Increase of Intraocular Pressure After Topical Administration of Prostaglandin Analogs

Several prostaglandins have been demonstrated to reduce intraocular pressure (IOP) in normal, hypertensive, and glaucomatous eyes.1-3 Two different prostaglandin analogs are commercially available: unoprostone (Rescula; Ciba Vision Ophthalmics, Duluth, Ga) and latanoprost (Xalatan; Pharmacia Inc, Columbus, Ohio). We observed an inverse reaction after topical administration of both analogs.

Report of a Case. A 29-year-old woman had retinitis pigmentosa with typical ophthalmoscopic findings, a ring scotoma, and a flat electroretinogram. Juvenile glaucoma was diagnosed at the age of 12 years. Because of the characteristic malformation of the anterior segment it was classified as Rieger syndrome. The initial IOP at the time of glaucoma detection was 50 mm Hg. Both eyes underwent Elliot operation. The left eye required an additional cryocoagulation of the ciliary body. After these operations, the IOP of the right eye was between 8 and 14 mm Hg without further medication. The IOP of the left eye was below 21 mm Hg until the patient was 26 years old. The IOP then began to increase, and a second cryocoagulation was performed. After the second cryocoagulation, the IOP varied between 0 mm Hg (without therapy) and 41 mm Hg OS (with maximum tolerated medical therapy without prostaglandin analogs). At this time visual acuity was 6/30 OD and 6/12 OS.

After a 9-week period of IOP values between 30 and 34 mm Hg OS, we decided to try an additional treatment of 2 drops of unoprostone, 1 in the morning and 1 in the evening. In less than 24 hours, the IOP increased to 56 mm Hg, accompanied by corneal edema. After withdrawal of treatment with unoprostone, the IOP returned to 15 mm Hg. During the following weeks the IOP again ranged between 1 and 35 mm Hg. Five months after this trial with unoprostone, another prostaglandin analog, latanoprost, became available. At this time, the IOP again was about 30 mm Hg despite maximum tolerated medical therapy without prostaglandin analogs. As with unoprostone, the IOP immediately increased to 55 mm Hg after 2 drops of latanoprost. This increase of IOP was again accompanied by corneal edema and a decrease in visual acuity. With intravenous 20% mannitol, the IOP rapidly dropped to 20 mm Hg and later returned to 30 mm Hg.

We now decided to perform a stepwise diode laser cyclophotocoagulation. After 4 treatments with 2 burns each, the IOP ranged between 10 mm Hg and 20 mm Hg OS. However, 5 months after the last laser treatment, IOP decreased to 0 mm Hg and remained at this hypotonous level for 3 weeks. Treatment with systemic and local steroids failed to increase IOP, and visual acuity was only 6/120. This was the reason why we now tried to elevate IOP using prostaglandin analogs. In fact, after 2 drops of unoprostone, IOP increased to 55 mm Hg within 36 hours and visual acuity increased to 6/20 (Figure).

There were no signs of acute anterior segment inflammation after the prostaglandin applications. A marked atrophy of the ciliary body was observed with high-resolution ultrasound biomicroscopy.

Comment. In the literature, we could not find any other reports of serious, reproducible IOP increase after unoprostone or latanoprost administration. These prostaglandin analogs are known to be safe and effective in reducing IOP.1-3 It is presumed that they facilitate the uveoscleral outflow, whereas trabecular outflow may be slightly impaired.4 One might speculate that, in our patient, uveoscleral outflow was considerably altered by the disease itself (Rieger syndrome and retinitis pigmentosa) or by the cryoprocedures. The atrophy of the ciliary body supports this theory. Because of these alterations, prostaglandins perhaps could not further improve uveoscleral outflow. Thus, the slight impairment of trabecular outflow could have caused the IOP increase.

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Transient Visual Loss and Decreased Ocular Blood Flow Velocities Following a Scleral Buckling Procedure

Scleral buckling procedures with encircling elements have been shown to decrease blood flow velocities in the central retinal artery but, in most cases, leave the ophthalmic artery unaffected. Although these hemodynamic changes are well documented with otherwise successful scleral buckling procedures, they are rarely symptomatic. We report the case of a young woman who developed episodes of posturally related transient visual loss following a scleral buckling procedure with an encircling element.

