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Surgical treatment of cataract and glaucoma in nanophthalmic eye – case report

Postępowanie chirurgiczne w przypadku zaćmy i jaskry u pacjenta z małooczem – opis przypadku

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Summary: Work presents surgical strategy in case of cataract and glaucoma in patient with simple nanophthalmus. Moreover optimal manner of calculation of the IOL and treatment in case of occurrence of complication from disturbed structure of eyeball. Surgical procedure included removal of cataract by using phacoemulsification, implantation of artificial Acri.Lyc Extreme lens, account from Haigis rule. Because of high intraocular pressure in early postoperative period we decided to implant Ahmed devices – children's type with drain placed in posterior chamber combined with posterior vitrectomy. We obtained stabilization of intraocular pressure and effective visual acuity.

Słowa kluczowe: jaskra, zaćma, małoocze, zastawka Ahmeda.
Key words: glaucoma, cataract, nanophthalmos, Ahmed valve.

Introduction

Nanophthalmus is a relatively rare disorder of eye structure always occurring bilaterally with the frequency from 0.046% to 0.11% (1). It can sporadically be inherited in a family – autosomally, recessively or dominantly (2). Nanophthalmus comprises a heterogeneous group of diseases which includes various anomalies of the anterior and the posterior segment. The following terms can be distinguished: the simple microphthalmus, or nanophthalmus, used in nomenclature as synonyms, the complex nanophthalmus, the partial nanophthalmus, the posterior nanophthalmus and the relatively anterior nanophthalmus. By the simple, or isolated, nanophthalmus we understand partial nanophthalmus without any other disorders in the structure of the eyeball or systemic disorders. Its primary feature is the shortened axial length of the eyeball (at least two standard deviations as compared to the norm for a particular age), without any accompanying anatomical deviations. In the simple nanophthalmus the horizontal diameter of the cornea should be below 11 mm on average (9.5-11 mm), the axial length of the eyeball – below 20 mm (14.5-20.5 mm), while the depth of the anterior chamber should be below 2 mm (1-2.7 mm), with refraction amounting to approx. +13.6 Dsph (+7.25-20.0 Dsph) (2,3,4,5,6). The lens is of the normal size or relatively larger, from 4.20 mm to 7.26 mm (6). The sclera is usually finer due to the abnormal collagen structure. The eye with the above structure is usually placed deeply in the normal orbital cavity. Due to a relatively larger size of the lens, the method which can be chosen for the treatment of the cataract in this kind of eye is a small incision phacoemulsification. The simple nanophthalmus is usually accompanied by the closed angle glaucoma, cornea guttata and the pseudoexfoliation syndrome. The term "complex nanophthalmus" means nanophthalmus with other anatomical disorders including the coloboma of the iris, the choroidea and the retina or the optic disc dysplasia. The relatively anterior nanophthalmus is characteristic for an eyeball with normal axial length and a disproportionally small anterior segment of the eye (3). The horizontal diameter of the cornea in this kind of eye amounts to 11 mm ± 0.34 mm or less, there are no morphological deviations, the length is above 20 mm ± 0.94 mm, the depth of the anterior chamber is below 2.2 mm ± 0.9 mm, and the thickness of the lens – 5.05 mm ± 0.45 mm (7). In this group the most common disorders are glaucoma, cornea guttata, the pseudoexfoliation syndrome, posterior synechiaes, while post-surgically there is a partial oedema of the cornea and the glaucoma malignant (7). The posterior nanophthalmus is characterised by a disproportionately small posterior segment of the eye, while the anterior segment is of normal size. In the case of a cataract surgery, the nanophthalmus is connected with an increased risk of the occurrence of intraoperative and postoperative complications such as a suprachoroid haemorrhage, the shallowing of the anterior chamber, an incarceration of iris, a malignant glaucoma, difficulties in stabilising the intraocular pressure (IOP), an exudative detachment of the retina or the choroidea and a long sustained oedema of the cornea.

