

Sleep Apnea

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ABSTRACT

Objective. To study clinical presentation of sleep disordered breathing (SDB) in children, their causative factors and response to treatment.

Methods. A retrospective study of clinical data and results of overnight polysomnography done at baseline and after therapy were reviewed in 56 patients under 18 years of age.

Results. Of the 56 patients included in the study 23(41%) cases were positive for SDB. 12 (52.1 %) patients had craniofacial abnormalities, 4 (17.3%) had neuromuscular and skeletal disorders, 2 (8.6%) had adenotonsillar hypertrophy, 1(4.3%) had bilateral vocal cord palsy and 3 (13%) had sleep apnoea associated with multisystemic disorders. Post-operative data showed improvement in all 6 cases of craniofacial abnormalities and both cases of adenotonsillar hypertrophy. Positive airway pressure treatment was useful in cases with obstructive sleep apnea (OSA) due to vocal cord palsy, thoracic scoliosis, systemic disorders and central hypoventilation.

Conclusion. 41% of suspected cases were detected to have SDB. Craniofacial abnormality was the leading cause of OSA in the present study. Surgical correction improved symptoms apnea-hypopnea index (AHI) and desaturation in cases of craniofacial disorders and adenotonsillar hypertrophy. Vocal cord palsy, thoracic scoliosis, hypoventilation and systemic disorders associated OSA responded to positive airway pressure ventilation. [*Indian J Pediatr* 2006; 73 (7) : 597-601]
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Key words : Sleep disorders breathing; Polysomnography; Obstructive sleep apnea; Apnea-hypopnea index

Sleep is a major physiological drive. The average child spends almost one half of his or her life asleep, thus respiratory disorders during sleep are of particular importance during childhood. Although some respiratory disorders such as sleep apnea occur only during sleep, virtually all respiratory disorders including upper airway obstruction, central hypoventilation, and chronic lung disease are worse during sleep than wakefulness. Obstructive sleep apnea (OSA) occurs approximately one-third as often as asthma in children. The peak prevalence of childhood OSA is 2-8 years of age, the age when the tonsils and adenoid are the largest in relation to the underlying airway size.¹ Though the vast majority of cases of OSA in children are associated with adenotonsillar hypertrophy, OSA also occurs in children with upper airway narrowing due to craniofacial abnormalities or

those with neuromuscular abnormalities such as hypotonia or muscular incoordination. Obese children are at a higher risk for OSA and its severity is proportional to the degree of obesity, though most children may not be obese but fail to thrive.² Surgery is the main modality of treatment in children with OSA especially for adenotonsillar hypertrophy and craniofacial abnormalities as the cause. Other treatment options include positive airway pressure therapy with continuous positive airway pressure (PAP) ventilation or Bilevel PAP ventilation. Role of oral appliances, uvulopalatopharyngoplasty and supplemental oxygen therapy has not been studied in pediatric patients having OSA.

Thus, aim of this study was to analyze the clinical presentation of sleep disordered breathing (SDB) in children, their causative factors and response to treatment.

MATERIALS AND METHODS

A retrospective study of 56 patients less than 18 years of age patients referred for sleep disordered breathing (SDB)

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from various specialty units mainly neurology, maxillofacial surgery and pediatrics from January 1998 to July 2004 was carried out in a tertiary care referral center in Mumbai, India. Baseline limited polysomnography (PSG) using DENSA DMS-200 (UK) was done with nighttime recording of respiratory variables. The parameters considered in the study were:

- Nasal airflow detected using nasal flow sensor.
- Heart rate and arterial oxygen saturation measured by finger oximetry
- Snoring by a snoring probe.
- Thoracic, abdominal and paradoxical movements using thoracoabdominal respiratory bands.
- Limb movements using limb sensors.

