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RAZMATRANJE TRIGONOCEFALUSA KAO PRIMERA KRANIOSINOSTOZE

TRIGONOCEPHALUS CONSIDERING AS EXAMPLE FOR CRANIOSYNOSTOSIS

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Kratak sadržaj

Kraniosinostoze, koje se javljaju zbog prevremenog spajanja jednog ili više šavova, najčešće su kraniofacijalne abnormalnosti. Tipični kranijalni deformiteti posledica su inhibicije rasta koja se javlja pod pravim uglom u odnosu na zahvaćene šavove. Funkcionalno oštećenje je uzrokovano direktno, razvijanjem patološkog oblika ili indirektno, povećanjem intrakranijalnog pritiska. Tipičan primer sinostoze je trigonocefalija. Trigonocefalični deformitet nastaje zbog prevremenog okoštavanja metopičnog šava. Rezultat sinostoze je ispupčenje čela. Oči se približavaju, što dovodi do hipotelorizma.
Indikacija za hiruršku intervenciju je potreba za povećanjem intrakranijalnog volumena i korekcija estetskog oštećenja u prednjem delu. U planiranju operativne procedure hirurg mora da razmotri orbitalne probleme.
Pregled opisuje različite oblike trigonocefalije i specifične hirurške pristupe korekciji.

Ključne reči: *kraniosinostoza, trigonocefalija, frontoorbitalno pomeranje*

Abstract

Craniosynostoses due to premature fusion of one or more craniofacial sutures are the most frequent craniofacial abnormalities. The typical cranial deformities result from growth inhibition perpendicular to the affected sutures. Functional impairment is caused either directly by the pathological growth pattern or indirectly by the increased intracranial pressure. Once typical Synostosis is the trigonocephaly. The trigonocephalic deformity is created by the premature ossification of the metopic suture. Synostosis results in a bulging of the forehead area. The eyes move closer together, resulting in hypotelorism.
Indications for surgery are the need to increase the intracranial volume and to correct esthetic impairments in the front. When planning the operating procedures, the surgeon has to consider the orbital problems.
This review describes the different disease patterns of trigonocephaly and the specific surgical approaches for the correction.

Key words: *craniosynostosis, trigonocephaly, Frontoorbital advancement*

Uvod

Pored prevremene sinostoze, koja obuhvata najveći broj slučajeva[1], grupa kraniofacijalnih malformacija obuhvata malformacije orbite, regije nosa i ušne školjke,

Introduction

Apart from premature synostoses, which represent the majority of cases[1], the group of craniofacial malformations comprises malformations of the orbitae, of the region of nose and

meningoencefalokele, facijalne rascepe, dizostoze maksile i mandibule kao i kraniofacijalne tumore.[2]

Orbitalna regija je od velikog značaja za izgled lica. Zbog toga, malformacije ove regije rezultiraju ne samo funkcionalnim, već i estetskim oštećenjima. Iz tog razloga, rekonstruktivne mere ne treba fokusirati isključivo na funkcionalnu, već i na estetsku rehabilitaciju pacijenta.[3,4]

Prevremeno spajanje kranijalnih šavova ima veoma teške posledice po orbitalnu regiju. Glavni simptomi su uni-ili bilateralni eksorbitizam kao hipo-ili i hipertelorizam.[5] Simptomi zavise od lokacije, broja i opsega sinostoza. Virchow je uvideo da prevremene sinostoze sputavaju vertikalni razvoj kosti u odnosu na zahvaćeni šav i da, u isto vreme, postoji intenzivnije napredovanje ka zahvaćenoj suturi. Po Virchowu, rezultirajući deformiteti lobanje prate poznati šablon, čija analiza objašnjava izvođenje zaključaka o zahvaćenom šavu.

U svom klasičnom članku "Uber den Cretinismus, namentlich in Franken, und uber die pathologische Schadelformen" on je opisao morfološke oblike bolesti, ali i njihov društveno istorijski značaj.[6] U prošlosti, specifičnosti malformacija u oblasti lica i lobanje bile su tumačene kao odraz mračnih sila i lošeg ljudskog karaktera. Ti ljudi su bili vređani i nazivani "poturenom decom" ili "čudovištima".[7] Vodili su skriveni život i kretali se samo u krugu porodice, ili su bili smeštani u ludnice. Često su spektakularno u javnosti bili predstavljani kao nakaze i čudovišta i njihova pojava pobuđivala je čuđenje ili prezir.[2]

