

# Widefield Imaging-guided Treatment of Pediatric Vitreoretinal Diseases

BY YOSHIHIRO YONEKAWA, MD; AND R.V. PAUL CHAN, MD, MSC, FACS

Recent years have ushered in many advances in retinal imaging. From spectral-domain optical coherence tomography to ultra-widefield fundus imaging, the new modalities are proving to be an integral part in the evaluation of pediatric retinal conditions. One of the biggest perceived hurdles in managing pediatric retinal patients is the challenging clinical examination; most adult patients can cooperate with the dilated fundoscopic examination more easily than a 2-year-old in the office. The traditional approach to examining young children would be examination under anesthesia (EUA) and retinal drawings to document and monitor retinal pathology. This general paradigm remains common practice, but advanced imaging technologies are now able to complement the EUA, and, perhaps most appealing, can be employed in the outpatient setting for selected disorders and age groups.

## CONTACT WIDEFIELD IMAGING

RetCam (Clarity Medical Systems), now in its third rendition, allows widefield fundus photography and fluorescein angiography (FA). A coupling agent is applied onto the cornea, and a hand-held camera with several choices of widefield lenses (up to 130°) are applied directly on the ocular surface. RetCam is commonly used for documentation during EUAs and retinopathy of prematurity (ROP) screenings. ROP telemedicine programs have shown that nonphysician operators can be trained to obtain RetCam images for remote expert interpretations.<sup>1-3</sup> Images from premature infants are usually acquired with only topical anesthesia, but careful and efficient exams have been shown to cause minimal distress for the infants.<sup>4</sup> Photographic documentation in ROP can also be a valuable asset for medicolegal and educational purposes.<sup>5</sup>

## NONCONTACT ULTRA-WIDEFIELD IMAGING

The Optos widefield imaging system uses confocal scanning laser ophthalmoscopes with ellipsoid mirrors to create images of up to 200° of the retinal periphery and

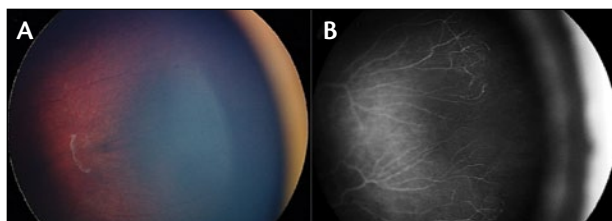
can be utilized through a nonmydriatic pupil. The 200Tx version also allows angiography and autofluorescence. In adults, ultra-widefield photography serves to document, but does not always add new diagnostic information beyond a good dilated retinal examination. However, Optos images can be of great value in pediatric patients, in whom only fleeting views of the fundus may be possible in the office. Given the rapid acquisition of the retinal image, children may be more amenable to Optos photography than clinical examinations, and very often we have found this to be the case in our clinical practice.

Ultra-widefield fluorescein angiography (UWFA) with Optos can provide invaluable information for a retina specialist evaluating children. Studies in adults,<sup>6</sup> and more recently in children,<sup>7</sup> have shown that significant pathology often exists beyond the standard 7 ETDRS fields. Older children may be able to tolerate fluorescein injections, but for younger children we recommend using oral fluorescein. Children too young will require RetCam FAs during EUAs. A recent paper suggests using UWFA for ROP imaging by holding up preemies onto the Optos interface in clinic,<sup>8</sup> but the safety of this technique must be evaluated, because babies requiring treatment are often the youngest, smallest, and sickest neonates, frequently on life-sustaining devices.

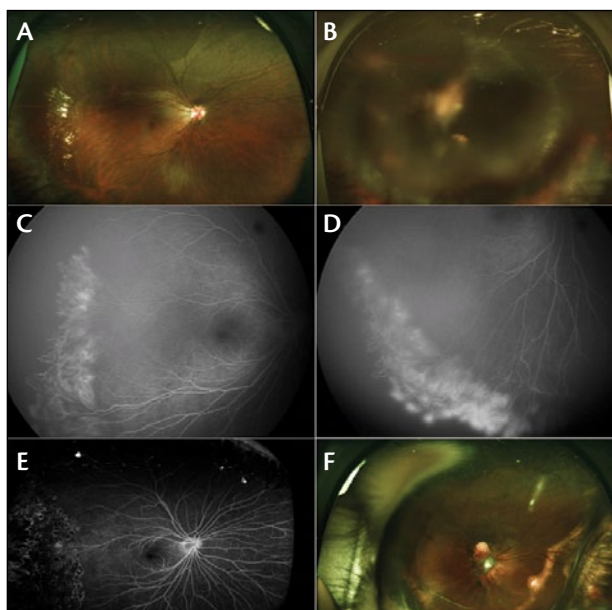
## WIDEFIELD FLUORESCEIN-GUIDED TREATMENT

### Retinopathy of Prematurity

Image-guided laser treatment for ROP has been well described.<sup>9-11</sup> RetCam FA allows appreciation of fibrovascular proliferation and precise delineation of avascular retina that requires photocoagulation. Both can be deceiving on clinical exams or RetCam photographs alone. Intravitreal anti-VEGF therapy is a treatment option for ROP, but children may need to be examined more frequently and for longer periods after treatment due to the possibility of delayed normal vascularization of the retina with poten-



**Figure 1.** A premature infant was treated with intravitreal bevacizumab for zone 1 stage 3 + retinopathy of prematurity. Two years later, the peripheral retina remains avascular (A), which is more easily seen with RetCam fluorescein angiography (B).

