BEST PRACTICE

SLEEP APNOEA

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In recent years there has been a dramatic increase in the recognition of sleep related breathing disorders in children, and a consequent but less dramatic increase in the availability of specialist provision for the diagnosis and treatment of such problems. However, the clinical awareness of these problems and their possible consequences remains patchy. The purpose of this article is to review the causes and consequences of sleep apnoea in children, to guide clinicians in the recognition of those children who need assessment, and to outline the management options.

Sleep apnoea in children may include obstructive sleep apnoea or hypopnoea and sleep apnoea or hypoventilation due to central causes. By far the most common of these pathologies is obstructive sleep apnoea. Obstructive sleep apnoea has been defined as a "disorder of breathing during sleep characterised by prolonged partial upper airway obstruction and/or intermittent complete obstruction that disrupts normal ventilation during sleep and normal sleep patterns".[1] In this article we will concentrate on obstructive airway problems leading to sleep apnoea, although high risk groups with mixed pathology will be discussed. We will not deal with purely central causes of sleep apnoea or hypoventilation, such as congenital central hypoventilation syndrome, nor with apnoea or apparently life threatening events of infancy.

AETIOLOGY

Apnoea or pathological hypoventilation during sleep may result from a decrease in central drive, a reduction in muscle power, or most commonly from upper airway obstruction.

The newborn infant spends 16–18 hours of the day asleep, and the majority of this is rapid eye movement (REM) sleep. The pharyngeal airway is maintained by mental and submental muscles such as genioglossus, and these relax during sleep, facilitating upper airway collapse. During REM sleep these effects are most pronounced, and intercostal muscles are also relaxed, leading to a reliance on diaphragmatic breathing and paradoxical inward movement of the chest wall during inspiration. Other consequences of REM sleep are a reduced and less stable functional residual capacity, reduced minute ventilation, and an increased hypoxic arousal threshold. The stage is therefore set for airway and breathing problems during sleep, which are likely to be particularly severe in REM sleep.[2]

Although airway obstruction is exacerbated by muscle tone changes during sleep, there are usually underlying anatomical factors which predispose to the problem in the first place. These may include craniofacial abnormalities, obesity, and adenotonsillar hypertrophy. The presence of neuromuscular disease increases the risk of sleep related breathing disorders including sleep apnoea. Underlying conditions which predispose to obstructive sleep apnoea are listed in table 1.

Often airway obstructions occur in series, when the increased inspiratory pressure required at each obstruction will enhance the degree of collapse at more proximal narrowings, so that the effects of several obstructions in series may be greater than the sum of their parts.

There is evidence that obstructive sleep apnoea and infant apnoeas have common familial factors, which may be related to both upper airway anatomy and to control of muscle tone and breathing during sleep.[3] It has therefore been hypothesised that children with obstructive sleep apnoea are at higher risk of similar recurrent problems later in life.

Two problems make identification of the problem more difficult. Firstly, some children may have gas exchange abnormalities during sleep without frank obstructive sleep apnoea, but with a pattern of obstructive hypoventilation. Secondly some children will have symptoms from upper airway obstruction that are not associated with demonstrable gas exchange abnormalities but are caused by frequent arousals and fragmentation of sleep. Both are more difficult to demonstrate or characterise on physiological monitoring.

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EPIDEMIOLOGY

One of the major challenges in delineating sleep related breathing disorders (SRBD) is the definition of abnormality. Normal reference values for apnoea in childhood have been published, and differ from those used in adults.[4] From these data we can state that it is abnormal for a child over the age of 1 year to have obstructive apnoea lasting longer than 10 seconds, or to desaturate

Table 1 Causes of obstructive sleep apnoea

- Adenotonsillar hypertrophy
- Obesity
- Down syndrome*
- Craniofacial abnormalities*
  - poorly formed jaw (e.g. Pierre Robin sequence, Treacher Collins syndrome)
  - midfacial hypoplasia (e.g. Apert syndrome, Crouzon syndrome)
- Achondroplasia
- Mucopolysaccharidoses
- Prader-Willi syndrome
- Neuromuscular disease*
- Cerebral palsy

*Obstruction may evolve in very early infancy.

below 90% during sleep. On the other hand, obstructive apnoeas occur to a greater or lesser extent in 18% of normal children and we do not know the exact cut off which predicts pathological consequences. Various authors have documented obstructive sleep apnoea causing gas exchange abnormalities to occur in 0.7–2.9% of pre-pubertal children.[5–7]

Habitual snoring occurs in 12% of 4–5 year old children in the UK[5] and in 10% of 8–10 year old German children.[8] Recent studies have suggested that snoring in primary school children is a predictor of later academic performance.[9] Although abnormal gas exchange has usually been taken to indicate a significant abnormality, a large population based survey indicated that snoring is a better predictor than nocturnal hypoxaemia for poor academic performance.[8] We do not yet know which physiological abnormalities are the best predictors of a good response to treatment.

