5.32	1.15
MA (n=14)	3.97	0.94	2.75	0.40	3.12	1.13	3.69	0.67	4.84	0.91	4.90	0.96

A one-way Anova was performed for each measure, which revealed that there was a significant difference between groups on Activity level ($F(3,42) = 4.130$, $p < 0.05$), on Distress and Latency to Approach Sudden or Novel Stimuli ($F(3,42) = 3.755$, $p < 0.05$), and on Duration of Orienting ($F(3,42) = 4.948$, $p < 0.05$). No significant differences were found between groups on Distress to Limitations ($F(3,42) = 0.874$, n.s.), Smiling ($F(3,42) = 0.666$, n.s.), or Soothability ($F(3,39) = 0.270$, n.s.). For Activity level, the significant difference was between the WS group and the DS group, the DS group scoring higher than the WS group (Tukey's HSD, $p<0.05$). For

Distress and Latency to Approach Sudden or Novel Stimuli, the difference was again between the WS group and the DS group (Tukey's HSD, $p<0.05$), the DS group scoring higher than the WS group. For Duration of Orienting, the MA group scored significantly lower than both the DS and the CA groups, but was not significantly different from the WS group (Tukey's HSD, $p<0.05$).

5.4 Discussion

The aim of this study was to examine temperament in infants with WS, DS, and chronological and age matched control groups, using the IBQ. Of particular interest were items relating to attention, such as duration of orienting. Results show that the DS and CA groups scored higher on duration of orienting than the MA group, but that the WS group was not significantly different from the MA group. Thus these results are not consistent with the finding of greater duration of orienting in DS in the Rothbart and Hanson (1983) study. This may indicate that by the time the child with DS has reached the age of two, the delay in processing visual information has reduced, so that looking behaviours are comparable with chronological age matched controls. However, it may also be interpreted as less report bias, in that parents of children with Down's syndrome no longer have unrealistic expectations of their child, which Emde, Katz, and Thorpe (1978) claim tends to happen once mothers of infants with Down's syndrome perceive the shift from infancy to toddlerhood.

This pattern of results is of interest when compared to the results on sustained attention from Chapter 4, which showed that DS infants had shorter duration of sustained attention than WS infants, and fewer periods of sustained attention than MA infants. Thus if duration of orienting can be viewed as a measure of attention, there would appear to be a discrepancy between the results from the sustained attention study, and the results from the temperament study. However, it could also be argued that these measures are not tapping the same abilities. The duration of orienting items in the Infant Behavior Questionnaire typically ask questions about how often the child

looked at books, or other objects for a certain length of time. Looking at an object does not necessarily equate to the enhanced processing thought to be taking place during periods of sustained attention. Simply looking at an object may reflect periods of casual attention. If duration of orienting can be viewed as a measure of casual attention, then the results of the two studies of attention reported here would seem to indicate that infants with WS appear to be better at sustained attention, while infants with DS show more casual attention. However, this interpretation must be treated with caution, and further work is required to tease out the different elements within attention in the measures reported here.

Furthermore, the use of the IBQ may also be questioned in terms of whether it is an appropriate scale for the age groups studied here. The IBQ was selected because of the mental ages yielded by the DS and WS infants on the Bayley Scales of Development. However, this may not have been appropriate, and with hindsight it may have been better to use a toddler temperament scale, such as the Toddler Behavior Assessment Questionnaire (Goldsmith, 1996).

The results also showed that the DS group scored higher than the WS group on the fear measure, distress and latency to novel and sudden stimuli. Although the DS infants did not differ significantly from control groups on this measure, they did score higher than other groups, a trend which would appear to be consistent with the higher fear scores from the Rothbart and Hanson (1983) study.

Finally, the DS group scored higher than the WS group on activity level. The Rothbart and Hanson study found no difference between DS infants and controls on this measure, but this may have been because infants tested in their study were much younger. Alternatively, it may be that the difference occurred due to lower activity levels by the WS group. It should be noted that both WS and DS tend to exhibit delayed motor development, and while motor skills as such were not assessed here, it

is unlikely that differences in motor ability could account for this finding. It is interesting to compare these scores on activity level with the results from the sustained attention study. One of the global measures of sustained attention, and consequently a criterion used to judge observed periods of sustained attention, is amount of extraneous movement. On this measure, infants with DS also scored higher than infants with WS. In some respects this is not an entirely fair comparison, in that activity level on the IBQ is not a task-specific measure, but relates to amount of movement observed by parents in normal domestic situations, for example, when in a car seat or during feeding. Nevertheless, there would appear to be a relationship between attention and movement in the two clinical groups across these tasks.

In conclusion, the results of the study presented here are consistent in part with the results from the earlier study using the IBQ in younger children with Down's syndrome. Of more interest is the differences between the infants with Williams and Down's syndromes, and the way in which these differences relate to other abilities. These relationships will be further explored and discussed in Chapter 7.

6

FACE PROCESSING

6.1 Introduction

Face processing in infancy and early childhood is an area that has been extensively researched. A number of sophisticated abilities in new-born infants have been reported, and will be reviewed in this section. However, demonstrations of particular competencies have been interpreted in a number of conflicting ways, which typically address the issue of domain-specific innateness, or otherwise, of general cognitive abilities. In adults with Williams syndrome, face processing has been reported as being spared in the presence of deficits in other visuo-spatial skills. However, there is some question as to whether this ability is actually 'spared', i.e. whether it equates to face processing in the normal adult population. Little, if any, work has been reported on face processing in infants with Williams syndrome, but it may be that research in this area can assess whether the development of face processing in individuals with Williams syndrome occurs in an atypical manner. The aim of this section is to present a review of the literature on face processing in typical infancy, and in adults and infants with Williams syndrome and Down's syndrome. Data will then be presented from a study of the formation of prototypes in face processing in infants and toddlers with Williams and Down's syndromes, as well as in chronological and mental age matched control groups.

6.1.1 Face Processing in Typically Developing Infants

Human infants demonstrate a strong interest in faces from a very early age, and a large amount of research has led to the claim that new-borns show a preference for faces over other stimuli. Goren, Sarty, and Wu, (1975), and Johnson, Dziurawiec, Ellis, and Morton (1991) have demonstrated that new-borns will track face-like stimuli in preference to various equally complex but scrambled stimuli. However, Maurer and Young (1983) claimed that there was no general face preference, as one of the scrambled stimuli in their study was followed as much as the face-like stimuli. Bremner (1994) suggests that face preference at birth will only be elicited by moving stimuli, and perhaps this may account in part for the negative result found by Maurer and Young, as the velocity of moving stimuli has been shown to affect the infant's ability to track smoothly (Aslin, 1981).

The role of movement in the perception of faces is also demonstrated in a study that used moving points of lights to represent facial movement. Stucki, Kaufmann-Hayoz, and Kaufmann (1987) found that 3-month-old infants were able to discriminate faces on this basis. This has been related to the representation of a body schema; Bremner (1994) suggests that infants identify the human face and body through complex, dynamic relational information. Some support may be found in a study in which 3 month old infants could discriminate body movements represented by light points (Bertenthal, Proffitt, and Cutting, 1984).

Johnson (1997) argues that standard preference test procedures tend to use static stimuli, and thus fail to demonstrate face preference in the new-born because they do not make demands upon the critical brain regions, which he proposes are subcortical at birth. Morton and Johnson (1991, and Johnson and Morton 1991) propose that

new-born infants make use of a subcortical face orienting mechanism (CONSPEC). There is also a cortical mechanism that takes over at 2 months (CONLERN). This theory states that new-borns are predisposed to attending to face-like stimuli, but need experience to learn about details of those stimuli. In terms of the mechanisms involved in their theory of face processing, Johnson and Morton propose that biases exist within the sub-cortical, orienting system, which relay the salient information to the developing cortex. The theory is partly based on tracking behaviour in infants less than 2 months of age. While infants do not attend to faces more than other stimuli when presented in the central visual field, they will track faces further, by making head or eye movements, when presented in the peripheral visual field (Goren, Sarty, and Wu, 1975; Johnson, Dziurawiec, Ellis, and Morton, 1991). However, this preference for tracking faces is displayed only when faces are presented in the temporal, and not the nasal visual field. This suggests that the retinotectal rather than the geniculostriate pathway is involved (Simion, Valenza, Umilta, and Barba, 1998).

Evidence for the two-process theory of face perception comes from the failure to find face preference in new-borns using static stimuli, which would not require use of the temporal visual field, which feeds mainly into the subcortical visual pathway. Also, the decline of face tracking during the first three months (Johnson, Dziurawiec, Ellis, and Morton, 1991) could be interpreted as the implementation of the new, cortical face processing system. Anatomical evidence can be adduced from the failure to find lateralization of face processing in early infancy (de Schonen & Mathivet, 1989). Lateralization in face processing is found in adults, as well as in infants from 4-5 months (de Schonen & Mathivet, 1990), and is thought to be characteristic of cortical functioning. Gaze detection, which is also thought to be mediated by cortical circuits, is not found in infants under 4 months (Vecera & Johnson, 1995).

Johnson and Morton (1991) claim that the preference for faces in new-borns is prompted by the gross composition of the face-like form, but that by 5 months infants prefer faces with moving internal features. This is also proposed as an account of the ability of very young infants to recognise individual faces, normally their mothers (e.g. Field, Woodson, Greenberg, and Cohen, 1982). However, Walton, Bower, and Bower (1992) found preference for mothers' faces in new-borns when gross features such as colouring and hairline were matched. Although Johnson (1994) accounts for this in terms of a non-specific pattern-learning mechanism, it would seem that some learning not consistent with a subcortical mechanism is already occurring.

Johnson and Morton have drawn comparisons between their two-process theory of face processing in new-borns, and a two-process theory of filial imprinting in the domestic chick. New-born chicks form an attachment for the first conspicuous object they see after hatching. Several hours after this, they show a strong preference for the object originally seen, providing it is moving, and of a suitable size. However, in the chick's natural environment, it is highly likely that the first object encountered will be the adult hen. It is suggested that imprinting in the chick is controlled by two brain systems. The first predisposes the chick to orient towards stimuli which resemble an adult hen (Horn, 1986). This directs the second system, the function of which is to gather information and learn about the selected stimuli. It is further claimed that these two systems are supported by largely independent neural substrates. Although the brain structures of human infants and chicks are dissimilar in many ways, it is claimed that underlying biochemical and neural principles are the same for cortical and chick forebrain plasticity, and that constraints on plasticity are also likely to be similar (Johnson, 1993).

The principal alternative to Johnson and Morton's two process theory of face preference in new-borns is the revised sensory hypothesis (e.g. Kleiner and Banks, 1987). The basic concept of this theory is that faces are not processed differently from any other visual stimuli, but are preferred by new-borns because the properties of faces are a better match for their age appropriate contrast sensitivity function than other stimuli. However, Valenza, Simion, Cassia, and Umiltà (1996) demonstrated that new-borns still show a preference for face-like stimuli when presented together with stimuli which have optimal spatial frequency for the new-born, in terms of their age appropriate contrast sensitivity function. These results support Johnson and Morton's hypothesis, and therefore reject the sensory hypothesis. Valenza et al. also reported findings that were not consistent with the Johnson and Morton theory, in that only measures that indicated detection rather than orienting to the stimuli were found to produce reliable face preference. Valenza and colleagues argue that if face preference in new-borns is supported by a subcortical mechanism, then it would be expected that orienting measures would be more reliable in demonstrating face preference. However, in a more recent study, this group of researchers present results which do demonstrate orienting to facelike stimuli (Simion, Valenza, Umilta, and Barba, 1998), thus supporting the proposal of activity mediated by a subcortical mechanism.

In summary, a large body of recent research acknowledges the presence of face preference in new-borns, which appears to change during the first few months of life to a process which supports face recognition as well as face preference. Initial face preference may be subcortically mediated, giving way to a cortical mechanism at around two months, but there remain several problems with this proposal. One way in which the problematic results concerning new-borns' recognition of mothers faces

may be reconciled with the Johnson and Morton theory, concerns the types of representations formed and stored during face processing, and the creation of prototypical representations of faces.

6.1.2 The Creation of Prototypes in Face Processing

One of the mechanisms proposed to underlie face processing is the abstraction of information from individual faces, which is then used to create a prototypical face. New faces that are then encountered are coded as to how far they deviate from the stored prototype. Several studies have demonstrated robust prototype creation in adults, often using a paradigm which examines the effect of distinctiveness of faces on recognition time (e.g. Valentine and Bruce, 1986). The logic behind this type of comparison is that distinctive faces will be further removed from the prototype, and are therefore easier to judge as being familiar or not.

