Severe Obstructive Sleep Apnea Syndrome with Secondary Pulmonary Hypertension after Palatoschisis Operation

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Key Words

Obstructive sleep apnea syndrome
Cheilognathopalatoschisis
Pulmonary hypertension

Abstract

We report on a 12-year-old boy with bilateral palatoschisis and obstructive sleep apnea syndrome. Patients with schistasis should be considered a high-risk group with regard to the development of obstructive sleep apnea syndrome.

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Schweres obstruktives Schlaf-Apnoe-Syndrom mit sekundärer Hypertonie nach Gaumenspaltenoperation

Es wird über einen 12 Jahre alten Jungen mit doppelseitiger Gaumenspalte und einem obstruktiven Schlaf-Apnoe-Syndrom berichtet. Patienten mit Spaltbildung sollten als Hochrisikogruppe für obstruktive Schlaf-Apnoe-Syndrome angesehen werden.

Syndrome des apnées du sommeil severe avec hypertension pulmonaire secondaire après operation d'une palatoschizis

L'auteur rapporte le cas d'un garçon de 12 ans présentant une fente palatine bilatérale et un syndrome d'apnée du sommeil obstructif. Les malades porteurs d'une fente palatine doivent être considérés comme un groupe à haut risque pour un syndrome d'apnée du sommeil.

Introduction

Obstructive sleep apnea syndrome after surgery (velopharyngeal plastic) to improve the speech of patients with cleft palate has rarely been reported in the literature. Because of abnormal development and in consequence of some of the operation procedures in patients with cleft palates, the middle part of the face shows reduced growth. Because of additional narrowness in the nasooropharynx, patients with schistasis have a high risk of developing nocturnal breathing disorders.

Case Report

A 12-year-old boy was examined to evaluate a systolic heart murmur (fig. 1). At the age of 4 and 5 years, an adenotonsillectomy and velopharyngeal plastic surgery, respectively, were carried out for sub-

total bilateral palatoschisis. Since this operation with a pharyngeal pedicle flap, a number of abnormalities have been observed including mouth respiration during the day, heavy snoring, restless sleep with dorsireclined head or in the knee-elbow position, enuresis and sweat outbreaks

during the night. Moreover, there was mental retardation. Extreme hypermotility, behavior disorders, and enuresis nocturna had already resulted in largely unsuccessful inpatient child-neuropsychiatric treatments.

On admission, the 12-year-old boy showed a good general condition. There were hypoplasia of the middle part of the face, defective positions of the teeth in the upper jaw (fig. 2), a wide pedicle of the velopharyngeal plastic, and nearly immovable velum palatinum.

Moreover, a chronic purulent rhinopharyngitis and a recurrent purulent conjunctivitis were stated. The right nostril was wider than the left. An X-ray of the paranasal sinus revealed no pathological findings. In the audiogram a low-grade conductive deafness was seen bilaterally, and in the tympanogram the curves showed a low course. Mouth breathing as well as a hyporhinophonia were noticed. All nasal sounds were of poor resonance. Sound mistakes were not recorded. There was a systolic murmur.

In the sleep laboratory a severe obstructive sleep apnea syndrome with nocturnal hypoxemia and pulmonary hypertension was diag-

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Fig. 1. A 12-year-old boy with bilateral palatoschisis. a Mouth breathing, b Hypo-plasia of the middle part of the face.

Fig. 2. a Defective positions of the teeth.

b Bilateral palatoschisis after surgery.

nosed. Therefore, loosening of the velopharyngeal pedicle was indicated.

Polysomnographically, 170 phases of obstructive apnea with a maximum duration of 84 s and a fall of oxygen saturation to 74% were observed preoperatively. Only 77% of all oxygen saturation values were > 95%. The respiratory disturbance index, i.e., the number of all apnea and hypoventilation phases per hour during sleep, was indicative of pathology. The normal value is < 15.

After loosening the velopharyngeal plastic, a significant improvement was noted.

Polysomnographically, 37 phases of obstructive apnea with a maximum duration of 68.5 s and a minimal oxygen saturation of 87% were measured. 98% of all oxygen saturation values amounted to more than 95%; the respiratory disturbance index was 9.2.

Echocardiography showed lessening of the pulmonary artery pressure. Secondary pulmonary hypertension with significant insufficiency of the tricuspid cardiac valve was determined as the cause of the heart murmur.

Because of the continuous nocturnal snoring, fragmental sleep by numerous arousals, and the hypoplasia of part of the middle face, treatment by continuous positive nasal airway pressure was indicated. This resulted in the normalization of the oxygen saturation. During sleep, 100% of all oxygen saturation values were > 95%. In the same way, the pulmonary artery pressure normalized. Nocturnal sweating, day time hypermotility, and irascible outbreaks disappeared.

Concentration also improved.

Conclusions

This case report shows that patients with surgically treated cheilognathopalatoschisis should be asked about nocturnal snoring and sweating, tiredness during the day, and concentration.

Attention should be paid to heart murmurs as a possible sign of a beginning cor pulmonale. If such symptoms are seen, an ambulatory or inpatient examination of the respiratory function is advisable to diagnose obstructive sleep apnea syndromes and to treat them early to avoid such protracted courses.

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