In high risk children the risk of sleep apnoea or other sleep related breathing disorders is much higher and warrants routine evaluation even in the absence of obvious symptoms, since snoring and poor performance are common in many of the conditions listed.

CONSEQUENCES AND COMPLICATIONS

As discussed above, there is considerable evidence that sleep apnoea is associated with worse academic performance, concentration, and behaviour. This may be due to sleep fragmentation, and although intervention may improve behaviour and concentration, effects on performance may be seen long after the symptoms have disappeared. Other consequences of sleep apnoea include failure to thrive, systemic hypertension with increased left ventricular mass, cor pulmonale, and pulmonary oedema. All of these complications reverse when the underlying problem is treated. Pulmonary hypertension may rarely complicate sleep apnoea in children, and may be irreversible. There are occasional reports of coma and death resulting from undiagnosed obstructive sleep apnoea, though this is an extremely rare complication. Children with obstructive sleep apnoea have increased health care utilisation even after allowing for the direct management of the sleep apnoea, suggesting that it may act as an amplifier of other health problems.

IDENTIFICATION, ASSESSMENT, AND DIFFERENTIAL DIAGNOSIS

Excessive daytime sleepiness is less common in children with SRBD than it is in adults. In children without underlying disorders, the presence of nocturnal obstructive symptoms is usual if obstructive sleep apnoea is present. However, symptoms have poor specificity, and many children with symptoms will not have polysomnographic abnormalities. Symptoms and signs which occur more commonly in obstructive sleep apnoea are listed in table 2.

Table 2 Symptoms and signs seen more commonly in obstructive sleep apnoea

Night time
- Snoring and snorting
- Gasps or witnessed apnoeas
- Restlessness and unusual sleep postures (e.g. extended neck)
- Laboured breathing
- Early morning headache
- Excessive sweating
- Enuresis

Daytime
- Irritability, behavioural problems
- Poor concentration
- Failure to thrive
- Upper airway obstruction/mouth breathing
- Harrison's sulci

In children with suggestive symptoms, further physiological assessment may be indicated. A number of end points can be measured, as described below.

Oxygenation

The simplest assessment method is overnight pulse oximetry. This can be done in the home, ensuring a more typical night's sleep compared to a hospital study. Several pulse oximeters now have storage and analysis software available to enable quantification of hypoxaemic dips. Dips in saturation of $>4\%$ from baseline and falls below 90% saturation can be quantified and compared to normal values.[10] Abnormal oximetry is specific for SRBD, but normal oximetry does not exclude sleep apnoea on polysomnography. Figure 1 shows oximetry recordings on a 2 year old child with failure to thrive from obstructive sleep apnoea. Note the normalisation of oximetry after adenotonsillectomy, and the lower heart rate during sleep.

Respiratory events

A more detailed method of assessment is limited polysomnography, measuring heart rate, respiratory effort, and airflow and movement as well as oximetry and a measure of arterial pCO_2. This can give a more detailed estimate of impaired gas exchange, and differentiate between central and obstructive events. This sort of cardiorespiratory assessment is imperative if the clinical problem is not a straightforward obstructive sleep apnoea requiring adenotonsillectomy, but it does not help in the assessment of electroencephalogram (EEG) arousals without impairment of gas exchange. Arousals associated with significant movement can, however, be seen. It has the advantage that it is relatively easy to set up, is not expensive, and has more potential to be performed in the child's home.