Reports were analyzed with respect to the apnea-hypopnea index (AHI) and nocturnal desaturations. AHI of >5 and desaturations of $<92\%$ were considered as positive for diagnosis for obstructive sleep apnoea (OSA). Medical records of the patients with positive AHI were evaluated in detail. Symptoms noted in the records were history of snoring, nocturnal choking, arousals during sleep, excessive daytime sleepiness and nocturia. Other symptoms included abnormal movements during sleep, psychological or mood changes, delayed milestones, scholastic backwardness or poor concentration. History of atypical manifestations such as mouth breathing, irregular pattern of breathing, apneic episodes, anger spells, nocturnal enuresis, sleepwalking, speech disturbances and excessive weight gain were also available. Height, weight, body mass index, blood pressure, spirometry (available in 12 cases) with a flow volume loop (FVL) and 2-Dimensional echocardiography was obtained from the medical records. Post treatment PSG was available to 12 cases. Surgical treatment consisted of surgical release of the temporomandibular joint (TMJ) ankylosis, coronoidectomy either unilateral or bilateral, osteoentrotomy with interpositional arthroplasty with temporal fascia grafting, mandibular distraction, mandibular advancement and genioplasty either single surgery or a combination in cases of craniofacial abnormality, while those with adenotonsillar hypertrophy were subjected to tonsilloadenoid resection (TAR). Medical treatment was given in the form of positive airway pressure ventilation in cases where surgical treatment could not be offered as sleep study with these devices showed improvement in AHI.

RESULTS

Of the 56 patients included in the study below 18 years of age, 23 (41%) patients were detected to have SDB with age range of 4 to 17 years (mean age of 12.1 years). 15 (65.2%) were males and 8 (34.7%) were females. 12 patients (52.1%) were underweight, 2(8.6%) patients were overweight and only 1 (4.3%) was obese. Snoring during

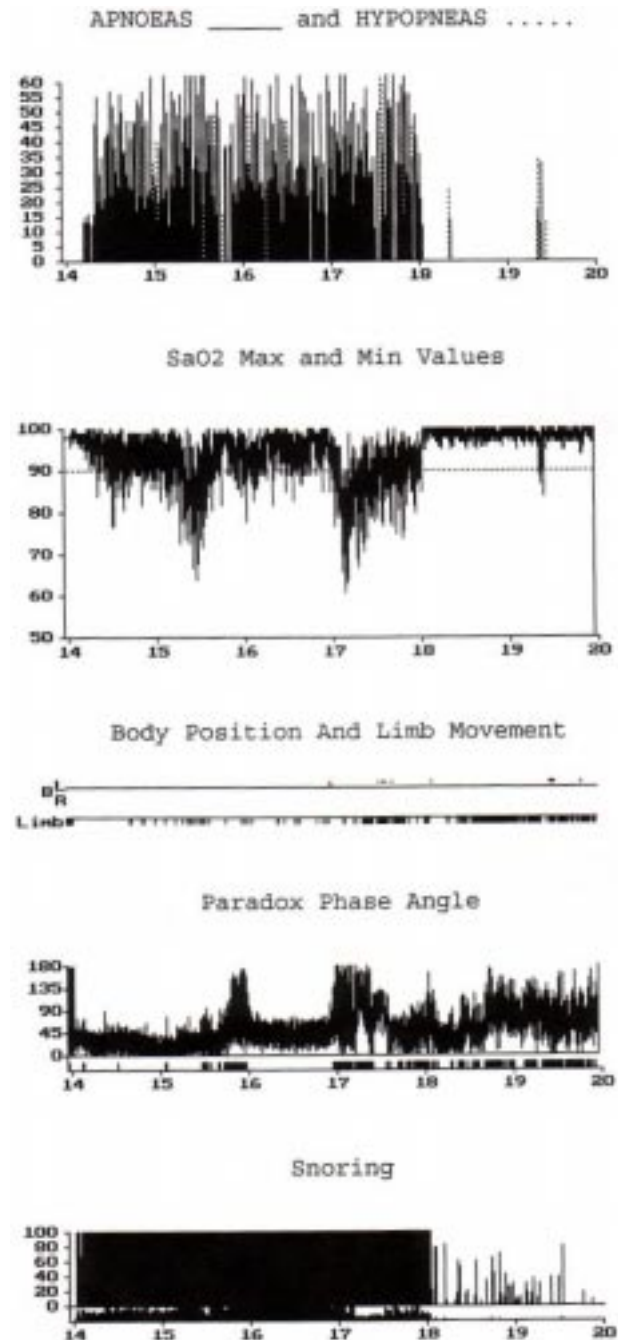


Fig. 1. Pre-operative sleep report of a case of obstructive sleep apnea (OSA) due to temporomandibular joint (TMJ) ankylosis with retrognathia.

