detected early in life, the tractional forces of the epiretinal membrane were associated with severe retinal microarchitecture compromise. Future studies are needed to evaluate the effect of earlier detection and de- lamination of the epiretinal membrane in prevention of permanent retinal disorganization and visual loss. Optical coherence tomography can provide important information regarding the vitreoretinal interface of this tumor and could influence surgical decisions. 

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Dynamic Atypical Optic Nerve Coloboma Associated With Transient Macular Detachment

Though infrequently encountered by most ophthalmologists, optic nerve pits and optic nerve colobomas (typical and atypical) are well known cavi- tary optic disc anomalies.1,2 Maculopathy characterized by macular schisis, outer layer detachments, and occasionally outer layer holes are fre- quently associated with cavitary op- tic disc anomalies.3,4 The etiology of the maculopathy and source of the subretinal fluid are unknown but both systemic and intraocular fac- tors have been proposed.5,6

Fluctuations, and even sponta- neous resolution, of the described maculopathy can occur but are un- common.1,3,10 Though other cavi- tary optic disc anomalies have rarely been reported to fluctuate in appearance,11-13 we are unaware of any previous reports of fluctuating optic pits or atypical colobomas. We present 3 unique cases in which the fluctu- ating appearance of an atypical op- tic nerve coloboma is associated with spontaneous resolution of the associ- ated maculopathy. 

Report of Cases. Case 1. A 40-year- old white woman sought care because of a 1-week history of de- creased vision in her right eye. The family history was negative for colobo- mas or optic nerve disease but 1 brother had a history of cataract and retinal detachment. Best-corrected acuity was measured at 20/200 in both eyes. The visual acuity of the left eye had slowly deteriorated 4 years prior due to progression of low tension glaucoma. At initial exami- nation, she was using Timoptic-XE (Merck & Co Inc, Whitehouse Sta- tion, NJ) and Azopt (Alcon Laborato- ries Inc, Fort Worth, Tex) in both eyes. Previous neuroimaging re- sults with computed tomography and magnetic resonance imaging were both normal. When first ex- amined, the optic nerve of the right eye was normal in size and color and contained a small slit-like cup (Figure 1A). No optic pit or peri- papillary pigmented changes were seen. A large macular schisis with a large outer layer detachment was noted. The fovea appeared cystic and no outer layer hole could be iden- tified. A posterior vitreous detach- ment could not be identified. Late frames of the fluorescein angio- gram showed mild disc hyperfluores- cence. The left disc was of normal size without peripapillary pigmentary changes and contained a large, uniformly deep cup (cup-disc ratio of 0.95). The macula ap- peared normal. We could identify the base of the cup and no distinct excavation was seen. Observation was recommended. Over the next 6 months, the acuity of the right eye improved to 20/40. The schisis and outer layer detachment resolved, but macular retinal pigment epithelial changes corresponding to the area of the schisis were still present. A dis- tinct optic nerve cup containing a central excavation became visible (Figure 1B). Funduscopic examina- tion findings of the left eye were un- changed throughout the course of observation. Optical coherence to- mography (OCT) was not avail- able. The optic nerve and macular appearance could not be altered by changes in body position or by light stimulation of the eye.

