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ECTOPIC LENS EXTRACTION IN CHILDREN

KIRURŠKO ZDRAVLJENJE EKTOPIČNE LEČE PRI OTROCIH

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Key words: *ectopia lentis; surgical technique; Cionni endocapsular tension ring; dry irrigation aspiration of lens material; IOL implantation*

Abstract – Background. *Ectopia lentis continues to be a therapeutic challenge for ophthalmologists. It can occur as an isolated condition, after ocular trauma, in association with other ocular disorders, as part of a systemic mesodermal disease or a complication of general metabolic disorders. Minimal subluxation of the lens may cause no visual symptoms, but in more advanced cases serious optical disturbances arise. The most important is amblyopia. Surgical treatment options include iris manipulation, lens discission, aspiration, intracapsular or extracapsular extraction, and pars plana lensectomy. The choice of surgical technique remains controversial, in part because of the historically poor visual results and high rate of perioperative complications, including vitreous loss and retinal detachment.*

Methods. *We describe a surgical technique based on the use of the Cionni endocapsular tension ring, dry irrigation aspiration of lens material, centration of the capsular bag and foldable intraocular lens implantation into the bag. With mentioned surgical technique 8 patients were operated; 4 boys and 4 girls, together 11 eyes.*

Results. *The final BCVA after follow up period improved in 9 eyes and it remained the same as before operation in one eye. Statistical comparison of preoperative and postoperative visual acuities showed significant improvement. On the other hand there was no correlation between preoperative and postoperative visual acuity.*

Conclusions. *This surgical procedure is an alternative approach in solving this challenging cases of ectopia lentis with good postoperative visual rehabilitation.*

Ključne besede: *ektopična leča; kirurška tehnika; Cionni endokapsularni tenzijski obroč; suha irigacija in aspiracija lečnega materiala*

Izvleček – Izhodišča. *Zdravljenje ektopične ali nepravilno lokalizirane leče danes še vedno predstavlja velik problem. Ektopična leča se lahko pojavlja kot izolirana nepravilnost, po očesni poškodbi; pridružena je lahko drugim očesnim boleznim. Pojavlja se lahko kot sistemska bolezen mezodermalnega izvora ali kot komplikacija splošne metabolne bolezni. Minimalen premik leče je lahko brez težav z vidom, pri napredovali obliki pa nastopijo nepravilnosti v optični osi. Najpogostejša posledica je slabovidnost ali ambliopija. Kirurške možnosti zdravljenja so naslednje: premik šarenice ali leče, aspiracija leče bodisi intrakapsularno ali ekstrakapsularno ter pars plana odstranitev leče. Odločitev o vrsti kirurškega posega je še zmerom vprašljiva predvsem zaradi slabih pooperativnih rezultatov v preteklosti in visokega odstotka kirurških zapletov. Najpogostejša sta uhajanje ali prolaps steklovine in odstop mrežnice.*

Metode. *V članku smo opisali kirurško tehniko operacije: odstranitev ektopične leče z uporabo Cionni endokapsularnega tenzijskega obročka, suha irigacija in aspiracija lečnega materiala, centracija kapsularne vrečke in vstavitev upogljive intraokularne leče v kapsularno vrečko. Z omenjeno kirurško tehniko je bilo zdravljenih osem bolnikov; štirje dečki in štiri deklice, skupaj 11 oči.*

Rezultati. *Končna vidna ostrina s korekcijo se je po sledenju bolnikov izboljšala pri devetih primerih, v enem primeru je ostala enaka kot pred operacijo. Statistična primerjava vidne ostrine pred in po operaciji je pokazala statistično značilno izboljšanje. Nismo pa zasledili povezave med vidno ostrino pred in po operaciji.*

Zaključki. *Opisana kirurška tehnika predstavlja možen način zdravljenja ektopične leče z uspešno rehabilitacijo vidne ostrine po operaciji.*

Aim of the study

Ectopia lentis, or displaced lens, is perhaps the most common congenital lenticular anomaly other than cataract. This usually bilateral condition may be caused by an extensive malformation of zonular fibers, resulting in the displacement of the lens in a direction opposite the area of the affected zonules. The displacement, commonly superior or superomedial, is generally the same in both eyes. Late spontaneous dislocation of the lens into the anterior chamber or into the vitreous may be an added sequela of ectopia lentis. Following spontaneous dislocation, a lens generally becomes cataractous, and further complications may arise with the development of elevated intraocular tension (1).

