ON CRYPTOHTHALMOS (2ND CZECH STUDY)

Krásný J.[1], Čakrtová M.[2], Kletenský J.[2], Novák V. (+)[3], Šach J.[4]

[1]Department of Ophthalmology, Královské Vinohrady University Hospital, Prague
Chief: prof. MUDr. P. Kuchynka, CSc.
[2]Department of Plastic Surgery, Královské Vinohrady University Hospital, Prague
Chief: doc. MUDr. M. Tvrdek, CSc.
[3]Department of Polymers, University of Chemistry and Technology, Prague
Head: doc. Ing. V. Schätz, Csc. (+) and prof. V. Dudáček, DrSc.
[4]Institute of Pathological Anatomy, Královské Vinohrady University Hospital and 3rd Faculty of Medicine, Charles University in Prague
Head: prof. MUDr. V. Mandys, DrSc.

Presented at the 9th and 11th Paediatric Ophthalmology Symposiums, Litomyšl 2009 and Olomouc 2013

Devoted to the memory of Dr. J. Otradovec, the founder of modern Czech orbitopathy

MUDr. Jan Krásný
Department of Ophthalmology, Královské Vinohrady University Hospital
Šrobárova 50
100 34 Praha 10,
jan.krasny@fnkv.cz

SUMMARY
ABOUT CRYPTOPHTHALMOS (2ND CZECH STUDY)

Aim: To get acquainted with the 2nd Czech study about cryptophthamos and with self-surgical methods.
Material: The boy with unilateral complete cryptophthalmos of left eye was treated from 2 to 20 years. The girls was treated from 4 month to 5 year yet for right abortive cryptophthalmos with microblepharon and left complete type still waiting for solutions.
Surgical methods and results: Authors present a surgical procedures for correction of the upper and lower eyelids and ocular anomalies both patients studied. Successful reconstruction of palpebral fissure took place in several stages at the boy. The surgical procedure gradually contained: the insertion of gradually increased convex concave circular-shaped implant (silicone ruber) due a modeling of palpebral fissure, an enucleation of rudimentary eye, a reconstruction of bottom palpebral fissure by retro-auricular skin graf and a releasing of the lower transitory fold by the cul-de-sac method. An adequate depth of palpebral fissure to allow perfect position of an aesthetic protesis. Enucleated eye was atypically shaped, reminding partly sand-glass clock. The cornea was replaced by thick fibrous membrane, the iris and the lens were not revealed. Gliomatic retina was detached nearly totaly and the optic nerv was rudimental. The repairing the upper lid coloboma of girl by a lid rotation flap reconstruction using the inferior eyelid was performed at the age 17 month. Corneal dermoid simultaneously removed (histologically verified). Upper conjunctival fornix was formed using the spherical covering foil (silicone rubber) before and after the reconstruction of the lid.
Conclusions: Plastic reconstructions required the need for patient access without trying immediate effect. An important role played silicone rubber implants (elastomer medical grade) which used temporarily.

Key words: cryptophthalmos, microblepharon, relief surgery, silicon ruber implants

Čes. a slov. Oftal., 71, 2015, No. 6, p. 278–286

INTRODUCTION

Developmental anomalies of eyelids have several different clinical pictures. These include above all ptosis of the upper eyelid, and there are also less frequent colobomas, mostly relating also to the upper eyelids. Both conditions can be treated surgically with a good result. Epiblepharon and euryblepharon, which are not treated surgically, also have a congenital basis. In the case of blaphophimosis and epicanthus, the surgical procedure depends on individual consideration. Ectropia and entropia on a congenital basis are rare, and the operation within the given time frame is determined by their scope and if applicable by present irritation of the eyeball. Reconstruction surgery is possible for these diagnoses, and the degree and extent of the defect is of decisive significance concerning the possibility not only of a cosmetically positive result, but also of the functional effect. From the perspective of the option of surgical cosmetic treatment without the possibility of restoring the functionality of the visual organ, serious conditions are represented by the clinical units outlined below. Microblepharon is an exceptional deformity of the eyelid, characterised by its truncation in the vertical axis. The clinical manifestation is determined by the width of the truncation, which is variable, and may lead to a colobomatous manifestation of such a size that only a rudimentary margin of the eyelid may remain. This therefore concerns an abortive form of cryptophthalmos in the sense of truncation mainly of the upper eyelid, with a deficit of closure of the palpebral fissure. Cryptophthlmos proper is a rare congenital disorder of the separation of both eyelids, which is characterised by defective development of the eyelids and palpebral fissure to a varying extent. Although the ectodermal layer is differentiated into the cornea and the conjunctiva, there are pronounced changes of its structure. The conjunctival sac is either not formed whatsoever or only partially formed. Another alternative is ancyloblepharon, in which the eyelids are originally formed, but the conjunctival sac has been obliterated in development. The eyeball itself is mostly markedly afflicted. A separate unit is represented by microphthalmos, which is accompanied by a reduction in the size of the palpebral fissure, though the function of the eyelids is retained. In extreme cases it may be so pronounced that it is sometimes difficult to differentiate from anophthalmia. In most cases it concerns a rare unilateral finding of sporadic occurrence. A bilateral finding is rare, and is generally linked with systemic abnormalities, primarily with Fraser's syndrome (see below). Also rare are findings of microblepharon and ablepharon, which are generally linked with deformities of the ocular structures located beneath them. Ablepharon is a manifestation of complete failure in the development of the eyelid. Its incidence is even rarer, and is accompanied by severe ocular and also general abnormalities. All disorders in the development of the eyelids with simultaneous severe alterations of the ocular structures located beneath them are frequently referred to in summary as crypotophthalmos, with diverse variants. Anophthal-

mia is another rare developmental anomaly of the eyeball, which has an incidence of 5/100 000. Congenital anophthalmia is divided into primary, secondary and degenerative, its etiology is multifactorial. Primary anophthalmia is always bilateral, and originates on a basis of differentiation of the optic disc. The orbits are generally afflicted, and the eyelids markedly truncated. The secondary form is the result of complete suppression or severe anomalous development of the entire anterior part of the neural canal. It is accompanied by an absence of the orbits. The third form is degenerative anophthalmia, which is mostly unilateral, originates on a basis of atrophy of an already established optic vesicle, and rudimentary extreme microphthalmos is generally present. Severe brain abnormalities are not present. The orbit and adnexa are usually only slightly afflicted.

