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BRIEF COMMUNICATION

Anterior displacement of Bruch's membrane: a useful sign for intracranial hypertension in craniosynostosis

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Craniosynostosis is characterised by the premature fusion of the cranial sutures. Craniosynostosis is often associated with intracranial hypertension (IH), which can damage vision and neurocognitive function if unaddressed. Direct measurement of intracranial pressure (ICP) is invasive and involves hospital admission, surgery and associated risks. By contrast, optical coherence tomography (OCT) has demonstrated good potential as a surrogate, non-invasive measure of ICP in children with craniosynostosis [1, 2]. However, it is unclear how IH may affect Bruch's membrane in children with craniosynostosis, a question unaddressed in a recent systematic review [2]. Here, we assessed the configuration of Bruch's membrane in this target population.

This was a cross-sectional study of children with craniosynostosis recruited from the Craniofacial Surgery Center at the Children's Hospital of Philadelphia (CHOP), analysed in partnership with the University of Leicester and Great Ormond Street Hospital for Children, London. Inclusion criteria were as follows: age under 18 years, clinical/genetic diagnosis of craniosynostosis (syndromic/non-syndromic), direct on-table ICP measurements and OCT images of the optic nerve head (ONH) both obtained according to CHOP's published protocol, with IH defined as > 15 mmHg [1]. Bruch's membrane was graded according to Sibony et al. [3]: V-flat, W-shape, S-shape and D-shape (dome-shape). To assess intergrader reliability, SRR (trained OCT grader) and JDB (untrained OCT grader) independently graded Bruch's membrane configurations, masked to all other clinical data.

Fifty children met inclusion criteria: predominantly male ($n = 31$, 62%) and non-syndromic ($n = 37$, 74%). Mean age at OCT was 40 months (standard deviation: 36 months). Bilateral OCTs of adequate quality were available in 41 patients (82%) and unilateral OCTs in 9 patients (18%). Twenty-five patients had IH (prevalence: 50%). Intergrader reliability was 100%, following one query from JDB, resolved by SRR, regarding a blood vessel in close proximity to Bruch's membrane. Five patients (10%) demonstrated anterior displacement of Bruch's membrane (Fig. 1). In this cohort, we observed bilateral dome-shape, $n = 1$; only left eye scanned, dome-shape, $n = 1$; right eye dome-shape and left eye S-shape, $n = 1$; right eye V-flat and left eye W-shape, $n = 1$; right eye S-shape and left eye dome-shape, $n = 1$. Respective ICPs were 22 mmHg, 17 mmHg, 23 mmHg, 15 mmHg and 3 mmHg—the latter had long-standing optic atrophy. All other patients demonstrated bilateral V-flat configuration.

Our findings suggest that the anterior displacement of Bruch's membrane on OCT represents a useful sign for IH in children with craniosynostosis. This likely reflects vector forces at the level of the lamina cribrosa, pushing Bruch's membrane anteriorly.

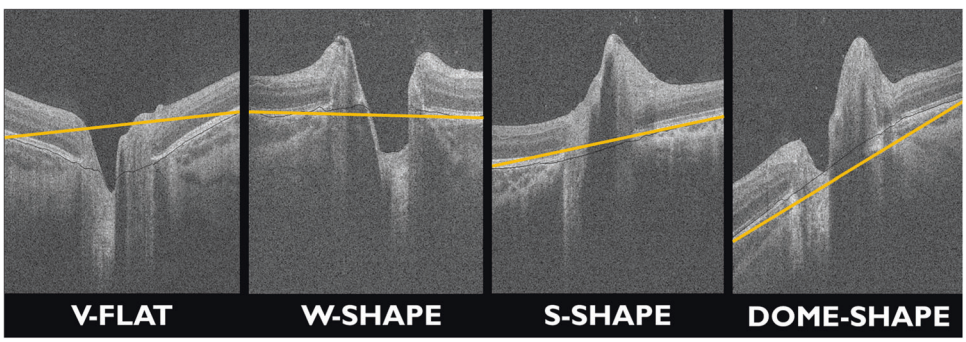

Fig. 1 Sample configurations of Bruch's membrane. The yellow reference line connects either end of Bruch's membrane (BM). V-Flat: BM lies on or below the reference line; W-shape: symmetrical anterior displacement of the inner BM margins; S-shape: concave BM curvature nasally and convex BM curvature temporally; Dome-shape: symmetrical dome-shaped anterior BM displacement.

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Interestingly, the patient whose ICP was 15 mmHg at the time of surgery had subtle W-shape developing only in the left eye (Fig. 1) – had this child not undergone surgery and the ICP risen further, this displacement may have become more pronounced. With regards to the patient with optic atrophy, although anterior displacement of Bruch's membrane is most frequently seen in IH and papilloedema, it has also been reported in nonarteritic ischaemic optic neuropathy and optic neuritis [4, 5]. Bruch's membrane configuration should ideally only be assessed on untilted OCT scans [3]. Overnight ICP monitoring was not feasible in this perioperative setting [1]. Further prospective work is needed to evaluate the optimal combination Bruch's membrane configuration with quantitative OCT measurements to maximise sensitivity and specificity for IH.

DATA AVAILABILITY

The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

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AUTHOR CONTRIBUTIONS

SRR: conceptualisation, methodology, formal analysis, writing – original draft, visualisation; JDB: data curation, formal analysis, writing – original draft, visualisation; CK: methodology, data collection, writing – review and editing; RA: conceptualisation, methodology, writing – review and editing; NUOJ: conceptualisation, writing – review and editing; FAP: conceptualisation, methodology, resources, writing – review and editing; JWS: conceptualisation, methodology, resources, writing – review and editing, obtaining funding, supervision.

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COMPETING INTERESTS

FAP reports personal fees from Leica Microsystems to run a virtual clinical symposium for hand-held optical coherence tomography, outside the submitted work. All other authors report no competing interests.

ADDITIONAL INFORMATION

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