Ectopia lentis can occur as an isolated condition (2); after ocular trauma (3); in association with other ocular disorders, such as ectopia pupillae (4, 5), aniridia (6), congenital glaucoma (7), and megalocornea (8); or as part of a systemic mesoder-

mal disease, as in Marfan (9) or Weill-Marchesani syndrome (10); a complication of general metabolic disorders, such as homocystinuria (11), hyperlysinemia, and combined xanthine and sulfite oxidase deficiency (12). It also has been reported with Ehler-Danlos syndrome (13), Sturge-Weber syndrome (14), Stickler (Marshall) syndrome (15), and many others (16). Both simple ectopia and complicated ectopia lentis have strong hereditary tendencies. The former is, for the most part, transmitted as a dominant trait, whereas the latter is usually recessive. Of these systemic disorders associated with ectopia lentis, Marfan syndrome, Weill-Marchesani syndrome and homocystinuria account for over 75% of the observed lens displacement (17).

Ectopia lentis continues to be a therapeutic challenge for ophthalmologists. Minimal subluxation of the lens may cause no visual symptoms, but in more advanced cases serious optical disturbances arise. The most important is amblyopia. Indications for the lens surgery include besides uncorrectable visual acuity, dislocation of the lens to the anterior chamber, lens opacity, lens-induced uveitis or glaucoma, and imminent complete luxation of the lens (18, 19). The most common indication for surgery to correct vision is bisection of the pupil by the lens (20, 21).

Methods

Surgical technique

We describe a surgical technique based on the use of the Cionni endocapsular tension ring (22–25), dry irrigation aspiration of lens material, centration of the capsular bag (26) and foldable intraocular lens implantation into the bag. Temporal anterior limbal approach is used. The 2.5 mm slit knife is used to create the incision, later two side port incisions are performed. Depending on the extent of mydriasis intracameral adrenalin can be used. Viscoelastic is injected into the anterior chamber. Overpressure should be avoided. With a band needle anterior capsule is punctured and Utrata forceps is used to perform capsulorhexis. It should be rather smaller than bigger. The edge of the capsule is pulled into the centripetal direction to avoid centrifugal tear, same as in normal congenital cataract. In the case of extremely poor zonular support contra action to the pulling Utrata forceps is performed with O'gava lens dialler.

Hidrodissection is performed. The hidrodissection canulla is pointed under the anterior capsule then elevated and BSS (buffered salt solution) is instilled exactly under the capsule to achieve total separation of the cortex. The dry irrigation and aspiration of lens material is done while maintaining the anterior chamber by injecting low viscosity viscoelastic. All the time the capsular bag is maintained with viscoelastic. The iris extension hooks can be used to maintain capsular shape. Double armed 10.0 Vycril suture with a straight needle is guided through the small hole in a Cionni capsular extension ring. The ring is after that implanted into the capsular bag with IOL (intraocular lens) implantation forceps and Sinsky hook, rotated with the ear to the area of the biggest zonular defect and left in place. In this area conjunctiva is cut and sclera exposed. One and half millimetre behind the limbus 27 gauge needle is introduced through sclera into the anterior chamber. From opposite side the straight Vycril needle is guided through paracentesis opening into the anterior chamber and further into the lumen of 27 gauge needle. Both needles are pulled out. The procedure is repeated with second Vycril needle. The sutures are tied and Cionni ring and the capsule are centred.

