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Cerebral palsy and sleep disordered breathing

Case report

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Introduction

Sleep breathing issues in children have been described for some considerable time and it is well established that children with neurodisability such as cerebral palsy are at increased risk of sleep disturbances [1, 2] when compared to the general population. However, there are concerns that awareness amongst clinicians remains patchy, and that many children with neurodisability have sleep breathing problems that remain undiagnosed and untreated. This case study illustrates that these issues remain challenging, highlighting the multifactorial approach required when dealing with sleep in this patient subgroup.

Cerebral palsy is said to affect 1.5–3.0 people per 1000 live births in Europe. As a result of the primary brain injury, children with cerebral palsy have multiple risk factors that predispose them to an increased risk of sleep disturbance. Cerebral palsy children can have an impaired initiation of sleep; this may be linked to blindness or visual impairment where their light perception is altered causing timing and maintenance of sleep to be altered as a result of the effect on the release of melatonin [3]. Epilepsy is another potential influence on the sleep of a cerebral palsy child. Sleep problems associated with epilepsy can be a major complicating factor. Epilepsy disrupts sleep quality [1, 4], which in turn exacerbates the epilepsy. Furthermore, antiepileptic drugs may then adversely affect the quality of sleep attained. Sleep disordered breathing has also been seen to effect uncontrolled epilepsy.

Sleep quality can also be affected by other common elements of the condition such as generalised abnormality of muscle tone or an abnormal neuromuscular control of their upper airway that can leave the upper airway at risk of obstruction [5, 6]. Cerebral palsy children are also known to regularly encounter pain during sleep as a result of muscle spasms, postural equipment used overnight and gastro-oesophageal reflux.

The contribution of these various factors leaves a cerebral palsy child, as will be discussed in this case, at risk of developing a problem with their sleep and their breathing during sleep. Identification and treatment of these at risk patients is therefore paramount.

Case presentation

A 10-year-old female with quadriplegic cerebral palsy:

- was born prematurely at 35 weeks
- has a Gross Motor Function Classification System score of 4
- has post-haemorrhagic hydrocephalus with a ventriculoperitoneal shunt
- had historical epileptic spasms that have now resolved (medication no longer required)
- has progressive scoliosis
- has nonverbal communication with learning difficulties
- uses a wheelchair
- uses a sleep system to regulate posture overnight
- previous had a tonsillectomy for snoring (5 years before)

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Despite the known correlation between neurodisability and sleep disordered breathing, cases are still missed http://ow.ly/2pNS305KII3

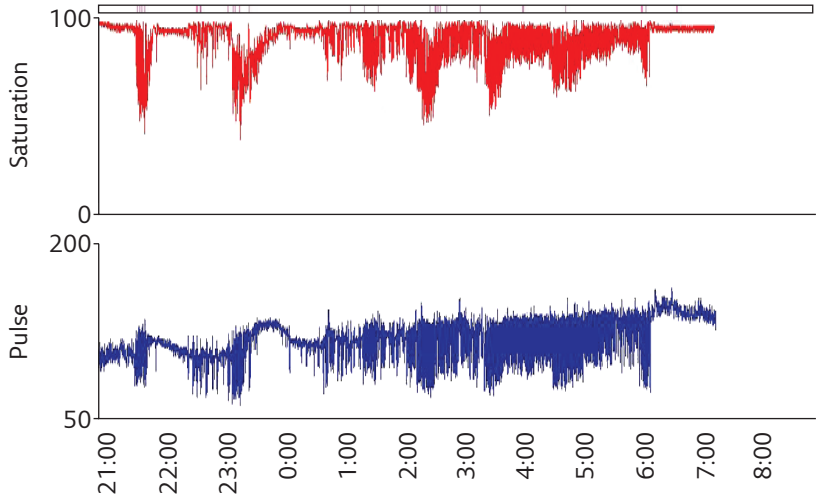


Figure 1 *Home oxygen saturation trace.*

Mean SpO_2:	87.7%		
		≥4%	≥3%
Dips per h:		85.3	122.0
Mean nadir:		76.8	79.0

Time spent	hh:mm:ss	% of analysis
<98%	10:15:34	99.69
<96%	09:26:10	91.69
<94%	07:01:50	68.32
<92%	05:17:20	51.39
<90%	04:28:58	43.56

