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OPTIMIZATION OF THE METHOD OF SURGICAL TREATMENT OF PRIMARY INFANTILE CONGENITAL GLAUCOMA IN CHILDREN

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Summary

Pediatric glaucoma is a term that encompasses several forms of congenital glaucoma and types of secondary glaucoma that differ in etiology, pathogenesis, and clinical presentation. Surgical treatment is the main method of IOP reduction in pediatric glaucoma. To date, no uniform method of surgical treatment of infantile glaucoma has been proposed, which makes the search for adequate methods of surgical intervention in this pathology most urgent.

The aim of this work was to evaluate the efficacy of surgical treatment of primary congenital infantile glaucoma in children.

Material and methods. *In the eye department of the clinic of the Tashkent Pediatric Medical Institute, the effectiveness of the method of surgical intervention in children from 3 to 10 years old with primary congenital infantile glaucoma was evaluated. Examined – 20 patients (40 eyes). Of these, 5 patients (10 eyes) entered the advanced stage, 10 patients (20 eyes) entered the far advanced stage, and 5 patients (10 eyes) entered the terminal stage, respectively. The patients underwent standard ophthalmological examination. The state of the optic nerve was investigated by optical coherence tomography Stratus OCT-3000 (Carl Zeiss Meditec), the visual fields – in 10 (20 eyes) patients aged over 8 years using standard automated and computer perimetry (SAP).*

Conclusions of the bioethical commission: *This article evaluates the effectiveness of the method of surgical treatment of primary congenital infantile glaucoma in children. And gives specific results of different operations at different stages of primary congenital infantile glaucoma. Surgical treatment was chosen according to the degree of goniodysgenesis and the stage of the disease, which showed a satisfactory result in stabilizing the glaucoma process.*

Statistical analysis method using Microsoft Excel and SPSS programs was used for processing the results of the ophthalmological study. Differences between the mean values ($M \pm \sigma$) were considered reliable at $P \leq 0.05$.

Results and Discussion. *All patients underwent surgical treatment, depending on the severity of morphometric changes in the anterior chamber angle, and in the postoperative period with an interval of 6 months – a course of neuroprotective therapy. According to the examinations, children with primary infantile glaucoma in advanced stage underwent non-penetrating deep sclerectomy in 10 eyes, with extensive – in 20 eyes and with terminal – in 10 eyes – sinusotrabelectomy ab externo, sinusotrabelectomy. In 2 eyes in the terminal stage, filtering surgery was performed according to the proposed method. Thus, the above methods of surgery reduce the number of intra- and postoperative complications by 12.5%, avoid reoperations, reduce IOP in 12.3% of cases, increase visual acuity by 0.15 compared to the original.*

Conclusion. *The above methods of surgical treatment were chosen according to the degree of goniodisgenesis and the stage of the disease, which showed a satisfactory result in stabilizing the glaucoma process.*

Key words: *Primary congenital infantile glaucoma; surgery; anterior-posterior axis of the eye; primary congenital glaucoma; ocular rigidity; optic disc excavation; myopia; intraocular pressure.*

Relevance

Pediatric glaucoma is a term that encompasses several forms of congenital glaucoma and types of secondary glaucoma that differ in etiology, pathogenesis, and clinical presentation [7,8,15,16]. Juvenile open-angle glaucoma (JOAG) is a form of open-angle glaucoma diagnosed in individuals older than 3 years and younger than 40 years of age. It is characterized by more severe elevations in intraocular pressure (IOP) and rapidly progressive visual field loss compared to adult primary open-angle glaucoma (POAG). Infantile (so-called delayed congenital) glaucoma manifests at the age of 3-10 years. Its pathogenesis and inheritance are similar to those of true congenital glaucoma. Gonioscopy shows signs of goniodysgenesis, but less pronounced than in true congenital glaucoma. The ophthalmotonus is increased, but the size of the cornea and globe are not altered, which is associated with greater scleral strength and resistance to IOP elevation compared to young children [7,3,25,9].