Report of a Case. A 26-year-old woman had undergone surgical repair of a 12-mm full-thickness corneoscleral laceration in the left eye 6 months previously. The laceration extended from the superior limbus to the inferior limbus. Two months after the ruptured globe repair, she developed an inferior macula-on retinal detachment, which was treated with pars plana vitrectomy and scleral buckling with a 42-style silicone encircling element (Labivrier, Oakville, Ontario). The intraocular pressure was normal until 4 weeks after the surgery, when it was measured at 44 mm Hg by the Tonopen tonometer (Mentor O&O, Norwell, Mass).

Treatment with topical 0.5% levobunolol and 0.2% bromonidine and oral nepotazene was initiated. The intraocular pressure stabilized in the low 20s and visual acuity remained counting fingers OS at 0.3 m due to a central corneal scar.

Approximately 3 months after the retinal detachment repair, the patient complained of a several-day history of episodic transient visual loss (to the level of bare light perception) in her left eye that occurred when she stood from a seated or supine position. She had recently resumed normal daily physical activities after being restricted in the postoperative period. The episodes were reproducible and she experienced up to 10 of these episodes daily, with each episode lasting 2 to 3 minutes.

Examination at that time revealed visual acuities of 20/20 OD and finger counting at 0.3 m OS. Goldmann applanation tonometry revealed an intraocular pressure of 23 mm Hg OS that was confirmed by the Tonopen tonometer. The anterior segment examination revealed a central corneal scar with a deep and quiet anterior chamber. The retina was completely attached deep and quiet anterior chamber. The retina was completely attached.

The retina was completely attached.

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also demonstrated the reversibility of the hemodynamic changes after removal of the encircling element or scleral buckle.\(^2,4\)

Although the decrease in retinal artery blood flow following scleral buckling procedures with an encircling element is well documented, the mechanism by which this occurs is unknown. Diddie and Ernest\(^5\) performed scleral buckling procedures on rabbits and found that encircling bands significantly reduced blood flow to the retina and choroid as measured by strontium 85–labeled microspheres.\(^6\) This effect was not seen in those eyes with segmental buckles or sham surgery. The authors speculated that the reduction of retinal and choroidal blood flow involved obstruction of choroidal venous drainage. Effects on the retinal circulation may be the result of direct or indirect increases in peripheral retinal vascular resistance from the buckle indentation.\(^1\)

In this case, a severe degree of blood flow velocity reduction was seen in the central retinal artery. In addition, the ophthalmic artery blood flow velocity was also reduced, a finding not encountered in the series of asymptomatic patients reported by Regillo et al.\(^1\) It is likely that the retinal, and possibly choroidal, perfusion was compromised to a much greater degree than what usually occurs following routine scleral buckle procedures with encircling elements. The markedly diminished ocular perfusion produced symptoms of transient visual loss precipitated by common postural changes in a young, healthy adult. Fortunately, the hemodynamic changes and symptoms were reversible, even months after the procedure.

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Retinal Pigment Mosaicism in Pallister-Killian Syndrome (Mosaic Tetrasomy 12p)

Pallister-Killian syndrome (PKS) was first reported by Pallister in 1977\(^2\) and later by Teschler-Nicola and Killian in 1981.\(^2\) This condition is caused by a mosaic tetrasomy of chromosome 12p, which is detectable in cultured fibroblastoid cells but generally not in peripheral lymphocytes. Systemic features in childhood are numerous and variable, and include most frequently “coarse” facial features, midface malformations, psychomotor delay, hypotonia, scalp hair sparsity, and variegated lightly and darkly pigmented skin.\(^1,5\) We describe a patient with the novel ocular manifestation of retinal pigmentary mosaicism, and confirm for the first time that the fibroblast cell line from the darkly pigmented skin contains the chromosomal tetrasomy.

Report of a Case. A 4½-month-old Hispanic boy was referred for evaluation of an irregular pupil. He was born at 35 weeks to nonconsanguineous parents; the pregnancy was complicated by maternal amphetamine use. The child was noted at birth to have bilateral inguinal hernias and dislocated hips, a cleft palate, a patent ductus arteriosus, and dysmorphic features. Whorled and streaky dark and light pigmentation was present on the skin of the trunk (Figure 1).

On examination, reasonable visual following (or fixation) behavior for age was demonstrated. There was hypertelorism and a low hairline. The right iris had an inferonasal coloboma; the anterior segments were otherwise normal. The fundus examination showed asymmetric patches and streaks of darkly and lightly pigmented retina (Figure 2), radiating outward from the optic discs in a petalloid fashion. There was concern about right optic nerve hypoplasia; the left optic nerve looked normal. The cycloplegic refraction was –6.25+1.00 × 90 OD and +4.25S OS. A subsequent visit showed a 40–prism diopter right esotropia.

