Surgical treatment of trigonocephalies and associated hypoteleorbitism

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Abstract

Premature closure of metopic suture is a relatively uncommon form of craniosynostosis with an estimated incidence of 0.3 per 1000 live births, comprising about 7% of surgical craniosynostosis referred to craniofacial centers. A broad phenotypical spectrum spreads from minor metopic ridges to severe trigonocephaly with pterional indentation supraorbital bar retrusion, temporal and parietal compensating bossings and hypotelorism.

Most of the cases arise spontaneously although autosomal dominant inheritance has been described and association with cromosomal abnormalities and different syndromes has been widely reported. Surgical correction has been attempted with good cosmetic results using several variations of the standard fronto-orbital advance. However there is still a number of questions to be solved in relation to this entity, mainly on its pathogenesis, but also on its development, natural history and treatment. Direct surgical approach to associated hypotelorism is a matter of argument when considering the reestablishment of normal interorbital distances.

We have conducted a retrospective analysis of our serie consisting of twenty-eight cases of trigonocephalies. Surgical correction of hypotelorism was attempted in eleven cases while the resting seven children remained "not treated". The objective was to review the functional outcome and cosmetic results comparing the different techniques applied to the frontal bone and to observe evolution of the hypotelorism after the treatment with or without osteotomies and grafting of the nasoethmoidal area.


Tratamiento quirúrgico de las trigonocefalias

Resumen

El cierre prematuro de la sutura metópica es una forma relativamente infrecuente de craneosinostosis con una incidencia estimada en torno al 0,3 por 1000 nacidos vivos y que comprende aproximadamente el 7% de todas las craneosinostosis referidas a una unidad de cirugía craneofacial. El espectro fenotípico es muy amplio y varía desde pequeñas crestas metópicas hasta formas severas de trigonocefalia con indentación pterional, retrusión de la barra supraorbitaria, prominencias temporales y parietales compensadoras e hipotelorismo.

En la mayoría de los casos surge espontáneamente aunque están descritos casos de herencia autosómica dominante y la asociación con anomalías cromosómicas y diferentes formas sindrómicas. El tratamiento consiste habitualmente en la corrección quirúrgica empleando diferentes formas de avance fronto-orbital. Los resultados cosméticos y funcionales son muy favorables. Existen sin embargo aún algunas cuestiones por resolver en relación con esta patología, principalmente sobre su patogenia, historia natural y tratamiento. El abordaje quirúrgico dirigido del hipotelorismo es un tema de discusión permanente al considerar el restablecimiento de las distancias interorbitarias normales.

Hemos desarrollado un análisis retrospectivo de una serie quirúrgica de veintiocho (28) casos de trigonocefalia. En once (11) casos se realizó tratamiento dirigido del hipotelorismo, mientras que en los restantes diecisiete (17) casos éste permaneció "sin tratamiento". El objetivo de este estudio es revisar la evolución desde el punto de vista estético y funcional comparando las distintas técnicas aplicadas al hueso frontal y observar la evolución del hipotelorismo tras el tratamiento con o sin osteotomías e injertos del área naso-ethmoidal.

Introduction

Premature closure of metopic suture is a relatively common form of craniosyostosis with an estimated incidence of 0.3 per 1000 live births, comprising about 7% of surgical craniosynostosis referred to craniofacial centers. A broad phenotypical spectrum spreads from minor metopic ridges to severe trigonocephaly with periorbital indentation, supraorbital bar retrusion, temporal and parietal compensating bossings and hypotelorism. Most of the cases arise spontaneously although autosomal dominant inheritance has been described and association with cromosomal abnormalities and different syndromes has been widely reported. Surgical correction has been attempted with good cosmetic results using several variations of the standard fronto-orbital approach. However there is still a number of questions to be solved in relation to this entity, mainly on its pathogenesis, but also on its development, natural history and treatment. Direct surgical approach to associated hypotelorism is a matter of argument when considering the re-establishment of normal interorbital distances.

We have conducted a retrospective analysis of our series consisting of twenty-eight cases of trigonocephalies. Surgical correction of hypotelorism was attempted in eleven cases while the resting seven children remained "not treated". The objective was to review the functional outcome and cosmetic results comparing the different techniques applied to the frontal bone and to observe evolution of the hypotelorbitism after the treatment with or without osteotomies and grafting of the nasoethmoidal area.

Patients and methods

In the period between January 1989 and December 1997, 349 cases of isolated or syndromic craniosynostosis were surgically treated in our Craniofacial Unit included in the Pediatric Neurosurgical Department.