As glaucoma progresses, optic disc excavation (D/E) increases and visual function decreases [23]. Due to the high rate of disability in children with infantile glaucoma, the study of the pathogenesis of this severe congenital disease and the mechanisms of visual impairment is an important task in pediatric ophthalmology. Glaucomatous

optic neuropathy (GON) in this disease is defined by a combined compressive effect of elevated IOP on the membranes, optic nerve and other ocular structures, causing their stretching, deformation and dystrophy. As infantile glaucoma develops and progresses, there are increasing changes in the optic nerve and retina, and morphologic abnormalities occur in all structures [5,3]. Surgical treatment is the main method of IOP reduction in pediatric glaucoma. To date, no uniform method of surgical treatment of infantile glaucoma has been proposed, which makes the search for adequate methods of surgical intervention in this pathology very urgent [6,23].

Aim. The aim of the work was to evaluate the efficacy of surgical treatment of primary congenital infantile glaucoma in children.

Materials and methods. We studied 20 patients (40 eyes), boys 14 (70%) and girls 6 (30%) aged 3-10 years old with primary infantile glaucoma at the eye department of the clinic of Tashkent Pediatric Medical Institute. Diagnosing was guided by the classification of N. A. Kachan, T. K. Toikuliya [9,20], children were divided into stages of ocular disease as follows: advanced stage – 5 (10 eyes),

very advanced stage – 10 (20 eyes), terminal stage – 5 patients (10 eyes). Primary infantile glaucoma was associated with various degrees of myopia in 7 patients (14 eyes). Pigment dispersion syndrome was diagnosed in 1 patient (2 eyes). Anatomical classification of anterior chamber angle (AAC) according to Jr. Hoskins clinically determined anatomical defects of AAC development in patients [11]. All patients underwent surgical treatment and a course of neuroprotective therapy every 6 months for 1.5 years in the postoperative period [19].

Once a child is diagnosed with glaucoma, the goal is to provide lifelong vision if possible. Medications are an important part of management; they are usually the first line of treatment for most glaucomas, they temporize IOP control prior to surgery, for example in primary congenital glaucoma (PCG), and they are often required as adjunctive therapy after partially successful surgery for IOP control [17,22]. However, surgery is the mainstay of treatment for childhood glaucoma and is often unavoidable in a child's lifetime [4,15].

Recognition of the importance of measuring the impact of disease, disability and treatment from the patient's perspective has led to the development and evaluation of patient-reported outcome measures and patient-reported experience measures for use in pediatric ophthalmology [12,2], but there is a paucity of vision-specific instruments. It is hoped that the fact that patient-reported assessment of

the impact of disease has developed a high profile in health service planning and policy in some countries will lead to increased funding for this research [13,24,14,1].

Patients underwent a standard ophthalmic examination. The condition of the optic nerve was examined by optical coherence tomography Stratus OCT-3000 (Carl Zeiss Meditec), visual fields – in 10 (20 eyes) patients older than 8 years by standard automated and computer perimetry (SAP).

Statistical analysis using Microsoft Excel and SPSS programs was used to process the results of the ophthalmologic study. Differences between means ($M \pm \sigma$) were considered reliable at $P \leq 0.05$.

Results and discussion. The study showed that the average visual acuity of the patients before surgery was 0.3 ± 0.56 . IOP was 26.8 ± 2.01 mm Hg in advanced glaucoma, 28.8 ± 1.01 mm Hg in very advanced glaucoma, and 34.01 ± 1.53 mm Hg in end stage glaucoma. The average anteroposterior dimension (APD) of the eye was 23.6 ± 1.2 mm in the advanced stage, 24.6 ± 2.1 mm in the most advanced stage, 25.7 ± 2.2 mm in the final stage. On gonioscopy: isolated trabeculodysgenesis was found in 10 eyes (25%) in the advanced stage, anterior iris attachment in 20 eyes (50%) and posterior iris attachment in 10 eyes (25%) in the most advanced stage. Optic disc excavation (D/E) was 0.4 ± 0.05 in advanced stage, 0.7 ± 0.1 in advanced stage and 0.95 ± 0.002 in terminal stage (Table 1).

