

SURGICAL SOLUTIONS AND ETHICAL DILEMMAS IN CHILDREN SUFFERING FROM DOWN'S SYNDROME AND CONGENITAL CATARACT

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Abstract: Congenital cataract is a transparency anomaly of the lens present at birth. Statistics in literature report a frequency of about one case in 2000 newborns, most of which have visual disturbances. Along with systemic anomalies, children suffering from Down's syndrome can also have ocular modifications, including congenital cataract.

Surgical approach in congenital cataract is much more complex than in adults because congenital cataract occurs in the process of natural development of the visual function. Relying on our experience, shared by most ophthalmologists, we believe that per primam implantation of an IOL in the posterior chamber is a rule in the surgery of congenital cataract as it ensures optimal conditions for the treatment of amblyopia and yields the best visual results. However, the selection of the IOL power that is based on rough biometrical formulas, is still unpredictable.

Therefore, up to two years of age, surgical aphakia in bilateral congenital cataract remains an acceptable alternative. In clinical practice we encounter borderline cases, between surgical pseudophakia and aphakia, whose complex ethical implications are shown in the paper below. We present the conclusions of our investigation carried out on a group of 14 children suffering from Down's syndrome and congenital cataract, i.e. on 26 eyes operated at the Clinical Emergency Eye Hospital Bucharest between 1.01 2010- 1.01 2020 as well as two illustrative cases. We also bring forward the ethical issues, e.g. the benefit for the patient, non-maleficence, informed agreement, standardization of the procedures when we choose pseudophakia or aphakia, with the aim at achieving the best result.

Key words: congenital cataract, Down's syndrome, pseudophakia, aphakia, moral dilemmas.

INTRODUCTION

Surgical treatment of congenital cataract, that requires clearing the visual axis as a first step, is a challenge for the eye surgeon who needs to be well acquainted with the peculiarities of child's eye. A good visual function is obtained with difficulty and over time, after a long cooperation with the toddlers and their families, as children's visual system develops continually up to 10-12 years of age [1]. It is essential for their families to know and understand the benefits of the surgery. Families should be correctly informed about the potential visual outcomes which may not reach their expectations, if the timing for a successful surgery is not adequate or the treatment of postoperative amblyopia is not tenaciously instituted.

The ophthalmologist often needs to face difficult therapeutic decisions in pediatric patients with Down's syndrome and congenital cataract that requires surgery. Regaining transparency of the visual axis, i.e. the extraction of the cataracted crystalline lens under general anesthesia results in surgical aphakia. This needs to be corrected by per primam implantation of an IOL, an attitude regarded as "gold standard", associated with spectacles or contact lenses [2]. In order to get the best visual function, essential and compulsory surgery (when there are no ocular contraindications, e.g. microphthalmia, congenital glaucoma, absence of photomotor reflex) should be followed by treatment of amblyopia [3]. The treatment of amblyopia takes a long time and requires very good cooperation between children and their families and the ophthalmologist [4]

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and should be initiated as soon as possible after surgery.

We share the same opinion with most of our colleagues [5] that per primam implantation is beneficial for pediatric patients with Down's syndrome even younger than 2 years of age, ensuring satisfactory vision for these children that have major difficulties in handling spectacles or contact lenses. Such patients can have single or multiple [6] systemic and/or eye anomalies. In our study group there were 14 children, i.e. 26 eyes; twelve patients had bilateral and 2 unilateral cataracts. All patients were hospitalized and operated under general anesthesia at the Clinical Emergency Eye Hospital Bucharest between 1.01.2010 – 1.01.2020 by the same eye surgeon. The children presented a number of systemic anomalies (delayed psychomotor development – 14 cases, congenital heart malformations – 9 cases, thyroid disorders – 4 cases, gastro-intestinal malformation – 1 case, obesity -7 cases, obstructive apnea – 1 case) as well as associated eye anomalies (significant anisometropia, important refractive errors, strabismus, nystagmus).

