Abstract:
The purpose of this report is to illustrate the clinical applicability of Optos Panoramic 200MA ultra-wide-field Fluorescein angiography (Optos fa) imaging in early diagnosis and treatment of new onset sickle cell proliferative retinopathy.

Details:
Case history:
An asymptomatic 27 year old African-American female presented to the Ocular Disease Clinic of the SUNY State College of Optometry for retinal consultation. The patient had been lost to follow-up for 1 year. The ocular history was significant for low myopia in both eyes, a history of posterior vitreous detachment in the right eye and sickle cell retinopathy in both eyes with possible proliferative changes in the right eye. Her medical history was significant for sickle cell disease with Hb SC genotype and a positive family history of sickle cell abnormalities. Her current medication was Folic Acid for sickle cell anemia.

Pertinent findings:
The patient’s best corrected visual acuities were 20/20 in each eye with a -1.25 sph prescription in each eye. Pupils were reactive to light with no afferent pupillary defect and confrontation fields were full. Anterior segment examination was unremarkable except for grade 1 Giant Papillary Conjunctivitis (GPC) in both eyes. Intraocular pressures were 14mmHg in the right eye and 14mmHg in the left. Posterior pole, optic nerve head and the macular region appeared to be normal in both eyes. Peripheral fundus examination of the right eye revealed a presence of posterior vitreous detachment, areas of scattered old sunburst lesions and fibroglial proliferation with surrounding dilated vessels in the temporal retina. Peripheral fundus examination of the left eye was noted to have areas of scattered old sunburst pigment proliferation in the temporal and superior-temporal retina. No neovascularization was visualized in the left eye. Both eyes were free of holes, tears or retinal detachments.

The images of both eyes were captured with Optos Panoramic 200 at SUNY, results of which demonstrated small areas of anomalous vasculature likely to be a surface neovascularization in the far temporal periphery of the right eye and in the superior-temporal quadrant of the left eye. Areas of capillary avascularity in the far periphery of both eyes were also highly probable, but this diagnosis required fluorescein angiography imaging.

The patient was then scheduled for the next available ultra-wide-field fluorescein angiography by Optos Panoramic 200MA (Optos fa) imaging to detect suspected focal areas of surface neovascularization, possible leakage and zones of capillary avascularity. Optos fa imaging of the right eye clearly revealed fluorescein dye leakage in the temporal far periphery, along with
peripheral areas of avascularity 360 degrees and a focal area of non-perfusion surrounded by telangiectatic vasculature in temporal far periphery. Optos fa imaging of the left eye demonstrated an area of fluorescein leakage in a sea fan pattern in a far periphery superior-temporally and three small focal areas of fluorescein leakage far-periphery at 3’, 8’ and 10’ o’clock along with areas of avascularity far periphery 360 degrees with surrounding abnormal telangiectatic vasculature. Those images allowed a clinician to visualize about a 200-degree view of a fundus at different time-intervals of fluorescein angiography with high precision and image quality and to confirm the diagnosis of capillary non-perfusion and proliferative sickle cell retinopathy.

Differential diagnosis:
Primary/leading: Proliferative sickle cell retinopathy OU
Others: Coats disease, Eales disease, proliferative diabetic retinopathy, BRVO/CRVO, Sarcoidosis and Systemic Lupus Erythematosus

Diagnosis and discussion:
Sickle cell disease is the most common genetic disease worldwide. Increased life expectancy of sickle cell patients has led to higher prevalence of blinding complications in the advanced stages of the disease. Proliferative sickle retinopathy (PSR) is a severe ocular change of the disease that may lead to blindness secondary to development of vitreous hemorrhage and retinal detachment. It is thought that pathogenesis of the disease is the series of peripheral vascular occlusions that lead to hypoxic events which in turn trigger growth of new vessels in a “sea fan” pattern. These signs are often peripheral and visualization can be difficult by traditional fluorescein angiography (FA) or observation alone. The technology at present can allow only a 50-degree field of view of posterior pole imaging or 75-degree field of view with composite images. There had been reports on using contact lens-based systems to increase the field of view in traditional FA; however, good patient cooperation and sedation are often required and images often contain significant peripheral blur. Optos fa is a new non-contact method that is able to avoid these complications by using the virtual point technology with tri-frequency scanning laser beam; thus, allowing visualization of up to 200 degrees or 82% of the retina in a single capture. At present, it is the most effective method in visualizing and diagnosing peripheral vascular abnormalities. This case had demonstrated that Optos fa was imperative in detection of peripheral abnormal vasculature in both eyes; and a definitive diagnosis of proliferative sickle cell retinopathy (PSR) was thus able to be obtained.

Treatment/management:
Optos fa images were reviewed by an experienced vitreo-retinal surgeon who opined that timely treatment was essential. The patient then underwent a scattered sectoral photocoagulation argon laser treatment of proliferative neovascularization of the right eye, followed by the scattered sectoral photocoagulation treatment of the left eye. Studies had shown that the rate of spontaneous regression of sea fans is about 32%-45.7% and it is more common in SS patients than in SC patients. Scattered sectoral photocoagulation is thought to be the most effective treatment for new vessel occlusion with a rate of regression of about 81.2%, which is significantly higher than the rate of spontaneous regression. Another treatment that is well-described in the literature is 360 peripheral circumferential scatter photocoagulation that results in 78% of PSR regression. However, due to the lack of controlled
trials, this treatment is currently reserved for extensive peripheral neovascularization. There has been a report on successful use of Avastin injection in regression of PSR. Comparisons of photocoagulation to anti-VEGF treatment are not yet reported.

**Conclusion:**
This case demonstrates the value of ultra-wide-field fluorescein angiography by Optos Panoramic 200MA (Optos fa) in early detection and prompt treatment of peripheral proliferative sickle retinopathy in a Hb SC sickle cell patient.