THE OPPORTUNITIES OF ULTRASOUND BIOMICROSCOPY IN DIAGNOSIS AND MONITORING OF IRIDOCORNEAL ENDOTHELIAL SYNDROME

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The aim of the investigation is to estimate the opportunities of ultrasound biomicroscopy in diagnosis and monitoring in various types iridocorneal endothelial syndrome.

Materials and Methods. The study was carried out on the basis of a detailed analysis of scanning images: ultrasound eye biomicroscopy was performed on 29 patients with iridocorneal endothelial syndrome.

Results. There were revealed ultrasound microscopic features reflecting pathological changes in the anterior eye bulb.

Conclusion. The ultrasound microscopy value is that it enables to assess to the fullest extent the degree of anatomic structures involvement into pathological process as it is the only way to obtain life-time high resolution images of eye bulb "latent" zones. Moreover, using the technique iridocorneal endothelial syndrome can be differentiated from other pathological states with similar clinical picture. The authors recommend using the method for diagnosis and monitoring of various types of iridocorneal endothelial syndrome.

Key words: iridocorneal endothelial syndrome, ultrasound eye microscopy.

The term iridocorneal endothelial syndrome (ICES) was first offered by M. Yanoff and H.G. Scheie in 1975 [1]. It unites three clinical manifestations of one pathological process — iris-nevus syndrome (Cogan-Reese syndrome), progressive (essential) iris atrophy and Chandler syndrome. As a rule, one eye is involved, and typical patients are young and middle-aged women. The etiology of the process is unknown. The pathological process is initiated by the changes in corneal endothelium (the so called endothelial epithelialization): pathologically changed cells of posterior corneal epithelium migrate through the angle of anterior chamber (endothelization of the angle of anterior chamber) into iris surface. The contraction of migrated membrane-like iridocorneal endothelial tissue leads to the formation of sectoral iridocorneal synechias and goniosynechias, distortion and ectopic pupil, pigmentary border ectropion, iris defects. On anterior iris surface there can form nodules and nevi (it is more typical for Cogan-Reese syndrome), iris atrophy (even via openings can develop that is the characteristic of essential iris atrophy), corneal edema develops (more severe in Chandler syndrome) [2]. And finally iridocorneal endothelial syndrome (ICES) in most patients (according to some authors, in 46–82% of cases) results in secondary refractory glaucoma [2, 3]. If untreated appropriately, secondary refractory glaucoma causes blindness.

Despite typical clinical signs, in some cases it is difficult to diagnose ICES. The cause of incorrect and/or late diagnosis is, on the one hand, that a doctor frequently has no suspicion due to the fact the disease is rare, and on the other hand — ICES variable clinical picture resembling to other pathological states with similar changes of anterior eye (e.g. iridociliary tumours with pupil distortion and pigmentary border ectropion, conditions after iritis or iridocyclitis with formation of anterior, and later — posterior synechias, deforming the pupil, posttraumatic conditions, various congenital anomalies). In a number of cases the diagnosis difficulty is worsened by corneal edema masking the zone of the angle of anterior chamber (AAC).

The imaging possibilities of ocular anterior chamber structures not easily accessible to be examined using conventional methods can be significantly widened applying ultrasound biomicroscopy (UBM).

The aim of the investigation is to estimate the opportunities of ultrasound biomicroscopy in diagnosis and monitoring in various types iridocorneal endothelial syndrome.

Materials and Methods. There have been analyzed the examination results of ICES patients followed up in the clinic of Eye Diseases Scientific Research Institute of RAMS from 2003 to 2010. There were studied 29 patients aged from 31 to 66 years (average age: 45.1±9.3), 26 female and 3 male patients. All patients had monolateral eye lesion.

Clinical examination included the evaluation of a patient's complain, history taking, diagnostics using traditional methods (test eyesight, tonometry, tonography, perimetry, biomicroscopy, gonioscopy). Photographic registration was used for objective follow-up. UBM was performed during epibulbar anesthesia using OTI HF 35–50 Ultrasound System (OTI, Canada). Funnel-like eye speculum filled with

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immersion fluid (saline solution) was put into conjunctival cavity. Ultrasonic transducer was immersed into the solution, and underlying tissues were scanned in datum plane. There were used axial, longitudinal and tangential scanning algorithms. Ultrasonic radiation frequency was 35 and 50MHz, scanning depth — 5x5 and 15x15 mm, measuring accuracy — 60 and 40 micrometers. The evaluation criteria of anterior chamber state were surface contour relief, reflectivity, homogeneity, quantitative parameters and spatial relations of anatomical structures. Colour reproduction in a number of cases enabled to show better the details of reflectivity. In reexaminations quantitative and qualitative characteristics of scanning images were compared with previous ones. To reveal the corneal changes typical of ICES, confocal microscopy method was used.