The child was lost to ophthalmologic follow-up until age 2 years, by which time a diagnosis of PKS had been suggested although not confirmed by cytogenetic testing. An examination under anesthesia was performed, and punch biopsies of both lightly and darkly pigmented skin were obtained. Chromosome studies performed on fibroblasts cultured from the darkly pigmented skin revealed a mos 47,XY,+i(12)(q10)/46,XY karyotype in 27 of 41 metaphase spreads; 14 of 41 metaphase spreads showed a 46,XY karyotype. Fibroblasts cultured from the lightly pigmented skin biopsy specimen revealed a normal 46,XY karyotype in all 50 metaphase spreads. The karyotype derived from the dark (hyperpigmented) skin is consistent with PKS.
Comment. To our knowledge, we are the first to describe retinal pigment mosaicism in PKS. This finding is similar to the characteristic variegated skin color. If we assume that the retinal pigment mosaicism is caused by karyotype mosaicism, this suggests that the short arm of chromosome 12 contains a gene that affects retinal pigment epithelium (RPE) pigment production. We cannot explain why this striking fundus feature was not noted in previous case reports, except to postulate that the level of mosaicism in the RPE was sufficiently low (whether the karyotype was predominantly normal or abnormal) that sharp demarcations between affected and unaffected RPE could not be seen.

An extensive review of the literature revealed only one other diagnosis with similar cutaneous and fundus findings: hypomelanosis of Ito. This rare neurocutaneous disorder consists of skin pigmentary mosaicism, patchy retinal hypopigmentation, and seizures. Other ocular abnormalities overlap with those of PKS (Table), and include strabismus, hypertelorism, epicanthal folds, scleralization of the cornea, iris heterochromia, and microphthalmia. The karyotype in hypomelanosis of Ito is normal.

Our case suggests that infants with dysmorphic facial features, other congenital abnormalities, and variegated skin pigmentation would benefit from a fundus examination. The presence of patchy variability in RPE pigmentation may be consistent with PKS. In this case, standard karyotyping of peripheral blood lymphocytes is inadequate to rule out the diagnosis. Fibroblasts cultured from a biopsy of the abnormally hyperpigmented skin would reveal the mosaic tetrasomy 12p.

Recurrent Poststreptococcal Uveitis

Poststreptococcal syndrome (PSS) involves the development of systemic nonsuppurative inflammation after a streptococcal infection. The inflammation is sterile and thought to represent an autoimmune reaction between streptococcal-sensitized lymphocytes and host tissue because of “molecular mimicry.” Common manifestations of PSS include acute rheumatic fever, reactive arthritis, and acute glomerulonephritis. Recently, uveitis was described as a sign of PSS. This intraocular inflammation also develops after the bacterial infection. One report claims PSS uveitis can be recurrent. Herein, we confirm the findings of that single case and report that the length of time between episodes may be as long as 27 months.

Report of a Case. A 10-year-old white boy developed photophobia, ocular redness, and blurred vision bilaterally following an episode of streptococcal pharyngitis. Medical history was otherwise noncontributory. Examination noted vision of 20/80 OU with 3+ cells in the ante-
rior chambers. Workup revealed an elevated antistreptolysin O (ASO) titer (760 IU/mL) (normal range, 0-125 IU/mL) and an elevated erythrocyte sedimentation rate (27 mm/h) (normal range, 0-15 mm/h). Titers for antinuclear antibodies, rheumatoid factor, toxoplasmosis, Lyme disease, and angiotensin-converting enzyme were normal. Chest x-ray films, urinalysis, complete blood cell count, and chemistry panel were unremarkable. Symptoms resolved with cycloplegics and topical steroids. Twenty-seven months later he developed blurred vision, ocular tenderness, and photophobia bilaterally 10 days after streptococcal pharyngitis. Medications included a 10-day course of a combination product consisting of amoxicillin and clavulanate (500 mg twice daily). He denied other PSS symptoms. On examination, visual acuity was 20/50 OU. Pupils, motility, visual field, and tonometry were normal. Slitlamp examination revealed mild bulbar conjunctival hyperemia in both eyes. Both anterior chambers had 2+ cells and flare. Fine white keratic precipitates were present on the inferior cornea bilaterally. Irids demonstrated no nodules or atrophy. Anterior vitreous contained mild cells in both eyes. Fundus examination disclosed disc hyperemia with a few fine peripheral vitreous precipitates bilaterally, but no “snow-banking” or vascular sheathing. Blood testing revealed an elevated ASO titer (753 IU/mL), an elevated erythrocyte sedimentation rate (27 mm/h), and an HLA-DR2 haplotype. Complete ophthalmologic examination disclosed masses coating the cerebral convexities (Figure 1, A), the acoustic nerves, gasserian ganglia bilaterally, the right hypoglossal canal, the posterior tuberculum sella, and planum sphenoidale. These findings were considered diagnostic of sporadic NF2. A lumbar puncture revealed a normal opening pressure and the cerebrospinal fluid (CSF) formula was normal. Results of an ophthalmologic examination, including visual acuity, visual fields, and ophthalmoscopy, were entirely normal.