The evaluation of the surgery/benefit ratio (improved visual acuity) was professionally and ethically difficult. Although the selection of the adequate IOL dioptric power is still controversial, per primam implantation is the surgical approach that seems most beneficial [7] to the pediatric patients with Down's syndrome; however, there are cases in which aphakia is the most successful approach [8]. In supporting our point of view, we report two clinical cases from the study group, one patient in which per primam implantation was performed and another in which aphakia was the best approach. Details regarding professional and ethical therapeutic decisions are given.

CASE PRESENTATION

Case 1 is a 16-year old male with congenital bilateral cerulean cataract (Fig. 1), pathological myopia and Down's syndrome (psychomotor delay, bicuspid aortic valve). On hospitalization the patient had a BCVA (best corrected visual acuity) OD= 0.15 and BCVA OS= 0.2, refraction: OD: -9.50/-4.00 x 11 and OS: -8.25/-1.75 x 85. Surgery with per primam implantation under general anesthesia [9] is performed in the weakest eye with highest sphericity, i.e. the right eye. After surgery BCVA OD = 0.3 (with a correction of -2.20/-2.25 x 151, SE= -3.25); the patient and the family are satisfied with the visual outcome. Then surgery of the left eye is performed; a personalized toric IOL for the correction of the astigmatism (Fig. 2) [10] (identified by biometry) was required.

Surgery entailed a new, extremely careful general anesthesia after the cardiological evaluation as the patient had bicuspid aortic valve. During surgery, following the implantation of the IOL within the posterior chamber, the absence of half of one of the IOL haptics is observed, a fact that makes its positioning impossible. An unexpected professional and ethical dilemma arises: should we remove the IOL by enlarging the incision to 5.5 mm, which would induce a high astigmatism, although the surgeon's initial intention was to correct the patient's preoperative astigmatism by using a personalized toric IOL?

Neither correction by spectacles can be taken into consideration because of the important anisometropia (IOL implanted), nor contact lenses because the patient is unable to deal with them (delayed psychomotor development). The surgeon chooses to

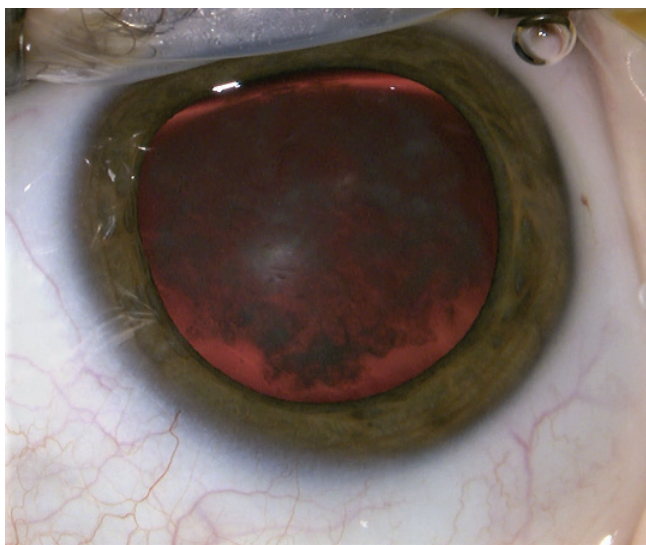


Figure 1. Cerulean cataract.

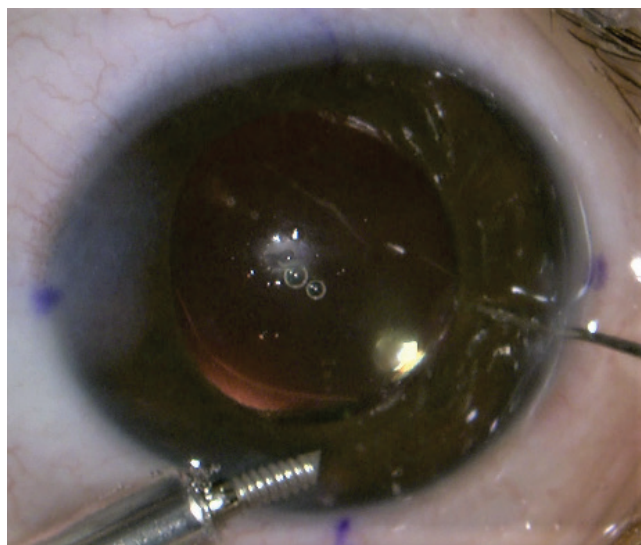


Figure 2. Intraoperative image of the toric implant.

