Journal of Plastic, Reconstructive & Aesthetic Surgery (2019) 72, 676–684

Management of median and paramedian craniofacial clefts☆

Eva Meia Rüegg[a,*], Andrea Bartoli[b], Bénédict Rilliet[b], Paolo Scolozzi[c], Denys Montandon[a], Brigitte Pittet-Cuénod[a]

[a] *Division of Plastic, Reconstructive and Esthetic Surgery, Department of Surgery, Geneva University Hospitals and Faculty of Medicine, 4 Rue Gabrielle Perret-Gentil, 1211 Geneva 14, Switzerland*
[b] *Division of Neurosurgery, Geneva University Hospitals and Faculty of Medicine, Geneva, Switzerland*
[c] *Division of Maxillofacial Surgery, Department of Surgery, Geneva University Hospitals and Faculty of Medicine, Geneva, Switzerland*

Received 25 July 2018; accepted 6 January 2019

KEYWORDS
Craniofacial cleft;
Box osteotomy;
Facial bipartition;
Medial faciotomy;
Nasal reconstruction;
Canthopexy

Summary *Background:* Median and paramedian craniofacial clefts are associated with hypertelorism, anterior encephalocele, positional abnormalities of the maxilla, and nasal deformity. Cleft lip and palate, eyelid coloboma, and widow's peak are frequently present.
Methods: The authors collected data from 30 patients (mean age, 5.8 years; range, 4 months to 18 years) operated between 1986 and 2017 with median or paramedian craniofacial clefts of differing degrees of severity. Malformations of the different anatomic units and their surgical treatment were assessed, as well as complication rates.
Results: All patients presented nasal malformations and either telecanthus (n = 16) or hypertelorism (n = 14). Most patients (n = 23) had anterior encephalocele. All patients underwent nasal corrections, and most of them had medial canthopexy (n = 24). Excision of encephalocele was associated with fronto-orbital remodeling. Medialization of the orbits was performed in 11 patients, mainly by box shift (n = 9). Patients from outside Switzerland (n = 23) were operated at an older age than those in the native patient group. Because of staged reconstruction, 13 patients had more than one operation. Surgical complications included three infections and one expander exposition. One patient had bone resorption of a frontal bone flap. Nasal correction needed more than one procedure in 5 patients, and medial canthopexy had to be repeated in 7 patients. Esthetic results were satisfactory, permitting social integration.

☆ Parts of this article have been presented at the following meetings: 1. 10th European Craniofacial Congress, Gothenburg, Sweden, 24-27 June 2015. 2. Annual Congress of the Société Française de Chirurgie Plastique, Reconstructrice et Esthétique, Paris, France, 19-21 November 2015.3. European Society of Craniofacial Surgery: Biennial Congress, Birmingham, UK, 23-24 November 2016. 4. The Joint American Society of Craniofacial Surgeons-American Society of Pediatric Neurosurgeons Symposium, Hawaii, USA, 23-25 January 2017. 5. European Plastic Surgery Research Council. Hamburg, Germany, 23-25 August 2018.

* Corresponding author.
E-mail address: eva.ruegg@hcuge.ch (E.M. Rüegg).

https://doi.org/10.1016/j.bjps.2019.01.001

Conclusion: Median and paramedian craniofacial clefts need adapted and carefully planned corrections respecting the growth of anatomic units. The quality of the medial canthal and nasal reconstruction is to a large extent responsible for the overall result.

Introduction

Median and paramedian craniofacial clefts are rare disorders. The incidence of craniofacial clefts is estimated at 1.4 to 4.9 cases per 100,000 live births.[1] These malformations can currently be detected with prenatal ultrasound during the second trimester of pregnancy.[2] However, even if fetal surgery is practiced in specialized centers for occipital encephaloceles,[3] craniofacial clefts are currently beyond the scope of prenatal surgery. In countries where prenatal ultrasound is available, most embryos or fetuses presenting severe craniofacial malformations may undergo termination of pregnancies. According to the time of the embryological accident, clinical manifestations are variable. Primary or true clefts occur between the 4th and 8th weeks of gestation because of the failure of fusion between the different facial processes.[4] Secondary or pseudo-clefts occur later. They concern mesenchymal differentiation and correspond to the term of dysplasia.[5] In both situations, the future growth potential is diminished compared to the rest of the face.[6,7]

