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Congenital anomalies of the optic nerve head – review

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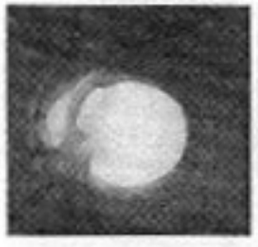
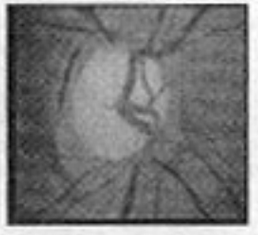
Abstract

Purpose: To show the topographic features and specific measurements of the different optic nerve head congenital anomalies. *Material:* Confocal tomography was performed to the following optic nerve head congenital anomalies: Morning Glory Syndrome. Optic Disc Coloboma, Peripapillary Staphylomas, Megalopapilla, Optic Disc Pit, Tilted Disc Syndrome and Pseudopapilla. *Methods:* Examinations were performed with the Heidelberg Retina Tomograph (long wave: 680 nm), using the new standard reference plane. Each eye was also examined with fundus biomicroscopy. *Results:* The examinations with confocal tomography show great differences and specific features in each anomaly. Topographic data and stereometric measurements are showed for each case. *Conclusion:* Confocal tomography is able to demonstrate and separate the different optic nerve head anomalies, and the measure O. N. H. parameters.

MEGALOPAPILLA

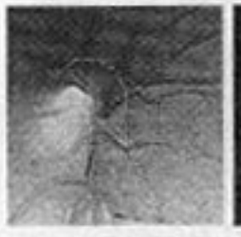


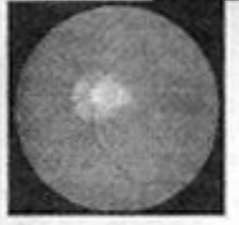



TYPE I :
- Surface > 2 .5 mm2.
- Normal configuration / higher cup/disc ratio
- Pale neuroretinal rim
- Bilateral

TYPE II :
- Surface > 2 .5 mm2.
- SUPERIOR DECENTERED CUPPING.
- Bigger NRR at the inferior part.
- Unilateral

OPTIC DISC PITS



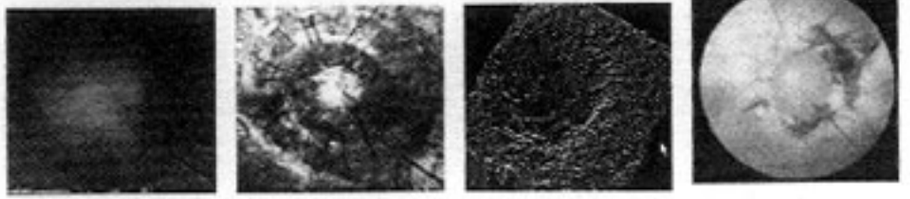



OPHTHALMOSCOPY
- Round or oval depression, grey, white or yellow.
- Temporal location.
- Peripapillary pigmentary changes.
- One or two cilioretinal arteries start at the pits.
- Generally unilateral, (15 % bilateral).

VISUAL FIELD
- The defects correlate with the pit's position.
- Arcuate scotoma, central scotoma.

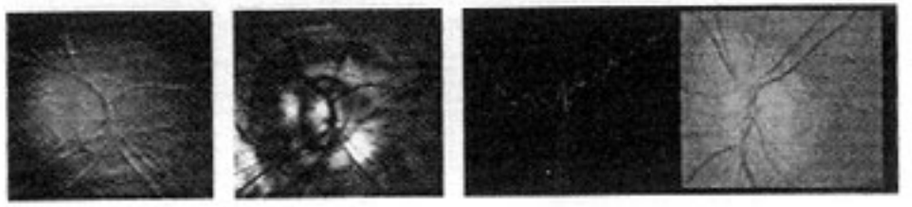
RETINAL DETACHMENT
- The macular serous detachment is related to the pit.
- Macular hole, retinal detachment surrounding it.

MORNING GLORY



- Increased number of vessels and difficult to distinguish
- Veins from arteries.
- White central veil (glia).
- Macular capture / retinal detachment, small holes near the O.N.
- Communicacion between vitreous body and subarachnoid space.