Figure 2 *Oximetry statistics. SpO_2: oxygen saturation measured by pulse oximetry.*

At a routine annual review by the community paediatricians the parents disclosed that the patient's school had noticed that she was falling asleep in class. Further enquiry elicited symptoms of snoring, restlessness during sleep, gasping and pauses in breathing. A home oxygen saturation monitor study was requested to screen for any abnormalities.

The result of the oximetry study showed the patient to be profoundly hypoxic overnight (figures 1 and 2). The profile of the oxygen trace bore a striking resemblance to that of an adult obstructive sleep apnoea (OSA) trace.

- Mean oxygen saturation measured by pulse oximetry (SpO_2): 87.7%.
- Desaturation index: dips ≥4%, 85.3 per h; dips ≥3%, 122.0 per h.
- Time spent with SpO_2 <94%: 68.32% of the time studied.
- Heart rate profile: climbing baseline heart rate suggestive of stress and increased work of breathing.

Task 1
What would you suggest the clinician should do in response to the oximetry result?

Answer 1

Refer the patient to a specialist centre for more detailed sleep investigation to assess for an obstructive or central component to the breathing abnormality and for evidence of underlying lung disease: cardiorespiratory sleep study (CRSS) or polysomnography.

The patient was urgently referred to the tertiary centre for further investigation. Although a polysomnography investigation is still commonly regarded as the best diagnostic clinical test [2, 6, 7], a CRSS was performed in this instance. It was thought that when combined with the strong clinical history, the CRSS provides an appropriate level of complexity to both identify and quantify any sleep disorder.

The CRSS (figure 3) showed the patient to be suffering from significant sleep disordered breathing with prolonged obstructive events resulting in sleep fragmentation and repeated episodes of hypoxia (figure 4).

As a CRSS was performed, it was not possible to assess for sleep staging or respiratory disturbance index; however, using the 2012 American Academy of Sleep Medicine guidelines [8] to score respiratory events, an apnoea–hypopnea index (AHI) was calculated (fig. 3).

- AHI: 101.4 events per h, showing close correlation to desaturation index.
- 95.7 of these events per hour were obstructive in nature.
- Mean S_{pO_2}: 93.3%.
- Desaturation index ≥3%: 93.7 dips per h.
- Time spent with S_{pO_2} <94%: 40.53% of time studied.
- Mean transcutaneous carbon dioxide: 7.0 kPa.

The majority of the events scored were obstructive in origin with a small number of central and mixed events. The transcutaneous carbon dioxide was raised with cyclical increases in the baseline noted throughout the study in association with clusters of severe obstruction.

Sleep profile

Analysis duration:	10:28:54	Sleep latency:	1:00:54	Stage 1:	0
Wake duration:	2:12:24/21.1	REM latency:	N/A	Stage 2:	0
Total sleep time:	8:16:30	Non REM time:	8:16:30	Stage 3:	0
		REM time:	0:00:00	REM:	0
		Sleep efficiency:	78.9%		

Respiratory events

	Number	Index
Obstructive:	785	94.9 /h
Mixed:	36	4.4 /h
Central:	11	1.3 /h
Hypopnoea:	7	0.8 /h
HYP: OBS	7	0.8 /h
HYP: CENTRAL	0	0 /h
Total (AHI):	839	101.4 /h
RERA:	0	0.0 /h
Total (RDI):	839	101.4 /h