Arousals

More detailed polysomnography includes limited EEG, electro-oculogram, and submental electromyogram to allow sleep staging and a measure of neurological arousals resulting from respiratory events, but this is technically demanding to set up and score, and there are relatively few

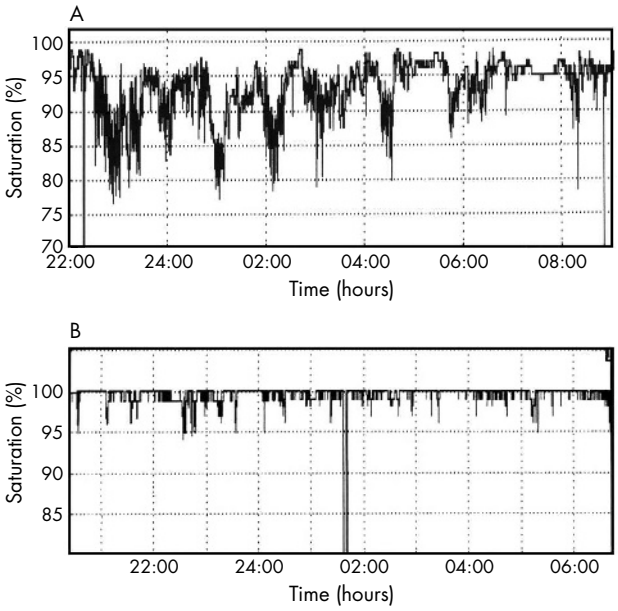

Figure 1 Oximetry recordings on a 2 year old boy with failure to thrive caused by obstructive sleep apnoea. (A) before adenotonsillectomy. (B) Two weeks after surgery.

paediatric facilities in the UK which offer this investigation. Behavioural arousals can be estimated from movement and video. Recently the pulse transit time (PTT) has been used to assess arousals. PTT is the time between the QRS complex on the electrocardiogram (ECG) and the peak of the plethysmographic waveform on the pulse oximeter, and is inversely related to peripheral vasomotor tone. A fall in PTT can be used as a measure of autonomic arousal and may be even more sensitive than the EEG arousal at detecting obstructive events.[11]

High risk children

In high risk children some form of regular screening for sleep apnoea is imperative. Almost all children with Down syndrome have a degree of sleep related breathing disorder,[12] and these children are at particular risk of pulmonary hypertension even with trivial cardiac lesions, so every effort should be made to prevent nocturnal hypoxaemia. Children with Down syndrome or craniofacial problems should have some assessment of gas exchange during sleep in infancy and thereafter at least every 6–12 months until the age of maximum adenotonsillar growth at 5–7 years. Children with Duchenne muscular dystrophy are at particular risk of sleep related breathing problems, especially when they become wheelchair dependent, and they should have assessment of gas exchange (preferably including capnography) from this age onwards on at least a yearly basis. Other neuromuscular conditions may also lead to sleep hypoventilation, especially if there is diaphragmatic involvement, and clinicians should have a low threshold for investigation.

MANAGEMENT

Sleep apnoea without an underlying condition

In nearly all cases this will be due to adenotonsillar hypertrophy. The presence of nocturnal hypoxaemia or of complications such as failure to thrive, cardiac effects, or chest deformity mandate urgent treatment in the form of adenotonsillectomy. Adenotonsillar size alone is not a good indicator of obstruction or the response to surgery. It is important that both tonsils and adenoids are removed, as adenoidectomy alone or adenomonotonsillectomy are associated with a higher failure rate.[13] In children with significant hypoxaemia at night there is an increased risk of perioperative complications,[14] and overnight oximetry should be performed as a preoperative screen to identify these high risk children. A decision pathway is shown in fig 2.

Nasal steroids have been used as a short term measure to improve overnight airway patency in adenotonsillar hypertrophy,[15] but there are no studies of their long term efficacy.

Oral jaw positioning devices have been shown to improve airway obstruction in children with malocclusion[16] but are not widely used in the UK.

In children with symptoms suggesting sleep apnoea but with normal oximetry the issues are much less straightforward. On the one hand there may be sleep fragmentation with impacts on learning and behaviour as well as growth, but on the other hand it is clearly undesirable to subject a child to unnecessary adenotonsillectomy when the problem may be self limiting. More detailed assessment of the frequency of arousals and severity of sleep related breathing disorder is indicated in these cases using either limited or full polysomnography, but it should be reiterated that we do not yet have good evidence about which abnormalities predict a good outcome from intervention.

If sleep apnoea persists despite surgical treatment, full polysomnography should be undertaken in a specialist centre. This situation is unusual, and we consider that it warrants detailed evaluation before committing a child to further intervention. Nasal continuous positive airway pressure (CPAP) or bi-level positive airway pressure (BIPAP) ventilation at night may be necessary.

Sleep apnoea with an underlying condition

In most cases of upper airway obstruction caused by an underlying condition, adenotonsillectomy should still be considered as first choice treatment, but it is less reliably effective.

