



Adaptive functioning in Williams syndrome and its relation to demographic variables and family environment



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ABSTRACT

This study assessed adaptive functioning in children and adults with Williams syndrome. The aims were to: (1) profile adaptive functioning; (2) investigate the relationship between adaptive functions and gender, CA, and IQ; (3) investigate the relationship between levels of adaptive functioning and family environment characteristics. In line with predictions: (1) there was extensive variability in adaptive functions; (2) neither gender nor IQ were significantly related to adaptive skills, but Communication skills and Interpersonal Relationship skills failed to make appropriate gains relative to same aged peers and (3) adaptive functioning was significantly related to family environment. Practical and clinical implications are discussed.

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1. Background

While research into the cognitive and intellectual functioning of individuals with Williams syndrome (WS) has received a lot of attention to date, there has been less research into adaptive functioning and any factors which may impact on the development of these skills.

Adaptive functioning refers to the learned conceptual, social and practical skills performed by an individual in their day-to-day lives (Tasse et al., 2012). Information about adaptive functioning is important to guide early intervention, educational and employment experiences and would benefit families and carers as they endeavour to support individuals with WS to live as independently as possible. For example, as adaptive functioning measures 'real-life' skills, it can be a good indicator of the level of support an individual will require in learning situations, suitable employment and recreational options, and the assistance they may need in managing legal decisions, health care, transportation and finances (APA, 2013).

Understanding adaptive functions also has implications for the diagnosis and classification of intellectual disability (ID), eligibility for funding support and services and in determining the least restrictive environment and level of supervision required for individuals with WS (Dixon, 2007). This is particularly pertinent with the recent revision of the Diagnostic and Statistical Manual of Mental Disorders [Fifth Edition (DSM-5), published by the American Psychiatric Association (APA, 2013)], where adaptive functioning has become a more prominent component of the diagnosis and classification of ID.

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The scant literature available regarding adaptive functioning in WS remains inconclusive in terms of relative strengths and weaknesses of adaptive skills and in terms of the relationship between adaptive functioning and gender, chronological age or level of intellect. Also, no research to date has explored how family characteristics, (family attitudes and family culture) might influence daily independence. This study aimed to investigate adaptive functioning in WS, exploring the profile of adaptive functioning skills and how this relates to demographic variables and family characteristics. As WS is commonly associated with extensive clinical variability (e.g. [Davies, Howlin, & Udwin, 1997](#); [Pezzini, Vicari, Voltera, Milani, & Ossella, 1999](#); [Porter & Coltheart, 2005](#)), both group patterns and individual profiles were explored.

1.1. *Williams syndrome*

Williams syndrome is a genetic disorder resulting from a hemizygous microdeletion on chromosome 7 at the location 7q11.23 ([Ewart et al., 1993](#)). The prevalence of WS is estimated to be approximately 1 in 7500 births ([Strømme, Bjørnstad, & Ramstad, 2002](#)). The disorder is multi-systemic and results in cardiovascular, connective tissue and neurodevelopmental deficits ([Pober and Dykens, 1996](#)). Cognitively, there have been reports of specific strengths and weaknesses in the cognitive profile of WS, characterised by relative strengths in certain verbal abilities (receptive vocabulary, grammatical abilities, verbal short-term memory), but with considerable weaknesses in visuospatial abilities, relation/conceptual language and pragmatics ([Mervis & Klein-Tasman, 2000](#); [Mervis & John, 2010](#)). Behaviourally, individuals with WS are noted for their outgoing and hypersociable personalities ([Jones et al., 2000](#)). At the same time, they have difficulty forming and especially maintaining friendships with same age peers ([Dimitropoulos, Ho, Klaiman, Koenig, & Schultz, 2009](#)).

The recognised cognitive and behavioural phenotype of WS has connotations for adaptive functioning outcomes. For example, their outgoing personalities may be expected to result in higher scores on scales measuring social skills, while difficulties in the area of maintaining friendships may perhaps be expected to negatively influence these scores. Likewise, reported verbal strengths are juxtaposed with pragmatic language impairments, which may have different influences on communication scales of adaptive functioning.

1.2. *Intellectual disability in WS: IQ and adaptive functions*

Intellectual functions are typically in the mild to moderate range in WS, but the level of severity varies widely, from severe (up to four standard deviations below the population mean) to average (within one standard deviation of the population mean) (for a review see [Mervis & John, 2010](#)).

Intellectual profiles also seem to vary in WS. There have been reports of significant differences between verbal and nonverbal (spatial) intellect in WS, yet other studies have failed to find a significant difference between verbal and nonverbal domains (for a review see [Martens, Wilson, & Reutens, 2008](#)). In some studies verbal intellect is reported as a significant strength compared to nonverbal intellect (e.g., see [Searcy et al., 2004](#)), but there are also reports of significantly higher nonverbal than verbal intellect ([Howlin, Elison, Udwin, & Stinton, 2010](#); [Searcy et al., 2004](#)). It is now recognised that not all individuals with WS demonstrate the same cognitive level or intellectual strengths and weaknesses. This highlights the need to go beyond group averages and to explore individual profiles of ability. It also highlights the need to explore why this variability occurs. Variability is likely to reflect inherent and environmental factors.

1.3. *Adaptive functioning in WS*

Adaptive scales, like IQ tests, render a standard score with a mean of 100 and standard deviation of 15. Using this metric, studies have reported that the majority (typically around 75%) of individuals with WS have impairments of adaptive functioning (i.e., more than two standard deviations below the population mean) ([Mervis & Klein-Tasman, 2000](#)). Typically, the mean level of adaptive ability falls within the mild to moderate range, which is consistent with the majority of studies on IQ levels in WS ([Greer, Brown, Pai, Choudry, & Klein, 1997](#)). However, as with IQ, there is evidence of heterogeneity, with some individuals functioning at an extremely low level, while others are functioning at a chronologically age appropriate level. This again highlights the need to consider individuals case-by-case.

1.4. *Strengths and weaknesses in adaptive functioning*

Adaptive functioning is considered a multidimensional construct, and while researchers continue to report a unified composite score, the trend has become to evaluate differences on the specific domain scores ([Dixon, 2007](#)). This is particularly relevant for WS with its peaks and troughs in ability. From the available studies of adaptive functioning in WS, children and adolescents were rated highest on their Socialisation compared to the domains of Communication and Daily Living Skills ([Dimitropoulos et al., 2009](#); [Fisch et al., 2007](#); [Greer et al., 1997](#); [Mervis, Klein-Tasman, & Mastin, 2001](#)). Of note, studies involving adults with WS have found slightly different profiles of adaptive functioning to studies of children with WS. While Socialisation remains a relative strength in adults, Daily Living Skills become the next highest skill, and Communication becomes the weakest area of functioning ([Cherniske et al., 2004](#); [Davies et al., 1997](#); [Howlin, Davies, & Udwin, 1998](#); [Howlin et al., 2010](#)). These differences were confirmed as statistically significant in two of these studies ([Howlin et al., 1998, 2010](#)).

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