Obstructive sleep apnoea syndrome in children

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Obstructive sleep apnoea syndrome in children is characterised by recurrent complete and/or partial upper airway obstruction during sleep. Owing to the limited number of studies, the exact prevalence rate has yet to be determined, but an estimate as high as 1% to 3% has been suggested. Recent studies have shown that obstructive sleep apnoea syndrome in children is different from that in adults in its distinct clinical presentation, diagnostic criteria and treatment. Polysomnography is now recommended as the standard investigation for confirming the diagnosis. The majority of patients may benefit from removal of the enlarged tonsils and adenoids. Although nasal continuous positive airway pressure therapy has been the mainstay of treatment for adults with obstructive sleep apnoea syndrome, experience of its therapeutic effect on children is limited; recent reports are encouraging. Further efforts are needed in advancing both the clinical management of and research into this disorder.

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Introduction

The association between snoring, upper airway obstruction and lethargy has been recognised since the last century. It has also been recognised that upper airway obstruction in children can lead to cor pulmonale which can be reversed upon removal of the obstruction through tonsillectomy. Polysomnography allows the detailed recording of abnormal breathing events during sleep by simultaneously monitoring the electroencephalogram (EEG), electro-oculogram (EOG), electrocardiogram (ECG), electromyogram (EMG), airflow, and chest wall movement. One of the first documented reports of obstructive sleep apnoea syndrome (OSAS) in children was written by Guilleminault in 1979 where he stressed the importance of polysomnographic findings. Since then, although there have been more studies of OSAS in children, many areas of this disorder remain poorly understood. It was only recently that the distinct diagnostic criteria for polysomnographic findings in children were further defined. In addition, the clinical presentation of OSAS in children was found to be quite different from that in adults. In the light of these unique clinical, diagnostic and therapeutic requirements, some authors have recommended that children with OSAS should be regarded as a distinct entity, rather than a junior form of adult OSAS.

OSAS in children is much more common than was once thought. As most symptoms of their condition occur during sleep, it can be easily overlooked. There are a limited number of studies which investigate the prevalence of snoring and OSAS in children. Based on questionnaire studies, up to 7% to 10% of children snore regularly. In a study investigating almost 1000 children in the United Kingdom, subjects in a high-risk group were subjected to home-video and oximetry monitoring and approximately 0.7% were found to have significant sleep-related breathing disorders. Another study in Iceland employing a similar two-stage design with initial questionnaire screening followed by polysomnographic study has estimated that the prevalence of OSAS in children could be as high as 3%. However, larger scale, better designed epidemiological studies are needed to determine the exact prevalence of OSAS in children across different ethnic and age groups.

The most common cause of OSAS in children is adenotonsillar hypertrophy, most cases of which are amenable to surgical treatment. Other conditions include obesity, Down’s syndrome, neuromuscular disease, pharyngeal flap surgery, laryngomalacia, and craniofacial anomalies such as Treacher Collins syn-
Some of these conditions can be extremely difficult to treat and may need expertise from various types of specialist including those from the ear, nose, and throat, maxillofacial surgery, paediatric pulmonology, and sleep departments.

**Definition of apnoea and hypopnoea in children**

Similar to adult OSAS, sleep apnoea in children is defined as being of the central, obstructive or mixed type. Central apnoea occurs when there is complete cessation of airflow at the nose and/or mouth in the absence of any apparent respiratory effort, whereas obstructive apnoea is associated with vigorous respiratory efforts which are ineffective due to lack of airway patency (Figs 1 and 2). Mixed apnoea is diagnosed when both central and obstructive elements occur at the same apnoeic episode without interruption by effective respiration. The definition for pathological

**Fig 1. Polysomnographic tracing for obstructive sleep apnoea in children**

Recording of a 13-month-old boy showing recurrent obstructive sleep apnoea associated with oxygen desaturation and cyclical brady-tachycardias. Calibration: 100 µV/10 mm for 1, 2 and 3; 25 µV/10 mm for 4; 1 MV/10 mm for 5