The ability of infants to abstract information and create prototypes has also been investigated. Several studies have demonstrated that infants as young as 2-3 months are able to form categories or prototypes on the basis of shape (Slater 1997), or types of animals (Quinn, Eimas and Rosencrantz. 1993). Recent studies have also begun to suggest that 6 month old infants have the ability to categorise faces, and furthermore that they can create face prototypes (Rubenstein, Kalakanis and Langlois, 1999).

The ability to abstract information from faces and store this in the form of a prototypical face can be applied to findings that were problematic for the Johnson and Morton two-process theory of face processing, as outlined above. The fact that new-borns are still able to recognise their mother's faces when gross facial features are

matched with controls (Walton, Bower, and Bower, 1992) was difficult to account for in the original form of the theory, which argued that face processing in the first few months of life is largely sub-cortical, and consists mainly of a face orienting mechanism. However, it has been proposed that face recognition at birth may be supported by an early hippocampal-based memory, which stores single exemplars of stimuli, but does not allow for comparison or interaction between such exemplars, such as would be required to create a prototypical face (de Haan, Johnson, Maurer, and Perrett, submitted). De Haan et al. argue that the ability to relate information between faces and thereby create a prototype may not be present from birth, and would only appear with the development of the cortical system, at around the age of 2 months. This was tested in their study, which examined prototype formation in 1 and 3 months olds. Infants were familiarised to individual faces, and then tested for preference of one of the faces used during familiarisation versus a computer generated average of all the faces seen. Infants in both age groups were able to recognise individual faces, but only the 3 month olds showed a preference for the familiar against the averaged face, indicating that the familiar face was more novel for them than the prototypical face, despite the fact that they had never seen the prototypical face, but rather created it in memory. This implies that these infants were able to abstract information from individual faces and create a face prototype. Thus the results from this study would appear to support the revised version of the Johnson and Morton theory.

6.1.3 Face Processing in Williams Syndrome and Down's Syndrome Adults

As mentioned above, individuals with Williams syndrome have been reported as demonstrating preserved face processing abilities, while exhibiting seriously impaired

performance on other visuo-spatial tasks. A number of studies have investigated face processing in adults and adolescents with Williams syndrome. Bellugi, Bihrle, Neville, Jernigan and Doherty (1992) demonstrated that adolescents with Williams Syndrome perform significantly better than Down's Syndrome matched controls, and do not differ from normal adult controls, on the Benton Test of Facial recognition. This test requires participants to select which of three or six faces, which differ from the target on lateral rotation or shadowing, match the target face for identity. On the Mooney Closure Test, in which participants classify high contrast faces as old/young and male/female, again adults with Williams syndrome performed as well as normal adult controls, and significantly better than Down's syndrome matched controls (Bellugi et al. 1988). By contrast, WS performance on tests of closure with non face-like stimuli tends to be impaired, indicating that something special may be occurring in the case of face processing. Wang, Doherty, Rourke, and Bellugi (1995) argue that this profile, in comparison with the more homogeneous profile of Down's Syndrome, may map onto the functional dichotomy between the ventral and dorsal pathways in the cerebral cortex.

An alternative approach to the investigation of face processing in people with Williams syndrome, the study of the development of these processes, may reveal more than examination of the end state. Karmiloff-Smith (1997) suggests that the fact that older children and adults with Williams Syndrome perform behaviourally like normal controls on the Benton Test of Facial recognition does not necessarily equate to the same starting point, or the same cognitive processes. Individuals with Williams Syndrome focus more than normals on facial features (Karmiloff-Smith, 1997), and also show less inversion effect. Recognition of faces is normally severely affected when faces are inverted (Yin, 1969), but people with Williams syndrome are less

perturbed by this type of manipulation. Individuals with Williams syndrome have also been reported as describing their strategy when performing such tasks as concentrating on feature matching, rather than on global configuration (Wang et al., 1995).

Face processing in individuals with Down's Syndrome is more in line with their general cognitive profile, than is found in people with Williams syndrome. Performance on discrimination of upright faces is poor, as is performance with inverted faces (Bellugi et al., 1992). However, these results have been found when testing older children and adults with Down's Syndrome. As with Williams syndrome, by solely examining adult abilities, the processes involved in the development of these abilities may be missed.

6.1.4 Face Processing in Williams Syndrome and Down's Syndrome Infants

Although little work has been done on face processing in very young infants with Down's Syndrome, several interesting results have been found. Legerstee and Bowman (1989), demonstrated that, unlike normally developing infants, it was not until they were older than 6 months that infants with DS discriminated between active and passive faces of adults, or between the face of the mother or a stranger. However, ERP recordings reveal that differences between infants with and without Down's Syndrome may be more subtle than are found in later childhood. Karrer, Wotjascek, and Davis (1995) recorded event related potentials (ERPs) from 6-month-old infants with and without Down's Syndrome, who were tested on a visual recognition memory task using faces. While both groups demonstrated similar ERP waveform morphology, the authors claim that amplitude differences between the groups may implicate less frontal attentional processes in Down's Syndrome. This result, in

conjunction with the finding that response decrements over trials were found for the central cortex in DS, were taken by the authors as an indication that habituation in DS may take place in some neural processes, but not others. Down's Syndrome infants also exhibited similar visual fixation as controls, as measured by corneal reflection, suggesting that the differences exhibited by older children with Down's Syndrome and controls may not yet be firmly entrenched in infants with Down's Syndrome.

Although several studies have focused on face processing in children and adults with Williams syndrome, to date none have been presented which examine this ability in infants with Williams syndrome.

6.1.5 Prototype Creation in Williams and Down's Syndromes.

To date, no studies have been published relating to the ability of people with Williams syndrome to create prototypes, or categories. However, it might be argued that results from some of the studies in face processing could be interpreted as indicating that people with Williams syndrome do not use prototypes as a basis for recognising individuals. The findings that people with Williams syndrome show less inversion effect, as well as self reports of feature matching, may indicate that instead of creating prototypes, they rely on exemplar learning and comparison, similar to that proposed to occur in very young infants.

Although there are no studies of prototype creation in WS, a recent study of concept formation in WS may be of relevance. When tested on conceptual repertoire, or intuitive knowledge about the biology of plants and animals, people with Williams syndrome did not differ from controls matched on verbal mental age, on concepts normally found in pre-school children (Johnson and Carey, 1998). However, when

tested on concepts normally found in children of 6 years and older, the WS group were impaired relative to controls. Although this tests animal knowledge, and the formation of concepts relating to biological entities, and thus cannot be generalised to faces and the formation of prototypes, it may indicate a possible problem in categorical representations, which might be extrapolated to the creation of prototypical faces in face processing.

Most of the work on categorisation or concept formation in people with Down's syndrome has been in the domain of language. However, several studies have been conducted using object or picture sorting tasks. Tager-Flusberg (1985) found that 11 year olds with Down's syndrome formed the same categories as adults and typically developing children and, as with typically developing children, those with Down's syndrome showed category extensions which were not identical to adults.

Further investigation of early face processing abilities in infants with Williams and Down's Syndromes may help to resolve some of the inconsistencies within the two-process theory of infant face recognition, as proposed by Johnson and Morton (1991). Face processing in Williams Syndrome is apparently spared, in comparison to other visuo-spatial abilities. However, this may develop in an atypical fashion, due to early differences, possibly in terms of the proposed subcortical mechanism. Comparison with early face processing abilities in Down's Syndrome will hopefully add to knowledge about both syndromes, as well as to normally developing face processing. To this end, the study reported here aimed to examine face processing in these groups of infants.

The experiment reported in this chapter looks at the formation of prototypes in face processing, and replicates the study of de Haan, Johnson, Maurer, and Perrett (submitted), as described previously. After a familiarisation phase with several faces, infants will then be presented with the computer generated averaged face and a familiar face. If infants are forming a prototype during the familiarisation phase, then the computer generated averaged face should be more familiar than any one of the familiar faces, as the prototype contains components of all faces seen. Therefore, when tested, they should prefer to look at the familiar face, as this would be relatively more novel. Novelty preference will also be tested. It is hypothesised that the infants with Williams syndrome may not create prototypes in face processing, and will therefore prefer to look at the average face, as this would be more novel if a prototype has not been formed. Other groups should exhibit prototype formation, and should therefore prefer to look at the familiar face in the test of averaged versus familiar face. It is also predicted that all groups will be able to recognise the familiar faces, and will therefore exhibit a preference for the novel face when presented with a familiar face.

6.2 Method

6.2.1 Design

A between-subjects design was used, with 4 groups; Williams syndrome (WS), Down's syndrome (DS), chronological age-matched controls (CA), and mental age-matched controls (MA).

6.2.2 Participants

11 WS infants, 15 DS infant, 14 CA infants and 12 MA infants were tested from the main sample described in chapter 2. Age ranges and mean age for each group are presented in Table 6.1

Table 6.1 Mean and range of ages for each group.

	Mean	Minimum	Maximum
WS (n=11)	31 months 19 days	23 months 16 days	37 months 7 days
DS (n=15)	29 months 25 days	23 months 25 days	36 months 25 days
CA (n=14)	30 months 11 days	23 months 14 days	36 months 29 days
MA (n=12)	15 months 11 days	11 months 24 days	20 months 15 days

6.2.3 Stimuli

Nine colour pictures of female faces were used; faces 1-4 in the familiarisation phase, faces 5-8 were novel faces used in the test trials, and face 9 was the computer generated averaged face. Hair was deleted from the pictures, and faces were presented against a grey background. The faces were 12 cm high by 10 cm wide, and 14 by 17 visual degrees when viewed from a distance of 40 cm. The averaged face was created from the four faces used in familiarisation. The average position of 224 feature points was calculated for the four faces, and each face was then morphed into the average shape. These were then combined by averaging colours and intensities of corresponding pixels. The stimuli described here were provided and created by David Perret (Rowland and Perret, 1995).

6.2.4 Apparatus

Figure 6.1 presents a representation of the Fagan box and the preferential looking set-up. A toddler car seat was used to seat the infants, which positioned infants in a semi-reclining position, such that the angle of gaze was approximately 45 degrees. A testing booth was constructed based on the visible preference apparatus developed by Fagan (1970). The booth consisted of grey painted wood on two sides and on top, and the front of the box was grey cloth. The box was mounted on wheels and was moved over the infant when seated in the car seat. A door was positioned directly in line with

the infant's gaze, and was used to display stimuli in two slots on the inside of the door. A central peephole was used to record looking times to left and right by a trained observer using two stopwatches. The door was opened to a horizontal position to change the stimuli after each trial, at which point a screen of high contrast stripes was positioned in the gap to ensure that the infant did not see the faces of the experimenter or observer. The screen also had a small peephole to allow the observer to ascertain that the infant was positioned centrally, and looking in the right direction and was therefore ready for the next trial.

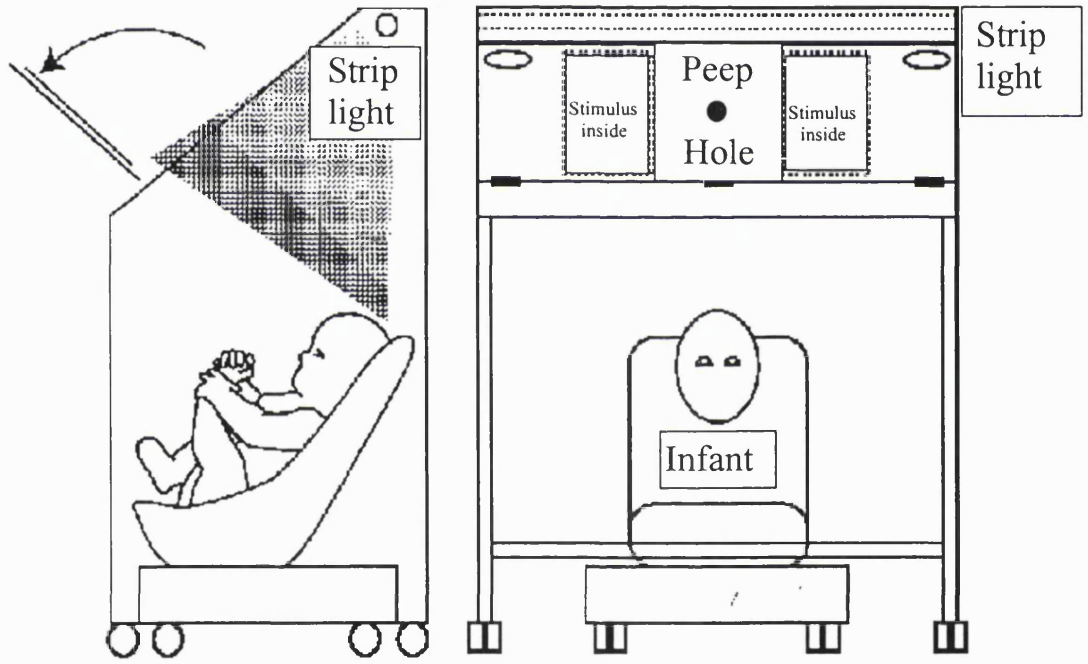

Figure 6.1 Side and front view of the preferential looking set-up.