keep this personalized IOL implanted and places it with the optical part behind the posterior capsulorhexis and the haptics within the sulcus, a daring solution that requires an experienced surgeon. The idea was that postoperative capsular fibrosis [11] would stabilize the IOL in the course of the following weeks.

After surgery BCVA OS =0.5 (with a correction of -2.25/-0.75 x 93). The patient and family are satisfied with the visual outcome. Five weeks later the patient is reevaluated and a decreased vision in OS is identified, because of the dislocation of the posterior chamber IOL (following a trauma). What approach to take in order to obtain an optimal visual result for the patient? Should we reposition the IOL with the broken haptic or remove and replace it [12] with a non-toric 3-pieces sulcus fixated IOL (toric variant of the implant within the sulcus was not available at the time). The surgeon chose the first approach that ensures good vision for the patient. After repositioning the IOL, the postoperative BCVA OS =0.5 and the patient and his family are satisfied with the result. Four weeks later we found that the IOL with broken haptic was displaced again (slit lamp examination). It was removed [13] through an incision of 5.5 mm and a new IOL was implanted in the sulcus (3-pieces IOL, 11D). The functional result is poorer than in the initial approach but stable in time which is very important in a child suffering from Down's syndrome.

Case 2 is a 17-year old female with pathological myopia, congenital nystagmus, bilateral congenital cataract – OS cerulean, OD total (Fig. 3), Down's syndrome with marked mental retardation and atrioventricular septal defect (general anesthesia not recommended) [14]. Preoperative biometry to

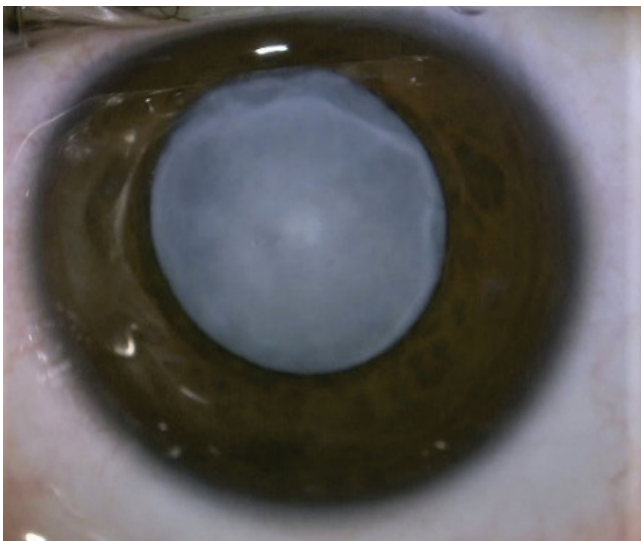


Figure 3. Total cataract.

estimate dioptric power of the IOL was not possible (very difficult cooperation with the patient, nystagmus present). Consequently, we chose to operate without implantation of an IOL and approximation of diopters, functional results being somewhat uncertain anyway. Postoperative refraction OD: -4.00/-0.50 x 67, i.e. good near vision, the patient was to wear spectacles as she was already used to wearing them. The patient and her family are satisfied with the visual outcome. The patient has not come in yet for the surgery of the left eye.

DISCUSSION

Down's syndrome patients might suffer from single or multiple eye anomalies, including congenital cataract along with systemic anomalies. Therefore, they are sensitive patients in which difficult therapeutic decisions are required, which entail ethical dilemmas more complex than in pediatric eye surgery in general.

