cyclosporine also can be an effective steroid-sparing agent to help relieve the ocular symptoms of SLK.[12]

Medical and surgical therapies aim to address the proposed mechanisms in SLK. Initial local treatment options consist of avoiding contact lens wear and topical treatments including lubricants, steroids, vitamin A, cromolyn sodium, and rebamipide.[1,10,11] Lacrimal punctal occlusion and autologous serum drops can be considered in patients with tear film deficiencies.[7]

Surgically, lax bulbar conjunctiva can be treated to promote adhesion of the conjunctiva to the sclera (amniotic membrane transplant, silver nitrate/thermocautery).[13] Patient 5 underwent a conjunctival excision with amniotic membrane transplant. Conjunctivoplasty involving resection of the redundant conjunctiva can be effective when other treatment options have failed and was performed in 1 patient in our series.[5]

In conclusion, clinicians should be aware of the possible association between ptosis surgery and SLK. SLK is an uncommon condition that may be missed because the symptoms are nonspecific, and the characteristic ocular lesions may be hidden under the upper eyelid. Early diagnosis and treatment have the potential to alleviate symptoms that otherwise may be incapacitating for patients.

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Footnotes and Disclosure

The authors have no proprietary or commercial interest in any materials discussed in this article.

Ultra-wide-field retinal imaging in tetralogy of Fallot before and after cardiac surgery

Congenital heart disease can be classified physiologically into 3 categories: volume overload, cyanotic, and obstructive. Cyanotic congenital heart disease (cCHD) includes lesions with right-to-left shunts or mixing abnormalities (e.g., transposition of the great vessels, persistent truncus arteriosus, tetralogy of Fallot).[1] Tetralogy of Fallot (ToF) encompasses a variety of anatomic abnormalities, including a large and unrestrictive ventricular septal defect, right ventricular outflow tract obstruction, overriding of the aorta, and right ventricular hypertrophy. The reported ocular findings in patients with cCHD are retinal vascular tortuosity, retinal hemorrhages, disc edema, papilledema, ischemic retinopathy, uveitis, and central retinal vein occlusion. Retinal vascular tortuosity is found to be the most frequent change.[2] The etiology of these vascular changes has been attributed to hypoxia and secondary erythrocytosis, even though systemic vascular endothelial growth factor (VEGF) levels are found to be elevated in these patients.[3] This article describes retinal vascular changes using ultra-wide-field (UWF) imaging in a case of ToF prior to and after surgical correction of ToF.

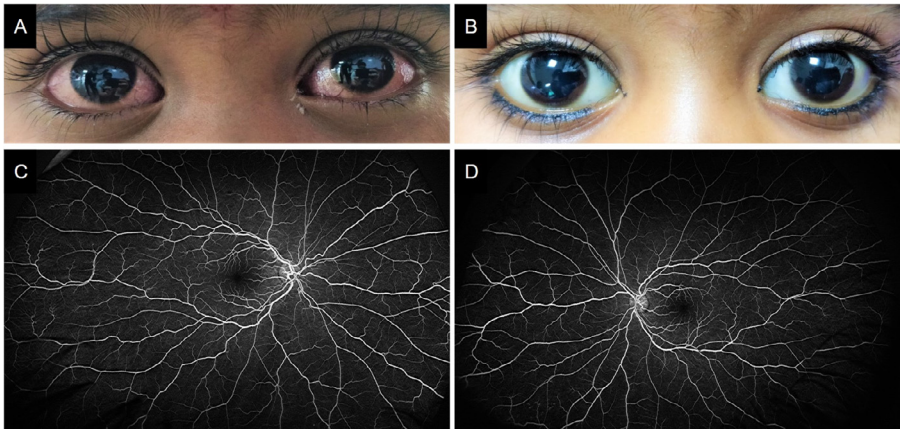

Fig. 1—Clinical picture of the eye before and after cardiac surgery (A, B) and postoperative UWF fluorescein angiography imaging (C, D). Dilated and tortuous conjunctival and episcleral vessels with congestion (A) and their resolution after cardiac surgery (B). Postoperative UWF fluorescein angiography revealed marked reduction in dilatation and tortuosity of the vessels and no evidence of peripheral capillary nonperfusion areas, neovascularisation, or disc edema OU (C, D).