Results. Diagnostic errors related to ICES are common and described in literature. From 29 patients studied, Cogan-Reese syndrome was clinically diagnosed in 16, progressive iris atrophy — in 9, and Chandler syndrome — in 4 patients. In 25 cases the ICES diagnosis was made for the first time; 14 patients from them had been followed up in other medical institutions for a long time. Among the misdiagnoses were tuour of iris and iridociliary zone, traumatic ectopic pupil, primary glaucoma, etc. The application of UBM made it possible not only differential diagnosis but also ICES UBM signs detection.

The corneal thickness was assessed in comparison with the paired eye. In 9 cases there was increase in its thickness in the centre by 80–120 micrometers; traditional pachymetry failed due to posterior epithelium edema. Maximum corneal edema with the increase of reflectivity and thickness in the centre up to 740 micrometer (510 micrometer — in intact eye) and in the periphery — up to 1100 micrometer was revealed in a female patient with Chandler syndrome (Fig. 1).

Pathological changes in AAC area were found in all patients. Scanning images of iridocorneal angle of 9 patients showed membrane-like tissue filling the vertex of AAC (Fig. 2, *a*). Iridocorneal synechias of various degree were in all patients with ICES, with goniosynechias being revealed in all eyes, in 14 cases with Cogane-Reese syndrome — sectoral planar anterior synechias (Fig. 2, *b*, 3, *c*), in 18 eyes — local iridocorneal synechias in a view of medium periphery or pupillary zone of iris, with AAC vertex remaining anatomically open (Fig. 2, *c*). It should be noted that it is impossible to estimate AAC state in gonioscopy in such cases. Due to iridocorneal synechias there was changing the iris position (on scanning images the iris profile was unsymmetrical, deviated from frontal plane, and had bridging profile on tangential view (Fig. 3, *a*, *b*), with iris middle part being soldered with cornea, and iridocorneal angle remaining open), the configuration of anterior and posterior chambers (the volume of the anterior chamber was decreased, and the volume of the posterior chamber — increased), there was deformed and ectopic pupil (Fig. 4, *a*, *b*), pigmentary border ectopion (on scanning images there was seen the junction of hyper-reflector strip of pigmented leaf through pupillary edge to iris anterior surface) (Fig. 3, *c*, 4, *c*).

Due to marked ectopic pupil, the depth of anterior chamber in the centre in 11 eyes was less by 500–680 micrometers in comparison with the paired eye. Iris atrophy was found in al patients, its degree varying. In some cases iris kept its initial profile on scanning images, but it was 1.5–2 times as thinner than the norm, and the surface relief was subdued (Fig. 3, *d*). In the presence of iridocorneal synechias, iris looked thickened on iris medium

