

New methods to predict visual acuity in children with Optic Nerve Hypoplasia (ONH)

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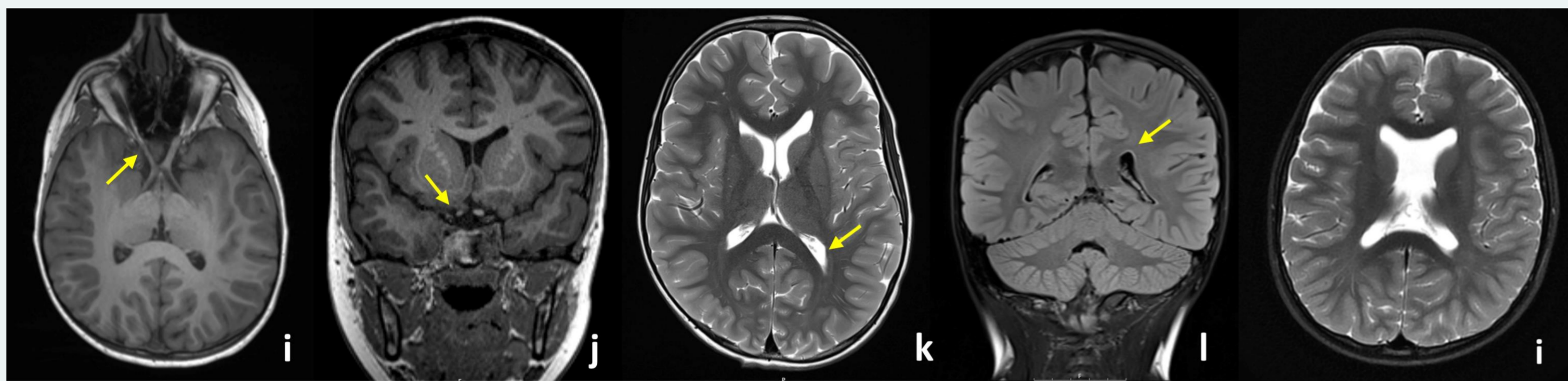
Conclusions:

- Optical Coherence Tomography (OCT) can facilitate the diagnosis of ONH.
- A thin peripapillary Retinal Nerve Fiber layer (RNFL) and/or macular ganglion cell complex (GCC) helps to confirm the diagnosis of ONH.
- Foveal hypoplasia is another biomarker that if present can aid in the diagnosis of ONH
- GCC and pRNFL thinning can indicate location and severity of visual field defects.

Introduction

Optic nerve hypoplasia (ONH) is the most common congenital optic nerve anomaly often associated with endocrinopathies, developmental delay and brain malformations.

Thin optic nerve on the right side Periventricular leukomalacia Absent septum pellucidum



