## **Abstract 30**

## PERIPAPILLARY PACHYCHOROID SYNDROME, A DIFFERENT PATHWAY IN VENOUS REMODELLING OF THE CHOROIDAL VASCULATURE

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Peripapillary pachychoroid syndrome (PPS) is a recently described syndrome belonging to the pachychoroid disease spectrum (PDS) group.

Our aim is to demonstrate with PPS, the presence of a venous collateral circulation which joins the peripapillary choroidal venous system with the central retinal vein via the circulation of the prelaminar region.

This collateral circulation reduces venous congestion of the nasal choroid secondary to outflow obstruction through the vortex veins or the choroidopial veins.

We describe the clinical evolution of patients affected by PPS over a 5-years period.

Two factors help diagnose this collateral circulation:

evidence from the fluorescein angiography showing dilated venules at the superficial nerve fibre layer and at the superficial prelaminar region, and evidence from the OCT-A 8 mm and automated segmentation level in deep mode, demonstrating congested vessels and increased vascular flow in the temporal juxtapapillary region.

These two factors become further evident in the more advanced stages of the disease.

It is also our intention to demonstrate with a retrospective analysis, the stages of development and possible spontaneous regression of the illness.

The increased outflow resistance via the vortex veins, as observed in patients affected by PDS, is the cause of congestion of the choroid and related chorioretinal manifestations.

Vortex veins anastomosis in the watershed zones develops to compensate stasis.

We believe that in the PPS cases we observed, and supposedly in each case of PPS, the increased venous choroidopial outflow resistance at the level of the nasal macular choroid determines the formation of a collateral circulation which develops between the peripapillary choroid and the central retinal vein, passing through the prelaminar region.

Furthermore, this study highlights how stages of disease activity can quickly be followed by major structural changes in the outer retina.

This suggests that treatment should not be delayed if the disease involves the central retina.

- [1] Phasukkijwatana N, Freund KB, Dolz-Marco R, Al-Sheikh M, Keane PA, Egan CA, Randhawa S, Stewart JM, Liu Q, Hunyor AP, Kreiger A, Nagiel A, Lalane R, Rahimi M, Lee WK, Jampol LM, Sarraf D. PERIPAPILLARY PACHYCHOROID SYNDROME. Retina. 2018 Sep;38(9):1652-1667. doi: 10.1097/IAE.000000000001907. PMID: 29135799.
- [2] Jane M. Olver Angioarchitecture of the Human Optic Nerve and Perioptic Area In: I.N.C. Innovation-News-Communication (ed.) Vascular system of the optic nerve and perioptic area. Roma 1998 p. 35-36
- [3] Ducournau D. Les artères ciliaires postérieures courtes para-optiques. Une entité anatomo-clinique [Short para-optic posterior ciliary arteries. An anatomo-clinical entity]. Bull Soc Ophtalmol Fr. 1982

Dec;82(12):1527. French. PMID: 7184651.

- [4] Kishi S, Matsumoto H. A new insight into pachychoroid diseases: Remodelling of choroidal vasculature. Graefes Arch Clin Exp Ophthalmol. 2022 Nov;260(11):3405-3417. doi: 10.1007/s00417-022-05687-6. Epub 2022 May 16. PMID: 35575932; PMCID: PMC9581833.
- [5] Xu D, Garg E, Lee K, Sakurada Y, Amphornphruet A, Phasukkijwatana N, Liakopoulos S, Pautler SE, Kreiger AE, Yzer S, Lee WK, Sadda S, Freund KB, Sarraf D. Long-term visual and anatomic outcomes of patients with peripapillary pachychoroid syndrome. Br J Ophthalmol. 2022 Apr;106(4):576-581. doi: 10.1136/bjophthalmol-2019-315550. Epub 2020 Dec 21. PMID: 33355149.