The role of plastic surgeon in complex cephalic malformations

Our experience

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AIM: *Our aim is focused on the field of action of plastic surgery in the cephalic malformation and on description of the surgical indications and techniques for their correction.*
MATERIAL OF STUDY: *We looked at 27 patients from 2006 to 2012 with cephalic district deformities. All patients underwent surgical and rehabilitative treatments.*
Results: *After a median follow-up from one to five years, in most cases we reached both morphological and functional reconstruction, alone or in a surgical team together with other surgical disciplines.*
DISCUSSION: *The correction of craniofacial malformations makes use of a surgical discipline particularly demanding, which must associate a basic surgical training with a learning techniques specific to the area and a knowledge of craniofacial malformation.*
CONCLUSIONS: *The surgical treatment of craniofacial malformations can be seen only through a joint neurosurgical and plastic-maxillo-facial surgery, guided by knowledge of the malformation, under the close supervision of anesthetists and pediatricians. This surgery is not limited to remodel the morphology but has its bases on the recovery of the functions, maintained by rehabilitation treatment.*

KEY WORDS: Complex cephalic deformities, Cephalic district malformations

Introduction

A malformation is an alteration of the body morphology due to an error occurred during the intrauterine development.
The complex malformations of the cephalic district include a variety of developmental abnormalities that, in addition to having considerable difficulty to be classified and setted, comprise forms that are sometimes strictly pertinent of specialist such as malformations of the eyeball, the middle or internal ear, etc..
These malformations can affect both soft tissues, such as the eyelids, and bones. Plastic surgery aims to restore injured or abnormal forms and to reestablish impaired functions being a multidisciplinary surgery, working both on surface and depth tissue. The eyelid district is particularly dear to the plastic surgery, which, moreover, has always been interested also on bone tissue and, therefore, on orbital of the district. In addition, treatment and follow-up of the surface abnormality, such as vascular malformations, neurofibromatosis and congenital nevi, is attributable to the plastic surgeon, even more in the cephalic region.

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Surgery of cephalic district complex malformations should not be limited to the restoration of morphology but must integrate the functions recovery that are maintained by rehabilitative treatments. Therefore remains a critical point the interdisciplinary and the close collaboration with other specialties. The pediatrician often is the first to deal with these anomalies and to get a diagnosis, but during the follow-up these patients are followed by a multidisciplinary team including neurosurgeons, otolaryngologists and ophthalmologists, besides to the essential geneticists and psychologists.

These patients are followed from the first visit to complete development and, in some cases, are treated very early in the case emerge breathing problems (maneuvers on tongue or bone distraction, possibly preceded or associated with tracheotomy).

The orbital morphology is altered by craniofacial malformations due to the close relationship it has with the facial and cranial structures during the pre-and post-natal care.

Surgery of complex malformations of the district cephalic requires prior knowledge of the characteristics of the malformations and of how they would develop.

Therefore, is necessary to keep in mind that complex malformations of the cephalic district might involve soft tissues with subsequent bone involvement (vascular malformations, neurofibromatosis, congenital nevi, craniocephalic malformations, cerebrofacial deformities), or bone structures with subsequent soft tissues involvement (craniofacial malformations, synostosis, dysostosis, cleft) [1,2].

Vascular malformations are errors of development consisting of dysplastic vessels lined by a quiescent endothelium. They are usually evident at birth, do not regress and often expand. There are also complex-combined vascular malformations associated with excessive soft tissue and bone growth. They are classified according to the type of vessels and divided into "slow flow" and "fast-flow"; are classified as slow-flowing the capillary malformations (CM), including telangiectasias, lymphatic (LM) and venous (VM) malformations, while as fast-flow the arterial or arteriovenous malformations (AVM).

Lymphatic malformations contain lymph, the others blood.

Congenital nevi are benign tumors represented by melanocytic or nevocytic nevi. The congenital nevus or giant pigmented nevus is histologically an intradermal nevus with diffuse foci of proliferative activity around the adnexa and the vascular and nervous structures, even beyond the limits of the dermis in the context of the subcutaneous fat. Clinically, it is a nevus of considerable size, being able to affect large areas of the face, trunk or limbs, greatly pigmented, often hairy and seborrhoeic, with irregular surface and definite but fringed margins, soft in consistence, totally asymptomatic but easily complicated by infectious and maceration. It is often associated with other malformations such posterior dysraphia and angiomas. The widespread present of junctional outbreaks of activity explains the major impact, valued at more than 10%, in the context of developing of a malignant melanoma from the nevus.

Cerebrofacial malformations are due to a deficit of the sensory placodes to induce the formation and development of the corresponding facial district. A lesion of the olfactory placode determines the arhinencephaly, aplasia/hypoplasia of the eye determines the anorbitism and microrbitism.