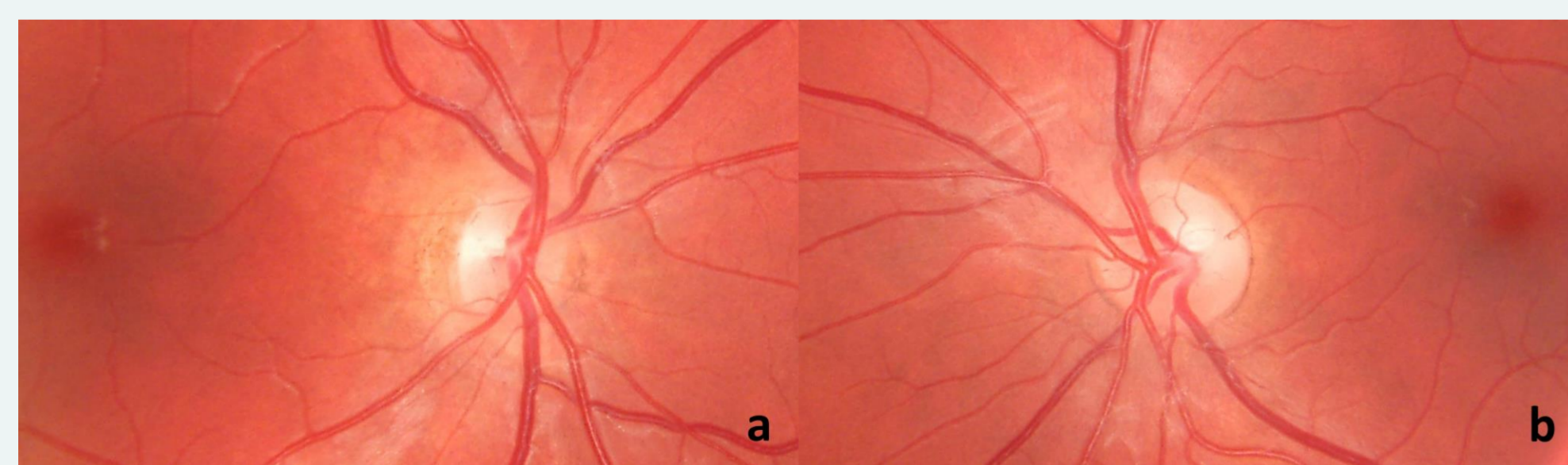
Clinical diagnosis of ONH

Fundus examination

ONH is characterized by the tetrad of :

1. small and pale optic disc
2. peripapillary “double-ring sign”
3. thinning of the nerve fibre layer
4. Torturous or straight blood vessels

None of these is pathognomonic

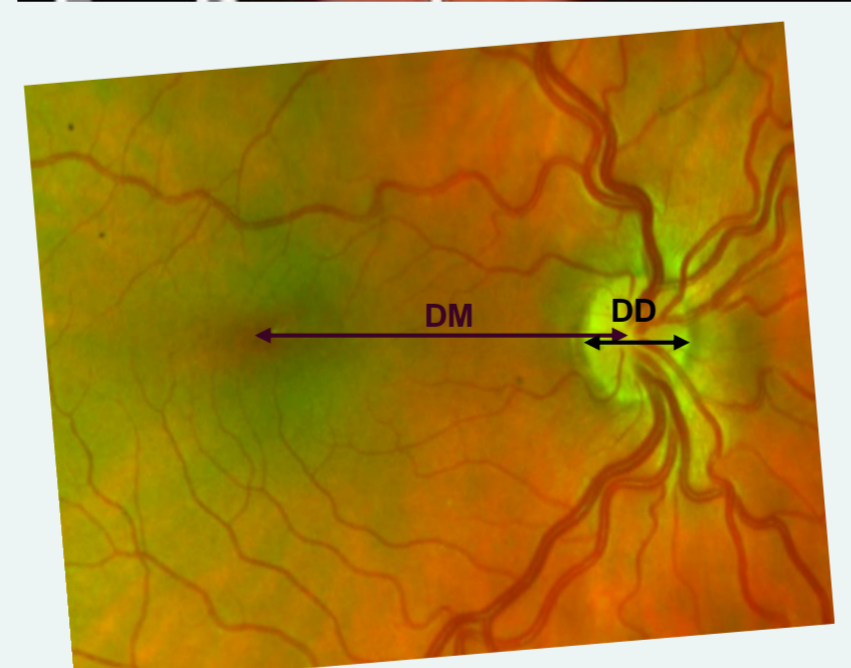
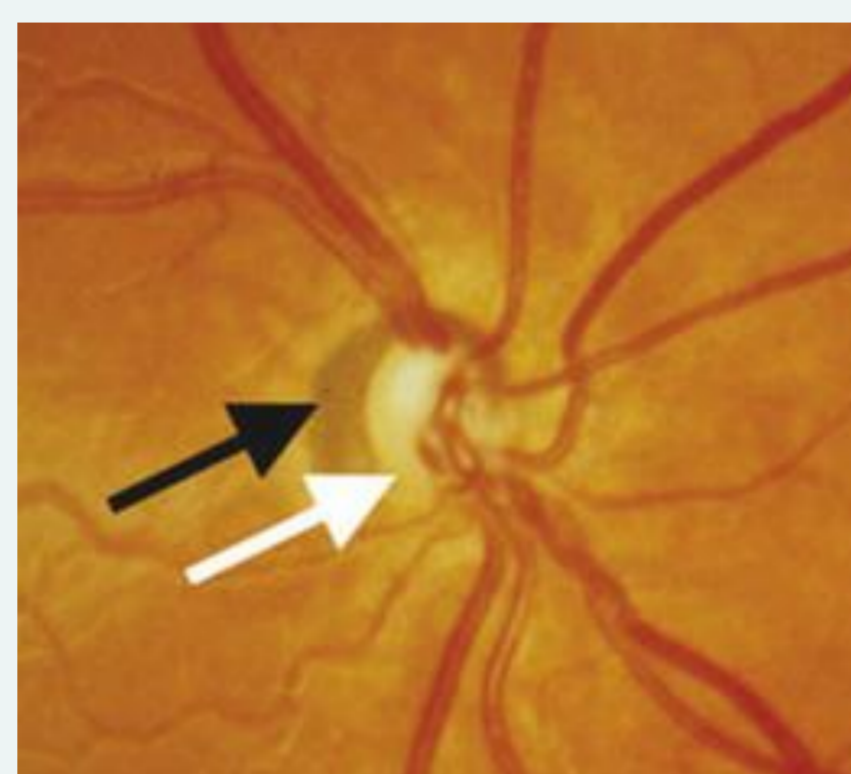


