Facial Reanimation in a Child with Branchio-Oculo-Facial Syndrome

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ABSTRACT

OBJECTIVES:
1. Review the features of branchio-oculo-facial syndrome (BOFS), including congenital facial paralysis and cleft lip.
2. Outline the surgical options for rehabilitation of bilateral congenital facial paralysis and how these may or may not be influenced by concurrent perioral scarring and microstomia following repair of bilateral cleft lip.
3. Describe the outcome following bilateral transoral orthodromic temporalis tendon transfer in a child with BOFS.

METHODS: Case report and literature review.

RESULTS: Branchio-oculo-facial syndrome is a rare condition characterized by colobomas, orofacial clefting, microtia, and characteristic facial features that may include congenital facial paralysis. We discuss a child with BOFS who presented for bilateral cleft lip revision at age 8 and after being followed for several years, ultimately underwent bilateral orthodromic temporalis tendon transfer through a transoral approach at 12 years of age. One centimeter fascia-lata jump grafts were used on either side. Following the operation the parents and child both reported improved speech intelligibility as well as the establishment of a dynamic smile.

CONCLUSION: Even in the context of perioral scarring related to repaired and revised bilateral cleft lip, a transoral approach to bilateral orthodromic temporalis tendon transfer was possible in this child and allowed for successful creation of a symmetric and dynamic smile.

INTRODUCTION

Branchio-oculo-facial syndrome (BOFS) is a rare, autosomal dominant condition with variable expressivity, caused by mutations in the TFAP2A gene. It affects the facial and neck structures in particular due to insufficient development of the first and second branchial arches. It is characterized by malformations of eyes and ears, with distinct branchial, ocular, and craniofacial characteristics including cleft lip and palate. Along with these deformities, BOFS often involves facial paralysis. As a consequence, patients with BOFS have difficulty producing intelligible speech and dynamic facial expression.

There are a number of surgical options for treatment of bilateral congenital facial paralysis. These include free tissue transfer, regional muscle transfer, nerve grafting, and nerve anastomosis. In recent years, orthodromic temporalis tendon transfer (OTTT) has become a popular surgical technique for facial reanimation. Even though OTTT is well studied and developed in adults, very few surgeons have employed it in children.

DISCUSSION

While many other facial reanimation procedures require multiple steps, OTTT has become increasingly popular as a single-step option that affords nearly immediate results. Orthodromic transposition techniques in particular, have gained favor owing to improved muscle excursion and power. Additionally, operative time is shortened, owing to the surgery’s relative simplicity. In adults, OTTT is commonly performed through an incision in the nasolabial fold with or without a fascia-lata graft. As children typically lack a well-developed nasolabial fold, a transoral approach is favored in the pediatric population.

CONCLUSIONS

This case report suggests that a history of perioral scarring and microstomia in the context of multiple prior cleft lip surgeries may not limit the functional outcome of OTTT, and that a transoral approach was still possible within the context of limited mouth opening.

CASE REPORT

A 12-year-old boy with Branchio-Oculo-Facial syndrome underwent endoscopically-assisted OTTT surgery to reanimate the paralyzed face. His past surgical history was notable for bilateral cleft lip repair as well bilateral excisions of branchial and preauricular sinus tracts in infancy. As a toddler he had undergone cleft rhinoplasty. After presenting to the senior author’s clinic at age 8 he underwent a major revision cleft lip repair with a complete take down. Even within the context of relative microstomia owing to multiple prior lip and nose procedures, the OTTT surgery was attempted via a transoral approach. A 1 cm fascia lata jump graft was utilized on either side with tissue harvested from the left upper leg. A transoral incision was made down to the ascending ramus with dissection toward the coronoid process and sigmoid notch (Figure 1). The temporalis tendon was isolated and coronoidectomy was then performed. Fascia lata grafts were secured to the temporalis, tunneled to perioral incisions, and secured to the modiolus of the orbicularis oris muscle (Figure 2). A Peña muscle stimulator was used to simulate the temporalis muscle and verify function. A similar procedure was performed on the contralateral side.

Following the operation the patient and his parents reported markedly improved speech intelligibility and the establishment of dynamic smile (Figures 3, 4A, and 4B). The beneficial effects of this surgery have persisted through the 2 year follow up interval.

REFERENCES