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Diagnostic of Optic Disk Drusen in Children

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Abstract

Optic disc drusen have a compression impact on the glial tissue and blood vessels within the scleral canal which causes pathological changes in the optic nerve and retinal peripapillary, as well as causing hemodynamic instability, which, in turn, is accompanied by a reduction of visual function. Severity of functional disorders depends on the amount of drusen and their localisation [1, 2, 3, 4, 5, 6]. To-date there are no classification criteria with regard to drusen effect on the visual function and methods of treatment.

Keywords: optic disc drusen, classification, diagnostic, children

Acknowledgement: We thank the volunteers for their participation.

Introduction

Optic disc drusen are viewed as a polyetiology disease with unclear mechanism of formation, the lack of a clear diagnostic and classification criteria and effective treatments. Optic disc druse are deposits of chondroitin sulfates in the depth of the optic nerve, that are prone to progressive calcification. Their appearance is associated with impaired axonal transport [7], mitochondrial calcification, structural features of the scleral canal and vascular architectonics [8, 9, 10, 11, 12].

Since the central vision in patients with optic disc drusen are rarely reduced, their diagnosis is usually limited to ascertaining the existence of drusen (Fig. 1, 2) without functional changes in the retina and optic nerve. However, there is pronounced polymorphism of the drusen structural changes, as well as retinal and functional parameters of the visual system.

Study purpose was to detect morphological and functional criteria in changes of the visual system in children with optic disc drusen, determining severity of the pathological process; to assess their diagnostic value in predicting disease outcome and to justify diagnostic and monitoring algorithm.

Method

Clinical studies were conducted in accordance with the World Medical Association Declaration of Helsinki – *Ethical Principles for medical research involving human subjects* (1964, amended in 2000) and *Rules of clinical practice in the Russian Federation* (Order of the Health Ministry of the Russian Federation No. 266 from 19.06.2003). Parents and guardians of the surveyed children gave their consent for the diagnostic tests.

We tested a total of 23 children (46 eyes) with drusen of the optic nerve at the age from 8 to 15 years; the ratio of girls to boys was 3:2. In all cases, the process was bilateral, data from both eyes tested included in the study.

To exclude concomitant ophthalmopathology, clinical group included children with refraction from +3.0 to -3.0 dioptres, corrected visual acuity not less than 0.8. Ultrasound scans of the eyeball determined study inclusion and exclusion criteria.

The study group included children with b-scan identified drusen as hyperechogenic sites in the the optic disc area. The exclusion criteria were the expansion of perineural space [13]. Depending on the drusen topography on optic coherence tomography (OCT), patients were divided into two groups.

The first group included 15 children with drusen located on the edge of the disc. The second group included eight children with drusen located in the centre of the disc. The control group included 30 healthy children of the approximately the same age.

All patients underwent complex ophthalmological testing. Standard testing included:

- visometry,
- eye biometrics,
- ultrasound b-scan of the posterior pole and orbit.

Additionally we performed:

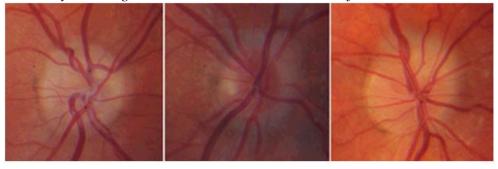
- OCT and optic coherence tomography angiography (OCTA),
- standard automated perimetry,
- visual evoked potentials registration and
- electroretinography.

Structural changes, such as degree of disc swelling, drusen localisation, retina change, were identified through the analysis of 3-D OCT images, line and frontal scans in EnFace mode. They were compared with functional changes in perimetry, electrophysiology and optic coherence tomography angiography results. Study data was statistically processed using the Mann-Whitney test (indicators were considered true at p <0,05).

Results

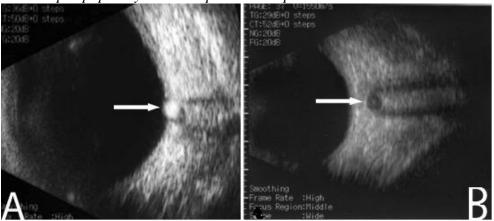
Diagnosis of superficial drusen is not hard. They are registered as small rounded sites with white or yellowish opalescence on the edge of the disc. Certain difficulties arise in diagnostic of deep seated drusen as they are not visualised ophthalmoscopically and often mimic the picture of stagnation, leading to the incorrect tactics in diagnostic and treatment (Figure 1).

Figure 1. Optic disc drusen. Ophthalmic findings. Optic disc swells in the vitreous cavity, disc edges are blurred, retinal vessels are full and curved.



