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.

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

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 subtotal 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.
Polysonomographically, 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.
Polysonomographically, 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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