Najraniji spisi koji opisuju ove malformacije se vezuju za Herodota (490–425 pre n.e.). On je opisao glavu koja se sastojala od samo jedne kosti, što je očigledno bio slučaj pansinostoze, posebno okoštavanja kranijalnih sutura.[8]

Naziv kraniosinostoza uveo je Bertoleti 1914. godine.[9] On je definisao kraniosinostozu kao prevremeno koštano spajanje sutura lobanje što dovodi do opšte ili transkribovane inhibicije rasta lobanje. Kraniosinostoze podležu dinamičnom procesu. Za vreme rasta javlja se nesrazmernost između kranijalnog volumena i mozga u razvoju što dovodi do povećanja intrakranijalnog pritiska. Kod kraniosinostoze sa jednom zahvaćenom suturom postoji 5–30%

auricle, meningoencephaloceles, facial clefts, dysostoses of maxilla and mandible as well as craniofacial tumors.[2]

The orbital region is of great importance for the appearance of the face. Therefore, malformations of this region result not only in functional but also in esthetic impairments. Therefore, reconstructive measures must not concentrate exclusively on the functional, but also on the esthetic rehabilitation of the patient.[3, 4]

Premature fusions of cranial sutures have very different consequences for the orbital region. Main symptoms are uni- or bilateral exorbitism as well as hypo- or hypertelorism.[5] The symptoms depend on the localisation, number and extent of the synostoses. Virchow realised that premature synostoses hamper bone development vertically in relation to the affected suture and that there is, at the same time, an increased expansion towards the affected suture. According to Virchow, the resulting skull deformities follow a set pattern, the analysis of which allows conclusions to be drawn about the affected suture.

In his classic article "Ueber den Cretinismus, namentlich in Franken, und ueber die pathologische Schädelformen" (Cretinism, namely in Franconia, and pathologic skull shapes) he described not only the morphologic disease patterns but also their socio-historical significance.[6] In the past, distinctive malformations in the regions of face and skull were considered as expressions of the powers of darkness and of the affected people's bad character. They were insulted as "changelings" or "monsters".[7] Their fate was a hidden life restricted to the immediate family, in lunatic asylums or institutions. Often they were made a spectacle to the public as monsters or curiosities and aroused astonishment or derision.[2]

The earliest writings describing these malformations go back to Herodot (ca. 490–425 BC). He described a head that consisted of one single bone, obviously representing a case of pansynostosis, i.e. the ossification of all cranial sutures.[8]

The term craniosynostosis was introduced by Bertolotti in 1914.[9] He defined craniosystosis as premature bony fusion of the sutures of the skull leading to general or transcribed inhibition of skull growth. Craniosynostoses are subject to a dynamic process. During growth, a discrepancy emerges between cranial volume and growing brain causing an increase in intracranial pressure. In craniosynostoses with one affected suture, there is a 5–30% risk of de-

rizika od povećanja intrakranijalnog pritiska, što zavisi od lokalizacije suture. Ako je zahvaćeno nekoliko sutura, rizik je > 90%.[10] Međutim, inhibicija rasta usled prevremene sinostoze nije ograničena na neurokranijum. Ona takođe vrši uticaj i na razvoj viscerokranijuma. Crouson je 1912. g.[11], opisao takvo oboljenje koje zahvata viscerokranijum.[10] Nekoliko autora[12-17] je napravilo kategorizaciju kombinacija kraniosinostoza koje su praćene dodatnim malformacijama, kao što je sindaktilizam, u definisane sindrome.

Simptomi bolesti, kao i njene ozbiljne posledice, rezultiraju indikacijama za hiruršku intervenciju. Fronto-orbitalno pomeranje napred, danas se smatra najsigurnijom tehnikom lečenja prevremene kraniosinostoze, čiji je razlog aktivno povećanje intrakranijalnog volumena putem napredovanja, pri čemu se istovremeno vrši korekcija malformacije. Funkcionalni i estetski rezultati postignuti ovom tehnikom mnogo su bolji u poređenju sa onima koji su postignuti ranijim hirurškim tehnikama.[5, 18]

