Original article

Clinical result of the treatment of a child with congenital pathology of the iris and lens

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ABSTRACT

Relevance. Congenital aniridia is a rare ocular malformation. Its occurrence in the world ranges from 1: 40,000 to 1: 96,000.
Purpose. To study clinical and functional characteristics of both eyes and the effectiveness of treatment in a child with congenital aniridia, congenital cataract and lens subluxation.
Material and methods. The article presents the results of the clinical case of the child T., born in 2006, diagnosed with: OU – Subluxation of the lens congenital III degree. Cataract congenital intrauterine polymorphic. Aniridia is congenital hereditary. Glaucoma congenital intrauterine initial subcompensated (m). Myopia congenital high with astigmatism. Nystagmus horizontal fine-caliber constant. Surgery was performed on both eyes – FAC + IOL + ICR with implantation of the complex «Artificial iris» + Anterior vitrectomy. The article presents an analysis of a clinical case with familial congenital aniridia in a child in order to increase the awareness of treating ophthalmologists about the complexity and systemic manifestations of this congenital malformation and the need for a systematic approach to its treatment.
Results. Surgery and postoperative period without complications. Visual acuity after surgery was – OD – 0.08 w/c cyl (+) 1.0 D = 0.2; OS – 0.1 N/c. The position of the Artificial Iris complex is correct. IOL in the back chamber, in the capsule bag. A month after the operation, the visual functions are preserved, the IOP is compensated, the horizontal nystagmus has decreased
Conclusion:
• The use of the «Artificial iris» complex with a one-stage IOL implantation to correct the elimination of congenital aniridia makes it possible to achieve high cosmetic and functional results.
• The child and the child's parents are satisfied with the cosmetic effect after the operation.
• A child after undergoing correction of aniridia with the «Artificial Iris» unit requires constant dynamic observation by an ophthalmologist.
• It is necessary to take into account the high risk of postoperative ophthalmic hypertension in the child and the possibility of repeated interventions.
Key words: *congenital aniridia, congenital cataract, lens subluxation, cataract phacoaspiration with IOL implantation, intracapsular ring, Artificial iris complex*

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Клинический случай лечения ребенка с врожденной патологией радужки и хрусталика

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РЕФЕРАТ

Актуальность. Врожденная аниридия – редкий порок развития глаза. Его распространенность в мире варьирует от 1:40 000 до 1:96 000.
Цель. Изучить клинико-функциональные особенности обоих глаз и эффективность лечения у ребенка с врожденной аниридией, врожденной катарактой и подвывихом хрусталика.
Материал и методы. В статье описан клинический случай. Ребенок Т., 2006 г.р., с диагнозом: OU – врожденный подвывих хрусталика III степени. Полиморфная врожденная катаракта. Врожденная наследственная аниридия. Начальная субкомпенсированная врожденная глаукома. Врожденная близорукость высокой степени, астигматизм. Горизонтальный мелкокалиберный постоянный нистагм. Операция была проведена на обоих глазах: факоэмульсификация катаракты + интраокулярная линза (ИОЛ) + внутрикапсульное кольцо с имплантацией комплекса «искусственная радужка» + передняя витрэктомия. В статье представлен клинический случай семейной врожденной аниридии у ребенка с целью повышения осведомленности практикующих офтальмологов о сложности и системных проявлениях этого врожденного порока развития и необходимости системного подхода к его лечению.
Результаты. Хирургия и послеоперационный период – без осложнений. Острота зрения после операции составила –

OD – 0,08 w/c, cyl (+) 1,0 D = 0,2; OS – 0,1 н/к. Положение комплекса искусственной радужки правильное. ИОЛ в капсульной сумке. Через месяц после операции зрительные функции сохранены, внутриглазное давление компенсировано, горизонтальный нистагм уменьшился.

Выводы:

• Использование комплекса «искусственная радужка» с одномоментной имплантацией ИОЛ для устранения врожденной аниридии позволяет достичь высоких косметических и функциональных результатов.

• Ребенок и его родители довольны полученным косметическим эффектом.

• Ребенок, перенесший коррекцию аниридии с использованием комплекса «искусственная радужка», нуждается в постоянном динамическом наблюдении офтальмолога.

• В подобных случаях у детей необходимо учитывать высокий риск послеоперационной офтальмологической гипертензии и возможность повторных вмешательств.