The malformation may concern brain, bone, and soft tissues either together or isolated. Bone malformations concern the forehead, orbits, ethmoidal cells, the maxilla, and the palate. Regarding the central nervous system, meningoceles or meningoencephaloceles occur most frequently. In encephaloceles, various degrees of bone defect, meningeal defect, cerebrospinal fluid, and brain herniation may occur. According to the anatomical location of the bone defect, encephaloceles can be divided into three types: the cranial vault, frontoethmoidal (or sincipital, anterior), and basal encephaloceles. Occipital encephaloceles, a subtype of cranial vault encephaloceles, appear to be the most common in North America, whereas anterior encephaloceles are more frequent in South East Asia and Central Africa.[8,9] Basal encephaloceles are less common.[10] The pathogenesis of anterior encephaloceles remains unclear, but it seems to be related to the late period of neurulation in the 4th week of gestation. On the basis of previous studies, Hoving proposed in 2000 that the disorder could be explained by a disturbance after the separation of the neural and surface ectoderms following the closure of the neural folds, as these encephaloceles are always covered by the skin.[11] Encephalocele content varies between normal brain parenchyma to fibrous gliotic tissue and cerebrospinal fluid. In most cases, the herniating content is deemed neurologically nonfunctional.[12] Hydrocephalus can also be associated with encephaloceles.[13-15]

Median and paramedian facial clefts are generally associated with hypertelorism, anterior or basal encephalocele, positional abnormalities of the maxilla, and nasal deformity. Soft tissue malformations may be also present, such as cleft lip and palate, eyelid coloboma, and widow's peak. There are different classifications describing craniofacial clefts, but the most common is the one proposed by Tessier.[16,17] In 2008, Fearon described a simplified classification for true facial clefts by regrouping these clefts as median (0-14 Tessier cleft), paramedian (1-13, 2-12 Tessier cleft), orbital (3-11, 4-10, 5-9 Tessier cleft), and lateral (6-8 Tessier cleft), which is easily applicable according to the planned procedures.[7] Van der Meulen et al. described a classification related to embryology,[4] similar to Allam et al., who proposed a classification for median craniofacial dysplasia by dividing three groups of hypo-, normo- (or cleft, dysraphia), and hyperplastic malformations into further subdivisions.[18] The coexistence of numerous classifications shows the difficulty to obtain a consensus. In our study, we chose to use the Tessier and Fearon classifications.

Importantly, surgery needs to address the aforementioned lesions and the reconstructive plan has to deal with the growth and social interaction of the disfigured child. The present report reviews the median and paramedian craniofacial clefts operated in our unit by analyzing the associated lesions and the reconstruction strategy in complex lesions, as well as the surgical revision rate during long-term follow-up.

Patients and methods

All patients operated between 1986 and 2017 in our unit for median and paramedian craniofacial dysplasia with varying degrees of severity were included in the study. Most patients ($n = 23$) came from a foreign country with a limited medical infrastructure and were referred to our hospital by different Swiss humanitarian organizations. Treatment costs for these patients were fully covered by the organizations, which benefited from particularly low preferential rates from our public hospital. A governmental authorization from the country of origin was obtained for each patient transfer and, consent was obtained from the child's parents or guardians. Upon arrival in Switzerland, patients underwent a general health check-up in the pediatric or internal medicine department, followed by a preoperative clinical examination in our unit. Initially, radiographs of lesions were obtained, but from 2003 onward, most operated patients had preoperative computed tomography (CT) scans.

Surgery was individually adapted but according to the following steps: removal of interorbital obstacles; reduction of the interorbital distance by orbital medialization; restoration of the continuity of the cranial base; and soft tissue and nasal reconstruction. Timing of surgical procedures was related to facial growth whenever possible. A multidisciplinary surgical team approach was used and comprised plastic surgeons and neurosurgeons for most patients, as well as maxillofacial surgeons for corrections of

