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Purpose: Parry Romberg's Disease is an enigmatic craniofacial disorder characterized by progressive facial atrophy. Early age of onset is associated with significant skeletal involvement and jaw abnormalities in 30-65% of patients, with up to 31% of patients requiring osseous reconstruction. ^{1,2,3,4} Standard reconstructive treatment allows the disease to "burn out" with 2 years of no change prior to reconstructive intervention. We have found that early intervention in active disease is beneficial.

Methods: All pediatric free tissue transfer cases (1989-2013) performed by the senior author (J.W.S.) were reviewed. Free tissue transfer of a circumflex scapular variant flap was performed to treat deformities arising from Romberg's hemifacial atrophy.

Results: In our series, 25 patients between the ages of 3-7 were operated on even in the presence of aggressive active disease. Follow up ranged from 1- 23 years. There were no flap takebacks or flap loss. There was one limited hematoma that was treated with bedside suction evacuation. There was one patient with preauricular incision dehiscence at the time of suture removal requiring resuturing. Flap revisions and secondary procedures were performed at 6 months following free tissue transfer including tissue rearrangement, debulking procedures, resuspensions, lip reconstructions, and minimal autologous fat transfers. In all patients the disease process seemingly halted and no disease process has recurred to date.

Conclusion: Microsurgical correction of Parry Romberg's Disease in young children is safe, reliable, and effective. It allows for correction of both severe and subtle deformities. Facial skeletal growth was markedly better than expected. These young children with severe disease would have classically gone on to have more severe skeletal deformities. Although one child had orbital repositioning, no other children who have undergone early free flap reconstruction have subsequently gone on to require any further osseous reconstruction. We theorize that the interposition of normal healthy tissue may halt or alter the disease process, preventing the skeletal hypoplasia that is classically seen. Biologic studies are currently underway to validate this surprising outcome. Ultimately, we feel strongly that young children with severe rapidly progressing disease should undergo early reconstruction with free tissue transfer to spare them the sequelae of a disease process that has been allowed to burn out, or the repetitive treatments required for fat grafting over a prolonged period of time.

References

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Pre and Postoperative Photos