Congenital Abnormalities of the Temporomandibular Joint

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KEYWORDS

• Temporomandibular joint • Condyle • Mandible • Hypoplasia • Aplasia • Craniofacial • Congenital • Syndrome

KEY POINTS

- Congenital growth disturbances of the mandibular condyle can result from a variety of disorders. Contemporary management of these disorders involves a team-based approach to address associated anatomic, functional, and psychosocial issues.
- Although incidence varies widely, some of the most common disorders include mandibulofacial dysostosis (Treacher Collins syndrome), hemifacial microsomia, oculoauriculovertebral syndrome (Goldenhar syndrome), oculomandibulodyscephaly (Hallermann-Streiff syndrome), and Nager syndrome.
- The severity of the temporomandibular joint and associated craniofacial deformities, facial growth patterns, and psychosocial development dictate management and reconstruction.

INTRODUCTION

Congenital deformities of the temporomandibular joint (TMJ) complex present as a heterogeneous continuum of growth disturbances of the mandibular condyle, articular eminence, and temporal bone.[1] Such disturbances may occur in utero, specifically late in the first trimester. Any disruption in condylar development has an effect on TMJ morphology, which can result in a degree of aplasia or hypoplasia of the condyle.[2] Although complete aplasia of the mandibular condyle is a rare anomaly, congenital condylar hypoplasia is more common. It is characterized by unilateral or bilateral underdevelopment of the mandibular condyle resulting from systemic or nonsystemic interruption in growth.[3] Various molecular and developmental errors precipitate this undergrowth phenomena. Systemic occurrences are most frequently associated with the first and second branchial arches, with data demonstrating extracellular matrix proteins such as transforming growth factor-β (TGF-β) playing a pivotal role in Meckel cartilage being a template for normal mandibular development, specifically in regulating chondrogenesis and osteogenesis.[4] This article discusses diagnosis and management of several syndromes with congenital condylar deformity such as mandibulofacial dysostosis (Treacher Collins syndrome, TCS), hemifacial microsomia (HFM), oculoauriculovertebral syndrome (Goldenhar syndrome), oculomandibulodyscephaly (Hallermann-Streiff syndrome), and Nager syndrome. The extent of TMJ malformation dictates the timing and techniques of mandibular reconstruction.[5]

TREATMENT CONSIDERATIONS

Initial Evaluation

At the time of birth, treatment priorities should focus on airway stabilization, feeding, hearing,

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and vision. The airway may be compromised as a result of choanal atresia, mandibular micrognathia, glossoptosis, pharyngeal dysfunction, or laryngomalacia.[6–8] Depending on the severity of these conditions, treatment can range from special infant positioning, tongue lip adhesion, early mandibular distraction, or tracheostomy.[9–11] Patients may also require continuous apnea or oxygen saturation monitoring. A formal sleep study is necessary to document the severity and type of apnea.

Some children may be unable to maintain nutritional goals secondary to compromised deglutition or anatomic variations. This may require the use of specialty feeders, gavage-assisted feedings, or even placement of a gastrostomy tube.[12,13] Speech language pathologists and feeding specialists can help clarify the extent of swallowing and feeding dysfunction. A swallow study may be required in certain cases. Airway and feeding compromise are most common in patients with TCS.

Evaluation by the pediatric otolaryngologist, in combination with initial audiologic testing (auditory evoked brain responses), is essential to clarify the presence of neurosensory hearing loss. When moderate-to-severe skeletal malformations are present, a full craniofacial computed tomography (CT) scan (axial and coronal slices) should be obtained during the initial 6 months of life. CT should include the superior aspect of skull through the cervical spine with 3-dimensional reformation. Special focused views through the petrous (temporal) bones are required to document external auditory canal and middle and inner ear anatomy.[14]

A pediatric ophthalmologic evaluation is useful to assess extraocular muscle function, corneal exposure difficulties, visual acuity, and adnexal region malformations. Depending on the degree of eye exposure, early tarsorrhaphy may be necessary to prevent corneal scarring, ulceration, and blindness.[14]

Treatment Sequence

Once the child is stable (ie, feeding and breathing without significant obstruction and with adequate corneal protection), the remaining craniomaxillofacial dysmorphology can be addressed using a staged, coordinated approach.

At about age 3 months, epibulbar limbal dermoids associated with OAV are commonly removed to obviate the development of amblyopia.[15] The current approach to the correction of craniofacial deformities is to stage reconstruction to coincide with facial growth patterns, visceral function, and psychosocial development. Precise morphologic analysis of each patient and recognition of the need for a staged reconstructive approach serve to clarify the objectives of each phase of treatment for clinicians and family.[16] Cranio-orbitozygomatic bony reconstruction is typically performed at 7 years of age when bony maturation of this region is near completion. Orbitozygomatic reconstruction for patients with moderate-to-severe forms of TCS can be carried out using full-thickness calvarial bone grafts. Surgical exposure is achieved using a coronal incision without the use of periorbital incisions. The cranial bone grafts are contoured to form a new zygomatic complex and reconstruct the orbits. Once adequate reconstruction is achieved, bone grafts are fixated with miniplates and screws. The cranial vault donor sites are reconstructed with fixed split-thickness autogenous grafts or bone substitutes.[16]

Kaban classification, altered facial height, horizontal mandibular deficiency, and chin dysplasia guide timing and technique of final jaw reconstruction. When excessive clockwise rotation of the maxillofacial complex results in an angle class II malocclusion with anterior open bite, orthognathic surgery is required.[17–23] This takes place during early skeletal maturity (on average, 13–15 years of age). Occasionally, orthodontic preparation includes extraction of mandibular and/or maxillary first premolars to improve dental crowding and normalize the inclination of the incisor teeth in preparation for repositioning of the jaws.[16,20,24]

Kaban types I and IIA mandibular deformities are best reconstructed at the time of early skeletal maturity (13–15 years) via sagittal ramus osteotomies of the mandible in combination with a Le Fort I osteotomy and genioplasty. In cases of Kaban type IIB deformities, the anomaly may be severe enough to consider early anterior repositioning via intraoral sagittal split osteotomies, extraoral inverted L ramus osteotomies, or mandibular distraction[25–32] (**Figs. 1–3**). Even in cases where early mandibular distraction is performed, patients should expect an additional mandibular procedure following growth maturation.[16] Nasal reconstruction should be performed as the final reconstructive procedure.

The Kaban type III condyle/ascending ramus deformity is generally present in conjunction with glenoid fossa hypoplasia. Reconstruction of the glenoid fossa and ramus/condyle unit is best performed in conjunction with zygomatic and orbital reconstruction with the use of a bilateral costochondral graft (CCG). During the operation, the distal mandible is repositioned anteriorly and held in place by intermaxillary fixation through a