JIDR Journal of Intellectual Disability Research
Published on behalf of mencap and in association with IASSID

Journal of Intellectual Disability Research doi: 10.1111/j.1365-2788.2011.01499.x

VOLUME 56 PART 6 pp 600–608 JUNE 2012

Sleep in children and adolescents with Angelman syndrome: association with parent sleep and stress

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Abstract

Background Sleep concerns are common in children with Angelman syndrome, with 20–80% of individuals having a decreased sleep need and/or abnormal sleep–wake cycles. The impact of these sleep behaviours on parental sleep and stress is not known.

Method Through the use of standardised questionnaires, wrist actigraphy and polysomnography, we defined the sleep behaviours of 15 children/adolescents with Angelman syndrome and the association of the child/adolescents sleep behaviours on parental sleep behaviours and parental stress.

Results Both children/adolescents and their parents exhibited over 1 h of wake time after sleep onset and fragmented sleep. Prolonged sleep latency in the child was associated with parent insomnia and daytime sleepiness. Additionally, variability in child total sleep time was associated with parental stress.

Conclusions Poor sleep in children/adolescents with Angelman syndrome was associated with poor parental sleep and higher parental stress. Further work is warranted to identify the underlying causes of the poor sleep, and to relate these findings to daytime functioning, behaviour and the family unit.

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Institution where work was performed: Vanderbilt University Medical Center.

Keywords Angelman syndrome, child–parent sleep, parental stress, sleep problems

Introduction

Angelman syndrome, a neurodevelopmental disorder caused by the absence or non-functioning of the maternal *UBE3A* gene, occurs in an estimated one in 12 000 to one in 20 000 individuals (Williams *et al.* 2006; Pelc *et al.* 2008). There are four main genetic mechanisms underlying this disorder: (1) deletions of the 15q11–q13 region on the maternal chromosome (approximately 70% of the cases); (2) parental uniparental disomy in which there are two copies of the 15th chromosome inherited from the father and none from the mother (2–3% of the cases); (3) mutation in the *UBE3A* gene on maternal chromosome 15 (5–10% of the cases); and (4) an imprinting defect which causes the chromosome 15 inherited from the mother to have the paternal imprint for gene functioning silencing maternal genes in the 15q11–13 region (3–5% of the cases). An additional 10–15% of the cases are diagnosed according to clinical criteria but have no recognisable genetic mutation. Characteristics include developmental delay, intellectual disability (ID),

recurrent seizures, lack of speech, and disorders of gait and balance (Williams *et al.* 2006; Williams 2010).

Sleep disturbances are common in Angelman syndrome, and are included in the diagnostic criteria established in 1995 and in the updated criteria published in 2006 (Williams *et al.* 1995, 2006). The prevalence of sleep problems in Angelman syndrome ranges from 20% to 80% (Williams *et al.* 2006). This variation in prevalence may relate to how sleep problems are defined (Pelc *et al.* 2008). Sleep problems appear more prevalent, and severe, in childhood between the ages of 2 and 9 years; however, in some individuals the sleep problems will persist (Clayton-Smith 1993; Bruni *et al.* 2004; Didden *et al.* 2004; Pelc *et al.* 2008).

Symptoms of insomnia, in particular difficulty initiating or maintaining sleep, are primary concerns (Clayton-Smith 1993; Zhadanova *et al.* 1999; Bruni *et al.* 2004; Berry *et al.* 2005; Walz *et al.* 2005); however, there have been reports that children with Angelman syndrome may need less sleep than typically developing children (Clayton-Smith 1993; Zhadanova *et al.* 1999; Bruni *et al.* 2004; Berry *et al.* 2005). Poor sleep in these children does not necessarily impact daytime alertness. Few studies have objectively investigated the specific sleep abnormalities present in this disorder. Miano *et al.* (2004, 2005) found reduced sleep efficiency, a lower percentage and duration of rapid eye movement sleep, and a higher percentage of slow wave sleep in children <8 years when compared to normal controls. Additional research is needed in this area.