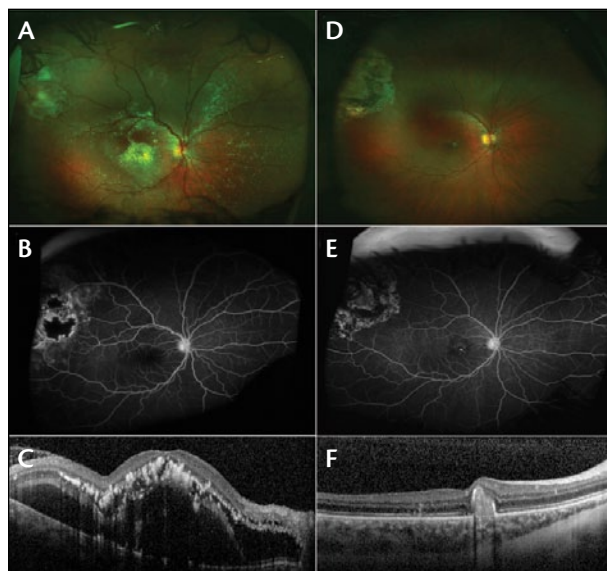


**Figure 3.** A 12-year-old boy was referred for neovascular glaucoma and vitreous hemorrhage of the left eye. On examination and Optos imaging, the right eye had temporal vascular dragging and peripheral exudations (A). There was no view into the left eye due to vitreous hemorrhage (B). An EUA was undertaken, and RetCam FA of the right eye showed peripheral nonperfusion with fluorescein leakage at the avascular-vascular zone (C-D). The patient was diagnosed with familial exudative vitreoretinopathy (FEVR), and the right eye was treated with fluorescein-guided laser photocoagulation (E), and the left eye underwent pars plana vitrectomy after bevacizumab intravitreal injection (F).

tial for late recurrence.<sup>12</sup> In Figure 1, a RetCam FA shows avascular peripheral retina in a child with ROP treated with intravitreal bevacizumab (Avastin, Genentech).

### Coats Disease

The mainstay of treatment for Coats disease is ablation of the telangiectatic vessels to reduce exudation.



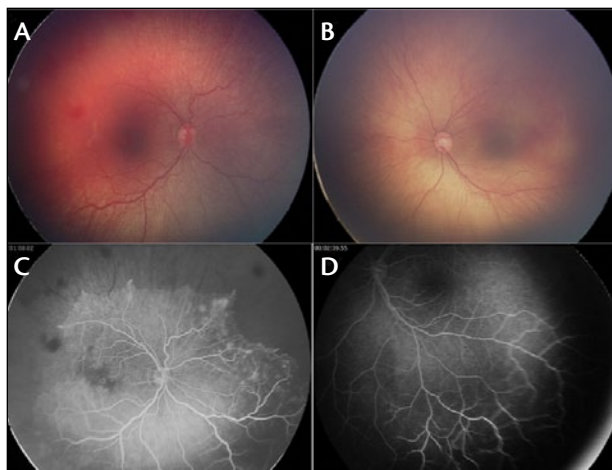
**Figure 2.** A 9-year-old boy presented with Coats' disease. Optos photography (A) and fluorescein angiography (B) revealed diffuse exudations with peripheral vascular telangiectasia in the superotemporal periphery. Spectral-domain optical coherence tomography (C) also shows subretinal fluid accumulation. Laser photocoagulation was delivered to the telangiectatic vessels, and the exudation gradually improved (D-F). Adopted from Kang KB et al. Ultra-widefield imaging for the management of pediatric retinal diseases.

Anti-VEGF agents have also been used. Laser delivery is most accurately planned based on angiographic findings. RetCam FA and UWFA are excellent modalities to identify the abnormal vasculature that requires treatment (Figure 2).

### FEVR and IP

Widefield angiography is a strong diagnostic tool for familial exudative vitreoretinopathy (FEVR; Figure 3) and incontinentia pigmenti (Figure 4). Both entities present with abnormal retinal vascular development, and fluorescein-guided laser ablation of peripheral nonperfusion is a key treatment strategy; avascular peripheral retina can be difficult to fully appreciate without angiography. The pathology may be located in the far periphery in subtle presentations, in which cases UWFA would be very useful. However, a detailed EUA may be necessary if the diagnosis is unclear. In Figure 3, we present a patient referred for neovascular glaucoma and dense vitreous hemorrhage. RetCam FA of the contralateral eye during an EUA revealed findings consistent with the diagnosis of FEVR. Pediatric retina patients often do not present until vision is severely affected in one or both eyes. The view may be compromised, making it difficult to determine the patho-

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**Figure 4.** A 4-month old girl with incontinentia pigmenti was admitted for seizures and intracranial hemorrhage. It may be difficult to appreciate the peripheral nonperfusion with RetCam photography alone (A-B), but the findings become clear with RetCam FA.

