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Cross syndrome comparison of sleep problems in children with Down syndrome and Williams syndrome

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ABSTRACT

Based on previous findings of frequent sleep problems in children with Down syndrome (DS) and Williams syndrome (WS), the present study aimed to expand our knowledge by using parent report and actigraphy to define sleep problems more precisely in these groups. Twenty-two school-aged children with DS, 24 with WS and 52 typically developing (TD) children took part in the study. Each child wore an actiwatch for a minimum of four nights and parents completed the Children's Sleep Habits Questionnaire (CSHQ). Sleep problems were common in both developmental disorders. Children with DS had the greatest sleep disruption, with frequent and longer night wakings as well as restlessness. Parents reported symptoms of sleep-disordered breathing and a range of other problems including grinding teeth, bedtime resistance and sleep anxiety. Children with WS had problems initiating sleep and parents also reported bed-wetting and body pain. Despite these problems, the mean actual sleep time, as measured by actigraphy, did not differ between the three groups. CSHQ reports were in agreement with actigraphy for children's sleep duration, but this was not the case for sleep latency, restlessness and the night wakings variables. Sleep problems in DS and WS are common and appear to be syndrome-specific. Due to the inaccuracy of parent report, it is recommended that children at risk undergo objective measures of sleep assessment.

1. Introduction

Sleep problems in TD children are common, with around one third experiencing some kind of sleep disturbance (Mindell & Owens, 2003; Owens, Spirito, McGuinn, & Nobile, 2000; Pegg, 2006). These range from behaviourally-based problems such as behavioural insomnia, to physiological problems such as sleep disordered breathing (SDB) and periodic limb movement disorder (PLMD). Parasomnias such as nocturnal enuresis (bed-wetting), somnambulism (sleep walking), bruxism (grinding teeth) or sleep terrors are also common but are generally outgrown by mid childhood.

Sleep is an important aspect of development and poor quality sleep has a detrimental effect on physical, cognitive and social functioning. Poor quality of sleep may manifest in behaviours such as daytime sleepiness, irritability, hyperactivity and impulsivity and have a negative effect on school performance (see Fallone, Owens, & Deane, 2000 for a review). Sleep

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problems can exert great pressure and distress on the family but are often amenable to treatment, which can improve the child's and family's quality of life (Merrell & Shott, 2007; Wood & Sacks, 2004).

It is now well established that sleep problems are also common in individuals with developmental disorders, yet there is relatively little information available characterising sleep patterns in these groups. This is perhaps surprising when considering the high frequency of sleep problems in such children and the importance of good sleep for healthy physiological and psychological development. For these reasons, reliable and valid methods for studying sleep in atypically developing children are crucial. The gold standard technique for the diagnoses of many sleep disorders is polysomnography (PSG): a detailed assessment in a sleep laboratory of physiological and neural activity during sleep. While this is a key diagnostic tool for sleep disorders that alter neurophysiology, such as PLMD, or respiratory function, such as in SDB, it only provides a single time point assessment that fails to capture dimensions of sleep associated with behaviour and usual sleep environment (Stores,Wiggs, & Campling, 1998). Furthermore, it is not always appropriate for children with developmental disorders who may become distressed at sleeping in a strange environment.

Relatively recently, actigraphy (movement monitoring) has emerged as a reliable and valid alternative which can be used to continuously assess activity levels in a home setting over a prolonged period of time. It yields more than 80% agreement with PSG in the prediction of wake and sleep for typical groups (Sadeh, Hauri, Kripke, & Lavie, 1995), but in comparison is inexpensive, non-intrusive, and less time consuming to analyse. In addition, it is more likely to be tolerated by children with developmental disorders. Its use can be supported by the use of questionnaires completed by parents to provide rich information on sleep habits and characteristics.

The aim of the current study is to use actigraphy supported by parent report to investigate common and syndrome-specific sleep problems in children with Down syndrome (DS) and Williams syndrome (WS) compared to a typically developing (TD) control group.

1.1. Down syndrome

DS is the most common sporadic genetic developmental disorder (1/700 live births) usually associated with the presence of three copies of chromosome 21 (trisomy 21). It is the leading genetic cause of intellectual disability, yielding an average IQ of around 50 points, but with wide individual variability (Roizen & Patterson, 2003). Individuals with DS often experience sleep disturbances (Carter, McCaughey, Annaz, & Hill, 2009; Cotton & Richdale, 2006). The most common of these is obstructive sleep apnoea syndrome (OSAS), a condition where the upper airway occludes during sleep, causing both disruption of sleep and intermittent hypoxia. Data from prospective community studies (Dyken, Lin-Dyken, Poulton, Zimmerman, & Sedars, 2003; Shott et al., 2006) and from children referred for clinical examination of SDB (Marcus, Keens, Bautista, Von Pechmann, & Ward, 1991) suggest that OSAS affects around two thirds of children with DS and is likely to be attributed to physical features associated with DS, such as craniofacial and upper airway abnormalities, obesity, tonsil and adenoid encroachment, and generalised hypotonia (Churchill, Kieckhefer, Landis, & Ward, 2011). Problems with settling, sleep maintenance and early morning waking have also been described from parent report studies (Breslin, Edgin, Bootzin, Goodwin, & Nadel, 2011; Cotton & Richdale, 2006; Stores, Stores, Fellows, & Buckley, 1998).

Much research assessing sleep in children with DS relies on parent reports, which may not always be as comprehensive and accurate as using objective methods. For instance, Shott et al. (2006) conducted an overnight PSG study with 56 young children with DS. They found evidence of SDB in 80% of their sample but when compared to parent report, it emerged that parents had both over- and under-reported problems, with only 23% being accurate. The high incidence of OSAS and the inaccuracy of parent reports together underline the importance of objective methods for studying sleep in individuals with DS.

1.2. Williams syndrome

WS is a rare neurodevelopmental disorder caused by a deletion of some 28 genes on the long arm of one copy of chromosome 7 at q11.23 (Donnai and Karmiloff-smith, 2000). Individuals with WS are inclined to be overly sociable and their performance is relatively proficient on language tasks, despite having an average IQ of 56 (range: 50–70) (Bellugi, Wang, & Jernigan, 1994; Mervis et al., 2000).

Similarly to the DS population, much previous data on sleep in WS has been acquired solely from questionnaire studies. These reported settling problems and night waking (Udwin, Yale, & Martin, 1987), as well as bed-wetting, getting up for the bathroom, and sleep anxiety (Sarimski, 1996). More recently, difficulties at bedtime, long sleep latencies, night waking as well as sleep anxiety have been reported (Annaz, Hill, Ashworth, Holley, & Karmiloff-Smith, 2011), and PLMD has found to be common (Arens et al., 1998; Goldman, Malow, Newman, Roof, & Dykens, 2009; Gombos, Bódizs, & Kovács, 2011). PSG has also demonstrated significant differences in sleep architecture, specifically decreased sleep efficiency, decreased REM sleep and increased slow wave sleep (Gombos et al., 2011; Mason et al., 2011). Although research thus far indicates significant sleep problems in WS, more in-depth research expanding this area is critical.

A cross-syndrome comparison of sleep problems in children with DS and WS will determine whether there are characteristic patterns specific to each disorder, or whether problems are common to both groups and could therefore be common to developmental disorders generally. Based on previous research, we expect to find significant sleep problems in children with DS and WS relative to TD children, but that there will be discrepancies between parent-report and actigraphy