In late 1993, mild bilateral optic disc edema was noted without any new symptoms or abnormalities of visual function (Figure 2, A). Another MRI scan showed no changes. He was diagnosed as having papilledema related to impaired CSF outflow owing to obstruction by diffuse convexity meningiomas. Because he had normal visual function, no further diagnostic or therapeutic intervention was undertaken. Subsequent semiannual ophthalmologic examinations during the next 2 years showed no new findings.

After being lost to ophthalmologic follow-up for 2 years, he returned in September 1997 with a 2-week history of orthostatic transient visual obscurations. Visual acuity was still 20/20 OU, but automated visual fields showed generalized depression and mildly enlarged blind spots in both eyes, with an in-

Comment. We believe this patient had recurrent PSS uveitis. However, we are aware of only 1 other case of recurrent PSS uveitis. In our patient, both episodes of inflammation were preceded by a culture-positive streptococcal pharyngitis with elevated ASO titers, and demonstrated findings and a clinical course typical for PSS uveitis. The patient’s HLA-DR2 typing is interesting, as this haplotype has been seen in association with rheumatic fever and other PSS. Recurrent PSS uveitis is not surprising since other PSS entities may be recurrent, even with a subclinical infection.

Confirmation that PSS uveitis may be recurrent could be important for its diagnosis, prevention, and treatment. We agree with Leiba et al that all patients with ocular signs and symptoms of PSS and active streptococcal infection or a history of PSS should receive eye examinations. Examining blood ASO titers may be useful in patients with idiopathic uveitis and a history of a streptococcal infection.

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Visual Loss Secondary to Increased Intracranial Pressure in Neurofibromatosis Type 2

Neurofibromatosis type 2 (NF2), previously known as central neurofibromatosis, is defined clinically by the presence of bilateral acoustic nerve schwannomas and genetically by a mutation in the long arm of chromosome 22. Schwannomas of other cranial and spinal nerves are often present, as well as meningiomas. Patients typically have hearing loss in the second or third decade of life. About 50% of cases are inherited in an autosomal dominant fashion.

Among the many ophthalmic signs that have been described in NF2, papilledema has not been highlighted. We report 2 cases in which the dire consequences of sustained elevated intracranial pressures were not recognized in time to prevent visual loss.

Report of Cases. Case 1. A 31-year-old man reported a 2-year history of worsening bilateral hearing loss and dizziness in 1992. Bilateral sensorineural hearing was documented. Results of a neurological examination were otherwise normal. A magnetic resonance imaging (MRI) scan disclosed masses coating the cerebral convexities (Figure 1, A), the acoustic nerves, gasserian ganglia bilaterally, the right hypoglossal canal, the posterior tuberculum sella, and planum sphenoidale. These findings were considered diagnostic of sporadic NF2. A lumbar puncture revealed a normal opening pressure and the cerebrospinal fluid (CSF) formula was normal. Results of an ophthalmologic examination, including visual acuity, visual fields, and ophthalmoscopy, were entirely normal.

In late 1993, mild bilateral optic disc edema was noted without any new symptoms or abnormalities of visual function (Figure 2, A). Another MRI scan showed no changes. He was diagnosed as having papilledema related to impaired CSF outflow owing to obstruction by diffuse convexity meningiomas. Because he had normal visual function, no further diagnostic or therapeutic intervention was undertaken. Subsequent semiannual ophthalmologic examinations during the next 2 years showed no new findings.
inferior nerve fiber bundle defect in the left eye (Figure 2, B). The optic disc edema was more pronounced in each eye. An MRI scan showed interval growth of the cerebral dural convexity masses (Figure 1, B). There was no venous sinus thrombosis or ventriculomegaly.

