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Review article

Management of upper airway obstruction in syndromic craniosynostosis: A lifespan approach from childhood to adulthood

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ABSTRACT

Introduction: Syndromic faciocraniosynostoses are malformations of the skull and face caused by premature closure of one or more cranial sutures, characterized by hypoplasia of the maxilla, leading to upper airway obstruction.
Objective: The aim is to provide a comprehensive review on the management of upper ventilatory disorders in the context of craniosynostosis.
Methods and results: Initial management should look for upper airway obstruction through questioning, clinical examination with flexible laryngoscopy, and, if in doubt, sleep recording to look for obstructive sleep-disordered breathing. Treatment consists of re-establishing airway patency through non-surgical treatments such as non-invasive ventilation and correcting the obstruction through various surgical procedures: fronto-facial advancement, adenoidectomy, turbinoplasty, septoplasty, etc. A tracheotomy may be necessary during early childhood, pending permeabilization of the upper airways. The age at which surgery is performed varies, depending on the individual patient, and it is not uncommon for patients to require surgery in adulthood, either as part of the initial management or for additional surgeries to improve obstruction.
Conclusion: The multidisciplinary approach is the key for managing craniosynostoses, from diagnosis to treatment, from childhood to adulthood.

1. Introduction

Craniosynostoses are malformations affecting the skull and face, caused by premature closure of one or more cranial sutures. The typical facial anomalies are characterized by more or less marked hypoplasia of the upper jaw, which hinders ventilation to varying degrees and can lead to exophthalmos and anomalies of the ocular fissures [1].

The most common syndromic craniosynostoses are Crouzon syndrome, Apert syndrome, and Pfeiffer syndrome [2]. Crouzon syndrome [2] is characterized by progressive (often multiple) craniosynostosis that usually appears around the age of two, and facial dysmorphia combining hypertelorism, exophthalmos (linked to the double retraction of the upper jaw and forehead), and an inversion of the dental articulation (Fig. 1). Apert syndrome [2] is a malformation combining craniosynostosis, bone and membranous syndactylies of the four limbs, and mental retardation. The bicoronal craniosynostosis is visible from birth, and the upper jaw is very hypoplastic, with inversion of the dental articulation. The face is broad, with a beak-like nose, constant hypertelorism, and sometimes significant exophthalmos (Fig. 2). Pfeiffer syndrome [2] combines craniosynostosis (especially bicoronal), hypoplasia of the middle stage of the face, and malformation of the hands and feet, with highly variable clinical expression. It is generally diagnosed in the neonatal period. Most affected patients present many other associated manifestations.

The objective is to provide an update on the management of upper airway obstruction in the context of syndromic craniosynostoses, from childhood to adulthood. The interest here is to emphasize the link between the initial management and long-term follow-up of patients. For

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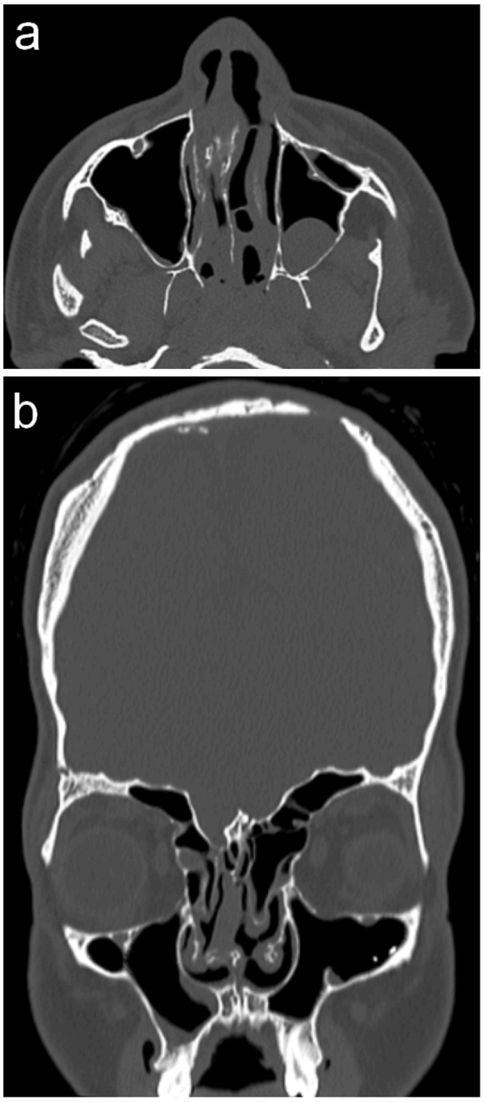

Fig. 1. Example of a septal deviation in a patient with untreated Crouzon syndrome during the growth phase, on an axial (a) and coronal (b) CT scan slice of the facial massif. It is noteworthy the midfacial hypoplasia with a narrow and deep palate, and overall narrow nasal fossae.

this, an English literature review in PubMed was carried out, using the keywords 'craniosynostosis' OR 'Crouzon' OR 'Apert' OR 'Pfeiffer' AND 'OSAS' OR 'sleep apnea' OR 'obstruction'.

