Surgical Planning and Correction of Median Craniofacial Cleft

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ABSTRACT

Median facial cleft (Tessier 0-14 cleft) is a very rare deformity which affects one child in 100,000 live births. It varies from small median cleft of the upper lip or a small midline notch of the nose to extreme median craniofacial cleft involving the cranium and face with severe orbital hypertelorism. We report on 9 cases of median facial clefts with varying degrees of severity. The importance of staged well planned and timed operations is discussed. Correction of the cleft lip is done with correction of the soft tissues of the bifid nose by Z or V-Y plasties, while the cleft palate is done at earlier time to prevent speech problems. Orbital hypertelorism is corrected by facial bipartition operation as early as 2 years of age with placement of costochondral graft for nasal augmentation in addition to meningocele excision if present. We present our limited experience with these cases with discussion of the cosmetic and functional results and the complications encountered.

INTRODUCTION

Tessier is credited for the creation of the craniofacial surgery since he laid the foundation of the modern craniofacial surgery by systemically analyzing facial clefts and describing facial osteotomies. In addition to that, he simultaneously used an extra and intra-cranial approaches to correct craniofacial anomalies and eradicate craniofacial tumors [1]. Traditional descriptive embryology based upon the interaction of frontonasal, lateral nasal and medial nasal prominences is incapable of explaining the three-dimensional development of the facial midline. Carsten [2] recently postulated that the internal structure of the nose and that of the oronasal midline can best be explained by the presence of paired A fields originating from the prechondral mesoderm, associated with the nasal and optic placodes, supplied by the internal carotid artery and sharing a common genetic coding with the prosomeres of the forebrain. Mesial drift of these fields leads to fusion of their medi-
between the maxillary central incisors, bifid nose of varying degrees, central alveolar cleft, cleft premaxilla, cleft palate, duplicated or thickened nasal spine and septum, the nasal cartilages are displaced laterally and usually hypoplastic, the nasal bones are widely separated, the ethmoidal air sinuses are wide and the air cells are increased in number and there is increased distance between the medial orbital walls (orbital hypertelorism). The cranial part of the cleft may manifest itself as hypoplasia of the central cranial structures which may lead to interfrontal or frontoethmoidal meningoencephalocele [5-8].

Orbital hypertelorism is defined as increased distance between the medial orbital walls (IOD), while telecanthus is increased distance between the medial canthi. 70% of the IOD is attained by the age of 2 years and the mean normal values are between 15 mm to 23 mm at the age of 12 years [9]. Treatment of orbital hypertelorism is an integral part of dealing with this problem. The classic procedure is the facial bipartition operation which entails simultaneous intracranial and extracranial approaches to accomplish osteotomies to free the supraorbital rims and the medial orbital walls with the adjacent portion of cranial base and moving this orbital bones medially, downwards and posteriorly followed by fixation [10-12].

An existing meningoencephalocele is dealt with if present. These occur either with facial clefts number 0-14 or 1-13 in which they are associated with orbital hypertelorism and when they are not associated with facial cleft there might be telecanthus with displacement of the medial orbital walls and deformation of the orbits rather than true orbital hypertelorism [13,14].

Correction of the bifid nose deformity is achieved by thorough understanding of the bifid nose anatomy, careful planning of incisions and meticulous surgical technique [15]. Given the rarity and the unique nature of the facial clefts, the treatment plane cannot be standardized but this depends on the individual assessment of each case. Nevertheless, there are general principles by which the sequence and timing of staged osseous and soft tissue reconstruction can proceed [5]. The complexity, severity and multiplicity of the cosmetic, functional, educational and psychological problems in these patients dictates the cooperation of a craniofacial team to achieve satisfactory results.

**PATIENTS AND METHODS**

This is a retrospective study conducted on patients with median facial clefts (Number 0-14 Tessier clefts) who have been treated in Menoufiya university by the craniofacial team in the period between 1994 and 2002. These patients were presented to the plastic surgery clinic during this period. According to the existing morphological features and extent of the deformity, these patients can be divided into four groups:

**Group 1 (3 cases):** Includes patients who have major median facial cleft involving the upper lip, the alveolus, the palate and the whole nose in addition to orbital hypertelorism. Case number 1 of this group is characterized by severe orbital hypertelorism, frontonasal Hamartoma and atresia of the pyriform openings leading to nasal obstruction (Fig. 1a). Case number 2 is characterized by moderate orbital hypertelorism, bifid nose, cleft palate, cleft vermillion of the upper lip and atresia of the nasal pyriform (Fig. 2a). Case number 3 is similar to case number 2 but without nasal obstruction. CT scan examination reveals enlarged and increased number of anterior ethmoidal air cells and features of nasal affection.