A 5-year-old female child with cCHD (ToF) was referred for fundus evaluation. The child had central and peripheral cyanosis with oxygen saturation (SpO_2) of 68% on room air. Hemoglobin was elevated to 25.8 g/dL, red blood cell count was 8.74 million/μL, and hematocrit was 77.7%. Visual acuity was found to be 20/20 OU by the Lea symbol chart. Intraocular pressure was within normal limits OU. Bilateral dilated vessels were seen in the upper eyelids with telangiectatic conjunctival and episcleral vessels (Fig. 1A). UWF fundus imaging using Optos cSLO (confocal scanning laser ophthalmoscopy; Optos PLC, Dunfermline, UK) revealed dilated and tortuous arterial and venous vessels OU associated with a cyanotic hue (Fig. 2A, B). Three days later the patient underwent an intracardiac repair with anterior septal tricuspid commissuroplasty and right ventricular outflow tract patch plasty with autologous pericardium with patent ductus arteriosus clipping. Postoperatively, on day 14, SpO_2 was 80% at room air with hemoglobin of 13.3 g/dL, red blood cell count of 4.94 million/μL, and a hematocrit of 44.9%. Ophthalmologic examination showed a visual acuity of 20/20 with dramatic resolution of upper eyelid, conjunctival, and episcleral vessel tortuosity bilaterally (Fig. 1B). UWF imaging showed marked reduction in the calibre of vessels, vessel tortuosity, and branching and a more reddish hue in the colour of the vessels. A perivascular dark impression on the retinal surface corresponding to the preoperative cyanotic vessel width also was noted (Fig. 2C, D). At 6-month follow-up, it was found that normal retinal vascular anatomy was maintained and that the perivascular dark impression started showing segmental clearing OU (Fig. 2E, F). UWF fluorescein angiography revealed findings consistent with fundus evaluation with no capillary nonperfusion areas, neovascularisation, or disc edema (Fig. 1C, D).

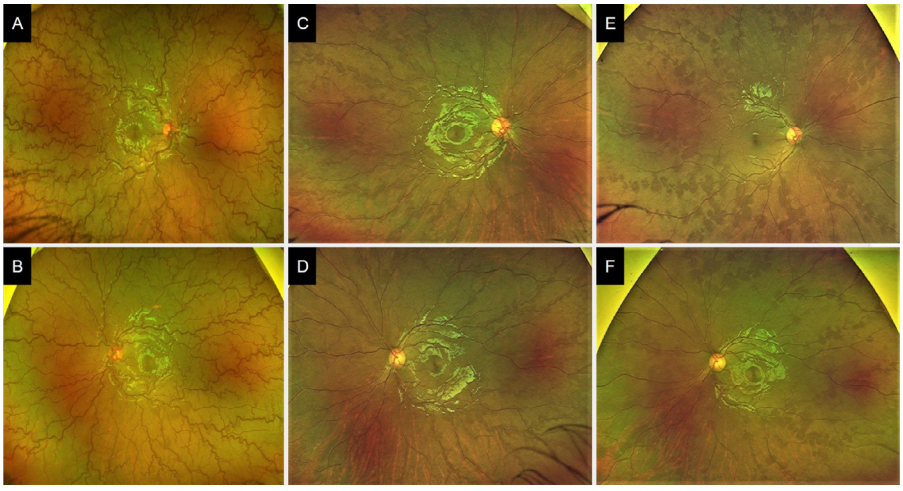

Fig. 2—Ultrawide field retinal imaging before and after cardiac surgery. Dilated and tortuous arterial and venous vessels with a cyanotic hue OD (A) and OS (B). After surgical correction, a marked reduction is noted in the calibre of vessels and vessel tortuosity with a more reddish hue in the colour of the vessels. A perivascular dark impression on the retinal surface corresponding to the preoperative cyanotic vessel width is also noted OD (C) and OS (D). At 6-month follow-up, segmental clearing of the perivascular dark impression is noted OU (E, F).