At the same time, ultrasound examination of the eyeball and orbit allows to quickly make a differential diagnosis between the optic disc drusen and stagnation, which are accompanied by an expansion of perineural space (Figure 2).

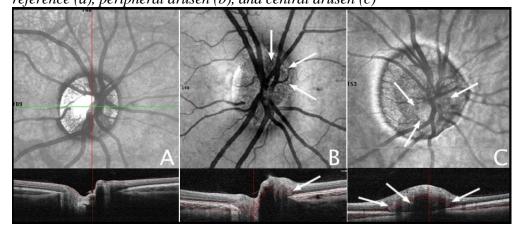
Figure 2. Optic disc drusen. Ultrasonic findings. A – scleral department, B – orbital department. Optic disc swells in the vitreous cavity. On the edge of the disc there is a hyperechoic inclusion with clear boundaries; no changes noted in peripapillary retina or perineural space



In the analysis of structural changes in the retina and optic disc according to OCT revealed swelling, expanding boundaries, changing physiological excavation of the disc, thinning layer of retinal ganglion cells.

Peripheral drusen are located on the edge of the disc beyond the scleral ring, note the decrease in the volume of physiological excavation. Central drusen are detected in the middle layers of the optic nerve head, they are in contact with a vascular bundle and are greater in size, thereby physiological excavation is absent (Figure 3).

Figure 3. Frontal (1) and linear (2) OCT scans of the optic disc at reference (a), peripheral drusen (b), and central drusen (c)



The height of the swelling of the optic disk in patients with drusen in 1.3 times higher than the rate in the control group. Statistically significant differences in both clinical groups have not been identified (see Table 1).

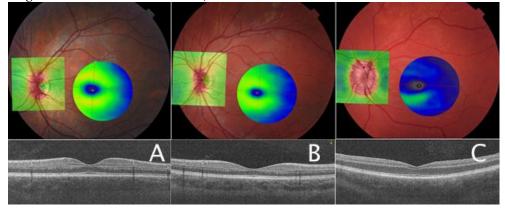
Table 1. Comparative analysis of structural and functional changes indicators of the visual system in healthy children and patients with optic disc drusen

Index		Group 1 *	Group 2 *	Group 3 *	Mann-Whitney U		
		n=30	n=16	n=30	p1-p2	р1-р3	р2-р3
Visual Acuty		$0,93\pm0,12$	$0,86\pm0,09$	$0,92\pm0,08$	0,34	0,82	0,34
Pattern-VEP	1,0 deg (M) p100 ms	107,05±14,68	113,20±13,81	104,40±3,91	0,16	0,67	0,16
	N75-p100	$17,57\pm9,82$	$10,59\pm5,18$	$24,72\pm12,03$	0,01	0,13	0,01
Oscillattory	P2 (ms)	23,35±0,44	23,28±0,31	22,95±0,55	0,18	0,06	0,18
potentials	P3 (ms)	30,86±0,81	30,61±0,66	30,28±0,81	0,42	0,12	0,42
	P4 (ms)	39,95±2,44	37,66±0,87	39,30±0,01	0,02	0,65	0,01
Photopic 3,0	a-wave (ms)	14,46±0,92	13,96±1,37	14,30±0,90	0,61	0,71	0,61
ERG (GF)	a-wave (μV)	$23,26\pm7,38$	27,38±7,87	21,35±6,45	0,15	0,56	0,15
	b (ms)	29,94±0,91	29,95±1,11	29,30±0,83	0,25	0,12	0,25
	b-wave (μV)	90,54±17,83	71,10±21,85	134,63±22,23	0,0002	0,02	0,0002
Flicker 30 Hz	P1 (ms)	60,88±1,04	60,29±1,31	61,05±1,59	0,34	0,73	0,34
ERG (GF)	N1 - P1	85,20±21,11	73,13±20,95	109,36±24,47	0,01	0,21	0,01
SAP	MS	25,04±3,34	$20,48\pm2,70$	25,03±4,48	0,04	1,00	0,04
	MD	$1,02\pm1,57$	$8,26\pm2,74$	$1,82\pm2,44$	0,0007	0,31	0,0007
	sLV	$2,19\pm0,67$	5,61±1,38	$2,40\pm1,04$	0,0005	0,53	0,0005
OCT	Inferior	417,87±106,53	450,00±74,77	371,17±15,30	0,43	0,30	0,03
Peripapillar	Superior	425,23±89,00	457,38±77,40	371,17±21,07	0,36	0,15	0,02
Thickness	Nasal	423,27±91,96	482,75±79,86	372,67±26,27	0,10	0,19	0,01
	Temporal	350,90±81,98	340,38±32,68	299,00±9,47	0,73	0,14	0,01
ONH Cube Vol.	mm ³	11,15±0,23	$11,48\pm0,18$	$10,7\pm0,2$	0,006	0,003	0,0000
GCC		95,72±13,72	83,60±10,43	105,21±2,22	0,04	0,25	0,009

Notes: * Group 1 – peripheral drusen, group 2 – central drusen, group 3 – control group.