The impact of sleep problems on parental sleep is understudied in typically developing children, and even less described in children with developmental disorders. Sleep problems in one family member often impact other members (Dahl & El-Sheikh 2007). In the general population, parents who perceive their child to have a sleep problem are more likely than those who do not to report having too little sleep (National Sleep Foundation 2004). Furthermore, the impact of the child's sleep on the parent's sleep is often the reason why a child's sleep is brought to the attention of a medical professional (Polimeni *et al.* 2005). Robinson & Richdale (2004) found that parents of children with developmental disabilities, who reported sleep problems in their children, were more likely to note how the child's sleep negatively affected theirs. Parents of children with autism were found to have poorer sleep quality and shorter sleep quantity than parents of typically developing children (Meltzer 2008). Parents of children with chronic illnesses were found to have sleep disruptions, which possibly explain their higher rates of negative daytime functioning (Meltzer & Moore 2008). Furthermore, mothers of children with poor sleep were found to have more mood problems, and stress problems, than those whose children did not have poor sleep (Meltzer & Mindell 2007). While evidence of poor night-time sleep in the child affecting parental sleep can be seen in other populations with developmental disabilities, minimal information is available on the association of disturbed sleep in children with Angelman syndrome and the impact of disturbed sleep on the primary caregiver.

In this study our goals were to: (1) obtain additional objective and subjective data on sleep in children/adolescents with Angelman syndrome; and (2) relate these sleep patterns to the primary parental caregivers sleep, daytime sleepiness and stress.

Method

Participants and procedure

The study population consisted of 16 children/adolescents, and the parent considered as the primary caregiver, participating in the Angelman Natural History Study at Vanderbilt University who volunteered to participate in the sleep protocol. The sleep protocol consisted of a sleep interview with a sleep clinician, overnight polysomnography (PSG) concurrent with wrist actigraphy, 28 nights of in-home wrist actigraphy and sleep diary, and a standardised sleep questionnaire. All caregivers provided consent, and the protocol was approved by the Institutional Review Board at Vanderbilt University.

Children, accompanied by the parent, were admitted to the Vanderbilt Sleep Research Core, located within the Vanderbilt Clinical Research Center (CRC) to undergo one night of PSG and actigraphy monitoring. After being admitted to the CRC, an actigraph was placed on the non-dominant wrist of the parent and child. The parent

was instructed in the use of the devices including that they were to be worn nightly starting several hours before bedtime until waking the next morning, and as tolerated during the day for the next 27 days. Participants were allowed to fall asleep according to their usual bedtime routine and to awaken spontaneously. Lights out (time child first attempted to fall asleep) and lights on (time child awoke) were recorded by the PSG technologist at the beginning and end of the study.

Instrumentation

Actigraphy, used extensively in sleep research and clinical practice, has been validated as a highly reliable method to differentiate sleep from wake (Sadeh & Acebo 2002; Ancoli-Israel *et al.* 2003). The actiwatch is a small-computerised wristwatch-like device used to detect movement and can serve as a surrogate of sleep and wake (Sadeh & Acebo 2002). Actigraphy measurements were obtained with an Actiwatch AW-64® manufactured by Philips Respironics (Bend, OR, USA). Data from the actigraphs were downloaded to a personal computer where all sleep intervals were manually placed on an actogram. Sleep variables were calculated using Actiware V5 software (Philips Respironics, Bend, OR, USA). The time in bed was calculated from lights out until lights on the next morning. The total sleep duration was the sum of all sleep epochs within the interval between the time set on the actogram for night-time sleep and morning wake time. Sleep efficiency was calculated as the ratio of total night-time sleep duration to the total time in bed. Sleep latency was calculated as the time required for sleep onset after lights out (first attempt to go to sleep). Wake after sleep onset was measured as the sum of all wake epochs during the sleep period and reflects the number of minutes scored as wake that exceeded the sensitivity threshold. The fragmentation index captures all movement regardless of the intensity of the movement.