2. Initial management

The management strategy for craniosynostoses begins at birth and continues into adulthood. Its objectives are to correct the craniostenosis, decompress the brain to avoid intracranial hypertension and its complications (such as blindness), correct the faciostenosis to improve ventilation, correct dental articulation disorders and orbital anomalies such as exophthalmos and hypertelorism (Fig. 2) [3]. Management is always multidisciplinary, involving at least a neurosurgeon, maxillofacial surgeon, ENT, reconstructive surgeon, orthodontic surgeon, ophthalmologist, and pneumo-pediatrician.

The search for upper airway obstruction, especially type III obstructive sleep apnea-hypopnea syndrome (OSAHS), must be systematic, through regular interviews with parents, physical exams by a pediatric ENT, including flexible laryngoscopy, and sleep recordings [4–7]. In cases of pathological sleep recordings, or when there is a discrepancy between clinical examination, history, and sleep recordings,

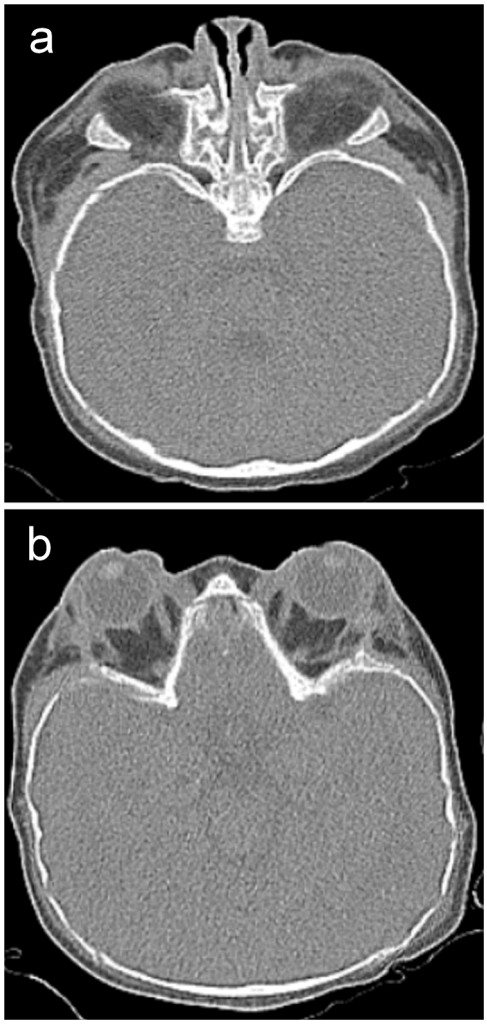

Fig. 2. Example of severe hypoplasia of the midface in an Apert syndrome in an infant, on two axial CT scan slices. (a) The nasal cavities are completely closed. (b) The exophthalmos is very significant and required appropriate management.

a morphological assessment under induced sleep—such as drug-induced sleep endoscopy (DISE)—can help identify obstructive sites and guide tailored surgical treatment if necessary. The examination under induced sleep can differ significantly from the findings observed during an outpatient consultation [8].

OSAHS is extremely common, from 7 to 67 % according to studies [6,7]. The literature lacks reliable prospective and systematic data on the prevalence of OSAHS in craniosynostoses. OSAHS can lead to growth retardation, with a low body mass index [5]. The obstructive respiratory disorder can be progressive, especially in the context of Crouzon syndrome, in which the evolution of faciostenosis is difficult to predict. Sleep recordings, and DISE if needed, must be carried out during the first weeks of life—in practice, as soon as it is feasible within the institution—, and whenever symptoms suggestive of OSAHS are observed. Ideally, polysomnography should be performed in an expert center in young children, while home polygraphy can be an alternative in older children [9,10]. Central apneas are no more frequent than in the general population [11]. The follow-up of children with craniosynostosis must be regular, in order not to overlook the onset of an obstructive respiratory disorder. Flexible laryngoscopy and sleep recordings should be repeated if the clinical history or symptoms become suggestive of obstructive respiratory disorder.

The therapeutic management of upper airway obstruction depends

on the patient's age, the craniosynostosis, comorbidities, imaging of the airways, flexible laryngoscopy, and possibly DISE [12].

In the neonatal period and early childhood, upper airway obstruction most often requires a combination of non-surgical and surgical treatments to ensure proper cerebral oxygenation and avoid irreversible brain damage or patient death.

