ters, and the bacteria were partially identified as coagulase positive the next day.

Thirty-six hours later, he had recurrent pain, decreased vision, reaccumulation of vitreous opacities, and 30% hypopyon. Following the EVS guidelines for re-injection, pars plana vitrectomy was performed and intravitreal antibiotics were administered. The next day, the initial vitreous and AC cultures identified S. aureus resistant to methicillin, vancomycin, ciprofloxacin, levofloxacin, and tetracycline but sensitive to chloramphenicol, quinupristin/dalfopristin, and linezolid (determined by Vitrek 2 system; BioMerieux, Inc.). He had persistent pain, worse vision, and recurrent 20% hypopyon. After informed consent for off-label use, intravitreal quinupristin/dalfopristin (0.4 mg/0.1 mL) and amikacin sulfate (0.4 mg/0.1 mL) were administered. Oral linezolid, 600 mg, and minocycline hydrochloride, 100 mg, were given for 21 days followed by rifampin and minocycline for a total of 3 months. Within 2 days there was significant clearing of vitreous debris and view of secondary vessels. During the next 3 months there was complete resolution of vitreous, AC, and corneal inflammation and return of visual acuity to his baseline of counting fingers.

**Comment.** Antibiotic treatment for bacterial endophthalmitis must be started promptly for optimal success, and the empirical antibiotic chosen should be active against the suspected bacteria even though sensitivities may not be available for up to 72 hours later. Clinicians need to be knowledgeable of the likely causes and sensitivity patterns of the suspected bacteria because delay in treatment can lead to poor outcomes. While all of the gram-positive bacteria in the EVS were sensitive to vancomycin, recent resistant organisms have been reported with increased frequency and are a concern.

Synercid is a streptogramin antimicrobial resulting from the combination of semisynthetic pristinamycin derivatives, quinupristin, and dalfopristin, in a 3:7 ratio. The combination targets both early and late stages of protein synthesis, resulting in synergistic activity, and is bactericidal. Linezolid is an oxazolidinone antibiotic that inhibits protein synthesis by binding to the 50S ribosomal subunit. It is bacteriostatic and has potential adverse effects of irreversible optic neuropathy from systemic use, which could potentially be even greater with intraocular use, although to our knowledge this has not been studied.

Antibiotic resistance is evolutionary and the genes responsible can be transferred between bacteria, leading to increased resistance. If resistance to established antibiotics becomes more common, newer treatment regimens need to be considered for continued successful treatment or there will be poor outcomes in those resistant cases. The EVS showed that intravitreal anti-infectives are effective with or without use of systemic antibiotics for bacterial endophthalmitis. Clinicians should be aware of alternative intravitreal antibiotics when vancomycin-resistant bacteria are present or highly suspected. Although prolonged oral antibiotics have not been necessary in endophthalmitis, a 3-month course of systemic therapy was recommended in these cases of vancomycin-resistant organisms to eradicate any nonocular reservoir of remaining bacteria. The timing of ocular improvement in these patients demonstrates that the effectiveness of intravitreal treatments is more rapid than the reported cases treated by oral linezolid alone. In conclusion, 2 cases of vancomycin-resistant endophthalmitis were successfully treated with intravitreal quinupristin/dalfopristin and oral antibiotics. Clinicians should consider intravitreal quinupristin/dalfopristin for vancomycin-resistant bacterial endophthalmitis.

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**Spectral-Domain Optical Coherence Tomographic Characteristics of Autosomal Recessive Isolated Foveal Hypoplasia**

Foveal hypoplasia, also referred to as foveal planum, is a congenital condition that can be associated with other ocular abnormalities such as aniridia, albinism, microphthalmos, and achromatopsia. Isolated foveal hypoplasia (IFH) is an even rarer disorder, with similar clinical findings in the fovea. The characteristic findings of patients with IFH are nystagmus, poor visual acuity, absent or abnormal maculofoveal reflexes on ophthalmoscopy, and variable and incomplete filtering of the choroidal fluorescence in the macular area on fluorescein angiography. No single hereditary pattern has been established for patients with IFH. Reported cases include patients with autosomal dominant and autosomal recessive inheritance patterns as well as sporadic cases. Only recently has a grading system for the...
spectral-domain (SD) optical coherence tomographic (OCT) findings of foveal hypoplasia been reported.2

Herein, we report the first case series to describe the foveal characteristics on SD-OCT imaging of 5 affected individuals in a single family affected with autosomal recessive IFH with an absent PAX6 mutation.