6.2.5 Procedure

Infants were settled in the car seat and positioned centrally in the Fagan box. Four familiarisation trials were followed by four test trials. Trials began when the infant looked to either of the stimuli. Looking times to right and left were measured by an

observer who was blind as to positions of specific face types in the test trials. These times were recorded by the experimenter at the end of each trial.

Familiarisation trials: Infants had 4 familiarisation trials, in each of which a pair of matching faces was presented for 30 seconds. This time was based on the familiarisation time determined for 3 month olds in the de Haan et al study, and was piloted on several infants. If infants scored a total looking time of less than 10 seconds on any trial, faces were presented for a further 10 seconds. The order of the four faces used was counterbalanced across infants and between groups.

Test trials: Test trials were 10 seconds, unless infants failed to look for a total of 1 second, in which case faces would be re-presented for a further 10 seconds. Two tests in which the averaged face was presented with a familiar face, and then a left/right reversal, were followed by two tests in which a novel face was presented with a familiar face, and then reversed. Averaged and novel faces were presented on the left during the first presentation for half of each of the four groups. Each of the familiar faces was paired with both the averaged face and the novel face on an equal number of occasions.

6.3 Results

6.3.1 Familiarisation

Average looking times for each trial in the familiarisation phase for all four groups is shown in Table 6.2, which also includes total average looking time across trials for all four groups. This breakdown in data was unavailable for one of the WS infants.

Average looking times for all groups was less on trial 4 than on trial 1, although the differences are very small.

Table 6.2 Average looking time in seconds

	TRIAL 1		TRIAL 2		TRIAL 3		TRIAL 4		TOTAL	
	Mean	SD	Mean	SD	Mean	SD	Mean	SD	Mean	SD
WS (n=10)	15.8	3.7	14.2	3.52	15.8	4.96	15.11	3.18	60.45	10.51
DS (n=15)	18.9	4.28	18.2	5.05	17.1	3.97	18.27	5.27	72.43	11.84
CA (n=14)	19.0	5.56	16.6	6.91	16.1	5.08	18.93	5.47	70.94	17.84
MA(n=12)	15.8	6.46	13.8	3.39	13.2	2.56	14.27	3.13	56.59	9.98

To determine whether there was a significant decrease in looking times over trials, which would indicate habituation to the stimuli, paired samples t-tests were carried out for each group, comparing length of looking time on Trial 4 against looking time on Trial 1. None of the groups showed a decrease in looking times across trials (WS: $t(9) = -0.60$, n.s.; DS: $t(14) = -0.38$, n.s.; CA: $t(13) = -0.05$. n.s.; MA: $t(11) = -0.86$, n.s.). Although these results might seem to indicate that habituation may not have been achieved, infant experiments of this type, particularly those which use faces as stimuli, do not always show habituation across trials. Furthermore, the time allowed for familiarisation was suitable for three month olds in the de Haan et al. study, and was therefore deemed to be adequate for the age groups tested here.

As comparison of trials failed to yield results indicative of habituation, total looking time, as presented in Table 6.2, was examined, to determine whether there was any difference between groups in terms of total looking time. The logic of such a comparison is that if any groups look less than others, this may be another indication of habituation, as it might be interpreted as boredom in the infants in that group. Average total looking times (i.e. across familiarisation trials) for all four groups

shows that the DS group had the longest total looking time, and the MA group the shortest. A one-way Anova was performed, which revealed that there was a significant difference between groups ($F(3,50) = 3.830$, $p<0.05$), and that the difference occurred between the MA and the DS groups, (Tukey's HSD, $p<.05$).

6.3.2 Novel Face v. Familiar Face

The average proportion of time spent looking at the novel face when presented with a familiar face is shown in Table 6.3. The figures in Table 6.3 are averaged from the two tests of novel versus the familiar face.

Table 6.3 Mean looking time to novel face (seconds)

	MEAN	SD
WS (n=11)	56.5%	18.3
DS (n=15)	47.7%	10.8
CA (n=14)	43.7%	14.8
MA (n=12)	53.5%	24.2

To test for novelty preference, paired samples t-tests were performed for each group, comparing looking time to the novel face against chance. None of the groups showed a significant preference for the novel face (WS: $t(10) = 1.17$, ns; DS: $t(14) = -0.83$, n.s.; CA: $t(13) = -1.59$, n.s.; MA: $t(11) = 0.5$, n.s.). In addition, a one-way Anova revealed that there was no significant difference between groups ($F(3,48) = 1.195$, n.s.).

6.3.3 Averaged Face v. Familiar Face

Table 6.4 shows the mean proportion of time spent looking to the averaged face when presented with the familiar face, for each of the four groups. Again, these figures are the average of the two tests of the averaged versus the familiar face. The DS group

spent the least proportion of time looking at the averaged face, while the WS group spent slightly more time looking to the averaged face.

Table 6.4 Mean looking time to averaged face (seconds)

	MEAN	SD
WS (n=11)	60.6%	23.8
DS (n=15)	51.8%	14.2
CA (n=14)	55.9%	15.1
MA (n=12)	60.4%	15.2

To determine whether any group showed a significant preference for the averaged face, paired samples t-tests were performed for each group, comparing looking time to the averaged face against chance. None of the groups showed a significant preference for the averaged face (WS: $t(10) = 1.48$, n.s.; DS: $t(14) = 0.49$, n.s.; CA: $t(13) = 1.37$, n.s.; MA: $t(11) = 2.06$, n.s.), and a one way Anova revealed no significant differences between groups ($F(3,48) = 0.49$, n.s.).

Table 6.5 Mean looking time to averaged face (seconds) for 3 month-olds.

	MEAN	SD
3mo (n=24)	32.16%	23.4

When compared with data from 3 month olds from de Haan et al. (see Table 6.5), a one-way Anova showed that there were significant differences between groups ($F(4,71) = 6.786$, $p<0.001$). Tukey's HSD revealed that these differences lay between the 3 month olds, and all the other groups ($p<.05$), with the 3 month-olds looking at the averaged face significantly less than the other groups.

6.3.4 Novel Face Preference >50%

The above results appear to indicate that not all of the infants were able to demonstrate novelty preference, and that they may have failed to familiarise to the stimuli. This probably affected their choice when presented with the averaged and the familiar face. Therefore infants were omitted who demonstrated less than 50% novelty preference to the novel individual face. Table 6.6 shows the mean proportion of time spent looking to the novel face under this criterion.

Table 6.6 Mean looking time to Novel face for infants with > 50% novelty preference.

	MEAN	SD
WS (n=6)	71.17%	5.98
DS (n=7)	57.00%	3.74
CA (n=5)	58.80%	9.58
MA (n=7)	70.14%	11.85

A one way Anova revealed that there was a significant difference between groups ($F(3,21) = 4.97$, $p < 0.05$). Tukey's HSD revealed that there was a significant difference between the DS group, and the WS and MA groups ($p < 0.05$). Paired samples t-tests were performed to determine whether any groups showed a statistically significant novelty preference. This was found to be the case for the WS group ($t(5) = -8.67$, $p < 0.0001$), the DS group ($t(6) = -4.95$, $p < 0.05$), and the MA group ($t(6) = -4.50$, $p < 0.05$), but not for the CA group ($t(4) = -2.06$, n.s.).

6.3.5 Averaged Face v. Familiar Face for infants with Novel Face Preference >50%

Table 6.7 shows the mean proportion of time spent looking to the averaged face for the infants who demonstrated greater than 50% novelty preference in each group. Although the WS group spent the longest proportion of time looking to the averaged face, paired samples t-tests revealed that no group looked significantly longer at the

averaged face, (WS: t(5) = 1.47, n.s.; DS: t(6) = 0.91, n.s.; CA: t(4) = 0.44, n.s.; MA: t(6) = 1.51, n.s.), when only infants who scored greater than 50% novelty preference were included. In addition, no significant differences were found between groups when a one-way Anova was performed (F(3,21) = 0.619. n.s.).

Table 6.7 Mean looking time to averaged face (seconds) for infants demonstrating >50% novelty individual face preference.

	MEAN	SD
WS (n=6)	66.5%	27.52
DS (n=7)	54.86%	14.10
CA (n=5)	53.00%	15.20
MA (n=7)	58.00%	14.00

6.3.6 Novelty Preference: First Test only.

The test for the preference for the novel face involved two presentations of the novel face with the familiar face. To further explore the failure to find any preference for the novel face, data were extracted from the first presentation, as it could be argued that infants may have habituated to the new face by the second presentation. The data from the first presentation of the novel and familiar face are presented in Table 6.8.

Table 6.8 Mean looking time to novel face on first test only

	MEAN	SD
WS (n=10)	55.81%	30.64
DS (n=15)	53.38%	30.40
CA (n=14)	51.06%	24.72
MA (n=12)	48.28%	29.41

A one-way Anova revealed no significant differences between groups ($F(3,47) = 0.142$, n.s.), and paired sample t-tests revealed that no groups demonstrated a significant novelty preference when compared with chance (WS $t(9) = -0.60$, n.s.; DS $t(14) = -0.431$, n.s.; CA $t(13) = -0.16$, n.s.; MA $t(11) = 0.202$, n.s.).

6.3.7 Average Preference: First Test only.

The same logic regarding presentation of stimuli and possible habituation by the second test was applied to the data for preference for averaged versus familiar face. Data from the first presentation of the averaged and familiar face are presented in Table 6.9.

Table 6.9 Mean looking time to averaged face (seconds), for first presentation only.

	MEAN	SD
WS (n=10)	70.36%	31.8
DS (n=15)	54.43%	22.2
CA (n=14)	55.21%	29.0
MA (n=12)	53.86%	28.7

A one-way Anova did not reveal significant differences between groups ($F(3,47) = 0.881$, n.s.).

6.3.6 Relationships between factors

Applying Pearsons product moment correlation to the proportion of time spent looking at the averaged face, the novel face, total looking time during familiarisation, and groups, revealed a significant negative correlation between total looking time during familiarisation, and proportion of time spent looking at the averaged face ($r = -0.3561$, $n = 51$, $p < 0.05$).

6.4 Discussion

The aim of this study was to determine whether infants with Williams syndrome differed from infants with Down's syndrome, and chronological and mental age matched infants, in terms of creating a prototype from a group of individual faces. It was predicted that the WS group would not create prototypes during face processing, and would therefore prefer to look at the computer generated averaged face, as this would be more novel for them after familiarisation with individual faces. The results failed to support the predictions, and questions regarding the suitability of the task are addressed here.

6.4.1 Familiarisation Times and Habituation Results.

The results from the test for habituation revealed that none of the groups showed a decrease in looking times over the familiarisation trials. This could be interpreted as a failure to habituate to the stimuli by all of the groups. While this might be argued for the WS and DS groups, as they may require longer looking times during familiarisation, this would appear to be unlikely in the case of the control groups, given that the time allowed for familiarisation was based on that determined for 3 month olds in the de Haan, Johnson, Maurer, and Perrett (submitted) study.

The overall behaviour of the infants during this task may reveal more about their performance. In general, the infants did not appear to enjoy the task, as it required sitting still for a moderate length of time and, with the exception of some of the DS and WS infants, all were able to walk at the time of testing, and seemed to find it tedious to have to remain seated. Although this behaviour was not apparent in any of the other tasks reported in this thesis, the procedure in this task differed from others in that the infants were behind a screen, and were being presented with static stimuli.

Although the observer could and did talk to the infants during testing, it may not have provided enough social interaction for them. The study on which this was based tested much younger children (one and three month olds), and it is possible that it was not stimulating enough for older children.

