Dragged Fovea Diplopia Syndrome After Epiretinal Membrane Peel Surgery
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ABSTRACT
Background: Dragged fovea diplopia syndrome is a condition involving intractable binocular diplopia associated with macular pathology often involving epiretinal membrane and macular pucker. The prevalence of this condition is unknown. There is no known cure for this condition but effective treatment for the symptoms is available. Case Reports: Two cases of intractable central binocular diplopia of 2 to 3 years' duration that were not correctable with prism are presented. Both patients had a history of epiretinal membrane and had undergone membrane peel with vitrectomy. No extraocular muscle dysfunction or paresis was evident in either patient. Both responded favorably to partial monocular occlusion with semi-transparent tape. Conclusion: Dragged fovea diplopia can be extremely frustrating for both patients and doctors. An understanding of this condition will allow practitioners to detect it and treat the symptoms in order to allow patients to function normally.

INTRODUCTION
Binocular diplopia is most often due to a defect of the ocular motor system, involving the muscles, nerves or supranuclear pathways. Less commonly, binocular diplopia can be caused by retinal pathology at or near the macula. Some of the reported retinal pathologies known to cause diplopia are epiretinal membrane (ERM), choroidal neovascular membrane, and central serous retinopathy. In the case of ERM, diplopia can occur due to wrinkling of the macula or following surgical intervention such as vitrectomy and membrane peel.

Dragged fovea diplopia syndrome is a condition in which the fovea in one or both eyes is displaced causing a loss of correspondence between foveas and thus diplopia which is not correctable with prism. We present two cases of intractable diplopia following vitrectomy and epiretinal membrane peel along with their treatment. We also propose ocular coherence tomography as a means of visualizing the dragged fovea and suggest that further investigation into this technique is warranted.

CASE REPORTS
Case 1
EN is an 83-year-old white male. He presented to our clinic after seeing numerous doctors in the past 4 to 5 years. He claimed to be experiencing double vision that was worse in the distance and none of the treatments he had received involving prism provided any lasting relief. EN had been suffering from intractable binocular diplopia since his first epiretinal membrane peel surgery on the right eye 5 years earlier. He had since undergone the same surgery on the left eye as well but the diplopia persisted despite many variations of prism correction. He denied any dizziness, pain, or monocular diplopia but was having trouble judging distances. He was not taking any ocular medications or culpable systemic medications. He denied any allergies and had not endured injury to either eye. He had a history of cataract extraction with posterior chamber intraocular lens (PCIOl) implantation on both eyes. Family history was unremarkable but EN had a history of hypertension.

Distance visual acuity was 6/12-1 (20/40-1) in the right eye (OD) and 6/15+2 (20/50+2) in the left eye (OS) with habitual correction of pl-1.00 x 060 2 base down OD, and -0.75-1.00 x 09 4 3 base up OS. Pinhole acuities over habitual spectacles were 6/9-1 (20/30-1) OD and 6/9-2 (20/30-2) OS. Near acuity was 6/6-1 (20/20-1) OD and 6/9+1 (20/30+1) OS through a near add of +3.50 in both eyes (OU). Pupils were round and reactive without relative afferent pupillary defect, motilities were full and unrestricted OU, and confrontation fields were full to finger counting. Cover test with habitual correction revealed a 4 prism diopter (PD) exophoria and a 3 PD intermittent left hypertropia at distance and a 14 PD
exophoria at near. The vertical prism in his spectacles appeared to be too strong since the patient’s left hypertropia with correction converted to a left hypotropia when tested without correction. On Amsler grid, EN reported a missing area on the far nasal field of the right eye but no metamorphopsia or scotoma on the left eye’s field. Manifest refraction was +0.75-1.00 x 162° (20/30) OD and -0.25-1.25 x 098° (20/30) OS.

External appearance and adnexa were unremarkable. Biomicroscopy revealed corneal arcus OU, normal conjunctiva, deep and quiet anterior chambers with flat irides and stable PCIOls OU with clear or opened posterior capsule. Intraocular pressures were 13 mmHg OD and 8 mmHg OS by applanation. Both optic nerve heads exhibited deep cups with visible lamina and cup to disc ratios of 0.6 and 0.5 OD and OS respectively with healthy pink neuroretinal rim. An epiretinal membrane was present in each eye with a slightly darker area over the macula that resembled a pseudohole. Watzke-Allen sign was negative in both eyes. Otherwise the maculae were flat. Retinal periphery was flat without holes, tears, or retinal detachments OU.