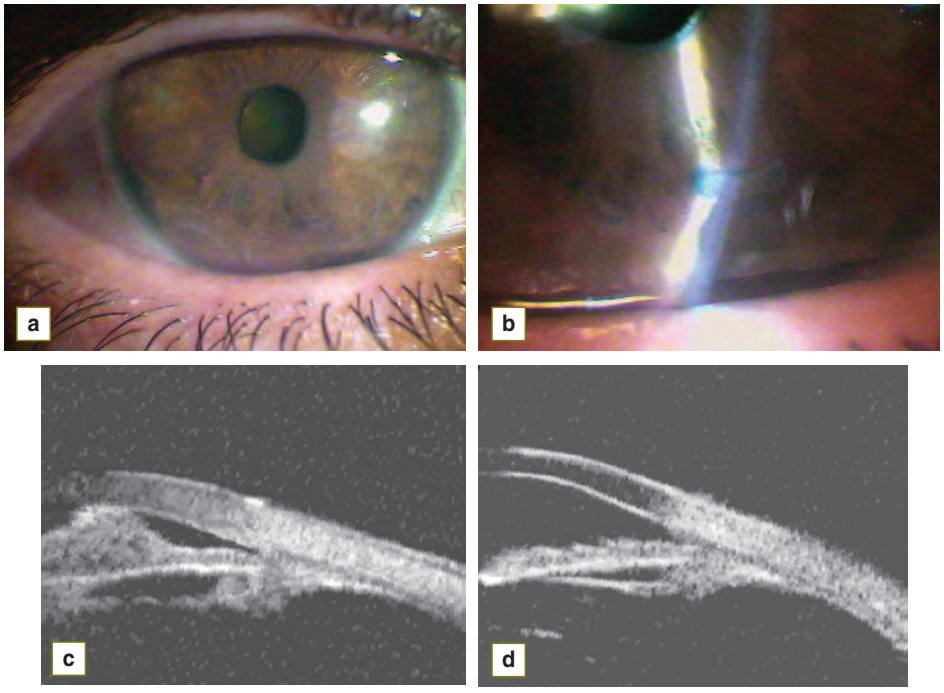

Fig. 1. Chandler syndrome: *a, b* — microscopic view of an affected eye; *c* — UBM picture of an affected eye, *d* — UBM picture of intact paired eye for comparison

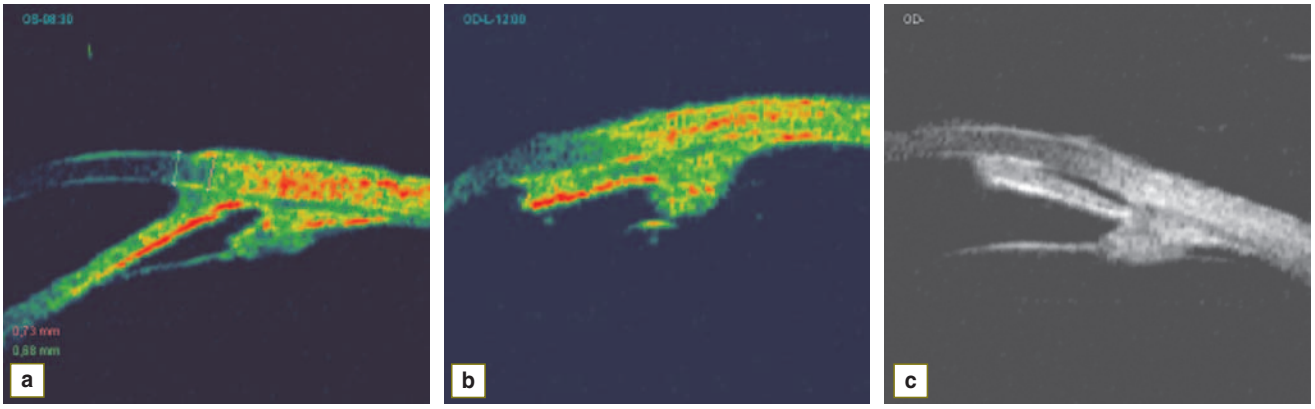

Fig. 2. UMB picture of iridocorneal synechias (ICS): *a* — membrane-like tissue filling the vertex of AAC; *b* — sectoral planar synechia; *c* — local synechia in pupillary edge view

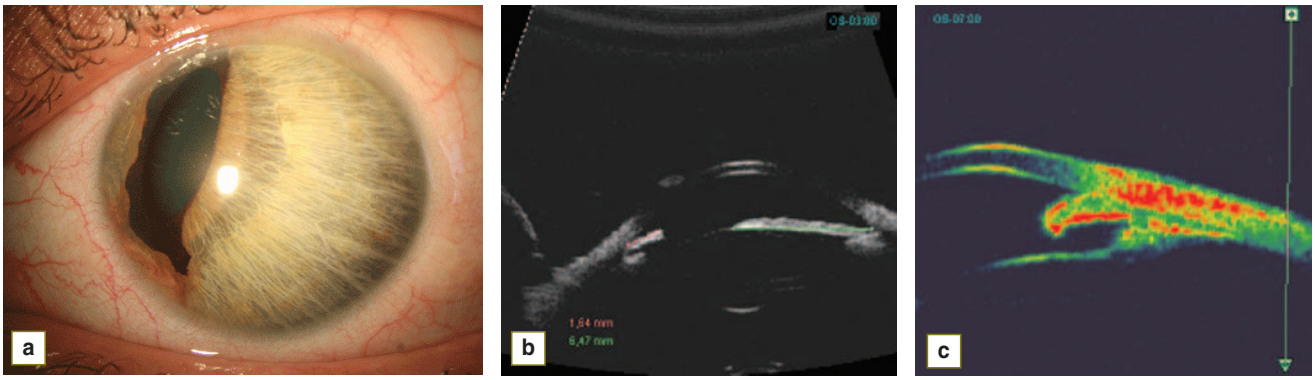

Fig. 3. Cogan-Reese syndrome: *a* — biomicroscopic view of the eye; *b*, *c* — UBM picture