Twenty four cases were diagnosed as isolated trigonocephalies. This accounts for 6.87% of all the cases. Syndromic or multiple synostoses were excluded of our study. Sixteen cases were males and 8 females with a ratio of 2/1. It takes the third place in frequency after sagittal synostosis (44.41%) and anterior plagiocephaly (17.47%). The average age at time of surgery was 6 months (range 3 to 60 months). The average follow-up period at the end of our study was 27 months (range 14 to 60 months)

Objective

To review the functional outcome and cosmetic results comparing the different techniques applied to the frontal bone. To observe evolution of the hypotelorbitism after the treatment with or without osteotomies and grafting of the nasoethmoidal area.

Preoperative evaluation

All cases were retrospectively reviewed on the basis of their medical chart and outpatient interview. Preoperative photographs were obtained from relatives or realized before and during the surgical procedures. Anamnesis related to pregnancy and labour was detailed. Genetic counseling was part of the evaluation to ensure recognition of syndromic trigonocephaly. In seven cases cariotype was also obtained, although it showed no abnormalities. Funduscoppy was performed routinely to discover papilledema or atrophic optic nerve.

Morphometric studies and examination showed characteristic dismorphic features consisting of a palpable bony ridge over the metopic suture with premature closure of the anterior fontanelle in more than 55% of the cases, widening of biparietal diameter and dimpling of the pterions bilaterally with the typical appearance of keel-shaped forehead. Frontal bones were retracted and hypoplastic. Supraorbital bandeau was therefore oblique and very often the presence of hypotelorbitism obvious, with intercrestal distance below the average measures. Anterior fossa was foreshortened and its volume decreased with compensating increase in biparietal width. Sometimes hypotelorbitism could be hidden by the presence of medial eipicanthus. Only 40% of our cases showed eipicanthus.

Preoperative assessment included measurement of the intercanthal width and the interorbital distance (internal intercrestalis) according to the standard values by Waitzman et al. Intercanthal width was considered a poor parameter for interorbital distance evaluation due to the fact that hypotelorbitism is often masked by a co-existing eipicanthus. It is also more difficult to obtain reliable measurements from non collaborative children. The same is true for interpupillary distance which can be also altered in patients presenting with strabismus. Hypotelorbitism was then diagnosed when the intercrestal distance was below 18.5mm or 20.5mm respectively to the first or second year of life.

Telerradiographies were obtained in all the cases. CT helicoidal scanning with 3-D reconstruction was performed routinely in all the patients since 1993.

Methods

Patients were reviewed in outpatient consulting at 1 month, 6 months, 1 year and 2 years postoperatively. In 15 cases was possible to obtain postoperative CT studies. Measures were taken from conventional X-rays films in those children when CT was not disposasible. Postoperative photographs were included in the medical chart. At 12 months follow up, measures were taken on plain radiographs or CT films to obtain (a) intercrestal width; (b) intertemporalis distance; (c) lateral orbital distance; (d) biparietal and; (e) nasion projection distance. The values were com-
pared to age-matched normal subjects (Gunter) pre and postoperatively and analyzed taking into account if medial osteotomy and grafting was performed or not to treat hypotelororbitism during surgical procedure.

All the children were evaluated by three different members of the unit and included in groups for cosmetic result. Bartlett categories are commonly used in our department because of its objectivity and reproducibility. Any case with discordance among the evaluating members was assigned to the lowest category. In the same way, when a patient deteriorated after the course of the first year of follow up he was included in the worst group.

Results and inclusion in categories were analyzed according to the treatment of the frontal area. We intended to observe different outcome features in relation to the surgical technique employed on the frontal bone reconstruction.

**Surgical technique**

The timing for surgical intervention depended on the arrival of children to our unit. They were commonly referred by pediatricians involved in primary care and sometimes by neurosurgical services without experience in the treatment of craniofacial deformities. Children presented thus with a broad range of ages, from 3 to 60 months. One case with a mild form of metopic ridge was operated at the age of 5 years for recontouring of the frontal bone without advancement and is not included in the series. In the resting cases surgery was electively performed between 3 and 6 months of age.

A standard bicoronal approach is scheduled with retroauricular incision and elevation of a bifrontal flap in the loose areolar plane between periostium and galea pericrania. Frontal branches of the facial nerve run inside the external fascia of temporalis muscle so care must be taken in order not to damage them. However these patients often show a low implantation of the temporalis muscle and dissection under the deep fascia of the muscle opens a broad exposition of the pterional and lateral wall of the orbits area. Periostium is incised at this level and dissection proceeds in the subperiosteal plane, exposing both frontal and pterional areas, lateral walls of the orbit with partial elevation of temporalis muscle to expose greater wing of the
sphenoid and superior wall of the orbits up to the internal canthus which is usually preserved.

