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Characterisation of sleep problems in children with Williams syndrome

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ABSTRACT

Sleep is critical to optimal daytime functioning, learning and general health. In children with established developmental disorders sleep difficulties may compound existing learning difficulties. The purpose of the present study was to evaluate the prevalence and syndrome specificity of sleep problems in Williams syndrome (WS), a neurodevelopmental disorder affecting around 1 in 20,000 live births.

Parents of 64 children with WS, aged 6–12 years, and 92 age matched healthy controls were surveyed about their child's sleep habits. The Child Sleep Habits Questionnaire, general health and background information were collected from the parents. Ninety seven percent of parents reported that their children had sleep problems and reported a high prevalence of sleep difficulties: greater bedtime resistance, sleep anxiety, night waking and daytime sleepiness. This is the first study to our knowledge to survey sleep problems in a large cohort of school age children with WS. Sleep problems in children with learning difficulties are often amendable to treatment if diagnosed early. Furthermore the negative impact of sleep disturbances on daytime behaviour and learning should be measured before diagnoses of behaviourally defined disorders are considered.

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1. Introduction

There is now a substantial body of evidence indicating that sleep is important for the healthy development of children and contributes to optimal social and neuropsychological function (Hill, Hogan, Karmiloff-Smith, 2007). Sleep requirements alter through life as a function of biological and environmental changes with age, and there are distinct differences between adult and child sleep patterns. The prevalence of sleep problems in typically developing school-aged children is relatively low (around 6–10%). However, in direct contrast, the existence of a high rate of sleep disturbances (34% to 86%) has been reported in children with developmental disorders such as Autism (Richdale, 1999; Richdale & Schreck, 2009; Stores & Wiggs, 2001), Angelman syndrome (Pelc, Cheron, Boyd & Dan, 2008), Attention Deficit Hyperactivity Disorder (Paavonen et al., 2009) and Down syndrome (Carter, McCaughey, Annaz, & Hill, 2009). Although the existence of sleep disorders in developmental disability is well established, their causes and syndrome specificity are yet to be identified and quantified.

There is abundant evidence to suggest that good night-time sleep leads to improved daytime behaviour in typically developing children and in children with developmental disorders (e.g., Maas et al., 2010; Scher, Hall, Zaidman-Zait, & Weinberg, 2010). Sleep disturbances can give rise to severe behavioural difficulties during the day, such as aggression,

Abbreviations: CSHQ, Child Sleep Habits Questionnaire; SRE, sleep-related enuresis; TD, typically developing children; WS, Williams syndrome.

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screaming, tantrums, non-compliance, and impulsivity (Stein, Mendelsohn, Obermeyer, Amromin & Benca, 2001) and predict maternal mood, stress, and fatigue family functioning, including maternal depression and familial stress (Meltzer & Mindell, 2007). Persistent sleep problems have been also linked to immune-system dysfunction (Franck et al., 1999), impairments in cognitive functioning resulting in a possible lack of understanding of the differences between wakefulness and sleep, inability to self soothe between sleep phases, and general anxiety (Sheldon, Ferber & Kryger, 2005).

Williams syndrome is a rare genetic disorder caused by a hemizygous microdeletion of some 28 genes on chromosome 7q11.23, with an approximate incidence of 1 in 20,000 live births (Schubert, 2009; Tassabehji, 2003). It results in a complex physical, cognitive and behavioural phenotype that includes an uneven cognitive profile, with relatively proficient face recognition and language skills alongside poor numerical and visuo-spatial skills compared to overall mental age, and overall IQs in the 50s to 60s range (Donnai & Karmiloff-Smith, 2000; Searcy, Lincoln, Rose, Klima & Bavar, 2004). Physically, the WS phenotype includes a dysmorphic face, growth retardation, congenital heart disease (typically supravalvular aortic stenosis occurring in 75% of cases), premature ageing, weakness of connective tissue, renal anomalies and reported infantile hypercalcaemia. Facial symptoms include a wide mouth with full cheeks and full lips and periorbital fullness with a broad forehead.

Although parents of children with WS informally report that their children suffer from sleep disturbances, research into the specific sleep problems in WS is scarce, however, there are now several research groups investigating the issue. Early studies described settling problems and night waking (Einfeld, Tonge, & Florio, 1997; Udwin, Yule & Martin, 1987), as well as bed wetting and sleep anxiety (Sarimski, 1996). In another study, Arens et al. (1998) undertook an eight-question telephone survey of 28 families of children with WS (age range: 1.5–10 years olds) and reported sleep problems such as night awakenings, restless sleep and difficulty initiating sleep. Polysomnography was performed on seven of these children with WS and compared to 10 typically developing controls. Periodic limb movement index was fivefold greater in children with Williams syndrome. However, the small sample size and wide age range make the results difficult to generalise to WS as a whole. A more recent study focused on a group of adolescents and adults with WS, using a sleep questionnaire and wrist actigraphy (Goldman, Malow, Newman, Roof & Dykens, 2009). The authors reported reduced sleep efficiency, prolonged sleep latency, increased number of night wakings and elevated movements during the night. However, no direct comparisons to controls or previously normative values were performed.

The aims of the current study were to (1) determine the prevalence of sleep problems in school-aged children with Williams syndrome and compare with TD children; (2) investigate the association between different types of sleep problems and (3) explore the relationship between sleep problems and child characteristics, including medication use, age, gender, ethnicity, child's health history and socioeconomic status. This was carried out by targeting a large population of children with WS within a narrow age range and primary school-aged children in the UK, by means of widely used sleep questionnaires.

2. Methods

2.1. Participants

Parents of primary school-aged children with WS were identified from the full database provided by the UK Williams Syndrome Foundation. Of the 82 invited parents, 64 completed and returned both questionnaires, yielding an overall response rate of 82%. Reasons for non-participation included: nine families unobtainable despite repeated contact attempts, seven changed addresses, with only two declining to participate. This cohort represents over half of all school-aged children with WS in the United Kingdom. All children with WS had been diagnosed clinically, as well as by means of the *fluorescence in situ hybridisation* (FISH) genetic test for deletion of one copy of the Elastin gene. The control group comprised 92 TD children sourced from Hampshire schools, UK. Background data from schools and parents were collected to ensure that the control group did not have any clinical diagnoses or learning difficulties before they were included in the study. Prior to recruitment of the participants, ethical approval was granted by the Middlesex University London Ethics Committee, the University of Southampton, School of Psychology Ethics Committee and the Williams Syndrome Foundation, UK.

2.2. Measures

The *Child Sleep Habits Questionnaire*: Parents completed the Child Sleep Habits Questionnaire (CSHQ) (Owens, Spirito & McGuinn, 2000), a screening instrument for school-aged children based on common clinical symptom presentations of prevalent sleep disorders. The CSHQ consists of 33 items rated on a 3-point Likert scale (never/rarely, sometimes, and usually). It yields scores on 8 sub-scales: sleep anxiety (e.g., "afraid of sleeping in the dark"), sleep duration, bedtime resistance, sleep onset delay, night waking, parasomnias, sleep disordered breathing and daytime sleepiness. It provides a total sleep disturbance score, as well as a problem-specific breakdown. Psychometric properties indicate that the CSHQ can identify sleep problems in a clinical sample and can distinguish between clinical and community samples (Waumans et al., 2010).

In addition, parents of children with WS completed developmental history and health habits sections of *The Pediatric Sleep Clinic Questionnaire* (Owens, http://www.kidzzzsleep.org). Demographic data were obtained from both groups.

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