Polysomnography

Polysomnography data were obtained using the Nihon-Kohden 9200 sleep acquisition system. Monitoring included 21-channel electroencephalography (to evaluate for epileptic seizures), electrooculography, chin and bilateral leg (tibialis anterior) electromyography, nasal thermistor, nasal pressure transducer, electrocardiography, thoracic and abdominal effort, and pulse oximetry. Video was recorded simultaneously as part of the PSG recording. Sleep parameters were calculated for each PSG study by the sleep software package (Polysmith Acquisition and Review software, version 4.0.17.0; Neurotronics, Inc.). Total sleep time was the total time comprised of epochs of sleep, with sleep efficiency calculated as the ratio of total sleep time to total time in bed. Sleep latency was calculated as the time to the first epoch of sleep after lights out, and wake time after sleep onset, as the total time comprised of epochs of wake after sleep onset. Arousal index was calculated based on the American Academy of Sleep Medicine definition of an electroencephalography arousal (Iber *et al.* 2007). Following acquisition, PSG data were manually scored by a single sleep technologist, using standard criteria (Iber *et al.* 2007) and reviewed by the senior author.

Questionnaires

Children's Sleep Habits Questionnaire

Sleep behaviours were evaluated using the Children's Sleep Habits Questionnaire (CSHQ). The CSHQ is a validated parental questionnaire consisting of 45 items relating to sleep complaints over the past month. The majority of questions are answered on a 3-point scale (1 = rarely, 2 = sometimes, 3 = usually). A total score is calculated, as well as sub-scale scores that measure insomnia-related aspects of sleep including sleep anxiety, sleep duration, sleep onset delay, night wakings, bedtime resistance and other dimensions such as sleep-disordered breathing, parasomnias and daytime sleepiness (Owens *et al.* 2000). The CSHQ has been used in children with a variety of neurodevelopmental disorders (Malow *et al.* 2006; Goodlin-Jones *et al.* 2008) with demonstrated validity and reliability in these disorders.

Parental sleep and stress

Three self-administered questionnaires were used to evaluate the parents sleep behaviours and stress. The Insomnia Severity Index (ISI; Morin 1993) was

used by the parents to evaluate the severity of their own insomnia. The ISI has been validated as a measure to detect changes in perceived sleep difficulties in adults with insomnia (Bastien *et al.* 2001). The ISI contains five questions reflecting areas such as the severity of the insomnia problems and how they interfere with daily functioning. The Epworth Sleepiness Scale (ESS; Johns 1991) was used to evaluate the general level of daytime sleepiness in the parents. The ESS consists of eight questions representing typical daytime situations commonly found in daily life. It has been validated as a reliable method to distinguish normal individuals from those with a wide range of sleep disorders. The Parenting Stress Index Short Form (PSI-SF; Abiden 1995) was used to assess parenting stress. The PSI-SF measures the parental stress arising from factors inherent to the child, inherent to the parent and factors associated with parent–child interaction. It is widely used for measuring parental stress in autism spectrum disorders (Zaidman-Zait *et al.* 2010). The PSI-SF consists of 36 items that measure the key constructs of parental distress, parental–child dysfunctional interactions, and the extent the parent considers the child to be difficult and a total score. It also includes a scale that evaluates the extent the parent answers the questionnaire with a bias to present themselves in the most favourable manner and to minimise the problems or stress in the parent–child relationship (defensive responding).

Statistical analysis

Descriptive statistics were conducted on all major variables. Actigraphy data for parent and child were averaged across nights for each participant. Spearman rank correlations (r_s) were used to evaluate associations between PSG sleep variables, actigraphy sleep variables and subjective sleep measures. Mann–Whitney *U*-tests were used to determine overall difference between parent and child on the sleep variables. Results are presented as mean (standard deviation) and given the pilot nature of the study, we did not pursue formal multiple comparison adjustment. A *P*-value of less than 0.05 was considered statistically significant. Analyses were performed using SAS V9.2 software (Cary, NC, USA).

