

Running head: EDUCATIONAL NEEDS WILLIAMS SYNDROME

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**Addressing the educational needs of children with Williams Syndrome: a rather
neglected area of research?**

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EDUCATIONAL NEEDS WILLIAMS SYNDROME

Abstract

Williams Syndrome (WS) is a rare neurodevelopmental disorder associated with physical health problems, limitations in cognitive abilities, and increased risk of mental health difficulties. This profile of complex needs may make it challenging to support children with WS in schools. Surprisingly, in the current international move for inclusion, limited research exists on the type of educational provision and academic achievements children with WS obtain, including any literature on their voices and the perspectives of key stakeholders. This letter calls for additional research on the risk and protective factors associated with the educational outcomes of these children, the perspectives of the children themselves, and the development of the evidence-base about the effectiveness of education intervention programs.

Keywords: Williams Syndrome, educational provision, academic achievements, transition, interventions

EDUCATIONAL NEEDS WILLIAMS SYNDROME

Williams Syndrome (WS) is a rare genetic neurodevelopmental disorder that affects 1 in 20,000 births (Morris & Mervis, 1999) and is caused by a micro-deletion of at least 27 genes on chromosome 7q11.23 (Koehler, Pabst, Pober, & Kozel, 2015). It is generally diagnosed based on the clinical profile as well as by a blood test to confirm the genetic deletion, either through a fluorescent in situ hybridization (FISH) test or micro-array analysis. Children and young people with WS experience associated physical health problems, including renal abnormalities and cardiovascular difficulties in the light of impaired cognitive development, which ranges between mild to severe intellectual impairment (Martens, Wilson & Reutens, 2008). Children and young people with WS have varied and uneven cognitive and behavioural profiles. Although they show strengths in face perception and auditory short-term memory, have a high drive for social interaction, and affinity with music, they face challenges concerning various cognitive skills, including delayed and atypical language development (especially verbal comprehension), spatial cognition, and number development (Van Herwegen, 2015). Additionally, they are at increased risk of developing behavioural and mental health difficulties including anxiety, attention deficit and hyperactivity disorders, and affective disorders (Riby et al., 2014).

The difficulties described above suggest that supporting the complex needs of these children in education settings may be a challenging task for educators, professionals, and families. However, to date there is a dearth of information concerning: 1) where children with WS are educated; 2) the academic achievements and needs of these children, especially during critical developmental periods, such as the transition from primary to secondary education; 3) evidence-based interventions that can support their educational and psychosocial development. The limited practical research and evidence-base concerning the educational needs/ achievements of children and young people with WS is rather surprising, if one takes into account that their cognitive profile has been the focus of extensive research

EDUCATIONAL NEEDS WILLIAMS SYNDROME

over the last 40 years (Howlin, Davies, & Udwin, 1998; Howlin, Elison, Udwin, & Stinton, 2010). This letter briefly reviews the existing literature to date with regards to these issues and calls for the development of a relevant research base, especially in the light of the need for evidence-based targeted educational interventions and various international policy calls for inclusion and raising the achievements of all children and young people with special educational needs.

Educational provision for children and young people with WS

Although there are still ongoing debates about the rights of children with special educational needs and disabilities (SEND) to be educated in mainstream settings, there is an increasing international trend for pupils with SEND to attend mainstream schools (DfE, 2017; Lindsay, 2007). WS is a rare developmental disorder, but it is still rather surprising that little is known about the context that children with WS are being educated and the support they receive in these placements in the light of the ongoing climate of commitment to inclusive education regardless of disability. One notable recent exception concerns a comparative study in the UK, which examined the educational provision of children with rare developmental disorders, including WS, based on parent and teacher reports. The findings suggested that a reduced number of children attend mainstream secondary education when compared to primary (Reilly, Senior, & Murtagh, 2015). However, the results of this study, except for the type of provision attended, provided limited insight about the level of support that the children with WS were receiving in order to have their needs met.

Additionally, even though a few studies have examined the views of parents and professionals (Fidler, Lawson, & Hodapp, 2003; Fisher & Morin, 2017; Reilly, Senior, & Murtagh, 2015; Reilly, Murtagh & Senior, 2015), there is a shortage of information on the perspectives of key stakeholders about the education provision of children with WS,

EDUCATIONAL NEEDS WILLIAMS SYNDROME

especially about the particular facilitators or barriers that affect their inclusion. Studies on the views of key stakeholders in WS are essential, as previous research in other developmental disorders has now established that professionals' knowledge and understanding are key factors in determining the successful inclusion in school settings (Humphrey & Lewis, 2008; Lindsay, Proulx, Thomson, & Scott, 2013). With exception of Fisher & Morin (2017), studies are literally non-existing concerning the voices of the children and young people with WS themselves regarding various aspects of their educational lives. The need for the views of these children to be heard about their education, health and care needs is imperative for policy, research, and practice internationally (DfE, 2015; Individuals with Disabilities Education Act, 2004).

