



Limited Vision Attributed to an Congenital Optic Nerve Anomaly

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Introduction:

In two unrelated patients, their vision is limited to CF by a congenital optic nerve anomaly. (Figure 1) Their fellow eyes are normal. Their personal and family histories are unremarkable.

Differential Diagnosis:

These clinical photos are most consistent with the presentation of morning glory syndrome, however the differential diagnosis can overlap with other disc abnormalities, the most common of which is optic disc colobomas. Both conditions occur due to incomplete closure of the posterior aspect of the fetal fissure. It is important to differentiate the two because of different systemic associations. Optic colobomas can be associated with CHARGE syndrome (coloboma of the eye, heart defects, choanal atresia, growth retardation, genitourinary abnormalities and ear abnormalities) and thus requires different workup.¹ Morning glory syndrome has different associations discussed below. Optic pits can also present with peripapillary excavations but lack the straightening of the retinal vasculature.

Clinical Findings:

Morning Glory syndrome is a rare sporadic

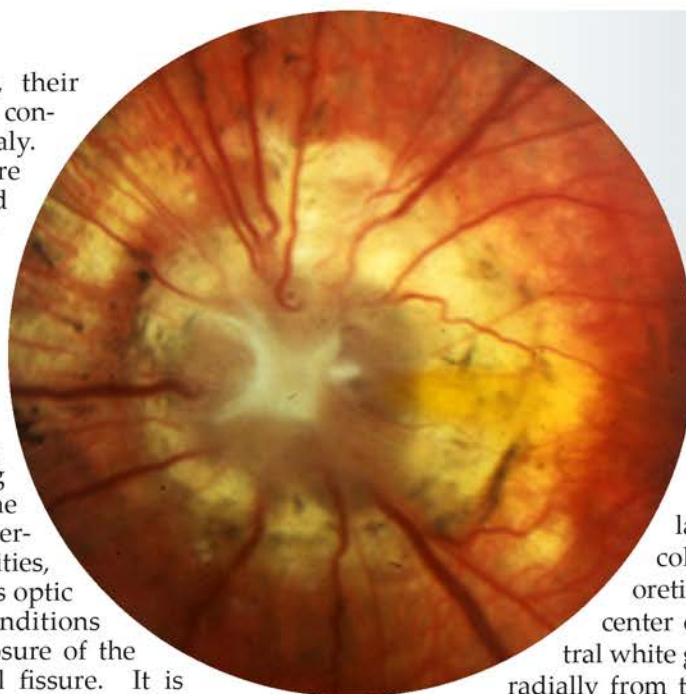
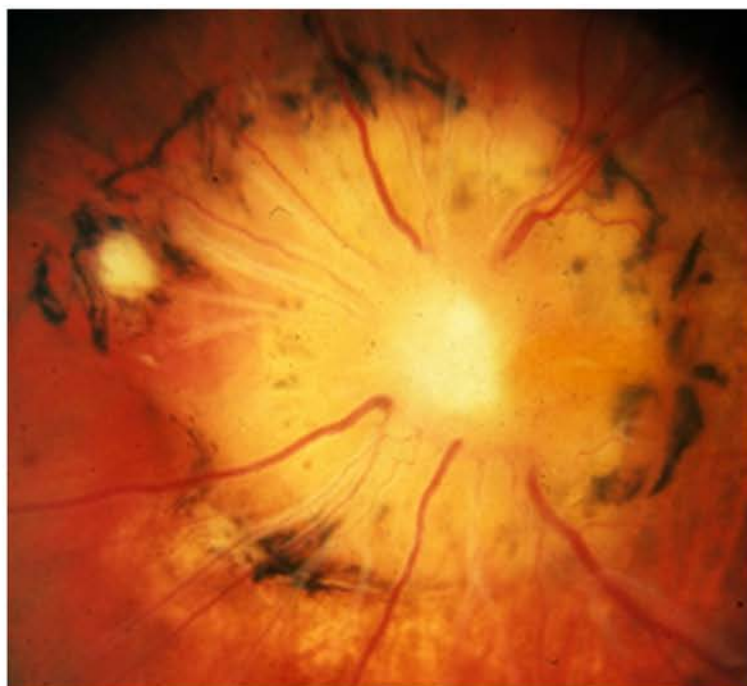


Figure 1 (A,B) – Photographs of morning glory discs demonstrating large peripapillary excavations with pigment, radial vasculature, and central white tufts.



congenital malformation first described in 1970 by Kindler. It is commonly described as a funnel-shaped excavation centered around the optic disc.² The optic disc generally appears large, and orange or pink in color with peripapillary chorioretinal pigmentary changes. The center of the disc can contain a central white glial tuft. Blood vessels extend radially from the disk instead of the classic curved branching patterns. Kindler named this syndrome morning glory due to its resemblance to the morning glory flower. (Figure 2) When the macula is involved, it is described as “macular capture”.

Visual acuity is generally poor but can range from 20/20 to count fingers vision depending on the extent of the anomaly and the development of amblyopia. Visual field defects are commonly found on testing. The condition is generally unilateral, which can result in an afferent pupillary defect.

Management and Treatment:

As the condition is congenital, vision must be optimized and monitored carefully to prevent amblyopia. Most importantly, these disc abnormalities can have systemic associations which can be dangerous if not found early. Brain imaging should be obtained to rule out Moyamoya disease, basal encephaloceles, and other vascular abnormalities.³



Figure 2. A photograph of the morning glory flower. Note the resemblance of the morning glory disc central white tuft and radial lines to those in the flower.

This beauty was grown in Dr. Krummenacher's garden expressly for this Case of the Month.

Regular exams should be performed to monitor for retinal detachments. OCT imaging has demonstrated abnormal communications between the sub-arachnoid space and the subretinal space.⁴ Additionally, tissue stretching around the peripapillary conus can result in slit-like retinal breaks that can lead to retinal detachments that may be addressed with vitrectomy techniques.⁵

References:

1. Heidary G. Congenital optic nerve anomalies and hereditary optic neuropathies. *J Pediatr Genet.* 2014 Dec;3(4):271-8.
2. Ceynowa DJ, Wickström R, Olsson M, Ek U, Eriksson U, Wiberg MK, Fahnehjelm KT. Morning glory

disc anomaly in childhood - a population-based study. *Acta Ophthalmol.* 2015 Nov;93(7):626-34.

3. Fei P, Zhang Q, Li J, Zhao P. Clinical characteristics and treatment of 22 eyes of morning glory syndrome associated with persistent hyperplastic primary vitreous. *Br J Ophthalmol.* 2013 Oct;97(10):1262-7.
4. Inoue M. Retinal complications associated with congenital optic disc anomalies determined by swept source optical coherence tomography. *Taiwan J Ophthalmol.* 2016 Jan-Mar;6(1):8-14.
5. Zhang Y1, Ou H2, Zhu T3. Surgical treatment for the proliferative retinal detachment associated with macular hole in the morning glory syndrome. *Eye Sci.* 2013 Mar;28(1):7-10.

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