sleep was seen in 21 (91.3%) cases, associated nocturnal choking and awakening in 4 (17.3%), excessive daytime somnolence (EDS) with early morning headache and nonrefreshing sleep in 4 (17.3%) and nocturia in 1(4.3%) case. 8 (34.7%) patients had neurocognitive symptoms in the form of scholastic backwardness in 3 (13%) cases, psychological disturbance with mood changes, abnormal sleep movements and delayed milestones in 1 case each (4.3%) and mouth breathing, apneic spells, nocturnal

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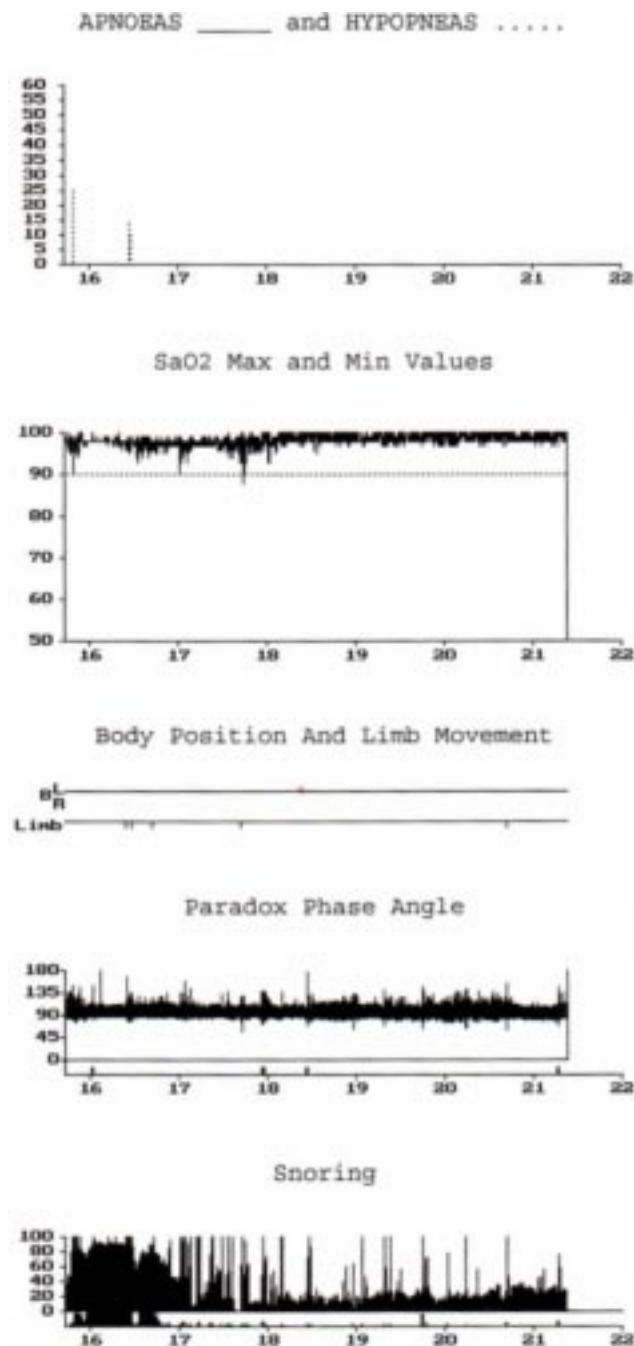


Fig 2. Post-operative sleep report of a case of obstructive sleep apnea (OSA) due to temporomandibular joint (TMJ) ankylosis with retrognathia.

enuresis, sleep walking, speech stuttering in 2 (8.6%) cases. Normal blood pressure was recorded in all cases.

