Sleep disturbances in the disabled child

A case report and literature review

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Sleep disturbances in children are common. In children with intellectual disabilities sleeping problems are more common. This may result in increased burden of illness, additional parental stress, and day time behavioural difficulties. This article illustrates the problems that sleep disturbances create for children with disabilities and discusses methods for managing sleep problems.

Case study – AB

AB, 10 years of age, is a boy with Mowat-Wilson syndrome, a genetic disorder resulting from a deletion on the long arm of chromosome 2.[1] He presented with the primary features of this syndrome, which include severe intellectual disabilities, microcephaly and seizures. He also presented with the associated features of Hirschsprungs disease, hypospadia, bowel obstructions, urogenic bladder and agenesis of the corpus callosum.

AB's sleeping pattern is one of frequent sleep and settling difficulties during the night with a prolonged awake time during the day. Frequently he will wake up early in the morning and then remain awake for the rest of the day. Naps are rarely taken.

This has major impacts on the family. Both his mother and grandmother who care for him have significant disruption of their own sleep patterns. His mother drives a special education school bus and finds this difficult after sleep deprivation. Apart from his medical conditions, the sleep disorder has the most severe impact on the family's functioning and wellbeing.

In the general population, up to 20% of children aged 2 years and 14% of children aged 3 years are reported to wake up regularly.[2,3] Numerous studies have described the prevalence of sleep disturbances among children with intellectual disabilities. Quine et al[4] described 200 severely mentally handicapped children of which 51% had settling problems and 67% had frequent night waking. A survey by Wiggs et al[5] found severe sleep problems in 43% of intellectually disabled children. Barlett et al[6] showed that intellectually disabled children aged less than 6 years have a prevalence rate of 86% for sleep problems. Richdale et al[7] determined that 59% of children with an intellectual disability had a sleep disturbance that persisted for more than 2 years.

Effect of sleep disturbances

It is difficult to define what constitutes 'normal' sleep. The sleep requirements of an intellectually disabled child are even more difficult to define. If there are variations, the basic problem still lies in the impact of their sleepless nights. Normal sleep requirements decrease as age increases (*Table 1*). Obtaining a scattered, interrupted night of sleep negatively affects the child and can also have a major impact on caregiver(s). In the child it is common to see day time behavioural issues such as hyperactivity and aggression.[8] In both the child and caregiver, day time fatigue and decreased cognitive functioning are apparent.[9] Mood disturbances in the caregiver have also been associated with sleep deprivation.[10] The caregiver's ability to function is extremely difficult when sleep is compromised, particularly when involved with work, providing care for other family members and carrying out functional duties as the primary caregiver for their child. A study of parents with disabled children highlighted too little rest and sleep as being the major problems in dealing with their children. They suggest it makes the parents feel as if they are in a prison with an infant who will never grow up.[10]

In addition, sleep deprivation also has direct physical effects. Sleep deprived individuals allowed 4 hours of sleep per night for 6 nights in comparison to controls produced only half the normal serum level of antibodies after immunisation with influenza vaccine.[11] The endocrine system is affected with rising cortisol levels, and an increase in sympathetic outflow. Increased insulin resistance has also been shown.[11] It has also been shown that with excessive sleep loss in adults (less than 6 hours sleep per night) reactions become similar to that of someone with a blood alcohol level of 0.1%.[12]

Pathophysiology

Sleep is defined as a reversible behavioural state of perceptual disengagement from and unresponsiveness to the environment.[13] There are two significant stages of sleep: non-REM (rapid eye movement) also known as slow wave sleep, and REM sleep. Non-REM occurs predominately in the first half of sleep and REM predominately in the second half. Non-REM sleep is defined as a relatively inactive state, yet the brain is actively regulating in a movable body. In REM sleep the brain is highly active but the body is paralysed. Non-REM sleep is known for increases in growth hormone levels and restorative functions of the immune, nervous, muscular, and skeletal systems. REM sleep has been indicated for memory consolidation.

What causes sleep is a range of factors with our circadian rhythms overriding the entire system. Zeitgebers, which are environmental time cues, tune our biological clock to a 24 hour sleep-wake cycle. The strongest cue is the light-dark cycle but other nonphotic cues include melatonin, exercise and social cues. When these are disturbed due to disease, blindness and environmental factors such as time zone differences, our body's internal clock becomes skewed and has to readjust with the help of zeitgebers.[14]

Treatment

Behavioural therapy and medication are options found to be effective in treating sleep disturbances.[9,15–20] The following therapies have been found to work in children with intellectual disabilities. It is beneficial to identify the particular cause of the sleep disturbance either by thorough history taking and/or through functional assessments in order to choose the most appropriate therapy (*Table 2*). Didden et al[9] described functional assessments as a range of procedures used to identify antecedent and consequent variables that control problem behaviour in natural environments.The aim is to identify the nature of the sleep disturbance and the trigger(s). In difficult cases this might be complex and therefore require formal assessment in a sleep disorder clinic.