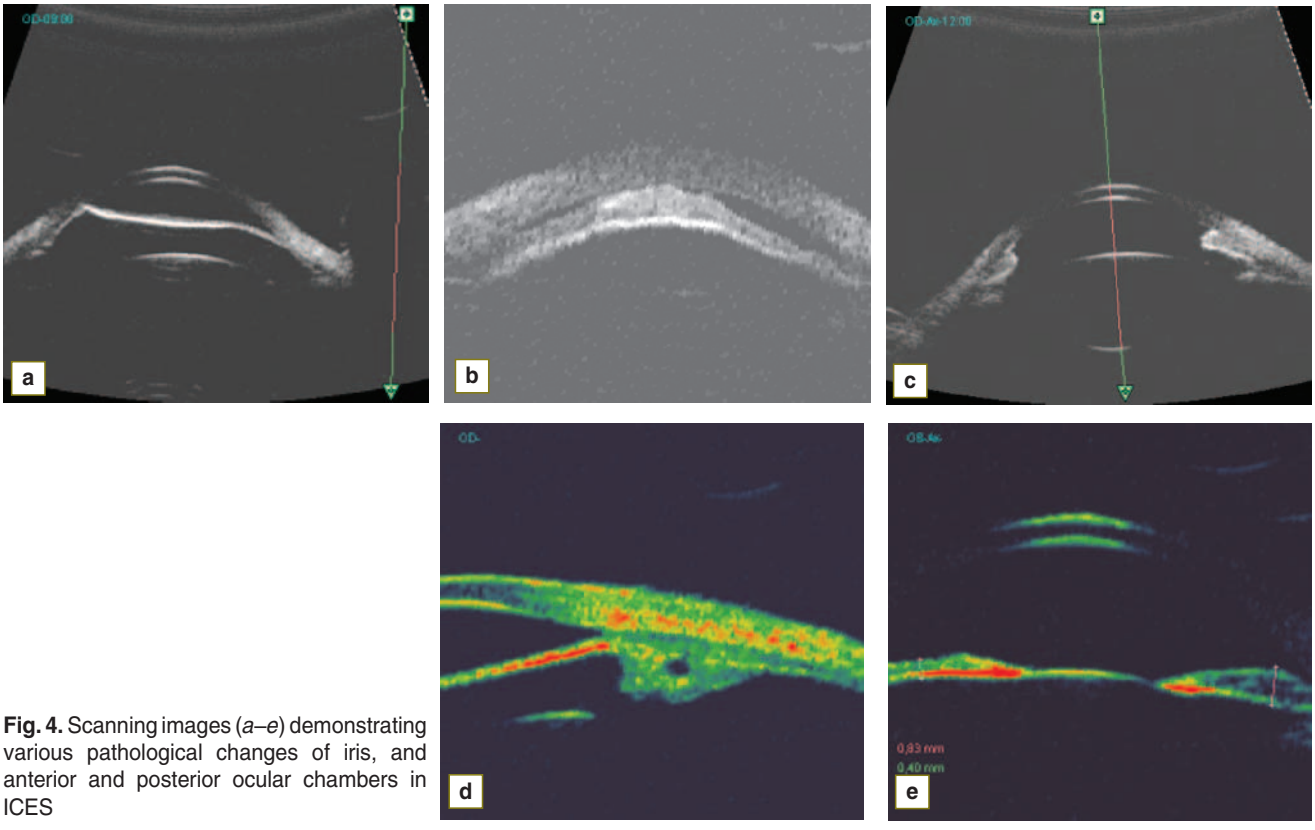

Fig. 4. Scanning images (*a*–*e*) demonstrating various pathological changes of iris, and anterior and posterior ocular chambers in ICES