Report of Cases. The affected individuals in our cohort are all siblings with healthy parents (no history of consanguinity), with 5 of 12 progeny having IFH (Figure 1). Of the 12 siblings, 7 were male and 5 were female. Of the 5 with IFH, 4 were male and 1 was female. All 5 affected members manifested decreased visual acuity (mean, 0.82 logMAR; visual acuities of affected siblings were 20/100 OU in sibling 1, 20/70 OU in sibling 4, 20/70 OD

Figure 1. Pedigree of the family with isolated foveal hypoplasia. Both parents were healthy. Affected siblings with nystagmus, poor visual acuity, and isolated foveal hypoplasia (siblings 1, 4, 6, 8, and 10) are shaded in blue. Squares indicate males; circles, females.

Figure 2. Fundus photographs of 3 of 5 affected siblings with isolated foveal hypoplasia show normal optic discs with a poorly defined foveal region and loss of foveal pit. The central circular white reflex is an artifact. All 5 siblings had a normal anterior segment and normal multifocal electroretinographic findings. A, Fundus of the right eye in sibling 1, a 35-year-old man with visual acuity of 20/100 OU. B, Fundus of the right eye in sibling 4, a 30-year-old man with visual acuity of 20/70 OU. C, Fundus of the right eye in sibling 6, a 26-year-old woman with visual acuity of 20/70 OD and 20/100 OS.

Figure 3. Spectral-domain optical coherence tomographic scan showing the normal foveal anatomy and structural details. ELM indicates external limiting membrane; GCL, ganglion cell layer; ILM, internal limiting membrane; INL, inner nuclear layer; IPL, inner plexiform layer; IS/OS, inner photoreceptor segment/outer photoreceptor segment junction; NFL, nerve fiber layer; ONL, outer nuclear layer; OPL, outer plexiform layer; OPR, outer segment photoreceptor/retinal pigment epithelium complex; and RPE, retinal pigment epithelium. The dashed lines highlight the normal foveal pit (*), extrusion of plexiform layers (†), ONL widening (‡), and outer segment lengthening (§). Foveal grading is described as the following: grade 1, shallow foveal pit, presence of ONL widening, and presence of outer segment lengthening; grade 2, grade 1 but absence of foveal pit; grade 3, grade 2 but absence of outer segment lengthening; and grade 4, grade 3 but absence of ONL widening.3
and 20/100 OS in sibling 6, 20/80 OD and 20/100 OS in sibling 8, and counting fingers at 3 ft OU in sibling 10), pendular nystagmus with variable null points in each affected individual, low to moderate hyperopia (mean hyperopia, 3.3 diopters), dark hair color, normal anterior segment on slitlamp examination, and absent iris transillumination defects. Dilated retinal examinations of each of the affected individuals revealed normal optic discs, absent foveal depression, absent foveal pit, and a normal retinal periphery (Figure 2). Multifocal electroretinographic results were normal in both affected and unaffected family members. Imaging with SD-OCT (Cirrus high-definition OCT; Carl Zeiss Meditec) in all 5 affected siblings demonstrated absent foveal depression, absence of extrusion of plexiform layers, absence of foveal pit, absence of outer segment lengthening, and absence of outer nuclear layer widening, all features corresponding to grade 4 foveal hypoplasia as described by Thomas et al (Figure 3, Figure 4, video 1, and video 2 [http://www.archophthalmol.com]). Imaging with SD-OCT in the unaffected siblings demonstrated normal foveal anatomy. Mutation analysis for PAX6 was performed on saliva samples of both affected and unaffected siblings (Oragene DNA kit; DNA Genotek Inc). No PAX6 mutation was found.