Table 1

Biometric parameters of eyes at different stages of infantile glaucoma before surgical treatment ($M \pm \sigma$)

Stages indicator	Development (n=10)	Fare gone (n=20)	Terminal (n=10)
P_i (mmHg)	$26,8 \pm 2,01$	$28,8 \pm 1,01$	$34,01 \pm 1,53$
APD (mm)	$23,6 \pm 1,2$	$24,6 \pm 2,1$	$25,7 \pm 2,2$ mm
D/E	$0,5 \pm 0,05$	$0,7 \pm 0,1$	$0,95 \pm 0,002$
P	$\leq 0,05$	$\leq 0,05$	$\leq 0,005$

n – number of eyes

Patients with advanced stage primary congenital infantile glaucoma underwent non-penetrating deep sclerectomy in 10 eyes. In our opinion, such a procedure is appropriate for the advanced stage of primary congenital infantile glaucoma because it improves the outflow of aqueous humor through the uveoscleral pathway under the conjunctiva. In 20 eyes in the advanced stage and in 10 eyes in the terminal stage, a combined filtration surgery was performed: sinusotrabeulotomy ab externo and sinusotrabeulectomy, while the aqueous humor is absorbed into the supraciliary space through the opened trabecular meshwork. A filtering operation was performed on 2 eyes at the terminal stage according to the method we proposed: sinusotrabeulotomy, sclerectomy, cycloretraction with autoscleral drainage (Patent for the invention "Method of surgical treatment of congenital glaucoma" No. IAP 04890 dated 12.05.2014). In our opinion, this method is most indicated in the terminal stage, as it improves the outflow of aqueous humor through the trabecular network, the uveoscleral pathway into the intrascleral space and under the conjunctiva, and autoscleral drainage prevents scarring of the outflow pathways.

We observed the following intraoperative complications: partial hyphema in 5 (12.5%) eyes with advanced and end-stage glaucoma; vitreous prolapse and choroidal detachment in 2 (5%) eyes with end-stage glaucoma. Partial hyphema resolved after 4 days and choroidal detachment resolved after 6 days.

After surgery, IOP indicators were maintained for 6-18 months in eyes with advanced stage in the range of -21.8 ± 0.9 mmHg, with advanced stage -23.1 ± 0.2 mmHg, with final stage -25.2 ± 2.1 mmHg.

With optical coherence tomography (OCT), morphometric data of the optic nerve head (ONH) showed that retinal tomography is a sensitive method for early diagnosis (indicator of neuroretinal rim area and volume) of primary infantile glaucoma, especially with an initial and unstable IOP elevation. In the case of a combination of primary infantile glaucoma and myopia, the degree of damage to the OD may be underestimated, so the use of OCT is informative and highly suggestive.

For dynamic observation of retinal and OD changes, we used a glaucoma damage likelihood scale (DDLS) [21]. After surgery for 6-18 months, morphometric indicators of

the OD showed an increase in the ratio of excavation area to disc area (0.35 ± 0.09) in 7 (17.5%) eyes, and a decrease in the neuroretinal rim area ($0.12 \pm 0.013 \text{ mm}^2$) at the final stage. In 26 eyes (65%), the excavation area decreased ($0.153 \pm 0.003 \text{ mm}^2$) and the neuroretinal rim volume increased ($0.08 \pm 0.005 \text{ mm}^3$).

In 10 patients (20 eyes) over 8 years of age, during 6-18 months after surgery, we were able to evaluate visual field indicators using static automated perimetry (SAP) in the threshold strategy phase within 300 from the fixation point. In the advanced stage of infantile glaucoma with tolerable

IOP, the number of normal perceived points increased by 10 (25%) eyes to 16, in the advanced stage by 10 (25%) eyes to 6, and in the terminal stage by 4 (10%) eyes to 2 points. In the process of dynamic observation in terms of 6-18 months. there was an increase in the area of cattle by 4 (10%) eyes in the terminal stage of infantile glaucoma. This research method cannot be objective due to the age of our patients and the lack of sufficient number of observations before surgery.