Over the entire sixty one years of the history of this journal, only a single report from the 1960s has been devoted to the issue of cryptophthalmos. We decided to return to this theme on the basis of two observations, primarily from the perspective of potential reconstruction.

Own observations and methods of plastic treatment

One boy and one girl were included in the study cohort. We treated the patients' conditions progressively within the framework of long-term observations, with an interval of five years.

Patient no. 1. The boy was born in February 1986, from the 3rd physiological pregnancy (both siblings, 17 and 9 years older, are healthy). The birth had a physiological course, birth weight 3150 g. The family anamnesis was without encumbrance, without serious hereditary disorders and consanguinity. At birth a closed palpebral fissure was diagnosed in the left eye, with a partially formed eyebrow. The upper eyelid was partially differentiated, the lower eyelid was only a fold of skin, in the outer half covered by the conjunctiva, but the inner part of the eyelid was lacking (fig. no. 1A). A palpable moving spherical formation was evident beneath the skin accretion of the thus altered eyelids. A CT examination demonstrated a bordered and reduced bone casing of the orbit with a slightly oval cystic formation in the place of the eyeball, with a size of 10 x 20 mm. In the right eye the finding of the orbital structures was within the norm, the same as CNS. The surrounding area of the right eye and the eyeball itself did not manifest any deviation from a physiological finding. No other developmental anomalies were found in the boy.

We commenced the 1st phase of reconstruction at the Motol University Hospital when the boy was aged 2 years (1988) by modelling of the palpebral fissure and insertion of a special conjunctival implant. The surgical method was

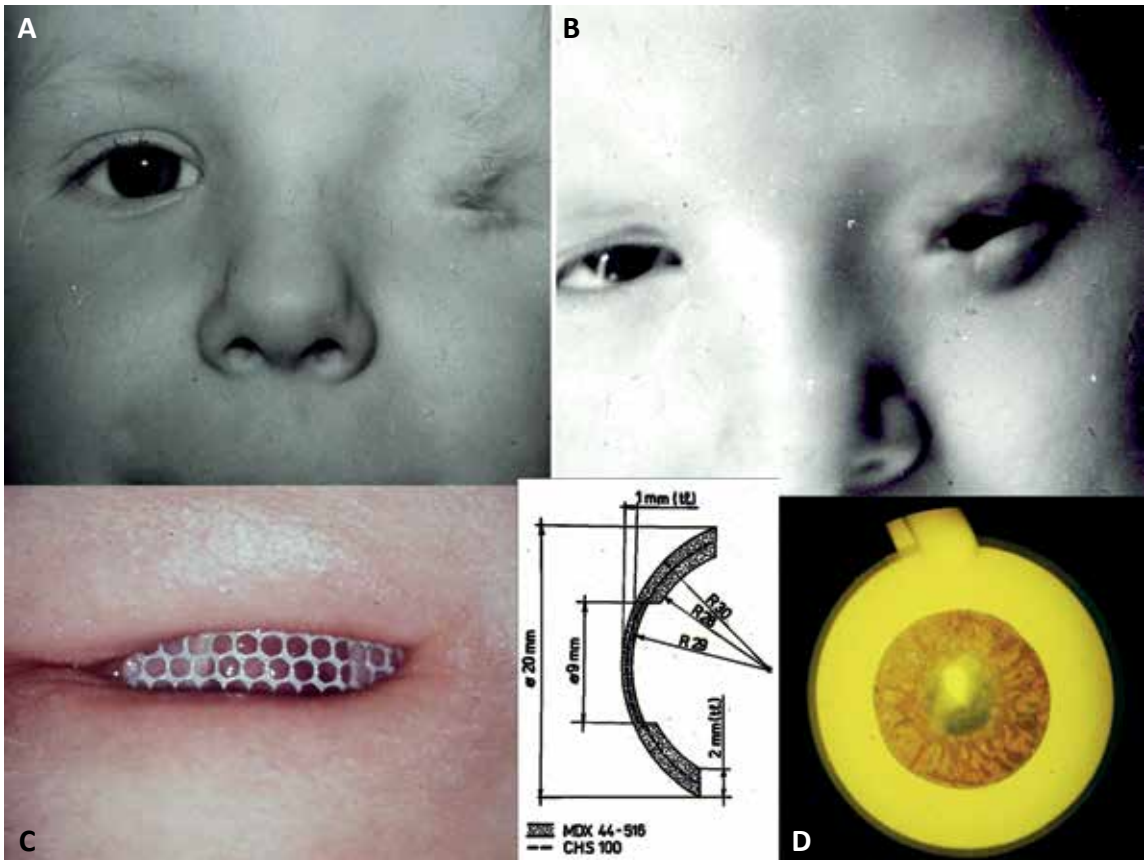

Fig. no. **1A. Two year old boy with cryptophthalmos in left eye**
1B. Formation of palpebral fissure by anophthalmic implant at age of four years
1C. Anophthalmic silicon dacron implant in palpebral fissure and diagram thereof (MDX 44-516: silicone rubber, CHS 100: dacron netting)
1D. Silicone implant with modelled anterior segment replacing prosthesis

similar to that which we used for plastic surgery of the conjunctival sac in the case of anopthalmos. During the course of the following six years we inserted a progressively enlarging implant a total of nine times for modelling of the area of the palpebral fissure (fig. no. 1B) in order to enable the insertion of the definitive prosthesis, furthermore the pressure of the implant supported the development of the orbital region. The implant was a circular shaped convex-concave disc (fig. no. 1C) produced from biomedical silicone rubber, which was originally reinforced with dacron netting.