Optical coherence tomography (Stratus OCT, Carl Zeiss Meditec, Inc.) was performed on both maculae. The scan for the right eye showed a normal flat macula, but the scan of the left macula appeared displaced with areas of thickening and thinning (Fig. 1). We also performed a modified version of the lights on-off test as described by De Pool et al by shining a target from a direct ophthalmoscope onto a black background and comparing fusion ability with the lights on and off. While wearing his habitual glasses EN was unable to fuse while looking at the target with the lights on but when the lights were turned off he reported that the targets would come closer together and he would almost see singly but then they would break further apart. We concluded that his habitual spectacles contained enough prism power to make fusion impossible.

We performed a few trials of differing prism strengths but none of the variations produced acceptable results. We tried monovision in spectacles with a +2.50 D addition over the left eye and the distance correction in the right eye but this was not comfortable for EN. Finally, we placed a piece of semitransparent plastic tape on the center of the left spectacle lens and EN reported single vision. He elected to keep the tape on his glasses lens as the form of treatment for his condition.

Case 2

JS is a 64-year-old white male. He first presented complaining of a blurred crescent shape obstructing his vision in the right eye for 6 or 7 months and that he would drive with his right eye closed. Best-corrected visual acuity was 6/12+2 (20/40+2) OD and 6/6 (20/20) OS. He had a history of a retinal bleed and a posterior vitreous detachment in the right eye. Amsler grid testing showed central metamorphopsia OD and normal OS. Wrinkling of the right macula was evident on ophthalmoscopy and he was diagnosed with an epiretinal membrane for which he underwent a vitrectomy and a membrane peel. Following the surgery he reported some residual blur as well as double vision with images separated vertically. Vertical diplopia was caused by a phoria which over time became an intermittent tropia. He received a prescription with vertical prism which would be altered numerous times over the next year without providing lasting relief from diplopia. His symptoms decreased for a short time after each change to the prism but would soon return. He reported that his peripheral vision appeared single but his central vision was double and still distorted, despite the surgery. One year after the vitrectomy he had a cataract removed from the right eye with implantation of a PCIOl. Following the surgery his best-corrected visual acuity was 6/7.5 (20/25) OD. He still noted metamorphopsia on Amsler grid testing OD and residual macular wrinkling could be observed on ophthalmoscopy. After a number of visits he was referred back to the surgeon who performed the membrane peel. The retinal surgeon assured JS that the surgery was a success, but she found that one of his spectacle lenses was decentered and tilted. She recommended that a new refraction be performed and his frames adjusted. Changes were again made to his glasses.

Fig. 1 Ocular coherence tomography scan of the left macula in Case 1 possibly showing foveal displacement.
including altered vertical prism without improvement. Finally, he was sent to a strabismus specialist who diagnosed him with dragged fovea diplopia and placed a piece of semitransparent tape on the center of his right lens to partially occlude the eye and eliminate his central diplopia. When he returned to our clinic in follow-up about 3 years after the initial presentation he was relieved to have single vision. We gave him a new prescription without prism and he continues to wear tape on his right lens. Both maculae appear normal without obvious wrinkling. OCT of the right eye showed a displaced fovea with normal thickness (Fig. 2). We performed the lights on-off test as described by De Pool et al.1 We presented an illuminated white 6/21 (20/70) optotype on a black background (Fig. 3) with the room lights on and JS reported central diplopia with single vision in the periphery. The room lights were then extinguished and a single white optotype was reported within 1 to 2 seconds.

DISCUSSION

Binocular diplopia in association with central retinal pathology has been reported in the past.4,5 The dragged fovea diplopia syndrome was coined by De Pool et al6 to describe a condition in which a person experiences central double vision that is not correctable with prism. Often when a patient with this condition is given prism in their glasses they will initially respond favorably but ultimately the diplopia will return. Sometimes the diplopia returns almost immediately, or it may take hours to days for symptoms to appear again.1 Usually a comitant, small-angle hyperdeviation is present without evidence of extraocular muscle dysfunction or paresis. The explanation for this intractable diplopia is a mechanical displacement of the fovea in one or both eyes disrupting the normal foveal correspondence and creating rivalry between central and peripheral fusional mechanisms.2,4,6 A small correcting movement of one or both eyes is made to allow central fusion, thus placing the peripheral retina of each eye in conflict. It is thought that the peripheral drive for fusion overrides the central drive because Panum’s fusional area is greater peripherally than centrally.1,8 Thus, central diplopia is manifest and since prism cannot exclusively influence the central retina, the diplopia remains despite prism treatment. Another possible contributing factor to diplopia, either solely or in conjunction with foveal displacement, is aniseikonia. Benegas et al9 proposed that separation or compression of the photoreceptors from macular disease may produce a difference in retinal image size significant enough to cause diplopia. This is another reason why prism does not relieve symptoms in these patients. We did not perform any tests to detect aniseikonia in our patients.