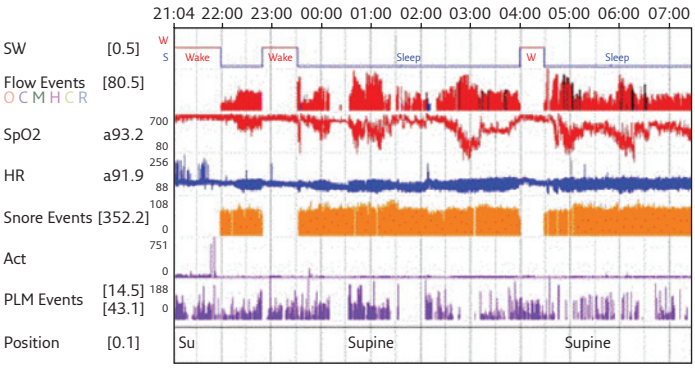
	Number	Index
Periodic breathing:	N/A	N/A

Snoring: 161.3 minutes and 32.5% of TST

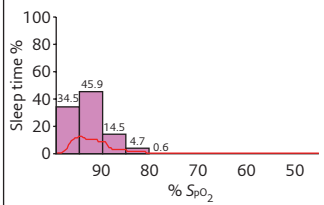
Transcutaneous monitor (TOSCA)

Mean S_{pO_2}:	93.3%
Dips ≥4%	61.8 /h
Dips ≥3%	93.7 /h
Mean P_{tCO_2}:	7.0 kPa

Event summary graph



S_{pO_2} summary (Nonin monitor)



S_{pO_2} time <90%:	19.5%	01:36:53
Average S_{pO_2}:	92%	
Deepest desaturation:	75%	
Longest desaturation:	120.3 s	
Average desaturation:	5.2%	
Number of desaturations:	384	
Desaturation index:	44.0 /h	(≥3%)

Figure 3 *CRSS report. REM: rapid eye movement; HYP: hypopnoea; OBS: obstructive; AHI: apnoea–hypopnoea index; RERA: respiratory event-related arousal; RDI: respiratory disturbance index; SpO_2: oxygen saturation measured by pulse oximetry; $PtcCO_2$: transcutaneous carbon dioxide tension.*

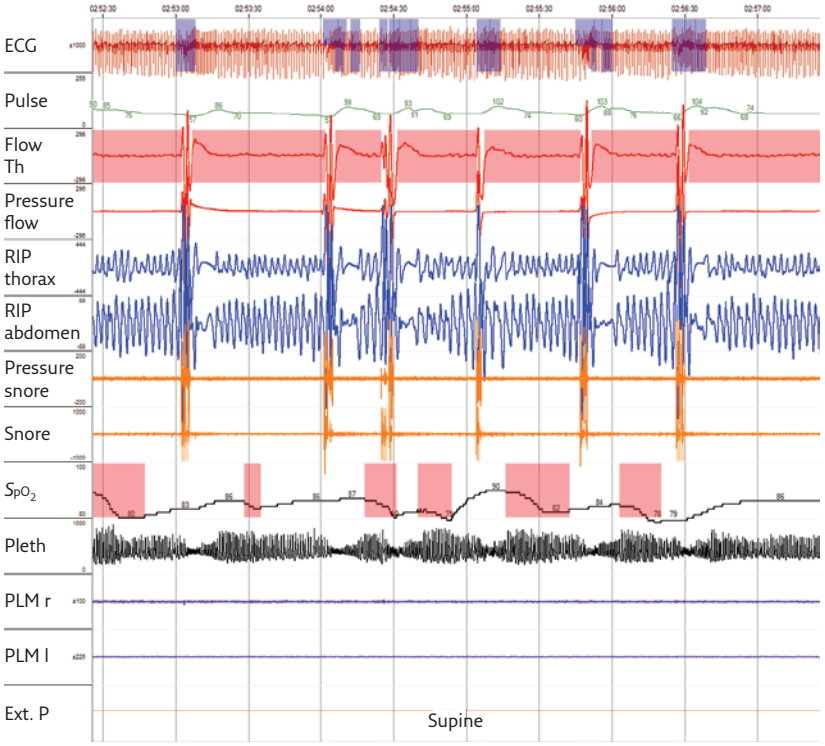


Figure 4 *2-min page of sleep channels showing prolonged obstructive events.*

The sleep physiologist supervising the overnight study documented, in addition to the obstructive events observed (red channel in figure 3), that the patient exhibited an increased work of breathing with significant sternal and subcostal recessions and tracheal tug, with the patient seen to be restless (purple channel in figure 3) and snoring throughout (brown channel in figure 3).