He was treated with acetazolamide (2 g/d) and furosemide (40 mg/d) but his visual acuity, visual fields, and optic disc edema worsened.

Figure 1. Case 1. A, Magnetic resonance imaging scan in 1992 shows diffuse thickening of the convexity dura by meningioma. B, Magnetic resonance imaging scan in 1997 shows interval growth of convexity meningioma.

Figure 2. Case 1. A, In 1993, mild optic disc edema is present (left), but visual fields are normal (right). B, In September 1997, optic disc edema has become more pronounced (left), and early visual field loss has emerged (right). C, In December 1997, optic disc edema has worsened (left), and visual fields are markedly constricted (right).
ened during the next 2 months. By December 1997, visual acuity had fallen to 20/80 OD and 20/40 OS. Visual fields now showed marked constriction in both eyes. Optic disc edema was even more severe (Figure 2, C). A lumbar puncture revealed an opening pressure of 32 cm H2O with a normal CSF formula. In the month that followed, he underwent optic nerve sheath fenestration in the right eye, high-dose intravenous methylprednisolone treatment, and a lumboperitoneal shunt, all of which failed to halt the downward spiral of visual loss. By February 1998, visual acuity had declined to hand motions OU. The optic discs were pale and flat (Figure 3). A shunt tap confirmed normalization of the opening pressure to 17 cm H2O.

Case 2. A 26-year-old man with a remote diagnosis of sporadic NF2 began complaining in 1997 of progressive visual loss in the left eye during the previous year and the recent onset of transient visual obscurations in that eye. His right eye had earlier been blinded by a retinal detachment.

He was totally deaf owing to bilateral acoustic schwannomas and had earlier undergone partial resections of a foramen magnum meningioma, a right frontal meningioma, and a left acoustic schwannoma. The acoustic surgery had resulted in left facial and trigeminal neuropathy with resulting neurotrophic keratitis in the left eye. A right acoustic schwannoma had remained unoperated. A brainstem stimulator, placed in the medulla in 1989 to provide some hearing, precluded further use of MRI.

Ophthalmologic examination in 1997 disclosed visual acuities of no light perception OD and 20/50 OS. Pupils measured 5 mm in dim illumination; the right did not react to direct light and the left was sluggish. A right afferent pupillary defect was present. Because of his deafness, neck weakness, and quadriplegia, visual fields could be assessed only by confrontation, showing a small temporal island in the left eye. Neurotrophic keratitis in the left eye had caused partial opacification of the cornea with pannus. Ophthalmoscopy of the left eye faintly disclosed chronic disc edema with some atrophy. The right fundus revealed a chronic total retinal detachment with no view of the optic nerve.

A computed tomographic scan of the brain showed considerable enlargement of the ventricles relative to an MRI in 1988 (Figure 4, A and B). Because of the foramen magnum lesion, lumbar puncture was considered contraindicated for fear of provoking brainstem herniation. The patient’s progressive visual loss was attributed to papilledema associated with chronic obstructive hydrocephalus from the posterior fossa masses. In preparation for a CSF shunting procedure, the patient was given acetazolamide (2 g/d) to avoid the visual loss associated with surgically induced sudden intracranial hypotension. Several weeks later, the patient’s visual acuity had fallen to 20/200 OS.

The patient underwent ventriculoperitoneal shunting that re-
vealed an opening CSF pressure of 33 cm H2O. Four weeks after shunting, visual acuity had improved to 20/70 OS. Confrontation visual fields showed persistent nasal loss with a preserved temporal island. Fundus examination disclosed significantly less optic disc edema in the left eye with mild pallor. A postoperative computed tomographic scan revealed normalization of ventricular size (Figure 4, C).

Comment. Our cases highlight chronic papilledema as a cause of severe visual loss in NF2. This manifestation has been mentioned in previous reports, but without emphasis or explanation of mechanism. For example, Bouzas et al2 mentioned that 4 (7.4%) of 54 patients with NF2 had progressive visual field constriction from long-standing papilledema, one of whom had loss of central acuity in one eye to 20/160. Kaye et al3 and Landau and Yasargil5 separately made passing references to 2 patients with NF2 and papilledema secondary to intracranial tumors. Furthermore, the National Institutes of Health guidelines2 include an initial ophthalmic assessment for detecting the presence of juvenile cataracts and for the “photography of papilledema.” No explanation accompanies this recommendation.

