STRAATSMA SYNDROME

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Triad of ipsilateral retinal nerve fiber myelination, myopia and amblyopia, with or without strabismus. The 45 degree fundus photos below show extensive unilateral myelination of the right fundus and a normal left fundus of a nine year old Chinese girl.

Refractive error measured -12.00DS OD and Plano OS with best corrected spectacle acuities of 20/400 and 20/15 in the right and left eyes respectively. There was no evidence of strabismus or nystagmus.

The success of case studies investigating the efficacy of occlusion therapy ranges from “unexpected good results” to “refractory.”2,3 Macular appearance may be a predictor of the success of occlusion therapy in these cases.4

The central OCT (top) in this case revealed normal macular architecture. The OCT outside the macula (bottom) reveals a hyper-reflective and thickened retinal nerve fiber layer.