Comment. Ishak and colleagues\(^3\) and May and colleagues\(^4\) each reported a case of endogenous endophthalmitis in patients with gingival disease that progressed to an abscess. In the latter case, the patient also had undergone a cavity filling 7 days before onset of symptoms.

Our case of endogenous endophthalmitis was in an immuno-competent individual who underwent routine dental cleaning 10 days before seeing an ophthalmologist. She did not undergo any procedures such as cavity filling or tooth extraction on that visit. As noted in previous case reports,\(^3,4\) periodontal disease is well documented as a potential cause of endogenous endophthalmitis. However, as far as we are aware, this is the first reported case indicating that a routine teeth cleaning without evidence of gingival disease or a focal infection, such as a periodontal abscess, can lead to endogenous endophthalmitis. Because α-hemolytic streptococci are known to reside as normal flora in the nasopharynx, we presume that transient bacteremia developed after the dental cleaning, which led to seeding of the organism into intraocular tissues. A lag time of 7 to 10 days before onset of symptoms appears to be consistent with previous reports in the literature.\(^4\) In short, a careful history in any patient with symptoms of uveitis should include inquiries regarding routine dental cleaning.

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**Chiasmal Enlargement and Optic Nerve Enhancement on Magnetic Resonance Imaging in Leber Hereditary Optic Neuropathy**

Findings on magnetic resonance imaging (MRI) of the brain and orbits are typically normal in patients with Leber hereditary optic neuropathy (LHON). We describe 2 patients with LHON who had abnormalities of the optic nerves and chiasm disclosed by MRI.

**Report of Cases.** Case 1. A 7-year-old boy had subacute painless decline in vision in both eyes in the middle of July 1999. An evaluation by a local ophthalmologist on August 23, 1999, revealed a visual acuity of 20/60 OD and 20/300 OS. Findings from the remainder of the examination were reportedly normal and the patient was given the diagnosis of functional visual loss. He was otherwise healthy with no other neurologic or systemic symptoms and no medical illnesses. Family history was unremarkable for ophthalmologic or neurologic disease.

Neuro-ophthalmologic evaluation was first performed on September 15, 1999, approximately 2 months after the onset of his visual decline. On examination he had a visual acuity of 20/100 OD and 20/400 OS. Kinetic perimetry showed central scotomas in both eyes. With Ishihara color testing, he identified 8 of a total of 14 plates with the right eye and 3 of a total of 14 plates with the left eye. Pupillary examination showed 4-mm, 2+ reactive pupils with a left relative afferent pupillary defect. Dilated fundus examination showed optic nerve pallor, greater in the left eye. The remainder of the ophthalmologic and neurologic examination findings were unremarkable.

Results from additional tests, including a complete blood cell count, erythrocyte sedimentation rate, blood chemistry, angiotensin-converting enzyme level, antinuclear antibody titer, syphilis serology, and chest radiograph, were normal.

An MRI of the brain and orbits with orbital fat suppression techniques following the administration of intravenous gadolinium was obtained on the day of evaluation (approximately 2 months after the onset of visual loss) and showed enlargement of the optic chiasm and enlargement and enhancement of the intracranial optic nerves (Figure 1, A and B). An optic pathway glioma and optic neuritis were initially considered as potential causes of this patient's visual decline. However, mitochondrial DNA testing disclosed a mutation at nucleotide position 11778 confirming the diagnosis of LHON.

A lumbar puncture showed an opening pressure of 100 mm H\(_2\)O and normal cerebrospinal fluid contents. Myelin basic protein and oligoclonal bands were not present. Results from the cerebrospinal fluid IgG index and cerebrospinal fluid cytology were normal.

Prior to obtaining the results of mitochondrial DNA testing, the patient was treated with high-dose systemic steroids (intravenous methylprednisolone followed by oral...
prednisone) for 2 weeks without improvement in his vision. Repeat cranial MRI 5 months after visual decline showed reduction in the amount of optic nerve enhancement. Cranial MRI 2 years after visual loss showed normal optic nerves and chiasm without enlargement or enhancement (Figure 1, C).

Case 2. A 19-year-old man noted unexplained loss of vision in his left eye 4 years prior to our evaluation. His vision was subsequently stable until 3 weeks prior to our evaluation when he noted progressive, painless visual loss in his right eye. The visual loss progressed over 2 weeks. He had no other neurologic or systemic symptoms and had no medical illnesses. Family history was unremarkable for ophthalmologic or neurologic disease.

On the day of his examination, neuro-ophthalmologic evaluation showed a visual acuity of light perception OD and of 20/400 OS. He was unable to identify Ishihara color plates with either eye. Confrontation visual fields and automated perimetry could not be plotted in his right eye and showed a central scotoma in his left eye. His pupils were equal, poorly reactive, and had a right relative afferent pupillary defect. Fundus examination findings showed bilateral optic nerve pallor, greater in the right eye. Findings from the remainder of the ophthalmologic and neurologic examination were normal.