Fundus photography and Zeki's ratio

The disc-macula distance to disc diameter ratio (DM/DD) is increased in eyes with ONH (>3)

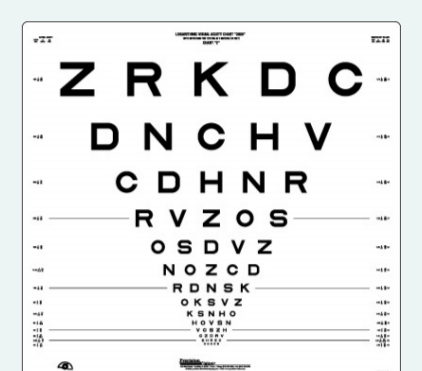
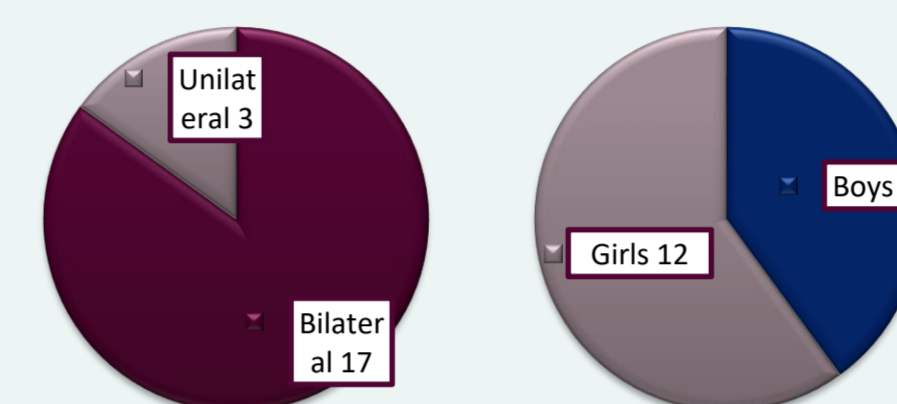
The DM is calculated by adding the vertical and transverse disc diameters and then dividing by 2. By adding half of the DD to the distance from the temporal margin of the optic disc to the fovea we obtain the 'disc-macula' distance (DM).

(Zeki et al in 1991)



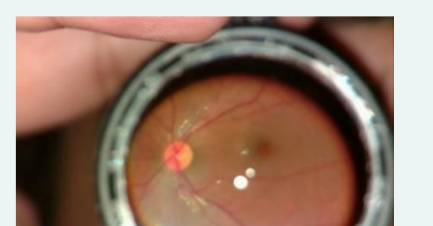
Material & Methods

- Cross-sectional cohort study
- Patients 2-18 years old with ONH in Stockholm 2020-2021
- 37 eyes from 20 patients with ONH