1 Electroencephalogram 2, 3 Electro-oculogram 4 Electromyogram
5 Electrocardiogram 6 Oxygen saturation 7 Chest impedance
8 Respiratory airflow
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Airflow cessation will depend on age and the type of apnoea. Whereas premature infants can have as long as 20 seconds of central apnoea, much shorter periods of obstructive apnoea (<10 seconds) may be considered abnormal in older infants and children. More important than such an arbitrary time is whether the patient has significant hypoxaemia or hypercapnia or arousal. In contrast to the adult condition in which apnoeic events are predominant, partial airway obstruction is more prevalent in children. However, while there is no consensus on the definition of these hypopnoeic events, it has been suggested that it should be defined as a reduction of 50% or more in the amplitude of the nasal/mouth airflow signal. These children often have increased respiratory efforts during sleep which are frequently accompanied by hypoxaemia, hypercapnia, arousal, and snoring. Oesophageal pressure monitoring may be more sensitive in detecting these respiratory events.

Fig 2. Polysomnographic tracing for central sleep apnoea in children

Recording of a 8-year-old boy showing recurrent central sleep apnoea associated with mild oxygen desaturation. Calibration: 100 µV/10 mm for 1, 2 and 3; 25 µV/10 mm for 4 and 6

1 Electroencephalogram 2, 3 Electro-oculogram 4 Electromyogram (chin)
5 Oxygen saturation 6 Electromyogram (leg) 7 Chest impedance
8 Respiratory airflow
Clinical features

In contrast to the adult condition, OSAS in children has an equal gender prevalence. The peak incidence is from ages 2 to 5 years, probably related to prominent lymphoid tissues of the upper airway and frequent inflammation in this age group. A careful history and detailed physical examination are the first steps in the diagnosis of OSAS. The two most common symptoms are snoring and difficulty in breathing during sleep. The snoring may be loud and harsh and be audible even in the next room. The parents may notice heaving of the chest wall, cessation of airflow and mouth breathing. In severe cases, the child may need to be woken up to re-establish breathing during some of these episodes. The child may sleep restlessly with neck extension or in a prone position with knees under the abdomen to relieve the airway obstruction. Excessive sweating and enuresis have been reported. However, the excessive daytime sleepiness which is a typical symptom of adult OSAS, is only seen in a third of these children. Behavioural, temperamental and learning difficulties have been reported, but their exact prevalence is unclear. Chronic rhinorrhoea is not uncommon. In younger children and infants, OSAS may present as failure to thrive. In extreme cases, cor pulmonale may be the presenting problem. Physical examination may reveal hyponasal voice, mouth breathing, enlarged tonsils, and pectus excavatum. Viral respiratory infection and allergic rhinitis will markedly exacerbate OSAS symptoms in affected children (Box).

Whereas obesity is commonly associated with adult OSAS, few children with OSAS are found to be obese. In fact, both low and increased prevalence of OSAS have been reported in obese children. Further study is clearly needed to clarify the relationship between OSAS and obesity in children, as obesity is becoming a major health problem in affluent societies such as Hong Kong.

OSAS in children may result in serious and potentially lethal complications. Recurrent episodes of hypoxaemia and hypercapnia may lead to cor pulmonale. As many as one half of subjects may have radiological and/or electrocardiographic evidence of right ventricular hypertrophy. Cardiac arrhythmia including sinus arrest, second degree heart block, and paradoxic tachycardia have been reported as being common in children with OSAS, but a negative study has also been reported recently. Systemic hypertension and left ventricular hypertrophy have also been reported as being associated with OSAS in children. As discussed above, some of these children may develop growth failure and neuropsychiatric complications such as behavioural, temperamental and learning difficulties. More importantly, these complications are potentially reversible upon relief of the upper airway obstruction. In some cases, cor pulmonale may be the presenting problem. If the condition is not recognised, however, these complications may progressively worsen and it is possible that sudden death may ensue. Although, the relationship between childhood OSAS and sudden infant death syndrome (SIDS) is not at all clear, recent family studies have suggested aggregation of OSAS in adults and near miss SIDS or SIDS.