Ключевые слова: *врожденная аниридия, врожденная катаракта, подвывих хрусталика, факоаспирация катаракты с имплантацией ИОЛ, интракапсулярное кольцо, комплекс искусственной радужки*

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Congenital aniridia can result from autosomal dominant inheritance from a diseased parent, from spontaneous mutation, or from WAGR, WAGRO, and mental retardation-related syndromes. In addition, studies have shown that congenital aniridia associated with changes in the PAX6 gene is accompanied by additional systemic changes (pathology of the endocrine and nervous systems, metabolic disorders). In this regard, PAX6-associated aniridia is more and more often described as «Aniridial syndrome», or «PAX6-syndrome». Unlike other congenital eye malformations, pathological changes in aniridia (cataract, subluxation, keratopathy, secondary glaucoma) progress throughout life and can cause complete blindness. There are several types of aniridia depending on the type of inheritance, the type often found: – Family aniridia (autosomal dominant). In this case, the probability of a child getting sick is 50% [1–5].

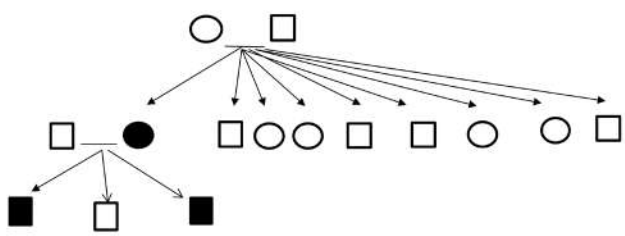
Case description Child T., born in 2005, with a diagnosis of: OU – Congenital lens subluxation, grade III. Congenital intrauterine polymorphic cataract. Aniridia is congenital hereditary. Glaucoma, congenital intrauterine initial subcompensated (m). Congenital high myopia with astigmatism. Horizontal small-bore nystagmus constant.

Anamnesismorbi: According to the child's mother, she notes low vision in both eyes from birth. At the place of residence they turned to an ophthalmologist. Diagnosed with OU – Congenital cataract. Congenital aniridia. According to the child's mother, they were incorrectly informed and did not undergo examination or consult about treatment until the age of 13. The child goes to a specialized school for the visually impaired.

As presented in schedule 1, child T., born in 2006, has a sick mother and a sick older brother *(fig. 1, 2)*. The parents of the mother, the rest of the brothers, the sisters of the mother of the child T., are healthy.

Clinical and instrumental examination of the child T. 2006:

Visual acuity on admission:



(Older brother, born in 1990) (Child, Born in 2006)

Patient geneology: sch 1. Family geneology tree of the child T., 2006, confirming the type of inheritance «Family aniridia»

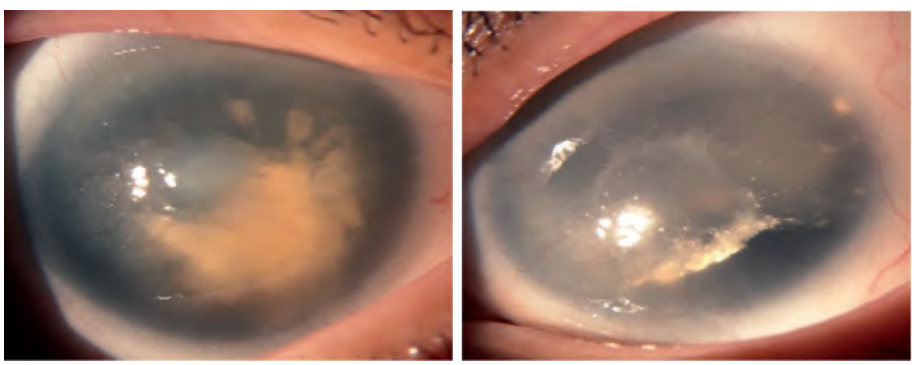


Fig. 1. Photo of the mother's right and left eyes, born in 1968. Diagnosis: OU – Congenital aniridia, grade III lens subluxation, congenital cataract, keratopathy. OU – 0,002 (hand movement near the face)

OD – 0.01 w/c sph (–) 5.0 D = 0.08;
OS – 0.04 w/c sph (–) 5.0 D = 0.08.
Intraocular pressure is contact:
OU – 19.0 mm Hg. Ultrasound: OD – 26.94 mm, OS – 26.94 mm.
OU – Vitreous destruction. The retina is attached.
Optical coherence tomography (OCT): OU – the relief of the macula is smoothed, the layers are not differentiated, thinning throughout.
Objectively: OD – Constant horizontal movement of the eyeball. The conjunctiva is pale pink. The sclera is white. The cornea is transparent. D = 10 mm. The anterior chamber is uneven, moisture is transparent. The remains of the root of the iris in the form of a ring. The lens is displaced upward and outwardly, diffusely turbid in all layers. From 3: 00 h until 9:00 Zinn ligaments are weakened. At 6:00 o'clock, complete separation of the zinc ligament. Deep media are not visible in detail *(fig. 3)*.
OS – Constant horizontal movement of the eyeball. The conjunctiva is pale pink. The sclera is white. The cornea is transparent. D = 10 mm. The anterior chamber is uneven, moisture is transparent. The remains of the root of the iris in the form of a ring. The lens is reduced in size, displaced upward and outwardly, evenly clouded in the cortical layers. From 3: 00 h until 9:00 Zinn ligaments are weakened. At 6:00 o'clock, complete separation of the zinc ligament. Deep environments are not visible in detail *(fig. 4)*.
An operation was performed on both eyes: OU – Phacoaspiration of cataract with IOL implantation of the «Artificial Iris» complex + Anterior vitrectomy *(fig. 5)*.