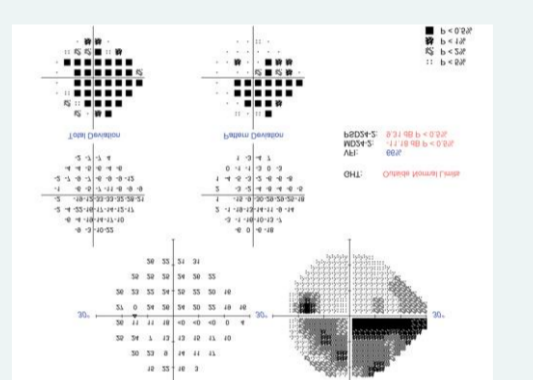
Ophthalmological assessments

- ✓ BCVA, ocular alignment, and tonometry
- ✓ Cycloplegic refraction
- ✓ Fundus examination and photography
- ✓ SS-OCT of the disc and macula
- ✓ Visual fields

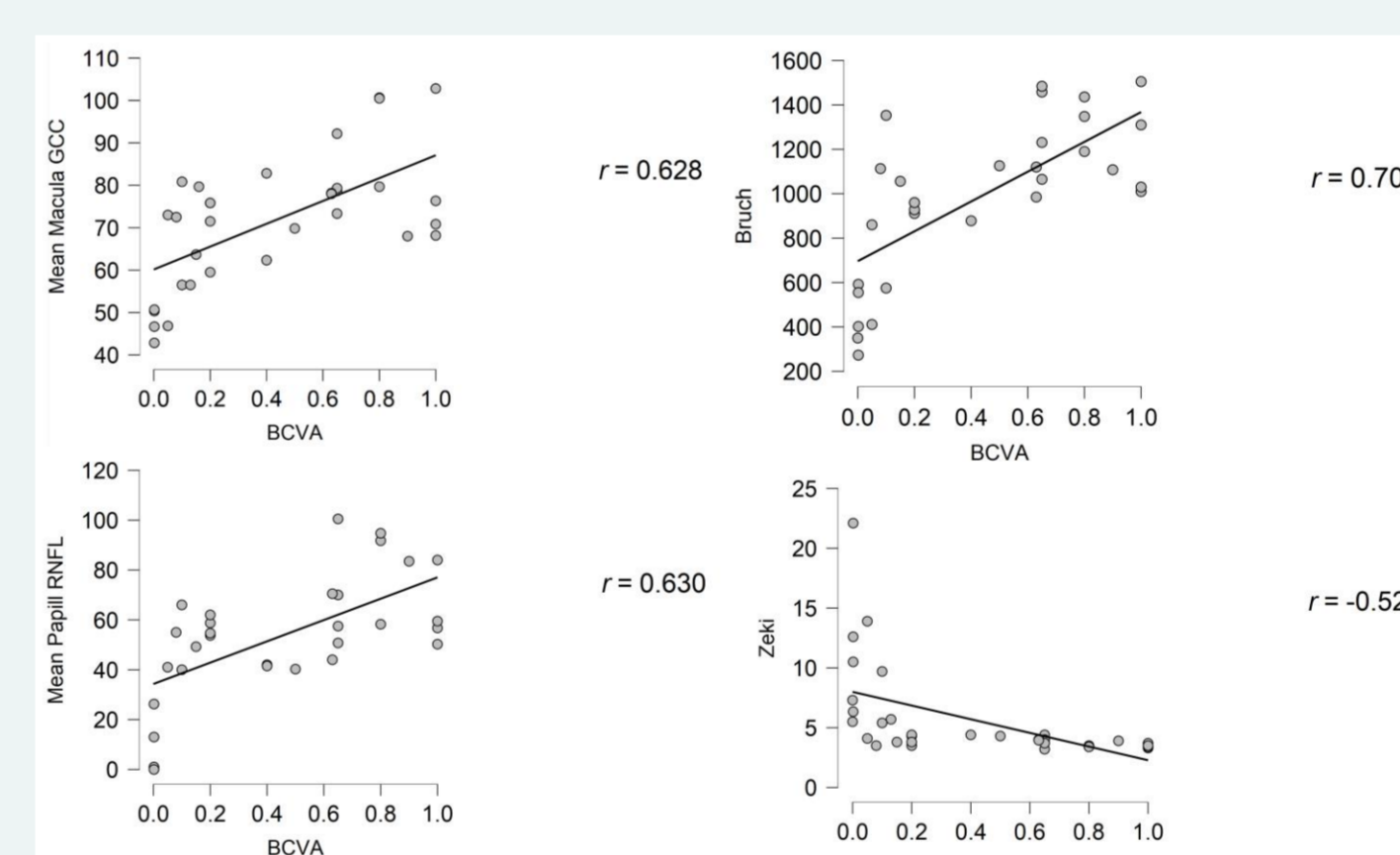


Reference data

- Healthy children aged 5-25 years born in term
- No ophthalmological, neurological problems
- Refractive errors between -6 D to +6 D
- No anisometropia, amblyopia or strabismus

