SECTION EDITOR: WALTER W. TUNNESSEN, JR, MD

Picture of the Month

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AN 8-YEAR-OLD GIRL was referred for evaluation of congenital facial deformities. She was born after an uneventful pregnancy during which her mother received no medications and was not exposed to radiation. There was no family history of congenital anomalies.

On physical examination the girl had unilateral macrostomia as a result of a transverse right stomal cleft and a severe deformity of the right auricle, including supernumerary auricular cartilage anterior to the tragus and absence of the right ear canal (**Figure 1** and **Figure 2**). The left ear was normal. There was no evidence of midfacial skeletal hypoplasia. The ascending ramus of the mandible and the temporomandibular joint were normal. The palate was intact. Conductive hearing loss was present in the right ear. The remainder of the physical examination results were unremarkable.

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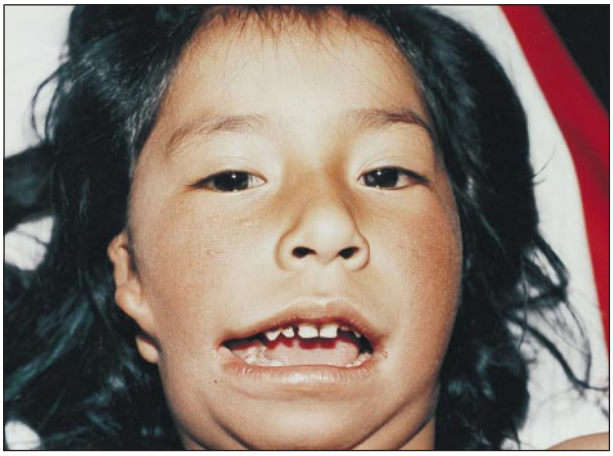

Figure 1.

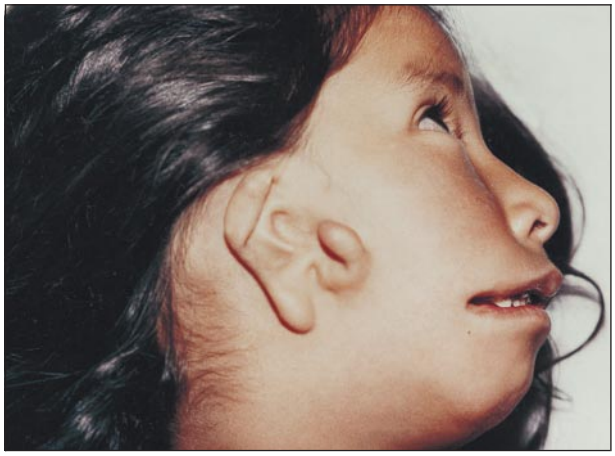

Figure 2.

Denouement and Discussion

First Branchial Arch Syndrome

Figure 1 and Figure 2. *The right corner of the mouth is extended in a cleftlike fashion. The right auricle is severely deformed and the ear canal is absent.*

Congenital macrostomia or transverse facial cleft is a rare congenital craniofacial anomaly usually associated with deformities of structures derived from the morphogenesis of the first and second branchial arches. Transverse facial clefts account for only 1 in 100 to 1 in 300 of all patients with facial clefts.[1] The first and second branchial arch syndrome was the name proposed to denote anomalies that include unilateral macrostomia, hemignathia, external ear deformities, and associated hypoplasia of the midfacial and temporal bones.[2] The ear deformity may involve the external, middle, and inner ear structures, and may vary from supernumerary auricular cartilages to total ear atresia.[3]

Various names have been applied to congenital defects involving structures derived from the first and second branchial arches in association with other anomalies, including the oculo-auriculo-vertebral spectrum, hemifacial microsomia, Goldenhar syndrome, and the facio-auriculo-vertebral spectrum.[4] Goldenhar syndrome is associated with epibulbar dermoid and vertebral anomalies in addition to the facial structural abnormalities.

PATHOGENESIS

Unilateral first and second branchial arch syndrome is almost always sporadic in occurrence.[4-6] Facial features begin to develop during the fourth week of embryonic life and differentiation of structures progresses through the eighth to ninth week. The first branchial arch is divided into maxillary and mandibular processes separated by a natural cleft, the stomodeum. Ventrally, the first arch components surround the primitive mouth. Gradually, the 2 processes fuse in a posterior to anterior manner and separate from alveolar processes to form the cheeks and lips. Some evidence suggests that disturbance in the embryonic blood supply, the stapedial artery that provides temporary blood supply to this area in the embryo, during mesechymal ingrowth could result in first arch deformities in the developing face.[6]

DIAGNOSIS AND MANAGEMENT

Regardless of the cause, the mouth deformity is one of muscle, subcutaneous tissue, and skin. The deformity is particularly evident when the affected child cries or laughs because the absence of the orbicularis oris muscle allows a wide gapping of the commissure. Supernumerary auricular cartilage anterior to the tragus and anomalies in the ipsilateral ear lobule are present in almost all individuals with this syndrome.[3,7]

The operative technique for repair of the deformity of macrostomia should be designed to reconstruct the muscular continuity of the orbicularis, preserve the rounded commissure with vermilion, and minimize scar formation. Various surgical procedures have been proposed for repair of the oral defect.[1,8,9] The standard correction, using an overlapping myoplasty at the commissure and a Z-plasty for the skin closure, is the best method to prevent postoperative scar contracture and accurate good lip symmetry.[3] If a severe ear deformity is present, reconstruction of the ear should be delayed until the child is old enough to understand the importance of protecting the reconstructed ear. A silicone rubber prosthesis or a free graft taken from costal cartilages could be necessary.[5,9]

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