The opinion we share with other specialists is that elective surgical approach in these patients should be one that has minimal postoperative complications. This implies performing an anterior capsulorhexis, a posterior one equal in size and a limited anterior vitrectomy to remove the anterior hyaloid. These surgical maneuvers are difficult to carry out on children's eyes, smaller in size and with an increased elasticity compared to adult eyes; the surgeon should be very experienced.

The most common complication is the re-opacification of the visual axis [15], which entails a new surgery under general anesthesia – a fact that rises many problems in these patients suffering from multiple systemic anomalies, especially cardiac diseases. Per primam implantation of an IOL is greatly beneficial, other means of correcting aphakia being very difficult. Implantation is, in its turn a very delicate maneuver, whether we perform the implantation of the IOL within the posterior chamber with its optic part behind the two capsulorhexes or as in BIL (bag in the lens) technique [16]. Implantation might involve inadequate correction because of the difficulties in selecting the suitable dioptric power, given the still controversial biometric formulae and complexity of the techniques. In children suffering from Down's syndrome, biometry yields uncertain results because of the frequent association with other eye anomalies and cooperation issues.

Aphakia is a potential solution, mostly in bilateral congenital cataract, but postoperative amblyopia management is more difficult and the visual results might be poorer.

Congenital cataract in children with Down's syndrome rises many dilemmas.

A professional dilemma regarding per primam implantation of an IOL (less predictable when the child is very young and there is a risk of inadequate choice of diopter) or aphakia with its multiple disadvantages.

The ethical dilemma regarding the benefit of the patient (implantation per primam with its advantages of easier and reliable treatment of amblyopia) and the principle of non-maleficence (surgical aphakia in order to avoid postoperative complications such as inadequate diopter and unpredictable refraction) is present at varying levels in children suffering from Down's syndrome, varying with the type of congenital cataract, in one or both eyes. When the benefit is not very clear, the procedure should at least not be harmful [18]. Hippocrates: "I will act for the good of my patients ... and never do harm to anyone" (Dorland's medical dictionary).

In bilateral congenital cataract the benefit for the patients means to use surgical procedures that should improve visual acuity in both eyes, so as to obtain stereoscopic vision that is of utmost help in the psychomotor development of such children. This approach satisfies the medical purposes of reaching a correct diagnosis and restoring the patient's health. The principle of non-maleficence in these cases means avoiding important and untreatable amblyopia.

In unilateral cataract the benefit for the patient is somewhat less obvious, as treatment of anisometric amblyopia may prove to be unsuccessful. Non-maleficence means to use safe and uncomplicated surgical procedures so as to avoid a new surgery under general anesthesia, a condition that is also satisfied by aphakia.

A correct decision is sometimes rather complicated ethically because we always want to obtain a long-term good visual acuity in both eyes and the evaluation of major complications that may endanger functional vision is always a great challenge. Per primam implantation is always an elective approach, both in bilateral and unilateral cataract; this medical technique implies the performing of a continuous, curvilinear capsulorhexis (Case 1). Per secundam implantation is not desirable, because we want to avoid another general anesthesia in such sensitive patients.

There is a risk of inadequacy of the IOL, but in most instances acceptable aerial corrections can be used. Aphakia in bilateral cataract can sometimes be acceptable, i.e. in high myopia, the patients obtaining a good near vision (Case 2). It is all about professionalism:

honesty and integrity, trust and responsibility, respect and compassion, professional improvement, knowing one's limits, cognition and cooperation, altruism and support [19].

A deontological dilemma may appear between standard procedures supported by most ophthalmologists - The health of my patient is my duty - Declaration of Geneva 1947 and "A physician shall act in the patient's best interest when providing medical care", International Code of Medical Ethics, World Medical Association 1949, in which the patients' major interests are stipulated. Standardization of therapeutic procedures reduces variability, increases patients' safety and improves the therapeutic results [20]. The major interest of patients in the above-mentioned cases is to get a good long term near and far visual acuity in both eyes and avoid important refractive postoperative amblyopia. In order to reach this result, the patients should be subject to low risks thus avoiding negative influence on their psychomotor development.

