Ultra-Widefield Imaging for the Management of Pediatric Retinal Diseases

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ABSTRACT

**Purpose:** To describe the utility of using ultra-widefield digital fundus photography and ultra-widefield fluorescein angiography (UWFA) in the pediatric patient population to evaluate peripheral retinal pathology and to manage Coats’ disease and familial exudative vitreoretinopathy (FEVR).

**Methods:** Retrospective review of pediatric retinal patients with FEVR or Coats’ disease who underwent ultra-widefield fundus photography and UWFA.

**Results:** Eight patients were included in this case series. Five patients had the clinical diagnosis of FEVR, and two eyes of two patients with FEVR received UWFA-guided laser photocoagulation. Three patients were diagnosed as having Coats’ disease and received UWFA-guided laser photocoagulation.

**Conclusions:** Ultra-widefield fundus photography and UWFA can be used successfully as an outpatient procedure in the pediatric patient population without the necessity of examination under anesthesia and can aid the physician in the documentation and evaluation of peripheral retinal pathology. UWFA can also assist in directing laser photocoagulation in the treatment of pediatric retinal diseases.


INTRODUCTION

Many proliferative and exudative retinal diseases in the pediatric patient population, including familial exudative vitreoretinopathy (FEVR) and Coats’ disease, involve abnormalities in the peripheral retina. Thorough examination of the peripheral retina by the physician is essential in the proper evaluation and early treatment of these diseases, because retinal detachment can develop in a large portion of eyes with advanced disease. Over the past few decades, fundus photography has served to complement physical examination by providing objective documentation of fundus findings to allow for comparisons over time and to monitor disease progression. In addition, fluorescein angiography has helped physicians evaluate retinal diseases by allowing for improved visualization of retinal non-perfusion, vascular leakage, microvascular abnormalities, and neovascularization. Traditional imaging techniques, including the conventional 7-standard...
fields (7SF), as described in the Early Treatment of Diabetic Retinopathy Study, have long been used to capture pathologies of the posterior pole. However, traditional imaging of the peripheral retina has been challenging, requiring a high level of photographic expertise and patient cooperation. This challenge can be especially true in the pediatric patient population.

In contrast to 7SF, which captures approximately 75 degrees of retina, a newer imaging technique first used in 2004, the Optos Optomap Panoramic 200A imaging system (Optos, PLC, Dunfermline, Scotland), can capture up to 200 degrees of high-resolution image in a single exposure. This imaging modality, known as “ultra-widefield,” has been used in the adult patient population to evaluate and manage various retinal diseases, including diabetic retinopathy, choroidal lesions, uveitis, retinal vascular occlusion, retinal detachment, and sickle cell retinopathy. However, the feasibility and utility of this imaging modality have not been well characterized in the pediatric patient population. In this case series, we demonstrate the utility of using ultra-widefield digital fundus photography and ultra-widefield fluorescein angiography (UWFA) in the pediatric patient population to evaluate peripheral retinal pathology and to assist in the management and treatment of Coats’ disease and FEVR.

PATIENTS AND METHODS

This case series was approved by the Institutional Review Board for human subjects at the Weill Cornell Medical Center. For the purpose of this case series, retinal imaging, retinal drawings, examination records, and surgical records of pediatric retinal patients who presented to Weill Cornell Medical Center from October 2010 to January 2012 were reviewed. The age, sex, gestational age at birth, presence of systemic conditions, and family history of patients were recorded. In all pediatric patients, the decision to perform ultra-widefield fundus imaging was determined by the attending physician. In eyes requiring laser treatment, the UWFA of the diseased eye was reviewed prior to laser photocoagulation to assist with targeted retinal photocoagulation.

Eligibility criteria for this case series included pediatric patients (younger than 18 years) who were seen at the Weill Cornell Medical Center and diagnosed as having either Coats’ disease based on Shields’ criteria or FEVR based on the diagnostic criteria provided by Ranchod et al. or pediatric patients who underwent ultra-widefield fundus photography and UWFA. Exclusion criteria consisted of media opacity limiting utility of UWFA and concomitant retinal diseases or pathology. No patients who met the above inclusion criteria were excluded from this case series.

RESULTS

Eight pediatric patients were diagnosed as having either FEVR or Coats’ disease at Weill Cornell Medical Center over the study period. Table 1 summarizes baseline demographic and surgical characteristics of patients and staging of diseased eyes. Five patients were diagnosed as having FEVR. Of note, 4 of the 5 patients were siblings whose mother also had a clinical diagnosis of FEVR. Of the patients with FEVR, laser photocoagulation was indicated in two eyes of two patients due to the presence of peripheral neovascularization. Laser photocoagulation was performed by a pediatric retinal specialist (RVPC). After treatment, neovascularization regressed and retinal vessel leakage improved. Both patients experienced stable visual acuity.

