ity. Bonnet3 as well as Postel et al9 reported eyes with optic pits and associated retinal detachment managed with intravitreal gas injection, in which gas bubbles migrated through a small break in tissue overlying the optic pits into the subretinal space postoperatively. Coll et al10 reported a communication between the subretinal space and the vitreous cavity in the morning glory syndrome.

Our case suggests that PFD should be avoided in retinal detachment surgery in eyes with optic pits. Furthermore, intraocular air/gas tamponade may be preferable to silicone oil tamponade if feasible to prevent the complications previously mentioned. The lower interfacial tension of silicone oil was not adequate to prevent bubbles of silicone oil from passing through the small retinal opening within the optic disc. Endolaser photocoagulation for formation of a barrier to fluid movement was effective here to prevent silicone oil from passing into the subretinal space. Such laser treatment appears prudent in similar cases when air/gas tamponade is insufficient to secure retinal attachment and when silicone oil must be used. The volume of both subretinal PFD and silicone oil increased during a short period of time. It is currently unknown which unidirectional forces are operative to efficiently move fluid from the vitreous cavity through the optic pit under the neurosensory retina. A pressure gradient must be assumed whereby gravity forces are unlikely because PFD and silicone oil owing to their specific weights would be expected to behave differently. It may be speculated that the pressure gradient is the difference between the intraocular pressure at the moment of migration and the pressure within the reservoir (ie, either the extraocular tissue pressure or the pressure within the subarachnoid space).

**Idiopathic Juxtafoveal Telangiectasis in Association With Celiac Sprue**

Idiopathic juxtafoveal telangiectasis (IJFT) is a condition of uncertain etiology that is characterized by retinal telangiectasias, superficial retinal crystalline deposits, right-angle venules, and intraretinal pigment plaques. It is capable of causing visual loss in otherwise healthy patients, and treatment remains controversial. It was first described by Reese in 19561 and has subsequently been divided into 3 groups by Gass and Blodi.2

We describe a patient with IJFT and celiac sprue. To the best of our knowledge, this is the first reported case showing a potential association between these 2 conditions.

**Report of a Case.** A 53-year-old woman with biopsy-proved celiac sprue reported a 10-year history of blurred central vision in both eyes.

**Her medical history was otherwise unremarkable. She denied a history of diabetes and had a normal blood glucose level, but she declined a formal glucose tolerance test. Her medical regimen included a multivitamin, hormone replacement therapy (estrogen-medroxyprogesterone), and a gluten-free diet. Her family history included no relatives with similar visual loss.**

Best-corrected visual acuity was 20/60 OD and 20/40 OS. Clinical examination showed clear media and symmetric optic discs. Macular edema and a perifoveal gray macular sheen were noted with apparent foveal thinning; no lipid or hemorrhage was present. A right-angle venule was noted in the temporal right macula (Figure 1).

Fluorescein angiography showed intraretinal telangiectasia with leakage for 340° in the right eye and nearly 360° surrounding the fovea in the left eye (Figure 2). Optical coherence tomography showed foveal thinning in both eyes with mild macular thickening (Figure 3).

**Comment.** Celiac sprue is characterized by malabsorption due to injury to the intestinal mucosa after ingestion of wheat gluten or related proteins. There is both clinical and histologic improvement on a strict gluten-free diet, and relapse occurs when dietary gluten is reintroduced.3 Until recently, celiac sprue was thought to be an uncommon condition in the United States, with an estimated prevalence of 1 in 3000. However, greater awareness of its manifestations and the advent of more accurate serologic tests have led to the realization that celiac sprue is relatively common, affecting approximately 1 of every 120 to 300 persons in North America.4

Celiac sprue is caused by an inappropriate T cell–mediated immune response against ingested gluten, causing damage to the intestinal mucosa, and the classic histologic finding of absent villi and hyperplastic crypts. There is a strong genetic component, with more than 95% of patients expressing the HLA-DQ2 heterodimer.5

Classically, celiac sprue is seen in infants with impaired growth and abdominal distention. The...
onset of symptoms is gradual, following the introduction of cereals into the diet. The condition is also increasingly being diagnosed in adults, in whom symptoms include diarrhea, flatulence, weight loss, and lactose intolerance. However, many patients have atypical symptoms or none at all.