Laboratory test results, including those for complete blood cell count, erythrocyte sedimentation rate, angiotensin-converting enzyme level, and syphilis serology, were normal. An MRI of the brain and orbits with orbital fat suppression techniques following the administration of intravenous gadolinium was obtained on the day of examination and showed enlargement of the optic chiasm without enhancement of the optic nerves or chiasm (Figure 2). Mitochondrial DNA testing revealed a mutation at nucleotide position 3460 thereby confirming a diagnosis of LHON. Examination findings 4 months after this initial examination remained unchanged.

Comment. Leber hereditary optic neuropathy is a maternally inherited optic neuropathy that typically affects males between the ages of 15 and 35 years.1 The classic funduscopic appearance is characterized by elevation and hyperemia of the
optic disc, tortuosity of the retinal vasculature, and peripapillary tel-angiectatic microangiopathy. The discovery of several mitochondrial DNA mutations responsible for LHON has broadened the clinical spectrum of this disease. Many patients with genetically confirmed LHON do not report a family history of visual loss and do not have the classic fundus appearance. Some of these patients are women and others are outside the typical age range. The patient described in case 1 was young, had no family history of ophthalmologic disease, and never demonstrated the typical fundus features of LHON. His first neuroophthalmic evaluation occurred 2 months after his visual loss and the fundus features of LHON may have been observed had he been evaluated at the time of his visual loss. The patient described in case 2 was atypical in that he had no family history of ophthalmologic disease, a 4-year interval prior to involvement of the second eye, and the typical fundus features of LHON were never observed despite the fact that he had had a neuro-ophthalmologic evaluation 3 weeks after visual loss in his right eye.

Magnetic resonance images of the brain and orbits are typically normal in patients with LHON. However, increased signal from the retrobulbar optic nerve on T2-weighted fast spin echo and short time inversion recovery sequences has been described. The increased optic nerve signal may not occur until several months after the onset of visual loss. Optic nerve sheath distention has been demonstrated with orbital ultrasonography, computed tomography, and MRI in a patient with LHON. Cranial MRI of our patients demonstrated enlargement of the optic chiasm in both cases and mild enlargement and enhancement of the proximal, intracranial optic nerves in case 1. The association of LHON with enlargement of the optic nerves and chiasm on MRI has not been reported. Although the classic fundus findings in LHON suggest pathology at the optic nerve head, chiasmal and perichiasmal pathology has been suggested. Weiner et al and Bair have described bitemporal visual field defects that respect the vertical meridian in patients with LHON. Imachi and Nishizaki noted arachnoid thickening and adhesions around the chiasm during surgery in 120 patients with LHON. Vaphiades and Newman described a patient with LHON who had enhancement of the retrobulbar optic nerves on MRI obtained several weeks after the onset of visual loss. To our knowledge, there are no other reported cases of optic nerve enhancement among patients with LHON, although many of the previous MRI studies did not obtain designated images of the orbits with orbital fat suppression techniques following the administration of intravenous gadolinium.

These cases demonstrate that MRI may show enlargement of the optic nerves and chiasm and optic nerve enhancement in patients with LHON. These neuroimaging signs may mimic an optic pathway glioma or optic neuritis. Leber hereditary optic neuropathy should be considered as a possible cause of any unexplained case of unilateral or bilateral optic neuropathy.

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Microcornea and Subluxated Lenses Due to a Splicing Error in the Fibrillin-1 Gene in a Patient With Marfan Syndrome

Marfan syndrome (MFS) is an autosomal dominant syndrome resulting from mutations in the fibrillin-1 gene (FBNI) on chromosome 15q21.1. Ectopia lentis is the major ocular criterion. Minor ocular criteria include flat corneas, increased axial length (>23.5 mm), and iris hypoplasia with miosis. We report the first genetically confirmed case of microcornea in MFS with a novel FBNI mutation.

Report of a Case. This female patient with MFS (including tall stature, positive wrist and thumb signs, joint hypermobility, highly arched palate, frontal bossing, and mitral valve prolapse) demonstrated progressive inferotemporal lens subluxations (Figure 1A and B) and increasing myopic astigmatism in both eyes. Her keratometry readings were 42.35 diopters OU, and axial lengths were 25.7 mm OU. She refused spectacle correction and became contact lens intolerant as a result of poor hygiene.

At age 14 years, she underwent lensectomy with insertion of an anterior chamber intraocular lens (AC-IOL) in both eyes. A suitable IOL was not used to help avoid retinal detachment and subluxation from suture degradation. Microcornea (horizontal diameter, 10 mm OU) was identified on the right eye during surgery because it was b oth