The most common ocular manifestations of cCHD include retinal vascular tortuosity, retinal vascular dilation, and darkening. Various hypotheses have been considered to explain these changes in the retinal vasculature in cases of cCHD. Peterson et al proposed that severity of retinopathy in cCHD is associated with hypoxia and polycythemia in 50% of the patients with cyanotic CHD.[3] They found no correlation of retinopathy with $PaCO_2$, pH, central venous pressure, type of cardiac abnormality, or age of the patient. Crowe et al reported a reduction in retinal vascular changes that occurred as a result of polycythemia and hypoxia in patients with cCHD after surgical repair.[3] The findings in our case corroborate these studies and demonstrate that these retinal vascular changes are reversible as early as 14 days after surgical repair of the cCHD. Mansour et al. noted that patients developed vascular tortuosity regardless of the hematocrit (high or low), thereby implicating low oxygen saturation as the major causative factor for vascular changes.[1] Retinal tissue has one of the highest oxygen consumptions per gram of tissue weight, and these vascular changes demonstrate the retinal vascular compensatory adaptations in response to hypoxia. Although compensatory angiogenesis in the form of systemic collateral vessels at other sites has been noted, these abnormal vessels have not been found to have any significant relationship with VEGF levels.[4] UWF imaging is quick and noninvasive and can be easily performed even in children. The presence of significant retinal changes probably can be a surrogate marker of disease severity (reflecting changes from hypoxia and polycythemia). Additionally, the rapid regression of changes could indicate surgical success, although a larger study is required to confirm this hypothesis.

The retinal and cerebral circulations have similar autoregulatory processes. Therefore, assessing the retinal circulation may help in assessing the status of the cerebral circulation. A decrease in white matter volume has been documented in adults with cCHD.[3] The electroencephalogram is often abnormal in patients with congenital cyanotic heart disease. The disturbance of cerebral function and structural changes may be due to a complex interplay of factors such as chronic oxygen desaturation or due to decreased cerebral blood flow resulting from polycythemia-related hyperviscosity.[5] Thus, detection of abnormal retinal findings can be important predecessors to detecting the effects of hypoxia on cortical function.

Retinal vascular dilation and tortuosity are the most characteristic findings in patients with cCHD and are attributed to hypoxia and/or secondary erythrocytosis. Systemic VEGF may not be responsible for causing retinal vascular changes, and neovascularization is not a frequent change seen in ischemia caused by cCHD. Improved imaging using UWF pseudocolor imaging and fluorescein angiography can help in documenting retinal vascular changes prior to and after cardiac surgical correction. These changes also may correlate with vascular changes occurring in the cerebral cortex.

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Preserved retinal sensitivity following spontaneous regression of soft drusen

Soft drusen are the clinical hallmark of intermediate age-related macular degeneration (AMD) and a major risk factor for late-stage disease. Soft drusen usually increase in size, area, and confluence with aging.[1] Their natural life cycle may involve retinal pigment epithelium (RPE) changes, collapse, and macular atrophy or spontaneous regression.[2] Pharmacologic and laser photocoagulation treatments intended to induce drusen regression have yet to show visual benefit.[1,3] Early treatment of intermediate AMD before progression to macular atrophy and/or neovascular complications is an area undergoing intense study and would be a breakthrough. However, whether soft drusen already represent an irreversible loss of retinal structure and function remains a topic of some debate. Confocal microperimetry has been demonstrated to be reliable in evaluating retinal function via threshold retinal sensitivity.[4] Herein we report the multimodal imaging including spectral-domain optical coherence tomography (SD-OCT) and confocal microperimetry of