The dynamics of visual acuity, IOP, retinal OCT, and OD indicators in patients after surgery are reflected in Tables 2,3,4.

Table 2

Dynamics of visual acuity in patients with primary infantile glaucoma ($M \pm \sigma$)

Observation period Stages of glaucoma	Before the operation	Postoperative period		
		6 months	12 months	18 months
Developed n=10	0,1±0,002*	0,2±0,04	0,3±0,07	0,4±0,01
Far gone n=20	0,03±0,01*	0,2±0,09*	0,3±0,15	0,3±0,008
Terminal n=10	2 ranks	3 ranks	*3 ranks	4 ranks

Note. * – significant differences in indicators ($P \leq 0.05$)

n – number of eyes – visual acuity indicators below 0.005 are given in the ranks; which were not subjected to statistical processing.

Table 3

Dynamics of IOP (mm Hg) in patients with primary infantile glaucoma ($M \pm \sigma$)

Observation period Stages of glaucoma	Before the surgery	Postoperative period		
		6 months	12 mec.	6 months
Developed n=10	26,8±2,01	21,0±0,04	21,5±0,07	21,8±0,9*
Far gone n=20	28,8±1,01	20,6±0,09*	22,5±0,15	23,1±0,2
Terminal n=10	34,01±1,53	22,5±1,3	24,9±,09	25,2±2,1*

Note. * – significant differences in indicators ($P \leq 0.05$).

n – number of eyes

Table 4

Dynamics of morphometric parameters of the retina in patients with primary infantile glaucoma ($M \pm \sigma$)

Observation period Morphometric indicators	Development (n=10)		Fare gone (n=20)		Terminal (n=10)	
	Before the surgery.	18 months after the surgery.	Before the surgery.	18 months after the surgery.	Before the surgery.	18 months after the surgery
The area of the discal nerve, mm^2	2,31±0,2	2,33±0,15	2,26±0,5	2,31±0,14*	2,25±0,41	2,30±0,2
Excavation area, mm^2	0,76±0,36	1,05±0,15*	1,1±0,36	1,44±0,4	1,3±0,36	1,51±0,3*
The ratio of the excavation area to the area of the discal nerve	0,6±0,12*	0,62±0,06	0,64±0,1*	0,66±0,1	0,74±0,1*	0,77±0,09
The area of the neuroretinal ring, mm^2	0,82±0,12*	1,26±0,42	0,65±0,12*	0,82±0,33	0,45±0,12*	0,65±0,08*
Volume of the neuroretinal ring, mm^3	0,22±0,08	0,28±0,08	0,18±0,08	0,21±0,1*	0,18±0,09	0,20±0,2
Average excavation depth, mm	0,42±0,12*	0,31±0,2	0,72±0,12*	0,61±0,12	0,92±0,12*	0,89±0,4

Note. * – significant differences in indicators ($P \leq 0.05$)

n – number of eyes

The observation of 20 patients (40 eyes) for 1.5 years showed that in the surgical treatment of primary infantile glaucoma it is necessary to take into account the degree of goniodysgenesis and to use an adequate method to improve the aqueous humor outflow. Thus, in the advanced stage (isolated by trabeculodysgenesis), improvement of aqueous humor outflow through the uveoscleral pathway and under the conjunctiva was achieved by non-penetrating deep sclerectomy. In the advanced stage (anterior attachment of the iris) – aqueous humor, outflow from the anterior chamber into the supraciliary space due to sinusotrabeulotomy ab externo and sinusotrabeulectomy. At the terminal stage (posterior iris attachment), the outflow of aqueous humor

through the trabecular meshwork, the uveoscleral pathway into the intrascleral space and under the conjunctiva improved when using the method we proposed. Thus, using the above mentioned methods of surgery, we managed to reduce the number of intraoperative and postoperative complications by 12.5%, avoid repeated operations, reduce IOP in 12.3% of cases and increase visual acuity by 0.15 of the initial one.