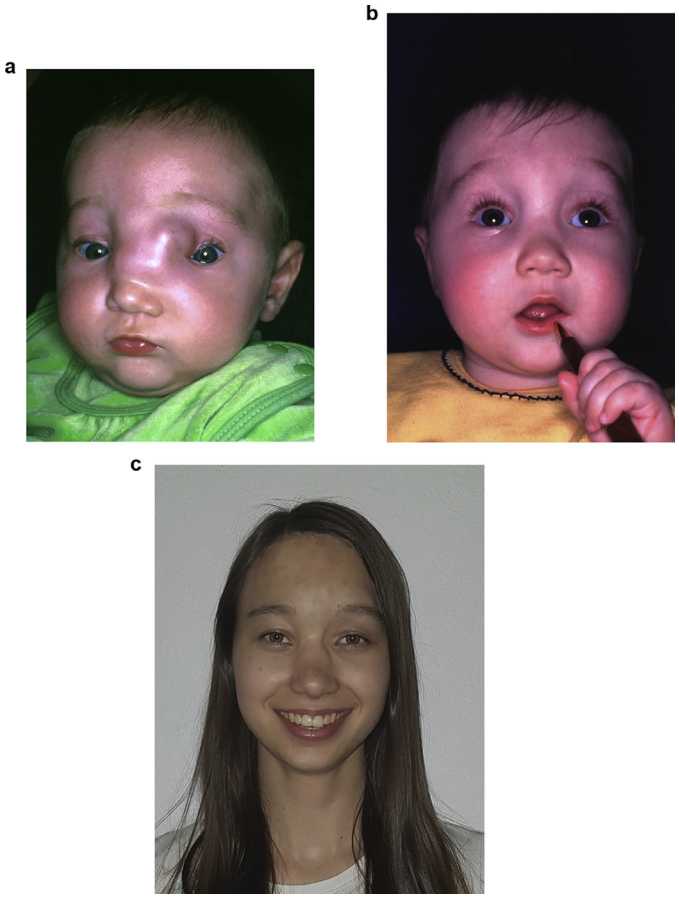

Figure 1 **a:** A 5-month-old girl from Switzerland with a paramedian left craniofacial cleft along with frontonasal encephalocele and left telecanthus. **b:** The same patient after resection of the encephalocele and frontal bone reconstruction by split calvarial graft of the frontal bone flap approach and left medial canthopexy. **c:** At 18-year follow-up with a stable result.

hypertelorism. Patients undergoing craniotomy were monitored in the intensive care unit during the first 24 h postoperatively. Intravenous antibiotics were continued for at least 2 days postoperatively.

Follow-up included clinical controls and photographs and was conducted either by our surgical team during surgical missions to the patient's country or by the local teams of the collaborating humanitarian organizations. Radiological follow-up was carried out when patients were retransferred to our hospital for further surgical procedures. Swiss patients had full medical coverage through the national insurance for disabled persons with regular clinical controls during 1 year postsurgery and a long-term follow-up until 20 years of age. Ethical approval to perform the study was obtained from the review board of the Medical Ethical Committee of the Canton of Geneva (project no. 2017-00528).

Results

Between 1986 and 2017, 30 patients (mean age, 5.8 years; range, 4 months to 18 years) were operated for median and paramedian craniofacial clefts with differing degrees of severity. Most patients (n = 19) came from Africa (Benin, Burkina Faso, Guinea, Madagascar, Morocco, Niger, Senegal, Tanzania, and Togo); 4 patients were from Lebanon, India, Cambodia, and Macedonia; and 7 patients were from Switzerland. According to Tessier's classification, there were 17 craniofacial midline clefts (0–14); of them, 2 were frontal only (14). Paramedian clefts were present in 13 patients (6 patients with cleft 13; 4 patients with cleft 1–13; and 1 patient with cleft 1). Two patients with medial orbital clefts (1 with cleft 3–11; 1 with cleft 3 and proboscis lateralis) were identified. The different malformations are described individually below.

Frontonasal encephaloceles

Our series included 23 patients with one or more sincipital encephaloceles. No dural sinus extension was found in any of the encephaloceles, whereas 3 patients had aberrant ventricular extension in the cele. Two patients had associated unilateral coronal synostosis. There were 13 midline clefts and 10 patients with paramedian cleft (4 patients with Tessier 1–13; 6 with Tessier 13). Patients underwent a coronal approach followed by excision of the encephalocele and then bony reconstruction of the calvarial defect with split calvarial bone graft and median transosseous canthopexy through a frontal bone flap approach. Primary dural closure was strengthened by pericranium flap and fibrin glue in all

