

# **Working Party on Sleep Physiology and Respiratory Control Disorders in Childhood**

## **Standards for Services for Children with Disorders of Sleep Physiology**

EXECUTIVE SUMMARY

September 2009



**Royal College of Paediatrics and Child Health**



# CONTENTS

## Foreword

<b>1.</b>	<b>Introduction</b>	<b>3</b>
1.1	Justification and remit	3
1.2	Developmental physiology	5
1.3	Behavioural sleep problems	6
1.4	General effects of sleep impairment	6
<b>2.</b>	<b>Methodology of assessment</b>	<b>7</b>
2.1	Adequacy of ventilation	7
2.2	Evaluation of respiratory disturbance	7
2.3	Assessment of cardiac rate/rhythm	8
2.4	Assessment of sleep state/arousals	8
2.5	Other measurements	8
2.6	Interpretation	8
2.7	Methodology of studies	8
<b>3.</b>	<b>Airway and breathing problems during sleep</b>	<b>10</b>
3.1	Obstructive sleep apnoea (OSA) and hypoventilation	10
3.1.1	Prevalence	10
3.1.2	Presenting features	10
3.1.3	Consequences	10
3.1.4	Identification	10
3.1.5	Assessment	11
3.1.6	Management	11
3.2	Specific conditions at high risk of SRBD	13
3.2.1	Down's syndrome	13
3.2.2	Neuromuscular Disease	14
3.2.3	Anatomical abnormalities	16
3.3	Congenital Central Hypoventilation Syndrome	18
<b>4.</b>	<b>Unexplained events in infancy – ALTE</b>	<b>19</b>
4.1	Specific conditions and ALTE	20
4.1.1	Gastro-oesophageal reflux	20
4.1.2	Breath holding attacks	20
4.1.3	Epilepsy	20
4.1.4	Child abuse	20
4.1.5	Intrinsic upper airway obstruction	21
4.1.6	Cardiac dysrhythmia	21
4.2	Consequences of ALTE	21
4.3	Discharge planning and interventions	21

<b>5.</b>	<b>Non Respiratory Causes Of Excessive Daytime Sleepiness In Children</b>	<b>23</b>
5.1	Narcolepsy	23
5.2	Idiopathic CNS Hypersomnia	24
5.3	Hypersomnia With Depression	24
5.4	Chronic Fatigue Syndrome / Fibromyalgia and Excessive Daytime Sleepiness	24
5.5	Insufficient Night Sleep	24
5.6	Delayed sleep phase syndrome	24
5.7	Non 24 Hour Sleep Wake Syndrome	25
5.8	Episodic Hypersomnia / Kleine Levin Syndrome	25
5.9	Restless legs syndrome / Periodic Leg Movement Disorder	25
<b>6.</b>	<b>Episodic behaviours in sleep after infancy</b>	<b>26</b>
<b>7.</b>	<b>Current provision of services</b>	<b>27</b>
<b>8.</b>	<b>Organisation of Services</b>	<b>29</b>
8.1	Training and education	29
8.2	Available facilities and expertise	30
8.2.1	Primary Care	30
8.2.2	Secondary Care	30
8.2.3	Tertiary Care	31
8.2.4	Quaternary Care	31
<b>9.</b>	<b>Quality control and Audit of services</b>	<b>33</b>
9.1	Local implementation	33
9.2	Quality Control	33
9.3	Resource implications	33
9.4	Key points for Audit	34
<b>10.</b>	<b>Declaration of Interests</b>	<b>35</b>

# **1. Introduction**

## **1.1 Justification and remit**

The sleep disorders dealt with in this report are those where sleep physiology is disturbed and/or breathing is impaired during sleep, and which may require assessment in a sleep laboratory. These include a wide range of conditions. Obstructive sleep apnoea occurs in around 2% of children, and is associated with a number of adverse physical and mental outcomes. Less common problems include the life-threatening disorder of congenital central hypoventilation syndrome, or the under-recognised and disabling problem of narcolepsy in children.

A recent survey of paediatricians disclosed a chaotic and unplanned structure of services for sleep disorders in children, often unfunded and frequently perceived as inadequate for local needs. This and a further survey have demonstrated wide variation in the availability and quality of facilities in the UK for the assessment and treatment of children with sleep disorders.

This report presents evidence-based recommendations for the diagnosis and management of disorders of sleep physiology and respiratory control in children, and the organisation of such services nationally in the UK. Guidelines already exist for the diagnosis and management of Obstructive Sleep Apnoea/Hypopnoea Syndrome in adults. Children are sufficiently different to justify a separate approach; they have more varied conditions presenting with sleep disordered breathing, with very different natural histories; they have far more protean and elusive symptoms; and they present different challenges in both diagnosis and treatment.

There are four main presentations which lead to the consideration of an underlying disturbance of sleep physiology or respiratory control. These are:

- symptoms suggesting airway or breathing problems during sleep;
- apparent life threatening events in infancy;
- diurnal symptoms suggesting disturbed sleep, including excessive daytime sleepiness;
- unexplained events during sleep in older children.

In addition, a number of conditions are known to be at high risk of such disorders even without suggestive symptoms.

The organisation of the clinical section of the report will therefore be according to these four presenting patterns of sleep and breathing impairment.

The report aims to aid parents, primary and secondary care physicians and surgeons to recognise the symptoms, to prioritise referral requests, to identify groups who require screening for abnormalities, and to understand which investigations and treatment modalities are

available and appropriate. It also aims to aid clinicians and health service managers involved in providing and commissioning services for affected children in prioritising such commissioning, and in organising pathways of care.

The main report is detailed and evidence-based. This document is a summary of the main points and conclusions of the report.

***GRADES OF RECOMMENDATION***

<b>A</b>	At least one meta-analysis, systematic review or RCT rated as 1 <sup>++</sup> and directly applicable to target population; <i>or</i>
	A body of evidence rated as 1 <sup>+</sup> consisting mainly of RCTs and directly applicable to target population, and consistent
<b>B</b>	A body of evidence including studies rated as 2 <sup>++</sup> directly applicable to target population, and consistent; or
	Extrapolated evidence from studies rated as 1 <sup>++</sup> or 1 <sup>+</sup>
<b>C</b>	A body of evidence including studies rated as 2 <sup>+</sup> directly applicable to target population, and consistent; or
	Extrapolated evidence from studies rated as 2 <sup>++</sup>
<b>D</b>	Evidence level 3 or 4; or
	Extrapolated evidence from studies rated 2 <sup>+</sup>

***GOOD PRACTICE POINTS***

- √ Recommended best practice based on clinical experience of working party.