Conclusion. The above methods of surgical treatment were chosen according to the degree of goniodysgenesis and the stage of the disease and showed satisfactory results in stabilizing the glaucoma process.

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ОПТИМІЗАЦІЯ МЕТОДИКИ ХІРУРГІЧНОГО ЛІКУВАННЯ ПЕРВИННОЇ ІНФАНТИЛЬНОЇ ВРОДЖЕНОЇ ГЛАУКОМИ У ДІТЕЙ

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Резюме

Дитяча глаукома – термін, що включає різні форми вродженої глаукоми та види вторинної глаукоми, різноманітні за етіологією та патогенезом, клінічною картиною. Хірургічне лікування є основним методом зниження внутрішньоочного тиску при глаукомі у дітей. На сьогоднішній день не запропоновано уніфікованого методу хірургічного лікування дитячої глаукоми, що робить пошук адекватних методів хірургічного втручання при цій патології найбільш актуальним.

Мета роботи – оцінити ефективність хірургічного лікування первинної вродженої інфантильної глаукоми у дітей.

Матеріал і методи. В очному відділенні клініки Ташкентського педіатричного медичного інституту проведена оцінка ефективності методу оперативного втручання у дітей від 3 до 10 років з первинною вродженою дитячою глаукомою. Обстежено 20 хворих (40 очей). З них 5 пацієнтів (10 очей) увійшли в пізню стадію, 10 пацієнтів (20 очей) увійшли в далеко запущену стадію і 5 пацієнтів (10 очей) увійшли в термінальну стадію відповідно. Пацієнти проходили стандартне офтальмологічне обстеження. Стан зорового нерва досліджували за допомогою оптичної когерентної томографії Stratus OCT-3000 (Carl Zeiss Meditec), полів зору – у 10 (20 очей) пацієнтів віком від 8 років за допомогою стандартної автоматизованої та комп'ютерної периметрії (SAP).

Висновки біоетичної комісії: У статті оцінено ефективність методу хірургічного лікування первинної вродженої інфантильної глаукоми у дітей. І наводить конкретні результати різних операцій на різних стадіях первинної вродженої дитячої глаукоми. Відповідно до ступеня гоніодизгенезу та стадії захворювання було обрано оперативне лікування, яке показало задовільний результат щодо стабілізації глаукомного процесу.

Для обробки результатів офтальмологічного дослідження використовували метод статистичного аналізу з використанням програм Microsoft Excel та SPSS. Відмінності між середніми значеннями ($M \pm \sigma$) вважалися достовірними при $P \leq 0,05$.

Результати і обговорення. Усім хворим проведено оперативне лікування залежно від вираженості морфометричних змін кута передньої камери, а в післяопераційному періоді з інтервалом 6 місяців – курс нейропротекторної терапії. За даними обстеження, дітям з первинною інфантильною глаукомою в пізній стадії виконано непроникаючу глибоку склеректомію на 10 очах; при далеко запущеній стадії – у 20 очах і при термінальній – у 10 очах – синусотрабекулотомія ab externo, синусотрабекулотомія. На 2 очах у термінальній стадії проводили фільтрацію за запропонованою методикою. Зазначені вище способи хірургічного втручання дозволяють зменшити кількість інтра- та післяопераційних ускладнень на 12,5%, уникнути повторних операцій, знизити внутрішньо очний тиск у 12,3% випадків, підвищити гостроту зору на 0,15 від вихідної.

Висновок. За ступенем гоніодизгенезу та стадією захворювання підбрано наведені методи хірургічного лікування, які показали задовільний результат щодо стабілізації глаукомного процесу.

Ключові слова: первинна вроджена інфантильна глаукома; хірургія; передньо-задня вісь ока; первинна вроджена глаукома; ригідність очей; екскавація диска зорового нерву; короткозорість; внутрішньоочний тиск.

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