Results

A total of 16 parents consented to participate in the study. Questionnaires were completed on 15 (94%) participants, overnight PSGs were performed on 14 (88%) participants, and 10 (63%) participants completed a minimum of seven night's in-home actigraphy. Seven (44%) participants had PSG and actigraphy data recorded on the same night, and actigraphy data were obtained from 13 (81%) parents. One parent withdrew from the study prior to the start of the PSG and another parent completed only the questionnaires, as she believed that the PSG and actigraphy would be problematic for her child to complete.

Children ranged in age from 2 to 16 years with a mean (standard deviation) of 6.5 (4.4) years. There were nine (60%) girls and six (40%) boys; 10 (67%) children were deletion-positive, three (20%) children had uniparental disomy, one child had an imprinting centre mutation and one child had an imprinting centre mosaic genotype (this child had only questionnaire data). All children were on at least one medication. The primary medications and number of participants using that medication were: melatonin ($n = 8$, 53%), clonazepam ($n = 7$, 47%), gastrointestinal medication ($n = 7$, 47%), topiramate ($n = 3$, 20%) and other medications ($n = 9$, 60%). All parents participating in the parental part of the study were mothers.

Sleep behaviours

Subjective sleep measures

Parents in this study all reported that their child had a sleep problem, and five (33%) parents reported some degree of co-sleeping with their child. Subjective measures of sleep are presented in Table 1. When compared to the normative sample published by Owens *et al.* (2000), children/adolescents with Angelman syndrome scored higher on all CSHQ sub-scales.

Objective sleep measures

Table 2 presents the PSG and actigraphy results of the child and the actigraphy results of the parent. Wilcoxon signed-rank tests to evaluate differences between each sleep variable obtained by actigraphy

Table 1 Children's Sleep Habits Questionnaire (CSHQ) sub-scale scores in children/adolescents with Angelman syndrome compared to normative sample

CSHQ sub-scale	Angelman syndrome ($n = 15$)	Normative sample* ($n = 469$)
Bedtime resistance	8.2 (2.9)	7.1 (1.9)
Sleep onset latency	1.5 (0.8)	1.3 (0.5)
Sleep anxiety	6.0 (1.4)	4.9 (1.5)
Sleep duration	5.6 (1.6)	3.2 (0.9)
Night wakings	5.8 (1.6)	3.5 (0.9)
Parasomnias	10.4 (1.4)	8.1 (1.3)
Sleep-disordered breathing	4.4 (1.4)	3.2 (0.6)
Daytime sleepiness	11.4 (3.2)	9.6 (2.8)
Sleep total	49.0 (7.3)	41

* Normative sample from Owens *et al.* (2000).

Table 2 Objective sleep measurements [mean (standard deviation)] of children/adolescents with Angelman syndrome and their parent

Sleep measurement/ measure	Sleep onset latency (min)	Sleep duration (min)	Night wakings WASO (min)	Fragmentation (index)	Time in bed (min)
Child – polysomnography	37.6 (50.3)	373.9 (132.8)	57.2 (91.5)	10.8 (14.4)	504.4 (107.0)
Child – actigraphy on polysomnography night	32.9 (33.1)	381.9 (100.5)	123.0 (85.0)	52.5 (21.2)	499.9 (149.5)
Child – in-home actigraphy	40.8 (23.3)	405.5 (54.8)	114.0 (32.5)	49.7 (6.1)	519.6 (63.5)
Parent – in-home actigraphy	31.7 (22.5)	379.5 (47.6)	63.8 (30.0)	35.0 (11.9)	443.4 (65.3)

WASO, wake after sleep onset.

on the night of the PSG and the average for all nights of in-home actigraphy showed no differences. There were also no differences found between the PSG sleep variables and the actigraphy-calculated sleep variables on the night of the PSG. No differences were found between parent and child sleep for average sleep latency, average total sleep time, standard deviation for time in bed and standard deviation of total sleep time. However, parent and child sleep differed for average time in bed, average wake after sleep onset, average sleep fragmentation, standard deviation for wake after sleep onset and the standard deviation for fragmented sleep.