<b>Table 1. Abbreviations used in main report</b>	
ADHD	Attention Deficit/Hyperactivity Syndrome
AHI	Apnoea/Hypopnoea Index
ALTE	Apparent life threatening event
BIPAP	Bilevel positive airway pressure
CCHS	Congenital Central Hypoventilation Syndrome
CPAP	Continuous positive airway pressure
ECG	Electrocardiograph
MSLT	Multiple Sleep Latency Test
NIV	Non-invasive ventilation
OSA	Obstructive Sleep Apnoea (includes hypopnoea)
P <sub>a</sub> CO <sub>2</sub>	Arterial Carbon Dioxide tension
P <sub>tc</sub> CO <sub>2</sub>	Transcutaneous Carbon Dioxide tension
P <sub>et</sub> CO <sub>2</sub>	End-tidal Carbon Dioxide tension
PIRCM	Paradoxical Inward Rib Cage Movement
PLMD	Periodic Leg Movement Disorder
PRS	Pierre Robin sequence
PSG	Polysomnography
PTT	Pulse Transit Time
PWS	Prader Willi Syndrome
REM	Rapid Eye Movement
SRBD	Sleep –Related Breathing Disorder
SpO <sub>2</sub>	Oxygen saturation measured by pulse oximetry
UARS	Upper airway resistance syndrome

## 1.2 Developmental physiology

The changes with increasing age, and the physiological changes during REM are summarised in Tables 2 and 3.

<b>Table 2. Changes in sleep with increasing age</b>
Reducing proportion of time asleep
Reducing quantity of REM sleep
Consolidation of sleep to night-time only

<b>Table 3. Physiological changes during REM</b>	
Metabolic response to cold stress	Vigorous (reduced in non-REM)
Ventilatory response to hypoxia	Reduced
Ventilatory response to hypercarbia	Reduced
Pharyngeal muscle tone	Reduced
Functional Residual Capacity	Reduced and unstable
Respiratory Rate	Increased variability
Heart Rate	Increased variability
Oxygen Saturation	Increased variability

### 1.3 Behavioural sleep problems

The prevalence of behavioural sleep problems, including bedtime resistance and sleep phase disturbances, is high. Moderate or severe sleep problems are reported in 17% of 1 year old children, and some form of sleep problem is present in 20% of 5 year olds and 6% of 11 year olds. There is a perceived lack of services for such problems. There can be considerable diagnostic difficulty between primary behavioural sleep disorders and those arising from sleep disordered breathing, and it is important that any centre which offers assessment of the latter should have some facilities to deal with behavioural sleep disorders either on site or by onward referral. However, the management of behavioural problems is outside the scope of this report, and will not be considered further here.

<b><i>1. Any centre which offers assessment of SRBD should establish some resource to deal with behavioural sleep disorders either on site or by onward referral.</i></b>	√
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### 1.4 General effects of sleep impairment

Sleep deprivation impairs cognition in adolescents. There is considerable epidemiological evidence that snoring, sleep disordered breathing or inadequate sleep in children are associated with behavioural and academic dysfunction, which is improved by intervention. There remains some uncertainty about the accurate identification of those children who will benefit from intervention, particularly in the more marginal cases.

#### ***Conclusion***

***Inadequate sleep duration or quality leads to impairment in attention, memory and behaviour, and worse school performance.***

## 2. Methodology of assessment

Three levels of investigation will be referred to:

**Screening studies** – used to screen for major abnormalities in a high risk population, or as a preliminary assessment of children with obstructive symptoms.

**Second-line studies** – used to assess children where the diagnosis is in doubt or where treatment decisions cannot be made on the basis of screening studies.

**Third line studies** – used to assess children where knowledge of sleep neurophysiology and architecture is important to decision-making or diagnosis.

### 2.1 Adequacy of ventilation

Oxygenation is not the most sensitive indicator of inadequate ventilation, but hypoxia is a clinically important end-point. It is best assessed by an overnight pulse oximetry recording.

<b>2. Oximetry recordings should only be performed by clinicians who are skilled in interpretation of the results, and systems should allow graphical inspection of recordings, with adequate facilities for artefact rejection.</b>	√
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Carbon dioxide is a more sensitive indicator of inadequate ventilation but is technically more difficult to assess accurately. End tidal capnography is useful if a plateau is obtained, but requires nasal or oronasal cannulae. Transcutaneous pCO<sub>2</sub> measurement is less invasive but results are less accurate in absolute terms.

<b>3. For any investigation of SRBD other than screening studies a measurement of CO<sub>2</sub> is essential, and the use of both end-tidal and transcutaneous modalities reduces the number of epochs with unobtainable data and is therefore recommended.</b>	√
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### 2.2 Evaluation of respiratory disturbance

To assess respiration requires a measure of airflow (oronasal thermistor, nasal cannulae with pressure transducer, ETCO<sub>2</sub>, or RIP) and effort (strain gauge bands or RIP). RIP can be used alone for both purposes, but is not as sensitive as other airflow methods in detecting apnoea.

## 2.3 Assessment of cardiac rate/rhythm

While the cardiac rate can be derived from the pulse oximeter, the rhythm and possible effects of hypoxia or airway obstruction can only be assessed using an ECG.

<b>4. <i>A single lead ECG is recommended as a minimum for second-line studies.</i></b>	√
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## 2.4 Assessment of sleep state/arousals

The minimum requirements are EEG (ideally central and occipital leads), submental EMG, and EOG. Arousals can be assessed behaviourally using movement (video or accelerometer), or autonomically by pulse transit time measurement.

## 2.5 Other measurements

Useful information may be gained from measuring body position, limb movements, oesophageal pH and video or sound recording. A laboratory undertaking second- or third- line studies should also have the facility for ventilator pressure measurement.

## 2.6 Interpretation

Adult criteria for identification of obstructive events should not be used for children as they may fail to identify clinically significant obstruction. The full report gives references for further information on normal values and criteria for respiratory events.

The optimum method for assessing arousals in children has yet to be determined. It is not yet clear which type of measured arousal is the best predictor of physical or neurocognitive effects.

<b>5. <i>Visual review of the complete recording should be undertaken by a competent observer before a report is issued.</i></b>	√
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## 2.7 Methodology of studies

Studies may be carried out at home or in a hospital depending on the complexity of the equipment required. Because both the equipment, the surroundings and the interpretation of findings are different in children, and because of the possible need for resuscitation in these patients, it is strongly recommended that all in-patient sleep studies on children are undertaken by staff with adequate training and experience in paediatrics, and in an environment where paediatric resuscitation facilities and skills are readily available. If children are studied in a primarily adult laboratory, it is strongly recommended that a paediatrician with expertise in

paediatric sleep medicine oversees the laboratory operations related to children and is involved in the interpretation of results.