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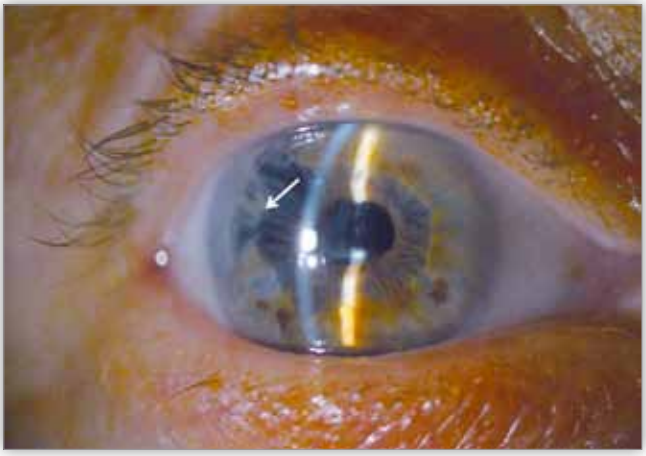
Case study

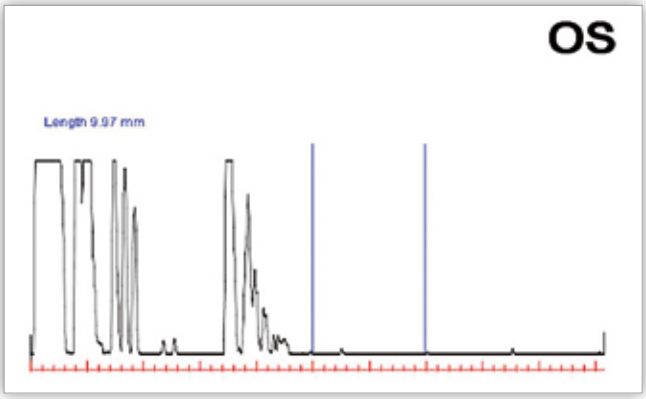
The patient, 60 years old, male, referred from the regional ophthalmology ward to the Eye Hospital of theUniversity in Lublin, due to the primary closed angle glaucoma of both eyes, nanophthalmus of both eyes, the complicated cataract of left eye (LE), the aphakia of right eye (RE), a high IOP, as well as the decline of the visual acuity in the LE. The general interview showed no contraindications. Due to a similar structure of the eye of the patient's sister, the case was qualified as the simple nanophthalmus which sporadically occurs within a family. The patient underwent a surgery in 2008 due to the cataract of RE , at another ophthalmology ward. During the surgery the lens could not be implanted due to its too large diameter in comparison to the anatomic conditions of the eyeball. The clinical examination showed the best visual acuity (VA) in the right eye of light perception without localization. Any correction did not improved VA. The visual acuity in LE was 0.3 with correction +12 Dsph. The autorefractometry was impossible to perform.

Ryc. 1. Nanophthalmus – the patient's eyes deeply set in the orbit.
Fig. 1. Nanophthalmus – oczy pacjenta głęboko osadzone w oczodołach.



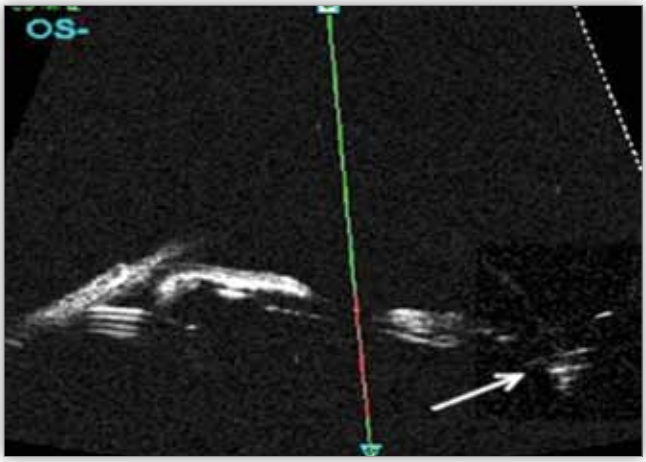
Ryc. 2. RE – anterior segment, anterior synechia.
Fig. 2. OP – przedni odcinek, zrost przedni.

Javal's keratometry in the RE amounted to K1 = 48, K2 = 48.5, while in LE respectively K1 = 48.5, K2 = 49. The cornea diameter measured by means of a compass amounted to: RE = 10.5 mm, LE = 10.6 mm. The visual field in LE narrowed to 6°, in RE impossible to define. The intraocular pressure in RE



Ryc. 3. USG – projection A – LE. ALX = 17.04 mm.
Fig. 3. USG – projekcja A – OL. ALX = 17,04 mm.