The foldable acrylic IOL (intraocular lens) is implanted into the bag. The lens is folded with haptics downwards and the haptics are captured between the optics. As the IOL unfolds the haptics are leaded directly into the capsular bag. The IOL canters perfectly in the bag. Careful irrigation and aspiration of the viscoelastic is performed. The incision is sutured if needed. At the end the Vancomycin is injected.

Patients

With mentioned surgical technique 8 patients were operated; 4 boys and 4 girls, together 11 eyes. Five patients have Marfan syndrome, 2 patients have idiopathic ectopia lentis with laxity of the connective tissue and one patient have homocystinuria. Mean age at surgery was 7.1 years (Tab. 1 and 2).

Tab. 1. *Preoperative direction of subluxation.*

Tab. 1. *Lokalizacija premaknjene leče pred operacijo.*

Temporal Temporalno	N = 4
Superotemporal Zgoraj – temporalno	N = 2
Superior Zgoraj	N = 3
Superonasal Zgoraj – nazalno	N = 1
Inferonasal Spodaj – nazalno	N = 1

Tab. 2. *Patient data: preoperative and postoperative findings (BCVA – best corrected visual acuity, CF – counting fingers).*

Tab. 2. *Podatki o bolnikih: pred in po operaciji (BCVA – najboljša vidna ostrina s korekcijo, CF – štetje prstov).*

Patient No. Št. bolnika	Age at surgery (years) Starost ob operaciji (leta)	BCVA before surgery BCVA pred operacijo	Post-operative BCVA BCVA po operaciji	Final BCVA Končna BCVA	Follow up (months) Sledenje bolnika (meseci)
1	6	CF	0.2	0.7	21
	6	CF	CF	CF	19
2	6	0.2	0.3	0.8	20
	8	CF	0.7	0.63	2
3	5	0.1	0.1	0.3	2
	5	0.05	0.3	0.5	7
4	4	< 0.1	0.3	0.4	2
5	11	0.2	0.5	0.63	2
6	13	0.3	0.4	0.5	14
7	7	0.4	0.4	0.4	17
8	6	0.05	0.2	0.6	9

Results

The final BCVA (best corrected visual acuity) after follow up period improved in 9 eyes and it remained the same as before operation in one eye. In one eye amblyopia was detected before surgery. There were no serious complications intraoperatively or postoperatively. Most of the implanted IOLs are well centered, in only one case we noticed mild, asymptomatic decentration. Follow up times ranged between 2 and 21 months (mean 10.5). Statistical comparison of preoperative and postoperative visual acuities showed significant improvement (Fig. 1). On the other hand there was no correlation between preoperative and postoperative visual acuity. However, the results of this study should be viewed in respect to small number of cases.

Discussion

Surgical treatment of ectopia lentis has traditionally been associated with a poor visual outcome and a high complication rate in the past. Numerous techniques were used in the past, but most of the presented results were not encouraging. Intra-

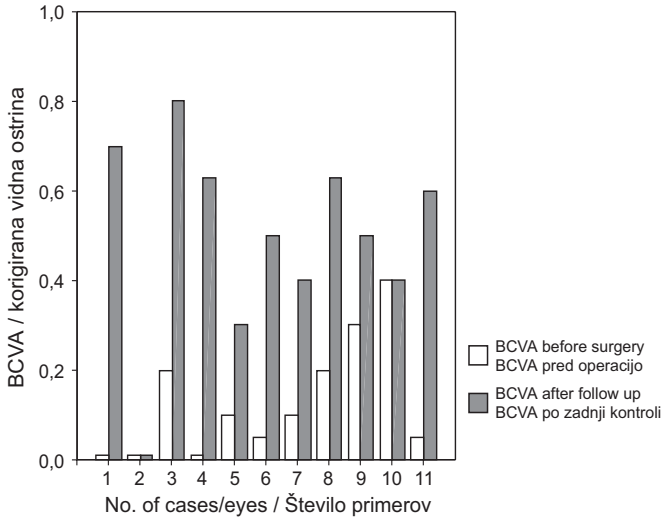

Fig. 1. *Statistical comparison of preoperative and final postoperative visual acuities after follow up period (BCVA – best corrected visual acuity).*