<b>Table 4. Recommendations for minimum standards of equipment available</b>		
	Minimum	Ideal
Screening studies:	Oximetry (adequate storage and replay, with good artefact detection)	CO <sub>2</sub> measurement Video and sound Arousal detection
Second line studies	Above plus: Airflow Effort (thorax and abdomen) ECG CO <sub>2</sub> measurement Video and sound Arousal detection	Sleep staging Body position
Third line studies	Above plus: EEG/EOG/EMG Assessment of PLMS Oesophageal pH	

<b>6. A study of the whole night is the recommended investigation to assess sleep disordered breathing. A minimum of 6 hours sleep is desirable.</b>	√
<b>7. A single night study is generally sufficient to assess SRBD.</b>	<b>B</b>
<b>8. Abnormalities in sleep architecture require a second night study for reliable diagnosis.</b>	<b>C</b>
<b>9. All in-patient sleep studies on children should be undertaken by staff with adequate training and experience in paediatrics and in an environment where paediatric resuscitation facilities and skills are readily available.</b>	√
<b>10. If children are studied in a primarily adult laboratory, a paediatrician with expertise in paediatric sleep medicine should oversee the laboratory operations related to children and be involved in the interpretation of results.</b>	√
<b>11. Sleep studies should only be done in a suitable, quiet environment where normal sleep is possible.</b>	√

### 3. Airway and breathing problems during sleep

#### 3.1 Obstructive sleep apnoea (OSA) and hypoventilation

##### 3.1.1 Prevalence

The prevalence of frequent or habitual snoring in children is 10-12% between the ages of 5 and 10 years. The prevalence of SRBD in children is 0.7-2.9%, rising to 13% in morbidly obese children. Diagnosis is often delayed, with one third of US children with SRBD being symptomatic for >4 years before diagnosis.

##### 3.1.2 Presenting features

<i>Table 5. Symptoms and signs associated with SRBD</i>	
<b>During sleep</b>	<b>In the day</b>
Snoring or snorts	Behaviour problems
Gasping or laboured breathing	Poor concentration
Witnessed apnoeas	Excessive tiredness (symptoms may be subtle)
Odd sleeping positions	Failure to thrive
Sweating	Morning headaches
Enuresis	Mouth breathing/adenoidal facies/nasal speech
	Harrison's sulcus

Although most children with SRBD have no underlying condition other than adenotonsillar hypertrophy, there are a number of conditions in which SRBD are common (see below 3.2).

##### 3.1.3 Consequences

OSA can cause reversible failure to thrive, and is associated with systemic hypertension and increased left ventricular mass. Life threatening complications in children include cor pulmonale or pulmonary oedema.

OSA is associated with impaired academic performance in children, even in the absence of nocturnal hypoxia. OSA is also associated with increased health care utilisation.

##### 3.1.4 Identification

Most children without underlying risk factors will be identified because of concern from a parent or health care professional. There are no data to suggest that screening for asymptomatic children with OSA is worthwhile. While primary snoring may be associated with impaired

cognitive and behavioural performance, there are insufficient data to recommend routine intervention in snoring children.

### 3.1.5 Assessment

<i>12. At present there is insufficient evidence to recommend intervention in children if primary snoring is the sole symptom.</i>	√
<i>13. Clinical history is a sensitive screen for OSA, but has low specificity and relates poorly to severity.</i>	B
<i>14. Second- or third-line studies are required to gauge correctly the severity of OSA and reliably to discriminate OSA from primary snoring.</i>	B
<i>15. Second-line studies may be satisfactory in uncomplicated children over the age of 2 years.</i>	C
<i>16. Saturation monitoring is useful as a screen in otherwise healthy children. If positive it is highly predictive of OSA. A negative result does not exclude OSA.</i>	B
<i>17. Adenotonsillar size or other craniofacial abnormalities cannot be relied upon to predict the presence or absence of OSA.</i>	B
<i>18. Other screening tests are as yet not sensitive or specific enough to make treatment decisions.</i>	√
<i>19. The symptoms of SRBD may be difficult to identify in children with the underlying conditions listed in Table 7, and screening should be offered in these children, even if apparently asymptomatic.</i>	√

### 3.1.6 Management

Although there are no randomised controlled trials of adenotonsillectomy for OSA, there are consistent observational studies showing improvements in physiology, symptoms, growth and cognitive functioning after intervention. If nocturnal hypoxaemia is present then treatment is urgent, but the procedure has a higher risk of peri- and post-operative complications and precautions should be taken.

Either adenoidectomy or tonsillectomy alone are less effective and are not recommended.

If surgery is unsuccessful, or contraindicated, then other treatments should be considered as listed below. In children without hypoxaemia, the degree of morbidity attributable to SRBD must be weighed against the risks and inconvenience of interventions.

<i>20. Children with proven OSA secondary to adenotonsillar hypertrophy should be referred for adenotonsillectomy.</i>	<i>C</i>
<i>21. Nasal steroids and/or leukotriene receptor antagonists may be considered in mild cases of OSA or where abnormalities persist after adenotonsillectomy.</i>	<i>B</i>
<i>22. Oral jaw positioning devices should be considered for OSA in malocclusion. Further data and experience are required before this can be recommended for routine practice.</i>	<i>D</i>
<i>23. Mandibular and maxillary advancement surgery may be helpful in the management of OSA in craniofacial syndromes especially in those where tracheostomy is the only alternative.</i>	<i>D</i>
<i>24. Uvulopalatopharyngoplasty cannot be recommended in children with OSA.</i>	√
<i>25. Oxygen may be used as a temporary measure for the management of OSA provided carbon dioxide levels are shown not to rise during treatment.</i>	<i>B</i>
<i>26. CPAP/BIPAP is an effective treatment for the physiological derangement of OSA and should be offered where adenotonsillectomy has failed or is contraindicated if symptoms or physiological disturbance are severe.</i>	<i>D</i>
<i>27. In children with severe OSA where all other options have failed tracheostomy may be required.</i>	√
<i>28. When a child with abnormal physiology has undergone treatment, a further study to ensure normalisation of the physiology is recommended; if abnormal gas exchange has been documented, this is mandatory.</i>	√
<i>29. Children with suspected OSA who have associated risk factors listed in Table 7 should only have adenotonsillar surgery in a centre with Paediatric Intensive Care facilities available.</i>	√
<i>30. Overnight pulse oximetry is a desirable method of assessing the operative risk in children without apparent co-morbidity who are being considered for adenotonsillectomy. If performed, a nadir of &lt;80% or baseline hypoxaemia should prompt referral to a centre with Paediatric Intensive Care facilities available.</i>	√