Ryc. 4. LE – anterior segment after operation TPPV+Phaco+ILCP+ Ahmed implant +irydectomy.
Fig. 4. OL – przedni odcinek po operacji TPPV+Phaco+ILCP+implant zastawki Ahmeda+irydektomia.



Ryc. 5. UBM LE after operations, arrow – tube of the Ahmed Vavle placed in the posterior chamber.
Fig. 5. UBM OL stan po operacji, strzałka – dren zastawki Ahmeda umieszczony w komorze tylnej.

was at the level of 16 mmHg, and in LE in the period of several months it varied from 26 mmHg to 40 mmHg. The patient received Xalacom and Pilocarpine to the left eye. The patient's eyes were visually small, placed deeply in the orbital cavities (Fig. 1). In the RE the presence of posterior adhesions, the atrophy of the iris and aphakia were observed (Fig. 2). In LE the shallowing of the anterior chamber, the filtration angle closed, posterior synechiaes were found. The A-scan ultrasound biometry revealed a short eyeball with axial length amounting to 17.11 mm in RE and 17.04 mm in LE (Fig. 3). The depth of the anterior chamber in both eyes was of 1.8 mm. The power of the lens was calculated on the basis of the Haigis formula. The patient was qualified for the surgery of cataract removal by means of the phacoemulsification method with an implant in the posterior chamber in LE. The surgical strategy: in the peribulbar anaesthesia a corneal tunnel at 11^{-th} hour, a paracentesis, a synechiolysis, a phacoemulsification of the cataract. A posterior-chamber Acri.Lyc Extreme lens was implanted with the power of 45 D and diameter 9.8 mm. In the early post-operative period a high intraocular pressure was observed. The paracentesis was performed. Due to the lack of improvement it was decided to place an Ahmed implant of the type designed for children in the upper temporal quadrant, with a drainage in the posterior chamber combined with a vitrectomy (Fig. 4, Fig. 5). An iridectomy was also carried out. At present the visual acuity in LE amounts to 0.4 with correction -1.0 Dsph. A refraction was impossible to perform. The intraocular pressure in the RE OP 19 mmHg and in the LE 19 mmHg. At present the patient does not receive any drugs.

Discussion

Prognostic and therapeutic problems resulting from the abnormal structure of the eyeball are related both to methods of measuring the eyeball, the choice of a calculation method for the lens, techniques of carrying out the procedure as well as the treatment in the case of an increased intraocular pressure or other postoperative complications. When measuring the length of the eyeball it needs to be remembered to change the calibration of the ultrasound probe which is set for the normal eye length. The measurement of the length of the eyeball in the operated patient was performed with the use of the immersion method. The advantage of this method is also the possibility to avoid flattening the cornea, which allows for a more precise measurement (8). The diameter of the cornea should also be taken into consideration since the effectiveness of the implant depends on its location, i.e. directly on the anatomic conditions of the eye before the surgery. The recommended methods of calculating the lens in microphthalmus are the following formulae: the Holladay–II, the Hoffer-Q and the Haigis formula (8,9). For calculating the lens, the Haigis method was used. The refraction error in this case amounts to $+0.51$ D $\pm$ 0.12 D. The Haigis formula can induce postoperative myopia more often than other methods. However, many authors claim it is the most appropriate in the case of a shortened length of the eyeball (9,10). Other formulae increase the refraction error: SRK/T $=$ -1.45 D $\pm$ 0.14 D, Hoffer Q $=$ -0.7 D $\pm$ 0.14 D and Holladay 1 $=$ -1.11 D $\pm$ 0.13 D (10). In the case of short eyeballs, postoperative hypermetropia is most common. In our case, the measurement error was within the error margins given above, taking into consideration the best corrected visual acuity of the patient, which before the cataract amounted to 0.3. Another important aspect is the choice of the intraocular lens, taking into account the available strength, the size of the haptic part as well as the size of the whole lens. The diameter of the optical part usually should be below 5.5 mm, while the diameter of the whole lens – below 12 mm. Here the posterior-chamber Acri. Lyc Extreme lens was used (dioptric power 45 D, diameter of the whole lens 9.5 mm). In the right eye the attempt to implant a standard size lens was not successful. In the preoperative procedure some authors suggest administering 40 mg prednisolon orally, 500 mg acetazolamid intravenously and 20% Mannitol in an enema (1). Our patient received 500 mg acetazolamid and 20% Mannitol directly before the procedure. Prednisolon was not used. The decision to remove the lens with the use of the phacoemulsification method was made due to the high fluctuation of intraocular pressure and the low effectiveness of pharmacological treatment. The procedure is recommended by many authors as a safe and effective method of lowering the pressure in a short eyeball (11). The surgical strategy did not differ from the standard treatment in the case of simple cataract. In the literature it is recommended to apply topical or general anaesthesia due to the possible increase of the IOP when the peribulbar anaesthesia is applied (1,7). Despite using the peribulbar anaesthesia, the patient did not demonstrate any increase in the intraocular pressure during the surgery. Shugar suggests a cut in the clear cornea from the temporal side due to a relatively deep orbital cavity (1). Taking into account the more frequent damage or the partial lack of the ciliary zonule, a capsulorexia should be performed as in the subluxed cataract.