We commenced the **2nd phase** of reconstruction with the patient aged ten years (1996). First of all plastic surgery of the inner corner was performed by a skin graft, and in the following years the base of the palpebral fissure was modelled twice with further skin grafts from both retro-auricular areas with regard to the similar character of the skin on the eyelids (fig. 2A). In order to deepen the palpebral fissure covered by the skin and to deepen the lower fornix, the rudimentary eyeball was enucleated when the patient was aged fifteen years (May 2001). The enucleated eyeball manifested pronounced atypical features. It had a shape reminiscent of an hour glass, with a front-to-back length of 22 mm, a mediolateral diameter of 10 mm and a craniocaudal diameter of 8 mm (fig. 3A). A detailed histological-pathological examination demonstrated that the cornea had been replaced by a fibrous membrane, the lens and iris were lacking (fig. no. 3B). The gliomatous retina was almost totally detached and the optic nerve was rudimentary (fig. no. 3C). Within the framework of the surgical procedure, we extended the lower fornix covered by the skin graft centrally up to the periosteum with the help of a traction fixation suture by the cul-de-sac method. Into the newly created palpebral fissure, replacing the classic conjunctival sac, we then inserted an individually produced conjunctival implant, which replaced the classic prosthesis (fig. no. 2B). This was again produced from silicone rubber: the iris-pupil area was modelled on a white coloured silicone background using acrylate paints, and covered with a transparent silicone foil (fig. no. 1D).

We supplemented the **3rd phase** of the reconstruction after an interval of five years (2006), when we completed the plastic surgery of the area of the newly created pseudoconjunctival sac (fig. no. 2C) by electroepilation of the hairs on the perfectly healed skin grafts, which were a source of irritation when attempting to insert the ocular prosthesis. For better fixation in the newly created base of the palpebral fissure, the previous plastic surgery of the fornix in the inner corner was supplemented by a further two traction sutures (cul-de-sac method) up to the periosteum. This condition

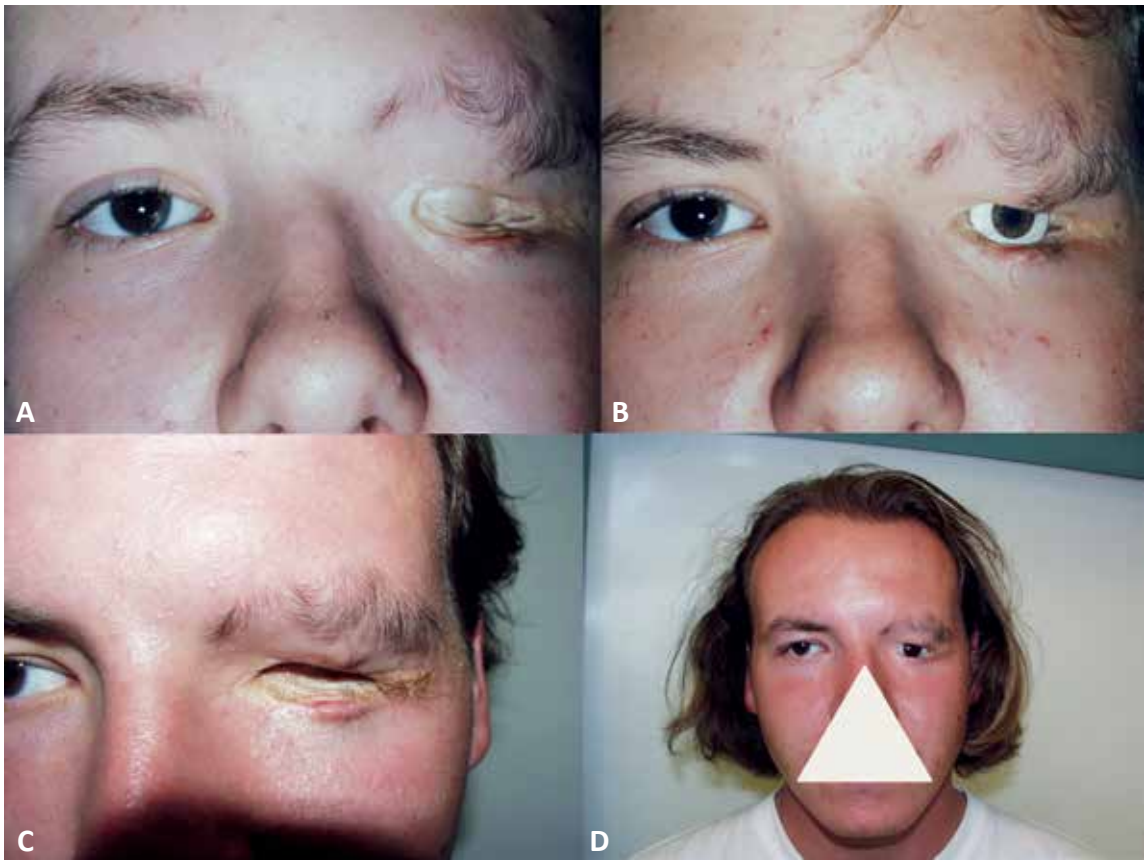

Fig no. 2A. Palpebral fissure in left eye after skin plastic surgery procedures at age of thirteen years
2B. Palpebral fissure in left eye with silicone implant (prosthesis) at age of thirteen years
2C. Definitive shape of palpebral fissure in left eye at age of twenty years
2D. Palpebral fissure in left eye with acrylate prosthesis at age of twenty years

