

Paradoxical thinning of the retinal nerve fiber layer after reversal of cupping: A case report of primary infantile glaucoma

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The circumpapillary retinal nerve fiber layer (RNFL) thickness was assessed by spectral domain optical coherent tomography (SD-OCT) before and after surgical reduction of intraocular pressure in an eye with primary infantile glaucoma. In this case, a postoperative reduction of cupping and a subsequent increase in neuroretinal rim area is associated with a paradoxical thinning of the RNFL. This is the first-known characterization of cupping reversal using SD-OCT.

Key words: Optical coherence tomography, pediatric glaucoma, reversal of cupping, retinal nerve fiber layer

Reversal of cupping is a common finding after surgical decompression of a hypertensive eye. In this case of primary infantile glaucoma treated with trabeculotomy, the successful surgical reduction of intraocular pressure was associated with marked reversal of cupping and a decrease of the circumpapillary retinal nerve fiber layer (RNFL) thickness as measured by spectral domain optical coherent tomography (SD-OCT). This is the first-known characterization of cupping reversal using SD-OCT.

Case report

A 5-year-old girl with no significant past ocular or systemic history presented to the (Bascom Palmer Eye Institute) complaining of decreased vision in the right eye (OD), first noted 2 weeks prior to presentation. On examination, best-corrected visual acuity was light perception OD and 20/30 left eye (OS). The eyes were orthophoric without extraocular motility limitations. Pupils were found to be asymmetrically

reactive to light, with a marked right afferent papillary defect. Intraocular pressure (IOP) by Tono-Pen (Reichert Inc., Depew, NY) was 35 mmHg right eye and 22 mmHg left eye. Externally, an asymmetry of palpebral fissures was noted. The corneal diameters were 13.5 mm right eye and 12 mm left eye, and

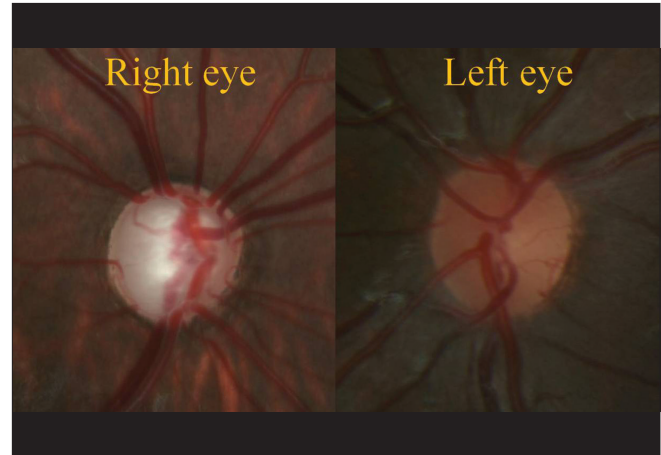


Figure 1: Optic disk photographs. Upon presentation, the right optic disk with near-complete cupping, while the left optic disk is normal in appearance

Cite this article as: Chang TC, Grajewski AL. Paradoxical thinning of the retinal nerve fiber layer after reversal of cupping: A case report of primary infantile glaucoma. *Indian J Ophthalmol* 2016;64:690-2.