Figure 4. Grade 4 foveal hypoplasia by spectral-domain optical coherence tomographic imaging in the right eyes of 3 of 5 affected siblings, including sibling 1 (A), sibling 4 (B), and sibling 6 (C). Grade 4 foveal hypoplasia is characterized by the absence of extrusion of plexiform layers, absence of foveal pit, absence of outer segment lengthening, and absence of outer nuclear layer widening. Scanning laser opthalmoscopic imaging also demonstrates the lack of a well-defined foveal area. I indicates inferior; N, nasal; S, superior; and T, temporal.
Comment. Isolated foveal hypoplasia is an unusual condition whose phenotypic findings may reflect several genotypes. Hence, variable inheritance patterns have been reported in the literature. The arrival of SD-OCT now allows us to potentially differentiate some of these subtypes, something that was not available to previous investigators.1,3,4 The ability to test for the presence of the PAX6 mutation is another potential way to differentiate between subgroups that was not available to earlier investigators.

Previous studies2,3 have reported SD-OCT findings in patients with IFH. Thomas et al2 described 14 patients with nonfamilial IFH, absent PAX6 mutation, a mean best-corrected visual acuity of 0.2 logMAR, and grade 1 foveal hypoplasia on SD-OCT imaging. Charbel Issa et al3 described 2 patients with foveal hypoplasia in their series, one associated with posterior embryotoxon and the second with IFH. Applying the recently published SD-OCT grading system2 to the images by Charbel Issa et al demonstrates foveal hypoplasia grades 3 and 4, respectively (correlating to their reported best-corrected visual acuities of 0.2 and 0.4 logMAR, respectively). The PAX6 mutation status was not reported for the patients in their series.3

In our case series, we describe 5 affected individuals with familial IFH with an autosomal recessive inheritance pattern and absent PAX6 mutation. Our patients have a mean best-corrected visual acuity of 0.82 logMAR and grade 4 foveal hypoplasia on SD-OCT imaging. Our cohort’s poorer best-corrected visual acuity and more severe grade of foveal hypoplasia demonstrated on SD-OCT imaging support the correlation of structure and function as well as the grading system described by Thomas et al.2

To our knowledge, this is the first case series to describe the foveal characteristics on SD-OCT imaging in a family with autosomal recessive IFH and absent PAX6 mutation. The uniqueness of the SD-OCT findings and the inheritance pattern suggest that our cohort likely represents a different subtype of IFH. The use of SD-OCT in assessing the foveal characteristics and correlating the findings with visual function in patients with IFH may lead us to better understand and isolate different subsets of this rare condition for additional study.

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Morning Glory Disc Anomaly With Peripheral Retinal Nonperfusion in 4 Consecutive Cases

In 1970, Kindler1 first described the morning glory disc anomaly in 10 patients with unusual congenital anomaly of the optic disc. Morning glory disc anomaly manifests with features of a congenitally enlarged optic disc with a central funnel-shaped excavation and overlying thin glial membrane. The anomalous disc is often surrounded by retinal pigment epithelial hyperplasia and occasionally surrounded by exudation and subretinal fluid. The descriptive name “morning glory” depicts the similarity of the recognizable retinal vascular pattern radially emerging from the optic disc, similar to the arrangement on the morning glory flower. These disc vessels are supernumerous and appear straightened and radially course to the retinal periphery. The morning glory disc anomaly can be associated with neurologic abnormalities, so recognition of the ocular finding and appropriate neuroimaging is important.

Peripheral retinal vascular nonperfusion in children has been recognized with several congenital conditions, such as retinopathy of prematurity, Coats disease, familial exudative vitreoretinopathy, facioscapulohumeral muscular dystrophy, and incontinentia pigmenti.2 To our knowledge, there have been no published reports on peripheral fluorescein angiographic findings in morning glory disc anomaly, perhaps because of the limited availability of fluorescein angiography in children and the difficulty in imaging the peripheral retina. Herein, we report the association of morning glory disc anomaly and peripheral retinal nonperfusion in 4 consecutive patients.

Report of Cases. There were 4 patients with morning glory disc anomaly, diagnosed at a mean age of 19 months (median, 11.5 months; range, 4-48 months). In all 4 cases, the morning glory disc anomaly was unilateral. There was no history of prematurity, low birth weight, nonperfusion in the opposite eye, or familial exudative vitreoretinopathy. Brain magnetic resonance imaging was normal in all cases. Demographic and clinical features are listed in Table 1. Retinal vascular features are listed in Table 2.

In all 4 cases, fundus findings, documented with the Retcam camera (Massie Industries) showed peripheral retinal nonperfusion involving 360° in 3 cases and 90°