Before starting osteotomies the obliquity of frontal bones, retraction of the lateral portions of the orbital bandeau, and pterional indentation are assessed. A bifrontal craniotomy is performed after a supraorbital bandeau is created. The marks are extended from the sphenosquamosal suture to its contralateral equivalent prepared for a tongue-ingroove advancement. The bifrontal flap includes the anterior part of the bregmatic fontanelle and both coronal sutures. Anterior fossa is exposed extradurally with visualization of the anterior two-thirds of the orbital roof.

Osteotomies on the orbitofrontal bandeau are now performed starting on the lateral orbital ridge over the fronto-zygomatic suture, and opening of the orbital roof including the portion over the cribiform plate at the mid-line. There is commonly a severe sphenoid thickening at the pterional level where osteotomies are the last step to obtain a free orbital bandeau. Pterions are resected up to the level of the anterior fossa. The bandeau is anteriorly displaced with rotation of the lateral extremes of the "bandeau" projecting forwards, maintaining the medial aspect of the bandeau in place. Orbital roofs and supraorbital ridge are aligned in proper position, and everything fixed in place with absorbable miniplates (Lactosorb®). After recontouring of the frontal bone closure proceeds in the standard way with anterior rotation and elevation of temporalis muscle bilaterally and covering of the field with periostium and galea. We place two subgaleal drains and close skin with subcutaneous absorbable sutures and staples.

**Treatment of hypotelorbitism**

Eighteen (18/28) of our patients presented with intercrestal distance below normal values. In 11 cases reduction of hypotelorbitism was accomplished. Intercanthal distance was increased expanding the naso-ethmoidal space by dividing internasalis suture and spreading of ethmoidal bone plates laterally. Displacement was kept in place by interposition of a wedge-shaped bone graft from calvarial donor-site. An absorbable miniplate from the frontal bone to the nasion helps to keep it in place. Concomitant expansion of lateral walls of the orbit is necessary to maintain proper orbital relation. In the resting seven patients no attempt was made to treat the hypotelorbitism.

There were no apparent inadvertent detachment of the medial canthal ligament. Lateral canthopexy was occasionally needed.

**Recontouring of frontal bone**

Different techniques were applied to the treatment of the frontal bone. In two cases with mild metopic ridges, the frontal bone was left attached and recontoured by simple drilling. Four cases received a "shell" technique recontouring as described by Di Rocco. In eight more cases two hemifrontal flaps were elevated with inversion and rotation of 1800 to obtain a normal shape of the definitive frontal bone. In the 10 resting cases a Marchac template served as a guide for grafting of a new frontal from parietal donor site.

**Results**

There was one case of mortality referred to a patient.
in whom unadverted tear of duramater during dissection of anterior fossa occasioned a fulminant meningitis in the third day of the postoperative period. Considering the whole surgical craniofacial serie total mortality was 0,5 \% (2/349). One case with a Marchac transposition presented with persistent craniolacunia after surgery and had to be reopened five years later to cover the defect. No other major or minor complications where related to the procedures. In the late cases of the serie, absorbable miniplates were used preventing complications related to the non-absorbable wires or miniplates.

**Cosmetic results**

All the children were evaluated by three different neurosurgeons, two staff of our surgical unit (other than the surgeon) and one senior resident and assigned to different categories according to the final postoperative appearance. The classification proposed by Barlett et al for the evaluation of cosmetic results in craniostenosis was elected because of its objectivity and reproducibility in evaluating different items. **Category I:** Included excellent result with no imperfections; **Category IIA:** Minor imperfections but no surgical revisions considered necessary; **Category IIB:** Minor imperfections. Soft tissue or lesser bone-contouring revisions advisable; **Category III:** Major alternative osteotomies or bone grafting procedure needed; **Category IV:** Major craniofacial procedure advisable, duplicating or exceeding the original procedure. Category I was considered as an excellent result (20 cases). Categories IIA (2 cases) and IIB (2 cases) were considered as good results, while categories III (3 cases) and IV (1 case) were considered bad results. In total 24 cases were classified as excellent or good results while 4 were categorized under poor result.