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/0301-4738.97092

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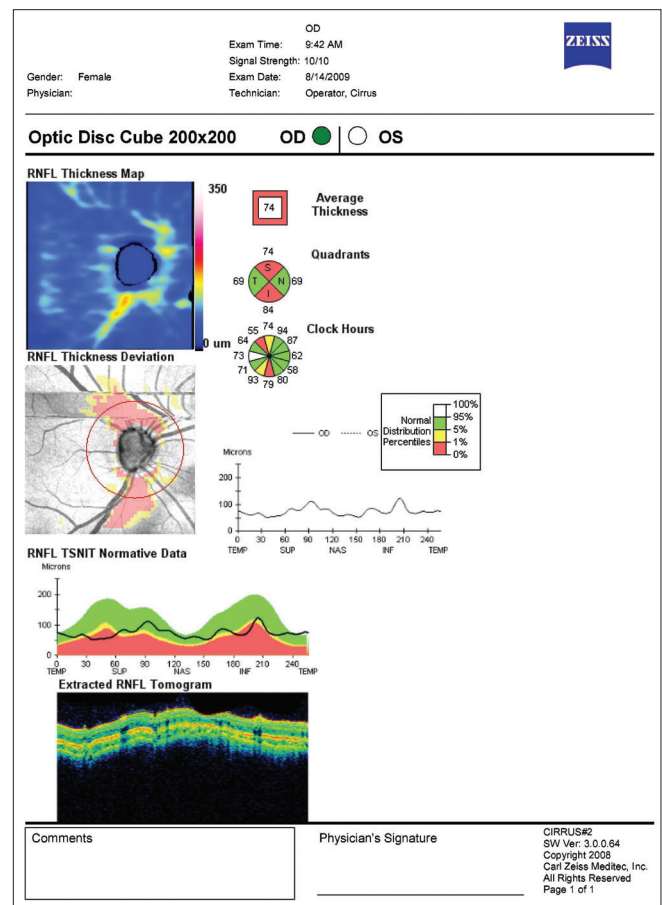


Figure 2: Spectral domain optical coherent tomography (SD-OCT) studies of circumpapillary retinal nerve fiber layers (RNFL) of the right eye upon presentation

were noted to be clear bilaterally with an intact Descemet’s membrane. Gonioscopy revealed grade IV angles bilaterally with indistinct features without an observed Barkan’s membrane. Fundus examination revealed significant optic disk cupping in the right eye and was within normal limits in the left eye [Fig. 1]. Measurement of the circumpapillary retinal nerve fiber layers (RNFL) by SD-OCT (Carl Zeiss Meditec, Inc., Dublin, CA) of the right eye revealed superior and inferior RNFL thinning [Fig. 2].

An examination under anesthesia was performed, during which additional IOP measurements by Tono-Pen under both light and deep sedation. The IOP was measured to be 49 mmHg and 42 mg Hg OD, and 32 mmHg and 24 mmHg OS respectively. The axial length by A-scan ultrasound is 24.3 mm right eye and 21.3 mm left eye. At the same session, a trabeculotomy was performed in the right eye, using a Harms trabeculotomy probe spanning 120° (10 to 2 o’clock meridians) superiorly.

On the first postoperative day, best-corrected visual acuity OD remained light perception, and IOP by Tono-Pen was 18 mmHg OD and 28 mmHg OS. Medical therapy was initiated with topical dorzolamide OS. Three months after the trabeculotomy, IOP was 17 mmHg OD and 24 mmHg OS by Goldmann applanation. Repeat disk photography demonstrated marked reversal of cupping and persistent pallor of the right optic disk [Fig. 3]. Measurement of the right RNFL by SD-OCT revealed paradoxical decrease of overall RNFL thickness compared to preoperative measurements [Fig. 4]. On the seventh postoperative month, IOP is 14 mmHg OD and 22 mmHg OS (on dorzolamide BID OS), and the optic disk appearance and SD-OCT RNFL measurements remain largely unchanged from the previous exam. Visual acuity remained light-perception in the right eye, and 20/30 OS.

Discussion

Primary infantile glaucoma has an incidence of approximately 1/10,000 live births, and contributes to approximately 22.2% of all childhood glaucoma cases.^[1] It is typically defined as ocular hypertension and associated optic neuropathy occurring

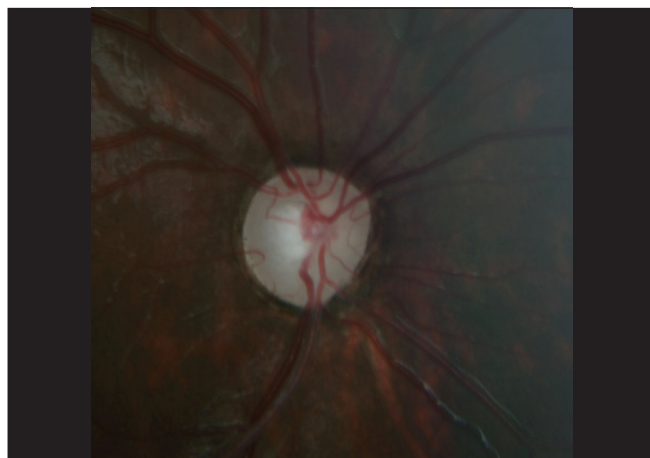


Figure 3: Photograph of right optic disk 3 months after successful trabeculotomy

prior to the age of 3, often accompanied with buphthalmic changes due to the elastic sclera of the infant eye.^[1] The definitive first-line therapy is angle surgery, with equal success found between trabeculotomy and goniotomy in one series.^[2] Circumferential trabeculotomy by filament cannulation of Schlemm’s canal may have an advantage over goniotomy as a primary procedure.^[3]