Klasifikacija

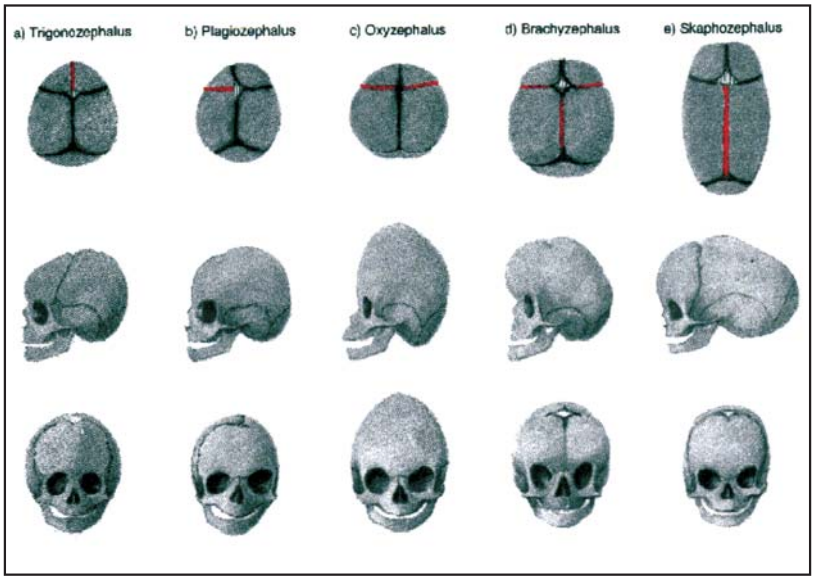
Oblici neuro- i viscerokranijuma su predstavljeni širokim spektrom pojava. Ekstremne varijacije od prirodnih varijacija oblika lobanje mogu da se definišu kao patološke. Po Marchac-u i Renieru[19], razlikuje se pet oblika lobanje u pogledu hirurških terapija. U zavisnosti od lokacije sinostoze razvijaju se tipične deformacije neuro- i viscerokranijuma (slika 1).

veloping an increase in intracranial pressure, depending on the localisation of the suture. If several sutures are affected, the risk is > 90%.[10] However, growth inhibition due to premature synostosis is not limited to the neurocranium, it also has an influence on the development of the viscerocranium. In 1912, Crouson[11] reported such a disease that affected the viscerocranium. Several authors[12-17] categorised combinations of craniosynostoses accompanied by additional malformations, e. g. syndactylism, into defined syndromes.

The symptoms of the disease, as well as its severe consequences, result in the indication for surgery. Fronto-orbital advancement to date is considered the safest technique for treatment of premature craniosynostoses, the reason being the active increase in intracranial volume, by means of the advancement, with concurrent correction of the malformation. The functional and esthetic results achievable by means of this technique are much better compared to those of earlier surgical techniques.[5, 18]

Classification

The shapes of neuro- and viscerocranium are marked by a broad and individual range of appearance. Extreme deviations from natural variations in skull shape can, however, be distinguished as being pathologic. For surgical therapy, five skull forms are distinguished according to Marchac and Renier.[19] Typical deformations of the neuro- and viscerocranium develop depending on the location of the synostosis (Figure 1).



Slika 1. Patološki oblici lobanje i klasifikacija po Marchacu i Renieru u Muhlingovom radu: Kirschnerishe allgemeine und spezielle Operationslehre, 1995.

Figure 1. Pathological skull forms and classification by Marchac und Renier in Mühling: Kirschnersche allgemeine und spezielle Operationslehre, 1995.

– trigonocefalija
– plagiocefalija
– skafocefalija
– oksicefalija
– brahicefalija

U sledećem koraku opisaćemo morfologiju i hiruršku proceduru u pogledu trigonocefalije.

