

Macular Unfolding for Managing Macular Folds in Infants With ROP

This surgical technique improved and restored vision in a small series of infants with regressed retinopathy of prematurity.

BY EHAB EL RAYES, MD, PhD

Tractional macular folds in infants are the result of continuous traction on the baby's elastic retina. This can occur in cases of regressed retinopathy of prematurity (ROP), peripheral toxocariasis, or exudative vitreoretinopathy. The problem is often seen in infants successfully treated for active or regressed ROP; however, if the fovea is involved in the folds they face the risk of loss of fixation.

Macular tractional folds in ROP are a result of the organization and contraction of the primary vitreous. Cells of the primary vitreous or tunica vasculosa lentis or both may also contribute cells to this hypocellular contraction of the vitreous, leading to creation of a vitreous strand (condensation stalk) that drags the macular tissue toward the periphery.^{1,2} This is clinically seen as the shifting of nasal blood vessels toward the macula and straightening of the temporal blood vessels, forming a fold in the macular area that causes the fovea to detach. The fold looks like a napkin in a wine cup because the stretched, elastic retina has more surface area than the underlying choroidal surface area (Figure 1). This condition is challenging to treat because the surrounding retina is attached. This article presents a technique for reattaching and redistributing the macula back in place to restore foveal fixation for the infant's developing vision.

TECHNIQUE AND CLINICAL EVALUATION

The surgical hypothesis for this technique is based on dissecting and opening of the central fold and undermining the edges to redistribute the tissue and ultimately reposition the fovea back into place. The repositioned tissue is redistributed outside the vascular arcades as excess tissue.

In this technique, which we have performed on a series of 16 infants (age range, 2 to 5 months) over the past

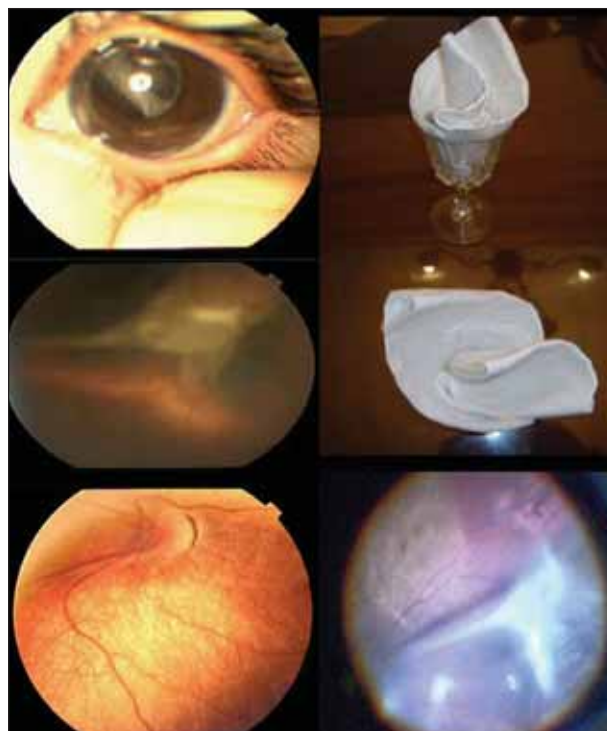


Figure 1. A composite picture comparing several forms of infant macular folds to a napkin in a wine cup.

6 years, lens-sparing vitrectomy is performed. After removing the core vitreous, I release the peripheral point of fixation at the trough where the fold is drawn by the vitreous strand, representing the pillar of the detachment. This is aided by peripheral scleral indentation by the assistant to avoid lens damage during peripheral dissection.

Next, I bimanually dissect the sheet of condensed primary vitreous of the fold, which usually appears as

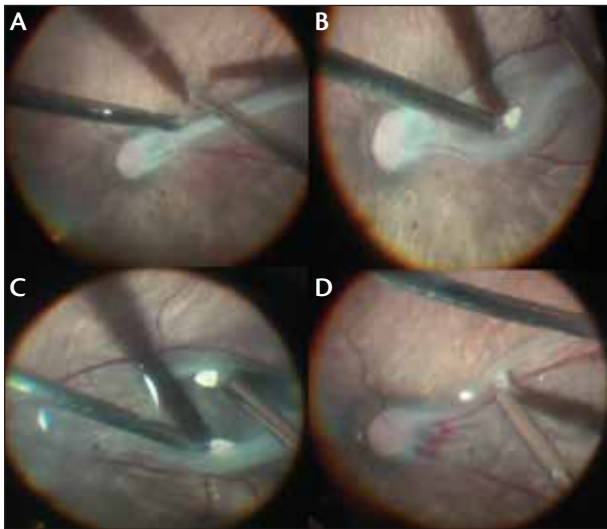


Figure 2. Bimanual dissection of the glistening condensed sheet encapsulating the macular fold (A,B). Cross-action redistribution of the macular tissue using two diamond-dusted scrapers (C). Displacing the recessed tissue peripherally after flattening of the fovea (D).

a glistening sheet coming off the retinal surface all the way to the periphery (Figure 2A and B). The dissection of the stalk off the fold enables me to manipulate and open the tissue in the fold. I then undermine the edges of the fold and redistribute the macular tissue over the macular retinal pigment epithelium (RPE) with a cross-action technique using two diamond dusted scrapers under perfluorocarbon liquid (Figure 2C). Care must be taken not to press or mechanically damage the underlying RPE while stretching the elastic retinal tissue.

