Optic pits are congenital excavations of the optic nerve head that may be associated with other abnormalities of the optic nerve and peripapillary retina. Optic pits occur in about one in 10,000 people, with no gender predilection, and are usually sporadic. Optic pits are usually incidental findings on fundus examination and remain asymptomatic unless complicated by macular lesions such as edema, schisis or serous detachment. A patient with macular involvement generally presents with visual acuity of worse than 20/70 in the affected eye, and 80 percent of these eyes loose visual acuity to 20/200 or worse. It has been suggested that these patients have a greater propensity to develop normal-tension glaucoma, although the arcuate visual field defects may be caused by the optic pit itself rather than by glaucomatous damage.

A 17 year old male patient presented with progressive defective vision of 6 months duration in his left eye. Ocular examination revealed a best corrected vision of 6/60 not improving further with glasses or pinhole. Ocular examination was within normal limits except for abnormal fundus findings. Fundus examination revealed the presence of congenital optic disc pit with a serous macular elevation in the left eye. (Fig 1)

**Laser photocoagulation was** used to produce several rows of laser burns between the area of the serous retinal detachment and the optic disc. The objective was to achieve a very light white laser burn with little collateral damage to the nerve fiber layer. This presumably creates a wall of scar tissue to block the passage of fluid from the optic pit to the inner retinal schisis cavity and subretinal space.

**Vitreous surgery and internal tamponade:** Combinations of posterior vitrectomy, photocoagulation and gas tamponade was suggested for treating optic pit–associated maculopathy. Successful macular reattachment and improved central vision can be achieved using vitrectomy with induction of PVD and gas tamponade.

**Discussion**

Congenital pits of the optic nerve head vary in size, shape, depth and location. They appear as small, hypopigmented, grayish, oval or round excavated depressions in the optic nerve head. They are usually about 500 μm in size and may be bilateral in 10 to 15 percent of cases. Optic pits are most commonly located on the temporal side of the optic disc, but they may be situated centrally or anywhere along the margin of the optic disc. Optic pits along the rim of the optic disc are most likely to lead to serous detachments of the retina, with associated full-thickness or laminar retinal holes, retinal pigment epithelium mottling and general cystic changes. The retinal detachments are usually confined between the superior and inferior vascular arcades and are contiguous with the optic disc, sometimes through a visible isthmus of subretinal fluid. The elevated retina contains cystic cavities in the outer plexiform layer.

**Optical coherence tomography** - OCT of an optic pit usually shows a schisislike separation...
between the inner and outer retina and a larger retinal detachment.

**Visual field testing.** Optic pits may be associated with visual field changes, which can be due to one or both of the following mechanisms:

- An optic pit, especially if large, may displace nerve fibers to produce an arcuate scotoma or may lead to an enlarged blind spot.
- Associated serous macular detachment may manifest as metamorphopsia or blurred vision, and visual fields may demonstrate central scotoma. However, unlike degenerative or reticular retinoschisis, there is no absolute scotoma in optic pit maculopathy.

**Fluorescein angiography** Fluorescein angiography is usually unremarkable in cases of optic pit. There is no dye accumulation in the area of the serous detachment, although there may be late hyperfluorescence of the optic pit. It has been suggested that vitreopapillary traction in this area may cause leakage from optic disc blood vessels.

**Electrophysiological testing** An electroretinogram (ERG) may show poorly defined and low-amplitude waveforms, consistent with schisis and serous detachment. Preoperative evaluation of macular function is important for predicting the likelihood of central vision recovery after successful macular reattachment. Patients with a poor ERG response are less likely to experience visual acuity improvement even after anatomical reattachment.

**Differential Diagnosis**

A dilated biomicroscopic fundus examination is essential for differentiating optic pits from the following conditions:

- Optic disc anomalies such as choroidal and scleral crescent.
- Tilted disc syndrome.
- Circumpapillary staphyloma.
- Hypoplastic disc.
- Glaucomatous optic neuropathy. (Any change in the appearance of the optic pit over time suggests that the lesion may be an acquired notch of the neuroretinal rim secondary to glaucomatous damage.)
- Central serous retinopathy and subretinal neovascular membranes. (These conditions are alternative considerations for serous macular detachment).

