**INTRODUCTION**

Craniofacial clefts are rare among facial anomalies, with an incidence of 1.5 to 5 per 100,000 births and 1 per 100 cases of cleft lip and palate. Tessier has provided a classification system for facial clefts, numbered 0-14. Many of these clefts involve the mouth, but the orbit is generally considered the referencing landmark when classifying these clefts. Transverse facial cleft (Tessier type 7), also known as congenital macrostomia, occurs with an incidence of 1:3000 to 1:5642 births. This is a result of failure of the maxillary and mandibular portions of the first branchial arch to unite. Males are more frequently affected than females. Bilateral involvement is rare (10-20% of cases). This finding has been termed hemifacial microsomia, craniofacial microsomia, first and second branchial arch syndrome, and otomandibular dysostosis. Clinical expression is variable. In microform cases, a preauricular skin tag can be present. In the complete form, the cleft begins as a macrostomia at the oral commissure and continues across the cheek toward a microtic ear. All soft tissues may be underdeveloped on the affected site. Osseous manifestations also cover a wide range.

The morphology of facial clefts can be explained on the basis of early embryonic development of the face. If the arrest occurs before the different processes have merged at the 17-mm crown-rump length stage, the lacrimal canal will fail to form, and a primary or transformation defect will be formed such as primary clefts, Tessier numbers 3 and 7. If the disturbance takes place after closure of the ectoderm of the face has been completed and a canal has been produced before the end of the differentiation phase at 60-mm CRL stage, a secondary or differentiation defect will result in secondary clefts, Tessier numbers 1, 4, 5, 6, 8, 9, 10, 13, and 14.

**CASE REPORT**

The patient is an African American male who was born at 40 weeks by vaginal delivery. He weighed 8 pounds at birth. His mother received prenatal care for the last 3 months of her pregnancy and she had no history of STDs. She was Group B Streptococcus positive but did receive antibiotics while she was in labor. There was no family history of facial deformities. The patient was hospitalized for 7 days at birth for feeding difficulties. He was then discharged home, where he lived with his parents and four siblings. He was readmitted at 15 days of age to the Pediatric service for stridor. He was noted to have persistent hypercarbia. His weight was in the 25th percentile, his height was in the 50th percentile, and his head circumference was in the 10-25th percentile. Nissen fundoplication and G tube were performed, and his stridor was noted to subsequently improve.

The Otolaryngology service was consulted for facial deformity, micrognathia, and microcephaly. On physical exam, he was noted to have a band of tissue that connected the right oral commissure to the oropharynx. It was felt that the vibrating of this band may have been contributing to his noisy breathing. The right auricle appeared normal, the mandible was intact, and there were no abnormalities of the facial nerve.

Echocardiogram was within normal limits. However he continued to have hypercarbia, and tracheostomy was performed at 4 months of age. At 8.5 months of age, the patient underwent direct laryngoscopy, bronchoscopy, and examination of the ears. He was noted to have a 1.5 cm cleft at the right oral commissure, a 1 cm wide band of keratinized epithelium on the buccal mucosa extending to the superior pole of the tonsil and soft palate (Figure 3), and bilateral acute otitis media. The hard palate appeared intact. Bilateral PET were placed in at one year of age for recurrent otitis media. The right one spontaneously extruded. Modified barium swallow at 1 ½ years of age showed no evidence of airway penetration or aspiration. He was seen in Craniofacial ENT clinic and noted to have 4+ tonsils and intact palate. At age 1 ½ years, the G tube was removed and mother noted choking spells since that time. Modified barium swallow showed no signs of aspiration. At age 2 years, the patient underwent repair of facial cleft, myringotomy, and placement of bilateral PE tubes. The facial cleft was repaired using the Skoog technique. The intracranal skin was excised and the mucosa was closed primarily. Z-plasty was performed of the right cheek skin. On postoperative day 1, the patient was tolerating oral intake. He continues to have his tracheostomy and speaks well with a passy muir valve. At the age of 3 years, the patient underwent placement of right PE tube, direct laryngoscopy, and bronchoscopy to evaluate for decannulation. There were no noted abnormalities on exam and he was to be decannulated in the recovery room. However, he failed decannulation secondary to hypercarbia. Flexible scope exam one month later showed no laryngomalacia or anatomic defects.

DISCUSSION

We describe a patient with Tessier 7, severe GERD, persistent middle ear effusions, and persistent hypercarbia requiring tracheostomy. This ignited our interest in the comorbidities that have been reported in association with Tessier 7. Among those found in a literature review are anophthalmia, preauricular skin tag, prominent ear deformity, type la hypoplastic mandibular ramus, absence of facial nerve function in the distribution of the marginal mandibular nerve, oblique clefts of the soft palate, symmetric abnormal structure of the zygomatic arch, absence of medial or lateral pterygoid plates, and Treacher Collins syndrome.

Other authors have noted associated anomalies with Tessier clefts (Table 1). Interestingly, Schienker examined 90 children with cleft lip and palate and noted a 23-27% incidence of abnormalities related to the eye and lacrimal apparatus, including colobomas of the iris. The great majority of patients with cleft lip, cleft palate, and facial cleft have normal intelligence. Senan reported an interesting case from India of a child with a Tessier 3 cleft. Tessier 30 involves lower midline facial cleft or the median mandibular cleft. Only 66 cases have been reported worldwide. This was diagnosed in a 13 year old boy who presented with a midline neck swelling of one year duration. Pathologic examination revealed gastric glands and lymphoid follicles in the submucosa of the specimen.

Sigler reported a case of a 6 month old boy with Tessier 2,3, and 7 clefts and a median lip pit. Lip pits, along with cleft lip and palate, are thought to be part of various syndromes, including Van der Woude syndrome. The primary feature in VDWS is symmetrical pits or eminences in the vermilion border of the lower lip. In 70% of cases, lip pits are associated with cleft lip with or without cleft palate or cleft palate alone.

Our literature review located reports of many anomalies of the head and neck region associated with Tessier clefts, but did not uncover any reports of respiratory illness or middle ear disease in patients with Tessier 7.