RESULTS

Visual acuity at discharge: OD – 0.08 w/c cyl (+) 1.0 D = 0.2; OS – 0.1 n/c.
IOP contact: OD – 19.0 mm Hg; OS – 16.0 mm Hg
OU – Conjunctiva pale pink. The cornea is transparent. The anterior chamber is medium, moisture is transparent. Artificial iris complex in the correct position. IOL in the posterior chamber, the position is correct *(fig. 6, 7)*.
In conclusion, in pediatric ophthalmology, it is important to determine the clinical signs that have a prognostic value for the course of the disease in the future. In this regard, early diagnosis

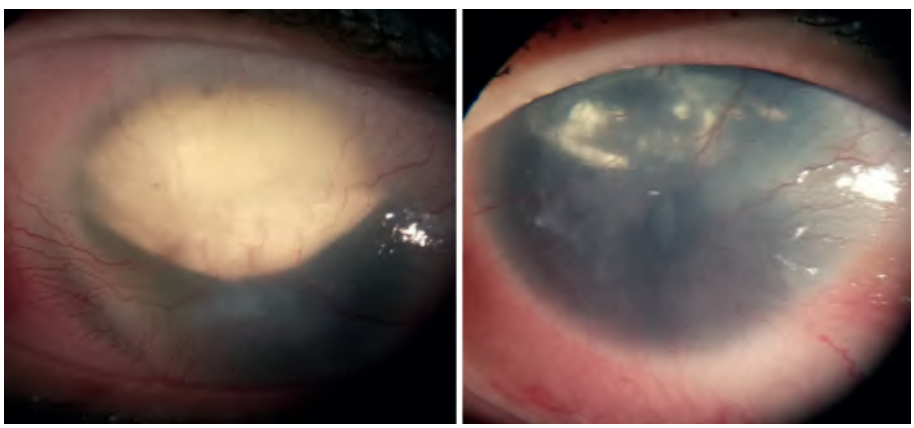


Fig. 2. Photo of the right and left eyes of the older brother, born in 1990. Diagnosis: OU– Congenital aniridia, grade III lens subluxation, congenital cataract, corneal neovascularization, congenital glaucoma