An alternative paradigm which could have been used in this type of study is the habituation paradigm, when infants determine their own level of reduced attention, rather than the familiarisation-novelty paradigm, where exposure is uniform for all subjects and is controlled by the experimenter. This would allow for greater certainty that each individual has habituated to the stimuli. However, the familiarisation-novelty paradigm is more useful for group comparisons, as the same procedure is used across groups, and it is also quicker to administer when running a large number of studies on the same day.

Recent research has yielded a suggestion for an alternative to the variables commonly measured in tests of preferential looking, such as total time or proportional time spent looking at stimuli, or duration of first look. Schafer and Plunkett (1998) argue that if an infant becomes less involved in the task as the trial proceeds, this may result in random behaviour that will mask any effect of preference. However, this type of behaviour would not be sufficient to hide the longest look. By recording the task on-line, the measure of longest look would be simple to extract, and should be considered in future studies using a preferential looking design.

6.4.2 Preference for Averaged Face and Novelty Preference Results.

During administration of the task, two tests of novel versus familiar face and two tests of the averaged versus familiar face were given; one presentation of each test pair

followed by a left/right reversal. In the initial analysis, which examined looking times over both test trials, no groups showed a significant preference for the novel face, and therefore no preference either for the familiar face. Similarly, the test for averaged versus familiar face preference showed no group preference for either averaged or familiar faces. A second analyses of preference for the novel face was conducted which included only those infants who had demonstrated over 50% novelty preference, in order to examine the performance of those infants who did demonstrate novelty preference. This analysis revealed that the WS, DS and MA groups showed a significant novelty preference, and that the DS group showed significantly less novelty preference than the WS and MA groups. However, as the level of novelty preference had been artificially manipulated, by only selecting those infants who had demonstrated greater than 50% novelty preference, this result was not surprising. Of greater interest was how these infants performed with the test of averaged versus familiar face preference. However, no significant preference was demonstrated in any groups. Next, only the first test of preference for the novel or the familiar face was considered, on the basis that infants may already have habituated to the test faces by the second presentation. Again there was no significant novelty preference in any of the groups, and no difference between groups. Finally, analysis of the first test only of averaged face preference did not reveal differences between groups.

This group of results indicate that there may be problems in interpreting the data, as neither of the typically developing control groups behaved as predicted. It was predicted that both typically developing control groups and the DS group would not show a preference for the averaged face, and by implication that they should prefer to look at the familiar face. However, this was not found in any of the tests of averaged versus familiar face preference. Furthermore, it was also predicted that all groups

should be able to remember individual faces, but the results of the test for novelty preference do not support this prediction. One explanation for this pattern of results might be found in the failure to find habituation during familiarisation. If the infants had indeed failed to process the stimuli sufficiently to produce a decrement in looking time during familiarisation, then this might account for failure to find novelty or preference, as the familiar face which was re-presented with the novel face would not be remembered. Similarly, failure to habituate could account for failure to find any preference in the test of averaged versus familiar face, as if individual faces were not remembered, then infants would not be able to create a prototype, or exhibit a preference for either face.

It could be argued then that the results presented here are consistent with those that would be expected when infants had failed to familiarise to the stimuli, due to inadequate looking time. However, as previously stated, it would appear to be unlikely that the 12 to 36 month old typically developing controls would require more time to familiarise to stimuli than the 3 month olds in the de Haan et al. study. Therefore an alternative explanation is required.

As stated previously, it may be that the task was not interesting enough for the age groups tested here. In addition to the fact that the infants were, for the most part, mobile, and appeared to dislike remaining seated for the task, the stimuli used may not have been engaging enough for these age groups. Static faces were used in the task, and may not have been interesting enough to capture the infants attention, in the way that moving faces may have been. Although the looking times in the familiarisation trials would appear to contradict this suggestion, as the minimum average looking time was 13.2 seconds, and each familiarisation trial lasted 30

seconds, the results presented do not reveal the occasions when infants looked for less than 10 seconds (when further time was allowed in order to reach this criterion). Several infants in all the groups required extra familiarisation time, which might imply that there was a lack of interest in the stimuli.

It must be acknowledged that the results from the study reported in this chapter are much less clear and informative than the results presented in the previous chapters. It is apparent that the task was not appropriate for any of the groups tested. Failure to demonstrate 'typical' performances in the control groups is problematic, and at one level would seem to justify removal of this study from the thesis. However, this chapter was retained for two reasons. First, it highlights the importance of good task design, particularly in the area of developmental research, where it is vital to ensure the task is age-appropriate. Second, as will be reported in chapter 7, an examination of the relationships between all tasks, and how they contribute to the structure of the groups, revealed an interesting finding in relation to one of the measures reported in this chapter.

7

GROUP STRUCTURE AND COHERENCE ACROSS DOMAINS

7.1 Introduction

The results of the experimental studies reported in this thesis suggest that infants with Williams syndrome and Down's syndrome differ from each other, and from typically developing control groups on a number of measures. Furthermore, the pattern of results suggests that infants with Williams and Down's syndromes do not necessarily present with the same cognitive profile as has been reported in adults. While this pattern of results goes some way to countering the adult neuropsychological model, as discussed in the introduction and which will be considered in detail in chapter 8, it is a valuable exercise to explore these results further here, in order to try to unravel the underlying developmental processes in WS and DS.

Given the paucity of studies which have researched psychological functions in Williams syndrome in infancy or early childhood, the collection of results presented in this thesis facilitates a unique examination of developmental processes in this rare syndrome at an early age. The purpose of this chapter is to take a more global perspective on cognitive functioning in Williams syndrome, by examining the relationships between measures reported from varying tasks, and how well this pattern of performance characterises specific groups. Therefore the focus in this section is on WS, although implications may be extrapolated to DS.

This exercise will also allow further examination of the proposal that there may be subgroups within the Williams syndrome population. The tendency to characterise syndromes as homogeneous groups has dominated research in developmental disorders. However, more recently, research is increasingly focusing on within-syndrome variability. Subgroups have been proposed and examined in several developmental disorders and syndromes, within a number of domains. The study of autism has generated several types of sub-groups, on the basis of social and cognitive skills (e.g. Prior, Eisenmajer, Leekam, Wing, Gould, Ong, and Dow, 1998). There are also thought to be sub-groups in Specific Language Impairment (Bishop, 1997). As mentioned in chapter 4, studies of attention in Down's syndrome have indicated possible sub-groups in this population (Green, Dennis, and Bennets, 1989). Often sub-groups within a syndrome are characterised in terms of comorbidity with other disorders, such as attention deficit hyperactivity disorder, and clearly a thorough clinical assessment will be of benefit in determining an individual child's psychological profile.

7.1.1 Sub-groups in Williams Syndrome

The suggestion that there may be sub-groups in the Williams syndrome population is based on the results from the study of spatial frames of reference, which was reported in chapter 3. The measure of interest was the ratio of egocentric saccades to all other saccades. Although there was no significant difference between the four groups examined on this measure, this comparison did reveal more variability within the Williams syndrome group than in the Down's syndrome, chronological age-matched controls, or mental age-matched controls. Two Williams syndrome sub-groups were generated from this finding: a high ratio and a low ratio group. Results from comparisons with other measures on this task indicate that there may be one group

which has a specific egocentric processing impairment, while the other group seems to have a more general target locating problem.

Several of the studies reported here have led to the emergence of a recurring theme concerning the interpretation of results, in that some findings could be influenced by an attention disengagement deficit in Williams syndrome. In the experiment on spatial frames of reference, it was proposed that such an attention disengagement deficit might account for fewer saccades made by the WS group. Similarly, the pattern of average duration of sustained attention in the WS group could be due to a failure to disengage from the stimuli. In a recent study, Pani, Mervis and Robinson (1999) have suggested that visuo constructive deficits in Williams syndrome may be due to a failure to disengage from a processing style once adopted. Thus a faulty disengagement mechanism could represent a defining characteristic of Williams syndrome. This would be an interesting area for future research. However, it may also prove fruitful to examine the impact of the attentional measures reported here, in terms of how well they characterise Williams syndrome.

It would be of interest to make direct comparisons across all the tasks reported in this thesis, in order to determine whether the proposed Williams syndrome sub-groups differed on any other measures, particularly in view of the finding that there was no difference between groups on scores on the Bayley Scales of Infant Development II. However, the fact that several infants did not complete all the tasks means that although there is a fair degree of overlap in terms of the individual participants for each task, when the data set as a whole is considered, there is a large quantity of missing values. Given that each of the Williams syndrome sub-groups (as proposed in chapter 3) consisted of six participants, this exercise would have resulted in

unfeasibly small groups. One solution to the issue of missing data is to substitute mean within group values for each measure. This will be covered in more detail in the method section.

7.1.2 Discriminant Function Analysis

In order to examine group structure and group performance, discriminant function analyses will be performed on the data set from previously reported tasks. It must be stated from the outset that this exercise is largely exploratory in nature. Although it offers the opportunity to test the hypothesis regarding subgroups in Williams syndrome, due to small and unequal sample sizes, and the volume of missing data, the robustness of any results cannot be assured. It will, however, prove valuable in generating hypotheses for future testing.

The purpose of discriminant function analysis in this set of studies is twofold. First, it allows predictions to be made regarding group membership, based on equations derived from discriminant functions, in order to determine how well the tasks classified the participants. Thus the extent to which the set of tasks administered act as a model for correct group identification can be established. In effect, it should be possible to determine how well the tasks characterise Williams syndrome as a group. Second, discriminant function analysis can also be used to identify those measures which are the best predictors of group membership, i.e. those variables which best distinguish between groups. By carrying out this type of procedure, it is possible to try to interpret the pattern of group differences found in the individual tasks.

7.2. Method

7.2.1 Participants

54.5% of all participants contributed scores on all measures. Of the 14 WS infants, two failed to provide five scores, all from the spatial frames of reference task, two failed to provide two scores from the IBQ, one infants failed to provide a score of number of periods of sustained attention, and one failed to provide a score for the looking time during familiarisation from the face processing task. In the DS group, 7 infants failed to provide scores from the IBQ, and 4 failed to provide scores for looking time during familiarisation from the face processing task. Eight of the CA group failed to provide scores for the IBQ measures, and one did not have a score for looking time during familiarisation trials. Finally, two of the MA infants did not have IBQ scores, one did not have any of the spatial frames of reference scores, and one did not have a looking time score from the face processing study. In each instance, missing values were calculated as within group mean values, and assigned to individuals without a score, for each measure entered into the analysis.

7.2.2 Procedure

A stepwise discriminant function analysis was used, in order to determine variables affecting each group. However, there is a danger that order of entry may be affected by relationships between variables which do not affect population differences, therefore a more liberal probability to enter criterion (.20) was adopted (Costanza and Affifi, 1979, in Tabachnick and Fidell, 1996).

As the number of variables entered into the analysis must be less than the sample size of the smallest group, the data set of all the task variables was reduced. This was done first by selecting only those variables which had produced a difference between

groups in the individual tasks. This reduced set was then entered into a preliminary stepwise discriminant function analysis, in order to determine the variables which would meet statistical criteria for entry into the analysis.

For the analysis examining sub-groups in Williams syndrome, the smallest group size was six, thus allowing a maximum of five variables to be entered into the analysis. Variables were selected as above, and on the basis of variables which had proved to be of interest in the first analysis with four groups. The egocentric to all second looks ratio, the measure on which the sub-groups were formed, was retained as a variable to be entered into the analysis, as it is of interest to determine how well this variable does distinguish the two WS sub-groups.

7.3 Results

7.3.1 Discriminant Function Analysis with Four Groups.

A stepwise discriminant function analysis was performed, using 11 task variables as predictors of membership in four groups. These variables were raw scores from the Bayley Scales of Infant Development II, five measures from the spatial frames of reference experiment (Looks to Target 1, Looks to Centre, Looks to Egocentric position, No Looks in second saccade, and Egocentric to All Second Looks Ratio), number of periods of sustained attention from the sustained attention study, three measures from the temperament questionnaire (Activity, Orienting, and Distress and latency to sudden or novel stimuli), and total looking time during familiarisation trials in the face processing task. The groups were Williams syndrome, Down's syndrome, chronological age-matched controls, and mental age-matched controls.