Some of the reported causes of binocular diplopia from macular pathology are: epiretinal membrane, choroidal neovascularization, central serous retinopathy, paramacular scars, and localized macular detachments.2,5 Both of our patients had a history of epiretinal membrane with subsequent vitrectomy and membrane peel. Diplopia did not ensue until after the surgery was performed. Silverberg et al10 also found this to be the case in 4 out of 7 patients they reviewed and attributed it to improved
vision in the operated eye which previously was reduced enough to preclude diplopia. The prevalence of this condition is not known but epiretinal membrane appears to be one of the more common causes. De Pool et al° reported it as the cause in more than half of the 87 patients they reviewed with dragged fovea diplopia.

**Pathophysiology of Epiretinal Membranes**

Epiretinal membranes (ERMs) are most commonly found in individuals over the age of 50 and are usually bilateral but often asymmetrical.°,10 ERMs are sometimes found in children, although this is rare. If an ERM is present in a child, it is commonly associated with trauma.11 Population-based studies of different demographics show an overall prevalence ranging from 2.2% to 18.5% and age-related prevalence of 2% in the 6th decade and up to 35% in the 8th decade.°,10,12-16 They can be classified clinically into an early, usually asymptomatic type called cellophane maculopathy caused purely by glial cell proliferation along the internal limiting membrane (ILM). There is also a more advanced type called preretinal macular fibrosis or macular pucker (Fig. 4A,B). The advanced type is usually more severe and is also caused by glial cells but with a fibrous component that causes traction and wrinkling of the retina.°,15 These membranes can also be classified as either primary or secondary depending on their underlying cause. Primary epiretinal membranes occur idiopathically or following posterior vitreous detachment (PVD). Secondary epiretinal membranes are associated with some kind of pathology or intraocular surgery; usually proliferative diabetic retinopathy, retinal tears or holes, macular holes, retinal detachment, or after cataract surgery.17 The histocytology of epiretinal membranes differs depending on the underlying pathology. Simple primary epiretinal membranes are formed when glial cells migrate through breaks in the ILM of the retina and begin to proliferate along the retinal surface. Snead et al° classified ERMs into three types: simple laminocyte ERMs, tissue repair ERMs, and neovascular ERMs. Simple ERMs form after PVD or idiopathically. Snead et al° described the glial cells in a simple ERM as laminocytes because they form a monolayer on the ILM as they proliferate and migrate along the vitreoretinal junction. The ILM in these membranes may become hyperconvoluted as the ERM contracts causing distortion of the retina and subsequent metamorphopsia and reduced visual acuity. Tissue repair ERMs develop after retinal tear, trauma, infection, or blunt injury. This membrane differs slightly from the simple type in that it also contains retinal pigment epithelial (RPE) cells, fibroblasts, and macrophages.°,18 RPE cells may be liberated when a retinal tear or break occurs, forming the characteristic tobacco dust in the anterior vitreous. Over time these cells settle onto the retina to cause a pigmented epiretinal membrane.19 Neovascular ERMs develop as a consequence of proliferative diabetic retinopathy, radiotherapy, or vasoformative tumors. This type of membrane is very different because it contains capillaries and acellular stromal tissue.° Kampik et al° found that ERMs also contained collagen and myofibroblasts and concluded that these features may account for the contractile properties of the membranes. Epiretinal membrane can be treated by surgical removal. Vitrectomy and membrane peel can achieve an increase in visual acuity of 2 Snellen lines or more in up to 90% of patients°,22 and can significantly reduce metamorphopsia. This reduction in

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Fig. 4 (A) Epiretinal membrane with macular pucker. (B) Note the radial striations from contraction of the membrane.
metamorphopsia may be an important benefit even if visual acuity is not noticeably improved.21

**Diagnosis and Treatment**

ERMs can be seen as a glistening sheen on the surface of the retina different from that seen in a healthy young fundus. This sheen appears less regular, like sunlight reflecting off disturbed water. The membrane may be clear or may have a greenish-grey appearance depending on the etiology. If macular pucker is present, radiating folds from one or more retinal contraction foci may be visible (Fig. 4A,B).24 After a membrane peel is performed, some residual retinal wrinkling may be evident and/or an apparent pseudohole may be seen. Alternatively, the retina may appear normal with no evidence of macular pathology on funduscopic examination. Case 1 had a visible remnant of epiretinal membrane and a pseudohole over the macula in each eye with a negative Watzke-Allen sign. In Case 2 some residual wrinkling was apparent in the operated eye.