The various craniofacial deformities resulting from an abnormal development of the first and second branchial arch that, around the 20th months of gestation, give origin to face bones and ears. These deformities are schematically divided into cranial and facial and are due to three pathogenetic mechanisms: the early welding of the joint areas between the bone segments which determines the synostosis; the intrinsic tissue alteration or dysplasia with subsequent dysostosis and the lack of facial buttons welding that leads to facial clefts. These defects include cleft lip and cleft palate, Treacher Collins (mandibulo-facial dysostosis) [3], Goldenhar (oculo-auriculo-vertebral dysplasia), Pierre Robin and Waardenburg syndromes, hypertelorism and deformity of the outer ear and middle ear. The majority of children with craniofacial anomalies have normal intelligence and development [4].

Gliomas are non-encapsulate glial cells accumulation located outside the CNS. There are several etiological hypotheses about their origin: seizure of glial tissue of the olfactory bulb (trapped during the cribriform plate fusion), ectopic neural cells, trapped encephalocele, inappropriate closure of the anterior neuropore (fonticulus frontalis). Intranasal forms can be located in the middle turbinate or the higher structures (miming a nasal polyp). 15% of gliomas connect with the dura throughout the foramen caecum or the fonticulus. The combined forms intra-extranasali have a typical shape of a handlebar. From the clinical point of view gliomas may manifest with: unilateral nasal obstruction, unilateral nasal mass, epistaxis, cerebrospinal rhinorrhea, canthi dystopia (extranasal forms), hypertelorism (extranasal forms). For the diagnosis CT and MRI may be useful and biopsies should be avoided. The treatment is crucial as it is useful in order to prevent meningitis and is based on surgical removal that can be carried out using an external approach for the extranasal masses, lateral rhinotomy for the intranasal masses or neurosurgical approach for the intrathecal masses.

The encephalocele is a neural tube defect characterized by sac-like protrusions of the brain and the membranes that cover it through openings in the skull and is similar to gliomas for its etiology. 20% of encephalocele arise in the skull and 15% are nasal forms.

Encephaloceles are generally classified as meningoceles (if they contain meninges), as encephalomeningoceli, containing brain tissue and meninges, encephalomeningocystocele if they are in communication with a ventricle.

From clinical point of view encephaloceles can manifest with history of rhinorrhea or recurrent meningitis, broad nose or with hypertelorism with dystopia of the canthi, with expansion during the Valsalva maneuver, with Furstenberg positive sign (stretching to compression of both internal jugular veins) or even discovered by transillumination. As well as for the diagnosis of a glioma, CT and MRI are useful while biopsies are contraindicated. Treatment consists of surgical excision and the reconstruction of the bone defect and often a craniotomy is necessary [5].

The cleft are a kind of malformations resulting from the failure of the frontal processes to develop properly or to join with the other nose processes. Among them the most common are cleft nose, the oro-orbital cleft and cleft palate [6].

The Proboscis Lateralis or tubular nose is a extremely rare congenital malformation and is caused by the failure of the frontal processes to develop properly or to join with other nasal processes (fusion of the maxillary process with the contralateral nasal process). Clinically it manifests with the absence of the nasal cavity and paranasal sinuses on one side (blind nasolacrimal duct) and may be associated with other congenital anomalies (in particular CNS ones). Excision of the tubular deformity and nasolacrimal duct reconstruction, started during the adolescence and repeated over time, are the treatments of choice for this malformation.

Methods and Materials

From June 2006 to January 2012 we treated 27 patients with complex cephalic district malformations. Patients had various tipe of malformations: 15 cases of vascular malformations, including 10 cases of arteriovenous malformation, 4 cases of capillary malformations and 1 case of Sturge-Weber syndrome, 5 cases of neurofibromatosis including 4 cases of neurofibromatosis type 1 (Von Recklinghausen's disease) and 1 case of neurofibroma oculopalpebrale (Fig. 1); 4 congenital nevi, of which 1 mole divided (Fig. 2), 3 cases of malformations cranioencefaliche including 1 case of arinencefalia, 1 case of trigonocephaly and 1 case of plagiocephaly.

The 10 cases of arteriovenous malformations were treated with arterial embolization with direct puncture of the nest and local arterial and venous compression, performed to obtain a temporary occlusion of the nest in preparation for surgical resection. Subsequently the AVM nest and the overlying skin were excised widely to prevent recurrence. The wound coverage was performed during the same surgical time and in 1 case required a free flap harvesting [7].

The 4 cases of capillary malformations were treated with pulsed dye laser while for the Sturge Weber Syndrome a surgical treatment was required to solve the soft tissue and skeletal hypertrophy and subsequent coverage with flaps [7].