Diagnosis

The use of questionnaires in diagnosing OSAS in children is limited due to difficulty in differentiating OSAS from primary snoring. To date, the most important and reliable diagnostic tool for childhood OSAS is the standardised polysomnographic study. This includes a multichannel (usually eight to 16) polysomnography with video camera recording, and consists of EEG, EOG, chin EMG, ECG, oxygen saturation monitoring, and assessment of oro-nasal airflow, chest and abdominal wall movement, and leg movement. Transcutaneous or end-tidal carbon dioxide (ETCO₂) may be useful in the assessment of the partial airway obstruction, increased physiological dead space, tachypnoea, and hypoventilation. In selected cases, oesophageal pressure monitoring has been recommended for detecting hypopnoea and increased respiratory efforts in the absence of airflow cessation. Oesophageal pH monitoring may be helpful in
assessing gastro-oesophageal reflux and aspiration, and its temporal relationship with apnoea or hypoxaemia.\textsuperscript{32} The use of oesophageal monitoring is, however, limited by its irritating effect on children and it is not recommended for routine use. Polysomnography can be performed as an overnight study that approximates the child’s usual sleep, or as a nap study in daytime for selected cases.\textsuperscript{24} The nap study should last at least 2 hours and include at least one period of rapid eye movement (REM) sleep. A positive nap study correlates well with the findings of an overnight study, but a normal nap study cannot exclude the presence of obstructive sleep apnoea.\textsuperscript{33}

Guilleminault introduced the term ‘sleep apnoea’ when he defined the condition as 30 or more apnoeic episodes of at least 10 seconds’ duration during a 7-hour sleep.\textsuperscript{34} Based on adult data, an apnoea index (number of apnoeas per hour) of ≥5 was initially proposed as the criteria for obstructive sleep apnoea in children, but it has become clear that adult criteria will fail to identify most cases of childhood OSAS.\textsuperscript{6,7} Rosen et al\textsuperscript{7} analysed a polysomnographic study of 20 children of age 8 months to 16 years with clinical evidence of upper airway obstruction during sleep together with episodes of hypoxaemia and hypercapnia. The mean apnoea index was only 1.9 and just three children had apnoea indices of ≥5. Marcus et al\textsuperscript{6} further defined normal polysomnographic values in children after studying 50 healthy children and adolescents aged 1 to 18 years. These children had a mean apnoea index of 0.1, and central apnoea was found to be frequent, although most cases were not associated with oxygen desaturation. This is further complicated by the fact that partial rather than complete airway obstruction is much more common in children. Abnormalities in gaseous exchange and paradoxical respiratory efforts would be better and more sensitive indices in diagnosing OSAS in children. Based on the available data, most authors and the diagnostic guidelines proposed by the American Thoracic Society recommend that normal polysomnographic values for children should have an apnoea index of less than 1.0.\textsuperscript{10,24} In addition, criteria measuring the arterial oxygen percent saturation (\textit{SAO2}) and \textit{ETCO2} may be useful in determining borderline cases. The oxygen desaturation index, defined as greater than 4% of \textit{SAO2} desaturation per hour of total sleep time, should be less than 1.4. No normal child should have a peak \textit{ETCO2} >53 mm Hg, or \textit{ETCO2} >45 mm Hg for more than 60% of total sleep time, or \textit{ETCO2} >50 mm Hg for >8% of total sleep time.\textsuperscript{5,24} As occasional central apnoea is commonly seen in normal infants and children, it has been suggested that central apnoea in children should be considered as abnormal if they are associated with \textit{SAO2} >4% or bradycardia, irrespective of the length of apnoea.\textsuperscript{24} Non-sigh and non-movement associated central apnoeas should be regarded as abnormal, if they are of greater than 20 seconds’ duration.\textsuperscript{24}