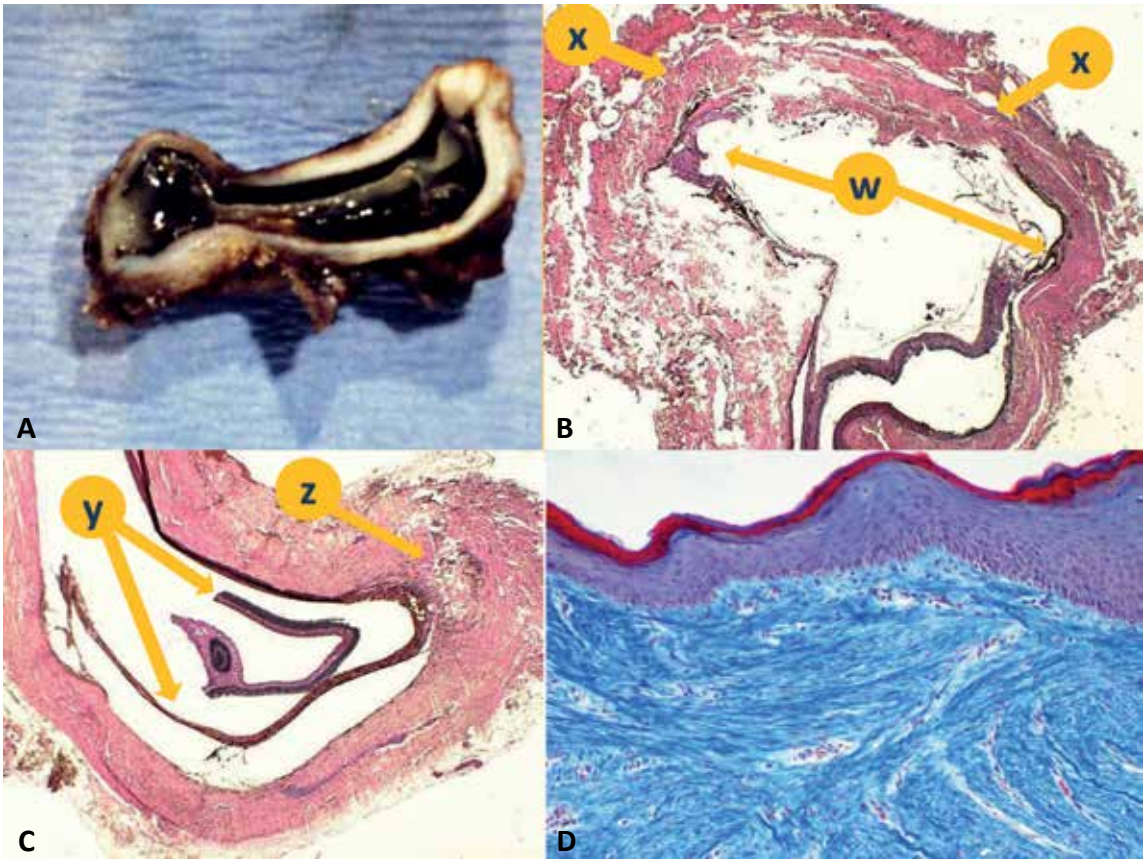

Fig. no. **3A. Cryptophthalmos – macro-image: in shape of hour glass**
3B. Cryptophthalmos – anterior segment: rudiment of plicated body "w" and between arrows "x" fibrous membrane of cornea. Colouring H.E., enlarged 20x.
3C. Cryptophthalmos – posterior segment: detached gliomatous retina "y" and rudimentary optic nerve "z". Colouring H.E., enlarged 20x.
3D. Epibulbar dermoid of cornea: keratinising (red) corneal tile-like epithelium (purple) and beneath den se, vascularised collagen ligament tissue (blue). Colouring blue trichome, enlarged 200x.

enabled free insertion of the specially produced acrylate prosthesis (fig. no. 2D).

Patient no. 2. The girl was born in May 2010 as the first child of young, unrelated healthy parents, and has no siblings. The course of the first pregnancy was physiological, the mother GBS negative. The birth took place in the 40th week spontaneously head first, birth weight 3500 g and length 50 cm, clear amniotic fluid, the baby was not resuscitated (Apgar 10 – 10 –10). After birth a bilateral developmental anomaly of the face was determined in the area of the palpebral fissures: microblepharon of the upper eyelid in the right eye connected with symblepharon of the conjunctiva toward the upper section turbid and thickened cornea, and cryptophthalmos in the left eye. The further overall development was commensurate, the baby gained weight. Although psychomotor development was influenced by the visual handicap, the child nevertheless walked unaided from the age of 15 months. A detailed screening examination for further congenital developmental anomalies demonstrated only a bone defect on the skull parietally on the left side. MR CNS and orbit: In the right the image of the eyeball, lateral eye muscles, optic nerve and orbit was physiological. In the left there was a deformity of the shallowed orbit without dissociation of the eyelids, which merged into a single skin structure. Present microphthalmos was accompanied by congenital aphakia. In the orbital cavity there was a present lachrymal gland, muscle structures and optic nerve. With the exception of the above-described bone defect, the intracranial finding was within the norm and both optical tracts were of a physiological course. The child was first treated in an ophthalmological clinical centre outside of Prague at the age of one month. Only tarsorrhaphy was performed using mattress sutures in the right eye, but without effect.

The patient was sent to the Department of Ophthalmology at the Královské Vinohrady University Hospital in Prague for assessment and solution of the ocular finding in September 2010 at the age of 4 months (fig. no. 4A). In the right eye the lower eyelid was normally developed, attached to the eyeball with a loose fornix, the lachrymal punctum was present. Towards the cornea with a diameter of 9 mm from above, between no. X up to no. I there was a an adherent colobomatously truncated conjunctival skin structure, passing through the symblepharon into the centre of the cornea and connecting to a whitish-grey undulating tumorous structure, which was above the niveau. The cornea it-

