Using developmental trajectories to examine verbal and visuospatial short-term memory development in children and adolescents with Williams and Down syndromes

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Williams (WS) and Down (DS) syndromes have been associated with specifically compromised short-term memory (STM) subsystems. Individuals with WS have shown impairments in visuospatial STM, while individuals with DS have often shown problems with the recall of verbal material. However, studies have not usually compared the development of STM skills in these domains, in these populations. The present study employed a cross-sectional developmental trajectories approach, plotting verbal and visuospatial STM performance against more general cognitive and chronological development, to investigate how the domain-specific skills of individuals with WS and DS may change as development progresses, as well as whether the difference between STM skill domains increases, in either group, as development progresses. Typically developing children, of broadly similar cognitive ability to the clinical groups, were also included. Planned between- and within-group comparisons were carried out. Individuals with WS and DS both showed the domain-specific STM weaknesses in overall performance that were expected based on the respective cognitive profiles. However, skills in both groups developed, according to general cognitive development, at similar rates to those of the TD group. In addition, no significant developmental divergence between STM domains was observed in either clinical group according to mental age or chronological age, although the general pattern of findings indicated that the influence of the latter variable across STM domains, particularly in WS, might merit further investigation.

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1. Introduction

1.1. Aetiological and cognitive characteristics of Williams and Down syndromes

Williams syndrome (WS) is a genetic condition occurring in between 1 in 7500 (Strømme, Bjørnstad, & Ramstad, 2002) and 1 in 20,000 (Morris, Demsey, Leonard, Dilts, & Blackburn, 1988) live births. It is caused by a microdeletion at the chromosomal locus 7q.11.23. This normally includes the elastin gene (e.g. Ewart et al., 1993; Lowery et al., 1995). Physically, WS is primarily characterised by distinct facial dysmorphia, and other musculoskeletal, cardiovascular, and renal

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abnormalities (e.g. Jones & Smith, 1975; Lenhoff, Wang, Greenberg, & Bellugi, 1997; Wang, Doherty, Rourke, & Bellugi, 1995). Psychological and behavioural markers include mild-moderate intellectual disability (ID; Udwin, Yule, & Martin, 1987), a sociable disposition (e.g. Jones et al., 2000), heightened anxiety (Dykens, 2003) and hyper-sensitivity to sound (e.g. Gallo, Klein-Tasman, Gaffrey, & Curran, 2008).

Down syndrome (DS) is the most common known genetic condition involving intellectual disabilities (Pennington, Moon, Edgin, Stedron, & Nadel, 2003), occurring in approximately 1 in every 700–1000 live births (Kittler, Krinsky-Mchale, & Devenny, 2008). It is caused by a triplication on chromosome 21 (Lejeune, Gautier, & Turpin, 1959). Physical characteristics include a distinctive facial appearance, heart and gastrointestinal anomalies, immunodeficiency, hearing problems, and precocious ageing (e.g. Korenberg et al., 1994; Zigman, Silverman, & Wisniewski, 1996). The most distinctive psychological features of the condition are moderate to severe ID (Pennington et al., 2003), and an increased risk of age-related cognitive decline (e.g. Rowe, Lavender, & Turk, 2006).

Both conditions have been associated with a fractionation of cognitive skills. Individuals with WS have shown relative verbal strengths alongside visuospatial impairments (e.g. Bellugi, Wang, & Jernigan, 1994; Udwin & Yule, 1991), with the latter particularly evident on tasks involving a constructive requirement (e.g. Hoffman, Landau, & Pagani, 2003). This is consistent with evidence for vulnerability of the dorsal stream (e.g. Atkinson et al., 2003; Galaburda & Bellugi, 2000), a visual cortical area involved in processing location and motion (Milner & Goodale, 1995). Although atypicality has been observed in a number of verbal sub-domains (see Brock, 2007, for a review) including pragmatic language (e.g. Reilly, Losh, Bellugi, & Wolfeck, 2004) and spatial grammar (e.g. Phillips, Jarrold, Baddeley, Grant, & Karmiloff-Smith, 2004), comparisons of the verbal and performance IQ scores of individuals with WS have generally shown a verbal superiority (e.g. Grant et al., 1997; Levy & Bechar, 2003). Individuals with DS usually display “flatter” profiles (Jarrold, Baddeley, & Phillips, 2007), but have often exhibited expressive language difficulties (e.g. Abbeduto & Chapman, 2005; Roberts, Price, & Malkin, 2007), especially with syntax (e.g. Chapman, 2003). In addition, verbal ability has been reported to be below performance, or overall, IQ levels (e.g. Vicari, Caselli, & Tonucci, 2000).

1.2. Mapping development

Most studies examining cognitive skills in WS and DS have done so by comparing the mean task scores of these populations with those of groups matched (individuality or overall) for chronological age (CA) and/or mental age (MA). This method usually collapses individual totals, age and ability levels, to give a group mean representative of overall performance level. While this approach enables the direct comparison of groups, it can be argued that it masks development, offering little indication of how skills change over time, how the clinical group may have arrived at that level of performance, and whether this differs from the typical pattern. An alternative approach is the developmental trajectories method, which attempts formally to encapsulate change in performance over time, normally by plotting it against chronological age (CA) and/or a measure of general cognitive ability. This has been claimed to provide a picture that is descriptively richer, in terms of categorising the types of delay or difference shown by clinical groups, than the binary distinction between these two concepts most readily predicated by matching (see Thomas et al., 2009).

The developmental trajectories approach has been employed, with regard to each group, to examine development within a number of domains. For instance, joint engagement behaviour in infants with DS has been mapped longitudinally (Adamson, Bakeman, Deckner, & Romski, 2009), while other authors have used cross-sectional approaches to plot the development of both lexical skills (Thomas et al., 2006), and facial processing abilities (Karmiloff-Smith et al., 2004) in individuals with WS. The latter authors observed that their sample, while often equivalent to TD controls in terms of overall performance on a number of facial processing tasks, showed patterns of development that were both delayed and deviant in comparison. This suggests that measured between-group equivalence may not always derive from similar underlying developmental processes. In addition, other authors have indicated that the skill development of different groups may converge and diverge at different stages. Paterson and colleagues (Paterson, Brown, Gödel, Johnson, & Karmiloff-Smith, 1999) reported similar levels of delay on a task tapping early vocabulary skills in infants with WS and DS. This parity is in marked contrast to the documented superiority of older individuals with WS, over older individuals with DS, in this domain (e.g. Paterson, 2001).

Findings such as these provide clear justification for an approach that accounts for change over time: in order to gain a more sophisticated understanding of skill profiles. This is important for an area such as facial processing, where it has been claimed that the skills of individuals with WS may proceed typically (e.g. Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003).

Despite the uneven ability profiles associated with WS, as well as the usefulness of profiling skills developmentally, only a limited number of studies have used developmental trajectories to compare how verbal and non-verbal/visuospatial skills may improve with development in this population. Jarrold, Baddeley, and Hewes (1998) plotted the performance of a group of sixteen individuals with WS (aged 6–28) using both the verbal British Picture Vocabulary Scale (BPVS; Dunn, Dunn, Whetten, & Pintilie, 1982) and verbal and non-verbal subtests, such as the visuospatial measure Pattern Construction, from the Differential Abilities Scale (DAS; Elliot, 1990) against CA. Verbal performance did not develop at a typical rate across time, but was faster to improve than non-verbal performance, with difference between the two domains increasing in line with verbal ability. Although these data were cross-sectional, and as such did not
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