Down syndrome disturbs breathing during sleep for several reasons. The low muscle tone predisposes to upper airway collapse; a large tongue tends to impede the oropharyngeal airway; midfacial hypoplasia reduces the nasal airway diameter; and the recurrent upper respiratory infections further impede nasal airway patency. To compound matters these children often have increased pulmonary blood flow because of congenital heart disease and are at particular risk of pulmonary hypertension.[17] In children with Down syndrome who do not respond to adenotonsillectomy, craniofacial or tongue surgery may be considered. Nasal CPAP is helpful in these cases, but may be difficult to achieve in practice. If there is concern about hypoxaemia leading to pulmonary hypertension then supplemental oxygen at night may be an effective palliative measure, although it does not correct the underlying problem. Oxygen supplementation may cause worse sleep apnoea in some children and should only be used if polysomnography shows that it does not affect breathing control.[18]

Children with obesity and sleep apnoea persisting after adenotonsillectomy often need nasal CPAP or BIPAP, as weight reduction can rarely be achieved in the time scale needed.

The management of obstructive sleep apnoea in craniofacial problems is ideally the correction of the craniofacial

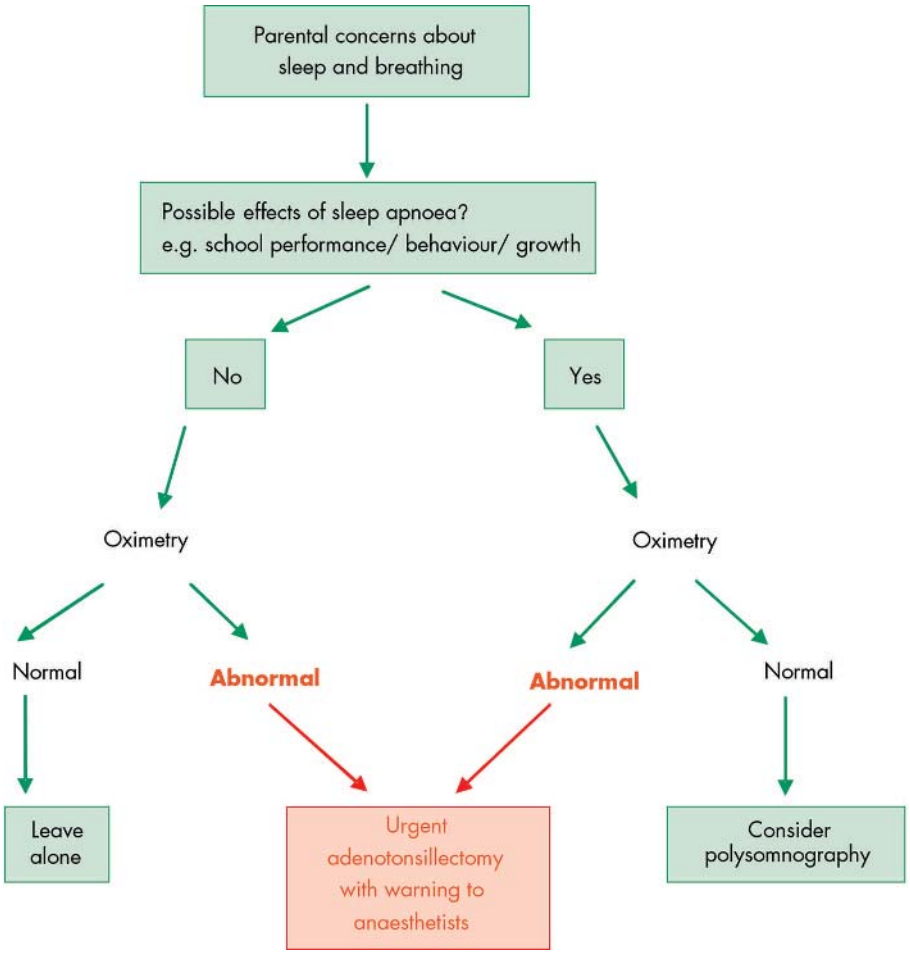

Figure 2 Decision pathway for the assessment of a child presenting with snoring, with no underlying high risk condition.

problem. In children with Pierre Robin sequence the placement of a nasopharyngeal airway can avoid tracheostomy and enable safe home management until the airway grows sufficiently.[19] In other craniofacial problems, if corrective surgery is not immediately possible then adenotonsillectomy is still helpful. Nasal CPAP remains an option, but is less effective, and tracheostomy may be necessary until correction can be achieved.