Associations between subjective and objective sleep measures

Table 3 presents the associations between the actigraphy-measured child and parent sleep parameters and the CSHQ domains. Actigraphy measures of night-time waking (wake after sleep onset) were highly associated between child in-home actigraphy and actigraphy on the PSG night ($r_s = 0.76$), and child in-home actigraphy and the PSG ($r_s = 0.64$). Additionally, a high association was found between the average amount of wake after sleep onset on in-home actigraphy and the actigraphy-measured time in bed on the night of the PSG ($r_s = 0.64$).

Associations between child's and parental sleep and stress

To evaluate the association of the child's sleep with the parent's daytime sleepiness, insomnia and stress, we examined Spearman rank correlations (r_s) between the average values obtained for the in-home actigraphy and the parent's subjective questionnaires. The child's average sleep latency was associated with a higher level of daytime sleepiness measured by the ESS ($r_s = 0.71$) and more symptoms of insomnia as measured by the ISI ($r_s = 0.66$). Total sleep time variability was associ-

Table 3 Spearman rank correlations (r_s) between sleep variables measured by actigraphy and parental reported sleep problems in children/adolescents with Angelman syndrome*

	Children's Sleep Habits Questionnaire sub-scales				
	Sleep anxiety	**Sleep delay**	**Sleep duration**	**Night wakenings**	**Parasomnias**
Child actigraphy – polysomnography night					
Sleep latency					
Wake after sleep onset	–0.95		0.82		
Fragmentation index		–0.76			
Time in bed	–0.86				
Child actigraphy – in home					
Sleep latency					
Fragmentation index			0.60		
Sleep duration				0.66	0.69
Parent actigraphy – in home					
Wake after sleep onset			0.59		
Fragmentation index			0.70		

* Only values with $r_s > 0.50$ and $P < 0.001$ are reported.

ated with the following scales from the Parental Stress Index-SF: Defensive Responding ($r_s = 0.64$), Parental Distress Scale ($r_s = 0.72$), and the Total Score ($r_s = 0.61$).

Discussion

Our study employed a multimodal approach to examine sleep in children with Angelman syndrome, and their primary caregiver, using subjective and objective measurements. Our results are consistent with previous research showing sleep problems in these children and highlighting the importance of understanding nocturnal awakenings and restless sleep. Furthermore, our findings showing disrupted night-time sleep of the parents in association with the parent's report of problems with the child's sleep duration suggest that the parents' sleep quality is affected by the child/adolescents sleep.

We found an association between a longer sleep duration on in-home actigraphy and parent report of parasomnias on the CSHQ. We also saw an association of the parasomnia scale with measurements of disrupted night-time sleep, wake after sleep onset and the fragmented index. Parasomnias, movement problems, daytime somnolence and disordered breathing have also been reported in one survey (Bruni *et al.* 2004). The parasomnia scale on the CSHQ is broad, including the items bedwetting, talking and walking in sleep, waking screaming or sweating, and restless sleep/excessive movement. A high frequency of bruxism, enuresis, sleep terrors and sleepwalking, as well as restless sleep, has been reported in another questionnaire study in Angelman syndrome (Bruni *et al.* 2004). It is not surprising therefore that given the inclusion of the question related to restless sleep/excessive movement on the parasomnia scale, this scale was significant in our study. Furthermore, compared to Owens *et al.*' normative sample of children (Owens *et al.* 2000), the parents of children in our study reported higher levels of parasomnias, sleep-disordered breathing and daytime sleepiness.