Sl. 1. *Statistična primerjava vidne ostrine pred operacijo in končne vidne ostrine po določenem časovnem obdobju (BCVA – najboljša korigirana vidna ostrina).*

capsular or extracapsular extraction, used in the times when automated vitrectomy was unavailable, often caused uncontrolled vitreous loss and retinal detachment (26–28, 30).
Varga (29) in 1971 reported on the long therm folow-up of 27 patients (46 eyes) undergoing surgery for ectopic lenses. In 31 eyes intracapsular lens extraction was performed, the rest being done by a variety of manual extracapsular techniques. Twenty-three eyes were in patients with Marfan's syndrome. An improvement in visual acuity was documented in 38,6% of patients with Marfan's syndrome at 5 years postoperatively. Visual results in patients with simple ectopia lentis were slightly better, with 8 out of 16 eyes having improved postoperative vision. Complications, including retinal detachments and glaucoma, occurred in 9 (40%) of 23 eyes of patients with Marfan's syndrome who underwent surgery.
In 1972, Jensen (27) and Cross described a surgical complication rate of 51% in treating dislocated lenses in patients with Marfan's syndrome using a variety of surgical techniques (ie, discision, aspiration, intracapsular removal, and inadvertent extracapsular removal). Retinal detachemnt occurred in 19% of aphakic eyes in which it was positively correlated to vitreous loss. Phakic retinal detachment occurred in 9% of patients, which was not significantly different ($p < 0.05$). Despite complication, including vitreous loss, iris prolaps, and corneal edema, the final visual acuity was improved in 27 of 43 eyes. Discission and aspiration is associated with lower incidence of complications (29, 31, 32), but these techniques can be used only for soft lenses.
The surgical techniques for subluxated lens removal have improved with the availability of automated vitrectomy. Peyman et al. (33) in 1979, and Seetner and Crawford (34) in 1981 reported on the first series of patients with ectopia lentis managed with pars plana lensectomy demonstrating good results and a few complications. Similar surgical results with a low complication rate and improved visual acuity were reported also by Syrdalen (35), Reese (36), Seetner (34), Behki (37), Plager (38), Hakin (39) and their coauthors. Visual acuity was in some cases limited only by amblyopia.
Girard (40) et al. described pars plana lensectomy by ultrasonic fragmentation for hard cataractous ectopic lenses in adults with good visual acuity outcome. The technique can be combined with the use of a phacoprosthesis (intraocular lens implant) in adults (41).
Recent developments in surgical techniques have meant that children with dislocated lenses can, more readily, be considered for lens removal. Many eyes with dislocated lenses have normal or near normal acuities after surgery (42). Hing et al. (43) in their study concluded that a child should have the lens removed and wear aphakic correction as soon as the level of vision is inadequate for normal life or the situation of the lens is such as to make amblyopia likely.
Problems related to subluxated lenses may not always be related to surgery. Optical and medical complications also occur. Complications related to optical problems can cause amblyopia, anisometropia, aniseikonia, prismatic effects, and loss of binocular function. These defects may occur with or without surgery. In patients with ectopia lentis is also important that this patients may have serious medical problems. Patients with Marfan's syndrome should be evaluated for mitral valve prolaps and other cardiac and large vessel anomalies. Patients with homocystinuria have a propensity for coagulation problems during and after general anesthesia.

Conclusions

This surgical procedure is an alternative approach in solving this challenging cases of ectopia lentis with good postoperative visual rehabilitation.
Because of the risk of amblyopia in young children, ectopia lentis should be regularly reassessed and surgical procedure considered as soon as uncorrectable optical errors become significant.

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