Case 2. A 30-year-old white man was seen for a 1-week history of blurred vision in the left eye. Fam- ily history was negative for colobo- mas or optic nerve disease. Cor- rected acuity measured 20/20 OD and 20/160 OS. The patient had previ- ously undergone bilateral my- opic photorefractive keratectomy and had a postkeratectomy uncor- rected acuity of 20/20 in both eyes. The optic nerve of the right eye ap- peared normal with a cup-disc ra- tio of 0.3. The floor of the cup was clearly visible and the macula ap- peared normal. The left optic nerve was normal in size without peripapil- lary pigment changes. The nerve was pink with slightly blurred mar- gins and contained a thin, centrally located, slit-like cup without a visible excavation (Figure 1C). A large macular schisis cavity with a smaller,
Figure 1. Fundus photographs. A, Case 1. Right eye showing normal-sized nerve with a small slit-like cup and associated macular schisis with outer layer detachment. B, Case 1. Right eye 6 months later showing an optic nerve cavitation while the macular schisis and outer layer detachment have resolved. C, Case 2. Left eye shows normal-sized nerve with a small slit-like cup and associated macular schisis with outer layer detachment. D, Case 2. Left eye 8 months later showing an optic nerve cavitation along with a much smaller schisis and outer layer detachment. E, Case 3. Left eye showing initial appearance of atypical optic nerve coloboma. Macular pigmentedary changes without macular schisis or detachment are present. Brightness and contrast of image have been modified. F, Case 3. Left eye 5 months after obtaining photograph in Figure 1E, showing normal-sized nerve with slit-like cup and associated macular schisis and outer layer detachment. Brightness and contrast of image have been modified. G, Case 3. Left eye 5 months later showing the optic nerve with an obvious temporal excavation and a diminished macular detachment. Retinal pigment epithelial changes are noted in the macula. Brightness and contrast of image have been modified.
centrally located outer layer detachment was observed. A posterior vitreous detachment could not be identified. Late frames of the fluorescein angiogram revealed increasing peri-papillary hyperfluorescence. Observation was recommended and 8 months later, the uncorrected acuity improved to 20/20. The schisis cavity and outer layer detachment were significantly diminished, but the optic nerve developed a temporal excavation (Figure 1D). Late frame disc hyperfluorescence was no longer present by angiography. Funduscopy examination findings of the right eye were unchanged throughout the course of observation. The optic nerve and macular appearance could not be altered by changes in body position or by light stimulation of the eye. Optical coherence tomography was unavailable.

Case 3. A 19-year-old white woman sought care for blurred vision in the right eye for 6 months. Best-corrected acuity was 20/60 OD and 20/25 OS. Macular pigmentary abnormalities suggestive of previous macular schisis and optic nerve excavations suggested of either large temporal optic pits or atypical colobomas were noted in both eyes (Figure 1E). Both optic nerves were normal in size and did not have any peripapillary pigmentary changes. We could not identify a macular schisis or an outer layer detachment in either eye. Five months later, this patient had an urgent examination for an acute drop in vision to 20/60 OS. We observed a pronounced change in the funduscopy appearance of the left eye. The optic nerve now had a slit-like cup and the macula contained a large schisis cavity with a centrally located outer layer detachment (Figure 1F). We also noted a macular schisis without an associated outer layer detachment in the right eye. We could not detect a change in the right optic disc or find an outer layer hole or a posterior vitreous detachment in either eye. Both eyes had late disc hyperfluorescence on angiography. We recommended observation and over the next 5 months, the acuity of the left eye improved to 20/40 while the right eye remained at 20/60. The macular schisis began to flatten bilaterally, but foveal retinal pigment epithelial changes were still noted bilaterally. A dramatic change in the appearance of the left optic nerve was seen, which was observed as early as 1 month after initial observation of the slit-like cup. The temporal optic nerve excavation originally noted had reappeared (Figure 1G). The right optic nerve was unchanged. We uncovered no systemic associations. The patient remained normotensive, and the optic nerve and macular appearance could not be altered with changes in body position or by light stimulation. Continued observation was recommended. Optical coherence tomography was unavailable.

Comment. Optic nerve pits and typical colobomas are probably embryologically distinct congenital excavations of the optic nerve.1,4 While typical colobomas are located inferonasally, pits are usually small, well demarcated, temporally located depressions in the nerve substance. Pits are sometimes even categorized as a subset of atypical optic nerve colobomas.5 The optic disc anomalies in our patients are similar to those reported by Johnson and Johnson,6 as well as observed in an autosomal dominant pedigree of atypical optic nerve colobomas and pits that were associated with macular detachments.1,4 Our patients lacked a family history of optic nerve disease.

In this series, we report the development of macular schisis in 4 eyes of 3 patients. Unlike the macular schisis typically associated with caviary optic nerve anomalies, in at least 1 eye of each of these 3 patients, an optic nerve excavation was definitely not seen at the onset of the macular schisis. As the macular schisis resolved in our 3 cases, simultaneous, ipsilateral optic nerve changes occurred. In 2 of the cases, only then did it become apparent that these eyes contained optic nerve excavations. In the third case, the patient was examined prior to the development of the macular schisis at which time a cavitory anomaly was observed. In this case, we documented the disappearance of the excavation along with the coinciding development of macular schisis. Subsequently, as the excavation reappeared, the macular schisis resolved. The third case is also unusual in that bilateral macular changes occurred simultaneously. In all cases, the optic nerve and macular changes occurred over several months, and in no case could the changes be reproduced by external stimulation.