**Table 6. Factors predicting need for PICU facilities in children with OSA**

Age < 2 years
Severe heart or lung disease
Neuromuscular disease
Craniofacial abnormalities
Severe neurodisability
Severe obesity (BMI Standard Deviation Score >2.5)

### 3.2 Specific conditions at high risk of SRBD

**Table 7. Conditions at high risk of Sleep Disordered Breathing**

Condition	Prevalence	Prevalence of SRBD	Other comments
Down's syndrome	1:1,000	70-100%	High risk of pulmonary hypertension, especially if co-incident heart disease
Neuromuscular Disease	1:3,000	42%	Difficult to detect clinically. Reduced life expectancy, reversible by treatment
Craniofacial abnormalities	1:7,000	Depends on severity; 100% in severe cases	
Achondroplasia	1:25,000	48%	
Mucopolysaccharidoses	1:40,000	>90%	Difficult to detect clinically
Prader-Willi syndrome	1:52,000	25-75%	Hypoxaemia common. Abnormal central ventilatory responses co-exist

#### 3.2.1 Down's syndrome

Children with Down's syndrome are at high risk of SRBD and nocturnal hypoxaemia, and the high incidence of congenital heart disease in these children makes pulmonary hypertension a significant risk. SRBD may be difficult to identify on symptoms in this group. Adenotonsillectomy may have a lower rate of success, but is still indicated. Other interventions including CPAP are effective but may be difficult to institute.

<p><b>31. All children with Down’s syndrome should be offered screening for SRBD, using at least oximetry; suggested screening ages are at least once in infancy then annually until age 3-5 years.</b></p>	<p>√</p>
<p><b>32. Children with Down’s syndrome with abnormalities on screening for SRBD, or where there is a clinical suspicion of a false negative screening test, should have polysomnography, including oximetry, airflow, effort and CO<sub>2</sub> measurement. Video should be included if possible.</b></p>	<p>√</p>
<p><b>33. If significant SRBD with hypoxia is present in children with Down’s syndrome, then appropriate treatment should be offered.</b></p>	<p>√</p>
<p><b>34. Further research is needed on the benefits and risks of screening for SRBD and Down’s syndrome.</b></p>	<p>√</p>

### 3.2.2 Neuromuscular Disease

There are two major patterns of sleep disordered breathing in neuromuscular disease: obstructive sleep apnoea due to loss of glossopharyngeal muscle tone and hypoventilation due to intercostal and abdominal weakness. If the diaphragm is involved then the hypoventilation is particularly severe during REM sleep. Bulbar palsy and scoliosis both increase the risk of respiratory failure and SRBD.

The overall prevalence of SRBD in neuromuscular disease is high, and in progressive conditions it is likely to occur at some stage in most patients. SRBD in neuromuscular disease is associated with increased pulmonary artery pressure. There is good evidence (level 2++) that NIV improves nocturnal and diurnal oxygen saturation and pCO<sub>2</sub> in neuromuscular disease patients who have SRBD, and evidence (level 2+) that NIV improves survival in these patients, particularly if hypercarbia is present. SRBD and respiratory failure are difficult to detect clinically, and screening in patients at high risk is recommended.

NIV should be part of a package of respiratory care in neuromuscular disease, which is aimed at preventing and effectively treating atelectasis and episodes of lower respiratory infection, optimising nutrition, and effective management of scoliosis.

### Assessment and screening

35. <i>Non-invasive ventilation is not indicated routinely in DMD in the absence of SRBD.</i>	B
36. <i>Overnight oximetry recordings should be carried out on all children with neuromuscular disease if there are symptoms of SRBD, impairment of diaphragmatic function, or a vital capacity below 50% predicted. In conditions such as myopathies, where the risk of early SRBD is particularly high, regular recordings should be carried out even in the absence of any of these indicators.</i>	√
37. <i>If feasible, CO<sub>2</sub> recordings should be performed in conjunction with oximetry in children with neuromuscular disease, as they may add useful information.</i>	√
38. <i>The optimum frequency of oximetry recordings in high risk children with neuromuscular disease is uncertain. At least annual recordings should be done, with more frequent recordings in higher risk situations.</i>	√
39. <i>Limited polysomnography (second-line study) should be performed in neuromuscular patients with abnormal oximetry, but in the presence of severe abnormalities treatment should not be delayed if polysomnography is not readily available.</i>	√

### Intervention

40. <i>If SRBD sufficient to cause hypoxaemia at night is demonstrated in an otherwise stable child with neuromuscular disease, then nocturnal NIV should be instituted.</i>	√
41. <i>If SRBD is associated with nocturnal hypercapnia in a child with neuromuscular disease then nocturnal NIV should be instituted.</i>	C
42. <i>A child with neuromuscular disease on NIV should have repeated studies with oximetry and CO<sub>2</sub> recordings to ensure optimal NIV settings.</i>	√

### 3.2.3 Anatomical abnormalities

#### 3.2.3.1 Craniofacial syndromes

Children with craniofacial syndromes are unusual in that they often present with SRBD in infancy. However, the airway obstruction may worsen with growth, particularly in midfacial hypoplasia.

In the absence of gas exchange abnormalities, intervention should be based on the presence of clinical symptoms suggestive of SRBD.

<p><b>43. All children with syndromes involving midfacial hypoplasia or micrognathia should be evaluated for SRBD with a minimum assessment of oximetry, preferably with a measure of CO<sub>2</sub>. This should be performed urgently if they have any clinical signs of airway obstruction, and within the first 4 weeks of life in any event.</b></p>	√
<p><b>44. Clinicians should be aware that infants with PRS may have worsening airway obstruction between 4 and 8 weeks and ascertain whether symptoms worsen at this age. If so, repeat assessment should be carried out.</b></p>	√
<p><b>45. Reassessment for SRBD in children with syndromes involving midfacial hypoplasia or micrognathia should occur at 3-6 monthly intervals in the first year of life, and subsequently should be dictated by clinical symptoms and signs.</b></p>	√
<p><b>46. In infants with PRS or other micrognathia syndromes and with significant airway obstruction or SRBD:</b></p> <ul style="list-style-type: none"> <li>• <b>A nasopharyngeal tube is the first line of treatment.</b></li> <li>• <b>If nasopharyngeal intubation is unsuccessful, nasal CPAP or BIPAP should be tried.</b></li> <li>• <b>Tracheostomy is necessary if other measures fail.</b></li> <li>• <b>Mandibular advancement surgery may have a role in refractory cases, taking into account the degree of expected mandibular growth.</b></li> <li>• <b>There is no evidence to support the practice of prone positioning.</b></li> </ul>	√
<p><b>47. In children with midfacial hypoplasia and airway obstruction or SRBD:</b></p> <ul style="list-style-type: none"> <li>• <b>A trial of nasal CPAP or BIPAP is indicated.</b></li> <li>• <b>If this fails, then surgical options include tracheostomy or surgical reconstruction. If the airway is significantly impaired then tracheostomy remains the immediate treatment of choice.</b></li> <li>• <b>The role of craniofacial surgery as an alternative to tracheostomy requires further evaluation.</b></li> </ul>	√

### 3.2.3.2 Mucopolysaccharidoses

Children with mucopolysaccharidoses, particularly Hurler, Hurler-Scheie and Hunter syndromes are at high risk for SRBD.