The fluctuation of the IOP constitutes a huge problem after the surgery of the cataract in the case of a short eyeball. In the early postoperative period we observed a rise in the IOP in the operated patient. Due to the lack of improvement after pharmacological treatment, the decision was made to place an Ahmed implant of the type designed for children in the posterior chamber combined with a vitrectomy. The drainage was placed in the posterior chamber since the anterior chamber was very shallow and the risk of cornea decompensation was high. A complete vitrectomy, in turn, was necessary for the proper functioning of the valve, maintaining the patency of the drainage as well as for restoring the normal flow of the aqueous humour. Taking into consideration both our own experience and the one of other authors, we claim that carrying out the trabeculectomy in the case of a shortened eyeball does not give the expected result of lowering the intraocular pressure. To the contrary, the shallowing of the anterior chamber and the malignant glaucoma occur more often than after other antiglaucoma procedures (12). First studies on the surgical procedures in the case of nanophthalmus brought not very optimistic results (12). The standard trabeculectomy in 60% of cases was unsuccessful and ended in the loss of vision in 86.6% of cases (12). After this procedure an exudative detachment of the choroidea and retina was more frequent, according to some authors even in 50% of operated patients, which was the main reason for the vision deterioration (12). The presence of a disturbed structure of collagen of the sclera also seems to be of considerable im-

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portance, the result of which is a decrease of its flexibility, an increase of the resistance of the aqueous humour flow and, at the same time, an improper functioning of the cusp.

In the literature there are no descriptions of the cases relating to implanting the Ahmed valve in the case of nanophthalmus. However, a drainage through the pars plana is recommended in patients with a very shallow anterior chamber, especially in the occurrence of a circular shallowing of the anterior chamber, in patients after a penetrating keatoplasty, as well as in the case of glaucoma, additionally accompanied with disorders of the vitreous body. Among the complications which may occur after such a procedure are detachment of the retina in 6% of cases, occlusion of the drainage due to left over the vitreous body – 9%, the occurrence of a preretinal membrane – 9%, a cystic oedema of the macula – 3% (13). Other procedures recommended by Jünemann and co-authors include, depending on the cause of the increase of the IOP, are cyclocriotherapy, vitrectomy, surgical iridectomy, or YAG iridectomy (14). When it comes to the cyclocriotherapy procedure suggested by Jünemann, it was performed only on the eyes of blind people, the vitrectomy – in the malignant glaucoma, while the surgical iridectomy and laser iridectomy was carried out in the primary stage of glaucoma (14). The vitrectomy alone can be effective in the case of glaucoma in patients with short eyeballs, however, there are no long term observations of the intraocular pressure in such patients. The vast experience resulting from the frequent use of Ahmed valves in different stages and types of glaucoma enables us to make an assumption that also in the case of a patient with nanophthalmus the intraocular pressure can be reduced to a satisfactory level. Presented patient was practically one-eyed, with an advanced optic neuropathy in the operated eye. The risk of deterioration or the loss of vision could rise after carrying out further surgical procedures, therefore we chose what in our opinion seemed to be the most effective and relatively safe surgical procedure, which turned out to be a new effective method of treating secondary glaucoma in a patient with nanophthalmus.

The surgical procedure in such a patient requires a special preparation to the planned surgery, a vast experience of the surgeon as well as the awareness of the possible complications of the surgical intervention. The availability of the appropriate type of lenses and improved surgical techniques made it possible to qualify the patient with nanophthalmus for the surgical procedure and, what is more, to achieve a usable visual acuity and stabilisation of the intraocular pressure.

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