**Group 2 (2 cases):** Includes patients with anterior meningocele and orbital hypertelorism. Case number one in this group was a 14 months old girl with neglected tense globular frontoethmoidal meningoencephalocele, which is 6 cm wide and 11 cm long with orbital hypertelorism, the overlying skin is very thin with impending rupture (Fig. 3a). Case number 2 in this group was a 3 year old boy with frontoethmoidal meningocele with a diameter of 2 cm and orbital hypertelorism. In both cases the cleft does not affect the lower third of the nose or the upper lip. CT scan examination of the first case indicated that the defect of the skull was replacing cisterna Galli and cribriform plates and about 3 cm in diameter (Fig. 3b). The other case scan shows 2 cm defect anterior to the cisterna Galli, cecum.

**Group 3 (3 cases):** These are patients with milder degree of bifid nose deformity with mild orbital hypertelorism. Case number one in this group is an 11 years old boy with bifid nose affecting mainly the lower half of the nose with absence of the nasal septum and mild orbital hypertelorism (Fig. 4a). The second case of this
group is a 12 year old female with milder degree of bifid nose and no orbital hypertelorism. Case number three is a 18 year old female patient with the cleft affecting mainly the upper two thirds of the nose leading to very widely separated nasal bones and very wide pyriform openings and orbital hypertelorism.

**Group 4 (1 case):** This case is characterized by median cleft of the upper lip and alveolus without affection of the palate and nose or orbital hypertelorism.

Preoperative CT scan, laboratory investigations and routine pediatric or medical consultation for pediatric and adult patients were done. In addition to that, routine ophthalmic examination for patients with orbital hypertelorism. Otolaryngological examination is also done for patients with nasal affection to examine the internal structures of the nose.

**Surgical technique:**

The objectives of surgical procedure are directed toward correction of the bifid nose deformity, nasal obstruction, median cleft lip, cleft palate, excision of an existing meningocele and correction of orbital hypertelorism.

**Bifid nose correction:**

In patients of group one, this is done by trying to approximate the widely separated nasal halves through careful planning of skin incisions. In case number 1 this could be achieved through Z plasty (Fig. 1b) and in case number 2 by V-Y flaps to bring the nasal tip down and in the same time reduce the soft tissues between the eye brows which helps to improve the result of skeletal correction of orbital hypertelorism (Fig. 2b). Costochondral grafts are used in a second sitting during facial bipartition to raise the nasal tip and the dorsum of the nose. This is used as spreader graft to its caudal part the upper and lower alar cartilages are fixed while the cephalic part is fixed in the glabellar region.

In group two patients, the nasal correction is achieved at the time of excision of the meningocele. We preferred at this stage to limit the nasal reconstruction by gathering the tissues together after excision of the thinned out abnormal skin to guard against any skin necrosis which might endanger the dural repair (Fig. 3f). In a second step the costochondral graft is used as stated before followed by excision of the previous scar.

In group three patients, a single step rhinoplasty by lateral nasal osteotomy to bring the nasal bones together and costochondral graft utilization for nasal augmentation as mentioned before (Figs. 4a,b). In case number 3 of this group, the widely opened pyriform openings were felt intraoperatively as a defect lateral to the nasal bones after the lateral osteotomy and this required placement of a bone graft from the costochondral graft.

Nasal obstruction has been encountered in the first and second cases of the first group. The atresia of the pyriform openings are dealt with by intraoral exposure and osteotomy to widen the narrowed openings. Postoperative nasal tubes are kept in place as splints for one week.

**Cleft lip and palate:**

Correction of the median cleft lip is done by excision of the central tissues at the edges of the cleft in the stage of correction of the bifid nose deformity. Then repair is done utilizing either Z-plasty to elongate a shortened lip-nose distance or by direct sutures if the lip-nose distance is normal. Cleft palate is encountered in the first group and repaired in a separate sitting utilizing the Von Langenbeck technique to prevent speech problems.

**Orbital hypertelorism:**

This is corrected in patients belonging to the first and second groups (5 cases). The technique of “facial bipartition” is utilized to correct the orbital hypertelorism and facilitate bifid nose correction.

