ELSEVIER

International Journal of Pediatric Otorhinolaryngology
49 Suppl. 1 (1999) S59–S61

INTERNATIONAL JOURNAL OF Pediatric Oto Rhino Laryngology

www.elsevier.com/locate/ijporl

Management of obstructive sleep apnea syndrome in children with craniofacial malformation

Hans L.J. Hoeve [a,*], Koen F.M. Joosten [b], Sandra van den Berg [b]

[a] *Department of Otorhinolaryngology, Sophia Children's Hospital, Erasmus Medical Center Rotterdam, P.O. Box 2060, 3000 CB Rotterdam, The Netherlands*

[b] *Department of Pediatric Intensive Care, Sophia Children's Hospital, Erasmus Medical Center Rotterdam, Rotterdam, The Netherlands*

Abstract

Children with craniofacial syndromes, especially those with midfacial hypoplasia, micrognathia, or deformation of the cranial base, are frequently suffering from obstructive sleep apnea syndrome (OSAS). It is important to recognize this condition. Diagnostic methods and therapeutic developments are discussed. Experience with 31 patients in the Sophia Children's Hospital is presented. The majority of these infants suffered from moderate or severe OSAS. Treatment varied from symptomatic (e.g. continuous positive airway pressure) to curative. These therapies could often prevent a tracheotomy. Still more curative treatment options are needed.

Keywords: Airway obstruction; Endoscopy; Polysomnography

1. Introduction

Children with craniofacial syndromes are frequently suffering from obstructive sleep apnea syndrome (OSAS). It is important to recognize this condition. In this presentation the diagnostic and therapeutic possibilities are discussed.

OSAS is defined by the American Thoracic Society as a disorder of breathing during sleep, characterized by prolonged partial upper airway obstruction and/or intermittent complete obstruction that disrupts normal ventilation during sleep and normal sleep patterns [1]. The obstruction implies collapse of the oropharynx (or the supraglottis) because that part of the airway lacks a framework. The only dilating force is a neuromuscular mechanism, and that function is diminished during sleep.

Five degrees of airway obstruction are distinguished:

- Grade 1: snoring.
- Grade 2: impeded respiration, disturbed sleep, arousal, but no apnea, hypopnea or hypoxemia (upper airway resistance syndrome, UARS).
- Grade 3: apnea or hypopnea, disturbed sleep without hypoxemia (OSAS).
- Grade 4: as grade 3, but with hypoxemia (OSAS).

* Corresponding author..
E-mail address: aukes@chis.azr.nl (H.L.J. Hoeve)

0165-5876/99/$ - see front matter
PII: S0165-5876(99)00134-2

- Grade 5, OSAS with right heart failure.

In mild OSAS two to five periods of apnea/hypopnea occur per hour, in moderate OSAS five to 24, in severe OSAS >24.

The localization of the obstruction in the craniofacial syndromes varies. In Down syndrome, the upper airway is narrow and compromised by chronic respiratory infections. Other factors are macroglossy and muscular hypotony. In craniosynostosis syndromes—Crouzon, Apert, Pfeiffer, Saethre-Chotzen—deformation of the cranial base and maxillary hypoplasia result in obstruction of the nasal and nasopharyngeal airway. Syndromes with micrognathia, such as Treacher Collins, Pierre Robin, Goldenhar and many others, are characterized by obstruction at the hypopharyngeal level. Achondroplasia implies deformation of the cranial base.

2. Diagnostics

Symptoms are related to sleep—snoring, obstructive apnea, enuresis and arousal—or related to daytime—drowsiness, cognitive function disorder, failure to thrive. ENT (ear, nose and throat) examination of infants with craniofacial disorders and suspected OSAS should focus on micrognathia, macroglossia, maxillar hypoplasia, a narrow nose or pharynx, cleft palate, tonsillar enlargement, etc.

Polysomnography in indicated to confirm the diagnosis of OSAS, to establish the degree of OSAS and the need for treatment. Cardiology consultation is indicated to diagnose heart failure. The next step is endoscopy of the airway to study the dynamics of the obstruction during spontaneous breathing, and to localize the obstruction; this is important in choosing the type of treatment. Magnetic resonance imaging (MRI), computed tomography (CT) scan and plain radiograms are of limited value, because they do not demonstrate the dynamics of respiration.

3. Therapy

Therapeutic options are oxygen, continuous positive airway pressure (CPAP) via a ventilation nose/mouth mask during sleep, decongestive or steroid nose drops, nasal surgery, surgery of choanal atresia, adenotonsillectomy, prone position, nasopharyngeal intubation, tongue surgery, anterior distraction of the maxilla or mandible and a tracheotomy.

Table 1
Diagnoses of 31 patients with OSAS and craniofacial malformation in Sophia Children's Hospital

Pierre Robin	7
Down	5
Micro/retrognathia	4
Nasal stenosis	4
Achondroplasia	1
Apert	1
Crouzon	1
Charge	1
Goldenhar	1
Hunter	1
Hurler	1
Noonan	1
Orofacial digit	1
Palatoschisis	1
Rubinstein Taybi	1

4. Patients

At Sophia Children's Hospital 31 infants with craniofacial syndromes and airway obstruction were diagnosed and treated in the period 1994–1998 (Table 1). The degree of OSAS was established by means of polysomnography in the majority of cases (Table 2). Further diagnostic evaluation included radiological examination when indicated, and endoscopy of the airway. This resulted in curative treatment if possible, or symptomatic treatment. Many patients had more than one type of treatment. In Table 3 the most essential treatment per patient is presented.

Table 2
Degree of OSAS in 31 patients in the Sophia Children's Hospital

Obstruction, no OSAS	2
Mild OSAS	5
Moderate OSAS	11
Severe OSAS	8
Clinical OSAS, degree unknown	5

Table 3
Therapy in 31 patients in Sophia Children's Hospital

Curative therapy	
Adenotonsillectomy	8
Nose surgery and stents	1
Mandibular surgery	1
Removal of pharyngoplasty	1
Symptomatic therapy	
Nose drops	5
Tracheotomy	4
Oxygen	2
Nasopharyngeal intubation	2
Prone positioning	2
CPAP	1
GE reflux medication	1
No therapy	3

5. Discussion and conclusions

While in the recent past tracheostomy was considered unavoidable in many cases, this has been replaced gradually by less invasive or even curative therapies, as oxygen administration, CPAP, adenotonsillectomy, etc. The efficacy of these therapies can be monitored and adapted by means of polysomnography. This development has improved the quality of life in many infants. However, the majority of infants are still treated symptomatically because we still have too few curative possibilities. Future progress is to be found in developing more curative therapies, such as mandibular and maxillary surgery.

References

[1] American Thoracic Society, Standards and indications for cardiopulmonary sleep studies in children, Am. J. Respir. Crit. Care Med. 153 (1996) 866–878.