Adduction on Attempted Abduction: The Opposite of Synergistic Divergence

Congenital fibrosis of the extraocular muscles (CFEOM) is a congenital ocular motility disorder that manifests as restrictive ophthalmoplegia with ptosis. Synergistic divergence (SD) is a deficit of adduction associated with simultaneous bilateral abduction on attempted gaze into the field of action of the affected medial rectus muscle. There has been no pathologic report of SD; however, magnetic resonance imaging showed that in 2 patients with CFEOM and SD, the oculomotor nerve was hypoplastic bilaterally and the abducens nerve was absent on the side exhibiting SD. To our knowledge, bilateral deficit of abduction associated with simultaneous bilateral adduction on attempted gaze into the field of action of the lateral rectus muscles, the counterpart of SD, has not been previously reported.

Report of a Case. A 39-year-old man was referred for the evaluation of ophthalmoplegia and ptosis since birth. He underwent a bilateral frontalis sling operation at age 29 years. Otherwise, medical and family histories were noncontributory.

Figure 1. Axial magnetic resonance images of the normal control. A, Cisternal segments of the normal right and left abducens nerves (black single arrows) as a linear dark signal emerging from the pontomedullary sulcus. The normal acoustic nerve (white arrows) and facial nerve (double-stemmed arrow) are seen in the left cerebellopontine angle cistern. The mean diameter of the right and left acoustic nerves is 1.5 mm. B, Abducens nerves (arrows) course in a superior oblique direction at the level of the lower pons. C, The right and left abducens nerves (arrows) finally enter into the clivus. D, Cisternal segments of the normal right and left oculomotor nerves (arrows) are found coursing in an anterior lateral direction at the level of the lower midbrain. The oculomotor nerves appear thicker than the acoustic nerves. The mean diameter of the right and left oculomotor nerves is 2.1 mm.

His corrected visual acuities were 20/60 OU. He had a chin-up posture, and both eyes were fixed in downgaze. He had 10 prism diopters (Δ) of esophoria in the primary position, which increased to an esotropia of more than 50Δ in right gaze, left gaze, and upgaze. He was unable to voluntarily elevate either eye to the primary position, and Bell's phenomenon was absent. Ductions were completely limited in both eyes, except for some residual adduction. During attempted right gaze, left gaze, and upgaze, both eyes adducted. He showed a limited retraction of the globe on attempted adduction, but he did not show upshoots or downshoots of either eye on adduction (Figure 1A). Both eyelids showed ptosis with a palpebral fissure of 2 mm. Both eyelid positions did not show any changes with gaze. Levator function was evaluated at 0 mm in both eyes. Neither eyelid showed movement accompanying mouth movement. Both lenses and fundi were normal. Both pupils reacted briskly to light without a relative afferent pupillary defect and did not contract with eye movement.

On thin-section magnetic resonance imaging, bilateral abducens nerves were absent, and bilateral oculomotor nerves were hypoplastic (Figure 1B-F). The superior rectus muscles of both eyes had severe atrophy, and the medial rectus muscles had mild atrophy (Figure 1G). The inferior and lateral rectus muscles of both eyes were of normal size. In 30 normal controls, the abducens and oculomotor nerves were identified bilaterally in all of them (Figure 2).

Figure 2. Congenital fibrosis syndrome with synergistic convergence. A, Ocular versions demonstrating complete limitation in both eyes, except for some residual adduction. During attempted right gaze, left gaze, and upgaze, both eyes adducted. On axial magnetic resonance images obtained at the levels of the pontomedullary junction (B), the lower pons (C), and the mid pons (D), the right and left abducens nerves are not identified. Compare with the normal abducens nerves in Figure 1. Arrows indicate the right and left acoustic nerves. Axial magnetic resonance images at the level of the upper pons (E) and lower midbrain (F) show small right and left oculomotor nerves (arrows). The mean diameter of the right and left oculomotor nerves (1.0 mm) was smaller than that of the acoustic nerves (1.4 mm), suggesting hypoplasia. G, Coronal magnetic resonance image of both orbits shows severe atrophy of the superior recti and mild atrophy of the medial recti compared with normal inferior and lateral recti (double-stemmed arrows). MR indicates medial rectus; SR, superior rectus; SO, superior oblique; and ON, optic nerve.
Comment. Our study included 1 patient with CFEOM and the counterpart of SD, namely, abduction on attempted abduction. Magnetic resonance imaging showed bilateral hypoplasia of the oculomotor nerve and bilateral absence of the abducens nerve. Based on the pathologic findings of CFEOM and Duane retraction syndrome, the absence of an abducens nerve may represent a specific finding of Duane retraction syndrome and bilateral hypoplasia of the oculomotor nerve, CFEOM. Because this finding was exactly the same as in those with synergistic divergence, we presume that this case is a rare variant of SD that could also be categorized as a neuro-pathologic disease with an aberrant innervation. This case shows that aberrant innervation supports a primary developmental abnormality of the cranial nerves in CFEOM.

