

## Correction of Enophthalmos in Progressive Hemifacial Atrophy: A Case Report

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**Summary:** This case report describes a technique used for correction of enophthalmos secondary to progressive hemifacial atrophy (Parry-Romberg syndrome). The only previous described technique utilized an orbital floor implant, but this method was apparently only partially successful in correcting the conditions, i.e., it did not correct both the enophthalmos and the pseudoptosis that occurs secondary to intraorbital fatty atrophy. In the present technique, the periorbita was cut at two equators, the globe and anterior periorbita advanced forward, and the resulting empty spaces filled with thin slices of radiated cartilage. Both the enophthalmos and the pseudoptosis were corrected in a single operation. There were no long-term complications and the correction has been maintained 3 years postoperatively. **Key Words:** Progressive hemifacial atrophy—Parry-Romberg syndrome—Enophthalmos.

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Progressive hemifacial atrophy, Parry-Romberg's syndrome, is a relatively rare condition, about which a great deal has been written. Several review articles discuss, in particular, ophthalmological aspects of this condition (1-3). Various treatments have been reviewed in several articles (2-4). In one article, enophthalmos was described as being the most common of the ophthalmological problems associated with the condition. These authors, however, made no attempt to correct the enophthalmos since they did not believe there to be safe effective procedure available at that time (3). However, in 1977, Smith and Guberina (4) at Manhattan Eye, Ear, and Throat Hospital had described correction of the enophthalmic globe by the use of a subperiosteal silastic block. The patient had had this placed in 1975 at the New York University Hospital. Interestingly, in this patient, a levator resection was necessary to correct the blepharoptosis (4).

This case report describes another treatment that corrects both the enophthalmos and the pseudoptosis associated with Parry-Romberg's syndrome,

using a single surgical procedure. The treatment lends itself to correction of the ptosis probably by anterior repositioning the orbital contents under the levator mechanism (muscle and tendon).

### CASE HISTORY

The patient, a 23-year-old, first noted a change in the right side of her face at ~16 years of age. She noted a slowing progressive grooving over the right chin soft tissues that extended up over the right cheek and onto the forehead. The right eye slowly became more deeply set.

Examination confirmed all of the above Fig. 1, **left**, and additionally demonstrated a small crown on the right maxillary lateral incisor, suggesting that her condition had started much earlier. The lateral incisor is usually exposed and, except for final root development, should be complete in growth by age 8-9 years (crown development, Nola stage five, usually complete by age 5) (5). She had no signs of any ocular complaints other than those affecting her appearance: enophthalmos and pseudoptosis. There was no diplopia or excessive tearing. She was, however, somewhat obstructed in superior gaze by the resulting ptosis of the upper lid seemingly secondary to the enophthalmos. An ophthalmological examination showed no other pathologic findings.

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**FIG. 1.** A–B: Preoperative view of the patient showing the enophthalmos and the right upper eyelid blepharoptosis. C–D: Two years postsurgery, long-term result, showing maintenance of correction of the enophthalmos and the blepharoptosis.

Magnetic resonance (MR) scans demonstrated enophthalmos, significant decrease in intraorbital fat, and a serpentine course of the optic nerve (Fig. 2). This suggested that, with the fatty atrophy, there was a posterior settling of the orbital contents causing the enophthalmos. No bony orbital changes

were noted in these scans unlike those in a previous report (6).

The corrective operation was approached through a bicoronal incision. The entire right periorbital area, except for the contents of the superior and inferior orbital fissures, the lacrimal sac, and



**FIG. 2.** MRI scan of the orbits showing loss of fat on the right and the associated serpentine course of the optic nerve.

the medial canthal ligament, was dissected back to ~0.5–1.0 cm from the optic nerve canal. The periorbita was then incised around the entire circumference of the orbit, exposing the underlying fat, at two equators (Fig. 3). Two (as opposed to one) circumferential cuts were made to facilitate pulling the anterior periorbita forward. It was believed that this would distribute the soft tissue over a greater area. The anterior incision was ~5 mm behind the orbital rim margin and the other ~6–8 mm behind the first. With alternating forcep traction at opposite polls, first at 3:00 and 9:00 o'clock then at 12:00 and 6:00 o'clock on the most anterior periorbita, the periorbita was pulled forward, stretching the orbital contents but only, hopefully, straightening the optic nerve. Then, to maintain the orbital contents forward and to compensate for the volume shift, multiple thin slices (each ~1 mm thick) of radiated rib cartilage were placed posterior to the equator of the globe around the orbital cavity. This was done until the right globe was overcorrected by ~2–3 mm when compared to the left.

Pericranial tissue was used to augment the clefts at the chin and forehead defects.

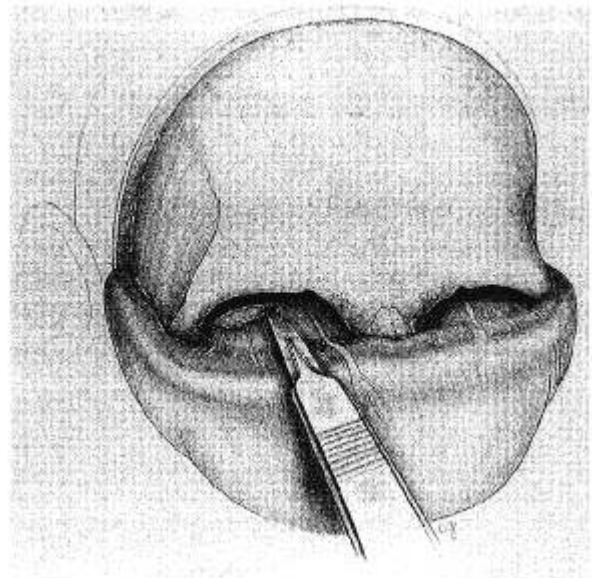
The postoperative course was totally uneventful, except for a brief period (~3–4 weeks) of some vertical diplopia in inferior gaze. This diplopia re-

solved spontaneously. The patient's final long-term result has been totally satisfactory with near-normal right orbital position and significantly improved appearance (Fig. 1 **B**). The patient has approved no follow-up, postoperative MRI scans as she remains asymptomatic and is happy with her appearance.