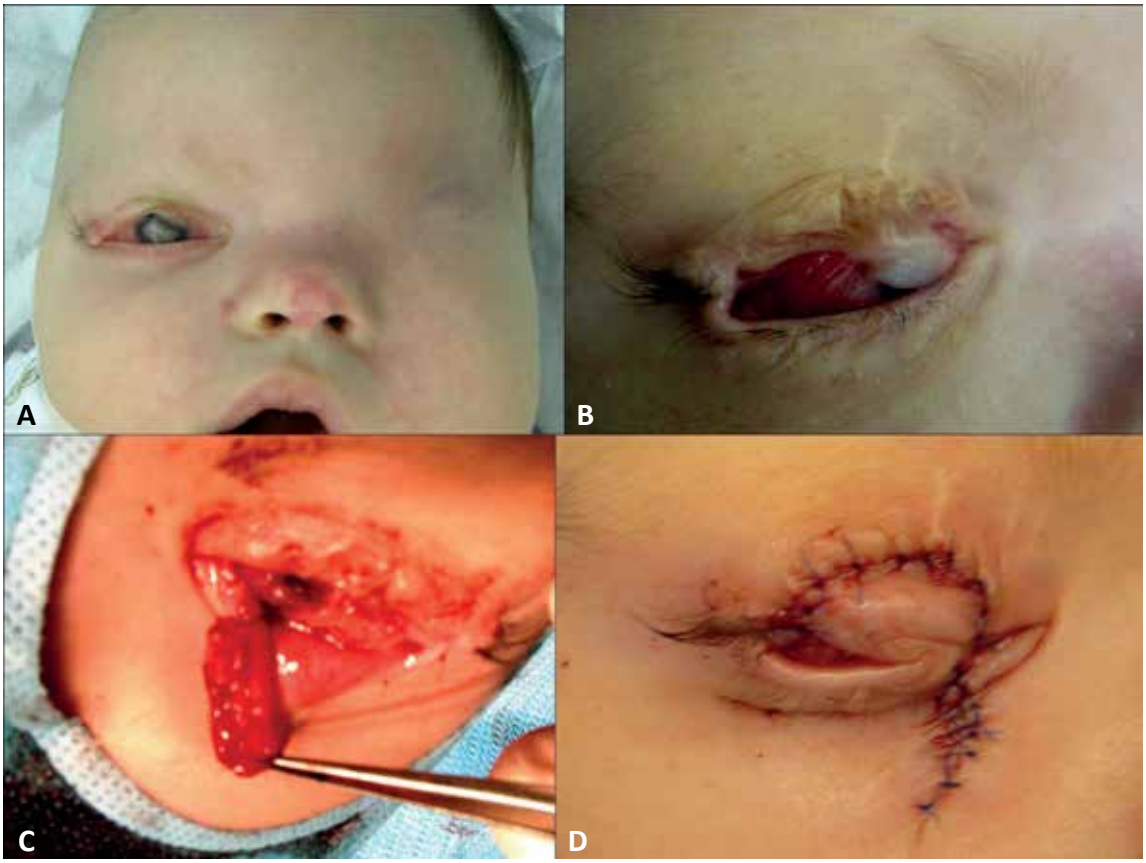

Fig. **4A. Four month old patient with abortive cryptophthalmos with microblepharon in right eye and classic cryptophthalmos in left eye.**
4B. Coloboma of upper eyelid with dermatoconjunctival symblepharon directed towards epibulbar der moid of cornea in left eye
4C. Preparation of rotation flap from lower eyelid in left eye
4D. Fixation of rotation flap in coloboma of upper eyelid and suture of lower eyelid

self was visible only in the lower half, and was leukomatously turbid, preventing differentiation of the other structures. The symblepharon ensued from the pronouncedly vertically truncated upper eyelid throughout the entire width of the palpebral fissure. The upper eyelid was in the form of a banded skin structure with a width of 2 mm, which was further truncated in wedge shape by the apex towards no. XII. In the left eye the area of the palpebral fissure without eyebrow was covered by skin tissue, without any sign of structural differentiation, beneath which there was an arched spherical formation.

Phase 1. In September 2010 we separated the truncated upper eyelid from the cornea and released the indicated upper fornix without conjunctival coverage, inserted spherical covering foil and closed the palpebral fissure by tarsorrhaphy using two mattress sutures underlain with foam amotio fillings. The spherical covering foil was further replaced four times under general anaesthesia, always with renewal of tarsorrhaphy, and left until the beginning of May 2011. After this time we only checked on the patient in outpatient care until the next surgical procedure.

Phase 2. In October 2011 we removed the tumorous undulating tissue (fig. no. 4B) from the centre of the cornea. A solid epibulbar dermoid of the cornea was verified histologically, characterised by abundant cellular and dense collagen ligament tissue with vascularisation, on the surface with coverage of a well matured, in places keratinising tile-like epithelium of epidermal type (fig. 3D). At the same time, following dislocation of the symblepharons and release of the upper fornix, we created a rotation flap from the lower eyelid throughout the entire thickness, which we fixed to the colobomatous defect of the upper eyelid (fig. 4C, 4D). Subsequently, in January 2012 we detached the rotation flap in the place of the stem and modelled a reconstructed or newly created upper eyelid (fig. no. 5A). The lower, original conjunctival fornix remained loose, and we again released the upper fissured pseudofornix (inserted spherical covering foil). It was only in October 2012 that we rotated the excess bulbar conjunctiva in the upper quadrants, which was formed by the pressure and tensile force of the spherical covering foil, in the direction beneath the original skin of the newly created upper eyelid, and fixed it to depth with transcutaneous sutures made of absorbent material (Vicryl 6-0). By this method the modelled actual upper fornix was practically covered from within throughout its entire scope by the conjunctival tissue, and simultaneously jointly fixed

