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Pediatric Cataract

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Introduction

A study in Sweden showed the incidence of all congenital cataract cases to be 36/100,000 (Abrahamsson et al. 1999). In addition, a few hundred children develop juvenile cataract each year in Sweden. Congenital cataract is considered to be the most common cause of treatable blindness in children. It is present at birth and may be unnoticed until the visual function is affected or a whitish pupil is seen. If the babies do not have surgery quickly they develop irreversible amblyopia. Pediatric cataract surgery is nowadays an increasingly safe procedure, although there are some complications to the surgery. Visual axis opacification is the most common complication, which threatens the vision again and can lead to amblyopia if not managed. Secondary glaucoma is the most feared complication and can lead to blindness and a cosmetically disturbing eye. Developmental cataract, which is not dense at birth, is more common and could be operated on much later. This would lead to fewer complications and a better outcome.

Congenital cataract

Congenital cataract is hereditary in approximately one third of the cases. It is often inherited autosomal dominant but can also be inherited autosomal recessive or X-linked. In approximately one third of the cases other diseases can be found. Metabolic disorders, such as galactosaemia and hypocalcaemia are rare causes. Intrauterine infections such as rubella, toxoplasmosis, herpes, varicella and syphilis can cause congenital cataract. Genetic syndromes such as trisomy 21 or Turner's syndrome and a variety of neurological disorders are often associated with congenital cataract. Other ocular anomalies such as iris coloboma,

aniridia, microphthalmia, retinopathy of prematurity, or persistent foetal vasculature (PFV) are often combined with cataract. In the rest of the patients, the congenital cataract is idiopathic. In unilateral cases, the cause is most often idiopathic and in a clinically healthy child, or if the cataract is inherited, there is no need for an extensive pre-operative evaluation. 5-20% of childhood blindness worldwide is caused by cataracts.

There are different types of cataract; nuclear, lamellar, sutural, polar, lenticonus, membranous and those associated with PFV. The size, density, laterality of the cataract and the presence of associated ocular abnormalities decide how strong the indication is for surgery. The more central and the more posteriorly located, the more visually significant the cataract will be. Since there are more complications after early surgery as discussed below, it is important to wait if the cataract is not visually significant.

In cases with dense congenital cataract the cataract surgery must be performed early to prevent irreversible amblyopia and nystagmus. At the same time the risk of secondary glaucoma increases in these very small children, the earlier the surgery is performed. It is therefore very important to find ways to reduce the risk for secondary glaucoma. Also, the visual axis opacification formation is much more pronounced in the youngest children.

Secondary glaucoma

The earlier the surgery is performed, the greater the risk of secondary glaucoma. The risk also seems to be greater in small eyes, as in microphthalmus with persistent fetal vessels. It is yet not clear what causes the secondary glaucoma. However, an IOL seems to decrease the risk. Arsani et al (Asrani et al. 2000) found a much higher rate of glaucoma following cataract surgery in patients who were left aphakic (14/124 patients), than if they were implanted with an IOL (1/377 patients). They also reviewed the literature and found no reported case of open-angle glaucoma in the over one thousand pseudophakic patients from the studies. In one of our studies, no eye out of 31 children implanted with a small acrylic SA30AL IOL at 2-28 months of age developed secondary glaucoma (Kugelberg et al. 2006). In a rabbit study we performed with 20 three week old rabbits, no eye implanted with an AcrySof SA30AT IOL developed secondary glaucoma, but three aphakic eyes did during the follow-up time. However, in other animal studies with different IOLs the frequency of secondary glaucoma was similar in aphakic and pseudophakic eyes. It might also be due to the increased inflammatory response in young children and infants, compared to older children.

Treatment of postoperative aphakia

Implantation of an IOL is now a common and accepted management of the postoperative aphakia even in the smallest children. For the very small eye of an infant most of the commercially available IOLs are too large. The myopic shift that occurs in the child's growing eye is also a great concern. Lately, after surgery for unilateral cataract, many surgeons implant an IOL also in infants. However, for optical correction in bilateral cases with congenital cataract, contact lenses are probably still most often used in the Western World. Contact lenses can cause infection, are sometimes hard to handle, tedious for the family and expensive. Compliance is often also poor regarding the use of contact lenses. Therefore it would be of great interest if it were possible to develop an IOL that fits the small eye. We have performed some studies evaluating different smaller lenses in a rabbit model and in small children. They seem well tolerated by the small eye.

Visual axis opacification

Visual axis opacification or after-cataract occurs when lens epithelial cells (LECs) migrate and proliferate from the anterior capsule and the equator of the lens capsule, onto the posterior capsule. The visual axis is then obscured, and vision blurred again. Children develop more and faster visual axis opacification than adults. This opacification, or posterior capsule opacification as it is called in adults, can be removed in a second procedure in adults, using Neodymium: YAG (Nd:YAG) laser. An opening is then created in the posterior capsule, and the vision is clear again. In most cases, visual axis opacification in children can not be removed with only Nd:YAG laser, because the LECs will continue to grow on the anterior vitreous surface. The opacification has preferably to be removed in a second surgical procedure with anterior vitrectomy, often via the pars plana. However, in the very small children the LECs can grow also on the posterior surface of the IOL, even after a vitrectomy is performed.

To diminish visual axis opacification in children, most cataract surgeons perform posterior capsulorhexis at surgery. It is also debated whether or not to do an anterior vitrectomy at primary surgery. It could be performed through the pars plana, or through the anterior chamber after the posterior capsulorhexis and implantation of an IOL. When the anterior vitreous has been removed, the LECs most often cannot grow on the remaining vitreous. It

seems that anterior vitrectomy is necessary at least in children below 5 years, to avoid rapid visual axis opacification.