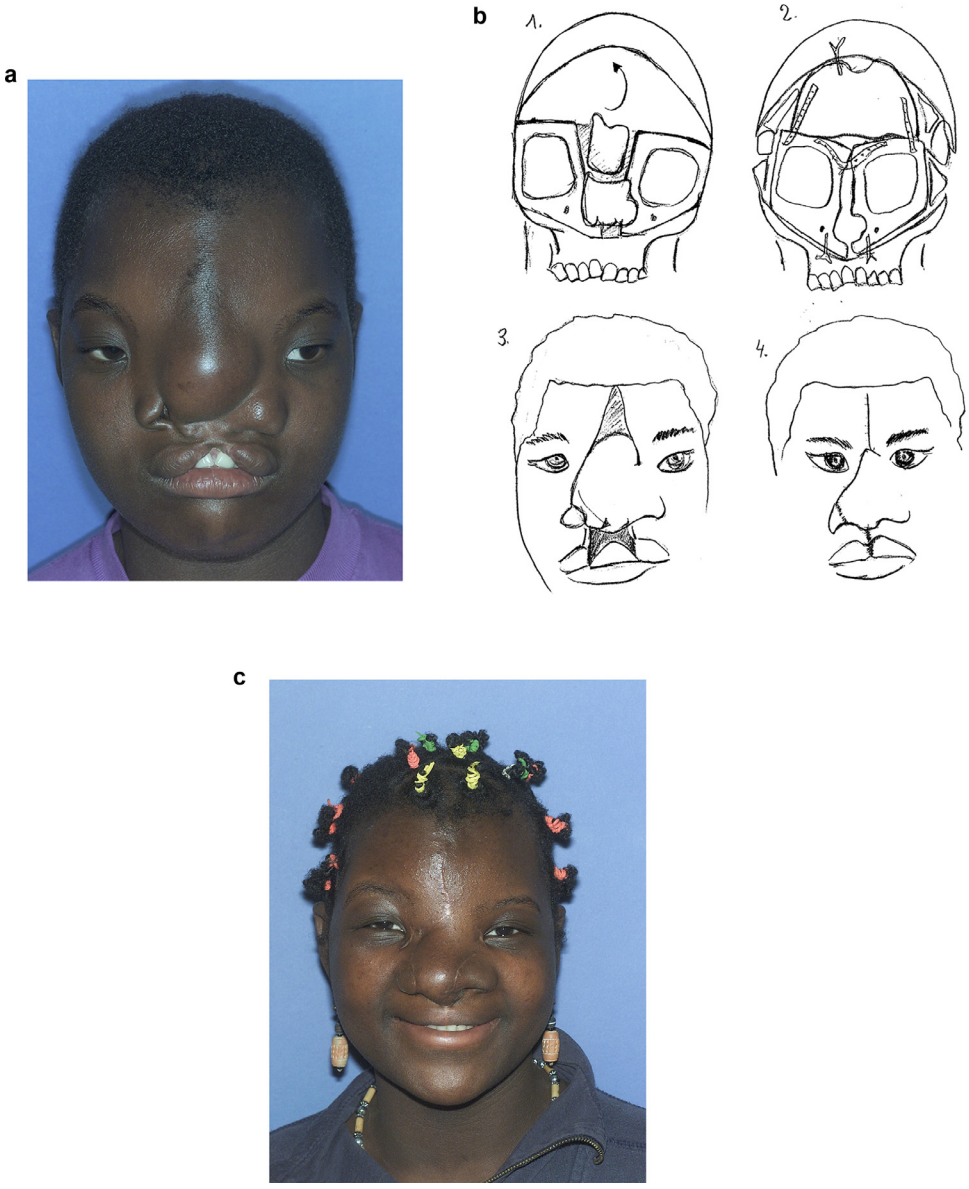

Figure 2 **a:** An 18-year-old girl from Burkina Faso with a craniofacial midline cleft with frontonasal encephalocele, severe hypertelorism (60 mm interorbital distance), and incomplete median cleft nose and lip. **b:** The surgical plan of the procedure with box shift medialization of the orbits by a frontal bone flap approach and use of the midline skin and nostril remnants for nasal reconstruction with an osseocartilaginous rib graft in a one-stage procedure. **c:** The result before returning to her country.

encephalocele repairs and orbital medializations to correct hypertelorism.

In clefts with large bone defects, frontal bone reconstruction was performed, with a parietal bone flap used as a split calvarial bone graft. Midline skin excision was necessary in 10 of 14 patients with midline cleft and encephalocele, mostly an H-excision over the nasal root (6 patients). All other patients required more complex skin excisions and soft tissue remodeling, generally used for nasal reconstruction. One patient needed a temporary lumbar drain for cerebrospinal fluid leak after anterior skull base reconstruction and medialization, but none needed a permanent cerebrospinal fluid diversion, including those who had abnormal ventricular extension in the encephalocele.

Telecanthus

Sixteen patients presented telecanthus in combination with encephalocele. The surgical procedure consisted of an internal orbitotomy and medialization of the internal orbital walls on both sides (n = 9), completed by transnasal internal canthopexy (n = 16). This procedure was generally performed at the same time as the resection of the encephalocele (Figure 1).

Hypertelorism

Orbital hypertelorism was present in 14 of 30 patients. Most patients had craniofacial midline clefts (n = 9); 5