Table 7.1 Pooled within-groups correlations between discriminating variables and canonical discriminant functions (ordered by size of correlation within function)

Variable	Function 1	Function 2	Function 3
Bayley Raw Scores	**.936***	-.104	-.091
Looks to Centre	-.142	**-.548***	.543
Looks to Egocentric Position	.160	**.508***	-.332
No Second Looks	-.123	**-.441***	.163
Distress to Novel Stimuli	.097	**.413***	.285
Looks to Target 1	*.018*	*.378**	*-.189*
Ego to All 2nd Looks	*.101*	*.280**	*-.237*
Activity Level	*-.025*	*.213**	*.026*
Looking Time	.141	.255	**.411***
Orienting	.194	.094	**.385***
Number of periods of Sustained Attention	*.020*	*.058*	*-.223**

* denotes largest absolute correlation between each variable and any discriminant function. Figures in bold represent eligible correlations from functions with a significant association between groups and predictors. Figures in italics represent variables not used in the analysis.

Three canonical discriminant functions were computed, with a combined chi-square (21) = 201.452, $p < 0.001$. Significant association remained between groups and predictors after removal of the first and second functions (After removal of first function, chi-square (12) = 94.902, $p < 0.001$; after removal of second function, chi-square (5) = 39.736, $p < 0.001$). The three discriminant functions accounted for 67%, 20%, and 13% of between group variability respectively.

Correlations between predictors and discriminant functions are presented in Table 7.1. Loadings less than 0.33 are not considered eligible to be interpreted (Tabachnick and Fiddell, 1996). The variable with the highest correlation, and the only significant correlation with the first discriminant function, is Bayley raw scores.

Examination of group centroids on the first two discriminant functions (see Figure 7.1) reveals that the first function, along the x-axis, provides the best separation of the CA group from the WS, DS, and MA groups. Thus the best predictor for distinguishing the CA from all other groups is performance on the Bayley. This is not an unexpected finding, as higher level Bayley item sets were administered to the CA group compared to the other three groups. The second function, on the y-axis, is a largely a measure of variables from the spatial frames of reference study, namely looks to the centre, and egocentric positions, and failure to make a second look, but is also contributed to by the fear measure from the IBQ. Examination of group centroids reveals that this second function does seem to discriminate the WS group from other groups, but that the separation achieved is not great. The third function is a measure of orienting, and looking time on familiariasation trails time, but only accounts for 13% of between group variability.

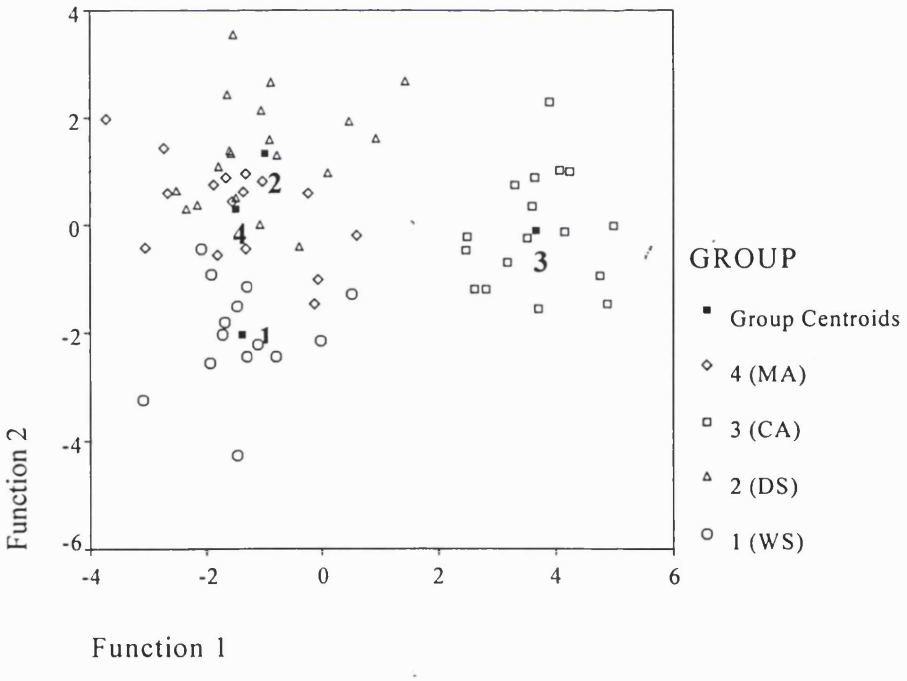

Figure 7.1 Group centroids on the first two discriminant functions computed from data from 11 task variables as predictors of membership in four groups, WS, DS, CA, and MA.

As the first discriminant function was largely a measure of Bayley raw scores, which served to distinguish the CA group from other groups, it was felt that including this measure did not prove helpful in investigating the WS group. This factor accounted for 67% of the variance between groups, and may therefore have been masking relationships between more interesting variables and their ability to predict group membership. Therefore a further analysis was performed, in which Bayley raw scores were excluded.

7.3.2 Discriminant Function Analysis with Four Groups, excluding Bayley scores.

A second discriminant function analysis was performed, using the same task variables as previously, with the exclusion of Bayley raw scores. In addition, one extra variable could be entered, due to the omission of Bayley scores, so total duration of sustained attention was added. This analysis revealed three discriminant functions, with a combined chi-square (21) = 116.951, $p < 0.001$. After removal of the first function, there was still a strong association between groups and predictors (chi-square (12) = 50.457, $p < 0.001$), but this was not the case after removal of the second function (chi-square (5) = 9.715, n.s.). The amount of between-group variability accounted for by the two significant discriminant functions was 64%and 31% respectively.

Figure 7.2 presents the centroids of the four groups on the first two discriminant functions. The first function, along the x-axis, provides the best separation of the WS group from the DS, CA, and MA groups. The second function, on the y-axis, discriminates the MA group from other groups.

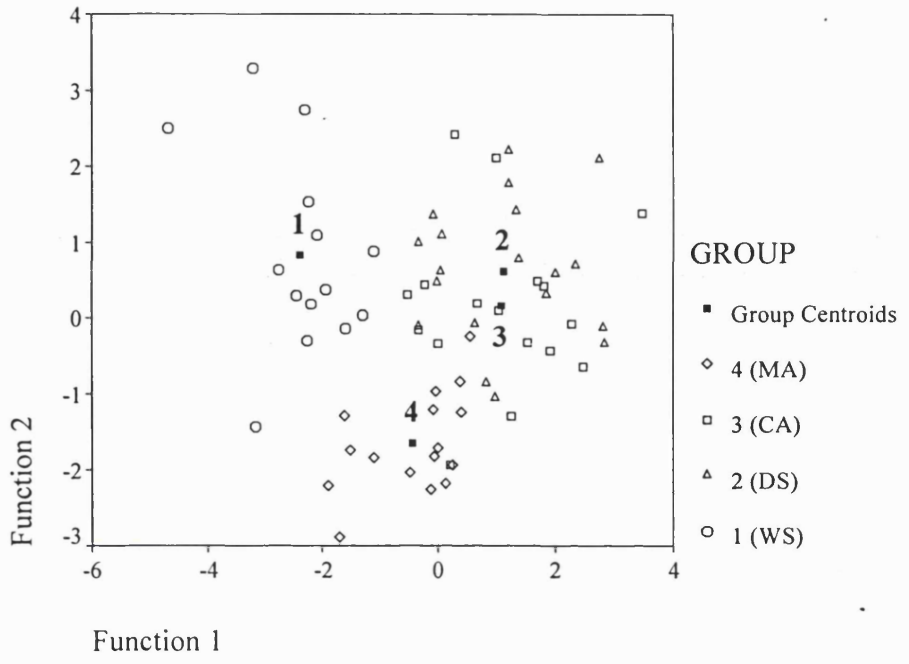

Figure 7.2 Group centroids on the first two discriminant functions computed from data from 11 task variables (excluding Bayley raw score) as predictors of membership in four groups, WS, DS, CA, and MA.

Correlations between predictors and discriminant functions are presented in Table 7.2. The variable with the highest correlation with the first discriminant function is looks to the egocentric position, from the spatial frames of reference study. Almost as high are correlations with failure to make a second look, also from the frames of reference study, and distress and latency to novel or sudden stimuli, the fear factor from the Infant Behavior Questionnaire. Thus the best predictors for distinguishing between the WS, and all other groups, are second saccade response types and a measure of fear. The second discriminant function, which largely serves to distinguish the MA group from other groups, correlates with looks to the centre, and looking time during familiarisation in the face processing study. It should be noted that looks to the centre also has a high loading on the first discriminant function, and is therefore also contributing to distinguishing the WS group from others.

Table 7.2 Pooled within-groups correlations between discriminating variables and canonical discriminant functions (ordered by size of correlation within function).

Variable	Function 1	Function 2	Function 3
Egocentric Looks	.478*	-.393	.173
No 2nd Look	-.411*	.228	-.007
Distress to Novel Stimuli	.399*	.195	-.321
Target 1	.293*	-.245	.137
Activity Level	.219*	-.020	-.216
Looks to Centre	-.478	.610*	-.155
Looking time	.329	.372*	.030
Ego to All 2nd Looks	.255	-.267*	.137
Sustained Attention Duration	-.199	-.051	.746*
Orienting	.253	.409	.512*
Number of periods of Sustained Attention	-.113	-.216	.464*

* denotes largest absolute correlation between each variable and any discriminant function. Figures in bold represent eligible correlations from functions with a significant association between groups and predictors. Figures in italics represent variables not used in the analysis.

It should be noted that the DS and CA groups have very similar group centroids (see Figure 7.2) in terms of the first and second discriminant functions, which suggests that they are not distinguishable from each other on the basis of the predictors included in this analysis. Given that these groups did not differ from each other on most of the variables entered into this analysis, this is not an unexpected result.

The results of group classifications are presented in Table 7.3. This allows comparison of original group membership with that predicted by the discriminant functions. Overall, 83.3% (55 individuals) of the original grouped cases were classified correctly, compared to 25% (16.7 individuals) who would be correctly

classified by chance alone (chance is calculated by multiplying each group number by prior probability, and summing all products). 100% of the WS group were correctly classified, and no members of other groups were predicted as members of the WS group. Nearly 70% (68.4%) of the DS group were correctly classified, with almost one third of the group incorrectly classified as CA. 82.4% of CA infants, and 87.5% of MA infants were correctly classified. Lower accuracy of classification for the DS, and to some extent the CA participants, reflects the pattern revealed in examination of group centroids. As stated previously, this finding is attributable to the variables entered into the analysis, which were measures on which the WS infants differed most. The overall pattern of results indicates that the discriminant functions identified are more likely to correctly classify WS infants than infants from any other group.

Table 7.3 Group classification results (percentages in brackets)

Group		Predicted Group Membership				Total
		WS	DS	CA	MA	
Original	WS	**14 (100)**	0	0	0	14 (100)
	DS	0	**13 (68.4)**	6 (31.6)	0	19 (100)
	CA	0	2 (11.8)	**14 (82.4)**	1 (5.9)	17 (100)
	MA	0	1 (6.3)	1 (6.3)	**14 (87.5)**	16 (100)
Cross-validated	WS	**10 (71.4)**	1 (7.1)	1 (7.1)	2 (14.3)	14 (100)
	DS	0	**12 (63.2)**	6 (31.6)	1 (5.3)	19 (100)
	CA	0	7 (41.2)	**7 (41.2)**	3 (17.6)	17 (100)
	MA	0	2 (12.5)	2 (12.5)	**12 (75)**	16 (100)

Cross-validation is performed in discriminant function analysis to provide a measure of reliability and stability of the classification procedure. In cross-validation, information about original group membership is withheld, and each case is classified by the functions derived from all cases other than that case. In other words, each

participant is classified based on information from all other participants, but excluding information about group membership. The results of cross-validation are included in Table 7.3. In total, 62.1% of the cross-validated groups were correctly classified, with WS and MA groups both having high correct classification percentages (71.4% and 75% respectively). Although classification has decreased for the cross-validation sample, a reduction in correct classification is usually the case in cross-validation (Tabachnick and Fiddell, 1996), and the figure reported does indicate a good degree of consistency in the classification scheme.