## DISCUSSION

Enophthalmos, whether it is the result of trauma (loss of fat or an increase in bony orbital volume), radiation (e.g., for retinoblastoma), congenital (e.g., hemifacial microsomia), or a developmental condition (e.g., this case), has been, in my experience, of considerable concern to the patient. Each cause has its appropriate treatment e.g., for trauma, reconstruction of the bony orbital cavity (7).

To correct enophthalmos secondary to a developmental problem in which the orbital walls, but not the orbital contents, are normal is a significant problem. To begin with, the basic pathologic problem cannot be corrected. The fat cannot be returned to the orbital contents. Autologous fat injections are only partially successful at any site, but injection of fat into the orbit, behind the globe, would seem to be extremely unwise and risky. To correct the soft tissue volume deficiency by decreasing only the bony orbital volume (bringing in the lateral orbital wall—the varus maneuver), could work, but



**FIG. 3.** This illustration shows the beginning of the first circum-periorbital incision with extrusion of fat. This excision extended completely around the periorbita. A second incision was made more posteriorly by ~6–8 mm.

would be difficult to control, is a more extensive procedure, and it may cause a lateral orbital rim deformity. Since, in this particular case, the condition started in childhood, based on her tooth development, there probably was some periosteal or other soft tissue deficiency. Decreasing the bony orbital volume alone by adding a hard material probably would not have allowed the orbital contents to be forced forward as desired; for example, in Smith's case (4), the blepharoptosis remained uncorrected following placement of a silicone block. Therefore, it was my opinion that the anterior periorbital should be repositioned, i.e., the periosteum should be incised, and the orbital contents delivered forward by traction on the periosteum (of course, not so much as to injure the optic nerve) and maintained there by decreasing the bony orbital volume (by use of the radiated rib cartilage). The true soft tissue orbital volume has not been changed but rather, has shifted forward and is maintained there by the hard tissue (cartilage) grafts.

Various materials have been utilized to reconstruct the orbital walls. Bone is now, generally, the honored material, especially cranial bone (7,8) but in my and others (9) experience, the long-term results are not predictable due to irregular and excessive bone graft resorption. For several years, I have used radiated cartilage for this maneuver. Thin slices of radiated cartilage can be placed nicely in the appropriate position in the posterior orbit. Radiated cartilage is bacteria and vira-free, and I have not seen any infections around it. Furthermore, it does not seem to resorb. In this case and one other, a single piece of the slippery cartilaginous material migrated forward and became obvious at the lateral infraorbital rim, such as frequently occurs with silicone sheeting, and had to be trimmed.

More recently (in the past year), I have been using thin pieces of Medpor, 0.85 or 1.5 mm thick. A recent article by Romano et al. (9) has shown their encouraging results from the use of this material. It is, presently, my material of choice as a substitute for intraorbital bone augmentation in most (but not all) cases. Medpor, however, seems to cause a period of increased edema and could, possibly, be problematic in some cases. This prolonged edema from Medpor has been noted in several of my patients in whom it has been used for cheek augmentation or nasal reconstruction.

The pseudoptosis was self-correcting, as would be expected, as the orbital contents came forward,

with tissue advancement under the levator muscle, tendon, and upper eyelid. To treat the ptosis before advancing the orbital contents would seem to be the wrong treatment based on the wrong diagnosis. Two negatives would in this case not yield a positive. It is difficult for me to understand how the levator palpebral muscle could possibly lengthen during the progression of this condition when most everything else is getting smaller and shorter.

It has been 3 years since this woman's orbit was reconstructed. There have been no long-term problems and the result has been maintained. The only short-term problem was that of diplopia, which corrected on its own and has remained so. It is possible, in a procedure in which a hard material is used posterior to the globe equator to force it forward, that some degree of diplopia might remain. If this were to occur, an extraocular muscle balancing procedure might be necessary. This method of treatment for enophthalmos secondary to fatty atrophy in Parry-Romberg's syndrome would seem to be most appropriate approach, since it is associated with both functional and aesthetic improvements.

#### SUMMARY

Progressive hemifacial atrophy has, as one of its common findings, enophthalmos. The enophthalmos in this case appeared to be secondary to intraorbital fatty atrophy. Another possible cause of the enophthalmos is enlargement of the orbit through bony changes, but computed tomography (CT) scans did not show any in this case. Pseudoptosis, which is also a common finding, probably occurred along with the enophthalmos as the soft tissues migrated posteriorly under the levator muscle, thus causing the small amount of ptosis. There were no other ophthalmological problems noted. The only previous treatment described for the enophthalmos associated with this condition is placement of a subperiosteal orbital floor silastic block followed by ptosis repair.

In this patient, treatment consisted of circumferential incision of the periorbital at two equators, and advancing the periorbital with the contained orbital contents forward and maintaining it forward by compensating for the fat atrophy by filling the now-empty space with thin slices of radiated cartilage. Both the enophthalmos and the pseudoptosis were thus corrected in a single maneuver. There were no long-term complications from the surgery, and the

long-term result has been maintained now for 3 years.

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