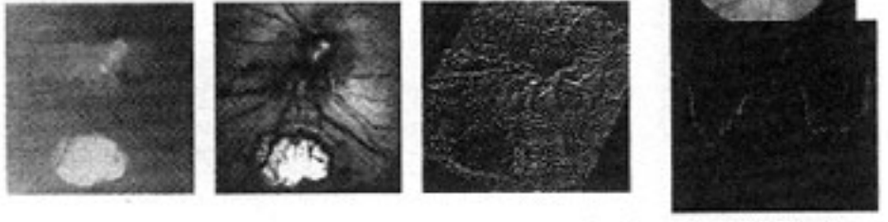
PERIPAPILLARY STAPHYLOMA



OPHTHALMOSCOPY:
- Deep cupping surrounding the O.N.H. (specially inferior) Surrounded by a pigment hale, (= M.Glory). There is no central glical white veil. Unfrequent / Unilateral

VISUAL ACUITY :
- Very reduced, though normal sometimes.
- Usually associated with iris, retinal and ciliary body colobomas.

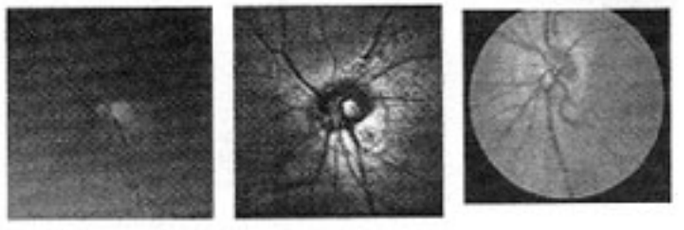
OPTIC NERVE COLOBOMAS



- Abnormal coaptation of the proximal part, optical slit.
- Big O.N., definite edges, white, deep cupping.
- CUP DECENTERED TOWARDS THE INFERIOR PART.
- Absent NRR at the inferior part.
- Inferior extension : "CHOROIDAL COLOBOMA" and/or iris coloboma.

- Posterior pole cupping (= M. Glory),
- bilateral (= M. Glory)
- The V.A. depends on the integrity of the papillo-macular bundle.
- Serous macular detachment / Normal retinal vessels

TILTED DISC SYNDROME



OPHTHALMOSCOPY
- Superonasal overelevated O.N.
- Inferonasal O.N. with posterior displacement.
- Oblique major axis.
- Situs Inversus of the papillary vessels..
- Congenital inferonasal conus.
- Retinal epithelium and inferonasal choroid thinning.
- Inferonasal albinism

PSEUDOPAPILLA

HERVOUET, 1958 : HYPOTHESIS: IT IS FEASIBLE FOR A DOUBLE OPTIC NERVE TO DEVELOP IF THERE IS A DOUBLING OF THE OPTICAL PEDICULUM, WITH THE DICHOTOMIZATION OF THE HYALOID ARTERY.

OCULAR MANIFESTATIONS

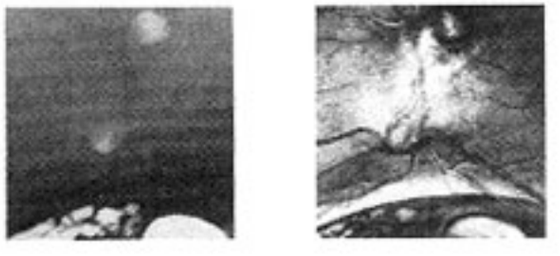
THERE ARE 2 CLINICO-OPHTHALMOSCOPIC FORMS:

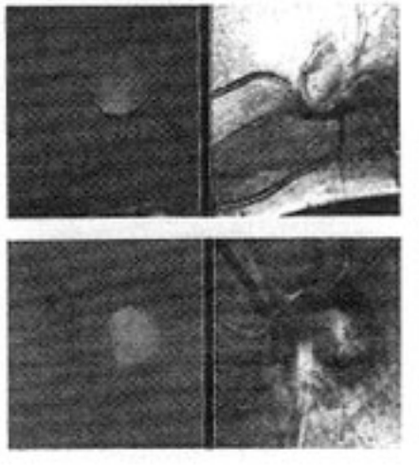
A) OPTIC DISCS JOINED BY THE VERTICAL MERIDIAN: where the supernumeric optic disc joins the inferior part of the main optic disc. Vascularization is shared.

B) OPTIC DISC DOUBLING: the requirements are :
- existence of a double optic disc contour.
- existence of double vascularization of a central type.
- existence of two joined discs without pigmentary elements between them.

C) VASCULAR PSEUDOPAPILLA: there is no actual second optic disc, but just a colobomatous scar through which retinal veins enter and choroidal arteries emerge. *

VASCULAR PSEUDOPAPILLA





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