7.3.3 Discriminant Function Analysis with Five Groups.

In order to examine the structure and validity of the proposed sub-groups in Williams syndrome, a second stepwise discriminant function analysis was performed, using five task variables as predictors of membership in five groups. Variable selection is as outlined in the procedure. The variables entered into the analysis were two measures from the spatial frames of reference experiment (Looks to Centre, and Egocentric to All Second Looks Ratio), two measures from the temperament questionnaire (Orienting, and Distress and latency to sudden or novel stimuli), and total looking time during familiarisation trials in the face processing task. The groups were Williams syndrome high egocentric ratio, Williams syndrome low egocentric ratio, Down's syndrome, chronological age-matched controls, and mental age-matched controls.

Four discriminant functions were calculated, with a combined chi-square (20) = 118.219, $p < 0.001$. After removal of the first function, there was still a strong association between groups and predictors (chi-square (12) = 60.277, $p < 0.001$). This association was also significant after removal of the second function (chi-square (6) =

25.991, $p < 0.001$) but this was not the case after removal of the third function (chi-square (2) = 3.819, n.s.). The amount of between-group variability accounted for by the three significant discriminant functions was 56%, 26%, and 15% respectively.

Correlations between predictors and discriminant functions are presented in Table 7.4. The variable with the highest loading on the first discriminant function is looking time during familiarisation trials from the face processing study.

Table 7.4 Pooled within-groups correlations between discriminating variables and canonical discriminant functions (ordered by size of correlation within function)

Variable	Function 1	Function 2	Function 3	Function 4
Looking time	**.453***	.230	-.105	.173
Looks to Centre	-.255	**.781***	.556	-.053
Ego to All 2nd Looks	.418	**-.643***	.598	-.179
Orienting	.397	.282	-.310	-.687*
Distress to Novel Stimuli	.414	.074	-.213	.681*

* denotes largest absolute correlation between each variable and any discriminant function. Figures in bold represent largest eligible correlations from functions with a significant association between groups and predictors.

Examination of the group centroids on the first two discriminant functions in Figure 7.3 reveals that although the first function is the best predictor for distinguishing the low egocentric ratio WS group from other groups, the distances between groups are actually quite small. The greatest separation occurs between the WS low ratio group and the DS group, with all other groups in between. However, this function does appear to have achieved a separation between the two WS groups. Looks to the centre has a high correlation with the second discriminant function, and the ego to all second

looks ratio has a high negative correlation with this function. Inspection of the group centroids again appears to reveal that this function best separates the low ratio WS group from other groups, although the separation between low and high ratio WS groups is less than for the first function. The third function identified also appears to be largely a measure of looks to the centre and the ego to all second looks ratio, and as such does not have loadings with any variables which are the highest for that variable.

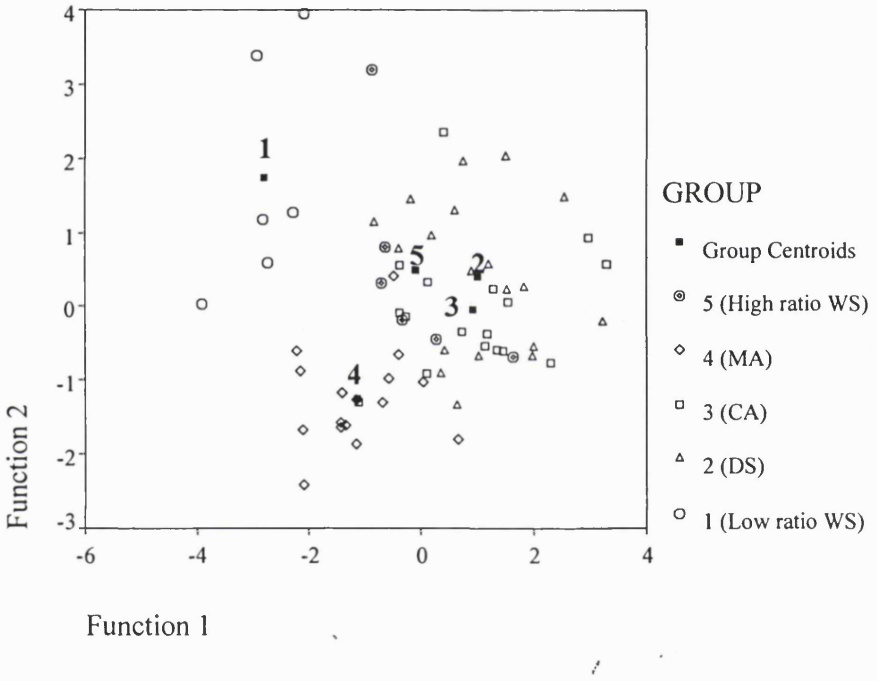

Figure 7.3 Group centroids on the first two discriminant functions computed from data from 5 task variables as predictors of membership in 5 groups, WS low egocentric saccades ratio, WS high egocentric saccades ratio, DS, CA, and MA.

As in the previous analysis with four groups, there is little separation achieved between the DS and CA groups, but again this is because there is little difference between these groups on the variables entered into the analysis. What is interesting to note is the fact that when plotted against the first two discriminant functions (see

Figure 7.3), the high ratio WS group centroid appears to be in very close proximity to the CA and DS groups, suggesting that the scores of the WS high ratio group on the variables in this analysis are somewhat similar.

The results of group classifications are presented in Table 7.5. Overall, 71.9% (47 individuals) of the original grouped cases were classified correctly, compared to 15% (10 individuals) who would be correctly classified by chance alone. 100% of the low ratio WS group and 66.7% of the high ratio WS group were correctly classified.

57.8% of the cross-validated groups were correctly classified, with both WS groups, and the MA group having high correct classification percentages (66.7% for each WS group, 81.3% for the MA group). Incorrectly classified WS high ratio group members are classified as DS, while incorrectly classified WS low ratio group members are classified as belonging to the MA group. DS and CA group members are equally likely to be incorrectly classified as CA or DS group members (respectively), as they are to be classified in their original group.

Table 7.5 Group classification results (percentages in brackets)

Group		Predicted Group Membership					Total
		WS low	WS high	DS	CA	MA	
Original	WS low	**6 (100%)**	0	0	0	0	6 (100%)
	WS high	0	**4 (66.7%)**	2 (33.3%)	0	0	6 (100%)
	DS	0	1 (5.3%)	**13 (68.4%)**	5 (26.3%)	0	19 (100%)
	CA	0	0	7 (41.2%)	**9 (52.9%)**	1 (5.9%)	17 (100%)
	MA	0	0	1 (6.3%)	1 (6.3%)	**14 (87.5%)**	16 (100%)
Cross-validated	WS low	**4 (66.7%)**	0	0	0	2 (33.3%)	6 (100%)
	WS high	0	**4 (66.7%)**	2 (33.3%)	0	0	6 (100%)
	DS	0	2 (10.5%)	**9 (47.4%)**	8 (42.1%)	0	19 (100%)
	CA	0	0	8 (47.1%)	**7 (41.2%)**	2 (11.8%)	17 (100%)
	MA	0	0	1 (6.3%)	2 (12.5%)	**13 (81.3%)**	16 (100%)

7.4 Discussion

The purpose of this chapter was to explore the results of the previously reported studies as a set of predictors for group membership. This analysis also facilitates examination of group structure and coherence, which in this case allowed examination of the proposal that there are sub-groups in Williams syndrome. The results demonstrate that the variables entered into the analysis achieved good group classification for Williams syndrome when four groups were predicted, and also offered some support for the existence of distinctive sub-groups in Williams syndrome when five groups were predicted. However, as stated in the introduction, for methodological reasons, these results cannot be considered to be very robust, and are therefore interpreted with caution.

Initial analysis revealed a main discriminant function, which accounted for 67% of between group variability, was largely a measure of mental age (Bayley raw scores), and served to distinguish the CA group from other groups. As this factor accounted for a large proportion of between group variability, and may have concealed factors more important in contributing to WS group membership, a second analysis was performed, from which Bayley raw scores were omitted. This second discriminant function analysis, with four groups, resulted in two significant functions. The first function, which was largely a measure of second saccade response types from the spatial frames of reference task, and a measure of fear from the Infant Behavior Questionnaire accounted for 64% of between group variability, and was the best predictor for distinguishing between the WS and all other groups. Although the measure of looks to the centre during the first saccade in the spatial frames of reference task was most highly correlated with the second function, it also had a high loading on the first function, and therefore can be interpreted as contributing to the

factors separating the WS group from other groups. The second function, which was mainly a measure of looks to the centre, was also contributed to by the amount of looking time during familiarisation on the face processing task, and this function was the best predictor for distinguishing the MA group from all other groups, accounting for 31% of between group variability.

Group classification results from this first analysis revealed that classification rates were much higher than would have been predicted by chance, and the highest classification rates were reported for the Williams syndrome group. Both the original classification rates and results reported from cross-validation revealed that infants in the DS and CA groups were less successfully distinguished by the variables entered into the analysis. As stated, this was because scores for the CA and DS groups on these measures were not dissimilar.

In summary, the discriminant functions generated in this analysis suggest that when the tasks reported in this thesis are considered as a set, the variables which best characterise Williams syndrome as performing differently from the other groups, are looks to the egocentric position, failure to make a second look, and looks to the central position, from the spatial frames of reference task. The fear measure, distress and latency to sudden or novel stimuli, also appears to play a significant role in characterising Williams syndrome. However, it appears that the measures that best classify the WS group from all other groups are largely visuo-spatial.

In considering comparisons between the variables which have been identified as best characterising Williams syndrome in infancy, and the cognitive profile in the adult Williams syndrome phenotype, performance on the spatial frames of reference task

could equate to visuo-spatial abilities, which are reported as poor in adults. However, the fear measure from the Infant Behavior Questionnaire, which also contributed to the best predictors for the Williams syndrome group, is less easily interpreted. As reported in chapter 5, the Williams syndrome group had the lowest score on this measure, and were significantly lower than the DS group, which had the highest score. This would seem to indicate that low fear is a predictor of Williams syndrome. However, adults with Williams syndrome are frequently reported as being highly anxious and fearful. Therefore it would appear that there is a mismatch between the infant and adult phenotype, which can be related to the issue of comparing developmental disorders and adult neuropsychological models. This issue will be addressed more fully in chapter 8.

It is also interesting to consider those variables which do not appear to have been identified as predictors of Williams syndrome. None of the measures of attention reported in this thesis were significantly correlated with either of the main discriminant functions, even though the WS group differed from some of the control groups on several of these measures. However, the results for the WS group from the sustained attention study, and the attention measures from the Infant Behaviour Questionnaire are less distinctive than the predictor measures outlined above. In these studies the DS group tends to be identified as performing differently from the other three groups. Therefore it is not surprising that these measures are not identified as predictors for the Williams syndrome group.

A discriminant function analysis was also performed with five groups, in order to examine the proposal that there are two sub-groups in the Williams syndrome population. As stated in the introduction, this suggestion was based on the variability

within the Williams syndrome group on the ratio of egocentric to all second saccades, from the spatial frames of reference task. The purpose of the discriminant function analysis was to determine how well this measure acted to separate the two WS groups, whether any other variables were reliable predictors of these sub-groups, and how valid these sub-groups actually were.

The first discriminant function, which accounted for 56% of between-group variability, was the best predictor for distinguishing between the low ratio WS group and the high ratio WS group. The variable with the highest correlation with this function was looking time during familiarisation trials on the face processing task. In addition, although the egocentric ratio, and the orienting and fear measures from the Infant Behaviour Questionnaire, all had higher correlations with other functions, they also loaded onto this first function. The second function was largely a measure of looks to the centre, and the egocentric ratio, both from the spatial frames of reference task. This function accounted for 26% of between-group variability, and best distinguished the WS low ratio group from the MA group.

The results from group classification and cross-validation indicate good consistency in the classification scheme, and appear to support the classification of two Williams syndrome sub-groups, from the variables entered into the analysis. Lack of discrimination between the DS and the CA groups, as found in the first analysis, is also apparent here. Examination of the group centroids on canonical discriminant functions reveals that the high ratio WS group are more similar to the DS and CA groups, than to the WS low ratio group. Comparison of the classification rates for the original WS group, and the two WS sub-groups, reveals that while the original WS group, and the low-ratio WS subgroup, had 100% correct classification, a somewhat

lower figure (66.7%) was reported for the WS high ratio subgroup. A third of this latter group were misclassified as belonging to the DS group, which is consistent with the pattern revealed on examination of group centroids.

