lomas just beneath the RPE. None of these intraretinal granulomas were situated near arterioles, capillaries, or venules. The choiopicapillaris itself did not show any sign of acute or chronic vasculitis. Furthermore, generalized granulomas were found in lung parenchyma, lymph nodes, heart, liver, and spleen. One could argue that the generalized granulomas with multinculated giant cells are in line with advanced sarcoidosis. In favor of this idea are 2 case reports of APMPPE with probable sarcoidosis and posterior choroiditis. The granulomas in our patient contained no characteristic asteroid bodies or Schaumann bodies, which can be seen in sarcoidosis. Furthermore, the occurrence of stroke in sarcoidosis is extremely rare and the fluorescein angiogram shows a different pattern.

The absence of any signs of previous or present vasculitis in the choiopicapillaris does not support the hypothesis that APMPPE is caused by a choroidal vasculitis of the lamina choiopicapillaris. Instead, our findings indicate that APMPPE is caused by choroidal granulomas and can be part of a generalized granulomatous disease. The granulomas resemble those seen in sarcoidosis. However, its clinical presentation and the occurrence of a cerebral granulomatous vasculitis of large and medium arteries instead suggests that it may be a distinct multisystem granulomatous disease.

Recognition of this syndrome is important and our case illustrates that it can be rapidly fatal. Because cerebral vasculitis associated with APMPPE usually responds well to corticosteroid therapy, we propose that patients with APMPPE complicated by central nervous system manifestations should be treated immediately with intravenous corticosteroids.

### Macular Hole in the Shaken Baby Syndrome

Retinal hemorrhages have been reported in 50% to 100% of infants diagnosed as having shaken baby syndrome (SBS). Retinoschisis and circular perimacular retinal folds are associated with poor prognosis in SBS. Although these ophthalmologic findings have been well documented in the literature, macular holes have not been described. We present 5 cases of children who developed macular holes as a sequela to SBS.

Five patients were diagnosed as having SBS based on systemic, intracranial, and ophthalmologic findings. The median age of trauma was 9 months (range, 6-10 months), and the median age of macular hole diagnosis was 10 months (range, 8-12 months) (Table 1). All macular holes were unilateral, despite severe bilateral retinal disease. Four patients had severe vitreous hemorrhage and intraretinal hemorrhage, and 1 patient had diffuse retinal hemorrhage affecting all retinal layers (Table 2). The median size of the macular hole was 700 μm (range, 500-1500 μm). Three macular holes were centrally located; 2 macular holes were ectopically located (juxtafoveal). The diagnosis of macular hole was made during initial funduscopic examination in 2 patients, during vitrectomy in 2 patients, and after clearing of the vitreous hemorrhage, initially obscuring the visual axis, in the final patient.

Surgical intervention was performed in 4 cases to clear the visual axis of vitreous and subhyaloidal hemorrhage. Surgery included vitrectomy, internal limiting membrane peel, and tamponade (no tamponade in patient 1, silicone oil in patient 2, perfluoropropane 12% tamponade in patient 3, and air in patient 4). The median age at the time of vitrectomy was 11.5 months. Three out of 4 eyes had successful hole closure following surgery (that in patient 1 remained open). The median follow-up period was 12 months (mean, 12.4 months; range, 6-24 months). In all 5 cases, blood was seen at the base of the hole, occasionally plugging outward.

### References


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Visual acuity outcomes were poor: 1 patient was able to track objects with the affected eye, 1 patient had aphakia and amblyopia, and 3 patients could not be assessed owing to severe intracranial damage. There were no mortalities.

Report of a Case. Case 1. A 7-month-old boy (patient 4; Table 1) was diagnosed as having SBS. A computed tomographic scan showed intracranial hemorrhage resulting in hemiplegia. Initial fundus examination at the time of injury showed massive preretinal, intraretinal, subretinal, and vitreous hemorrhage in both eyes. Medical instability precluded further ophthalmologic evaluation for 4 months. His hemorrhages did not resolve, and he underwent lensectomy combined with vitrectomy in the left eye. One month later a lensectomy and vitrectomy were performed in his right eye, during which an ectopic macular hole with fluid cuff was noted inferotemporal to the fovea. The posterior hyaloid had spontaneously separated. An epiretinal membrane was seen and peeled with partial removal of the internal limiting membrane. Laser photocoagulation was subsequently placed around the edge of the hole (Figure). The hole remained visible but closed postoperatively. Postoperatively the patient was aphakic, wore a contact lens, and underwent patching for amblyopia.

Case 2. Lens-sparing vitrectomy was performed in the left eye of an 8-month-old boy (Table 1; patient 1) with SBS. The macula was detached with organized blood; a thick plume of old blood spouted through the fovea via a 700-µm hole. A skirt of retinal pigment epithelial atrophy, almost extending to the arcades, surrounded the submacular blood. Postoperatively the hole was closed. Fluorescein angiography demonstrated disruption of the retinal pigment epithelium with numerous window defects.