To our knowledge, this case represents the first reported association between celiac sprue and IJFT. Although the concurrence of the 2 disorders may be coincidental, it may represent a previously unrecognized association, due to the historic underdiagnosis of celiac sprue by the medical community.

The underlying pathophysiologic characteristics of IJFT are still largely unknown. Green and associates studied the histologic features of a patient with group 2A IJFT and found thickened retinal capillaries with narrowed lumens. This was theorized to be a result of endothelial degeneration and regeneration, with increased basement membrane production, and secondary pericyte loss. Gass and Blodi suggested that these might cause decreased metabolic exchange and increased endothelial permeability, thus leading to chronic nutritional damage to the juxtafoveal retinal cells.

Likewise, some studies suggest that the primary defect in celiac sprue may be an abnormally in-
increased permeability of the intestinal epithelium. A preexisting permeability defect may trigger the expression of celiac sprue by allowing ingested gluten to cross the epithelial barrier and incite a pathologic immune reaction in genetically susceptible individuals.

Thus, one possible explanation for an association between IJFT and celiac sprue is an underlying systemic permeability defect that is responsible for both increased gluten hypersensitivity in the gut and endothelial decompensation in the retina. As more patients with celiac sprue are identified, future investigations should disclose whether there is a true causal association between celiac sprue and IJFT.

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**Infantile Orofacial Hemangioma With Ipsilateral Peripapillary Excavation in Girls: A Variant of the PHACE Syndrome**

The PHACE syndrome is a neurocutaneous syndrome that includes the following primary features: posterior fossa malformations of the brain, large facial hemangiomas, arterial anomalies, cardiac anomalies and aortic coarctation, and eye abnormalities. It occurs almost exclusively in girls. Several recent reports have documented an association between orofacial hemangioma and excavated optic disc anomalies (morning glory disc anomaly and peripapillary staphyloma) in girls. Metry et al recently proposed that this association falls within the spectrum of PHACE syndrome. We document ipsilateral intracranial vascular abnormalities in 2 girls with juvenile orofacial hemangioma and excavated optic discs to provide further evidence that these patients fall within the spectrum of PHACE syndrome.

**Report of Cases. Case 1.** A 6-year-old girl was referred for evaluation of unilaterally decreased vision in the right eye. She was born full term, and her perinatal course was uneventful. At 2 weeks of age, she developed multiple hemangiomas involving the face, lip, parotid gland, and throat on the right side. At 1 month of age, these hemangiomas began to grow rapidly (Figure 1A). Magnetic resonance imaging confirmed massive hemangiomatous involvement of the right parotid gland and right side of the face. Results of a cardiology evaluation with echocardiography were normal. The hemangiomas continued to enlarge until the patient was 9 months of age. Treatment with oral steroids and multiple laser treatments to the gingival mucosa and the parotid gland produced regression of the tumor. At 1 year of age, she was noted to have an esotropia of the right eye.

**Visual acuity** was 20/160 OD and 20/20 OS. Both pupils reacted briskly to light, and there was a mild right afferent pupillary defect. Extracocular movements were full. Fixating near, she had 10 prism diopeters of esotropia and fixated eccentrically with the right eye. Results of slit-lamp examination were normal. Cycloplegic refractive error was +2.00 OD and +2.75 OS. Retinal examination disclosed a morning glory disc anomaly in the right eye with an elevated pigmented area of juxtapapillary scarring temporal to the disc (Figure 1B). Magnetic resonance angiography demonstrated marked tortuosity of the supraclinoid right internal carotid artery and narrowing of the proximal right middle cerebral artery (Figure 1C).

**Case 2.** A 6-week-old girl was evaluated for right-sided epiphora. Her perinatal course had been complicated by respiratory difficulties requiring temporary resuscitation. External examination disclosed a large capillary hemangioma on the right upper eyelid; tip of the nose; and right cheek, neck, and shoulder. In the ensuing weeks, the patient developed respiratory insufficiency requiring intubation. Results of a cardiology evaluation with echocardiography were normal. Magnetic resonance imaging disclosed several large hemangiomas in the chest, with extension to the upper aorta, displacement of the right upper lobe of the lung, and displacement of the trachea and larynx. The orofacial component of the hemangioma involved the right masseter muscle, parotid gland, orbit, and soft tissues of the tongue and lips on the right side.

Ophthalmologic examination of the patient at age 6 months showed esotropia and dense amblyopia of the right eye. A small con-