It is not possible to visualize the displacement of the fovea on funduscopy so this condition must be confirmed in other ways. The lights on-off test described by De Pool et al1 is a simple test that we have found very useful. A white illuminated optotype of about 6/21 (20/70) Snellen equivalent is presented on a black background (Fig. 3). In a positive result the patient reports central diplopia with the lights on but when the lights are extinguished and all peripheral cues to fusion are eliminated a single object is reported. Both of our patients responded positively to this test. When we attempted this test in Case 1, we did not have the setup exactly as described but we arranged a modified version with an illuminated target from an ophthalmoscope on a black background. Case 1 reported that the objects came closer together and almost became single but then broke further apart. We attribute this to his glasses which contained vertical prism of sufficient quantity to prevent fusion. In Case 2 we had an LCD screen with a black background and a white optotype as described which produced a definitive positive result. The synoptophore has been used in the past to confirm the disparity between central and peripheral fusion, but the lights on-off test seems to be a valid substitute.2 With the increasing availability of computerized visual acuity charts, the lights on-off test may be more readily available than the synoptophore.

We performed OCT (Stratus OCT, Carl Zeiss Meditec, Inc.) of the macula on both patients with interesting results. The image showed the foveal area displaced from the center of the reference lines (Figs. 1, 2). This was instructive because such a displacement is not generally seen when performing macular scans. We wondered if the image produced by the OCT was actually showing the displaced fovea. We recognize that there are inherent flaws in this assumption since fixation cannot be accurately monitored while performing the scans. Gupta et al2 were able to visualize the foci of contraction on the retina with macular pucker using OCT with scanning laser ophthalmoscopy (SLO). This instrument allowed coronal plane images from the OCT to be superimposed on the SLO image with point to point registration. This combined image revealed multiple centers of retinal contraction with retinal folds and areas of thickening easily referenced with the anatomical location on the retina. One wonders if it would be possible to measure the displacement of the fovea with this method in dragged fovea diplopia syndrome.

There is no known cure for dragged fovea diplopia, only palliative treatment. There is no way to align the foveas without causing peripheral diplopia or vice versa. Others have reported on the success of placing Bangerter foils of different densities over the affected eye in order to degrade the image for that eye and eliminate diplopia.1,3 The foil is cut down so it only affects the central vision and can be placed on the inside center of the spectacle lens. This allows the patient to fixate with the other eye and use peripheral retina to maintain fusion. Patients tend to find this acceptable. Another option described by De Pool et al1 is Scotch Satin tape (3M Co., St. Paul, MN). They found that this was more cosmetically appealing than the more opaque Scotch Magic tape (3M Co., St. Paul, MN) and less expensive than Bangerter foil. Both our patients responded very favorably to tape placed on the center of one lens. The patient in Case 2 has been wearing this tape for almost 2 years. We also attempted monovision in the patient in Case 1; however, he found this uncomfortable and was not able to tolerate it.

**Conclusion**

Dragged fovea diplopia syndrome may be more common than we currently realize. It can be very frustrating for both patients and doctors to encounter this condition if it is not understood. There is a simple subjective way to test for it using the lights on-off test. As technology advances perhaps an objective means of detecting this condition will become available. We suggest that further testing should be done with OCT to determine if the actual foveal displacement can be objectively quantified. This condition cannot be cured but the symptoms can be treated very inexpensively and with relatively good success by partially occluding one eye with semi-transparent tape or Bangerter foil.

**References**


The patient in Case 1 had an absence of the following clinical signs and symptoms, **except**:
- Ocular pain
- Difficulty judging distances
- Dizziness
- Sudden loss of visual acuity

The patient in Case 2 presented with which of the following signs or symptoms?
- Decreased left-sided visual field
- A floater
- Blurred shape obstructing his vision in the right eye
- Reduced vision at night

All of the following statements about dragged fovea diplopia syndrome are true, **except**:
- Diplopia may disappear and appear again
- It is not correctable by prism
- OCT is effective in determining if the foveal displacement can be objectively quantified
- Prism is usually effective in curing the condition
4. Which of the following best describes an individual with epiretinal membranes (ERMs)?
- 55-year-old female
- 60-year-old Caucasian male
- 65-year-old African-American male
- 76-year-old Asian female

5. What is the prevalence of ERMs according to population-basic studies?
- 1.2% to 4.5%
- 2.2% to 18.5%
- 5.3% to 9.4%
- 3.6% to 20.5%

6. Secondary epiretinal membranes are associated with all of the following, **EXCEPT**:
- Cystoid macular degeneration
- Macular holes
- Retinal tears
- Retinal detachment

7. Surgery is effective in achieving an increase of visual acuity of 2 Snellen lines in what percentage of patients?
- 60%
- 75%
- 90%
- 95%

8. Which of the following is the most likely cause of dragged fovea diplopia syndrome?
- Cataract extraction
- Vitreous detachment
- Membrane peel
- Family history of the condition

9. In what number of patients did diplopia occur after vitrectomy and membrane peel?
- 2 out of 3
- 4 out of 7
- 6 out of 9
- 8 out of 10

10. Which of the following treatments was effective in the two Case Reports presented in the paper?
- Partial monocular occlusion with semitransparent tape
- Care and regular patient monitoring
- Prism correction
- Monovision in spectacles