Apart from a comprehensive polysomnographic study, other investigations may also be helpful in the management of obstructive sleep apnoea. A chest X-ray and an ECG are needed to exclude right ventricular hypertrophy and cor pulmonale. A lateral neck X-ray may be helpful in checking airway obstruction, especially by enlarged tonsils and adenoids. Laryngoscopy can be performed to evaluate dynamic upper airway obstruction and adenoid hypertrophy. Cephalometric radiography has also been used to evaluate the nature of airway obstruction and facial bone abnormality in craniofacial anomalies.\textsuperscript{22}

**Pathophysiology**

The pathophysiology of upper airway obstruction in infants and children is complex and not completely understood. The paediatric airway is more susceptible to obstruction due to its smaller calibre, and decreased muscular tone and the high position of the larynx.\textsuperscript{23} Obstructive sleep apnoea occurs when the upper airway collapses during respiration. The most easily collapsible region in the human infant is at the entrance to the larynx, followed by the level at the aryepiglottic folds.\textsuperscript{33} Mechanical support for these compliant areas of the upper airway is derived from the muscles surrounding the pharynx. Such muscle tone has been shown to prevent airway collapse during neck flexion as well as during inspiration.\textsuperscript{35,36} Contraction of the genioglossus and geniohyoid muscles of the tongue also dilate the upper airway and make it more rigid and resistant to collapse by negative pressure.\textsuperscript{36,37} Pharyngeal airway maintenance thus depends on a dynamic balance between diaphragmatic contractile force, upper airway resistance (airway suction force), and the contractile force of the airway dilating muscles (airway patency force). Both functional and anatomical factors may tilt the balance.

Sleep is the most obvious functional factor that predisposes to airway obstruction. This is thought to be due to a reduction in airway muscular activity during sleep, particularly during REM sleep. Arousal from sleep has a potent stimulatory effect on the upper airway dilating muscles. This is thought to be the predominant factor responsible for spontaneous termination of obstructive apnoeic episodes. Drugs
such as narcotics, sedatives, and alcohol, and brainstem lesions, such as Arnold Chiari malformation, may exacerbate or lead to OSAS by depressing airway maintaining activity.

The leading cause of obstructive sleep apnoea in children is a prominence of lymphoid tissue which increases the upper airway resistive load. Narrowing of the aperture of the nasal or pharyngeal airway as seen in various craniofacial syndromes may displace the tongue posteriorly and result in an obstruction. Neuromuscular disease may cause hypotonia of the pharyngeal muscles and reduce airway patency. Obesity increases pharyngeal resistance and may decrease thoracic compliance through mass loading of the pharynx, thorax and abdomen. Neck flexion may also predispose to airway closure.

Some adults with OSAS have been shown to have abnormal ventilatory responses during wakefulness, but a similar ventilatory dysfunction is more doubtful in children. A recent study of 20 children with OSAS did not confirm the abnormal ventilatory responses by rebreathing techniques. However, the limited number of subjects and relatively milder cases included in this study precluded any conclusive statements.

Preliminary studies have suggested the familial aggregation of OSAS in children. A familial factor may predispose children to develop OSAS by affecting the upper airway resistive loads through craniofacial abnormalities and/or probable ventilatory control dysfunctions. More studies are needed, however, to confirm this familial tendency and any underlying pathophysiological mechanisms.

**Treatment**

The optimal treatment strategy for paediatric OSAS remains unresolved. Since enlarged tonsils and adenoids are a major cause of OSAS in children, the surgical removal of these can be curative. This may even be helpful for selected patients with other causes such as craniofacial abnormalities or obesity. Preoperative sedation should be carefully administered in severely obstructed cases and close monitoring should be implemented post-operatively, as oedema and sedation may also aggravate airway obstruction in the first 24 hours after surgery. It may take up to 6 weeks for symptoms to resolve. In some patients whose adenotonsillectomy cannot be immediately performed, overnight supplemental oxygen may be a safe and effective temporary treatment for OSAS. Overnight oxygen has also been shown to significantly improve nocturnal oxygenation, and to decrease apnoeas, hypopnoeas, and paradoxical breathing in a group of 16 children aged 2 to 8 years with mild to moderate OSAS. There was no definite deleterious effect of nocturnal oxygen on the ventilation drive in these children, which has been reported in adults with severe OSAS.