This is started by a bicoronal incision of the scalp extending to the preauricular area in both sides followed by subperiosteal dissection of the anterior scalp flap. When the deep temporal fat is reached laterally subperiosteal dissection of the temporalis muscle is done until we reach the zygomatic archs. The anterior scalp flap is then everted to expose the supraorbital ridges and the whole lateral orbital walls bilaterally. Exposure of the anterior cranial fossa is achieved by elevation of the bifrontal craniotomy. This is done through Burr holes which are placed bilaterally above the temporal crests and parasagittally posterior to the coronal suture and in the frontal bone. The anterior and lateral maxillary walls are exposed through intraoral
midfacial degloving incision saving the infraorbital nerves and vessels which lies 7 mm below the infraorbital rim. Orbital dissection is done by subperiosteal dissection of orbital soft tissues as deep as 20 mm from the orbital margins for 360 degrees. Taking care to preserve the nasolacrimal duct. Using oscillating saw osteotomy is performed in the following regions (Figs. 5a,b):

1- The orbital roofs 10 cm anterior to the optic nerve after backward retraction of the frontal lobes extending medially to the cribriform pates which are saved.

2- The lateral orbital walls and malar bones after retraction of the temporal lobes medially.

3- The infraorbital region, 10 mm below the infraorbital margin through an intraoral midfacial degloving incision preserving the infraorbital nerves and vessels.

4- The orbital floor by very fine osteotome.

5- The widened interorbital regions, by two vertical paramedian osteotomies to remove the central bony segment containing the ethmoid air cells with care to save the cribriform plate and olfactory nerves. This is followed by approximating the two orbits medially and any bony connection which might hinder the mobility is dealt with by very careful osteotomy with a chisel (Figs. 5a,b). Fixation is done in these cases by minimal number of miniplates and screws. The detached medial canthi are fixed through drills posterior and higher to the lacrimal grooves. The frontal bone flap is returned and fixed by miniplates and screws. In the first case of the first group no bicoronal bone flap was done and we could achieve satisfactory orbital mobilization after excision of the median segment which allowed osteotomy of the orbital roof (Figs. 1c,d,e,f,g).

In the second group, excision of the meningocele is done through simultaneous intracranial and direct approaches. The intracranial approach is done through bifrontal bone flap elevation and traction of the frontal lobes followed by exposure of the anterior cranial fossa, while the direct approach is done through incision in the stretched abnormal skin overlying the meningocele with every attempt done to remove this skin. Careful dissection is performed to expose the defect in the anterior cranial fossa and excise the meningocele. In one case, abnormal disorganized brain tissues in the meningocele was excised. The bone defect is closed by bone graft and the defect in the dura is closed carefully. The procedure of facial bipartition is performed as described before to correct the orbital hypertelorism. Preoperative ventriculoperitoneal shunt has been used in case number one with very tense meningocele one week before the surgery to decrease the intracranial tension in the perioperative period. This facilitates excision of the meningocele, traction of the frontal and temporal lobes during osteotomies and prevents postoperative increase of the intracranial tension and hence cerebrospinal fluid leaks (Figs. 3c,d,e,f,g).

RESULTS

The results of surgical correction of this multifactorial problem are very gratifying as regard the cosmetic and functional outcome. Despite the magnitude of the this multistaged surgery, the complications were not major indicating the unexpected compliance of children to this surgery. We did not encounter any mortality perhaps due to absence of anesthetic complications in addition to absence of bleeding catastrophe. Also neurological complications as CSF leak or brain injury were not encountered in the four cases with intracranial operations. Reconstructive complications were encountered as partial relapse of the orbital hypertelorism in case number one, which we contribute to severity of the orbital hypertelorism and to inadequate exenteration of the nasoethmoid complex (Fig. 1g). Cutaneous sinuses have been encountered in two cases. The first was due to partial costochondral graft necrosis in nasal correction and treated conservatively by repeated curettage and antibiotics until it healed and then additional costochondral graft was inserted after six months. The second case was due to extrusion of bone wax and treated by excision of the sinus and evacuation of the wax. Nasal deviation has occurred in one patient of the third group due to inadequate fixation of costochondral graft and this required revision surgery. Long nose deformity occurred in case number 4 (Fig. 3g). Chest infection has occurred in one child and was controlled by antibiotics (Table 1).
Fig. (1-A): Median facial cleft.

Fig. (1-B): Z-plasty to correct bifid nose with cleft lip correction.

Fig. (1-C): Preoperative planning of facial bi-partition.

Fig. (1-D): Excision of the central bony segment containing the ethmoids.

Fig. (1-E): Mobilization of the orbits medially to cross the gap after central bony excision.

Fig. (1-F): Fixation of the craniofacial skeleton by miniplates and screws.

Fig. (1-G): Same patient after completion of surgery.
Fig. (2-A): Median facial cleft with severe orbital hypertelorism.

Fig. (2-B): V-Y flap to bring the nose down and approximate the eye brows.

Fig. (2-C): Same patient after completion of facial bipartition and cartilage graft of the nose.

Fig. (3-A): Frontoethmoidal meningoencephalocele with impending rupture.

Fig. (3-B): CT scan of the same case.

