# Early Airway Intervention for Craniofacial Anomalies

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**KEYWORDS**

• Craniofacial anomalies • Airway obstruction • Airway intervention • Distraction osteogenesis • Tracheostomy

**KEY POINTS**

- The area of craniofacial skeleton involved in different syndromes is predictive of the airway problems typically encountered in affected patients.
- Most episodes of upper airway obstruction in children with craniofacial anomalies present in the immediate neonatal period.
- A majority of infants with Pierre Robin sequence (PRS) are able to be managed nonsurgically.
- The need for early endotracheal intubation is associated with an increased rate of subsequent surgical airway intervention.
- Tracheostomy rates are highest among children with combined midface and mandible hypoplasia.

## INTRODUCTION

The medical management of children with craniofacial anomalies is complex. Therefore, their care is best addressed by a multidisciplinary team consisting of a geneticist, pediatrician, otolaryngologist, craniofacial surgeon, neurosurgeon, dentist, orthodontist, oral maxillofacial surgeon, audiologist, speech-language pathologist, and social services provider. The otolaryngologist plays a critical role in the evaluation and management of the airway in these children.

This patient population is uniquely predisposed to upper airway obstruction. Children with craniofacial anomalies often present with varying degrees of respiratory insufficiency during the neonatal period, requiring acute intervention. The airway management of these patients should be tailored to the degree and anatomic site of obstruction. Possible interventions range from positioning maneuvers to surgical airway establishment. This article presents a classification system of craniofacial anomalies by the site of airway obstruction, along with algorithms to guide the initial management of these patients.

## DIAGNOSTIC EVALUATION

### *Bedside Clinical Assessment*

All patients with suspected craniofacial anomalies should undergo a complete head and neck physical examination, along with flexible fiberoptic laryngoscopy. The physical assessment should focus on the cranial vault shape and suture patency, maxillomandibular relation, palatal clefting, tongue position, presence of stertor,

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presence of stridor, and overall work of breathing.

An endoscopic examination is most useful to determine the anatomic location of the obstruction. Nasal endoscopy should always be performed bilaterally to ensure patency of both nasal passages. Difficulty passing the endoscope through the nasal vestibule is diagnostic of pyriform aperture stenosis, whereas the inability to pass it more posteriorly suggests choanal atresia. In children with micrognathia, endoscopy can be used to confirm the presence of tongue-base obstruction.

### *Laboratory Studies*

In select cases, laboratory studies may be helpful to diagnose subclinical respiratory insufficiency. Blood gas analysis can demonstrate elevated carbon dioxide levels, which is indicative of inadequate gas exchange and may predict impending hypercarbic respiratory failure.

### *Polysomnography*

Prior studies have demonstrated a strong correlation between obstructive sleep apnea (OSA) and craniofacial anomalies.[1] The overall incidence of positive OSA screening in children with craniofacial anomalies is 28.2%, but can affect at least 50% of children with particular craniosynostosis syndromes and facial clefts.[2,3]

In the authors' experience, however, polysomnography usually does not provide additional information that would alter management in the acute setting. The necessity of early airway intervention is most often a clinical decision based on a patient's degree of respiratory compromise and feeding ability, and is unrelated to sleep.

## CRANIOFACIAL SYNDROME CLASSIFICATION

Major craniofacial anomalies may be categorized into those with midface hypoplasia, mandible hypoplasia, combined midface and mandible hypoplasia, and midline deformities. This approach provides an anatomic classification of the most common craniofacial anomalies (**Table 1**).

**Table 1**
**Craniofacial classification**

| Site | Syndrome |
|---|---|
| Midface | Apert<br>Carpenter<br>Crouzon<br>Down<br>Pfeiffer<br>Saethre-Chotzen |
| Mandible | PRS<br>Nager<br>Stickler |
| Combined | Bilateral hemifacial microsomia<br>Treacher Collins syndrome |
| Midline | Choanal atresia<br>Midline cleft<br>Pyriform aperture stenosis |

## MIDFACE HYPOPLASIA

### *Pathophysiology*

Many craniosynostosis syndromes involve premature fusion of the cranial sutures with concomitant midface hypoplasia. The midfacial growth tends to occur more slowly and arrests prior to 10 years of age, resulting in a short anterior cranial base, acute cranial base angle, and class III occlusion.[4] Additionally, the maxilla is constricted and highly arched, which may impinge on the vertical dimension of the nasal cavity.

These anatomic abnormalities, along with the normal development of lymphatic tissue within the Waldeyer ring, leads to nasopharyngeal and oropharyngeal obstruction. The degree of obstruction can be so severe that it mimics bilateral choanal atresia during presentation.[5] In addition to a routine physical examination, all children with suspected craniosynostosis syndromes should undergo a nasopharyngeal evaluation with passage of a 5/6 French suction catheter or fiberoptic endoscope.

CT is the initial imaging modality of choice to evaluate the bony anatomy of the midface. Three-dimensional reconstructions can also be obtained to plan for future craniofacial surgery (**Fig. 1**). Finally, MRI is indicated to rule out associated central nervous system abnormalities, such as hydrocephalus or Chiari malformation.

### *Airway Management Techniques*

A previous study demonstrated that approximately half of patients with midface hypoplasia require some form of airway intervention.[6] Medical therapy with topical vasoconstrictors represents a minimally invasive intervention that can relieve mild nasal obstruction, particularly during periods of concurrent infection.

Nasopharyngeal airways, also known as nasal trumpets, offer another potential method to relieve midface obstruction. In the authors' experience, however, placement of a nasopharyngeal airway is not always possible nor beneficial in these patients. Nasopharyngeal airways themselves are space occupying and, therefore, compromise the