The use of nasal continuous positive airway pressure (nCPAP) can be effective for those who do not respond to adenotonsillectomy, for complicated cases such as craniofacial anomalies, or as an interim measure in infants and young children to enable a better planning of surgery when they are older. While experience in infants and children is limited, recent reports are encouraging with a success rate of up to 90%. The mean level of nCPAP pressure was similar to that of adult OSAS and pressure levels ranging from 4 cm to 20 cm H2O have been reported. The pressure requirements change with growth, and periodic reassessments are needed to adjust the pressure. It has been recommended that the pressure should be set to maintain the SAO2 above 95%, or such that the occurrence of both apnoeas and hypopnoeas is prevented, although further research is needed to establish a better criterion for selecting the optimal pressure. In some specialist centres, oesophageal pressure monitoring has been recommended as an additional criterion in titrating the nCPAP pressure. Failure is usually due to poor tolerance, but hypventilation during high nCPAP pressure has also been reported. This is thought to be related to the rebound in REM sleep upon immediate institution of nCPAP in severe cases.

Commercially available masks are suitable for most children, but special custom-made masks may be needed for younger infants. Periodical adjustments of the size of the nasal masks are needed for growing children. Careful explanation and instruction on the use of nCPAP with parental involvement are important to ensure successful trials. Long-term follow-up data on the use of nCPAP in childhood OSAS have recently become available in two specialised sleep centres in United States and Australia and suggest that this is a safe and well-tolerated treatment. There have been no reports of retardation of facial and maxillary growth in children regularly using nCPAP. Recently, the bilevel positive airway pressure (BiPAP) system also has become available to children, but further studies are needed to determine whether BiPAP will really improve tolerability and compliance. Our centre has begun to employ nCPAP for infants and children with OSAS related to craniofacial anomalies such as Treacher Collins syndrome and Crouzon’s disease.
and the results have been encouraging. Surgical correction of these craniofacial anomalies can then be planned at a later stage when the children are older. The use of nCPAP in these cases will buy time for optimising the surgery schedule and avoiding the necessity for tracheostomy.10,42 For example, choanal stenosis can be widened, mandibular advancement can be performed for Treacher Collins syndrome or Pierre Robin syndrome, and maxillary bone advancement can be carried out for Crouzon’s disease. Again, postoperative airway oedema may occur and these children need to be closely monitored in the immediate postoperative period.

Uvulopalatopharyngoplasty has been used as a mode of treatment in adult cases, but experience in infants and children is limited.42 Furthermore, there is uncertainty about the long-term effectiveness of this treatment in adults with OSAS. Tracheostomy should be the last resort if other modes of treatment fail. The tracheostomy site needs constant care, and the operation carries a definite mortality risk in infants and children. Repeat sleep studies should be performed after surgical treatment or after nCPAP to determine the effectiveness of the treatment modality.

Conclusion

The understanding and knowledge of childhood OSAS is evolving rapidly. It is a common form of paediatric sleep disorder with significant associated morbidity and mortality. Locally in Hong Kong, there is a dearth of data, highlighting the need to study this problem, especially in our paediatric population. Proper recognition of this condition is important to ensure effective management. Polysomnographic study requires expertise in the field and good technical support. It will require contributions from a multidisciplinary team including sleep specialists, ear, nose, and throat surgeons, paediatric pulmonologists, and maxillofacial surgeons. Further research is clearly needed in clarifying the diagnostic criteria, management and the prognosis of this common but frequently underdiagnosed sleep disorder.

References


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