being the reference. Analyses were adjusted for age, sex, complement factor H (CFH) Y402H polymorphism, and smoking—the latter two being other major risk factors for ARM.6

Of 963 subjects, 738 subjects had complete data. There were 1424 gradable eyes.

**Results.** As shown in the Table, by comparison with the GG genotype, the TT genotype was associated with very high risk for all types of ARM, with increasing odds ratios according to the severity of ARM (from 4.60 for early ARM1 to 23.63 and 16.15 for late atrophic and neovascular ARM, respectively). By contrast, associations of the different types of ARM with the GT genotype were modest (OR, 1.45-2.47) and reached statistical significance only for early ARM1.

Smoking and CFH Y402H remained independently associated with ARM, after controlling for ARMS A69S genotypes (data not shown).

**Comment.** This population-based study confirms the major contribution of the TT genotype of the ARMS2 A69S polymorphism in early and late ARM, independently from the other 2 major risk factors (smoking and CFH Y402H polymorphism). Associations with the GT genotypes were much weaker, suggesting a moderate codominant genetic mode of action. Strengths of this study include the population-based setting and the photographic assessment and detailed grading of ARM status.

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**Peripheral Ischemic Retinopathy in Adams-Oliver Syndrome**

Adams-Oliver syndrome (AOS) is an inherited congenital condition defined by aplasia cutis congenita and transverse limb defects. Additional variable involvement of the brain, eye, skin, and cardiovascular system has led to the consideration of this syndrome as a constellation of clinical findings resulting from early embryonic vascular abnormalities.2 Herein, we report a case of AOS with previously unreported retinal findings consistent with an ischemic-proliferative retinopathy.

**Report of a Case.** A full-term girl was delivered at 40 weeks’ gestation to a healthy, nonconsanguineous couple. Prenatal ultrasonography detected lower limb deformities. Weight, length, and head circumference at birth were between the 25th and 50th percentiles. Aplasia cutis congenita with a large scalp and skull defects in the vertex area together with upper and lower limb reduction defects were noted. Widespread cutis marmorata was also observed (Figure 1). Complete systemic examination disclosed a mild ventricular septal defect that spontaneously closed within the first 2 weeks of life.

Ocular examination of the right eye disclosed prominent iris vessels and posterior retinal arterial narrowing and venous dilatation with combined (arterial and venous) blood column segmentation (“boxcarring” appearance). Peripheral avascular retina with capillary dropout, arteriovenous anastomosis, and telangiectasia on the temporal side were noted. The left eye was slightly microphthalmic, with microcornea, a leukoma, and temporosuperior scleral erosion (Figure 2). Detailed left funduscop was not possible because of media opacity. The patient’s parents were thoroughly in-
formed about management options, and it was decided that peripheral retinal laser ablation would be performed in the right eye and conservative management would be implemented in the left eye. Two weeks after photocoagulation, microvascular anomalies in the right eye had regressed.

The exposed dura was covered with artificial dermis on the seventh day of life to prevent hemorrhage and infection. Postoperative complications included sepsis and repeated longitudinal sinus bleeding despite abdominal muscle fascia grafting. The patient died of disruption of the leptomeningeal membrane and brain herniation on the 87th day of life (Figure 1). The patient’s parents declined an autopsy.

Comment. Anomalies related to AOS are thought to result from genetically decreased stability of embryonic blood vessels and/or abnormal endothelial regulation, which compromise the vascular status in the form of reduced perfusion and ischemia.

Abnormal pericytal recruitment or coverage of the vasculature was demonstrated in the pathologic examination of a patient with AOS and pulmonary hypertension. Interestingly, the ocular pathology of this patient revealed incomplete retinal vascularization with gliosis and intravitreal neovascularization. The funduscopic findings of our patient together with this previous pathologic data may support an ischemic-proliferative retinal process in this syndrome.

Peripheral retinal ischemia in children may appear in a fully vascularized retina or in a primarily immature, avascular retina. The terminal nature of the retinal circulation makes the peripheral retina susceptible to small-vessel disease and ischemic conditions. The features of the retinopathy of our patient mimic the clinical findings of the primary immaturity or retinal vascular maldevelopment of infants with retinopathy of prematurity, familial exudative vitreoretinopathy, or Norrie disease.

The ocular findings suggestive of Peters anomaly and persistent fetal vasculature in the left eye of this patient may not have been coincidental because abnormalities in cellular apoptosis and migration may occur in this syndrome.

In conclusion, we have described previously unreported retinal findings consistent with an ischemic-proliferative retinopathy. These findings strengthen the small-vessel vasculopathy hypothesis in AOS.
mic examination and funduscopy in children with AOS are warranted. Retinal ablation to arrest the potential evolution from ischemia to neovascularization, further glial organization, and retinal detachment should be considered.

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Recovery of Vision From No Light Perception in Giant Cell Arteritis

Up to 50% of patients with giant cell arteritis (GCA) have visual symptoms early in the disease course, in most cases due to anterior ischemic optic neuropathy (AION). The vision loss from AION in GCA is often devastating, with the initial visual acuity being 20/200 or worse in more than 50% of patients. There is often, but not always, pallid optic disc edema and there is rarely a significant recovery, even with timely initiation of corticosteroids. We describe a patient with biopsy-proven GCA who had severe vision loss.

Figure 2. Ocular findings in a patient with Adams-Oliver syndrome. A, Prominent right iris vessels. B, Left microcornea with leukoma. C, Detailed right funduscopy showing arterial (arrows) and venous (arrowheads) "boxcarring." D, Retinal arterial narrowing (arrows) and venous dilatation (arrowheads). E, Peripheral avascular retina with arteriovenous anastomosis, telangiectasia (arrows), and venous beading (arrowheads).