Fig. (3-C): Collapse of the meningoencephalocele after ventriculoperitoneal shunt.
Fig. (3-D): The defect at the base of the anterior cranial fossa with the forceps passing from the nose of the anterior cranial fossa.

Fig. (3-E): Closure of the defect by bone graft with rigid fixation.

Fig. (3-F): Closure of the nasal defect by tissue gathering to prevent any skin lose which may endanger the bone graft or the dural closure.

Fig. (3-G): Postoperative of the same patient.

Fig. (4-A): Mild median facial cleft of the nose with mild orbital hypertelorism.

Fig. (4-B): Same patient after costochondral graft.
DISCUSSION

Craniofacial anomalies are grouped into two categories, those which involve failure of fusion or disruption of embryological units leading to facial clefting, or those which involve premature closure of cranial sutures leading to craniostenosis. Children with craniofacial anomalies suffer the severest degree of psychological and educational drawbacks, which is usually not due to any mental disorder but due to inability to interact normally with their community because of the apparent deformity [16,17]. Plastic surgeons have great responsibility toward their community as regard public education through the different media to help parents and teachers to detect and seek medical advice whenever they are confronted with these children.

The timing of the management is very crucial to avoid psychological disorders and pre-
and his colleagues have studied the maxillary cleft growth. In major midline facial cleft the cleft lip and palate repair is started with bifid nose correction. Cleft palate repair is usually done before referral of patients to us by other surgeons to avoid speech disorders. Bifid nose associated with facial cleft may present as a minimally noticeable midline nasal tip groove to a complete clefting of the osteocartilagenous framework resulting into 2 complete half noses [15]. We achieved favorable results by starting with the nasal soft tissue correction by either V-Y or Z plasties to rejoin the nasal halves as a first step, then the excess glabellar tissues excision and the orbital hypertelorism in a second step. In presence of meningocele we preferred the gathering of skin instead of the Z-plasty to guard against any necrosis of this thinned out skin. Necrosis of this skin may endanger the bone graft placed to close the defect in the base of the skull and the repair of the dural defect after meningocele excision. Excision of the gathered tissues is done later on when the bone graft is healed and the dural defect is sealed. It should be noted that early nasal reconstruction are unlikely to grow and serial augmentations and revisions may be necessary until facial growth ceases [13]. We also address the problem of nasal obstruction which was present in two cases and treated by intraoral osteotomy to widen the atretic pyriform apertures and application of postoperative splitting by nasal tubes with repeated dilatation. We have also anticipated and seen the long nose deformity in cases with meningoceles as described by Ortiz-Monasterio and Fuente Del Campo which results from depression of the cribiform plate and the nasal placode [18]. This could be improved as described by fixing thealar cartilages to the caudal end of the graft and fixing the cephalic end on the glabellar region. The orbital hypertelorism is corrected by the operation of facial bipartition which was described originally by Tessier in two stages with severing of the olfactory nerves. Later Converse and his associates developed a one stage procedure which preserves the cribiform plates and olfactory nerves [19]. The timing of the procedure is very controversial as many surgeons delay the operation till the age of 5 or six years, claiming that before that age, the craniofacial bones are thin and fragile which makes surgery more difficult in addition to the fact that early operation may impair the midfacial growth [20]. However, recently Monastreio and his colleagues have studied the maxillary growth in children after early facial bipartition in 9 patients and found out normal sagittal maxillary growth [21]. We have used rigid fixation by means of miniplates and screws and we have found them safer than using the stainless steel wiring which could penetrate to the dura during introduction. But the minimal amount of hardware is used to prevent restriction of growth of the craniofacial skeleton and migration of these hard ware during growth [22]. Based on these facts, we performed facial bipartision as a single stage with preservation of the olfactory nerves and as early as 2 years. However, in cases of meningocele early correction is mandatory to guard against rupture and secondary deformities. Also, we did not have any problem with the thin craniofacial bones, in contrary these allowed easier osteotomy and greenstick fracturing. Also, we preferred the intraoral approach in infraorbital horizontal maxillary osteotomy better than the subciliary approach as it permits more access to the region and allows correction of the nasal obstruction by widening of the atretic pyriform fossa which is not addressed by some authors.

In management of tense meningocele, we have found out that preoperative shunt decreases the intracranial pressure perioperatively and facilitates excision of the meningocele and prevents postoperative increase in intracranial pressure and CSF leak. We conclude that correction of median facial cleft can start by soft tissue correction of the bifid nose with correction of the cleft lip, then followed by facial bipartition to correct orbital hypertelorism with nasal costochonral graft at 2 or three years of age is very successful as regard the cosmetic and functional results and also in terms of children compliance to the procedure.

REFERENCES