Out of the 12 cases where spirometry could be performed, 5 (41.6%) showed variable extra thoracic upper airway obstruction on FVL. 2-Dimensional Echocardiography demonstrated pulmonary hypertension in 1(4.3%) case having severe micrognathia with OSA.

Craniofacial abnormalities like TMJ ankylosis, retrognathia, micrognathia or an overlap of each other was seen in 12 (52.1%) cases. Of these, 1 patient had TMJ ankylosis associated with arthrogryposis multiplex congenita. Neuromuscular and skeletal disorders causing OSA was seen in 4 of the 23 cases (17.3%). Of these 4 patients, 3 had idiopathic thoracic kyphoscoliosis and 1 patient had scoliosis associated with Marfans syndrome. Adenoid-tonsillar hypertrophy was seen in 2(8.6%) cases. Bilateral vocal cord palsy caused OSA in 1(4.3%) case. OSA in 3 (13%) cases of systemic disease was due to achondroplasia, Laurence-Moon-Bardet-Biedl syndrome, and Albright's osteodystrophy with insulin resistance diabetes mellitus. One patient with nocturnal hypercapnoea, hypoxemia, hypoventilation and hypothalamic disorder was diagnosed to have late onset congenital hypoventilation syndrome (LOCHS). All 6 cases of craniofacial abnormality where post-operative data was available showed improvement in symptoms and AHI. Both the cases of adenotonsillar hypertrophy improved post-operatively following tonsilloadenoid resection (TAR). Continuous positive airway pressure (CPAP) applied through a nasal mask was advised in patients with OSA due to vocal cord palsy, thoracic scoliosis and systemic disorders. Bilevel PAP ventilation with a backup spontaneous time (S-T) mode was advised in the case with LOCHS.

DISCUSSION

Symptoms, pathophysiology, polysomnographic findings, and treatment of pediatric OSA differ significantly from adult OSA. Adult OSA is usually associated with obesity whereas children with OSAS are usually underweight. This is because of failure to thrive and increased work of breathing during sleep. In our study 12(52.1%) cases were underweight and only 3 (12.9%) were overweight or obese. Children with OSA appear to have a deficit in arousal mechanisms. Studies have shown that these patients have elevated arousal thresholds in response to hypercapnia and increased upper airway resistance.^{3,4} As a result, sleep architecture is preserved in children with OSA and therefore excessive daytime sleepiness, the cardinal symptom of OSA in adults, is uncommon.⁵ In the present study also, excessive daytime sleepiness was observed only in 17.3% of cases. Although apnea-related EEG arousals are less common in children, sub cortical arousals and subtle disturbances in sleep architecture occur frequently.⁶ These factors may contribute to neurobehavioral and autonomic complications. Gozal *et al* have demonstrated that students of grade I in lowest 10th percentile of their class academically had an amazingly high proportion (18%) of home studies suggestive of sleep-disordered breathing, reflecting the effect of sleep-disordered breathing on intellectual function.⁷ In the present study too,

neurocognitive symptoms were seen in 34.7% cases.