<b>48. In children with Hurler, Hurler-Scheie and Hunter syndromes:</b>	
<i>a. Screening for SRBD should be offered, after discussion of the possible interventions and benefits.</i>	√
<i>b. Adenotonsillectomy should be considered if there is significant SRBD.</i>	√
<i>c. In significant SRBD where adenotonsillectomy is unsuccessful or not feasible, nasal CPAP should be considered.</i>	<b>D</b>

### 3.2.3.3 Achondroplasia

Children with achondroplasia are at high risk of SRBD. Pulmonary hypertension and cor pulmonale are significant risks in SRBD in achondroplasia. Interventions such as adenotonsillectomy and nasal CPAP are usually, but not always, associated with clinical and polysomnographic improvements.

<b>49. In children with achondroplasia:</b>	
<i>a. Screening for SRBD should be offered to all children with achondroplasia. Ideally this should include oximetry and capnography.</i>	√
<i>b. If initial screening is normal, the optimum frequency of subsequent screening is unclear, but should probably be every 6-12 months in the first 5 years of life.</i>	√
<i>c. If significant SRBD is discovered, then adenotonsillectomy should be offered.</i>	√
<i>d. A trial of CPAP should be considered if symptoms or significant gas exchange abnormalities persist after adenotonsillectomy.</i>	√

### 3.2.3.4 Prader Willi Syndrome (PWS)

Sleep disordered breathing and nocturnal hypoxaemia are common in patients with PWS, and nocturnal hypoventilation leads to cardiorespiratory failure. Respiratory failure occurring in PWS can be treated effectively by nocturnal NIV.

<i>50. All children with PWS should be screened with oximetry and capnography on an annual basis. Children with abnormal oximetry should have polysomnography.</i>	√
<i>51. In PWS with respiratory failure a trial of NIV at night should be initiated.</i>	<b>D</b>
<i>52. In PWS with significant nocturnal hypoxaemia a trial of NIV at night should be initiated.</i>	√
<i>53. Adequacy of breathing during sleep should be assessed formally in any child with PWS prior to starting growth hormone treatment.</i>	√

### 3.3 Congenital Central Hypoventilation Syndrome

Untreated CCHS is incompatible with long-term or intact survival.

The diagnosis of CCHS requires the following criteria:

1. Persistent evidence of hypoventilation during sleep [ $P_a\text{CO}_2 > 60$  mm Hg (8 kPa)]
2. Onset of symptoms usually in the first year after birth
3. Absence of primary pulmonary or neuromuscular disease
4. No evidence of primary heart disease

<i>54. Children with suspected CCHS should be referred to a specialist centre with adequate facilities and experience for confirmation of the diagnosis.</i>	√
<i>55. In children with CCHS:</i>	
<i>a. Ventilatory support is almost always essential for survival.</i>	<b>B</b>
<i>b. Management in the first few years of life usually requires tracheostomy, although other techniques have been used successfully in some specialist centres.</i>	√
<i>c. Diaphragmatic pacing should be considered in children requiring 24-hour ventilatory support.</i>	<b>D</b>
<i>d. Care should be supervised by a specialist centre with experience of CCHS management.</i>	√
<i>e. Oxygen saturation should be monitored continuously during sleep.</i>	√
<i>f. During acute illnesses children with CCHS require checks of oxygen saturation and carbon dioxide levels when awake.</i>	√
<i>g. Carers and children should be advised of the cautions required during exercise, and the specific dangers of alcohol or cannabis use.</i>	√

## 4. Unexplained events in infancy – ALTE

Conditions which present as possible respiratory control disorders in infancy are often described by the term “apparent life threatening events” or ALTEs. An ALTE has been defined as “an episode that is frightening to the observer and that is characterised by some combination of apnoea (central or occasionally obstructive), colour change (usually cyanotic or pallid), marked change in muscle tone, choking or gagging.”

**Table 8. Causes to which ALTE has been ascribed**

Respiratory
Infection – e.g. respiratory syncytial virus infection, pertussis, pneumonia*
Upper airway obstruction – e.g. retrognathia, laryngomalacia
Lower airway obstruction or closure – e.g. tracheo-bronchomalacia
Intrapulmonary shunting – e.g. cyanotic breath holding
Neurological
Epileptic - seizure induced *
Intracranial haemorrhage - vitamin K deficiency, child abuse*
Central hypoventilation - drugs, congenital*
Brain tumour*
Infective
Septicaemia, urinary tract infection, gastro-enteritis, *
Meningo-encephalitis*
Autonomic
Vasovagal
Gastro-oesophageal reflux
Skin pallor changes
Child Abuse
Illness fabrication *
Attempted suffocation*
Poisoning *
Cardiac
Tachyarrhythmias - Wolfe-Parkinson White and Long-QT syndrome
Congenital heart disease
Inborn areas of metabolism *
Miscellaneous
Carbon monoxide poisoning*
Cat smothering
Abnormal infant holding practices
Haemorrhagic shock encephalopathy syndrome
Unknown

\* Evidence of benefit from identification

## 4.1 Specific conditions and ALTE

### 4.1.1 Gastro-oesophageal reflux

A causative association between gastro-oesophageal reflux and ALTE has yet to be firmly established.