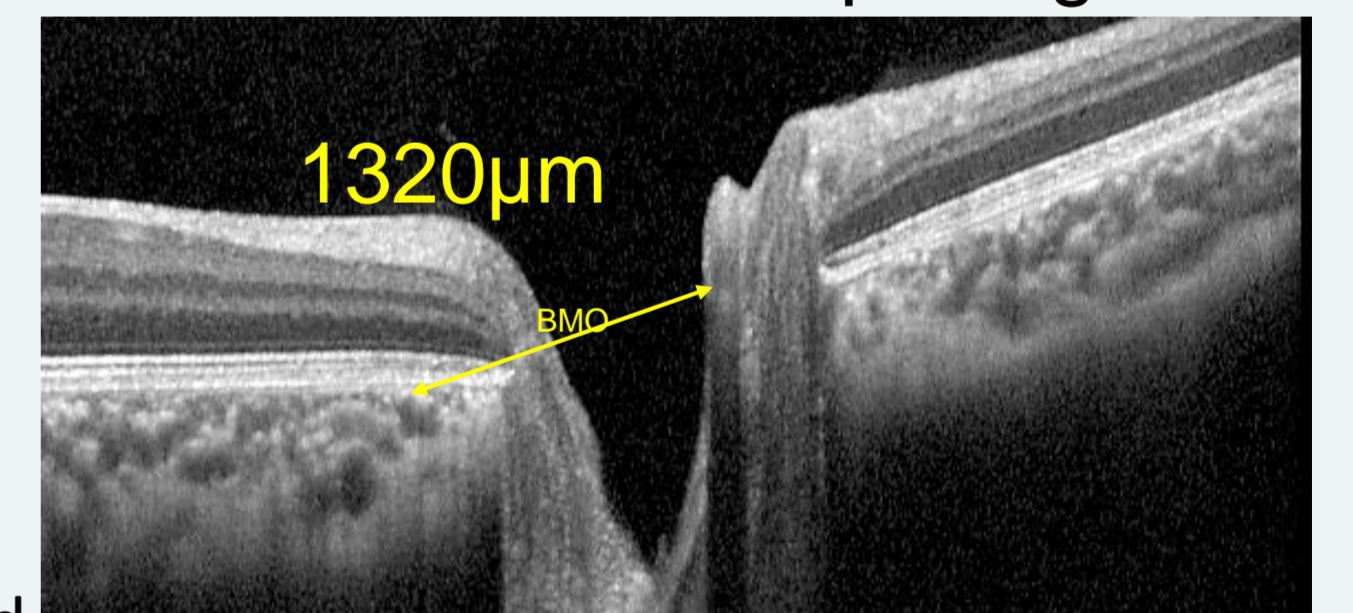


Results



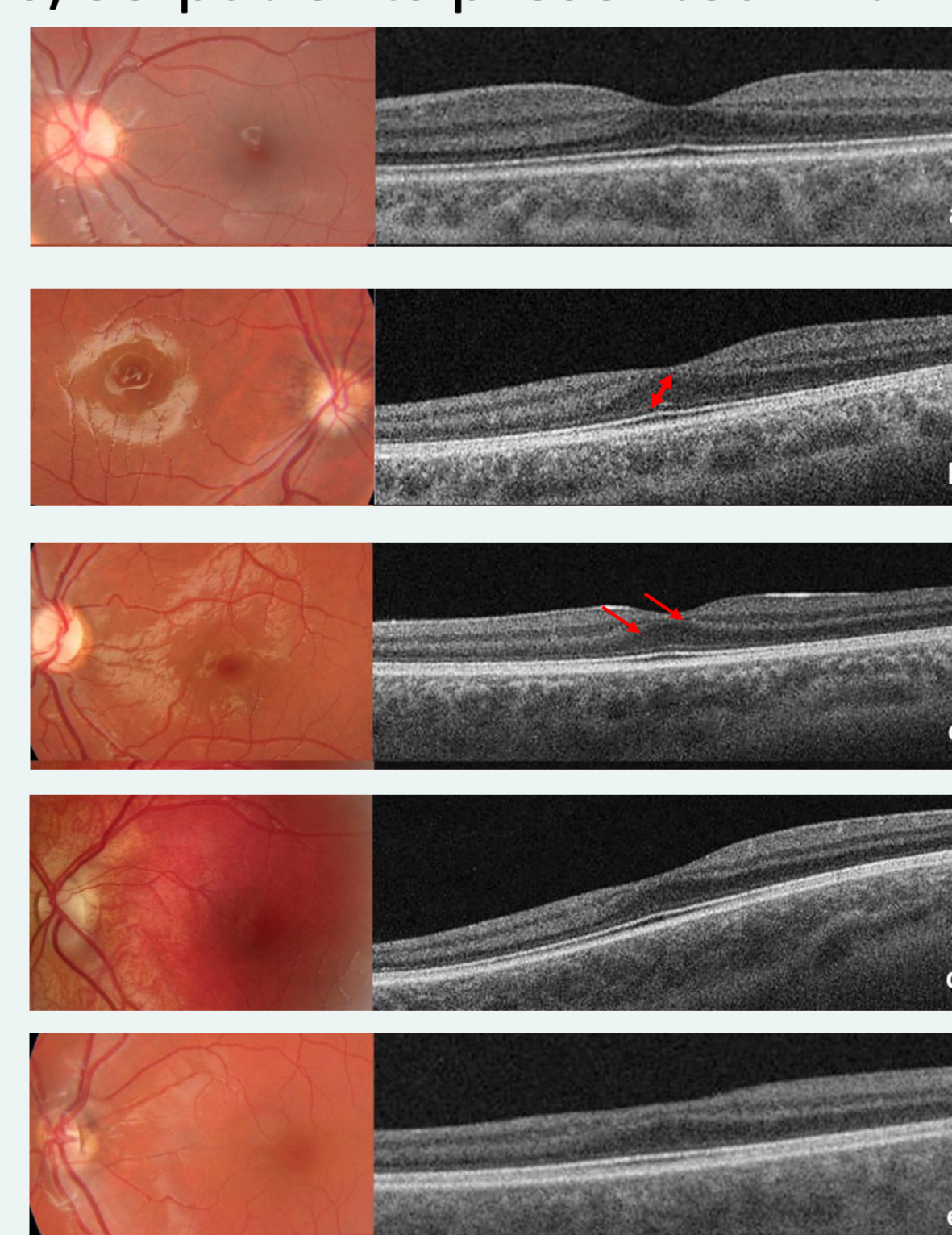
Sensitivity 74,6% Specificity 97,9%

Bruchs Membrane Opening



Moderate to strong correlation of BCVA with the pRNFL, the mGCC, Bruch's membrane opening and Zeki's ratio.

10/35 patients presented with a foveal hypoplasia



- Normal fovea (a).
- Subnormal fovea" with a shallow foveal pit because of the ganglion cell complex thinning in the parafoveal area but without continuation of the inner retinal layers under centre (b).
- Six eyes had FH grade 1 with a continuation of the inner retinal layers through the fovea and a shallow foveal pit (c).
- One eye had FH grade 2 without an identifiable foveal pit but with elongation of the photoreceptors under centre (d).
- Three eyes had FH grade 3 without any photoreceptor outer segment (OS) lengthening detected under centre but with thickening of the outer nuclear layer that defined the location of the fovea (e).



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