Of the three patients diagnosed as having Coats’ disease, laser photocoagulation was indicated in all cases due to the presence of telangiectasias and extrafoveal exudation. One patient received treatment at an outside hospital. One patient required re-treatment by the pediatric retinal specialist (RVPC). Of the patients treated at Weill Cornell Medical Center, one experienced improved visual acuity from 20/500 to 20/100 and the visual acuity of the other patient was stable. Neovascularization regressed and retinal vessel leakage improved in all of these eyes.

Case Reports

Ultra-widefield Imaging in the Evaluation and Management of FEVR. An 11-year-old boy reported decreased vision and floaters in both eyes and was referred to Weill Cornell Medical Center by his pediatric ophthalmologist with a diagnosis of vitritis. The patient was born full-term. His best-corrected visual acuity was 20/20 in both eyes. Intraocular pressures of both eyes were normal. There were no abnormalities in the anterior chamber. Fundus examination showed slight dragging of the retinal vessels temporally without neovascularization in the right eye. Fundus examination of the left eye showed vitreous debris inferiorly, loss of retinal blood
vessels temporally, and dragging of the retinal vessels temporally (Figures 1A-1B). No abnormalities were found on optical coherence tomography using Spectralis HRA+OCT (Heidelberg Engineering, Inc., Heidelberg, Germany) (Figure 1C). UWFA showed areas of non-perfusion in the superotemporal and inferotemporal retina, and neovascularization temporally (Figure 1E). A clinical diagnosis of FEVR was made, and laser photocoagulation with 810-nm laser was applied to areas of retinal non-perfusion in the periphery of the left eye. Six weeks after initial laser treatment, UWFA (Figures 1E-1F) showed residual retinal non-perfusion associated with vascular leakage temporally. Additional laser treatment was performed with treatment boundaries delineated by UWFA findings (Figures 1G-1H). After re-treatment, retinal neovascularization regressed. The patient experienced no complications or visual acuity loss. Of note, the patient's family was subsequently examined and underwent UWFA because the autosomal dominant form of inheritance of FEVR is common. The patient's mother and three siblings were diagnosed as having FEVR based on clinical and UWFA findings.

**Ultra-widefield Imaging in the Evaluation and Management of Coats’ Disease.** A 9-year-old boy presented with decreased vision in the right eye for 3 months’ duration associated with central and nasal visual field defects, as well as intermittent flashes of light. The patient’s best-corrected visual acuity was 20/500 in the right eye and 20/25 in the left eye. Intraocular pressures of both eyes were normal. There were no abnormalities in the anterior chamber. Fundus examination of the right eye showed diffuse macular edema, a significant amount of exudate in the fovea, telangiectatic vessels, and exudation in the superotemporal retina (Figure 2B). Optical coherence tomography of the right eye showed large cystic changes in the neurosensory retina and subretinal fluid (Figure 2A). UWFA showed areas of non-perfusion in the superotemporal retina, superotemporal retinal vessel telangiectasias, and aneurysmal dilatations consistent with the characteristic “light bulb” appearance of Coats’ disease (Figure 2C). Laser photocoagulation with 532-nm laser was applied to the right eye with treatment boundaries delineated by UWFA findings. After treatment, ultra-widefield fundus imaging showed improved subretinal and intraretinal fluid with complete obliteration of the telangiectatic

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RS = retinal specialist; ped oph = pediatric ophthalmologist; OS = left eye; OD = right eye; FEVR = familial exudative vitreoretinopathy; FA = fluorescein angiography; VA = visual acuity

*Patient did not receive laser treatment.*

*Patient was transferred to an outside hospital for care.*
Figure 1. (A) Ultra-widefield images of the left eye of a patient with familial exudative vitreoretinopathy. White rectangle indicates area magnified in 1B demonstrating area of retinal neovascularization. (B) Enlarged image (of white rectangle in 1A) showing area of neovascularization temporally (red circle). (C) Optical coherence tomography of left eye showed no abnormalities. (D) Ultra-widefield fluorescein angiography (UWFA) showing additional areas of non-perfusion in the superotemporal and inferotemporal retina. (E) Ultra-widefield fundus photography and (F) UWFA post-laser showing residual temporal peripheral retinal non-perfusion and areas of vessel leakage (delineated in red). (G) Ultra wide-field fundus photography and (H) UWFA showing additional laser performed with treatment boundaries delineated by UWFA findings.
vessels (Figure 2D). In addition, no new vessel leakage was observed on UWFA (Figure 2E). The patient’s visual acuity improved from 20/500 to 20/100 at the 1-month follow-up visit.