56. <i>Surgical treatment of gastro-oesophageal reflux should not be undertaken in patients presenting with recurrent apnoea or ALTE without evidence of the temporal association of the events with reflux.</i>	√
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### 4.1.2 Breath holding attacks

57. <i>In the presence of a clear history of breath holding attacks, treatment with iron should be considered, particularly if there is evidence of iron deficiency.</i>	B
58. <i>Further trials of the safety and efficacy of piracetam in breath holding attacks are needed before this drug can be recommended.</i>	√

### 4.1.3 Epilepsy

59. <i>In recurrent, unexplained ALTE continuous EEG recording should be undertaken (preferably simultaneously with other physiological monitoring) to attempt to capture an event.</i>	√
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### 4.1.4 Child abuse

60. <i>Child abuse, including fabricated or induced illness by carers, should be considered as a possible cause of ALTE in any of the following circumstances:</i>  <ul style="list-style-type: none"> <li>– <i>there is a history of severe or repeated attacks with a single witness of the attack onset</i></li> <li>– <i>petechiae or bleeding from the mouth or nose</i></li> <li>– <i>there is a history of ALTE or sudden death in siblings</i></li> </ul> <i>However, none of these features in isolation is diagnostic of child abuse.</i>	B
61. <i>In cases of ALTE where child abuse is considered a possible cause, referral to a specialist centre may be required for elucidation.</i>	√

#### 4.1.5 Intrinsic upper airway obstruction

<i>62. Polysomnography should be performed in infants with severe and recurrent ALTE.</i>	√
<i>63. In children with severe and recurrent ALTE due to OSA, a trial of CPAP is indicated.</i>	√

#### 4.1.6 Cardiac dysrhythmia

Cardiac dysrhythmias are a rare cause of ALTE.

<i>64. An ECG should be recorded, and QTc measured, in all infants presenting with ALTE.</i>	√
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### 4.2 Consequences of ALTE

Infants with ALTE where no serious underlying condition is found have a very low risk of subsequent death.

<i>65. Specialist assessment of ALTE is needed for infants with recurrent significant events; events where cardiopulmonary resuscitation has been needed; or those with a family history of unexplained childhood death.</i>	√
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### 4.3 Discharge planning and interventions

<i>66. There is no evidence to support the routine use of home monitors in ALTE.</i>	√
<i>67. If a decision is taken to issue a monitor, the parents must understand that there is no evidence that it will prevent subsequent death.</i>	√
<i>68. Cardiopulmonary resuscitation should be taught to any carers who take home an infant with a monitor.</i>	√

**Table 9. Suggested investigations in ALTE**

First line investigations

***If clinically well***

Full blood count  
Urine culture  
S<sub>a</sub>O<sub>2</sub> measurement/recording  
Urine organic acids  
Serum and urine amino acids  
Blood sugar †  
Arterial blood gas †  
Lactate †  
Ammonia †

***If unwell, may also require:***

Blood culture ± lumbar puncture  
Nasopharyngeal aspirate for viral immunofluorescence and culture  
Pernasal swab for pertussis  
Chest x-ray  
ECG

Second line investigations (severe/recurrent events)

Multi-channel physiological recordings / event recording\*  
EEG\*  
Oesophageal pH monitoring (simultaneous with physiological recording if possible)  
ENT assessment  
Cranial imaging (Ultrasound, CT, MRI)  
Echocardiogram  
Skeletal survey  
Urinary toxicology screen

Third line investigations

Covert video surveillance (if onset only ever witnessed by one person)

† if close to event/still unwell

\* of particular value for documenting pathophysiology during subsequent event

## 5. Non Respiratory Causes Of Excessive Daytime Sleepiness In Children

There are a number of causes of excessive daytime sleepiness which should be considered, some of which are rare. The conditions are listed here, but only the commoner conditions are discussed. Rarer conditions are dealt with in the full report.

### 5.1 Narcolepsy

**Table 10. Diagnostic criteria for narcolepsy**

- |  |
|--|
| <p>A. The patient has a complaint of excessive sleepiness or sudden muscle weakness.</p> <p>B. Recurrent daytime naps or lapses into sleep occur almost daily for at least 3 months.</p> <p>C. Sudden bilateral loss of postural muscle tone occurs in association with intense emotion (cataplexy).</p> <p>D. Associated features include:-</p> <ol style="list-style-type: none"><li>1. sleep paralysis</li><li>2. hypnagogic hallucinations</li><li>3. automatic behaviours</li><li>4. disruptive major sleep episode</li></ol> <p>E. Polysomnography demonstrates one or more of the following:-</p> <ol style="list-style-type: none"><li>1. sleep latency less than 10 minutes</li><li>2. REM sleep latency less than 20 minutes</li><li>3. multiple sleep latency test demonstrates a mean sleep latency of less than 5 minutes</li><li>4. 2 or more sleep onset REM periods</li></ol> <p>F. HLA typing demonstrates DQB1 0602 or DR2 positivity.</p> <p>G. No medical or mental disorders account for the symptoms.</p> <p>H. Other sleep disorders, for example, periodic leg movement disorder or central sleep apnoea syndrome maybe present but are not the primary cause of the symptoms.</p> |
|--|

A high index of suspicion and a careful history are required to recognise symptoms of narcolepsy in childhood.

<i>69. To confirm a diagnosis of narcolepsy, referral to a centre experienced at PSG and MSLT in children, with clinical experience of narcolepsy is necessary.</i>	√
<i>70. Management of narcolepsy should only be undertaken under supervision from a clinical service experienced in the condition.</i>	√

## 5.2 Idiopathic CNS Hypersomnia

## 5.3 Hypersomnia With Depression

## 5.4 Chronic Fatigue Syndrome / Fibromyalgia and Excessive Daytime Sleepiness

<i>71. Polysomnography may be necessary to rule out a primary sleep disorder in selected cases of suspected CFS where excessive daytime sleepiness is a prominent feature.</i>	√
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## 5.5 Insufficient Night Sleep

Sleep restriction is common in adolescents and is associated with poor daytime functioning.

<i>72. Assessment of sleep habits using diaries or actigraphy are essential in the evaluation of daytime sleepiness.</i>	√
<i>73. Polysomnography is indicated if a cause is not otherwise apparent.</i>	√

## 5.6 Delayed sleep phase syndrome

<i>74. Delayed sleep phase syndrome is best diagnosed by history and actigraphy.</i>	√
<i>75. Chronotherapy should be considered in the treatment of delayed sleep phase syndrome.</i>	<b>D</b>
<i>76. Melatonin may give short term benefits in delayed sleep phase syndrome.</i>	<b>B</b>
<i>77. The effects of melatonin in delayed sleep phase syndrome are not sustained after cessation of treatment.</i>	<b>D</b>

## 5.7 Non 24 Hour Sleep Wake Syndrome

## 5.8 Episodic Hypersomnia / Kleine Levin Syndrome

## 5.9 Restless legs syndrome / Periodic Leg Movement Disorder

<i>78. A history of restless legs or growing pains should be sought in children with daytime symptoms suggestive of a sleep disturbance, including attention problems.</i>	√
<i>79. The current evidence is not yet adequate to warrant screening for PLMD in children with ADHD.</i>	√
<i>80. Iron deficiency should be sought and treated in children diagnosed with RLS.</i>	C
<i>81. If PLMD/RLS is associated with significant functional disturbance, then treatment with levodopa, dopamine agonists, gabapentin or clonazepam should be considered.</i>	D

## 6. Episodic behaviours in sleep after infancy

These events come under the headings of arousal disorders, sleep-wake transition disorders, REM parasomnias, and epilepsy.

