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Features of Management of Children with Congenal Cataracts and Accompanying Visual Pathology

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Abstract An analysis of the case histories of 72 children (117 eyes) diagnosed with congenital cataracts, who were hospitalized in the eye department of the TashPMI clinic for the period from 2016 to 2023 at the age of 3 months to 14 years, was carried out. The study revealed that, children with congenital cataracts and concomitant pathology of the visual organ require a differentiated approach to patient management: in the presence of microphthalmos, early surgical intervention is necessary before 1 year - stage 1, including cataract extraction, followed by stage 2 - IOL implantation. Constant dynamic monitoring is necessary, given the high percentage of postoperative complications.

Keywords Congenital cataract, Concomitant pathology, Microphthalmia, IOL implantation

1. Introduction

One of the common causes of blindness and low vision in children is congenital pathology of the lens, namely cataracts, which account for up to 10-19.5% [5,7,9,16]. Congenital cataracts (CC) are observed in 5 cases out of 100,000 newborns and cause 10-38% of cases of childhood blindness [5]. Cataract is a persistent clouding of the substance of the lens or its bag, accompanied by a decrease in visual acuity from a slight weakening to light perception. Congenital cataracts are often combined with other pathological changes in the organ of vision, which are observed in 36.8-77.3% of children: strabismus, nystagmus, microphthalmos, microcornea and other anomalies of the cornea, as well as the vitreous body, choroid, retina and optic nerve. Strabismus is observed in 30.2-83.3% of children with congenital cataracts. More often it is converging, alternating, predominantly constant. Nystagmus is observed in 14.0-58.8% of children with congenital cataracts; it can be congenital or acquired, associated with a sharp decrease in vision. More often it is pendulum-shaped, horizontal, less often - jerky. Microphthalmos is a congenital anomaly that is often associated with congenital cataracts in 22.5% of cases due to its characteristic structures including a thickened sclera, a normal or slightly larger lens with a relatively small eye volume, a higher lens-to-eye volume ratio, and a shallow anterior chamber, narrow camera angle and short ocular axis. Microphthalmos can cause glaucoma and other complications affecting the retina and choroid. Congenital cataract with complex microphthalmos is often combined with ocular or systemic abnormalities, and patients after surgery are prone to secondary glaucoma, corneal opacification and other serious complications [10,11,14,15]. Depending on the size of the eye, there are 3 degrees of microphthalmos: 1 degree - a decrease in one or two of the above dimensions by 1.0-1.5 mm compared to the age norm; 2nd degree - decrease by 2.0-2.5 mm; 3rd degree - reduction by 3mm or more. Features of congenital cataracts with microphthalmia are predominantly complete opacities of the lens - 66.7% (zonular are 2.7%), a high percentage of capsulo-lenticular opacities - 67.1%, a narrow rigid pupil and its various anomalies, even absence. The most common change in the cornea in congenital cataracts is a change in its size - microcornea. Colobomas of the iris are often encountered, which vary widely in size and shape and are often located in the lower segment [4]. Severe anomalies are various manifestations of mesodermal dysgenesis. Cataract is one of the elements of Rieger's syndrome (dysgenesis of the iris and cornea) and Peters syndrome (mesodermal-ectodermal dysgenesis of the cornea and iris). CC is also part of the Persistent hyperplastic primary vitreous (PHPV). PHPV is associated with a disruption of the process of regression of the primary embryonic vitreous body. Changes in the retina and optic nerve of varying nature and severity, identified in 55% of cases, are one of the reasons for low visual acuity after extraction of congenital cataracts. More often, combined lesions were observed (40.5%), less often - isolated changes in the retina (5.0%) or optic nerve (9.5%). Pathologies of the optic nerve included partial atrophy and developmental abnormalities (reduction in size, change in disc shape, etc.). On the part of the retina, macular hypoplasia, myelin fibers, central and peripheral dystrophy, “old” chorioretinal lesions (multiple, small, in the periphery, or large in the central region) were detected [2,3]. Treatment of congenital cataracts in children remains a serious problem, which is associated not only with the anatomical

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features of the structure of the child's eye, but also with the frequency of concomitant pathologies, as well as with various complications caused by the hyperergic state of the child's immune system [7,6,8,16].

One of the most important questions remains about the timing of surgical intervention for congenital cataracts. Currently, most authors are of the opinion that complete, layered and central cataracts with an opacification area of more than 2.5 mm should be operated on within the first three months after the birth of the child [1,3], and for para central and central cataracts less than 2.5 mm in diameter - dynamic observation is possible. Proponents of a more radical approach insist on surgical treatment of congenital cataracts after a clinical diagnosis of the disease has been established [12]. However, despite different views, in most cases the problem of early surgical intervention is solved individually in each case, taking into account not only the degree of intensity of lens opacification, but also the presence and severity of combined pathology of the visual analyzer and the child's body as a whole [13,14].

Purpose of the study. To study the features of management of children with congenital cataracts and concomitant pathologies of the visual organ.

