Introduction: Since its debut in clinical practice, optical coherence tomography angiography (OCTA) has been updating our understanding of retinal and choroidal diseases. Due to its depth-resolved data acquiring capabilities, an unrivalled and non-invasive visualization of the retinal and choroidal vasculature became possible. Still, the clinical applications of OCTA in chorioretinal dystrophies remain largely unexplored field. The purpose of this study was to qualitative and quantitatively evaluate changes in the macular vascular networks and choriocapillaries in patients with a wide range of chorioretinal dystrophies.

Methods: Prospective study including sequential patients evaluated in the retinal dystrophies clinic at Centro Hospitalar e Universitário de Coimbra. All subjects underwent a complete ophthalmologic evaluation, complemented with OCTA using the AngioVue™ OCTA system (Avanti, Optovue, USA). The Angioanalytics™ software was employed to automatically determine several vascular parameters: choroidal flow, superficial retina non-flow (foveal avascular zone) and vascular density of the foveal and parafoveal sections (superior, inferior, nasal and temporal), and the results were compared to a normative database of healthy individuals.Additionally, qualitative changes, such as the presence of microvascular abnormalities or retinal deposits, were assessed both in the angiograms and en face OCT images.

Results: Patients with retinitis pigmentosa, cone-rod dystrophies, Stargardt disease, X-linked retinoschisis, Best vitelliform dystrophy, choroideremia, central areolar dystrophy, Bietti crystalline dystrophy, retinitis punctata albescens and fundus albipunctatus were included. The OCTA findings were diverse across the wide range of pathologies and included central atrophy of the choriocapillaries in patients with Stargardt disease, choroidal shadowing due to deposits in the outer retina in cases of Retinitis Punctata Albescens, increased foveal avascular zone in cone-rod dystrophies, etc.

In the subgroup of patients with RP (n=25 eyes), we observed a statistically significant decrease of both the superficial and deep retinal plexus densities (40.91±4.81 and 46.94±5.09, respectively, p<0.001). This finding persisted when each quadrant and the fovea were evaluated independently. Exploratory analysis showed a tendency towards sparing of the foveal superficial vasculature in younger patients: the 20-39 year-old subgroup presented a vascular density similar to healthy controls (30.06±6.67 vs 31.31±4.81 respectively, p>0.05), while older patients (40-59 year-old) had a significant decrease (23.34±6.91 vs 30.31±4.30, respectively, p<0.001).

Conclusion: The highly detailed information offered by OCTA has begun to revolutionize the way ophthalmologists evaluate the retinal and choroidal vascular networks. In the near future, this technology might provide new non-invasive ways to diagnose chorioretinal dystrophies and, most importantly, detect its progression, offering patients an objective, reliable and near instantaneous method to assess their visual function.