In contrast to adult sleep medicine, a positive study in paediatrics is often classed as having one or more obstructive events per hour, with the severity scale often classified as 1–5 events per h being mild, 5–10 events per h being moderate and ≥10 events per h being severe [2], although there is currently no formal consensus. Given the marked clinical symptoms and the severity of intermittent hypoxia, and the extremely high AHI of 101.4 events per h, treatment options were sought immediately.

Task 2
What are the treatment options?

Task 3
What treatment option would you take?

Answer 2

- Ear, nose and throat (ENT) department to review for tonsillar regrowth, and assess size of adenoids. Given that the patient had a tonsillectomy 5 years prior to the current presentation, is there sufficient remaining lymphoid tissue present to justify a further surgical procedure?
- Positive pressure therapy: continuous positive airway pressure (CPAP)/noninvasive ventilation (NIV) therapy
- Tracheostomy
- Oxygen therapy: this would not deal with the underlying abnormality of airway obstruction but would improve hypoxia, with care being taken to ensure that it does not lead to carbon dioxide levels rising further. For some children with severe neurodisability, a less aggressive form of management may be judged to be appropriate following discussion with the family or if the child is unable to tolerate other modalities of treatment.

Answer 3
Trial of positive pressure therapy

An ENT review was requested. Studies have shown that surgical approaches to the treatment of OSA in a cerebral palsy patient can prove successful, with symptomatic improvement reported [6, 9, 10]. The patient had a previous tonsillectomy 5 years prior to the current presentation. Upper airway examination did not reveal sufficient residual tonsillar or adenoidal tissue to warrant an operation.

When there is a high disturbance index to sleep there is a significantly higher risk of the complaint persisting despite any surgical procedure [2]. Therefore, it was decided that positive pressure therapy was the most appropriate option in this instance.

CPAP has been well described as an effective treatment for infants and children [6, 11], with the potential adverse effects being similar to those encountered in adult medicine. For children with neurodisability, who often have associated learning problems, establishing CPAP can be a significant challenge and additional resources such as psychology input or behavioural techniques have proven to be useful adjuncts to the process [6].

Due to the level of hypoxia and the severity of the OSA discovered, it was felt that the patient should be immediately commenced on treatment [2].

On this occasion, the patient was trialled on bilevel therapy with a backup rate due to concerns over comfort and the central component found on the sleep study. Although small in number, the central apnoeas were prolonged, causing deep desaturations.

Over the next two nights, the patient's pressures were titrated by the overnight sleep physiologist. On the second night, using a spontaneous/timed mode with pressures of 12/8 cmH_2O and a backup rate of 12 breaths per min *via* a full face mask (FFM), the gas exchange profiles were greatly improved.

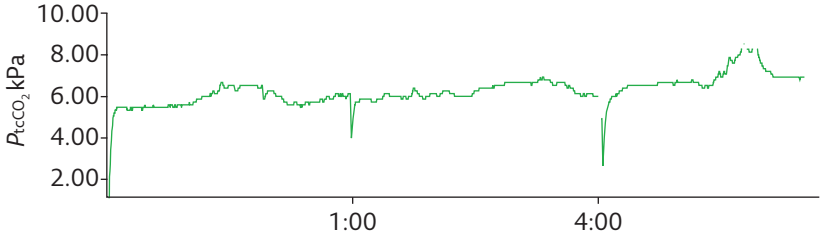
- Mean S_{pO_2}: 96.7%.
- Desaturation index: dips ≥4%, 5.6 per h: dips ≥3%, 14.5 per h.
- Time spent with S_{pO_2} <94%: 3.93% of time studied.
- Mean transcutaneous carbon dioxide: 6.3 kPa; however, there was a slight increase in baseline over the night, suggesting that treatment was still not optimal (figure 5).