<i>82. Most episodic events occurring during sleep are benign and do not warrant investigation.</i>	√
<i>83. Episodes which are frequent, or occur throughout the night, require more evaluation, including EEG and video monitoring to exclude epilepsy.</i>	C
<i>84. Persistent troublesome events during sleep may require full polysomnography, including video and EEG, to exclude treatable factors.</i>	√

## **7. Current provision of services**

A preliminary survey was conducted by the working party in 2002, with a questionnaire sent to all consultant paediatricians in the UK, asking them about sleep services in their area, and where they would send five exemplar cases (OSA, neuromuscular patient with suspected nocturnal hypoventilation, possible narcolepsy, ALTE, unusual night awakening.) The median number of different referral targets listed by doctors from a single Strategic Health Authority ranged from 2 to 3. Contradictory referral patterns were identified. In more than one area a tertiary recipient of referrals for a case would themselves refer the case elsewhere. In one area the neurology services said that they referred to the respiratory paediatricians and vice versa. Respondents were invited to add free text comments, and 88 (34%) did so. The commonest comment (86%) was that there was a large unmet need for sleep services in the area.

The points which emerged from this survey were:

- Poor awareness of local facilities, particularly if a full service is not available.
- Inconsistent referral patterns, with adjacent hospitals often referring to widely different centres.
- A widely perceived need for better provision and organisation of services.

Further to the 2002 survey, a more detailed and directed survey of paediatric PSG facilities was conducted by Dr Cathy Hill in 2005 (unpublished data). This identified 21 possible paediatric sleep centres from 3 sources: the British Sleep Society UK Provider Directory, information from commercial companies providing sleep systems and data from the original 2001 survey. A survey questionnaire was sent to all possible centres, of whom 18 (86%) responded, one of whom was not a provider of PSG services. 12 centres were offering full PSG, 2 were in transition to such a service and 5 were offering extended cardiorespiratory monitoring only. Some centres offered mainly electrophysiological investigation and some mainly cardiorespiratory investigation.

The number of PSG studies performed by each centre per year varied from 20 to over 500 (reported by a single centre). Studies were done in a variety of settings with only 6 centres having a specialised paediatric sleep laboratory in which to conduct studies. Two centres conducted home PSGs, two had mixed adult/paediatric laboratories, and one used HDU. Two centres could only perform studies in an open paediatric ward. A total of 10 specialised paediatric sleep laboratory beds were identified nationally.

Eleven centres have the ability for electrophysiological sleep staging of studies, 5 can do full EEG recording with sleep studies, and 7 can record leg EMG.

Ten centres reported that their studies were fully attended overnight, with others relying on

intermittent nurse surveillance. 8/17 centres employ a total of 22 sleep technologists or physiologists, but the other 9 centres are without any specialised staff for the PSG. Paediatricians in 5 centres were reported to be competent to set up, score, and report a PSG. Concern was expressed by many respondents about quality control, since there was no identified mechanism for external review of studies in any centre, and most are working in isolation.

The problems identified in the current provision of service are:

- Very variable quality and quantity of services in different geographical areas.
- Lack of awareness of tertiary facilities available within secondary care centres.
- Diagnostic sleep facilities generally poorly staffed, often with unqualified personnel, and in inappropriate clinical areas.
- Few arrangements in place for quality control of studies.

A list of the UK NHS centres currently believed to have the ability to provide full polysomnography with neurophysiological sleep staging (“third line” studies) is provided in Appendix 4. It should be noted that this is derived from self-reported information and no objective data are available on accuracy or on quality of service provided. It is not possible to make an accurate list of centres which can provide adequate tertiary-level studies (i.e. cardiorespiratory assessments and ventilation titrations), and there is a clear need for some form of quality control in centres providing second- and third- line studies (i.e. tertiary and quaternary centres).

## **8. Organisation of Services**

In the light of the information in section 7 there appears to be a clear perception that current services are not meeting the diagnostic and treatment needs of children. The literature includes descriptions of clinical investigation pathways but no comparative data of varying service models. What does exist is from North America, where health service organisation is not comparable to the UK. There is one UK review, which describes a recommended set of practices based on literature evidence.

Previous discussion in this report makes it clear that children with unrecognised sleep physiology disorders make heavy use of medical services, under-perform academically and behaviourally and derive measurable benefit from diagnosis and treatment.

The consequences of failing to address the current erratic and patchy paediatric sleep services in the UK can be deduced from much of the preceding report. These include:

- Continuing behavioural and cognitive problems (section 1.5)
- Continuing difficulties in assessing OSA (section 3.1.5)
- Failure to recognise severe OSA with increased peri-operative risk of ENT surgery (section 3.1.6)
- Continued inequality of services for children and adolescents with muscle disease (section 3.2.2)
- Continued inequality in services for infants/children with craniofacial problems, storage disorders, skeletal abnormalities and PWS (section 3.2.3)
- Potential difficulties in accessing investigation in cases of ALTE (section 4)
- Inadequate access to detailed studies to diagnose narcolepsy (section 5.1) and to differentiate other excessive daytime sleepiness (section 5.2 to 5.9)

Any recommendation of service has currently to be based on expert opinion of the shape of service that will minimise the chances of morbidity that could be addressed by efficient readily available diagnostic and treatment services. Ideally respiratory and neurology expertise will be available within the service. In addition paediatric sleep investigation services need to work closely with colleagues in ENT and airway surgery.

### **8.1 Training and education**

Effective provision of services in the field of sleep related physiological disorders in childhood will require the implementation of education and training for all clinical staff dealing with children at the primary, secondary or tertiary level, in order to identify those children for whom referral to secondary or tertiary services will be appropriate.

Detailed description of appropriate training and educational approaches is beyond the scope of this report, but as noted previously, a basic knowledge of sleep physiology and its development in childhood should be incorporated into undergraduate and postgraduate training prospectuses for a wide range of health care professionals. Appropriate multi-professional postgraduate training packages will also need to be developed at different levels for those taking part in the assessment and treatment of these conditions. These packages need to be specific for children.