– trigonocephaly
– plagiocephaly
– scaphocephaly
– oxycephaly
– brachycephaly

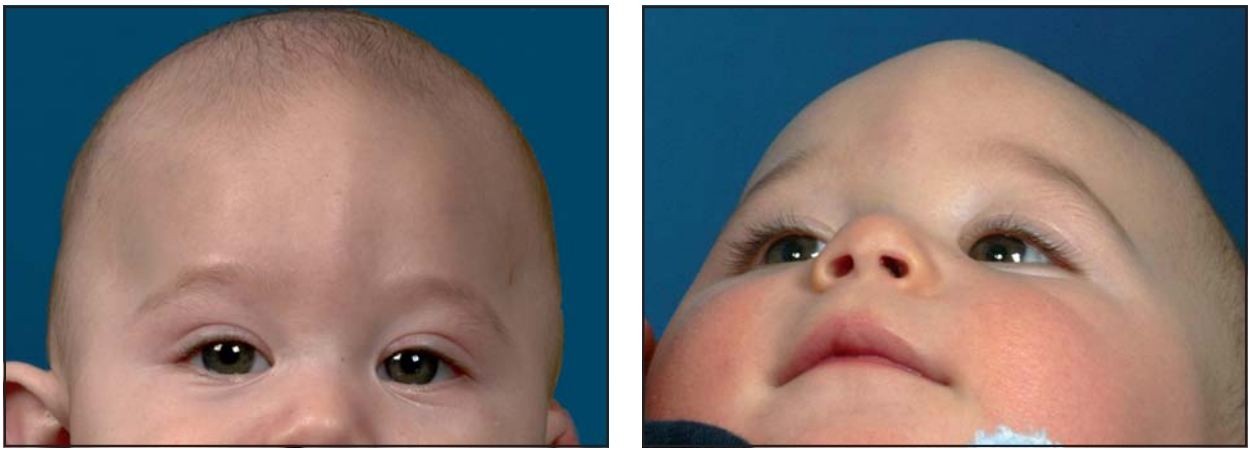
In the next step we will described the morphology and the surgical procedure considering as example trigonocephaly.

Trigonocefalija

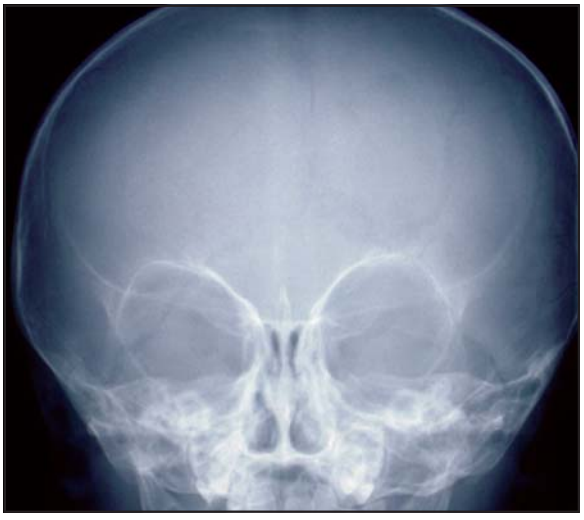
Razvoj trigonocefalije izazvan je prevremenim okoštavanjem metopične suture pri čemu čelo u horizontalnoj ravni dobija trouglasti oblik (slika 2). Izraženi obod kosti se može primetiti kod istog pacijenta. Istovremeno, frontolateralna regija se zaravnjuje. Čelo u celini je usko, dok okcipitalna regija izgleda prošireno. Oči se približavaju, što dovodi do hipotelorizma (slika 3). Rast srednjeg dela lica je ovde retko zahvaćen i uglavnom je regija srednjih režnjeva ograničena.

Trigonocephaly

The development of a trigonocephaly is caused by premature ossification of the metopic suture, the forehead taking a triangular shape in the horizontal plane (Figure 2). A prominent bone ridge can be observed in same patients. At the same time, the frontolateral region is flattened. The forehead as a whole is narrow, whereas the occipital region appears to be broadened. The eyes move closer together, resulting in hypotelorism (Figure 3). Midface growth is rarely affected and it is mainly the region of the frontal lobes that is restricted.



Slika 2. Trigonocefalus: prevremeno okoštavanje od metopične suture
Figure 2. Trigonocephalus: premature ossification from sutura metopica



Slika 3. Trigonocefalus sa hipotelorizmom
Figure 3. Trigonocephalus with hypotelorism

Hirurška tehnika

Na osnovu Tesierove[20] osnovne ideje i njenog daljeg razvoja od strane Marchaca i Reniera,[19] Muhling je definisao standardnu hiruršku šemu za lečenje kraniosinostoza i hipertelorizma.[1,3,5,18] Uz pomoć Muhlingovog standardnog fronto-orbitalnog napredovanja mnoštvo zbunjujućih aktuelnih hirurških tehnika se sada svelo na zajednički imenilac.

Tehnika fronto-orbitalnog napredovanja koristi lučni rez u hirurškom pristupu.