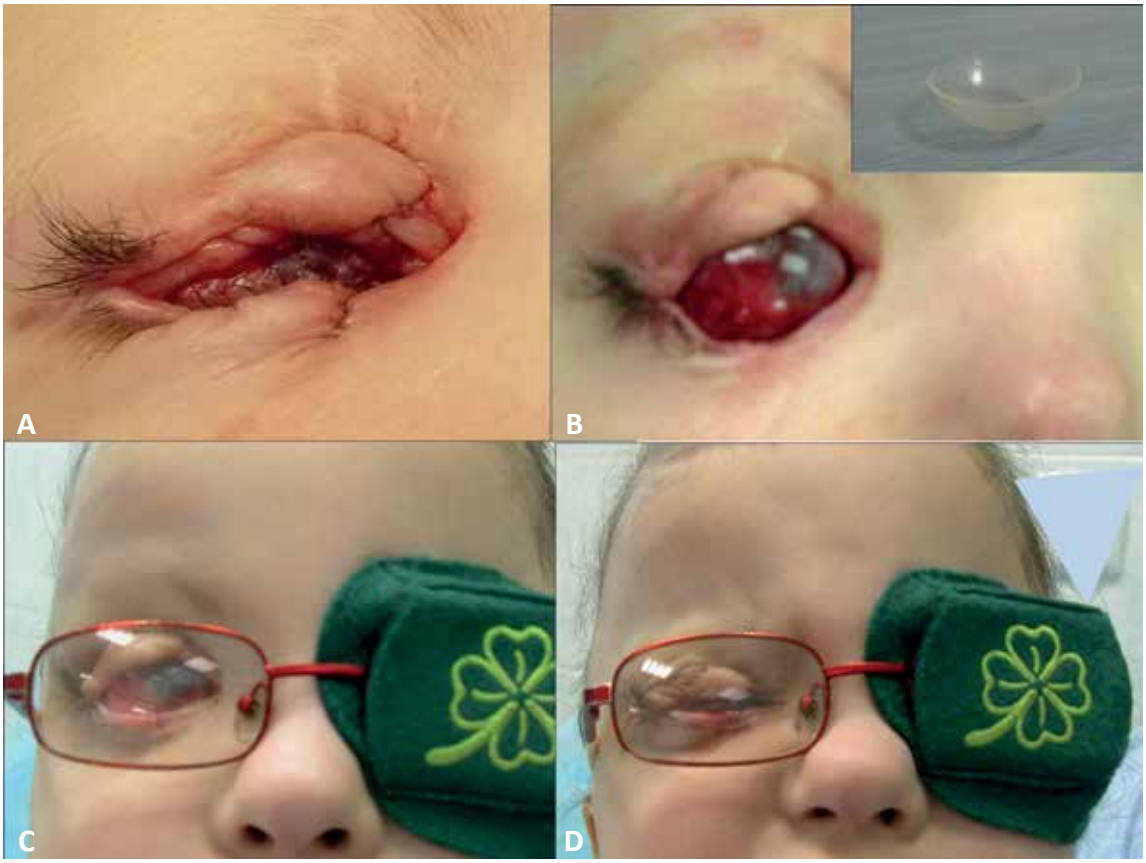

fig. 5A. Release of upper eyelid after transplantation – creation of palpebral fissure
5B. Plastic surgery of fornix using spherical covering foil in 3rd phase of surgical procedure
5C. Shape of right palpebral fissure at age of six years behind glasses with cosmetic occlusor on left eye
5D. Spontaneous closure of right palpebral fissure at age of six years, analogous cosmetic solution

by the inserted spherical covering foil.

Phase 3 In February 2013, in connection with the rotation of the lower eyelid into the area of the defect of the upper eyelid, the edge of the eyelid became loosened, ectropion occurred, and the child was unable to close the palpebral fissure. As a solution we chose suspension of the eyelid using a fascial graft (ZUM 43104: allogen fascial graft frozen from the Tissue Centre at Hradec Králové University Hospital) stretched from the outer corner, where it was fixed to the periosteum, to the inner corner, where it was fixed by a loop to the inner canthus, again using absorbent material (Vicryl 6-0). At the same time we fixed a further part of the bulbar conjunctiva above using transcutaneous sutures for deepening the fornix, which we again and repeatedly supported by pressure of the spherical covering foil modelling the palpebral fissure (fig. 5C). We had to repeat this procedure in the area of the upper fornix 2x in June 2013 and March 2015, since spontaneous loosening of the spherical covering foil after nine months and one year led to repeated symblepharons, as well as recurrence of a dermoid of the cornea. This time we performed fixation using the long-term absorbent material PDS 6-0. We again had to collect the tumour, increased mitotic activity was not histologically demonstrated, but again this concerned a dermoid.

The eyeball remained freely mobile. The patient was able to close the palpebral fissure spontaneous even during sleep (fig. no. 5C), the skin of the eyelids only in creases with a relative excess. Visual acuity in the right eye was so far defined on the level of severe purblindness (movement with light projection, with determination of basic colours from a few centimetres). Spatial orientation was retained, manual contact was required for definition of an object. From a cosmetic perspective a glasses frame with an occlusor was fitted to cover the left orbital area (fig. no. 5D).

CONCLUSION – EPICRISIS

Our own observation of the individual phases of treatment in the case of the male patient, up to the time when it was possible to conclude the condition definitively with a relatively successful cosmetic effect, lasted for a total of eighteen years. A perfect plastic surgical solution of cryptophthalmos is not possible, our endeavour was to improve the physiognomy of the face for adult age. Long-term observation and the individual phases of surgery demonstrate the necessity of a patient approach on the part of the medical team, without attempts to achieve an immediate effect. An essential prerequisite for success is complex co-operation, partially with the plastic surgeon and in our case the possibi-

lity of co-operation with a biotechnologist, who was able to propose and prepare special modelling preparations.

The female patient is and shall continue to be observed in outpatient care. In the right eye we created a relatively regular, slightly narrower palpebral fissure through reconstruction, lined with eyelids within the framework of interdisciplinary co-operation with a plastic surgeon. The perforating keratoplasty under consideration does not have a precisely determined time frame, since first of all there must be a long-term created and functional palpebral fissure with free fornices and bearing of the eyelids on the eyeball. Furthermore, here it shall be essential to ensure immunosupressive safeguarding of the surgical procedure. Plastic surgery is also being considered prospectively in the area of the cryptopthalmos in the left eye, similarly as in the case of the first patient.

DISCUSSION

The term "cryptophthalmos" was proposed by Manz in 1872 (from Greek: cryptos – hidden + ophthalmos – eye), within the framework of a joint observation with Zehender, who recorded the first clinical description [37]. The detailed literary analysis of cryptophthalmos covering the 90 years since Zehender and Manze's initial observation includes a total of 39 observations, including the author's from 1962 [29]. A detailed story of the first observation is also presented here: Healthy parents without cosanguinity brought an infant for examination at the age of six months, who had been born "without eyes or determinable sex". The child was bilaterally without eyebrows, the eyelids and palpebral fissures and the skin passed smoothly from the forehead across the entrance to the orbit onto the skin of the face, beneath the skin there were palpable mobile bulbs. Further malformations included an extensive abdominal rupture, syndactily on all the limbs and malformed genitalia with agnesis of the sphicters of the anus and urethra. After a few months the infant died and a histological examination was performed on one of the eyes [29].