Behavioural therapies

Behavioural therapy is most effective in children who are maintaining their disruptive behaviours due to social factors, ie. parents are reinforcing negative sleep behaviours with positive attention. There are two main ways of targeting behaviour in relation to sleep disturbances: extinction and desensitisation. The extinction technique consists of parents/caregivers ceasing their original methods for soothing their child to sleep and instead putting them to bed with a bedtime routine. After saying 'good night' they are not to re-enter the room. They are not to react to the child's behaviour and in the morning if the child had been quiet throughout the night the child receives positive reinforcement.

Desensitisation is often used in children who express anxiety about being left alone at night. This technique allows the parent to re-enter the room once the child has been put to bed if the child cries out for them. Each night the period of time lengthens before the parent is allowed to re-enter the room after the initial cry out. This desensitises the child to time alone at night. Initially behavioural therapy is quite stressful for the caregiver and requires ongoing support. Significant improvements can be expected in most cases within several weeks.[9]

Melatonin and other medications

Medications have been tested in aiding people with sleep problems. Melatonin is the most widely prescribed drug as it is a naturally occurring hormone secreted by the pineal gland in the brain. It is thought that exogenous melatonin can affect the sleep-wake cycle in three different ways:

- the mechanism may involve a phase shift of the endogenous circadian pacemaker
- a reduction in core body temperature, and
- a direct action on somnogenic structures of the brain.

Table 1. Sleep requirements defined by age group[22]

Age	Total sleep hours
Newborn – 1 year	14–16+
Toddlers	10–13
Pre-schoolers	10–12
Primary school age	10+
Pre-teens	9+
Teens	8–9.5
Adult	7–8
Elderly	8–9

Melatonin is given in doses ranging from 0.003–75.0 mg. The response to melatonin is similar in most individuals regardless of their age or sex. A 2004 review reported melatonin enhanced sleep onset in people with primary sleep disorders but had no clinically significant affect on sleep onset in people with secondary sleep disorders.[15] Another recent study has shown that add-on melatonin can be used in conjunction with valproic acid and has a significant decrease in an epileptic child's parasomnias. It did not however, decrease sleep fragmentation scores, but the positive effect it had on epileptic fits throughout the night suggests this to be a beneficial treatment.[16] Ross and Whitehouse[17] enrolled 40 children with sleep problems, who had either cerebral palsy, learning difficulties, epilepsy, autistic spectrum disorders or Down syndrome. They showed a 50% decrease in sleep disturbances from an average of seven interruptions per week to three per week once the children were administered melatonin at levels of 2.5 mg if under 5 years, and 5 mg if over 5 years of age. The dosage could be increased up to a maximum of 10 mg in children over 5 years of age. The number of hours of sleep per week increased by 10% for the children in the study.

Anticonvulsants may be useful in diminishing sleep disturbances in children with epilepsy.[21] Although some anticonvulsants may act more as a sedative, others may increase sleep disturbances. *Table 3* outlines some of the common anticonvulsant medications and their

Table 2. Aetiologies of sleep disturbances in various disorders

Disorder	Aetiology
Corpus callosum agenesis	Sleep is regulated by many different parts of the brain. The corpus callosum is part of the frontal cortex and is one area well known to control sleep. Slow wave activity in the frontal cortex during non-REM sleep impairs local regulation of sleep. This is due to altered neuronal connectivity, which is especially prevalent under conditions of increased sleep propensity[23]
Visual impairment	People with no conscious light perception often have a non-24 hour sleep-wake cycle. Their circadian rhythms (6-sulphatoxymelatonin which is the end product of melatonin and cortisol) induce good and bad sleep throughout the day[24]
Epilepsy	Epileptic activity may disrupt the normal organisation of sleep and cause sleep disruption. Partial epilepsy has effects on both sleep continuity and sleep architecture (non-REM and REM sleep are reduced)[25]

Table 3. Anticonvulsants and their effect on sleep[21]

	Delays sleep onset	Negatively affects REM	Sedative
Phenobarbital	No	Yes	Yes
Clonazepam	No	Yes	Yes
Phenytoin	No	Yes	No
Carbamazepine	No	Yes	No
Lamotrigine	Yes	No	No
Gabapentin	No	No	No
Valproate	No	No	No
Oxcarbazepine	Not yet known	Not yet known	Not yet known
Levetiracetam	Not yet known	Not yet known	Not yet known
Zonisamide	Not yet known	Not yet known	Not yet known

effect on sleep.

Recommendations for AB

In our case study, social attention is positively reinforcing AB's behaviour, therefore an extinction program may be very effective. It might also be useful to complement behavioural therapy with melatonin in order to enhance sleep onset and increase the number of hours of sleep per night. However, the impact of melatonin in regards to his epilepsy needs to be closely observed. Expectedly, melatonin will have a positive effect, however, in anecdotal reports an increase of seizures has been attributed to melatonin.

Conclusion

General practitioners and paediatricians will be confronted with the problem of sleep disorders and disturbances in children with intellectual disabilities. It is important that they elicit the signs and symptoms of sleep disturbances in the child and family and recognise that there are effective management strategies of sleep disturbance. Functional assessment may be required to assess possible underlying causes of sleep disturbance. It is important to elicit from the family what strategies have already been tried, and their effectiveness. Ultimately, behavioural therapy is the ideal way to treat sleep disturbances. More research needs to be done into this area of assessing sleep disorders.

Conflict of interest: none declared.

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