One of the most problematic groups of children with sleep related breathing disorders are those with cerebral palsy. Poor midfacial growth coupled with abnormal airway tone and bulbar dysfunction can lead to obstruction which is difficult to treat. Oral jaw positioning devices may have a role in this group. If adenotonsillectomy is ineffective then more aggressive treatment can pose both clinical and ethical problems.

Children with neuromuscular disease who have sleep related breathing disorders will almost always require overnight BIPAP. This has been shown to improve life expectancy dramatically in Duchenne muscular dystrophy in the presence of respiratory failure,[20 21] and is generally very well tolerated. It should be remembered that overnight respiratory support is only part of a package of interventions needed to

Sleep apnoea: key points

- Sleep apnoea and hypoventilation problems are common in childhood, with 10–12% of children habitually snoring and 0.7–2.9% showing significant gas exchange abnormalities
- Consequences of sleep apnoea include learning and behaviour problems, failure to thrive and life threatening cardiorespiratory effects
- The presence of symptoms is sensitive but not specific as an indicator of physiological derangement
- Overnight oximetry is specific but not sensitive as an indicator of physiological derangement
- Adenotonsillectomy is the first line treatment of obstructive sleep apnoea and is usually curative in uncomplicated disease
- Overnight oximetry should be performed in any child where adenotonsillectomy is planned for obstructive sleep apnoea
- Children with underlying conditions which predispose to sleep related breathing disorders should be screened with oximetry ± capnography on a regular basis
- If adenotonsillectomy is not effective or feasible then other treatments including nasal CPAP or BIPAP should be considered. These will require specialist referral

protect the respiratory health of children with neuromuscular disease; other measures include chest physiotherapy with assisted coughing, scoliosis correction, and prevention of reflux and aspiration.[22]

GUIDELINES, EVIDENCE, AND FUTURE RESEARCH

The American Academy of Pediatrics has produced a clinical practice guideline and technical report on the diagnosis and management of childhood obstructive sleep apnoea.[23 24] These guidelines are based on a systematic approach to the literature and formal grading of evidence. They have an emphasis on the gold standard of polysomnography in the diagnosis of obstructive sleep apnoea, which is somewhat tautological, since by definition the condition requires polysomnography to quantify it.

A Cochrane review of the role of adenotonsillectomy in obstructive sleep apnoea in children concluded that there were no acceptable randomised controlled trials supporting this intervention.[25] There are, however, a large number of observations of dramatic physiological and clinical improvement after intervention. The key question which remains unanswered is what degree and what type of physiological derangement is clinically important, and sufficient to warrant intervention.

CONTENTIOUS ISSUES

In addition to the major issue of not knowing what level or type of physiological derangement is an indication for intervention (discussed above), there are other contentious issues in this field.

Assessment of arousals

There are several methods of assessing arousals, described above. It is unclear which of these will be the best predictor of response to treatment.

When is a full polysomnography indicated?

The advantages of a full polysomnogram, including neurophysiological measures, is that it can be used to assess sleep architecture, including arousals and presence or absence of REM sleep. Since sleep apnoea may occur only during REM, this may be important in interpreting a negative study. If a child is referred because of daytime sleepiness or other symptoms thought to be consequent of sleep deprivation, then the sleep architecture may add useful information, particularly if no major breathing problem is found. On the other hand, it is expensive of resources, limited in availability, and much less physiological than a home study of limited cardiorespiratory parameters. Since the majority of questions can be answered with limited studies, we would recommend that full polysomnography is reserved for children with atypical symptoms, where there is continuing uncertainty after simple testing, or where initial surgical treatment of presumed obstructive sleep apnoea has been ineffective.

RECOMMENDED READING

- American Academy of Pediatrics. Clinical practice guideline: diagnosis and management of childhood obstructive sleep apnea syndrome. *Pediatrics* 2002;**109**:704–12.
- American Thoracic Society. Standards and indications for cardiopulmonary sleep studies in children. *Am J Respir Crit Care Med* 1996;**153**:866–78.