Although gas exchange within the normal range had not yet been established, it was felt that the patient would not, at this point, tolerate the increased pressures required to achieve this. This was due to worries over patient compliance and persistent leak around the mask. With the gas exchange profile now greatly improved, it was felt appropriate to discharge the patient home with a follow-up review planned for 4 weeks.

Ongoing management

On the three subsequent inpatient visits over the next 6 months, bilevel pressures were titrated with effective pressures found to be 18/14 cmH_2O. A trial of single-level CPAP was undertaken on one of the visits but this showed no therapeutic advantage over bilevel positive airway pressure, and the patient became agitated and did not tolerate the change in mode, so was returned to bilevel therapy.

In addition to the normalised gas exchange, the parents and school reported a significant improvement in the child's health and quality of life. She was more alert, no longer napping at



Mean P_{tcCO_2}			
5th/95th percentile:	5.47/7.20		
Time below	<4.0 kPa	00:00:00	0.00%
Time above	≥6.0 kPa	05:52:18	73.73%

Time spent	hh:mm:ss	% of analysis
<6.0 kPa	02:05:33	26.27%
<6.5 kPa	04:42:48	59.18%
<7.0 kPa	07:25:06	93.15%
<7.5 kPa	07:38:27	95.94%
<8.0 kPa	07:47:54	97.92%

Interpretation: See Above

Figure 5 *Transcutaneous carbon dioxide tension ($PtcCO_2$) trace on bilevel support.*

school, quiet and restful when sleeping, and now woke up without requiring repeated attempts to stir.

The parents also reported having an improved quality of life as a direct consequence of their daughter's improved sleep routine. With night awakenings now greatly reduced, they no longer felt that they had to sleep in the same room as the patient to attend to her during the night.

Issues with mask fit and leak persisted but in view of the clinical improvement, the family were happy to continue with the therapy despite this side-effect.

Discussion

Children need good sleep as sleep problems are known to impact upon growth and development, general health, mood, learning, and social and family interaction [2, 6]. The main issue that this case study highlighted was that even with a diagnosis of cerebral palsy, which is well known to increase the risk of sleep disturbance [1], and regular attendance at a variety of medical clinics, the patient was slow to be diagnosed as having a significant sleep disorder. It is likely that this was not caused by an individual factor but due to a combination of issues related to muscle tone, neuromuscular regulation of the airway, posture and chest deformity.

A key learning point to come from this case study is to review the screening pathway that was provided by the secondary and tertiary centres. Knowing which patients to screen for sleep disturbances, and when, is difficult. With resources under pressure, clinicians have to use diagnostic tests in a targeted manner but this leaves potential for patients to be missed. Patients known to the specialist centre to be at risk will generally receive routine follow-up. The more concerning group is the children with complex medical issues that may attend several clinicians whose focus is on specific aspects of their care. In children with complex needs, eliciting a clear sleep history may not be prioritised when there are multiple other medical needs, particularly in units without any sleep specialists. Routine diagnostic markers used to identify sleep disturbances are not as clear in this group of children. Changes to behaviour or levels of alertness can be difficult to differentiate when the baseline is abnormal and other factors such as pain, discomfort or seizures may themselves affect sleep patterns and quality. The local pathway for this child required the patient/parent to present a concern before an investigation was performed. This, however, requires the parent to be able to identify that there is a problem. For this group of patients, there is not always a formal screening pathway. This should be reviewed.