Bad results were analized individually in order to find an explanation for the poor final appearance. Among cases categorized as Barlett III, case 1 was the first operated in our unit. Surgical technique consisted only in inversion of both frontal hemiflaps without osteotomies of the orbito-nasofrontal "bandeau" and thus the technique was considered incomplete. The children was 24 months by the time of the operation and followed up for 7 years. Family was happy with final result and rejected new intervention for total reconstruction. Cases 12 and 13 obtained also Barlett III. Case 12 was operated at 32 months of age and case 13 with 5 years. Both cases were referred from distant centers without consultant pediatric neurosurgeon and delayed diagnosis. In both children a kind of syndromic disease was suspected because of peculiar characteristic phenotype and minor malformations, although syndromologist were not able to include them in any of the known categories. Both suffered from minor mental retardation and ulterior surgical treatment were also denied by the families. Initial surgical intervention consisted in 93shell94 technique for the frontal bone in case 12 and inversion of the hemifrontals.
in case 13. In both of them treatment of hypoteleorbitism was accomplished by nasion osteotomy and medial grafting. Failure was imputed to delayed treatmet and not to surgical kind of intervention.

The only case included as type IV deserves mention apart. It was a severe trigonocephaly responding to a type II of Di Rocco’s classification4, with severe pteronial indentation and hypoteleobitism. It was operated at 7 months of age. A parietal donnor site was employed as a new frontal and hypoteleorism not treated. Final result was considered very bad and a new procedure proposed and developed by the age of 2 and a half years. This time hypoteleorbitism was treated and the former frontal employed with splitting and rotation of both hemiflaps. This case made number 11 in the serie and our department had accomplished a wide experience in the treatment of craniosinostosis by that time. Unsatisfactory result had to be explained because of an incomplete and bad technique. Finally this boy was categorized as IIB.

**Hypoteleorbitism**

According to Waitzman recommandations31 different craniofacial skeletal measurements were made pre and postoperatively in the neuroimaging studies. Medial orbital wall protrusion, intertemporalis, intercoronoral, lateral orbital walls and medial intercanthalis (between both lacrimalis crestae) distances were elected as cephalometric assess-
Discussion

Closure of the metopic suture starts under normal circumstances at the end of the first year and may last until the end of the second year. When premature sutural closure occurs, usually before birth, a typical craniofacial malformation develops. The term trigonocephaly was first coined by Welcker in 1862. Since then it has been easily recognized and thoroughly operated. Several techniques have been described and good cosmetic results obtained. However there are still a number of questions to be solved in relation to this entity, mainly on its pathogenesis, but also on its development, natural history and treatment.

Trigonocephaly is considered a fairly common type of craniosynostosis. Formerly denoted as unusual, it represents nowadays about 10% of all patients related to craniofacial centers. Its incidence is estimated to be 1 in 2,500 births and there is a male predominance of 65-85%.

The premature arrest of growth of the metopic suture may present as a spectrum of manifestations, depending on the timing and extent of the sutural closure. In its mildest form it is a familial and ethnically inherited facial morphology. Milder forms of metopic synostosis consist only in a more or less prominent metopic ridge that does not need surgical intervention. In severe forms a characteristic keel-shaped forehead can be observed, with absence of frontal eminences, retruded orbital rims and hypotelorism and epicanthus giving the peculiar craniofacial appearance to those children. It seems possible that there are different degrees of involvement of the anterior chondrocranial structures like pre-ethmoid, mesoethmoid and the ectoethmoid sutures. Another different explanation for the broad phenotype of affected children could be a non-specific process acting at different evolutive ages. Trigonocephaly may appear either as an isolated anomaly or as part of syndromes involving prosencephalic or rhinencephalic structures (holoprosencephaly) such as Optiz C syndrome, Sav-Meyer syndrome or Frydman syndrome. Recently a new fronto-ocular syndrome has been described with trigonocephaly. Several genetic defects have been described associated to trigonocephaly. Abnormalities including deletions and duplications of chromosomes 3p, 9p, 1q, 13q have been published. Isolated trigonocephaly has an unknown etiology. Autosomal dominant inheritance with very low penetrance has been proposed in 2-5% of the cases for familial cases. The possible involvement of mutated FGFRs (fibroblast growth factor receptors) in newborns affected by trigonocephaly has recently been investigated by molecular screening, searching for mutations in FGFR2 which has been implicated in complex craniofacial syndromes. However none of the cases studied carried mutations except one evolving later towards a Crouzon-like profile.