In our first patient, increased intracranial pressure was likely caused by diffuse convexity meningiomas that interfered with CSF outflow across the arachnoid granulations. In this communicating hydrocephalus, the fact that there was no pressure gradient across the ventricular outflow routes explains why the ventricles were not enlarged. A similar mechanism has been described in a case of childhood meningiomas.3 Dural sinus drainage may also have been compromised by the meningiomas, but was not seen on MRI or documented by angiography. To the best of our knowledge, these mechanisms of increased intracranial pressure in NF2 have not been previously described.

In our second patient, papilledema was due to compression of the brainstem by tumor. This led to an obstruction of CSF outflow through the fourth ventricle. In this noncommunicating hydrocephalus, there is a pressure gradient between the ventricles and subarachnoid space, leading to ventriculomegaly. We speculate that this may be the more common cause of papilledema in NF2, referred to previously by other authors.3,4

The early recognition and treatment of papilledema in patients with NF2 can be difficult. As in our second patient, media opacification from coexistent neurotrophic keratitis can inhibit the detection of disc edema. Once recognized, it may be difficult to decide at what point the potential for developing chronic atrophic papilledema outweighs the risks of intervention. Whether early surgery in these patients is warranted is unknown. Regular ophthalmologic follow-up is essential in preventing visual morbidity. Visual loss can be particularly devastating to these patients, as they often have hearing impairment and other neurological disabilities.

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Negative Sinus Pressure and Normal Predisease Imaging in Silent Sinus Syndrome

We present the first case of silent sinus syndrome with both normal predisease imaging findings and documented negative maxillary sinus pressure, demonstrating unequivocally the acquired pressure and possible etiologic association with negative maxillary sinus pressure in at least some cases of silent sinus syndrome.

Report of a Case. A 27-year-old woman with painless, progressive sinking of her right eye over a 3-month period demonstrated 8 mm of enophthalmos and 4 mm of hypoglobus. Computed tomography showed a small, opacified right maxillary sinus with a depressed orbital floor (Figure 1), a new finding, as a magnetic resonance imaging study performed 3 years earlier (for new-onset seizures) was normal (Figure 2).

The patient underwent right orbital floor reconstruction with maxillary antrostomy. Before surgical manipulation, the maxillary os was found to be occluded, and an 18-gauge needle attached to a pressure transducer (model 90062A; Space-Labs Inc, Redmond, Wash) was inserted into the sinus. A pressure of −23 mm Hg was recorded.

Comment. Silent sinus syndrome is spontaneous enophthalmos and hypoglobus associated with a small, ipsilateral maxillary sinus.1 It develops over a course of days to years and is not associated with trauma. At presentation, the maxillary os may be patent or occluded, and the sinus may be partially or completely opacified.

One theory for the development of silent sinus syndrome is as follows. Occlusion of the maxillary sinus os forms an enclosed mucosal space where resorption of air creates negative pressure. Such negative pressure has been recorded in occluded rabbit maxillary sinuses2 and in humans.3 Negative sinus pressure may cause thinning and inward bowing of the sinus walls, including the orbital floor, resulting in hypoglobus. If the maxillary os opens, the sinus fluid, which initially may have provided some support for the thin orbital floor, may drain, allowing further depression of the orbital floor and globe. This may account for the rapid presentation of some patients, the variable patency of the os, and the variability of sinus fluid.

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In the more than 25 cases in our experience, and a nearly equal number in the literature, we are aware of only one case of silent sinus syndrome with normal predisease neuroimaging findings\(^4\) and only one report of documented negative sinus pressure.\(^3\) Our patient represents the first case of silent sinus syndrome with both normal predisease imaging and documented negative maxillary sinus pressure, demonstrating unequivocally the acquired nature as well as likely etiologic association with negative maxillary sinus pressure in at least some cases of silent sinus syndrome.

**Figure 1.** Computed tomographic scan (axial [left] and coronal [right]) at time of presentation shows marked depression of the right orbital floor with opacification of the right maxillary sinus.

**Figure 2.** Magnetic resonance imaging study 3 years before presentation shows normal orbits and maxillary sinuses bilaterally (left, anterior; right, posterior).

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