In this particular case, two sleep interventions had taken place, suggesting that there was a degree of awareness of sleep issues prior to the latest referral. A postural sleep system had been in use and a tonsillectomy was performed 5 years prior for snoring. It is likely that opportunities to highlight poor sleep and a potential sleep breathing problem were missed, and that increased local awareness of sleep problems in children with neurodisability might have allowed the OSA to have been identified still earlier. Education of health professionals and of families is a key learning point to be taken from this case study.

Parental reporting of sleep disturbance is thought to be one of the best predictive markers for OSA [7]. If the parent is unaware their child has a sleep issue, they are unlikely to raise any concerns with their general practitioner (GP) [12, 13], particularly when they have multiple other ongoing medical requirements. Parents of children with neurodisability have been known to relate observed disturbances in their child's sleep to "normal behaviours" or the inevitable consequence of their child's condition [13]. Their child may never have had a normal sleep routine, having always been a poor sleeper, tolerating multiple awakenings in the night where other parents would not. If a secondary sleep disorder such as OSA evolves, the patient or parent may be unaware [13] and certainly much less likely to identify any deterioration in what was always a fragmented sleep routine. This could, in part, explain why sleep problems in a child with neurodisability can be slow to present in primary care.

Do primary care providers know what to ask when looking for sleep disturbances [12–14]? Does the GP have or use appropriate tools when diagnostic investigations such as a polysomnography are not readily available to screen for sleep disturbance? Questionnaires have been shown to be helpful in screening for OSA, although currently there is not an accepted well validated questionnaire suitable for use with children with neurodisability.

As discussed, children with cerebral palsy have a complex sleep picture in which many of the classical diagnostic symptoms, such as fractured sleep, can occur due to non-sleep breathing issues, potentially masking a development such as obstructive sleep breathing. Further research is required to look at the education of parents and primary care givers [12–14]. Current tools such as questionnaires may be under-utilised or not sensitive enough in this population to consistently identify sleep breathing disorders. It may be that increased resources and education from tertiary care centres is required to improve identification of the sleep breathing issues that present in this population.

Alternatively, it may be the patients could be screened using more creative methods. Trained healthcare professionals who attend special needs schools could perform assessments or questionnaires. They already treat these complex children on a regular basis, and could use their medical background to identify and refer to the tertiary centre.

Another option may be to target the parents own sleep routine. Many parents of children with neurodisability report having a poor or disrupted sleep routine, often as a direct result of caring for their child overnight [12, 15, 16]. The effects a child with sleep issues can have on their parents are well described: reduction in quality of life, stress, fatigue and depression [12, 15, 17]. Given the appropriate education, could the parents identify their own sleep issues, which may have been clouding the overall picture in the home. An improved sleep routine would better equip them to tackle their child's sleep-related disturbances and the extra work involved in maximising quality of life for children with neurodisability.

Finally, once the sleep breathing issue has been identified and treatment selected, there is the issue of finding an appropriately sized paediatric mask. The range of masks available is heavily weighted in favour of the adult patient. There is still a shortage of paediatric-sized masks [18]. A poorly fitting mask will always reduce the effectiveness of the therapy and bring adverse effects of discomfort, marking of the face or even skin breakdown. In this case, significant pressures were required to overcome the obstructive episodes. The patient encountered a marked leak and other NIV-associated side-effects. Manufacturers have recently begun to tackle the issue of paediatric-sized masks but progress is slow. We should continue to encourage manufacturers to address properly the issue of paediatric-sized interfaces.

Conclusion

Patients with neurodisability, particularly those at the more severe end of the spectrum, are at increased risk of sleep disturbances. This case illustrates that despite this correlation being known, these patients are still being missed. I hope I have highlighted there is no "quick fix" to what is a complex problem. Multiple elements such as targeted education for clinicians and carers, increased access to clinical and diagnostic tools, and improved screening should be considered to aid future improvement. If we do not ask about sleep in these high-risk patients, we will never find the true numbers affected or be able to offer therapy that can often have a large impact on quality of life for the children and their parents/carers.

Acknowledgements

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Conflict of interest

Disclosures can be found alongside this article at breathe.ersjournals.com

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