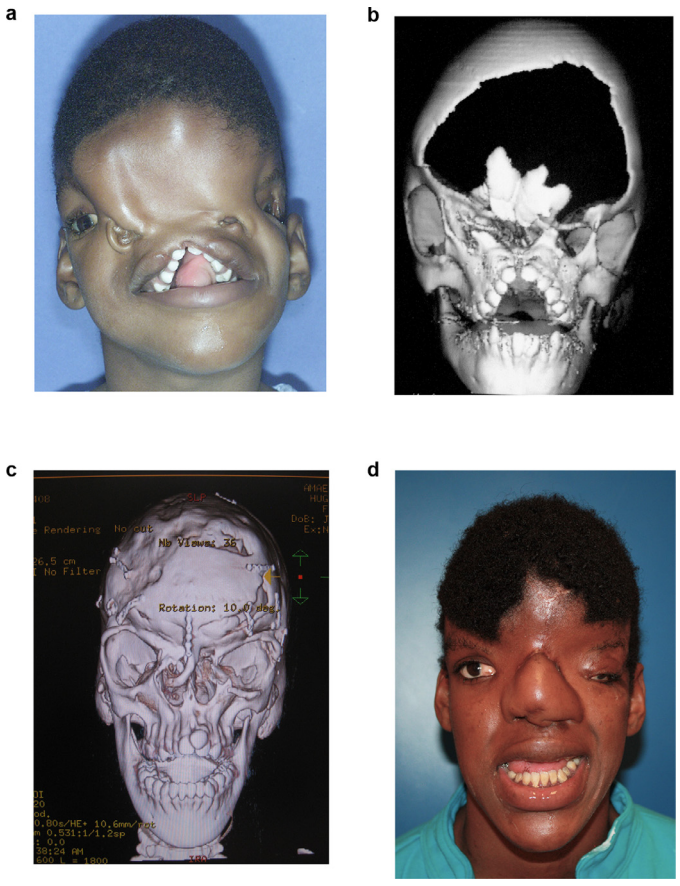

Figure 3 (Permission to reproduce figures 3a and 3b from reference 21. a: A 6-year-old girl from Nigeria with an extremely large median craniofacial cleft (Tessier 0-14) presenting several frontal encephaloceles and the absence of frontal bone, as well as a large widow's peak covering virtually the whole forehead. She had a severe hypertelorism of approximately 120 mm interorbital distance; a large left upper eyelid coloboma, which had led to corneal opacity; and a large centrofacial cleft with the absence of the nose (two nostril remnants being present) and a median cleft lip. Her palate was ogival and intact. She presented with left amblyopia. **b:** CT scan of the same patient showing the large craniofacial cleft of the midline, absence of the frontal bone, and midfacial cranial tilting, as well as extreme hypertelorism with lateral orientation of the orbits. Her palate was extremely arcuate. **c:** CT scan at 5-year follow-up showing good general bony consolidation, although some small bony defects persisted. The contour of the facial skeleton is greatly normalized with a persistent stable moderate hypertelorism of approximately 4 cm. **d:** At 11-year follow-up. Further, she had another nasal correction for deviation of the rib graft and left medial canthopexy 5 years after the initial surgeries. Note the good stability of the facial height, although bony correction was necessary during the period of facial growth.

patients had paramedian craniofacial cleft. In addition, 3 patients had anterior plagiocephaly due to unilateral craniosynostosis. One patient had supplemental anterior encephalocele.

Patients were mostly treated by box osteotomy (n = 9) (Figure 2).[19] Facial bipartition was used for 2 patients to correct a maxillary cleft (Figures 3 and 4).[20,21] The remaining 3 patients were either too young for surgery (1 patient) or not interested in surgical correction of the hypertelorism (2 patients; 1 requested a rhinoplasty to correct the bifid nose). All patients in this series presented severe hypertelorism according to the Tessier classification,[17] with a mean interorbital distance of 59 mm (range, 40-120 mm) before surgery. Mean reduction of interorbital distance was 33 mm (range, 18-60 mm).

Widow's peak

Three patients with wide craniofacial clefts presented an important widow's peak. In the patient with the most extreme hypertelorism of 12 cm interorbital distance, resection was not possible, as hair covered the entire forehead.[21] In 2 patients, the widow's peak was resected after frontal skin expansion, as bone stability allowed the use of expanders (Figure 5).

Palpebral coloboma

Six patients presented palpebral colobomas. The upper eyelid was concerned in 4 patients with median (1 patient, Tessier 0-14) and paramedian (3 patients, Tessier 1-13)

Figure 4 **a:** A 16-month-old boy from Guinea with paramedian craniofacial cleft (Tessier 1-13), presenting a frontal bone cleft measuring approximately 35 × 40 mm and a small widow's peak. He had a severe hypertelorism of 55 mm interorbital distance, a small left upper eyelid coloboma, and a large left nasal cleft including premaxilla. His palate was ogival with a bony cleft. **b:** The same patient after closure of the right upper eyelid coloboma, frontal bone reconstruction, and facial bipartition, permitting 30 mm of reduction of interorbital distance and closure of the premaxillary cleft, excision of the widow's peak, and soft tissue nasal reconstruction before returning to Guinea. **c:** At 15-year follow-up without having had other procedures in the meantime. Note a stable result of hypertelorism correction and soft tissue reconstruction.

clefts. The lower eyelid was affected in 3 cases with medial orbital clefts (1 case, Tessier 3-11 cleft; 1 case, Tessier 3 cleft, and proboscis lateralis). Direct closure was possible in most patients presenting a small size cleft and accurate vision. Two patients with large upper eyelid coloboma and loss of vision because of corneal exposition and coexisting severe hypertelorism were closed with local temporal flaps.