2. Material and Research Methods

An analysis of the case histories of 72 children (117 eyes) diagnosed with congenital cataracts, who were hospitalized in the eye department of the TashPMI clinic for the period from 2016 to 2023 at the age of 3 months to 14 years, was carried out. Boys made up 42%, girls 58%. A bilateral process is observed in 45 (62%), unilateral in 27 (38%) children. The patients underwent visometry, biomicroscopy, keratometry, ophthalmoscopy, tonometry, echobiometry, strabismus angle determination, and visual evoked potential (VEP) study. Preoperative preparation also included consultations with a pediatrician and neurologist; if necessary, children were consulted by a geneticist, an ophthalmologist, and other specialists.

3. Research Results and Discussion

According to the clinical and surgical classification of congenital cataracts [5], "layered" (nuclear, zonular) were observed in 22 (19%) eyes, "total" cataracts in 38 (32%), "atypical" cataracts (semi-resolved, calcified, anteroposterior capsular, posterior and anterior lenticonus) in 57 (49%) children. In patients, the range of concomitant pathologies was quite wide (Table 1).

Classical extracapsular cataract extraction (ECE) with simultaneous implantation of a soft IOL made of hydrophobic acrylic (Acrysof IQ, Acrysof) was performed in 84 (72%) eyes. In 33 (28%) eyes, ECE was performed without implantation due to the presence of high-grade microphthalmia and complications that arose during implantation (unplanned rupture of the posterior capsule), and the presence of pathology from the fundus of the eye (hypoplasia of the optic disc, stage IV-V, PHPV). In 72% of cases (42 eyes), the operation was performed through a small tunnel sclerocorneal incision; in 28% (17 eyes) of cases, tunnel self-sealing corneal incisions were used (in children over 3 years old) in the upper segment at 11-12 o'clock.

Table 1

Concomitant pathology of the organ of vision	Number of eyes * (n=117)	
	Abs.	%
Pathology of the adnexal apparatus	58	50
- strabismus	40	34
- nystagmus	18	16
Microphthalmos	45	39
I degree	21	18
II degree	15	13
III degree	9	8
Refractive errors (myopia)	18	15
PHPV	7	6
Hypoplasia of the optic disc and macula	17	15
Atrophy of the optic disc	9	8

**note - in some cases several types of pathologies were observed in one eye*

For children with microphthalmia, surgical treatment was carried out as early as possible. At the first stage, ECE was performed. After the first stage surgery, the patients' condition was monitored. At the second stage, an IOL was implanted with the lens placed in the ciliary sulcus if the eye condition was suitable.

In the postoperative period, an inflammatory reaction was observed in the form of a fibrous-exudative reaction that occurred in the first days in 49% of cases, which was stopped by frequent instillations of anti-inflammatory drugs. In 9% of cases, subluxation of the IOL was observed, which required repeated surgical intervention-reposition of the IOL. In 18% of cases, an increase in IOP was observed, which was stopped by the prescription of antihypertensive drugs.

The high percentage of complications can be explained by the presence of concomitant pathology of the organ of vision, especially microphthalmia and PHPV. Anatomical features of congenital cataracts with complex microphthalmos include reduced globe volume, reduced axial length (<18 mm), predominantly spherical lenses, the presence of possible microcornea and a shallow anterior chamber. Patients with congenital microcornea often have a corneal diameter <9 mm, corneal flattening, and are often associated with cataracts and coloboma of the iris or choroid. Congenital cataracts accompanied by complex microphthalmos or microcornea are often accompanied by a rigid pupil. Due to the structures of the eyeball characteristic of congenital microphthalmia, intervention for cataracts is very difficult and risky and can lead to postoperative complications such as severe inflammatory reaction, glaucoma, IOL luxation, retinal detachment and other serious complications.

Rehabilitation of children in the postoperative period included:

- Spectacle correction
- Treatment of amblyopia
- Conservative neuroprotective and nootropic therapy (with concomitant pathology of the optic disc)
- YAG laser capsulotomy for the development of fibrosis of the posterior capsule of the lens.

Rehabilitation measures must also be carried out in local clinics. Recommendations for improving the quality of rehabilitation of children with CC in outpatient settings include the following:

1. Increasing the level of knowledge of ophthalmologists in city clinics and regional centers in matters of diagnosis, treatment, clinical examination and rehabilitation of children with congenital cataracts and concomitant pathologies of the organ of vision.
2. Increasing the frequency of observations during the rehabilitation period - after discharge, 2 times a month for 2 months, then 1 time for 4 months, then as indicated. If the process is stabilized, it is necessary to examine children 1-2 times a year until they reach 15 years of age.
3. Increasing the frequency of studies performed during the rehabilitation period - during examination, it is necessary to monitor the density of lens opacity, measure the diameter of the cornea, determine the state of transparency of the lens capsule, determine acuity and IOP.

4. Conclusions

Thus, children with congenital cataracts and concomitant pathology of the visual organ require a differentiated approach to patient management: in the presence of microphthalmos, early surgical intervention is necessary before 1 year - stage 1, including cataract extraction, followed by stage 2 - IOL implantation. Constant dynamic monitoring is necessary, given the high percentage of postoperative complications. It is also necessary to carry out the following rehabilitation measures: glasses correction, measures aimed at combating amblyopia, if necessary, nootropic therapy, YAG - laser capsulotomy.

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