Educational transition and WS

Transition periods, such as the moving from one year to another but especially from primary to secondary school, places substantial social, intellectual, organisational, and emotional demands upon pupils and is considered to be one of the greatest challenges in a young person's educational career (Riglin, Frederickson, Shelton, & Rice, 2013). This may be particularly true for pupils with WS who except for the difficulties related to the elevated demands of the curriculum, may have to deal with the increased levels of anxiety caused by the changes in the structure and quantity of support they are receiving. For example, although people with WS are extremely sociable, they have difficulties with establishing meaningful peer-relationships. In addition, children and young adults with WS often experience higher levels of anxiety and limited adaptive behaviours (Mervis & Klein-Tasman, 2000). Due to these difficulties children and young people with WS rely more on a structured environment and thus, transitions which impacts on their daily routines, familiarity of the support staff and peers, can be more problematic for them.

EDUCATIONAL NEEDS WILLIAMS SYNDROME

However, to date little is known on the effect of transition from primary to secondary education on the educational and psychosocial outcomes of children with WS. Additionally, there is limited evidence how the important people in the lives of children with WS perceive this transfer and the risk and protective factors associated with successful transition. Further information about how transitions impact children with WS and their family is necessary in order to fully understand how such these transitions can be mediated and prepared for.

Lack of evidence-based educational interventions

Given the fact that WS is a genetic disorder that can be diagnosed early in life, it is in theory possible to intervene from an early age onwards. However, in practice there are very few interventions that target the difficulties in WS and the evidence-based evaluation of educational interventions and therapies for WS is limited. For example, although it has been suggested that based on their cognitive profile children with WS may benefit from the use of computers and calculators to access learning, and that verbal mediation strategies may improve academic outcomes (Dykens, Hodapp, & Finucane, 2000), there is currently a dearth of research evidence that backs up these suggestions. However, there is some evidence that using the cognitive strengths of people with WS can improve their learning difficulties. For example, music therapy has been shown to improve WS children's mathematical abilities (Reis, Schader, Milne, & Stephens, 2003) and verbal working memory abilities to learn new words (Dunning, Martens & Jungers, 2015).

Although it is well-established that a large proportion of children with WS experience overall cognitive impairments, especially visuo-spatial, working memory, language comprehension, planning, and number abilities (Howlin, Elison, & Stinton, 2009), there is a scarce of evidence-based information on the effect that these difficulties have on their literacy

EDUCATIONAL NEEDS WILLIAMS SYNDROME

(including reading, reading comprehension and writing), numeracy skills, and the academic achievements during primary and secondary education (Steele, Sverif, Cornish & Karmiloff-Smith, 2013). Understanding the factors that may predict, hinder or enable the educational achievements of children and young people with WS may be of crucial importance for developing and putting in place evidence-based interventions for children and young people with WS and for raising achievement for this group of children.

Although it can be argued that etiology-based interventions may have a larger impact on the learning outcomes in WS (Hodapp & Fisher, 2017), it may in reality not be possible for teachers and practitioners to put into place WS-specific educational interventions due to the rarity of the disorder. However, seeing the overlap between WS and other developmental disorders, such as Down syndrome (DS) and Autism Spectrum Disorders (ASD), further research is needed whether some of the interventions and educational programmes that have shown to be successful for these disorders would also benefit children with WS. For example, although children with WS and DS show different reading difficulties with children with WS having visual-spatial processing difficulties and those with DS showing poor phonological awareness and vocabulary, vocabulary was a significant longitudinal predictor of reading growth for both groups of children (Steele et al., 2013), and thus the Reading and Language Intervention for children with DS which includes a focus on reading and language development might also benefit reading abilities in those with WS (Burgoyne et al., 2012).

In sum, a better understanding is required of how the strengths and difficulties may benefit or hinder the educational outcomes of people with WS, in order to assess which existing interventions may benefit those with WS and which syndrome specific interventions need to be developed and evaluated through evidence-based studies.

EDUCATIONAL NEEDS WILLIAMS SYNDROME

Call for future research

Moving forward, it is essential that future research in WS starts building the evidence-base in relation to the educational outcomes of these children and young people and the interventions that benefit those with WS. Cross-sectional and longitudinal studies identifying the factors that predict the educational outcomes of these children and highlight potential facilitators and barriers, especially during educational transition times would be of particular importance in shaping relevant policy and practice in better addressing the needs of children with rare developmental disorders in school settings.

Another area that there is a paucity of research concerns the views of the stakeholders in relation to the educational needs children and young people with WS. Their knowledge and awareness of the strengths and difficulties in WS is essential in addressing the complex needs of these children in educational settings. Additionally, research on the voices of children and young people with WS would also be essential as involving them meaningfully in decision making may make them feel empowered to live as independently as possible.

To sum up, WS is a complex and rare developmental disorder which is characterised by a number of physical health problems, cognitive impairments, and mental health issues. Meeting the needs of these children in educational settings may pose a challenge for schools and professionals involved with supporting them. However, research in the educational outcomes of children and young people is still very much literally non-existing and yet essential in establishing the best educational provision and developing evidence-base intervention programs that can support their learning and raise their achievements.

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EDUCATIONAL NEEDS WILLIAMS SYNDROME

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EDUCATIONAL NEEDS WILLIAMS SYNDROME

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EDUCATIONAL NEEDS WILLIAMS SYNDROME

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