Nasal reconstruction

Nasal malformations were ubiquitous in this series. They were usually corrected during the excision of frontonasal encephaloceles or hypertelorism correction as described above with calvarial bone grafting to improve nasal support and projection and, if necessary, central skin resection. In patients with median and paramedian craniofacial clefts including complete nasal cleft, local and locoregional flaps using nasal remnants (10 patients) were used. Nasal lining was reconstructed using mucosal or hinge flaps of local tissue. The osseocartilaginous framework was reconstructed with either calvarial bone grafts (n = 15) or osseocartilaginous rib grafts (n = 4) used as cantilever grafts.

Staged reconstructions and timing

Most patients had more than one operation because of staged reconstruction (n = 13; range, 2-7). Patients from outside Switzerland (23 patients) were mostly operated at an older age than the age according to the traditional surgical algorithm,[22,23] i.e., encephalocele corrections and/or cranioplasty were performed at a mean age of 6.4 years (range, 6 months to 18 years). Five of these (mainly African) patients had been previously operated in their own country. The 2 Swiss patients were operated for encephalocele correction at 5 months of age and at 11 years. The second patient immigrated to Switzerland just before the procedure.

Box osteotomies were performed at a mean age of 9.6 years (range, 4-18 years) in non-native patients compared to 1 Swiss patient operated at 5 years of age. Facial bipartition was performed in 2 African patients at 2 years of age and 6 years of age. Palpebral coloboma was closed in a Swiss

Figure 5 **a:** A 17-year-old girl from Tanzania with right paramedian craniofacial cleft (Tessier 1–13), presenting frontal meningoencephalocele and a large widow's peak. She had a severe hypertelorism, a small right upper eyelid coloboma, and a large right nasal cleft. Her palate was intact and occlusion correct. She presented with right amblyopia. **b:** The surgical strategy included the use of two skin expanders to excise her large widow's peak. This image shows the situation just before excision of the widow's peak and orbital medialization by box osteotomies. **c:** The same patient after closure of the right upper eyelid coloboma, excision of her meningoencephalocele, frontal bone reconstruction and orbital box shift medialization (30 mm reduction of interorbital distance), excision of the widow's peak, and soft tissue nasal reconstruction before returning to Tanzania.

patient at 9 months of age, whereas the mean age at closure in foreign patients was 6.8 years (16 months to 17 years). Nasal remodeling was performed at a mean age of 5.5 years (range, 5 months to 18 years) for a first procedure in this mixed cohort. Refinements and rhinoplasty were performed after 16 years of age.

Outcome and Follow-up

The surgical procedure was well supported by all patients, and there were no occurrence of anesthesia-related or intracranial complications. Complications included 3 frontal infections requiring surgical debridement. One occurred at 2 years postoperatively after a banal trauma, requiring another transfer because of posttraumatic osteomyelitis. There were 3 retransfers: 1 for further nasal correction and 1 for correction of hypertelorism. The latter had bone resorption of a frontal bone flap requiring frontal revision with a titanium plate fixation. Concerning reoperations during the same transfer, 1 patient had expander exposition during skin expansion. In 7 patients, medial canthopexy had to be repeated and 5 patients needed further refinement for nasal reconstruction. One patient had transient lumbar decompression for nasal liquorrhea. Esthetic results were subjectively satisfactory for patients, permitting social integration. Long-term follow-up ranged from 1 to 30 years (mean, 16 years) and was performed mainly by local teams of humanitarian organizations for foreign patients and during our surgical missions to the country (Figures 3 and 4). Swiss patients had clinical follow-up at our institution (Figure 1).

Discussion

Different teams have published their long-term observations regarding patient groups with median or paramedian dysplasia and clefts, mostly analyzing their results regarding growth. In two long-term studies of median and paramedian clefts, the Rotterdam team described a worsening of their results with time after initial favorable results, attributed to intrinsic growth restriction, particularly in the central midface.[24,25] They proposed a timing guideline for the different procedures these patients typically need and

insisted on final midfacial and nasal corrections at the end of facial growth. Similar to Marchac et al., they observed stable growth in a large series of hypertelorism corrections operated at a young age.[23] We confirm their observations regarding the stability of hypertelorism correction and the importance of final nasal reconstruction at facial maturity. In addition, Ortiz-Monasterio highlighted the importance of using the esthetic subunit principle in the reconstructive strategy for these patients,[26] which we also used in our surgical plan.

Concerning the esthetic quality of the postoperative result, we observed that the anatomic position of the eyelids and a harmonious nasal reconstruction were responsible for the overall result to a large extent. Consequently, nasal refinement and canthopexies were the most frequent reoperations. The particularity of our series is the high number of foreign patients with severe malformations and their long-term follow-up because of an excellent long-term collaboration with humanitarian organizations.