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REFERENCES

1 **American Thoracic Society**. Standards and indications for cardiopulmonary sleep studies in children. *Am J Respir Crit Care Med* 1996;**153**:866–78.
2 **Gaultier C**. Cardiorespiratory adaptation during sleep in infants and children. *Pediatr Pulmon* 1995;**19**:105–17.
3 **McNamara F**, Sullivan C. The genesis of adult sleep apnea in childhood. *Thorax* 2000;**55**:964–9.
4 **Marcus CL**, Omlin KJ, Basinki DJ, *et al*. Normal polysomnographic values for children and adolescents. *Am Rev Respir Dis* 1992;**146**:1235–9.
5 **Ali NJ**, Pitson DJ, Stradling JR. Snoring, sleep disturbance, and behaviour in 4–5 year olds. *Arch Dis Child* 1993;**68**:360–6.
6 **Brunetti L**, Rana S, Lospalluti ML, *et al*. Prevalence of obstructive sleep apnea syndrome in a cohort of 1,207 children of southern Italy. *Chest* 2001;**120**:1930–5.
7 **Gislason T**, Benediktsdottir B. Snoring, apneic episodes, and nocturnal hypoxemia among children 6 months to 6 years old. An epidemiologic study of lower limit of prevalence. *Chest* 1995;**107**:963–6.
8 **Urschitz MS**, Guenther A, Eggebrecht E, *et al*. Snoring, intermittent hypoxia and academic performance in primary school children. *Am J Respir Crit Care Med* 2003;**168**:464–8.
9 **Gozal D**, Pope DWJ. Snoring during early childhood and academic performance at ages thirteen to fourteen years. *Pediatrics* 2001;**107**:1394–9.
10 **Urschitz MS**, Wolff J, von Einem V, *et al*. Reference values for nocturnal home pulse oximetry during sleep in primary school children. *Chest* 2003;**123**:96–101.
11 **Katz ES**, Lutz J, Black C, *et al*. Pulse transit time as a measure of arousal and respiratory effort in children with sleep-disordered breathing. *Pediatr Res* 2003;**53**:580–8.
12 **Marcus C**, Keens T, Bautista D, *et al*. Obstructive sleep apnea in children with Down syndrome. *Pediatrics* 1991;**88**:132.
13 **Shintani T**, Asakura K, Kataura A. The effect of adenotonsillectomy in children with OSA. *Int J Pediatr Otorhinolaryngol* 1998;**44**:51–8.
14 **Brown KA**, Morin I, Hickey C, *et al*. Urgent adenotonsillectomy: an analysis of risk factors associated with postoperative respiratory morbidity. *Anesthesiology* 1982;**99**:586–95.
15 **Brouillette RT**, Manoukian JJ, Ducharme FM, *et al*. Efficacy of fluticasone nasal spray for pediatric obstructive sleep apnea. *J Pediatr* 2001;**138**:838–44.
16 **Villa MP**, Bernkopf E, Pagani J, *et al*. Randomized controlled study of an oral jaw-positioning appliance for the treatment of obstructive sleep apnea in children with malocclusion. *Am J Respir Crit Care Med* 2001;**165**:123–7.
17 **Jacobs IN**, Teague WG, Bland JW Jr. Pulmonary vascular complications of chronic airway obstruction in children. *Arch Otolaryngol Head Neck Surg* 1997;**123**:700–4.
18 **Marcus CL**, Carroll JL, Bamford O, *et al*. Supplemental oxygen during sleep in children with sleep-disordered breathing. *Am J Respir Crit Care Med* 1995;**152**:1297–301.
19 **Masters I**, Chang A, Harris M, *et al*. Modified nasopharyngeal tube for upper airway obstruction. *Arch Dis Child* 1999;**80**:186–7.
20 **Simonds AK**, Muntoni F, Heather S, *et al*. Impact of nasal ventilation on survival in hypercapnic Duchenne muscular dystrophy. *Thorax* 1998;**53**:949–52.
21 **Eagle M**, Baudouin SV, Chandler C, *et al*. Survival in Duchenne muscular dystrophy: improvements in life expectancy since 1967 and the impact of home nocturnal ventilation. *Neuromuscular Disorders* 2002;**12**:926–9.
22 **Wallgren-Pettersson C**, Bushby K, Mellies U, *et al*. 117th ENMC workshop: ventilatory support in congenital neuromuscular disorders – congenital myopathies, congenital muscular dystrophies, congenital myotonic dystrophy and SMA (II) 4–6 April 2003, Naarden, The Netherlands. *Neuromuscular Disorders* 2004;**14**:56–69.
23 **American Academy of Pediatrics**. Clinical practice guideline: diagnosis and management of childhood obstructive sleep apnea syndrome. *Pediatrics* 2002;**109**:704–12.
24 **Schechter MS**. Technical report: diagnosis and management of childhood obstructive sleep apnea syndrome. *Pediatrics* 2002;**109**:e69.
25 **Lim J**, McKean M. Adenotonsillectomy for obstructive sleep apnoea in children. *Cochrane Database of Systematic Reviews* 2004;**1**.