Night wakings were noted both subjectively by parental report and objectively with actigraphy and PSG. Wake after sleep onset measured by PSG averaged almost 1 h over all children and, when measured with actigraphy, both in the laboratory and at home, averaged almost 2 h. These findings, along with the high fragmentation index in both the sleep lab and at home, highlight the disturbed sleep in these individuals. These objective measurements support previous subjective reports of other researchers who have noted frequent night wakings,

and sleep maintenance problems, to be severe and more prevalent than settling problems in children with Angelman syndrome (Didden *et al.* 2002). A prior case series of adults with Angelman syndrome using wrist actigraphy showed similar fragmented sleep to our children (Anderson *et al.* 2008). A high frequency of abnormal movements, including restless sleep, have previously been reported with the suggestion that these nocturnal movements might be part of the extrapyramidal symptoms of Angelman syndrome including tremors, myoclonus and dyskinesia (Bruni *et al.* 2004).

Our study is the first to our knowledge to look at the sleep patterns and parenting stress in parents of children with Angelman syndrome. Previous studies have shown that sleep problems in children with ID are related to family functioning (Richdale *et al.* 2000; Didden *et al.* 2002; Meltzer & Mindell 2007). High levels of disturbed sleep, such as wake after sleep onset and fragmented sleep, are often associated with increased stress and other medical problems in adults. Parents of children with developmental disabilities have been shown to experience higher levels of stress than parents of typically developing children (Doo & Wing 2006; Sung *et al.* 2008; Zaidman-Zait *et al.* 2010). While sleep is one possible source of parental stress, these causes are most likely multifactorial. Our study extends this finding to parents of children with Angelman syndrome. The association of child sleep, parent daytime sleepiness and insomnia found in this study suggests that in a caregiver population that is already stressed, attention to the caregiver's sleep is important to minimise potential adverse effects.

Similar to other neurodevelopmental disorders, the aetiology of insomnia in Angelman syndrome is most likely multifactorial rather than due to one specific component. In Angelman syndrome, biological abnormalities in the circadian control of sleep are a plausible cause of insomnia. The possibility of disturbed melatonin metabolism has been suggested based on the response to melatonin treatment in small trials (Braam *et al.* 2008). Psychological factors, such as reliance on parents to promote sleep (Walz *et al.* 2005), or associating the bedroom with activities other than sleep, are also likely to be involved. Other causes or contributing factors to insomnia in Angelman syndrome may include primary sleep disorders such as nightmares (Walz *et al.* 2005) or epileptic seizures (Clayton-Smith 1993). Independent of epileptic seizures, sleep continuity and architecture may be disrupted by frequent interictal epileptiform activity or by abnormal high-amplitude rhythmic slow wave discharges that are characteristic findings in the waking electroencephalographies of Angelman syndrome patients (Dan & Boyd 2003).

High wake after sleep onset has previously been reported in children with Angelman syndrome. Using PSG, Miano *et al.* (2005) showed a higher percentage of wake after sleep onset, and a higher rate of awakenings per hour, in children with Angelman syndrome than children with ID, or epilepsy.

The strengths of our study include the use of a validated questionnaire, wrist actigraphy and PSG that measure both objective and subjective sleep. Limitations are a small sample size across a large age range where sleep patterns may differ. There was also incomplete data, which was due in part to parents' concerns about their children's abilities to tolerate PSG. Additionally, participants were part of the Angelman Natural History Study and may not be representative of the general Angelman population.

In summary, our study shows the presence of high nocturnal waking and arousals in children/adolescents with Angelman syndrome, as well as evidence suggesting high fragmentation and night-time wakings of parents of these children/adolescents that are potentially associated with the sleep of their child. Research to identify potential causes of these arousals, for example, seizures or interictal epileptiform activity, is warranted. Improvement in the child's sleep not only will benefit the child's health and well-being, but also offers promise for improving the sleep and daytime vigilance of the parents.

Acknowledgements

This research was supported in part by a Donna and Jay Richards – Hobbs Discovery Grant from the Vanderbilt Kennedy Center for Research on Human Development, Vanderbilt University Medical Center, as well as in part by Vanderbilt CTSA Grant 1 UL1 RR024975 from the National Center for Research Resources, National Institutes of Health.

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Accepted 15 September 2011