Particularities of humanitarian transfers

More than two-thirds of our study cohort represents children from developing countries treated as humanitarian transfers. Patients were often seen at an older age than the normal start of medical treatment for craniofacial malformations in developed countries. Some presented with irreversible sequelae such as loss of vision because of eyelid coloboma or amblyopia in the case of hypertelorism. Palpebral coloboma may expose the patient rapidly to a loss of vision in the concerned eye because of an opacification of the cornea in large defects (Figure 3a). Therefore, the closure of coloboma should be performed as soon as possible. For small-to-medium coloboma-presenting defects of one-third up to one-half of the eyelid, direct closure is generally sufficient, sometimes associated with external canthol-ysis. In large colobomas, total palpebral reconstruction by heteropalpebral tarsoconjunctival flaps associated with skin grafts or local flaps from the temporal area may be necessary.[27-29] In our series, 2 patients presented with an already existing loss of vision because of corneal exposure. In the case of severe hypertelorism, we performed closure of the large coloboma several weeks before medialization of the orbits as the first surgical step to avoid excessive tension and functional problems of the upper eyelid. We had 4 cases of severe untreated hypertelorism arriving in Geneva with amblyopia. These patients had not been seen by an ophthalmologist in their country and could not benefit from occlusion therapy, a well-known and simple therapy to prevent the loss of vision of the nondominant eye.

Considering the older age for frontonasal encephalocele and hypertelorism correction in the transferred patient group, some patients had already achieved skeletal maturity of the face. The disadvantage is the impossibility of a spontaneous correction of collateral distortions, i.e., of the nose after correction of the supraorbital bar in anterior plagiocephaly due to unilateral coronal synostosis.[22] Thus, the surgeon has to deal with a definitive situation and correct the malformation sequelae of growth ideally at the same time as the basic standard correction. On the other hand, the advantage of older age is the better stability of surgical results as shown by two long-term studies on the results of hypertelorism.[23,30]

The nose - the center of the face

It is evident that median and paramedian craniofacial clefts always concern the nose. As in patients with cleft lip, closure and replacement of structures to the right place should be performed as soon as possible to permit orthotopic growth and to avoid further aggravation of distortion because of growth. Dissection should be limited to avoid growth disturbances, but the different anatomical structures and subunits, especially of the nose, need to be placed at their anatomical position. In our series, the mean age at initial surgery was 5.5 years. The high age for an initial surgery is explained by the adult age of some patients. In this cohort of mostly transferred patients, it had the advantage to limit the number of operations.

In hypertelorism, the base of the nose is always enlarged and the tip often bifid.[17] The enlargement is mostly treated by surgical reduction of the intercanthal distance.[31] When nasal bones are hypoplastic, bone grafting of the dorsum during hypertelorism correction may be necessary, as limited nasal growth is to be expected if untreated, especially if the malformation involves the septum.[23] Secondary corrections or rhinoplasty should be completed at the end of facial growth[25] and upon the request of the adult patient. Nostril retractions may be corrected by Z-, VY-plasties, or cartilage grafts.

Conclusions

Median and paramedian clefts present with highly variable degrees of severity and need adapted and carefully planned corrections. Surgery aims to re-establish facial integrity according to esthetic subunits and starts with skeletal malformations. The quality of nasal reconstructions and canthoplasties are of paramount importance and responsible for the overall result to a large extent.

Conflict of interest

The authors declare no financial or personal conflict of interest. None of the authors has commercial associations.

Acknowledgments

We extend our warmest thanks to the humanitarian organizations “Terre des Hommes,” “Sentinelles,” “Hymne aux Enfants,” and “Second chance’’ for their excellent care of our young patients and their extraordinary collaboration for so many years. We thank Geneva University Hospitals for supporting our humanitarian program by offering preferential low hospital rates for many years and providing free-of-charge medical services from 2013 onward. Finally, we thank Rosemary Sudan for editorial assistance.