Among the non-surgical treatments, non-invasive ventilation is most often retained when surgery is not indicated, insufficient, or has failed. Continuous positive pressure may be necessary during sleep to treat sleep disordered-breathing and OSAHS [3,13]. Appropriate interfaces are required to reduce the risks of pressure sores, eye irritation due to air leaks, and aggravation of facial deformation due to the pressure of the interface on the facial skeleton [14]. Nasopharyngeal cannulas aimed at bypassing nasal and nasopharyngeal narrowing by positioning the distal end of the tube in the oropharynx are an option to wait for surgical correction while avoiding non-invasive ventilation: they allow a reduction in the severity of OSAHS, but are not a long-term solution [15].

Surgical options are numerous for the management of craniosynostoses. In the first weeks of life, performing a tracheotomy to bypass the nasal and pharyngeal obstruction or manage an associated laryngotracheal anomaly may be essential [16–18]. The goal will then be to remove this tracheotomy when surgical treatments and the child's growth have allowed for the restoration of proper upper airway patency.

Fronto-orbital advancement surgeries can improve aesthetics, dental occlusion, exorbitism, but also the obstruction of the nasal cavities and pharynx. They are often indicated in Crouzon, Pfeiffer, and Apert syndromes, in which midfacial hypoplasia is severe [4,19–23].

Hypertrophy of the tonsils and adenoids is common, sometimes causing obstructive sleep respiratory disorders and OSAHS [24,25]. This hypertrophy is no more frequent than in the genera[l pediatric population, but its clinical impact is more significant due to the narrowness of the pharynx. A study showed that adenoid-tonsillectomy could reduce the apnea-hypopnea index by 5/h and the number of desaturations by 8.5/h, but rarely completely eliminate OSAHS in the context of syndromic craniosynostosis [25]. In cases of craniosynostosis with obstructive sleep-disordered breathing and adeno-tonsillar hypertrophy, an adenoid-tonsillectomy should be performed to improve breathing or to facilitate non-invasive ventilation, if required. If there is any doubt about the involvement of the tonsils and adenoids in obstructive sleep-disordered breathing during wakeful examination, a morphological assessment under induced sleep should be considered.

In case of nasal obstruction, other surgical procedures may be proposed based on imaging and endoscopy results: enlargement of the pyriform aperture [26] or nasal cavity, turbinoplasty [27] or turbinectomy for hypertrophy of the inferior turbinates, septoplasty for nasal septum deviation, etc.

3. Management after growth

Most of the management of craniosynostoses takes place during the growth period, and studies on adult management are rare, often being case reports or case series. Obstruction tends to regress with growth, with a reduction in the apnea-hypopnea index over time, except in patients with severe midfacial hypoplasia [7]. Once growth is complete, obstructive respiratory impairments are no longer progressive. However, it is rare for patients with severe craniosynostosis to have perfect upper airway patency.

Residual OSAHS is often present in adulthood. Management must therefore continue throughout the patient's life. This is a fundamental issue for these patients; adult follow-up must be carried out by practitioners who are fully familiar with these congenital malformative pathologies. If not managed at the time of consultation, OSAHS must be systematically sought through interview and physical examination. The physical examination must include flexible laryngoscopy to locate obstructions, which can be present at all levels of the upper airways. If OSAHS is suspected, a sleep recording must be systematically requested. In adults and older children, outpatient ventilatory polygraphy is possible, even if polysomnography remains the reference examination [9,10].

If the presence of OSAHS is confirmed, management must be adapted on a case-by-case basis and may include elements mentioned in the initial management chapter, depending on the location of the obstruction and the management already carried out. Imaging and DISE [28] are often essential complements to best tailor treatment. A correction of facial hypoplasia can be undertaken in adulthood if it was not performed in childhood [29,30]. Among the most frequent surgeries in adulthood performed, a septoplasty (Fig. 1) or turbinoplasty can improve nasal ventilation. Complete permeabilization of the nasal cavities is sometimes necessary [31]. Improving nasal ventilation helps to enhance the effectiveness and tolerance of continuous positive pressure ventilation if it proves necessary.

The literature does not provide clear recommendations on the monitoring frequency of airway obstruction in adults with craniosynostosis. It seems reasonable to assess the clinical status in consultation every two years and to repeat flexible laryngoscopy and sleep studies if symptoms suggestive of obstruction appear.

The quality of life of adult patients with syndromic craniosynostosis is impaired by the multitude of craniofacial abnormalities and overmedicalization. However, the impact of craniosynostoses on quality of life in adulthood is not clearly established; studies on this subject find disparate results [32–34].

4. Conclusion

Patients with syndromic craniosynostosis are most often managed during growth and undergo multiple surgeries. Some patients who did not benefit from an early and appropriate diagnosis and management will require interventions to correct facial hypoplasia in adulthood. Others, well monitored, may also require corrections in adulthood to improve ventilation. The multidisciplinary management requires precise anatomical assessment of the obstruction and medical-surgical management in specialized centres.

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Declaration of competing interest

The authors have no conflicts of interest to declare.

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