Comment. The macular holes seen in our patients with SBS represent a previously unreported finding. In all 5 cases of patients with SBS with macular holes, blood emanated from the base of the holes. We believe that the underlying retinal hemorrhage caused mechanical pressure leading to full-thickness macular hole formation in a structurally weakened retina. The causes of a weakened retina include necrosis due to underlying hemorrhage (demonstrated in pathologic specimens in a rat model by Lincoff et al) and mechanical forces of shaking (a break in the internal limiting membrane over an area of retinal hemorrhage has been reported).

Attempts to preserve vision and prevent amblyopia by macular hole repair are important. However, successful macular hole repair did not lead to good visual outcomes. In one case (patient 1), no tamponade was used because the surgeon believed that functional vision was not salvageable; the hole remained open in

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Abbreviation: NA, not applicable.

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Figure. RetCam (Clarity Medical Systems, Pleasanton, Calif) photograph of the right fundus following pars plicata vitrectomy, lensectomy, membrane peel, barrier laser photocoagulation, and air tamponade. The retinal pigment epithelium surrounding the visible but closed macular hole is hyperpigmented in response to laser photocoagulation (arrowheads).
that eye, and visual acuity is poor. Limited visual outcome in these patients can be attributed to severe intracranial damage (Table 2), severe retinal hemorrhages (Table 2), and aphakia with amblyopia.

The presence of a macular hole in SBS is a poor prognostic finding. Affected patients usually have severe visual acuity deficits, even with successful anatomical repair. Additionally, macular hole may be a predictor of severe neurologic injury in SBS, as was the case in 3 of the 5 patients we described.

Judy I. Ou, MD
Darius M. Moshfeghi, MD
Khaled Tawansy, MD
Jonathan E. Sears, MD

Correspondence: Dr Moshfeghi, Department of Ophthalmology, Stanford University School of Medicine, 1225 Crane St, Menlo Park, CA 94025 (dariusm@stanford.edu).

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Transorbital Penetrating Brainstem Injuries

Penetrating injury to the posterior fossa is uncommon, and when it occurs, it is typically due to projectiles that pierce the cranial bones. Morbidity and mortality from these injuries are high. We present 3 cases in which objects that were thrust through the orbit penetrated the upper brainstem. All patients survived their injuries.

Report of Cases. Case 1. A 21-year-old man was struck below the left eye by a wire 4 mm in diameter while laying concrete-reinforcing steel mesh. He had a puncture site evident on his left lower lid. He complained of vertical and horizontal diplopia, mild left ptosis, decreased coordination of the right side of his body, and slurred speech.

Examination showed visual acuities of 20/15 OU. There was mild ptosis with limited adduction, abduction, and elevation in the left eye. His pupils were symmetric and reactive without a relative afferent pupillary defect. There was no evidence of globe injury. Goldmann perimetry testing results were normal. He had poor coordination of the right side of his body. There was no weakness. Speech was dysarthric without aphasia.

A computed tomographic scan showed minimal hemorrhage in the region of the left inferior rectus muscle. No bony fracture was present. Magnetic resonance imaging revealed a linear lesion in the inferior midbrain that began in the cerebral peduncle and was directed back toward the aqueduct (Figure 1). There was no retrobulbar hemorrhage.

A follow-up examination 2 years later revealed residual diplopia and a mild right hemiparesis.

Case 2. A 38-year-old man was stabbed in the upper left orbit and immediately became paralyzed on his right side. He remained conscious. He also complained of an inability to see out of his left eye secondary to complete ptosis. He reported local orbital pain, but had no other sensory symptoms.

Examination revealed acuities of 20/20 OD and 20/100 OS. Color vision and confrontation visual fields were normal. A small entry wound was discovered in the left medial upper lid and measured less than 1 cm in length. The right eye moved normally but the left was immobile and had a fixed, dilated pupil without a relative afferent pupillary defect. Dilated fundoscopic examination did not reveal any abnormalities. He had a right central seventh nerve palsy and a right hemiparesis with upgoing toe.

Magnetic resonance imaging of the brain revealed a linear lesion in the superior pons angling from the left ventrolateral side medially toward the fourth ventricle (Figure 2). The lesion was dark on gradient echo imaging and bright on diffusion-weighted imaging. A computed tomography angiogram showed no evidence of a vascular injury.

The hemiparesis gradually improved and the patient walked into the clinic for his follow-up visit 3 months later. At that time, he had visual acuities of 20/15 OD and 20/20 OS. Ocular motility revealed minimal adduction, elevation, and infraction of the left eye. He had moderate abduction and the eye was intorted. The pupil was fixed and dilated and a subtle right hemiparesis persisted.

Case 3. A 13-year-old boy was struck below his right eye by a beach umbrella tine ⅙ of an inch in diameter. His mother reported that he did not lose consciousness but was