In conclusion, our study demonstrates the rare existence of the counterpart of SD. The radiologic findings of bilateral hypoplasia of the oculomotor nerve and the absence of the abducens nerve were the same as those of SD.

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Financial Disclosure: None.


Pegaptanib as an Adjunctive Treatment for Complicated Neovascular Diabetic Retinopathy

Cataract extraction in patients with proliferative diabetic retinopathy may exacerbate the disease. Iris neovascularization, neovascular glaucoma, disc and retinal neovascularization, and macular edema may occur. Sometimes, adequate treatment with laser photocoagulation may be impossible because of media opacities. Intravitreal injection of triamcinolone acetonide might reduce postoperative progression of the iris neovascularization presumably owing to its antiproliferative and anti-inflammatory properties. Recently, pegaptanib sodium, a more selective antiangiogenic agent, has been shown to be safe for intravitreal injections. Since antiangiogenic factors have caused regression of iris vessels in nonhuman primates, we considered intravitreal injection of pegaptanib in a patient with persistent iris neovascularization and media opacities.

Report of a Case. A 53-year-old man had received panretinal photocoagulation (>2000 spots in each eye) for proliferative diabetic retinopathy as well as macular grid laser treatment followed by intravitreal triamcinolone injections for macular edema in both eyes. His left eye had undergone pars plana vitrectomy for macular edema followed by cataract extraction. Subsequent development of neovascular glaucoma in that eye required Baerveldt tube placement and another vitrectomy. Despite aggressive treatments, his vision deteriorated rapidly to hand motion OS.

He returned 2 months later. Visual acuity was counting fingers at 1 ft OD and hand motion OS, with intraocular pressures of 20 and 15 mm Hg OD and OS, respectively. Slitlamp examination revealed extensive rubecution of the angle and peripheral anterior synchiae. Bilateral vitreous hemorrhages precluded the view of the posterior pole.

Salvaging vision in the right eye would have required cataract extraction and vitrectomy, but surgery carried the risk of intraoperative bleeding and exacerbation of neovascular glaucoma. In view of his situation, we discussed with the patient the off-label use of pegaptanib (Macugen; Eyetech Pharmaceuticals, New York, NY) to treat the iris neovascularization, explaining the lack of clinical trials. After obtaining consent, the patient received an intravitreal injection of 0.3 mg of pegaptanib sodium in his right eye. Within 9 days, the iris vessels resolved (Figure).

The patient underwent phacoemulsification with posterior chamber intraocular lens implantation followed by pars plana vitrectomy and endolaser treatment and received intravitreal pegaptanib 4 weeks after the first injection. Neovascularization of the disc was noted intraoperatively. No intraoperative bleeding occurred from the iris, but there was some bleeding from the optic nerve after peeling a neovascular membrane. Despite an initial favorable response, the patient developed postoperative vitreous hemorrhage and recurrent rubecution on postoperative day 7. He subsequently developed elevated intraocular pressure requiring a Baerveldt tube, repeat pars plana vitrectomy and endolaser treatment, and a third injection of pegaptanib 4 weeks after the first surgical procedure. Five weeks after the second surgical procedure, his visual acuity was 20/160 OD and his intraocular pressure was 17 mm Hg OD. The iris and optic disc neovascularization had regressed.

Eight weeks after the second vitrectomy, the iris neovascularization reappeared with a decrease of visual acuity to 20/200 and necessitated a fourth injection of pegaptanib 1 week later. The iris neovascularization regressed again, and visual acuity returned to 20/160.

Comment. Since the iris vessels regressed after the first injection of