**DISCUSSION**

The key findings of this series are (1) ultra-widefield fundus photography and UWFA can be used successfully in the pediatric patient population in the outpatient setting to evaluate peripheral retinal pathology and (2) UWFA may assist in directing laser photocoagulation in the treatment of certain pediatric retinal diseases.

Due to difficulty with patient cooperation in the pediatric population, adequate evaluation of the entire retina using fluorescein angiography often requires examination under anesthesia in the operating room. Although examination under anesthesia is generally a safe procedure, various complications may exist, including minor side effects from the administration of anesthetic agents such as nausea, vomiting, discomfort at the injection site, and rare adverse reactions such as alterations in blood pressure, anaphylaxis, respiratory depression, and cardiac arrest.\(^{24,25}\) In addition, pediatric patients with certain conditions (eg, severe cardiac...

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*Figure 2.* (A) Optical coherence tomography of the right eye of a patient with Coats’ disease showing cystoid macular edema and subretinal fluid. (B) Ultra-widefield fundus photography of the right eye showing exudates in the macula and superotemporal retinal exudates and vessel telangiectasias. (C) Ultra-widefield fluorescein angiography (UWFA) showing areas of non-perfusion and blockage in the superotemporal retina and aneurysmal dilatations consistent with the characteristic “light bulb” appearance of Coats’ disease. Seven-standard fields border is outlined in white. (D) Ultra-widefield fundus photography and (E) UWFA after treatment with laser.
and pulmonary diseases) may not be able to tolerate repeated examination under anesthesia. Examination under anesthesia in the operating room can also be more time-consuming for both the physician and the patient than examination in the office, and it can increase the number of unnecessary visits to the operating room.

Although laser photocoagulation treatment may eventually require the use of anesthesia in the operating room, in this case series, we demonstrate the feasibility of using ultra-widefield imaging in the pediatric patient population in physician offices without the use of anesthesia. Compared to other types of peripheral retinal imaging (eg, contact lens systems and 7SF), ultra-widefield imaging is a non-contact system capable of capturing 200 degrees of retina in a single exposure without the need for repositioning or refixation, and thus requires less patient cooperation.26,27 Due to these advantages, UWFA can help physicians to avoid performing fluorescein angiography in the operating room and limit anesthesia exposure to the pediatric patient. This technique may thus improve examination efficiency and decrease the delay in disease diagnosis and treatment. Although only cases of Coats’ disease and FEVR were reported in this case series, we believe that this imaging modality may facilitate the evaluation and management of retinal diseases in the cooperative pediatric patient population.

Not only does UWFA reduce the need to bring patients to the operating room, but it also allows visualization of peripheral retina findings that are difficult or impossible with other imaging techniques. A recent study on adult patients with diabetic retinopathy showed that UWFA covers more total retinal area, more areas of retinal non-perfusion, and more areas of neovascularization than the conventional 7SF.6 Similarly, we found UWFA may help to identify peripheral retinal pathology not detected by the conventional 7SF in the evaluation of pediatric retinal diseases. As shown in Figure 2C, UWFA illustrated additional areas of retinal telangiectasias, vessel leakage, and non-perfusion in the superotemporal retina in an eye with Coats’ disease, all of which was outside the border delineated by 7SF.

Because UWFA helps visualize peripheral pathology not seen by other imaging techniques, it can serve as a useful reference in the treatment of peripheral retinal diseases. UWFA allows physicians not only to visualize areas of retinal vascular leakage and neovascularization in the peripheral retina, but also to detect and delineate areas of retinal non-perfusion that may not be evident on examination. As demonstrated in Figure 1F, UWFA clearly outlines areas of continued retinal pathology after laser photocoagulation. As shown in this case series, it is possible to use these UWFA images as a reference during laser photocoagulation to specifically target areas of retinal non-perfusion and continued neovascularization.

There are certain limitations of the case series that should be considered. First, the number of patients included in the series was small (n = 8). However, the goal of the case series was to report the feasibility and potential utility of using ultra-widefield imaging in the pediatric patient population. It is possible that with a larger number of eyes, one can systematically analyze the benefit of using UWFA compared to traditional fluorescein angiography in the pediatric patient population, including patient outcome and cost-benefit analyses. Second, this is a retrospective case series. With a prospective study, certain characteristics of pediatric retinal diseases on UWFA and ultra-widefield fundus photography can be noted and followed in the long term. It is possible that these characteristic features seen on UWFA and ultra-widefield fundus photography can help predict prognostic features of diseases.

We have demonstrated that ultra-widefield imaging can reveal peripheral retinal pathologies and can serve as a useful tool for physicians in the evaluation and management of pediatric retinal diseases. Physicians may now better use ultra-widefield imaging in the cooperative pediatric patient population.

REFERENCES

8. Friberg TR, Gupta A, Yu J, et al. Ultrawide angle fluorescein an-