Mental delay may occur in isolated trigonocephaly and has been observed in as much as 10% of these children. Development delays may consist of subtle changes in the time of acquisition of head control or sitting position or include more severe forms of mental retardation, with or without intracranial hypertension.

Di Rocco has recently proposed two different subgroups on the basis of clinical and radiological findings. Group I presents with bilateral frontal bone hypoplasia associated with extreme retraction of the supraorbital margins. In this group hypotelorism is associated with abnormally deep position of the cribriform plate, giving ethmoidal region a hollow appearance. In these patients the nasion-pterional angle is severely restricted and the nasion-ethmoidal distance significantly increased. Group II shows also bilateral frontal hypoplasia with hypotelorism, supraorbital retraction and reduced naso-pterional angle.

However, the nasion-ethmoidal distance is almost normal and pterional evidence is scarcely evident. Moreover, patients in group II showed a lesser degree of temporal compensatory expansion. He postulates that in some patients a compensatory elongation of the nasion-ethmoidal distance and an incomplete synostosis of the fronto-ethmoidal sutures allowing for a partial lateral expansion of the anterior cranial fossa could diminish the necessity for posterior calvarial expansion. On the other hand, children with more severe involvement of the naso-ethmoidal sutures resulting in a diminished lateral expansion of the anterior fossa would determine the need for compensatory changes in the temporal and parietal regions. Patients in group II accomplished good correction of associated hypotelorism whereas patients in group I did not reach normal interorbital values when confronted to the same surgical procedure that did not include specific treatment of hypotelorism. Other authors have addressed the importance of the moment and degree of involvement of pathological changes in the anterior chondrocranial structures. Milder forms of trigonocephaly would affect only the upper metopic suture while more severe forms include involvement of pre-sphenoid, meso-ethmoid and ecto-ethmoid structures.

Since Tessier and his neurosurgical colleagues proposed a new surgical approach to craniofacial deformities different techniques have been proposed to treat this condition with reports of satisfactory results in many series. The standard approach consists of bifrontal craniotomy and remodelling and frontal-orbital advance with recontouring. However there is not a consensus yet when considering the hypotelorism correction. Several papers have addressed the importance of associated hypotelorism and the need for direct surgical approach. Different authors propose adding a nasofrontal osteotomy and an interpositional bone graft to the supraorbital bar and nasoethmoidal area to correct hypotelorism simultaneously with lateral orbi-
tary wall expansion or three-quarter orbital wall osteotomies. On the other hand there are authors who do not advocate direct approach to the hypotelorism in the believe that internasalis grafting widens only nasal bones without increasing interorbital width as far as osteotomies remain usually anterior and superior to nasoethmoidalis complex. On this basis they recommend standard approach without grafting and thus consider the latter ineffective and unnecessary. Both techniques have achieved statistically significant improvement in hypotelorism, greater than expected from normal growth curves. However undercorrection of hypotelorism and persistance of abnormally low interorbital distances is frequent when ordinary widening is not addressed.

Our serie shows postoperative improvent of every parameter in comparison to preoperative values as much in "treated" (corrected hypotelorbitism) as in "nontreated" patients.

Hypotelorbitism improves in all the cases although amelioration is higher in patients where nasofrontalis complex was treated. However a number of patients in both groups were not able to reach normal values. None of the children whose hypotelorism was left untouched reached normal values at two years follow up.

Ours is a retrospective non-categorized study and the number of patients too low to draw out definitive conclusions in a complex and debatable topic. However, and according to our cases review we favour the treatment of hypotelorbitism by splitting and grafting of nasofrontalis complex along with lateral orbital expansion. None of the "non-treated" group reached normal intercrestals distance while 62.5% of the treated group were in normal. This a question that in our understanding remains to be solved and further studies including strict preoperative categorizing and prospective randomized studies need to be multicentrically worked out.

Conclusions

Surgical treatment of isolated metopic synostosis is feasible with good cosmetic results and low morbidity. Outcome is much better when operation is promptly afronted. It is very important to rise inquietude in pediatricians and in primary care attention for early diagnosis that will undoubtedly favour final results.

The standard technique obtains good cosmetic results and armonization of the anterior fossa diameters independently of the treatment applied to the contouring of the frontal bone. Probably it is better to use a parietal grafting under Marchac pattern for children older than 1 year of age.

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The question of whether splitting of the nasoethmoidal area should be accomplished or not for the treatment of the associated hypotelorbitism in trigonocephaly remains a subject for discussion. Further prospective studies with categorized groups should be developed. Facilities for image transmission in the interhospitalary network era could help to establish a multicentric database.

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