Another surgical technique that has been studied is a so called optic capture, i.e. the IOL is pushed through the posterior capsulorhexis, while the haptics remain in the bag. However, the technique does not seem to fully prevent the formation of visual axis opacification, and it has been described that the anterior vitreous face became semi-opaque and that the LECs can grow also on the anterior surface of the IOL. However, optic capture might be a suitable technique in some instances, since it provides a good centration of the IOL, which is necessary in cases after trauma or an incomplete rhexis.

Removal of residual LECs during the primary surgery is a key factor in avoiding posterior capsule opacification. Several chemicals have been suggested in experimental settings to remove or destroy residual LECs, but it has to be kept in mind that the substances may be toxic to other ocular structures. Research is directed towards finding a device or substance that can selectively remove the LECs. There are always complications associated with touching the vitreous and breaking the posterior lens capsule. If this can be avoided, it would be highly advantageous. This however, generates the need for a lens capsule with no remaining LECs. Perhaps a sealed-capsule irrigation could be an option at least in pediatric patients.

Perfect Capsule

The sealed-capsule irrigation device, Perfect Capsule, invented by Anthony Maloof (Agarwal et al. 2003), consists of a silicone plate, 0.7 mm thick, 7 mm in outer diameter, and 5 mm in inner diameter. It has an inflow tube, an outflow tube, and a tube for creation of a vacuum. The device can be folded and introduced through a normal small incision. After the lens extraction, the device is placed over the anterior capsulorhexis, and a vacuum is created with a syringe. This produces a sealed system between the capsular bag and the device. The capsule can be irrigated with a substance through the inflow tube, and the substance does not come in contact with the other intraocular structures. The substance is washed out with balanced salt solution (BSS), the vacuum is released, and the device is withdrawn. An IOL then can be placed in the bag.

Human studies have reported the safety of the device in adults undergoing cataract surgery. In that study, the sealed system was irrigated with distilled deionized water, which did not prevent posterior capsule opacification development. Earlier in vitro studies had shown that the LECs succumbed from osmotic lysis when they were exposed to distilled deionized water. However, this evidently did not work in vivo.

We evaluated the Perfect Capsule in a young rabbit model. Our experiments showed that 5-fluorouracil 50 mg/ml was the most effective substance evaluated to prevent visual axis opacification (Abdelwahab et al. 2008) (Fig. 1 D). We also investigated a substance called thapsigargin, which has proven effective in in vitro studies of human LECs, but the substance was not effective in our model (Fig. 1).

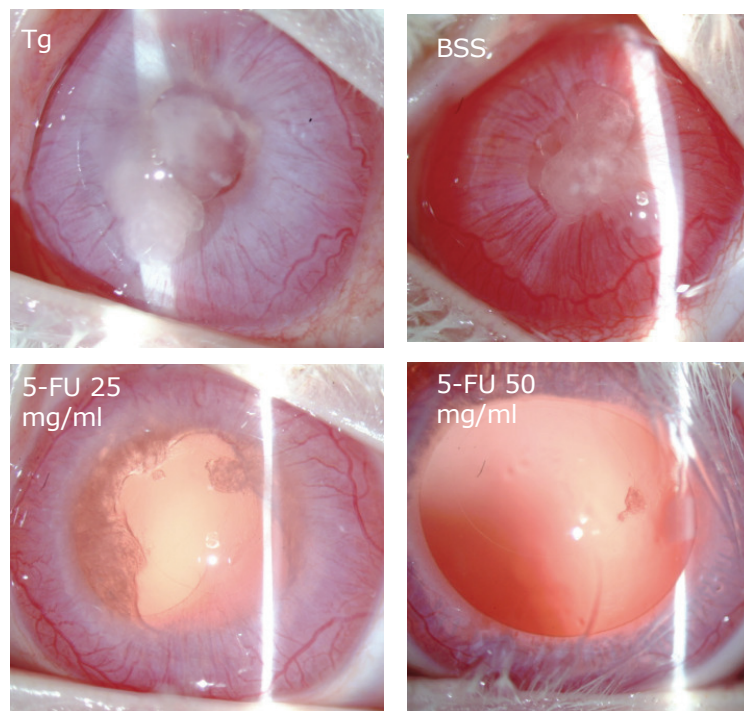


Figure 1. Rabbit eyes that have undergone clear lens extraction and irrigation with different substances in the sealed-capsule irrigation device Perfect Capsule. Conditions six weeks postoperatively. (Top left) Eye irrigated with thapsigargin. The eye shows synechia and visual axis opacification. (Top right) Eye irrigated with BSS; synechia and visual axis opacification is seen. (Bottom left) Eye irrigated with 5-fluorouracil 25 mg/ml. Some synechia is seen. (Bottom right) Eye irrigated with 5-fluorouracil 50 mg/ml. A clear visual axis and no synechia (Abdelwahab et al. 2008).

The safety of 5-FU 50 mg/ml as an irrigation substance with the Perfect Capsule device was investigated, no damage was seen in the corneal endothelium, trabecular meshwork, and central and peripheral retina. We did not detect any damage when looking at the posterior capsule with transmission electron microscopy.

Conclusions

In conclusion, pediatric cataract is still much more problematic than cataract in adults. However, nowadays, IOLs are implanted at a younger age, less secondary glaucoma is then seen, and methods are invented to diminish the visual axis opacification. Hopefully, the research can continue and provide even more information to aid the children in the future.

References

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