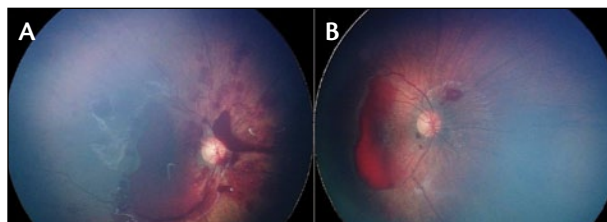
logical etiology. Clues in the contralateral eye can provide essential diagnostic information in such cases.

### OTHER USES

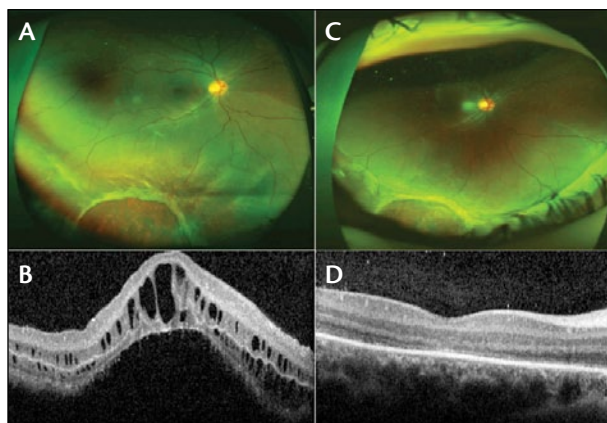
UWFA in clinic, or RetCam FA in the intensive care unit or during EUAs, can be used for myriad other pediatric retinal disorders.<sup>13</sup> Sick cell retinopathy in children and peripheral tumors such as capillary hemangiomas also benefit from widefield imaging. Abusive head trauma is important to document photographically (Figure 5), and studies have shown that peripheral nonperfusion can also be seen.<sup>14</sup> The technology is also useful for pre- and postsurgical assessment, such as in Figure 6, where we use ultra-widefield imaging to evaluate a pediatric patient with a retinal detachment.

### LIMITATIONS OF CURRENT TECHNOLOGIES

The RetCam uses a contact system, so children need to be under anesthesia during EUAs, or premature infants can be swaddled. The camera is also sensitive to media opacities, potentially leading to inferior image quality compared with other imaging systems. Small pupils are difficult to work around, and dark fundi generally produce poorer images. The widest possible angle is 130°, but peripheral pathology may be difficult to clearly photograph. Fortunately for ROP imaging, eyes that require referral for treatment or an indirect ophthalmoscopic examination often do not require imaging of the far peripheral retina.<sup>15</sup> It should also be noted that excessive pressure on the globe can mask plus disease in eyes with ROP and should be avoided.<sup>16</sup> A challenge during EUAs can be Bell's phenomenon if children are not



**Figure 5.** Photodocumentation is essential in the care of patients with abusive head trauma. Retinal findings include hemorrhages in different layers (A-B), retinal folds, and traumatic retinoschisis. RetCam imaging is appropriate in these patients who are often encountered in the neonatal or pediatric intensive care units.



**Figure 6.** Preoperative Optos image (A), and spectral-domain optical coherence tomography (B) of a 10-year-old boy with a chronic macula-off rhegmatogenous retinal detachment with an inferior retinal break. The corresponding images after a primary scleral buckle demonstrate reattached retina (C-D). The noncontact nature and fast acquisition times allow outpatient ultra-widefield imaging, even in postoperative children who may be in discomfort after surgery.

sedated enough. In such cases, anesthesia can be deepened, or an assistant can infraduct the globe.

Artifacts may also be common in Optos images, and the green and red laser wavelengths create unrealistic color renderings. The resolution of the posterior pole may also be lower compared with conventional fundus cameras. Optos cameras are currently exclusively used in the outpatient clinic setting because of their size and requirement for cooperation from an awake patient. The most recent Daytona model is smaller, but still not completely portable.

### CONCLUSION

Widefield imaging provides clinically meaningful data to help us better manage our pediatric retina patients. The accumulating evidence will allow us to solidify the

appropriate indications. Cost will hopefully decline, so that this versatile technology becomes more accessible to middle- and lower-income nations, where the need is likely greatest. Regardless of the incredible advances we have seen with the advent of these new imaging modalities, we should always have a low threshold to examine pediatric patients under anesthesia if the examination in the office is insufficient. ■

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