References

1. Ozaki W, Kawamoto H, editors. *Craniofacial Clefting*. Philadelphia, PA: Saunders; 2002.
2. Maarse W, Pistorius LR, Van Eeten WK, et al. Prenatal ultrasound screening for orofacial clefts. *Ultrasound Obstet Gynecol* 2011;**38**:434–9.
3. Cavalheiro S, da Costa MDS, Mendonca JN, et al. Antenatal management of fetal neurosurgical diseases. *Childs Nerv Syst* 2017;**33**:1125–41.
4. van der Meulen JC, Mazzola R, Vermey-Keers C, Stricker M, Raphael B. A morphogenetic classification of craniofacial malformations. *Plast Reconstr Surg* 1983;**71**:560–72.
5. Mazzola RF, Mazzola IC. Facial clefts and facial dysplasia: revisiting the classification. *J Craniofac Surg* 2014;**25**:26–34.
6. van der Meulen JC, Vaandrager JM. Facial clefts. *World J Surg* 1989;**13**:373–83.
7. Fearon JA. Rare craniofacial clefts: a surgical classification. *J Craniofac Surg* 2008;**19**:110–12.
8. Rowland CA, Correa A, Cragan JD, Alverson CJ. Are encephaloceles neural tube defects? *Pediatrics* 2006;**118**:916–23.
9. Naidich TP, Altman NR, Braffman BH, McLone DG, Zimmerman RA. Cephaloceles and related malformations. *Am J Neuroradiol* 1992;**13**:655–90.
10. Mulcahy MM, McMenomey SO, Talbot JM, Delashaw JB Jr. Congenital encephalocele of the medial skull base. *Laryngoscope* 1997;**107**:910–14.
11. Hoving EW. Nasal encephaloceles. *Childs Nerv Syst* 2000;**16**:702–6.
12. Simpson DA, David DJ, White J. Cephaloceles: treatment, outcome, and antenatal diagnosis. *Neurosurgery* 1984;**15**:14–21.
13. Mealey J Jr, Dzenitis AJ, Hockey AA. The prognosis of encephaloceles. *J Neurosurg* 1970;**32**:209–18.
14. Shokunbi T, Adeloye A, Olumide A. Occipital encephalocoeles in 57 Nigerian children: a retrospective analysis. *Childs Nerv Syst* 1990;**6**:99–102.
15. Chapman PH, Swearingen B, Caviness VS. Subtorcular occipital encephaloceles. Anatomical considerations relevant to operative management. *J Neurosurg* 1989;**71**:375–81.
16. Kawamoto HJ. The kaleidoscopic world of rare craniofacial clefts: order out of chaos (Tessier classification). *Clin Plast Surg* 1976;**3**:529–72.
17. Tessier P. Anatomical classification facial, cranio-facial and latero-facial clefts. *J Maxillofac Surg* 1976;**4**:69–92.
18. Allam KA, Wan DC, Kawamoto HK, Bradley JP, Sedano HO, Saied S. The spectrum of median craniofacial dysplasia. *Plast Reconstr Surg* 2011;**127**:812–21.
19. Tessier P, Guiot G, Rougerie J, Delbet JP, Pastoriza J. [Craniostenosis. Craniofacial osteotomies. Hypertelorism]. *Ann Chir Plast* 1967;**12**:103–18.
20. van der Meulen JC. Medial faciotomy. *Br J Plast Surg* 1979;**32**:339–42.
21. Pittet B, Jaquinet A, Rilliet B, Montandon D. Simultaneous correction of major hypertelorism, frontal bone defect, nasal aplasia, and cleft of the upper lip (Tessier 0-14). *Plast Reconstr Surg* 2004;**113**:299–303.
22. Marchac D, Arnaud E. Midface surgery from Tessier to distraction. *Childs Nerv Syst* 1999;**15**:681–94.
23. Marchac D, Sati S, Renier D, Deschamps-Braly J, Marchac A. Hypertelorism correction: what happens with growth? Evaluation of a series of 95 surgical cases. *Plast Reconstr Surg* 2012;**129**:713–27.
24. van den Elzen ME, Versnel SL, Wolvius EB, et al. Long-term results after 40 years experience with treatment of rare facial clefts: Part 2-Symmetrical median clefts. *J Plast Reconstr Aesthet Surg* 2011;**64**:1344–52.
25. Versnel SL, van den Elzen ME, Wolvius EB, et al. Long-term results after 40 years experience with treatment of rare facial clefts: Part 1-Oblique and paramedian clefts. *J Plast Reconstr Aesthet Surg* 2011;**64**:1334–43.
26. Monasterio FO, Taylor JA. Major craniofacial clefts: case series and treatment philosophy. *Plast Reconstr Surg* 2008;**122**:534–43.
27. Mustarde JC. The use of flaps in the orbital region. *Plast Reconstr Surg* 1970;**45**:146–50.
28. Mustarde JC. Congenital deformities in the orbital region. *Proc Royal Soc Med* 1971;**64**:1121–34.
29. Gumener R, Montandon D. [The tarsoconjunctival flap in palpebral reconstruction]. *Ann Chir Plast Esthet* 1983;**28**:115–20.
30. Raposo-Amaral CE, Raposo-Amaral CM, Raposo-Amaral CA, Chahal H, Bradley JP, Jarrahy R. Age at surgery significantly impacts the amount of orbital relapse following hypertelorbitism correction: a 30-year longitudinal study. *Plast Reconstr Surg* 2011;**127**:1620–30.
31. Arnaud E. [Some features of the nose in craniofacial malformations]. *Ann Chir Plast Esthet* 2014;**59**:585–91.