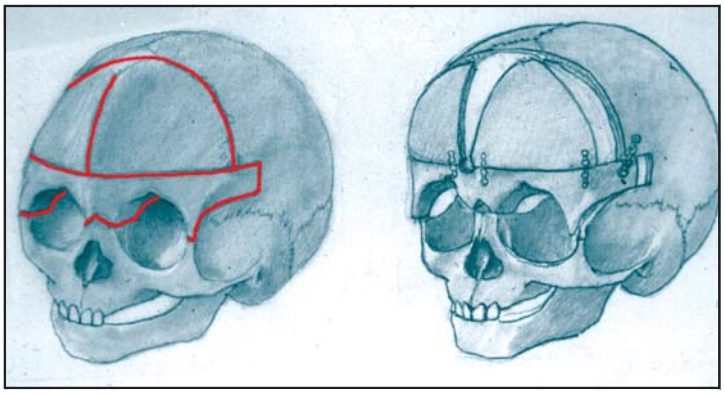
Postoji jedna standardizovana šema za osteostomije u pogledu svih patoloških oblika lobanje. Šema za osteotomiju i oblikovanje trigonocefalije prikazana je na slici 4.

Surgical technique

Founded on Tessier's[20] basic idea and its development by Marchac and Renier,[19] Mühling designed a standardized surgical scheme for the treatment of craniosynostoses and hypertelorism, respectively[1,3,5,18]. By means of Mühling's standardized fronto-orbital advancement the confusing variety of surgical techniques applied up to then was reduced to a common denominator.

The fronto-orbital advancement technique uses a bow cut for surgical approach.

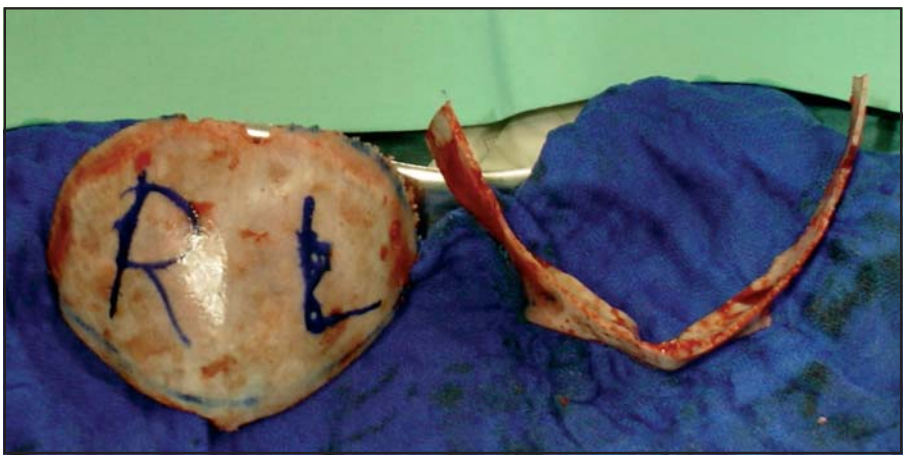
There is one standardized scheme for osteotomies for all pathological skull forms. The scheme for osteotomy and shaping of trigonocephaly is pictured in figure 4.



Slika 4. Hirurška korekcija pomoću trigonocefalije sa hipotelorizmom
Figure 4. Surgical correction of trigonocephaly with hypotelorism

Kraniotomija segmenta prednje kosti je praćena disekcijom i odvajanjem dure. Zatim sledi osteotomija orbitalnog segmenta. Nakon osteotomije i uklanjanja segmenta orbitalne kosti (slika 5), ovaj deo je oslabljen srednjom incizijom, klinastom ekscizijom ili redukcijom unutrašnjih kortikalisa. Zbog toga, segment se može saviti ili oblikovati, tako da se postiže normalni oblik čela. Dalje, orbitalni segment se

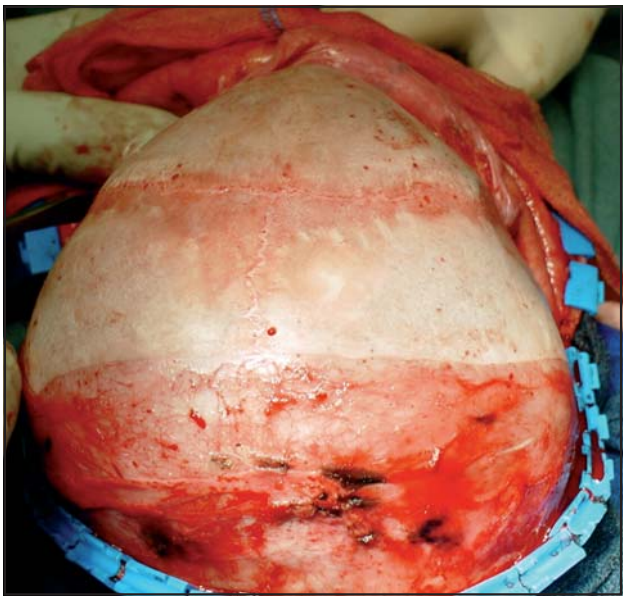
The craniotomy of the frontal bone segment is followed by dissection and detachment of the dura. Then follows the osteotomy of the orbital segment. After the osteotomy and removal of the orbital bone segment (Figure 5), this is weakened by median incision, wedge-shaped excision or reduction of its inner corticalis. Subsequently the segment can be bent and shaped so that the form of normal forehead is achieved.