**Pathophysiology**

Congenital optic pits result from an imperfect closure of the superior edge of the embryonic fissure. They are asymptomatic unless complicated by secondary macular changes. They typically lead to a two-layered maculopathy consisting of a primary inner retinal layer schisis and a secondary outer layer detachment. Although the exact mechanism by which optic pits cause macular detachment is not known, various theories about the source of fluid and the macular changes have been proposed, including:

**Subretinal fluid.** It has not been established conclusively whether the subretinal fluid originates from the vitreous cavity, from the subarachnoid space or from leakage from the retinal vessels around the optic disc. Studies involving intrathecal fluorescein injections and histological tissue analysis have failed to provide any evidence of the optic pit acting as a conduit between the subarachnoid and subretinal spaces. The lack of dye leakage from retinal vessels
makes it unlikely that the retinal vasculature is the source of the fluid. Brown and colleagues suggested that there may be a connection between the vitreous and the submacular fluid, based on the findings in their canine model of optic pit. Using India ink they found a direct communication between the vitreous, the optic pit and the subretinal space in three collie dogs with congenital optic pits.

**Two-layer separation.** Serous macular detachment associated with optic pit was thought to be due to direct communication between the optic pit and the subretinal space, facilitating fluid accumulation under the macula. However, Lincoff and colleagues suggested that the primary communication from the optic pit may be to the retina. Fluid may move into the retina, causing a schisislike separation of the inner and outer layers, with the neurosensory serous retinal detachment occurring secondary to this schisis. Recent OCT findings confirm this separation.

**Vitreous traction.** Vitreous traction appears to be an important factor in the pathogenesis of optic pit-related macular detachment. Traction, vitreomacular or vitreopapillary, may permit entry of fluid into the retina through the optic pit.

**Management**

Patients with asymptomatic optic pits need regular monitoring for the onset of any macular involvement. The management of optic pits with associated macular involvement is not well defined; various treatment modalities have been tried with variable success. Less-invasive treatments like laser photocoagulation should be tried initially, followed by a combination of vitrectomy, complete posterior vitreous detachment (PVD) induction and internal gas tamponade if symptoms persist.

When the optic pit is asymptomatic, the patient should be advised about the importance of regular comprehensive eye exams, including dilated retinal evaluations and threshold visual fields. Patients should be educated about the use of home visual acuity assessment and Amsler grid testing to monitor for the onset of maculopathy. They should be made aware of the signs and symptoms (e.g., blurred vision and metamorphopsia) of macular complications.

**Laser photocoagulation.** This is used to produce one or several rows of laser burns between the area of the serous retinal detachment and the optic disc. The objective is to achieve a very light white laser burn with little collateral damage to the nerve fiber layer. This presumably creates a wall of scar tissue to block the passage of fluid from the optic pit to the inner retinal schisis cavity and subretinal space (although the scarring may also involve peripapillary retinal tissue). While studies have reported successful resolution of the serous detachment in eyes that have been treated with photocoagulation, this does not always translate into improved final visual outcome.

**Macular buckling.** Macular buckling has been reported as a treatment option for serous detachment associated with optic pit. Scleral buckling converts the posterior hyaloid traction from an inward to an outward vector, promoting reattachment of the macula.

**Vitreal surgery and internal tamponade.** Combinations of posterior vitrectomy, photocoagulation and gas tamponade are used for treating optic pit–associated maculopathy. Successful macular reattachment and improved central vision can be achieved using vitrectomy with induction of PVD and gas tamponade. A complete PVD helps relieve vitreous traction. Indeed, spontaneous macular reattachment has been observed in eyes undergoing posterior vitreous separation. Gandorfer and Kampik advocate internal limiting membrane peeling in addition to removing the posterior vitreous for relieving all tractional components.

**Conclusion**

Maculopathy caused by optic pits has an overall poor prognosis, and long-term studies involving large groups of these patients are lacking. Given that the exact pathophysiology is still a matter of debate, management should be tailored to the visual disability and macular changes of the specific patient.

**References**