Though earlier reports described the association of macular schisis and optic pits,1,5 Lincoff et al5 were the first to detail the macular changes in this condition. Since the publication of the seminal article by Lincoff et al, reports describing the OCT findings of optic pit maculopathy have confirmed their clinical observations.16,17 The macular changes in our patients were identical to those described in these reports. All of our patients had evidence of a macular schisis and in 3 of the 4 eyes, an outer layer detachment was identified. In no case was there a posterior vitreous detachment. The optic nerve and macular appearance were very similar to those of our 3 patients at initial examination. This eye required multiple surgical treatments including vitrectomy, intraocular gas tamponade, and peripapillary laser to achieve macular reattachment. A disc photograph taken after successful macular reattachment showed pronounced optic nerve cavitation like those seen in our patients following spontaneous resolution of the macular detachment.

Spaide et al19 have even reported the clinical and OCT findings in a patient with macular schisis without a cavitory disc anomaly. In contrast to our 3 patients, Spaide et al did not observe any optic nerve changes in their patient in 4 years of follow-up.15 Additionally, Walsh and Hoyt20 described a 14-year-old white girl with central serous reti-
nopathy in which an optic pit was not seen during the first year of observation but later became apparent. Over a 9-year period, the authors did not document a fluctuation in the optic nerve appearance despite noting spontaneous clearing of the central serous retinopathy on 2 occasions. In our cases, the optic nerve and macular changes were documented over a 5- to 8-month period. Since patient 3 was under our care prior to the development of the optic nerve and macular changes, we were able to observe changes in the optic nerve that may not have been otherwise suspected. Despite the noted differences, it is possible that the cases reported by Johnson and Johnson, Spaid et al, and Walsh and Hoyt represent a variant of the condition we are herein reporting.

Moore and associates described a patient with autosomal dominant atypical optic nerve coloboma, unassociated with serous macular detachment, who developed progressively worsening optic disc excavation attributed to low-tension glaucoma over a period of 13 years. Our cases appear to represent a more acute process owing to the short time period over which disc excavation occurred (1-8 months), initial disc hyperfluorescence on angiography, and reversal of associated maculopathy. Case 3 also showed the reappearance of a disc excavation, suggesting the excavation was a baseline abnormality and not secondary to glaucoma progression.

The fluctuating appearance of peripapillary staphylomas, optic nerve colobomas, and morning glory discs have been reported previously. In all reported cases, these changes typically occur over a several-minute period instead of a several-month period as we report. In addition, to our knowledge, concurrent macular changes have not been observed previously except for in association with the morning-glory disc anomaly. Our cases lacked features typical of morning-glory disc anomalies, peripapillary staphylomas, or typical optic disc colobomas. Given the normal size and appearance of the nerve and associated vessels and peripapillary retina, our cases most likely represent atypical optic nerve colobomas.

The spontaneous resolution of the macular schisis in our cases is not unique; this phenomenon has been reported infrequently. In fact, many authors favor treatment of macular schisis associated with optic disc pits and other disc anomalies because of the poor natural history. However, the unusual manifestations in our patients and the variable response to treatment reported in the literature lead us to recommend observation in these cases. It is uncertain whether our patients would have had better visual outcomes or quicker visual recovery had a surgical procedure been performed.

Both the source of fluid and the underlying mechanism responsible for the development of optic pit maculopathy remain elusive. Though other authors have postulated that forces along the vitreoretinal interface are responsible for optic pit maculopathy, we were unable to convince ourselves that these forces were responsible for the macular and optic disc changes seen in our patients. Additionally, the development of bilateral simultaneous macular schisis in our third patient suggests at least a systemic influence despite our inability to determine one.