In the present study, craniofacial abnormality (52.1%) was the commonest cause for pediatric OSA, followed by skeletal (17.3%), adenotonsillar hypertrophy (8.6%) and bilateral vocal cord palsy (4.3%). 13% cases had OSA associated with multisystemic disorders like achondroplasia, Albright's syndrome (pseudohypoparathyroidism with insulin resistance diabetes mellitus) and Laurence-Moon-Bardet-Biedl syndrome. Only one case was detected to have a very rare and unusual form of SDB, diagnosed as late onset central hypoventilation. The vast majority of cases of OSA in children reported in literature are associated with adenotonsillar hypertrophy, however in the present study the leading cause was craniofacial abnormality. This is perhaps because our dental hospital is a referral center for corrective surgery of craniofacial abnormality and hence awareness regarding associated SDB is high. TM joint ankylosis, retrognathia or micrognathia can cause OSA due to either congenital or post traumatic anatomical defect leading to difficult mouth opening and upper airway narrowing because of reduced inter-incisal distance. Thoracic kyphoscoliosis is often associated with sleep apnea; the 'Quasimodo's syndrome' named after the legendary hunchback of Notre Dame. There is no upper airway obstruction in cases of Quasimodo's syndrome, however, the deformed and stiff chest wall causes reduced chest wall expansion leading to alveolar hypoventilation, which gets aggravated during sleep resulting in apneas. These patients also may have an abnormal central drive for respiration causing cessation of breathing. Although childhood OSA is associated with adenotonsillar hypertrophy, large tonsils and adenoids alone do not cause it. This is explained by the fact that these patients do not obstruct during wakefulness. Studies have failed to show a correlation between upper airway or adenotonsillar size and OSA and a small percentage of children with adenotonsillar hypertrophy with OSA is not cured by TAR. Guilleminault and colleagues reported a cohort of children who were cured of their OSA by adenotonsillectomy, but developed a recurrence during adolescence.⁸ Thus; it appears that childhood OSA is a dynamic process resulting from a combination of structural and neuromotor abnormalities, rather than from structural abnormalities alone.

Untreated OSA can result in serious morbidity. Early reports documented complications such as failure to thrive, cor pulmonale, and mental retardation.⁹ These severe sequelae are less common now, due to earlier diagnosis and treatment. Even though failure to thrive is an exception these days, children with OSA still tend to have a growth spurt following TAR. A recent study found an increase in insulin-like growth factor-I following TAR, suggesting that endocrine factors play a role in postoperative spurt in growth.¹⁰ Cor pulmonale with heart failure used to be a common mode of presentation

for children with OSA but is now rare. Tal and coworkers showed a reduced right ventricular ejection fraction in 37% of children with clinically diagnosed OSA, although only 7% had clinical evidence of pulmonary hypertension.¹¹ In the present study only 1(4.3%) patient had pulmonary hypertension having OSA due to severe micrognathia. Cor pulmonale can be reversed on treatment of OSA. Systemic hypertension is a well-described complication of OSA in adults, and has been reported in a few pediatric case series. A systematic study showed elevated diastolic blood pressure in children with OSA, which could be predicted by AHI, body mass index, and age.¹² None of the cases had elevated blood pressure in the present study.

Craniofacial surgery is appropriate for some children with craniofacial anomalies. In the present study out of the 6 patients who had followed up after surgical correction for craniofacial abnormalities, improvement in sleep symptoms and AHI was seen in all (100%) cases. In a study done for the role of distraction osteogenesis in the correction of micrognathia with OSA in 28 patients, all patients were shown to have improvement following surgery with a complete curative effect in 82.1% cases.¹³ Children suffering from tonsilloadenoid hypertrophy with OSA have both symptomatic and polysomnographic improvement following TAR.¹⁴ Similarly in the present study both patients showed complete resolution of symptoms and AHI following TAR. Nasal continuous positive airway pressure (CPAP) is the most widely used and perhaps the most effective treatment for adult OSA. Positive pressure to the upper airway acts like a pneumatic splint to maintain airway patency thereby increasing lung volume, which may improve oxygenation. However limiting factors for CPAP therapy in children include lack of adequate pediatric interfaces, inappropriate behavioral techniques and need for other equipment designs for children. Young or weak children frequently do not trigger bilevel ventilators. Children may also develop central apneas or hypoventilation at higher-pressure levels.¹⁵ This is presumably due to activation of the Hering-Breuer reflex by stimulating pulmonary stretch receptors. It can be remedied by placing the patient on bilevel ventilation with a backup rate. There is also concern among pediatric practitioners that the current nasal masks can cause midfacial depression when used in very young patients. Nasal deformities have also been noted in premature infants receiving CPAP *via* nasal prongs. Sleep study with continuous positive airway pressure (CPAP) ventilation improved symptoms and AHI in bilateral vocal cord palsy, thoracic scoliosis and systemic disorders associated with OSA. Also, therapeutic trial of bilevel PAP ventilation in LOCHS showed significant clinical improvement in the present study. However, patients could not be persuaded to take long-term treatment with these devices because of technical difficulties and their cost.

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