In the first case implantation of a personalized IOL in the left eye was chosen, although half of the haptic was missing; this approach was possible due to the surgeon's ability who made an implantation in the sulcus and captured the optic behind the capsulorhexes, avoiding explantation. The patient's major interest was satisfied (best visual acuity by correcting myopia and astigmatism) with minimal risks. Visual outcomes confirm the therapeutic solution, the family and the patient being satisfied. Unfortunately, child's behavior entailed repeated eye accidents which led to the removal and replacement of the personalized IOL, with a poorer functional result. The question arises whether it would have been more beneficial for the patient to resort to the removal of the IOL with the broken haptic from the very beginning and aphakia with per secundam implantation, although the specialist knows that the functional results would be below current therapy; the patient suffered another 2 surgeries performed under general anesthesia, thus ascertaining the truth that "the best is the enemy of the good" (Voltaire).

In the second case the specialist chose aphakia because biometry could not be done and the IOL diopter could not be reasonably estimated, thus the final result being highly unpredictable. The patient's interest was to have a reasonably good vision and this aim was achieved. Postoperative refraction of -4D enables good near vision whilst for distance vision the patient is using a significantly lower correction; the patient and the family are satisfied with the result.

According to professional independence,

the doctor is allowed to make personal therapeutic decisions, based on standards, rules, recommendations and their own experience and competence. The above mentioned cases illustrate this principle, every doctor being free to solve uncommon situations, not yet encountered in guidebooks [19]. These decisions must be presented to the patient in order to get his/her informed consent, in our cases the patients' parents. Parents' written approval of the procedure is compulsory, its absence is illegal. Informed consent does not diminish the doctor's professional responsibility. Patient's autonomy, i.e. his/her right to informed consent is a modern principle that goes along with social justice.

Patients autonomy alters neither the difficulties of the professional decision nor legal or social responsibilities [21]. Complete information of the patient on the surgery should include both the advantages of the procedure as well as the potential risks and complications [4]. This is an honest and transparent process given the fact that the family expects not only improvement of visual acuity but also progress in the psychomotor development of the child. We also informed the parents on our decision to use innovative technologies, the capsulorhexis by femtosecond laser and implantation of a new type of personalized toric IOL. A new issue arises, i.e. our responsibility of ascertaining the safety and benefit of innovative surgery for the patient before using it in everyday practice.

Ethical issues arise when these clinical instances are part of clinical research groups, with the observation of anonymity and confidentiality – essential in the relationship between patient and physician. The answer is far from simple, given the fact that randomized clinical studies are the most effective method of evaluating new surgical procedures and stability of artificial lenses. The analysis of these two cases of surgery on congenital cataract offers encouraging solutions in the use of new personalized toric IOL in patients in which biometry is not possible, i.e. Down's syndrome patients. In order to use new surgical procedures in pediatric patients, including the disabled ones, the doctor is supposed to think over quite a number of ethical principles and also reflect on questions like, i.e. is that procedure actually used in practice or is it only mentioned in research protocols? The physician is responsible for the use of innovative procedures in current practice and procedures insufficiently tested should be considered risky for the patient [22]. "The newest and most innovative procedure" may seduce

the patient and family but the specialist should be very careful in presenting the procedure in these terms.

In conclusion, in our opinion the surgical treatment of congenital cataract, including in patients suffering from Down's syndrome is still a professional and ethical challenge for the eye surgeon. These dilemmas can be worked out and we conclude that in Down's syndrome patients the per primam implantation of an IOL is the ideal approach, yielding optimal conditions for the treatment of amblyopia, as these children have difficulties in using spectacles or contact lenses. Per secundam implantation should be avoided because there are risks entailed by general anesthesia, given the associated systemic anomalies that might require surgery. In special situations (high myopia, bilateral or associated congenital cataract when biometry is not possible because of the patient's lack of cooperation) aphakia is preferred. In Down's syndrome patients suffering from congenital cataract, the major aim is to get long-time improvement of visual acuity, with further influence on their psychomotor development.

Conflict of interest

The authors declare that they have no conflict of interest.

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