Although the two Williams syndrome sub-groups were originally formed on the basis of egocentric ratio from the spatial frames of reference study, it should be noted that the measure which best distinguished the WS low ratio group from the WS high ratio group was looking time during familiarisation trials on the face processing task. Although the egocentric ratio was also identified as a predictor, this emerged on the second discriminant function, which accounted for less between group variability. The looking time data from the face processing study show that the WS group spent less time looking at the faces during familiarisation trials than the DS and CA groups. Unlike the egocentric ratio data from the spatial frames of reference study, the WS group do not show greater variability in terms of their looking time during familiarisation data, therefore it is less easy to interpret this finding. It is interesting to note that the measure of looking time from the faces study had a significant negative correlation with the amount of time spent looking at the prototypical face. As the results from the face processing study are largely inconclusive, it is problematic to tease apart the nature of the relationships which seem to be indicated here. However, it may be that this pattern of results is indicative of some face specific attention mechanism. This is clearly an area which requires further investigation.

To date, little research has been carried out on familiarisation time in Williams syndrome, although it might be compared to speed of processing in adults. Preliminary studies suggest that speed of processing in adults with WS is much slower, even when compared with adults with Down's syndrome (Anderson,

Hatzakis, Grant, and Karmiloff-Smith, unpublished manuscript). If this difference in processing time is robust, this could have implications in terms of identification and intervention strategies. It would also be of interest to investigate speed of processing in relation to the attention disengagement deficit, hypothesised elsewhere in this thesis.

The fear measure found to be a predictor for the two Williams syndrome groups was also identified as a predictor in the first analysis, for the WS group as a whole. However, the orienting measure, which has been equated to a measure of attention (see chapter 5), was not identified as a predictor for the original Williams syndrome group, but does appear to play a role in distinguishing between the two WS sub-groups. If this finding is robust, then this would imply that there are attentional differences between these groups.

In addition to exploring the relationship between the measures of visuo-spatial cognition reported here, it would also be of interest to combine these measures from other domains, such as language ability, in a similar type of analysis. This would allow a more broad-based comparison of reported areas of relative strengths and weakness, which would assist in the attempt to obtain a more global representation of cognitive abilities in Williams syndrome.

In summary, discriminant function analysis adds some support to the proposal that there are distinct sub-groups within the Williams syndrome population, and furthermore, that these sub-groups differ not only on the egocentric ratio, from which the groups were formed, but also in terms of speed of processing, and attention. However, as stated at the beginning of the discussion, these findings should be treated

as preliminary. Therefore further work on these types of measures would be required to validate these findings.

8

SUMMARY AND CONCLUSIONS

8.1 Introduction

This thesis has attempted to establish the abilities of infants and young children with Williams and Down's syndromes, in a number of areas within the domain of visual cognition. The rationale behind the set of studies reported here was fourfold. First, while Williams syndrome has attracted a great deal of research recently, most of the work previously carried out has focused on older children and adults. This work has established a now well-documented uneven cognitive profile in Williams syndrome, with relative strengths in language and face processing, while visuo-spatial skills are significantly impaired. However, there is little indication whether this profile is present from infancy. Therefore, it is important to establish cognitive functioning at an early age, to add to the body of knowledge on Williams syndrome, and how cognitive impairment develops over time. Second, while cognitive functioning in infants and young children with Down's syndrome is relatively well documented, a number of the tasks reported here have not previously been tested on infants with Down's syndrome, and are therefore of interest in adding to knowledge about this population. Third, comparisons between syndrome groups in infancy, as well as with adult phenotypes in both syndromes, will begin to shed light on the trajectory which influences development in these groups. Although adult groups were not tested in this thesis, comparisons made will be based on abilities within syndromes reported in the literature. Finally, this comparison also allows for the evaluation of two contrasting

approaches to the study of developmental disorders: the static, adult neuropsychological approach, and the developmental cognitive neuroconstructivist approach. It is proposed here that the adult neuropsychological approach to modelling developmental disorders is inappropriate, and that a more dynamic developmental approach is crucial to begin to unravel the developmental processes which not only govern atypical development, but which also take place in normal development. By adopting a broad approach examining performance across domains, it is possible to compare a range of abilities, and how they relate to one another, rather than examining domains in isolation.

In this chapter, the results from each of the studies reported in this thesis will be reviewed, both individually, as stand-alone studies of specific abilities, and collectively, in an attempt to unravel the overall pattern of behaviours seen in infants and young children with Williams and Down's syndromes. Comparisons between the findings from these infant studies and reported findings in the adult phenotype will also be considered. The two contrasting approaches to developmental disorders, as outlined above, and in more detail in the main introduction, will be assessed in view of the pattern of results found from the experimental studies reported here. Finally, implications for the populations studied, and directions for future work will be considered.

8.2 Summary of Task Results and Implications

Infants with Williams and Down's syndromes, and chronological and mental age-matched controls were tested on a range of tasks in the domain of visual cognition, and on two more general tasks. In chapter 2, results from administration of the Bayley Scales of Infant Development II were reported. This scale was used as a matching

tool, firstly to establish equivalence in terms of general level of development across the WS, DS, and MA groups, and secondly to ensure participants in the CA and MA groups were performing within normal limits. Analysis of the data also confirmed reliability between different testers, and different testing locations. It was hypothesised that the WS group might show a deficit on performance with visuo-spatial items, as visuo-spatial processing is impaired in adults with Williams syndrome. A subset of visuo-spatial items was identified by experienced administrators of the Bayley, and performance on this item set was compared across groups, but no difference was found between groups. Finally, it was acknowledged that there are several methodological and theoretical problems which must be considered when administering the Bayley with atypically developing groups.

In chapter 3, groups were tested on a measure of saccade planning. A double-step saccade paradigm allowed investigation of the spatial frames of reference used when planning eye movements. It was hypothesised that infants in the WS group would be impaired on this measure, as adults with WS have a deficit in visuo-spatial processing. To a certain extent, the results supported this hypothesis. The WS group differed from other groups on both first and second saccade measures. Although the ratio of body-centred looks between the WS group and all other groups failed to reach significance, there was some indication that a subgroup of the WS infants are impaired in using body-centred spatial frames of reference for saccades.

It is important to note that the DS group did not differ from the two typically developing control groups on any of the measures in the spatial processing task, therefore any differences for the WS group cannot be ascribed to general effects of developmental delay. The results appear to indicate that overall, infants with William

syndrome show a greater reliance on sub-cortical processing mechanisms. However, the existence of sub-groups within WS suggest that the outcome of this atypical processing style is variable; one group appears to exhibit a specific deficit using egocentric spatial frames of reference, while for the other group, the deficit is a more general location finding problem.

The proposal that there are distinct sub-groups within Williams syndrome, in terms of visuo-spatial processing skills, is intriguing, but not without precedent in developmental disorders, as similar suggestions have been made for other populations, such as the Down's syndrome population (e.g. Green, Dennis, and Bennets, 1989). While it is acknowledged that more work is required in order to investigate this claim further, this subdivision in WS highlights the need to study developmental processes within groups. In the same way that perhaps it is inappropriate to infer infant abilities from those seen in adults, it should not be assumed that the same, or similar starting state will necessarily result in the same outcome, even as early as a few years after birth.

The role of attention in developmental disorders was investigated in chapters 4 and 5. Problems in attention have been documented in Down's syndrome, but although anecdotal reports suggest attention may also be impaired in Williams syndrome, little empirical work on attention has been carried out with this population. The study of sustained attention revealed that the DS and the WS groups performed very differently on a number of measures, with the DS group exhibiting poorer attention in terms of both duration and number of periods of sustained attention. While the WS group did not differ statistically from the typically developing control groups, it was suggested that they may be demonstrating longer duration of attention. Results from

the Infant Behavior Questionnaire revealed that on duration of orienting, an attentional measure, the DS group performance was similar to that found in the CA group, and both scored higher than the MA group, while the WS group did not differ from any of the other groups. On the fear measure, distress to novel and sudden stimuli, the DS group scored higher than the WS group.

On first examination, the two studies of attention appear to present conflicting results; the sustained attention study implies better attention in infants with WS, while the IBQ reports better attention in infants with DS. However, it is suggested that attention measures in these tasks may be tapping different aspects of attention. The orienting measure from the IBQ does not necessarily represent intense processing which takes place during sustained attention, but may be a representation of casual attention, which is better defined by simple looking behaviour, rather than the complex pattern of behaviours thought to be indicative of sustained attention.

Face processing, reported to be a relative strength in Williams syndrome, was also investigated, and a study of face processing was reported in chapter 6. This study was based on the proposal that one of the mechanisms used during face processing is the abstraction of a prototypical face from exemplars of faces encountered. It was hypothesised that infants with WS would not create a face prototype, while all other groups would. Failure to extract prototypes might indicate a tendency for exemplar learning in WS, which could relate to the reported bias for featural rather than configural processing in this population. The results failed to support the hypothesis, and it was suggested that there were several methodological problems with this study. Failure to find reliable novelty preference across groups may suggest that the task was not age appropriate, and that infants did not find the stimuli engaging. This highlights

the problem of designing appropriate tasks, not only for use with atypically developing infants, but also when testing normally developing infants of different age groups. Failure to find novelty preference in this task means that any differences between groups in terms of prototype formation cannot be treated as reliable. However, one interesting finding did emerge from this task. When examined as part of the whole set of studies, the amount of time spent looking at faces in the familiarisation trials was highly predictive of membership of WS sub-groups. Indeed it was the most predictive measure. Analysis of the looking time data showed that the DS group spent significantly longer looking at the faces than the MA group. The WS group looked for slightly longer than the MA group, and less than the CA group. This would seem to indicate that although there is nothing noticeable about the performance of the WS group in relation to all the other groups on this measure, in some way this measure is acting as a descriptor for Williams syndrome. While it cannot be determined whether or not this effect was specific to faces, indications that there may be differences within the WS group in terms of face processing suggest that this is an area worthy of more work in the future, with a greater emphasis on task design.

8.3 Relationships Between Measures and Within Groups

While results from the individual studies reported above are of interest in themselves, it is important to consider how deficits or strengths in a particular area might impact on other areas of development. This requires taking a more global perspective on the overall pattern of results. To this end, three discriminant function analyses were performed, and reported in chapter 7. The purpose of this exercise was to examine how well the tasks reported in this thesis characterised Williams and Down's

syndromes, and to what extent the subgroups proposed from the frames of reference study were predicted from other measures.

This analysis was also performed in order to investigate a proposed effect of attention disengagement in Williams syndrome. Although not directly tested, several of the studies reported here yielded findings that could be indicative of an inability to terminate attention appropriately. Therefore it was of interest to examine in particular the role of measures of attention in identifying the clinical groups. However, it must be acknowledged that this analysis is not a true test of attention disengagement, as the measures entered into the analysis tap other dimensions of attention, namely orienting of attention, and sustained attention.

Overall, the results of the tasks entered into the analyses provided good predictions for group membership for WS infants. DS infants were less clearly defined, due to the fact that most of the measures in the analysis were those in which the DS group largely performed like the typically developing groups. The variables which best characterised Williams syndrome were measures from the spatial frames of reference task, and the fear measure from the IBQ. These findings suggest that visuo-spatial deficits are a major component in characterising Williams syndrome, but also highlight the importance of emotional factors, and their impact on general levels of functioning. This in turn stresses the importance of adopting a broad-based approach to studying development, in order to examine interactions across domains.

A further analysis appeared to provide some support for the distinction between sub-groups in WS, and indicated that the high egocentric ratio WS group resembled the CA and DS groups more than it did the low egocentric WS group. It was also revealed

that the best predictor for distinguishing between WS sub-groups was looking time during familiarisation trials. As stated previously, it cannot be determined if the difference within WS looking times is specific to the face stimuli, but this pattern of results may indicate the presence of face specific attention differences in WS sub-groups. This could relate to the proposed attention disengagement deficit, in that an inability to disengage from stimuli, such as faces, might lead to a tendency to focus on specific details within that stimulus. This type of processing might then go some way towards accounting for the reported preference for componential rather than configural processing in Williams syndrome.

None of the attentional measures appeared to act as reliable predictors of group membership. Thus the issue of a possible attention disengagement deficit in Williams syndrome cannot be addressed by this analysis. However, as stated in chapter 7, infants from the DS group performed more distinctively than other groups on these measures, therefore perhaps it is not surprising that they failed to emerge as good predictors of Williams syndrome. In addition, as stated above, the measures entered into the analysis reflect dimensions of attention other than attention disengagement.