Once the fold starts to open, I redistribute the fovea back in place and displace the recess tissue peripherally outside the arcades if possible (Figure 2D). That is, I flatten the central fold containing the fovea and form two small extramacular folds. While this maneuver is done, the surgeon may notice that a vascular pattern or bifurcation of retinal vessels appears out of the fold (Figure 3). The eye is left filled with air or sometimes with a bubble of nonexpandable SF₆, which helps to create shear vectors to stretch the macular area.

RESULTS

All 16 eyes in this series were followed for at least 1 year with clinical anatomic assessment and electrophysiologic assessment. Multifocal electroretinogram (mERG) was used to evaluate pre- and postoperative values for macular function, and nonverbal sweep visual evoked potentials (VEP) were used as a measurement of

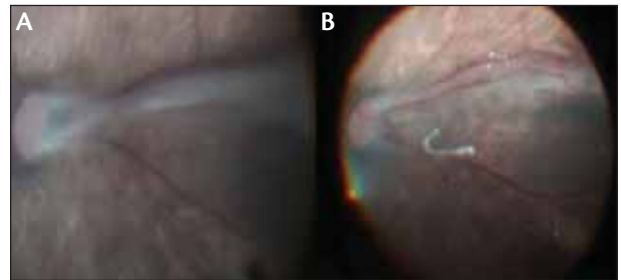


Figure 3. Early in surgery (A) and during dissection (B). Notice the appearance of a bifurcation of the upper temporal retinal vessels that were buried in the fold.

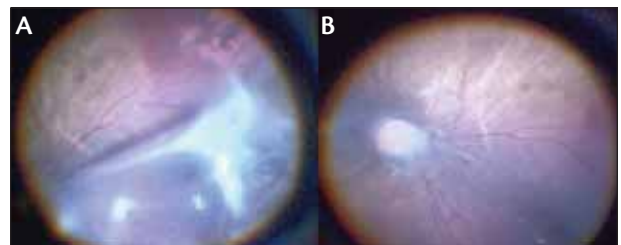


Figure 4. Preoperative color fundus photo of an infant with macular fold (A). At 1 year after surgery the macular area is flattened and the foveal anatomy is restored.

potential visual gain during follow-up.

Macular folds were flattened completely in 11 (68%) infants (Figure 4). Four (25%) infants had good central flattening with residual equatorial dragging, and one (6%) infant showed only partial improvement in the height and thickness of the fold, with incomplete central flattening of the fold.

Postoperative mERG of the macula showed improvement in pattern values compared with preoperative macula images in eight infants at 3 months. One more infant showed improvement at month 4. Subgroup analysis showed that the infants who improved were operated on before 3 months of age. Four infants showed no significant change up to 1 year after surgery, and sweep VEP showed improvement in visual function in 10 (62.5%) infants over the follow-up period.

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Figure 5. Composite picture of a child's fundus postoperatively. At 6 months, changes in RPE are observed in the macula (A). At 4 years, RPE is undergoing hypertrophic change (B) and fluorescein angiography shows good RPE under the fovea (C).

Hypertrophic RPE changes can occur postoperatively, but we have observed that they improve with time (Figure 5).

DISCUSSION

Macular folds involving the fovea can be a complication of regressed ROP, causing disruption of central fixation and the development of wandering eye movement early in life. In this case series, we attempted to relocate the fovea to see if this technique is useful and visually rewarding during these children's visual development.

From this series we know the following:

- foveal fixation is better than extrafoveal fixation;
- early intervention, before mechanical and ischemic changes happen to the photoreceptors included in the fold, is better than later;
- early intervention means a chance of restoring foveal fixation in the developing visual system; and
- it is possible to flatten folds.

What we do not yet know includes the following:

- whether anatomic attachment of the fovea results in visual benefit;
- the age limit in which there will be visual improvement if surgery is performed; and
- whether the chances are better for a child with bilateral folds vs unilateral folds for developing amblyopia.

Macular mERG demonstrated significant improvement in two out of three infants who were operated on at 5 months of age. These results are dependent on multiple factors including the availability of a good central nervous system for vision, minimum competition between eyes, the absence of subretinal fluid (tractional folds), and aggressive amblyopic therapy. Late-onset improvement in vision despite sensory deprivation has also been reported in cases of volcano-shaped retinal detachment.³ Improvement in vision was seen in infants with this form of detachment who underwent surgery at 5 months of age.

The improvement seen in sweep VEP, as a nonverbal estimate of visual function, suggests that there can be benefit from doing this type of surgery. Follow-up revealed central fixation development with fixation skills, as reported by parents and low-vision teachers. Subgroup age analysis of electrophysiologic results demonstrated that early intervention is better, despite improvements seen up to 5 months of age. Also, minimal manipulation over the foveal area when opening the folds helps to minimize damage to the RPE. Postoperative amblyopic treatment to regain a new fixation point is essential and important.

CONCLUSION

In infants with macular folds, macular unfolding is technically possible and can be considered as a surgical option for improving or restoring macular vision. Early intervention and aggressive amblyopic therapy are important for augmenting visual improvement. My hope is that this technique and others yet to be discovered can restore and save the precise macular vision these infants can lose over the course of the disease. ■

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