



# Deeper processing is beneficial during episodic memory encoding for adults with Williams syndrome



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## ABSTRACT

Previous research exploring declarative memory in Williams syndrome (WS) has revealed impairment in the processing of episodic information accompanied by a relative strength in semantic ability. The aim of the current study was to extend this literature by examining how relatively spared semantic memory may support episodic remembering. Using a level of processing paradigm, older adults with WS (aged 35–61 years) were compared to typical adults of the same chronological age and typically developing children matched for verbal ability. In the study phase, pictures were encoded using either a deep (decide if a picture belongs to a particular category) or shallow (perceptual based processing) memory strategy. Behavioural indices (reaction time and accuracy) at retrieval were suggestive of an overall difficulty in episodic memory for WS adults. Interestingly, however, semantic support was evident with a greater recall of items encoded with deep compared to shallow processing, indicative of an ability to employ semantic encoding strategies to maximise the strength of the memory trace created. Unlike individuals with autism who find semantic elaboration strategies problematic, the pattern of findings reported here suggests in those domains that are relatively impaired in WS, support can be recruited from relatively spared cognitive processes.

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## 1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder with an estimated prevalence of 1:20,000 live births (Morris & Mervis, 2000). Although there is significant heterogeneity of cognitive function, individuals with WS tend to function at the level of mild-moderate intellectual difficulty (Searcy et al., 2004). The disorder has attracted the attention of cognitive scientists primarily due to the distinctive cognitive profile (Meyer-Lindenberg, Mervis, & Berman, 2006). A wealth of literature has documented relatively impaired non-verbal and visuo-spatial skills (e.g. Jarrold, Baddeley, & Phillips, 2007; Vicari, Bellucci, & Carlesimo, 2005) compared with relative strengths in the verbal domain (Brock, 2007). This profile occurs against the general backdrop of cognitive impairment. Relative strength in the verbal domain is mirrored behaviourally, as individuals with WS (both children and adults) tend to be highly-social, exhibiting a strong desire to converse with others, clear verbal articulation skills, and speech fluency (Udwin, Yule, & Martin, 1987). However, these verbal abilities are far from

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'intact' and the development of language is far from 'typical'. Rather, the social demeanour shown by some individuals with WS may give a misleading impression of competence and ability, masking the extent of subtle communication atypicalities and language impairments. In a manner similar to the subtle atypicalities that characterise WS language, the spatial skills of individuals with WS not only show a general inaccuracy, but are characterised by subtle atypicalities in processing style, especially a deficit linking information into a coherent whole (Deruelle, Rondan, Mancini, & Livet, 2006). A neglected aspect of the WS cognitive profile is declarative memory and in particular how the component parts of this system, namely episodic and semantic memory interact to produce rich and coherent long-term memory representations.

Not only is the ability to make an association between extracts of information particularly problematic for many individuals with WS (Costanzo, Vicari, & Carlesimo, 2013), it is also a key attribute of episodic memory ability. Long-term episodic memory can be defined as the ability to remember rich details of previously encountered events, which would include not only memory for items but also any accompanying associations and contextual details. Importantly, episodic memory can be further divided into two component processes, namely; recollection and familiarity (Yonelinas, 2002, for review). Devenny et al. (2004) examined episodic and working memory in adults with WS compared to controls (developmental disability with unspecified aetiology). Using a free-recall paradigm, episodic memory was found to be impaired relative to controls and, importantly, age predicted the degree of impairment in the WS group. The lack of difficulty in the working memory domain led the authors to argue for specific problems in memory requiring the need to retrieve rich associative and contextual information. The data were also consistent with the 'accelerated ageing' hypothesis in WS, with a greater deficit in the older adults who had the disorder (mean 48.3 years of age). The suggestion that the cognitive decline emulates the pattern seen in 'normal' ageing would be consistent with impaired episodic compared to semantic memory, albeit with the decline occurring chronologically earlier.

Neuroimaging and studies examining other neurodevelopmental disorders have been informative regarding possible similarities in the profiles of older adults who have developed typically and WS adults, showing parallels in the nature of impairments of the hippocampal region, the key substrate of episodic memory. Meyer-Lindenberg et al. (2005) used multi-modal imaging to investigate structural (MRI) and functional (PET, functional and spectroscopic MRI) integrity and found similarities in structure (although subtle difference were observed) but reductions in resting blood flow and metabolic activity of the hippocampus. The authors argued that the region is critical in the processing of spatial and episodic information. As a final point, it is interesting to note comparisons in the neuro-cognitive profiles of Down syndrome (DS) and Williams syndrome (WS). For example, in research comparing cognition in DS and typical development, Pennington, Moon, Edgin, Stedron, and Nadel (2003) observed exaggerated deficits in those domains of cognition sub-served by the hippocampus (e.g. pattern recognition; paired associate learning) compared to frontal lobe measures (e.g. verbal and design fluency). Therefore, evidence of atypicalities and deficits of cognition in other disorders, even without direct comparison to WS, can be informative.

The behavioural and neuroimaging evidence concerning episodic memory ability in WS therefore suggests an array of atypicalities, especially when linking information in memory. Inspection of the literature regarding the second component of declarative memory, namely semantic memory, reveals less consistency with mixed results regarding more and less proficient areas of functioning. Tests of semantic fluency are informative and tell us much about semantic organisation. In a typical experiment participants are required to generate exemplars from a particular category in a set time (for example listing apple, orange, banana as types of fruits) and WS individuals tend to produce unusual and low frequency exemplars (e.g. Bellugi, Wang, & Jernigan, 1994). However, Jarrold, Hartley, Phillips, and Baddeley (2000) examined individuals with WS and vocabulary matched typical controls (arguably the most appropriate method given the nature of the tasks) on a category fluency task and examined the overall number of exemplars that were generated, how unusual the exemplars were, and grouping of semantic related responses. The findings demonstrated no evidence of the production of unusual responses. A key finding was that during the retrieval of exemplars, impairments in the monitoring of responses were evident; indexed by the number of repeated exemplars given (see Greer, Riby, Hamilton, & Riby, 2013 for a discussion of monitoring and executive control deficits in adults with WS). Therefore, it could be hypothesised that the atypicalities associated with performance on this type of semantic task, are not linked solely to memory or language skill but link to broader aspects of the cognitive capacity and executive function (e.g. see Greer et al., 2013; Rhodes, Riby, Matthews, & Coghill, 2011).

Elsewhere, Thomas et al. (2006) examined picture naming speed (e.g. in the categories of animals, body parts and household items) as a potential measure of the speed of access to semantic memory. Overall speed of naming was slower in participants with WS. However, equivalent performance or at least similar semantic organisation could be proposed since after controlling for this basic speed measure, naming was more difficult and less frequent items were equally problematic across participant groups. Likewise, in another arguably less demanding semantic task, semantic priming and naming speed (speed of access to semantic memory as a measure) was relatively well preserved when target words were preceded by a semantically related (e.g. apple/pear) prime, compared to unrelated (e.g. house/banana) prime (Tyler et al., 1997). A further behavioural finding is noteworthy and highlights semantic strategies employed during memory retrieval rather than simple naming speed. Indeed, Bellugi et al. (1994) reported that when individuals with WS were presented with exemplars from various categories to remember, recall performance was characterised by semantic clustering of the previously studied items (grouping items from the same category) and therefore they suggested that individuals with the disorder were successfully using semantic memory to aid episodic memory performance.

The aforementioned research suggests that when long-term memory requires the encoding or retrieval of rich item and contextual information, difficulties are observed for individuals with WS. However, much like the pattern seen in the

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