8.4 Comparing Infant and Adult Phenotypes

The results of the tasks presented above reveal that infants and young children with Williams and Down's syndrome perform differently on a number of tasks. Furthermore, when comparisons are made with the adult phenotype in each of these groups (reported elsewhere in the literature on WS and DS), it is apparent that the pattern of abilities in infant groups cannot always be predicted from adult studies. In

this section, the results of infants studies are compared with abilities which are, for the most part, well documented in the adult phenotype.

In terms of visuo-spatial processing, it is apparent that, as in adulthood, infants with Williams and Down's syndromes exhibit different abilities. The results from the study of spatial frames of reference indicate that impairments in visuo-spatial processing in adults with Williams syndrome may have correlates in infancy. Thus on first examination it would appear that inferring infant abilities from adults was successful in this domain. However, the proposal that there are subgroups within the WS infant population suggest that this is not necessarily the case. While it is possible that the infants in each of these subgroups differ on some more basic, as yet undetected measure, it is apparent that assumptions about infant and adult similarity may mask underlying differences, both within groups, and in terms of developmental trajectories. While further work is required to validate the existence of subgroups in the WS infant population, it would also be of interest to determine more closely whether such differences also exist in the adult population.

When comparisons are made between infant and adult phenotypes in relation to attention, once more the findings seem to indicate discrepancies between the starting state and the end state. The incidence of attentional problems, such as ADHD, appears to be greater in adults with WS than in DS. However, infants with WS show better sustained attention than infants with DS, thus the deficit found in adulthood does not appear to be present in infancy. Although the picture is less clear when other measures of attention, such as orienting, are considered, in that DS infant performance was better than WS, it should be noted that WS performance did not differ from

typically developing controls. Again, the trend in the adult phenotype does not reflect the infant status.

Although the main purpose of administering the IBQ was to examine measures of attention in temperament, this tool also revealed a further discrepancy between infant and adult phenotypes. Adults with WS are frequently reported as exhibiting anxieties and fears. This was not found in infants with WS, who had the lowest score on the fear measure of the IBQ.

Comparisons between infant performance on the study of face processing reported here, and face processing in adults with WS and DS are less easily made, due to methodological problems in the infant study. However, given that face processing is an area of reported proficiency in adults with WS, this is an area which warrants further testing in infants with WS, using a more suitable task, in order to make comparisons between infancy and adulthood.

8.5 Evaluating Approaches to Developmental Disorders

Two approaches to studying developmental disorders were contrasted in this thesis. The static, adult neuropsychological approach, tends to make inferences about the infant state based on the pattern of adult outcome, and concludes that the adult pattern of abilities indicate modularity of cognitive processes. The developmental cognitive neuroconstructivist approach claims that infant starting states cannot necessarily be inferred from the adult phenotype, and that apparent modularity in the adult is far less well defined in infancy. The studies reported in this thesis go some way to indicating that the former approach is inappropriate in the study of developmental disorders.

Discrepancies between cognitive functioning in adults and infants with Williams and Down's syndromes indicate that the infant state cannot necessarily be inferred from the adult state. Development is a much more dynamic and interactional process, and modularity of cognitive functions is much less pre-determined than is supposed by this approach. Furthermore, differences within syndromes in infancy, as found in the WS sub-groups, suggest that there may be small differences within phenotypes which can radically affect outcome, and that assumptions about homogeneity even within adult phenotypes may be misleading.

Claims concerning intact and/or innate modules lead to research which diminishes, or even overlooks the many and dynamic processes which occur during the course of development. Only by adopting a more developmental and interactional approach, charting the subtle changes which occur early on and during development, as well as considering the many influences which impact on developmental trajectories, can the processes underlying development be understood. The set of studies reported in this thesis makes some advances towards adopting a truly developmental approach. However, the design is cross-sectional, and in order to map out the developmental trajectory as described above, a longitudinal component would be necessary. This would allow individual changes and developmental pathways to be examined. Although a longitudinal component was originally included within the design for this thesis, practical considerations meant that too few infants were seen on a longitudinal basis, which meant that group comparisons across a longitudinal design were not feasible.

8.6 Conclusions and Future Directions

The studies reported in this thesis indicate that modelling developmental disorders such as Down's and Williams syndromes on adult neuropsychological studies is inappropriate. The importance of studying development from a very early age cannot be overstressed, although in practice this is often problematic. In the case of Williams syndrome, very young infants tend to present with significant medical problems, which makes neonate testing difficult. Furthermore, for parents of children with developmental disorders, there is often a period of coming to terms with a diagnosis, before which testing may be unwelcome or intrusive. However, early diagnosis is becoming more prevalent in WS, with the advent of the genetic test, and as more practitioners become aware of the syndrome. It is therefore hoped that testing of very young infants with WS will be possible in the future. Experimental design which is sympathetic to the medical problems in very young infants with WS will also expedite future research.

The studies reported here also highlight the importance of good task design. This is always particularly challenging when testing infants with developmental disorders, and typically developing control groups of different ages. However, infants with developmental disorders should be viewed as rare resources, and every effort taken to ensure that their contribution is as meaningful as possible.

The results of these studies of infants with Williams and Down's syndromes has raised a number of questions which will hopefully be addressed by future research. Heterogeneity within syndromes, as evidenced by sub-groups in WS, requires further investigation, in both adult and infant phenotypes. Research of this type could be of

benefit not only in terms of directing further research, but also in relation to the design and implementation of intervention strategies.

Results of the studies of attention, as well as those from other studies presented in this thesis, have raised the suggestion that attention disengagement is worthy of investigation in DS and WS. As mentioned previously, attention disengagement deficits have been reported in other developmental disorders, and should be investigated as a possible source of influence on development. If attention is terminated late, orienting to new, possibly crucial stimuli may be affected, thereby reducing the quantity or quality of exposure to environmental influences. If there is a bias in WS towards overly long periods of attention, it would also be of interest to investigate whether this has a relationship with the obsessive behaviours often seen in adults with WS. Failure or inability to shift attention may have some influence on the development of such obsessions. It may also be implicated in hyperacusis in WS, in that sufferers of hyperacusis may be unable to switch off from a sound source. Finally, as stated previously, an attention disengagement deficit may offer an account for the bias toward componential processing in WS. Further work is required to establish if there is such a deficit in WS infants, and if so, whether this is found only in relation to processing faces, or extends to all classes of stimuli.

Processes such as speed of processing should also be further investigated in WS and DS. This would be informative in terms of general processing styles and abilities, and would also be helpful in future studies employing habituation and preferential looking methodologies. Furthermore, studies of information processing in infants with developmental disorders could be informative, in view of recent suggestions that such

measures are better predictors of later cognitive abilities than standardised tests of development.

In conclusion, the studies reported in this thesis represent an emergent approach to developmental research which acknowledges the importance of development itself, and which tries to unravel the processes which take place in development. It is the first study in visual cognition to examine experimentally a large group of WS infants, and to compare them to infants with DS as well as MA and CA typically developing controls. By demonstrating that small differences at the outset may result in radically divergent developmental trajectories, it is hoped that the magnitude of the role played by developmental processes themselves will be acknowledged at all times in future research.

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APPENDIX 1: Bayley Raw Data Scores

Williams Syndrome	
Age	Bayley Raw Score
23 months 16 days	96
24 months 10 days	83
24 months 20 days	94
24 months 22 days	99
25 months 3 days	97
25 months 14 days	88
30 months 2 days	109
30 months 12 days	104
30 months 14 days	103
30 months 17 days	100
30 months 24 days	107
31 months 15 days	105
36 months 27 days	113
37 months 7 days	122

Down's Syndrome	
Age	Bayley Raw Score
23 months 25 days	93
24 months 2 days	95
24 months 5 days	88
24 months 9 days	96
24 months 11 days	94
24 months 13 days	91
24 months 19 days	88
25 months 2 days	100
30 months 0 days	95
30 months 3 days	99
30 months 5 days	102
30 months 9 days	117
30 months 22 days	99
31 months 0 days	99
31 months 14 days	100
35 months 15 days	121
35 months 22 days	116
36 months 15 days	112
36 months 25 days	112

Chronological Age-Matched Controls	
Age	Bayley Raw Scores
23 months 14 days	136
23 months 20 days	139
23 months 25 days	135
24 months 17 days	140
24 months 18 days	138
24 months 25 days	139
30 months 0 days	141
30 months 0 days	145
30 months 1 days	143
30 months 4 days	149
30 months 12 days	149
31 months 23 days	149
35 months 23 days	157
36 months 1 days	149
36 months 11 days	165
36 months 13 days	160
36 months 29 days	161

Mental Age-Matched Controls	
Age	Bayley Raw Scores
11 months 24 days	84
12 months 5 days	87
12 months 6 days	88
12 months 20 days	88
14 months 1 days	96
14 months 4 days	95
14 months 4 days	100
14 months 23 days	95
15 months 11 days	101
15 months 12 days	98
15 months 25 days	98
16 months 5 days	102
18 months 2 days	113
18 months 4 days	110
18 months 5 days	113
20 months 15 days	123

APPENDIX 2: Sustained Attention Scoring System, reproduced from material supplied by Holly A. Ruff

SUSTAINED ATTENTION CODING

When scoring, it is important to keep all of the following in mind:

* The coder will need a stopwatch, with a stop/reset function in order to determine the examining time for each trial.
* After each trial, pause the tape and record the time on the score sheet.
* Complete code all trials, REST, then code all trials again. Rest simply means get up, walk around, get a drink of water,..... Do not code the same trial two times in a row as biases tend to set in. Code the subject on two independent occasions.
* If a subject mouths, sucks, bangs, or throws the object, it is never counted as looking time. Even though sucking can give the infant some information regarding the object's features, such an assumption would depend on cross modal perception skills, particularly in cases in which one item is visually examined and the next is sucked on.
* If a subject holds an object in the air as if handing it to parent/experimenter, and may even appear to be looking at the item while doing this, it is not counted as looking time. The reason is that it is usually impossible for two coders to determine/agree if the subject is looking at the item or right past that item, to the person.
* Listen for any type of biases from parent or experimenter, 'Look at that! Look here!', tapping the object, and labelling the object (which inadvertently and automatically happens when the infant waves the object in the parent's face), instructing the subject to look at the item,.... Any and all of these should be

clearly indicated on the coding sheet by the coder. Biases will disqualify the data.

* Sometimes the size of the object can influence coding. If a subject holds a larger object directly in front of his/her face, you need to determine if the subject is examining the object, or the object is simply in his/her plane of view. Again, it is important to consider 'quality' of looking. Does the infant have a wide-eyed, glazed over look? Or is the infant engaged in inspection? It can be impossible to distinguish the two in cases where large objects are used. With large objects, the infants eyes will appear wide open because a wide visual angle is required to look at the object as a whole, particularly if the object is held close to his/her eyes. large objects can also get between the camera and the infants face, and make coding impossible. The ideal size of objects is too large to be swallowed or disappear into the infant's mouth, but small enough to be easily manipulated and scrutinized.

SUSTAINED ATTENTION CODING

(judgement based on an holistic/integrated basis - do not rate each episode for each feature. An episode of focused/sustained attention that may be characterised with a "1" in all categories would be judged as focused with a high degree of confidence.

A. Steadiness of Gaze

1. Steady gaze at toy or activity.
2. Moderately steady gaze with occasional very brief glances away
3. Frequent looks away, eyes searching, shifting gaze.

B. Facial Expression

1. Serious, intent (brows knit, mouth pursed or slightly open).
2. Relaxed.
3. Smiling, animated.

C. Position of Toys

1. Child lifts toy up and brings it close to eyes for inspection or some manipulation or leans down close to activity on table.
2. The toys are on the table in front of the child, but there is a moderate distance between eyes and toys.
3. The toys are at a distance; the child reaches to play and does not arrange them to facilitate play or exploration.

D. Self-Consciousness

1. Child does not seem to be aware of self.
2. There seems to be some awareness of self in the form of shyness.
3. Child seems very aware of self, with affected speech or dramatic gestures, concerned with the impression made on others in the room.

E. Amount of Extraneous Movement

1. Body movement has stilled except for that essential to the activity being focused on (may be slight but observable tension).
2. There is occasional movement, e.g. scratching, brushing hair aside.
3. Child is wiggling, lifting self up and down, restless.

F. Speed of Movement

1. Slow, deliberate.
2. Moderately fast.
3. Fast, careless.

G. Talking/Vocalising

1. None.
2. Some subdued sounds/talking to self.
3. A lot of talking, much of it directed to someone else.