The theory of the origin of cryptophthalmos was controversial. At the turn of the 19th and 20th centuries, the background of origin was considered to be coalescence of the amnion with the anterior surface of the embryo [17]. After World War I, cryptophthalmos was defined as a condition of a rudimentary eye, over which there is unbreached skin without the formation of eyelids and a palpebral fissure – ablepharon [13], and metaplasia of the corneal and conjunctival ectoderm was also considered [25]. According to current knowledge, in the 8th week the conjunctival sac forms a separating space between the cornea and the future eyelids from the semilunar folds of the ectoderm covered by the mesoderm [7]. Differentiation of the eyelids begins to take place on the embryo with a size of 32 to 37 mm, and is definitively concluded in the 6th month of pregnancy [9]. Actual cryptophthalmos with microphthalmos is linked with a developmental disorder of the eyelids in the 26th to 28th week of gestation, in which the eyelids separate from one another [2]. Congenital colobomas of the upper eyelid in connection with potential cryptophthalmos are divided into five degrees. The first three degrees are represented by actual coloboma of the eyelid without cryptophthalmos, or with abortive cryptophthalmos or complete cryptophthalmos. The further two degrees are represented by classic cryptophthalmos (absence of eyelid structures and the eyeball is completely covered by skin) and severe cyptophthalmos (with severe deformities of the nose and ectropia of the upper eyelid) [24]. According to this differentiation, it would be possible to classify our male patient as within the classic form of cryptophthalmos and the female patient as a combination of classic cryptophthalmos on one hand and abortive cryptophthalmos on the other. A morphological examination of the enucleated eyeball demonstrated considerable influence of the development on both the anterior segment of the eye and also the posterior segment. The presence of a lens was not demonstrated, which is in accordance with the previous nine out of ten histological examinations, here also the cornea was atypical, since the skin was a part of the wall and the retina was detached [29]. This reflects the foreseeable matter that a malfunction of contact and development was stated on the interface between the optical sac and the ectodermal surface during the course of embryonic development [1, 36].

Fraser's syndrome (F.s.) is an autosomally recurring pathology, which combines predominantly bilateral cryptophthalmos (93%) with general affliction of a range of organs. The most important in terms of numbers are syndactaly, described in 57% of cases, mental disablement in 50% and genital affliction (cryptorchidism, hypospadias, chordae penis) in 49 %. Life threatening conditions are renal hypoplasia or agenesis in 37% of cases and laryngeal stenosis or atresia in 21%, as a result of which 20% of infants born with this condition die within the first year of life. The data is based on a comparison of more than 100 observations [10]. the condition was first defined by the British geneticist George Fraser in 1962, and is sometimes referred to as Fraser-Francois syndrome [11]. 28 observations of cryptophthalmos in connection with multi-organ affliction have been recorded in the past, including the Czech case report [29]. An epidemiological study processed by 25 geneticists without a Czech contribution between the years of 1990 and 2008 detected F.s. in 26 individuals out of almost 13 million newborn infants within the European population (prevalence of 0.2/100 000 births) [3]. An analysis of 59 cases of F.s. in a total of 40 families demonstrated cosanguinity in 25 cases [32], which represents 42% of its representation. A comparison of individual previous observations states cosanguinity, but only in 4 cases in cryptophthalmos with affliction of organs in 28 of those afflicted, which represents 14% [29]. Prenatal diagnosis enables determination of multi-organ affliction through an analysis of the amniotic fluid [32]. A significant aid is ultrasonic examination of the foetus, which detects above all agenesis of the kidneys or multicystic affliction and stenoses of the respiratory tracts in the 16th or 23rd weeks of gestation [4, 12]. In one of these observations the face and brain structures were still without detectable changes. On the basis of the above examination, the parents decided to terminate the pregnancy and the diagnosis of F.s. without cryptophthalmos was determined by the section [4]. In addition to F.s., cryptophthalmos is described in oculodentodigital syndromes, which are without a ge-

netic burden (Loehmann's or Meyer-Schwickerath syndrome), in which there is associated cleft palate, changes of the dentia and syndactyly [9]. Cryptophthalmos has also been described in connection with atresia of the larynx, malformations of the turbinate bones, umbilical hernias [1, 9], malformations of the large blood vessels [15, 16] or brain: encephalocele [15] and schizencephaly with meningoenphalocele [20]. From our observation of 4 patients with anophthalmia [22] and a further five infants in the following years, of whom two were afflicted bilaterally, it ensues that cryptophthalmos was approximately five times rarer than anophthalmia over a nineteen-year observation period. The primary and secondary form of anophthalmia in differential diagnostics incorporates cryptophthalmos, cyclopia, synophthalmia and congenital cyst of the eyeball [34], in which magnetic resonance makes a significant contribution to diagnosis [6]. Only the third degenerative form of anophthalmia can also be corrected by reconstruction, e.g. with the use of a silicon forming implant [19] or plastic surgical procedure by the insertion of skin grafts [34].

In the surgical solution of the individual phases of reconstructing the upper eyelid and fornix in the observed female patient, commenced in infant age, we started out from our own experiences of plastic surgery treatment for cryptophthalmos on the first patient, which had taken place over a total period of fifteen years [20] and which was completed five years ago. A significant component of reconstruction of the palpebral fissure was partially supplementation of the skin cover with grafts, but also use of the cul-de-sac technique, which helps in deepening the lower transitory fold [28]. This technique was proposed at the beginning of the 20th century by Weeks [13], and we technically adjusted it into its present form [18]. In the early stages of treatment we used spherical covering foil in order to shape the upper fornix and for protection of the cornea [23]. Later in toddler age we supplemented plastic surgery with the used technique of a rotation flap from the lower eyelid [5]. Here it was essential to exercise patience and to be economical with all the naturally formed tissues of the eyelid and conjunctival sac, and upon transplantation an excess of skin was retained for further growth of the eyelids and the possibility of any applicable surgical correction [21]. The procedure was demonstrated to be beneficial, since the flap from the lower eyelid served for reconstruction of the upper eyelid in the case of incomplete cryophthalmos also for future surgeons [8, 26], when the buccal mucosa also was simultaneously used [8, 31]. The surgical solution included removal of an epibulbar dermoid of the cornea, which was described [14]. Other surgical techniques have appeared in the literature, e.g. insertion of conchal cartilage as a layer between the skin of the mucosa [33]. Upon solution of bilateral partial cryptophthalmos, a loose skin graft from the child's retro-auricular region was also used, the tarsal plate was replaced by a scleral graft from the tissue bank, and the buccal mucosa in the place of conjunctiva was implanted from the mother of the child [27]. In our female patient we did not replace the conjunctiva with other tissue. The transplant from the lower eyelid contained the lower tarsal conjunctiva on its inner surface, and we developed a part of the released part of the bulbar conjunctiva and fixed it to the remaining part of the skin of the upper eyelid, and further shaped the fornix with pressure from the inserted spherical covering foil [23]. The originality of our surgical solution lay in the insertion of the spherical covering foil, the development of which was appraised and included in the digital Slides Online Digital Library [35]. The aim of the surgical procedures was to create a functional upper eyelid and thus provide a basis for future keratoplasty, and at the same time to create the opportunity for sensory stimulation of the visual organ as a whole, with a normally functioning upper eyelid.