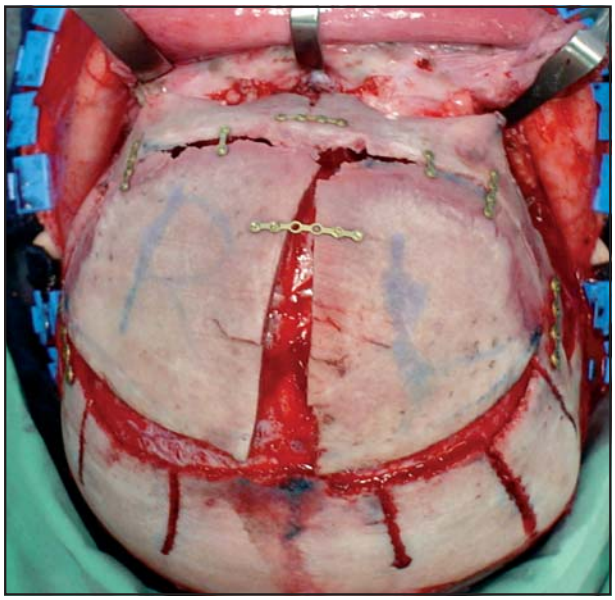


Slika 5. Uklonjeni fronto-orbitalni koštani segmenti trigonocefalijom
Figure 5. Removed fronto-orbital bone segments by trigonocephaly

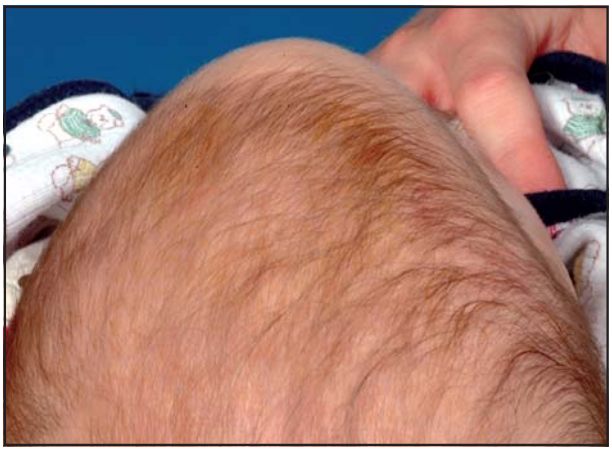
reimplantira uz pomeranje. Opseg koštanog pomeranja je uslovljen stepenom malformacije, naročito rastom oštećenja i spoljašnjim izgledom samog pacijenta. Po pravilu, radi se blaga, dodatna korekcija. Kao poslednji korak frontalni koštani poklopac se prilagođava novoformiranom orbitalnom segmentu, što ima za cilj formiranje skladne obline čela. To se postiže formiranjem koštanog poklopca uz upotrebu instrumenata za oblikovanje i fleksiju i ako je neophodno, njenom rotacijom. Segment je fiksiran titanijumskim pločicama (slika 6). Putem novoformirane intrakranijalne šupljine pritisak se može smanjiti odmah nakon operacije, jer sada postoji mogućnost da se mozak proširi unutar ove šupljine (slika 7).