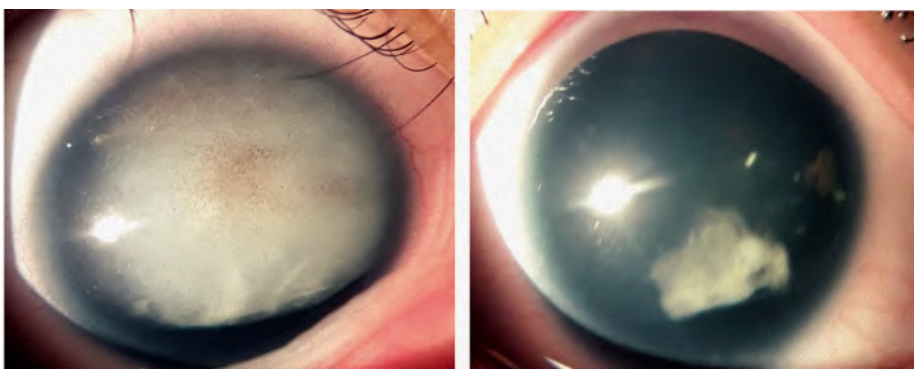


Fig. 3, 4. Photos of the right and left eye of the child T., 2006

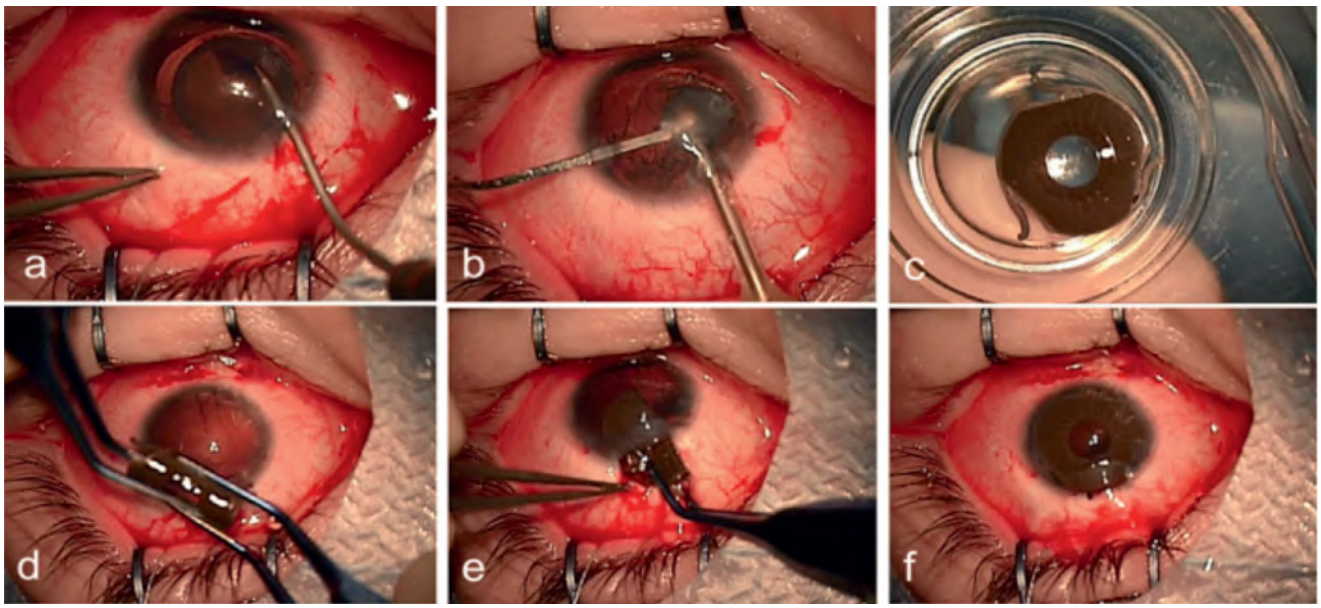


Fig. 5. Stages of surgery

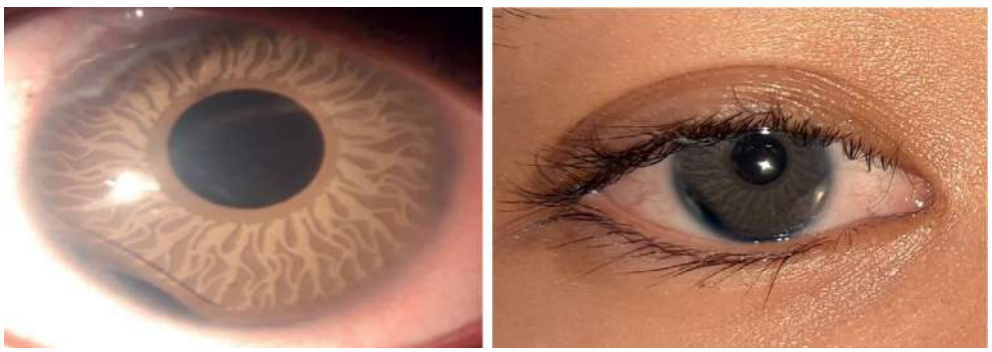


Fig. 6. Photo of the right eye of the child T., 2006, after operation

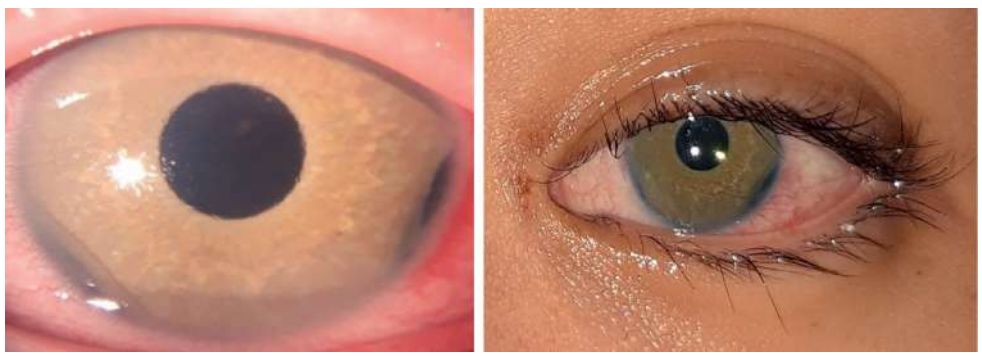


Fig. 7. Photo of the left eye of the child T., 2006, after operation

and early detection of complications are very important in order to preserve visual functions, visual fields, reduce the effect of damaging factors and ensure a better quality of life for aniridia patients.

LITERATURE

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