Reversal of cupping is a well-described phenomenon after incisional glaucoma therapy, occurring in approximately 6.6% of adults. This reversal is associated with a lower postoperative IOP, but not with improvement in either visual acuity or visual field.^[4] Prior studies have correlated decreased RNFL thickness with increased cup/disk ratio in children,^[5] and suggest no significant change in RNFL with lowering of IOP in adults.^[6] The SD-OCT characteristics of RNFL in reversal of cupping have not been previously described to date. Our case demonstrates a paradoxical decrease of RNFL thickness with photographically documented reversal of cupping. Although interexamination variability may contribute to this observation, this is less likely given that SD-OCT has a variability of 5.8-8.1 microns,^[7] smaller than the magnitude of observed thinning in our case (14 microns). We propose that in a hypertensive eye, blockage of axoplasmic flow at the scleral edge may contribute to axonal swelling and a greater

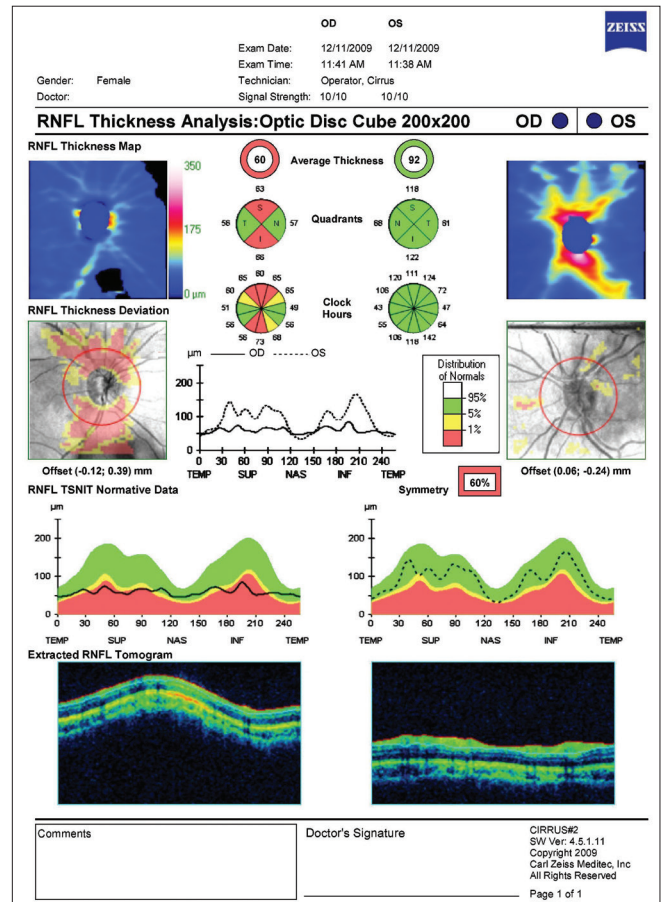


Figure 4: SD-OCT examination of the right optic disk on the third postoperative month

circumpapillary RNFL thickness. Decompression of the eye relieves this blockage at the scleral edge, thereby allowing more downstream axoplasmic flow to proceed, possibly explaining the thinning of circumpapillary RNFL measurements with the apparent decrease in cupping in this case. On the other hand, this may simply represent continued glaucomatous damage to RNFL despite well-controlled IOP. A further study of this phenomenon may be warranted to help elucidate the mechanisms of injury of glaucomatous damage in pediatric eyes.

Literature search

PubMed databases from 1975 to 2010 were searched for keywords *reversal of cupping*, *optical coherent tomography*, and *retinal nerve fiber layer*. The non-English literature was not included in our search.

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