## **8.2 Available facilities and expertise**

In order to meet the suggested standards for investigation, diagnosis and treatment of children with sleep related physiological disturbances set out in sections 1-6 of this report, relevant expertise and facilities will need to be provided at primary, secondary, tertiary and quaternary levels of care. An outline of the minimum recommended levels of expertise, staffing and facilities based upon these standards is set out below. These estimates include only those staff directly employed in the provision of the services, and must be fully supported by appropriate levels of administrative and secretarial staff, plus appropriate technical support for care and maintenance of the complex equipment required.

### **8.2.1 Primary Care**

Information on relevant symptomatology and possible consequences of disorders of childhood sleep physiology should be incorporated into training for health visitors, school nurses and those involved in developmental screening in childhood. The development and incorporation of appropriate questionnaires on sleep into routine developmental screening and more widespread recognition of the potential contribution of sleep disorders to poor school performance and behavioural problems are likely to increase the appropriate and early recognition and referral of affected children.

### **8.2.2 Secondary Care**

The high prevalence of many disorders of sleep physiology in childhood (e.g. OSA, ALTE) means that most children with suggestive symptoms will most appropriately be seen, investigated and treated by the local paediatric secondary care service. This will require, in addition to the appropriate level of training for consultant paediatricians, the availability of the necessary equipment (with robust artefact detection or signal extraction facilities) to carry out overnight recordings of pulse oximetry on children at home. Because of the limitations of non-observed home oximetry recordings (see section 3.1.5), some secondary care services will also benefit from the availability of facilities to make more detailed recordings – e.g. expired carbon dioxide and/or overnight infrared or low-light video recordings. In a secondary care Paediatric

service serving a population of 50- 60,000 children, with 3,000 births per year, a single recording oximetry system is likely to be sufficient for this purpose. Some provision for this service must be made in job plans for medical and support staff, though the time commitment is likely to be small (approximately one PA of consultant time per month). It is essential that the clinicians involved in this service work in liaison with the tertiary centre to ensure a smooth patient journey.

### **8.2.3 Tertiary Care**

Tertiary level investigational and treatment services for children with disorders of sleep physiology should be available in all tertiary care centres serving 2, 3 or more Strategic Health Authorities.

Facilities should include the full range of “second line” investigations (see section 2.7.3), together with appropriate staff and resources to conduct such investigations in hospital – on paediatric wards or preferably also in specialised sleep laboratories – and in the community – particularly for children receiving continuing treatments such as invasive or non-invasive ventilatory support.

The workload for such a tertiary care facility, serving a population of 3-4 million people will be such that dedicated consultant and support staff time will need to be identified and funded. From the workload of such centres currently undertaking this level of service provision this is likely to be in the range of 5 – 8 consultant PA’s per week, plus 2 – 3 WTE nurse specialists (or technical staff) for a centre that does not also provide quaternary level services (see below).

### **8.2.4 Quaternary Care**

Some (but not all) tertiary level services will also need to provide more complex investigational facilities and expertise (e.g. quantitative recordings of minute ventilation, combined neurophysiological and respiratory recordings), for children with complex neurological disorders affecting sleep physiology, and those with disorders of respiratory control (e.g. CCHS).

The staffing requirements for such quaternary services will be determined by the precise services and investigations provided, the complexity of the case-mix and the configuration of the sleep laboratory. For a centre providing detailed investigational and treatment facilities for children with complex neurological and respiratory control disorders from a population of 5-6 million people, the additional staffing required (based upon the current workload in Bristol at present) is approximately 1 WTE physiologist, plus 3 PA’s per week of consultant time, in addition to that specified for tertiary centres. Thus a centre providing full polysomnography

will require at least 1 WTE consultant, 2 nurses/technicians or equivalent, and 1 WTE physiologist as a minimum. The overall numbers will also depend on whether the centre is conducting attended studies, and the configuration of beds in the laboratory. Two nurses or technicians can set up a maximum of three studies in a laboratory per night, and a 2-3 bedded laboratory will allow a more cost-efficient service than a single bedded unit, particularly if studies are attended. These numbers would allow for attended studies on 2-3 patients for 3 nights of the week. Larger throughput would require a proportionate increase in staffing. Smaller laboratories would have a lower capacity with little reduction in staffing. Multiple Sleep Latency testing involves another full day of physiologist time, and a centre undertaking significant number of these tests would need extra staff for this purpose.

## **9. Quality control and Audit of services**

### **9.1 Local implementation**

These UK recommendations will present a challenge to those running existing services as well as to areas of the country where these clinical issues have not yet been as well addressed. The exact configuration of service will vary due to geographic issues and established referral routes. However, clinicians and managers have a responsibility to implement the recommendations to enhance the provision of NHS care for this group of children. A system of managed clinical networks is recommended forging relationships between the tertiary and quaternary centres in a region and those offering secondary care level services.

### **9.2 Quality Control**

There are at present no systems for quality control of diagnostic or therapeutic services in this field. Sleep laboratories are often limited to a single expert who is able to score and assess polysomnographic studies. This poses a considerable clinical governance risk and there is an urgent need to institute more robust systems. In the first instance it is recommended that any centre offering diagnostic sleep facilities should take part in an external quality control system. This should consist of an annual visit from clinicians and physiologists from another centre, who will review the equipment and algorithms used and the outcome and throughput data for the centre, comparing with the recommendations in this report. The visiting team will independently score five randomly selected studies (this may be done in advance of the visit) and compare results with the original scoring. A standardised report proforma will be used for each visit, with recommendations for development. It is hoped that a network of tertiary/quaternary centres will be developed, and where major discrepancies are demonstrated between two centres these can be resolved by others in the network.

### **9.3 Resource implications**

Our surveys have demonstrated that current provision across the UK falls well short of the standards described in this document. There is likely to be significant need for further professional time (consultant, nurse and physiologist/technician) and a more modest need for new equipment, in particular, the provision of modern recording oximeters with high quality artefact rejection. There needs to be clear designation of the Quaternary centres, which will require proper investigation facilities as described in section 8.3.4.

## **9.4 Key points for Audit**

- Availability of good quality pulse oximetry at secondary care level.
- Prompt referral to ENT services or tertiary care for those with positive or unclear diagnosis.
- Proportion of positive diagnoses from those tested.
- Local rate of adenotonsillectomy for OSAHS.
- Follow up assessment of children with abnormal physiology after intervention.
- Proportion of children deemed to need further treatment.
- Proportion of those deemed to need CPAP who are established on therapy for greater than 4 hours per night.
- Annual review of all those on non-invasive or invasive ventilatory support.
- Peer review of clinical service, outcomes, polysomnography raw data and reporting.

## **10. Declaration of Interests**

No funding was received by the working party from any commercial bodies.