Cavitary optic nerve anomalies are herniations of dysplastic retina through a defect in the lamina cribrosa, in which tissue sometimes extends posteriorly into the subarachnoid space. Even though histological evidence for a direct communication between the pit and subarachnoid space is lacking, experimental and some clinical evidence suggests it is conceivable. Alternatively, there is compelling clinical evidence that in some cases a direct connection exists between the vitreous and subretinal space via a defect in the excavation. Recently, Johnson and Johnson have proposed that the excavation could act as a bulb syringe and subsequent fluctuations in the intracranial pressure could lead to the development of macular schisis. We believe the hypothesis proposed by Johnson and Johnson can explain the optic nerve and macular changes noted in our 3 patients. Cavitary anomalies likely contain dysplastic tissue that is multilaminar and in some instances porous, which could provide interconnections between the vitreous, the subretinal space, and possibly the subarachnoid space (Figure 2A). As the formed vitreous liquefies with age, some fluid could migrate through a tissue defect possibly as a result of intracranial pressure fluctuations. At a later time, a sudden or pronounced increase in intracranial pressure could force the liquid vitreous anteriorly from a sac in the cavitation into the retinal tissue thus creating a schisis.
If some of this fluid became trapped under redundant dysplastic tissue normally lining the anomalous wall, anterior displacement of this tissue could change the appearance of the optic nerve (Figure 2B) as we observed in our 3 patients. In a similar manner, following vitrectomy and fluid-gas exchange in the aforementioned 24-year-old woman, Johnson and Johnson photographically documented the presence of a gas bubble trapped under tissue overlying the optic disc cavitation, which altered the optic nerve appearance. It is interesting that in our third patient, optic nerve changes were only seen in 1 eye. We suspect the constellation of histological features necessary for our observations is rarely present, perhaps further explaining why these findings have not been reported previously. Diagnostic technology like OCT might be helpful in confirming or refuting this hypothesis.

In summary, we present 3 patients with macular schisis and concomitant optic nerve changes. Though unproven, these changes could be the result of fluctuations in the pressure gradient between the subarachnoid space and the vitreous space. In particular, the simultaneous development of bilateral macular detachments in our third patient lends further evidence that systemic factors may play a role in the development of macular changes in patients with cavitary optic nerve anomalies. Clinicians should be aware that in cases of macular schisis in which an optic nerve excavation is not initially apparent, spontaneous resolution of the macular schisis is possible. Concurrent optic nerve changes should be sought and in some cases will reveal a cavitary anomaly. In the future, advances in imaging technology may allow further understanding of the pathophysiology in this unusual condition.

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11. Kral K, Svarc D. Contractile peripapillary chorioretinitis with circular “target-like” lesions scattered in the midperiphery and often arranged in a radial linear pattern has been a consistent feature. Other intraocular findings include mild iridocyclitis, vitritis, occlusive retinal vasculitis, and optic disc edema. Although patients may suffer a significant decline in vision initially, visual acuity tends to improve with medical therapy. Though iridocyclitis and vitritis are not unusual, and in some cases will reveal a cavitary anomaly. In the future, advances in imaging technology may allow further understanding of the pathophysiology in this unusual condition.

**Extensive Chorioretinitis and Severe Vision Loss Associated With West Nile Virus Meningoencephalitis**

West Nile virus is an arthropod-borne flavivirus that was first documented to affect humans in the United States in August 1999. Since that time, the infection has spread across North America, with 9862 human cases reported in 2003 alone. The majority of patients with these infections are asymptomatic, but approximately 20% develop an acute febrile illness accompanied by malaise, headache, myalgia, arthralgia, skin rash, and gastrointestinal symptoms lasting less than 1 week. Advanced age and the presence of diabetes mellitus have been identified as risk factors for severe neuroinvasive disease, including meningeal and encephalitis.

The common intraocular manifestations of West Nile virus infection have been described in several recent articles. Bilateral, multifocal chorioretinitis with circular “target-like” lesions scattered in the midperiphery and often arranged in a radial linear pattern has been a consistent feature. Other intraocular findings include mild iridocyclitis, vitritis, occlusive retinal vasculitis, and optic disc edema. Although patients may suffer a significant decline in visual function initially, visual acuity tends to improve with medical therapy.