Poděkování patří třem výše uvedeným inženýrům z Katedry polymerů VŠCHT, kteří se zasloužili o rozvoj oftalmologie vývojem různých implantátů. Řada z nich je stále ve výrobním sortimentu firma ELLA-CS v Hradci Králové.

LITERATURE

1. **Apple, D.J., Rabb, M.F.:** Ocular Pathology, 5th edition., Mosby, 1998 , St. Louis, p.16.
2. **Barishak, Y.R.:** Embryology of the Eye and Its Adnexa. Karger, Basel, 2001, p.132.
3. **Barisic, I., Odek, L., et al.:** Fraser Syndrome: Epidemiologic Study in a European Population. Am J Med Genet A, 30; 2013: 1012–1018.
4. **Berg, C., Geipel, A., et al.:** Prenatal Detection of Fraser Syndrome without Cryptophthalmos. Case Report and Review of Literature. Ultrasound Obstet Gynecol, 18; 2001: 76–80.
5. **Brazier, D.J., Hardman–Lea, S.J., Collins, J.R.:** Cryphtophthalmos: Surgical Treatment of the Congenital Symblepharon Variant Brit J Ophthalmol, 70; 1986: 391–395.
6. **Celebi, A.R., Sasani, H.:** Differentiation of True Anophthalmia from Clinical Anophthalmia Using Neuroradiological Imaging. Word J Radiol, 6; 2014: 515–518.
7. **Conlon, R.M., Sutula, F.C.:** Congenital Eyelid Anomalies. In Albert, D.M. & Jakobiec, F.A.: Principles and Practice of Ophthalmology, Vol. III., W.B. Saunders Co. Philadelphia, 1994, p. 1693–1699.
8. **Dibben, K., Rabinowitz, Y.S. et al.:** Surgical Correction of Incomplete Cryptophthalmos in Fraser Syndrome. Am J Ophthalmol, 124; 1997: 107–109.
9. **Duke-Elder, S.:** Congenital Anomalies of the Ocular Adnexa. In System of Ophthalmology, Vol. III. Congenital Deformities, H. Kimptom, London, 1964, pp. 827–835.
10. **Eibschitz-Tsimhoni, M.:** Ocular Manifestation of Inherited Diseases. In Wright, K.W., Spiegel, P.H.: Pediatric Ophthalmology and Strabismus. Springer, New York, 2003, p.1038 .
11. **Francois, J.:** Syndrome malformatif avec cryptophthalme. Ophthalmologica, Basel, 150, 1965: 215.
12. **Fryns, J.P., van Schoubroeck, D., et al.:** Diagnostic Echographic Findings in Cryptophthalmos Syndrome (Fraser Syndrome). Prenat. Diagn., 17, 1997: 582–584.
13. **Fuchs, H.E., Duan, A.:** Text-book of Ophthalmology. J. B. Lippincott Co., Philadelphia 1923, p. 437.
14. **Gerinec, A.:** Detská oftalmológia, Osveta, Martin, 2005, s. 104.
15. **Goldhammer, Y., Smith, J.L.:** Cryptophthalmos Syndrome with Basal Encephaloceles. Am J Ophthalmol, 80; 1975: 146–149.
16. **Hambire, S.D., Bhavsar, PP., Jayakar, A.V.:** Fraser-Cryptophthalmos Syndrome with Cardiovascular Malformations:

A Rare Association. Indian. Pediatr, 40; 2003: 888–900.
17. **Chalupecký, J.:** Oční lékařství. Hejda a Tucek, Praha, 1902, s. 316
18. **Krásný, J.:** Plastika anoftalmického spojivkového vaku upravenou metodikou cul-de-sac. Čes a Slov Oftal, 71; 2015: 37–43.
19. **Krásny, J. Autrata, R.:** Pedooftalmologie a strabismus. In Kuchynka, P. a kol.: Oční lékařství, Grada, Praha, 2007, s. 647, 653.
20. **Krásný, J., Čakrtová, M., Novák, V.:** Jednostranný kryptoftalmus–plastická úprava. Folia Strabol Neuroophthalmol; 10, 2009, suppl. I.: 86– 88.
21. **Krásný, J., Kletenský, J.:** Kombinace pravostranného horního mikroblefaronu s kongenitálním symblefaronem a levostranného kryptoftalmu. Folia Strabol Neuroophthalmol; 14, 2014, suppl. I.: 39–43.
22. **Krásný, J., Novák, V.:** Anophthalmus–spojivkový implantát. Čes a Slov Oftal, 59; 2003: 33–39.
23. **Krásný, J., Novák, V.:** Kulová krycí folie–silikonový implantát formující přechodné řasy spojivky. Čes a Slov Oftal, 63; 2007: 369–372.
24. **Nouby,G.:** Congenital Upper Eyelid Coloboma and Cryptophthalmos. Ophthal. Plast Reconstr Surg, 18, 2902: 373–377.
25. **Mann, I.:** Development Abnormalities of the Eye. H. Kimptom, London,1957, p.371.
26. **Morax, S., Herdan, M.L., Hurbli, T.:** Orbito-palpebral Reconstruction in Two Case of Incomplete Cryptophthalmos. Adv Ophthalmic Plast Reconstr Surg, 9; 1992: 81–91.
27. **Murthy, R., Gupta H.:** Novel Surgical Techniqe for Management of Partial Cryptophthalmos. Indian J Ophthalmol, 62; 2014: 1096–1098.
28. **Neuhas, R.W., Hawes, M.J.:** Inadequate Inferior Cul-de-sac in the Anophthalmic Soket. Ophtalmology, 99; 1992: 153–157.
29. **Otradovec, J., Janovský, M.:** O krytoftalmu. Čs Oftal, 18; 1962: 128–138.
30. **Oztoprak, I., Erdogan, H., et al.:** A Case Scizencephaly Presenting with Unilateral Cryptophthalmos. Br. J. Radiol., 2008, doi: 10.1259/bjr/56258419.
31. **Saleh, G.M., Hussain, B., et al.:** A Sur-