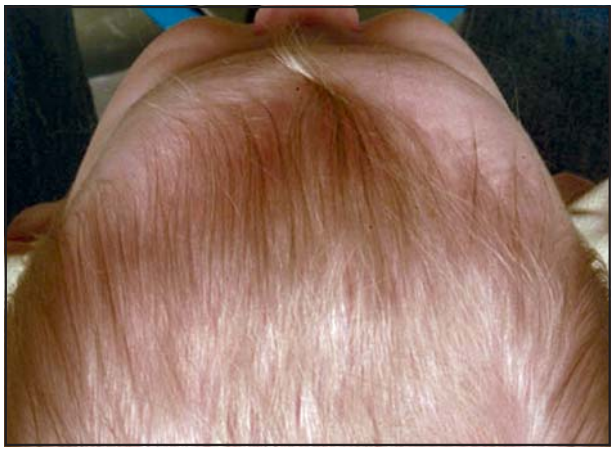
Next, the orbital segment is reimplanted with an advancement. The extent of the osseous advancement is determined by the degree of the malformation, i. e. of the growth impairment, and by the patient's outer appearance. As a principle, a slight over-correction should be performed. As a last step, the frontal bone cap is adjusted to the newly formed orbital segment, the aim being the formation of a harmonic curve of the forehead. This is achieved by forming the bone cap with the use of shaping and flexing instruments and, if necessary, by its rotation. The segment is fixed with titanium plates (Figure 6). By means of the newly formed intracranial cavity, pressure can be decreased immediately after surgery, as the constricted brain is given the opportunity to expand into this cavity (Figure 7).





Slika 6. Trigonocefalija pre- i postoperativno. Reponirani i pomereni fronto-orbitalni koštani segmenti, fiksacija mini- i mikropločicama preko kanala u vidu jezička
Figure 6. Trigonocephaly pre- and postoperatively. Repositioned and advanced fronto-orbital bone segments, fixation with mini- and microplates in the tongue-in-groove area





Slika 7. Trigonocefalija pre- i postoperativno, izgled čela
Figure 7. Trigonocephaly pre- and postoperatively, forehead appearance

Diskusija

Pomoću ove hirurške tehnike može se postići zadovoljavajuća korekcija malformacija ne samo u funkcionalnom, već i u estetskom pogledu. Kod starijih pacijenata postignuti rezultati se smatraju i konačnim. Međutim, kod dece, zbog dinamičnog procesa bolesti recidiv je uvek moguć, jer se pravi uzrok bolesti ne može eliminisati.

Zbog toga je dalje praćenje pacijenata od velikog značaja. Kontrolni pregledi se rade na šest meseci ili godinu dana i dužina tog intervala zavisi od stepena oboljenja. Praćenje uključuje kontrolu daljeg rasta lobanje kao i kontrolu intrakranijalnog pritiska. Vremenska odrednica druge operarcije ili ortodontskog lečenja kao i mere rehabilitacije se utvrđuju individualno.

Zaključak

Ciljevi operacije su i estetski i funkcionalni. Oblikovanje lica je neophodno zbog unapređenja društvene integracije deteta. Binokularni vid, adekvatna respiracija i razvoj govora su takođe neophodni za prihvatanje i razvoj.

Štaviše, ne smemo da zaboravimo da, uprkos tehnološkom napretku, naše operativne mere predstavljaju samo simptomatsku terapiju i ne uklanjaju uzrok bolesti. Stoga, buduća istraživanja ne bi smela da budu usmerena samo ka daljem razvoju hirurških tehnika, već i ka rasvetljavanju patologije ove bolesti. Možda će u budućnosti isključivo istraživanja poštedeti pacijente ovih komplikacija.

Discussion

By means of this surgical technique, satisfactory correction of craniofacial malformations – not only from the functional, but also from the esthetical point of view – can be achieved. In adult patients, the achieved result can be considered as being definitive. In children, however, due to the dynamic process of the disease a relapse is always possible, as the original cause of the disease cannot be eliminated.

Therefore, follow-up examinations are of utmost importance. The control examinations are performed in six-month or one-year intervals, the length of the intervals depending on the degree of the disease. Follow-up includes the control of the further skull growth as well as the control of the intracranial pressure. In this context, funduscopy and X-ray controls are important, in order to allow early recognition and surgical treatment of a relapse. Time points for secondary surgery or orthodontic treatment as well as all measures for rehabilitation have to be established individually.

Conclusion

The aims of the operation are both esthetic and functional. Shaping of the face is necessary to improve child's social integration. Binocular vision, adequate respiration, and speech development are also important for acceptance and development.

Furthermore, we must not forget that - despite all technical progress - our operative measures represent only a symptomatic therapy and do not eliminate the cause of the disease. Therefore, future research must not focus on further development of the surgical techniques alone, but also on elucidating the pathology of the disease. In the future, pure research may help to spare patients these complicated operations.

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