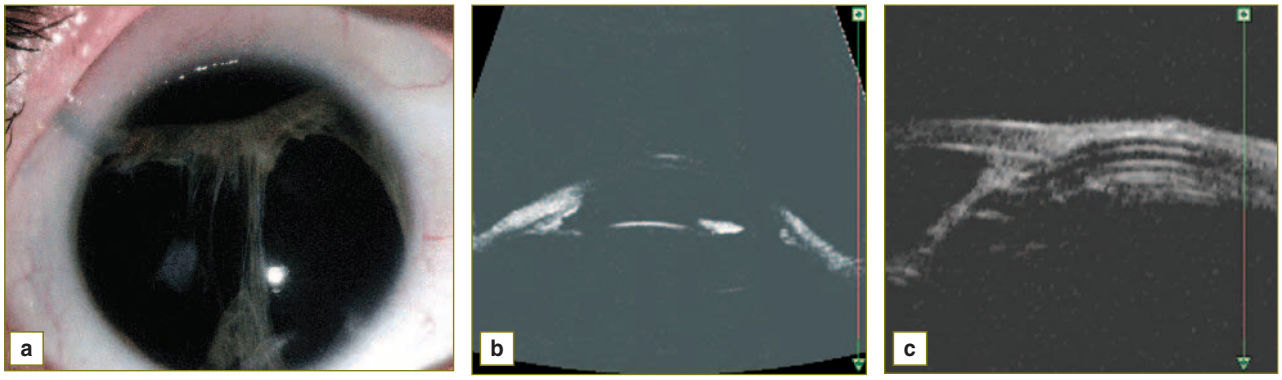

Fig. 5. Progressive iris atrophy: *a* — view of the eye in biomicroscopy; *b*, *c* — UBM picture

periphery view, though with low echodensity compared to normal reflectivity (Fig. 3, *e*). In severe cases, extensive iris atrophy combined with sectoral anterior synechias changed greatly the configuration and relationship of anatomical structures of the eye anterior chamber. In general, iris atrophy was more expressed in progressive iris atrophy.

2 patients with marked iris atrophy and, as a consequence, impaired iridolenticular diaphragm, had the change of lens position (the equator in one of the sectors shifted forward) that clinically resulted in lenticular astigmatism.

In ciliary body area, 12 patients had single iridociliary cysts (from 300–400 micrometer to 1.2–1.5 mm in size), in one case — numerous small unilocular cysts (up to 300 micrometer) and multilocular cysts that is typical for iridociliary dystrophies including ICES.

In some patients (8 eyes) UBM was re-performed (2–3 times every other year). The comparison of pathological changes (the length of iridocorneal synechias, the degree of spatial relations abnormality of anatomical structures) enabled to objectivize the follow-up of these patients.

The estimation of surgical results is of great importance in disease monitoring. Two patients from the group under study had been operated before. Primary open-angle glaucoma had been diagnosed and non-penetrating deep sclerectomy with collagen drainage implantation had been performed a year before being admitted to our institute. UBM revealed goniosynechias, AAC vertex (1/2 of the circle) was "covered" by membrane-like tissue with medium reflectivity that was the cause of intraocular pressure compensation absence, to our opinion. In the second case, in progressive refractory glaucoma, there was carried out silicone drainage implantation with a valve and amniotic insert (Fig. 5, *a*). UBM enabled to see proximal (pre-equatorial) part of a drainage tube, and goniosinechias along nearly the full AAC circle (Fig. 5, *b*, *c*). Due to marked atrophy, iris profile was deformed greatly and the most scanning images showed reduced residuals of iris root parts soldered with cornea.

Thus, UBM proved suspected ICES in all studied patients, and in a number of cases made it possible to rule out neoplasms and other pathological changes.

Conclusion. The analysis of surface profile relief, reflectivity, homogenicity, quantitative parameters and spatial relations of anatomical structures in iridocorneal endothelial syndrome based on high-frequency ultrasound biomicroscopy, provides objective and reliable view of pathologically changed eye anterior chamber. The technique enables not only to estimate the degree of pathological changes induced by the syndrome but also differentiate between the syndrome and other nosological conditions with similar clinical picture.

Ultrasound biomicroscopy (along with routine diagnostic methods, gonioscopy and confocal corneal microscopy) should be included into patients' examination algorithm to avoid diagnostic errors and to objectivize follow up in iridocorneal endothelial syndrome (including postoperative period).

References

1. Scheie H.G., Yanoff M. *Arch Ophthalmol* 1975; 93(10): 378–379.
2. Wilson M.C., Shields M.B. *Arch Ophthalmol* 1989; 107(10): 1465–1468.
3. Laganowski H.C., Kerr Muir M.G., Hitchings R.A. *Arch Ophthalmol* 1992; 110: 346–350.