

## Introduction

- Retinal dystrophy is a genetic disease which is characterised by the degeneration of retinal cells.
- Retinitis pigmentosa (RP) is the most common retinal dystrophy.
- In recent years, studies have shown significant changes in RP retina vasculatures when compared to controls by using optical coherence tomography angiography (OCTA).
- OCTA is fast, non-invasive and able to produce 3-dimensional images.
- Retinal vasculatures at the superficial capillary plexus (SCP), deep capillary plexus (DCP), choriocapillaris (CC) and area of foveal avascular zone (FAZ) were studied.

## Aim

- This review aims to summarise the vascular findings of RP patients which were observed with OCTA.

## Methods

- Search terms such as 'retinal dystrophy', 'vascular' and 'optical coherence tomography angiography' were keyed into five databases which were Ovid Embase, Ovid Medline, Web of Science, Cochrane Library and Scopus.
- The records found were screened to remove duplicates, case reports or studies written in languages other than English.
- Studies were included when they mentioned retinal dystrophy, vascular changes and OCTA.
- The results of included studies were then summarised based on the layers of retina.

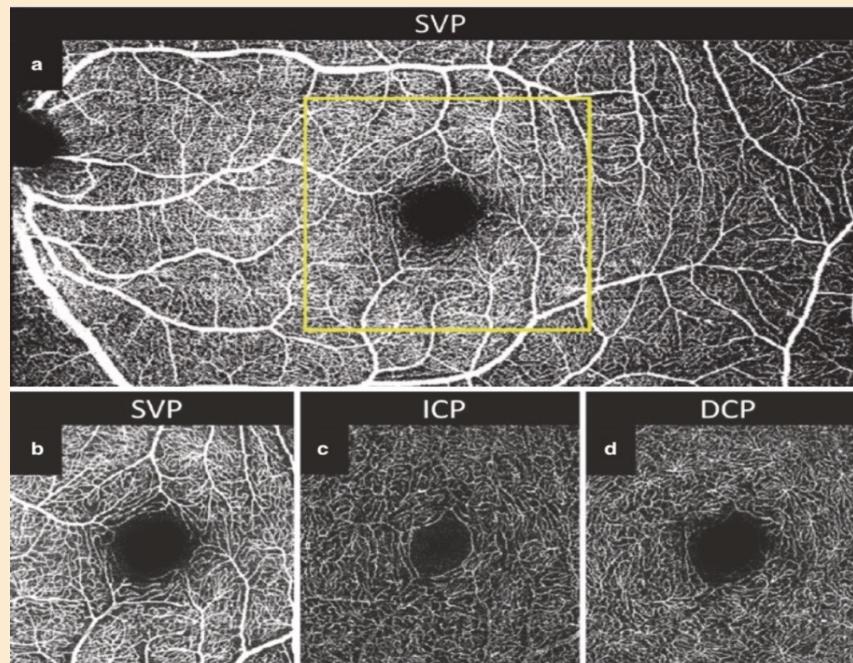


Figure 1: Examples of OCTA en face images of different layers in the retina. Image a) shows the retinal vessels in the superficial vascular plexus. The yellow box signifies the same area taken in image b) but magnified. Image c) and d) are en face images of intermediate capillary plexus and deep capillary plexus respectively<sup>1</sup>.

## Results

- This review consist of 33 articles, which included 2 meta-analysis, 3 reviews, 4 case series, 7 case-control studies and 17 cross-sectional studies.
- Most studies reported that SCP, DCP and CC vessel density (VD) were significantly lower in RP patients than in controls. However, some studies recorded non-significant differences between RP and control retinas in each layer.
- There were mixed results about the difference in FAZ area between RP patients and controls.
- Significant correlations were found between SCP VD, DCP VD and best corrected visual acuity (BCVA).
- CC VD did not significantly correlate with BCVA.
- There were heterogenous results about the correlation between FAZ area and BCVA.
- OCTA parameters were recorded to be significantly correlated to visual field.

## Discussion

- The included studies were commonly limited by their small sample size, lack of genetic characterisation, OCTA image artefacts and cross-sectional study design.
- Heterogenous results might be due to different OCTA models used, different methodologies in recruiting RP patients and various correlation analysis test used.
- Vascular changes were detected by OCTA before patient reported symptoms<sup>2</sup>.
- Hypothesis for vascular remodelling in RP retinas included photoreceptor cell atrophy<sup>3</sup>, increased endothelin-1 (ET-1) concentration<sup>4</sup> and migrating RPE cells form an extracellular matrix around vessels<sup>5</sup>.
- Future work will need to include genetic characterization of RP patients and longitudinal studies designs to study the progression of RP. Artificial intelligence can be utilised to provide better OCTA image quality.

## Conclusion

- OCTA has been proven to have more advantages than fundus fluorescein angiography and indocyanine green angiography, but it is not superior to other imaging modalities such as optical coherence tomography and fundus autofluorescence.
- OCTA can be used adjunctly with other investigative tests to provide multi-modal imaging to RP patients.

## References:

1. Rocholz et al., 2019, *High Resolution Imaging in Microscopy and Ophthalmology: New Frontiers in Biomedical Optics*. Cham (CH): Springer, doi: [10.1007/978-3-030-16638-0\\_6](https://doi.org/10.1007/978-3-030-16638-0_6)
2. Takagi et al., 2018, *Acta Ophthalmologica*, doi: [10.1111/aos.13680](https://doi.org/10.1111/aos.13680).
3. Eysteinnsson et al., 2014, *Acta Ophthalmologica*, doi: [10.1111/aos.12359](https://doi.org/10.1111/aos.12359).
4. Cellini et al., 2010, *Can J Physio Pharmacol.*, doi: [10.1139/Y10-036](https://doi.org/10.1139/Y10-036).
5. Li et al., 1995, *Ophthalmology*, doi: [10.1016/s0161-6420\(95\)30953-0](https://doi.org/10.1016/s0161-6420(95)30953-0)

## Abbreviations:

RP: retinitis pigmentosa. OCTA: optical coherence tomography angiography. SCP: superficial capillary plexus. DCP: deep capillary plexus. CC: choriocapillaris. FAZ: foveal avascular zone. VD: vessel density. BCVA: best corrected visual acuity. ET-1: endothelin-1.