A statistically significant reduction in the thickness of the GCC was revealed in patients with central drusen (Table 1, Figure 4 a, b). In one case, a child with giant drusen in central location (Figure 4 c) had thinning GCC up to 63 microns, which was 1.67 times lower than in control subjects.

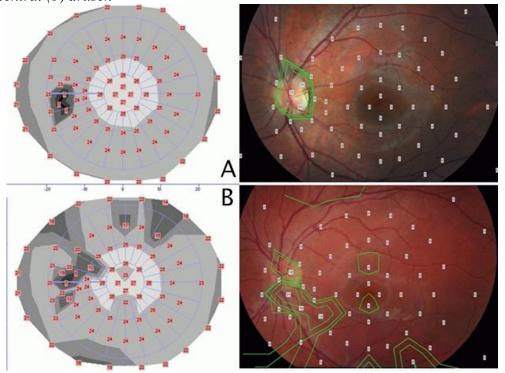
Figure 4. Changes in the retina and optic disc due to the drusen compression influence of various localisation (a–peripheral, b–central, c–in a patient with large drusen in central location)



Despite the lack of decrease in visual acuity, patients with drusen in central location showed a significant depression of the retina light sensitivity, as indicated by changes in MS and MD indicators (Table 1).

In children from the second clinical group average light sensitivity variation exceeded this value in those from the first clinical group by 8 times, and by 4.5 times in control group. In most cases, we observed paracentral relative scotomas, and scotomas fanning out from the optic disc along the nerve fibres in the vicinity of the drusen locations (Figure 5).

Figure 5. Changes the retina sensitivity in patients with peripheral (a) and central (b) drusen



The most informative indicators of the retina functional activity according to electrophysiological studies are:

- the amplitude of photopic ERG b-wave,
- increased inter-peak latency of oscillatory potentials, reflecting activity of the second order neurons (bi-polars and horizontal cells),
- disturbance of interneuronal relationship and
- the degree of ischemia of the retina inner layers, which is fed by the branches of the central retinal artery.

Decrease in P100 amplitude pattern in visual evoked potential (VEP) and rhythmic ERG point out the involvement of papillomacular beam and macular area of the retina, which is fully consistent with the change in perimetry parameters (Figure 5).

OCTA is the method of combining the study of the retina inner layers and the state of perfusion in its vessels. In assessing the changes we took into account the vessel density of surface and deep vascular plexuses. Patients with deep drusen showed decrease by 7.3% in foveal perfusion and by 13.5% parafoveally when compared with the control group and the first clinical group (Figure 6).

Figure 6. Changes in the vessel density (%) of the surface vascular plexus according to OCT angiography results in patients with peripheral (a) and

ensity (%)	Section	Thickness (µm)	Density (%)	Section	Thickness (µm)
50.36	Whole Image	N/A	42.20	Whole Image	N/A
35.56	Fovea	216	32.89	Fovea	251
51.37	ParaFovea	299	44.45	ParaFovea	314
51.28	- Superior-Hemi	300	43.63	- Superior-Hemi	314
51.47	- Inferior-Hemi	297	45.26	- Inferior-Hemi	313
54.21	- Tempo	293	46.45	- Tempo	298
52.47	- Superior	309	42.41	- Superior	322
48.40	- Nasal	289	45.86	- Nasal	318
50.46	- Inferior	303	43.08	- Inferior	317
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Conclusion

Inclusion in the examination algorithm of modern high-tech diagnostic methods allows not only to establish presence of drusen, but also to determine their precise location, level of morphological and functional changes, evaluate the severity and prognosis of the disease course.

Significant changes in the structural and functional parameters of the visual system in patients with deep central drusen are caused by mechanical compression of the optic disc and accompanied by:

- a blockage of retrograde axonal transport,
- a disturbance of the blood supply to the retina
- damage to glia and axons of ganglion cells.

This allows us to consider this pathology as a progressive neuroopticopathy. Identification of the triggers of this pathological process would determine search for effective treatments.

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