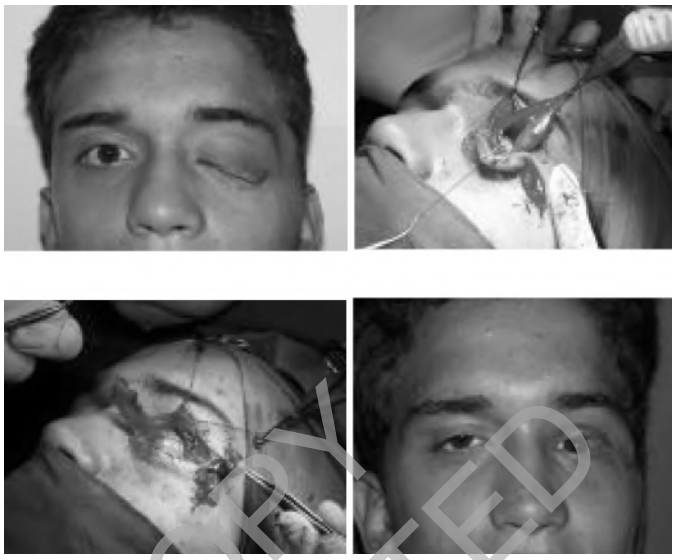

Fig. 1: Oculopalpebral neurofibroma.

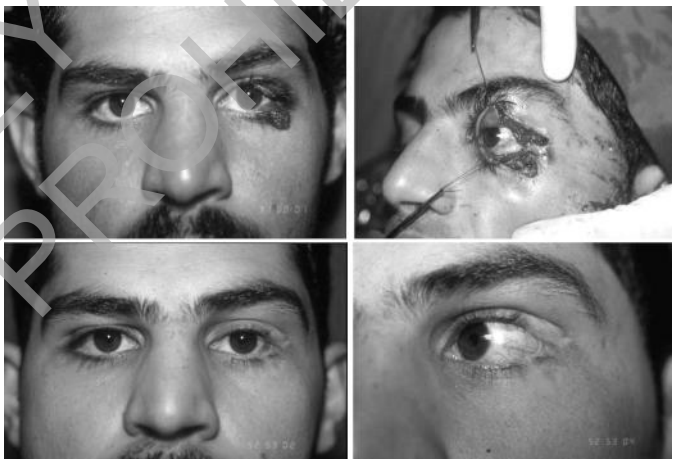

Fig. 2.

Four cases of neurofibromatosis were surgically treated for the neurofibromas removing where it was possible, while the oculopalpebral neurofibroma [8-10] was removed by CO_2 laser in order to reduce bleeding and obtain a better hemostasis; then we proceeded with reconstruction of the skin integrity.

The 2 congenital nevi were completely excised and the reconstruction was performed using local flaps. In one case it was necessary to use the partial serial excisions in three steps, utilizing the elasticity of the surrounding skin until the complete excision of the lesion. The divided nevus was completely excised and then repaired with local flaps [9].

Regarding cranioencephalic malformations, in the case of arhinencephaly we created new nasal cavity and external nose through local flaps and cartilaginous grafts; in the

case of trigonocephaly we performed surgical correction in the first year of life by opening the nasal-orbito-frontal region, then we have expanded the glabella and lowered the orbits and then lodged a triangular bone segment in the nose. The plagiocephaly was surgically corrected (only the pathological side) creating a fronto-orbital- repositioning with a 3D shifting whose rotation center was represented by the nose [11-13].

Results

After a mean follow-up ranging from one to five years, in all cases we have achieved the aims of a morphological and functional reconstruction.
Surgical exploration of such regions of the skull and face requires accurately selected accesses in order to limit the scarring, without however limiting the security afforded by a properly exposed operative field. The approach respects the structures and tissues of coverage, remaining extradural to protect the sub-periosteal brain during the exposition and the mobilization of the bone segments and extra-mucosal in the nasal fossa to prevent peri and post-operative care contamination. The conservation of sensory pathways, in particular those olfactory, must be considered a rule. The malformation aetiology places surgeon in front of specific problems: a therapeutic protocol is establishes, thus, after the malformation has been identify, estimated the severity of tissue alterations and evaluated the ability of growth of the affected territories. A as well as the remodelling of the morphology, the recovery of function and rehabilitation treatment are two key points. Finally, the presence of the teeth and occlusal problems frequently impose an orthodontic complement.

Discussion

Complex malformations of cephalic region mostly involve children or infants and therefore require a paediatric environment. The pediatrician is who, in most cases, recognise malformations, identify them as isolated or otherwise integrated into a more complex malformation syndrome and he has the responsibility to inform the parents about the genetic aetiology.
Craniofacial surgery combines neurosurgery expertise with facial surgeon one in a single way of thinking and acting, without which such an act could not be conceived. It 'is therefore clear that it is a surgery team.
The otolaryngologist, the ophthalmologist and the orthodontist bring their respective expertise in the preoperative assessment and controls in the short and long term. The anaesthetist-resuscitator, finally, must receive special training to deal with all the risks of this surgery that can reach positive results only if carried out with mutual trust between the various actors.

Conclusion

The correction of craniofacial malformations require a particularly exigent surgical discipline, which must associate a basic surgical training together with specific techniques learning for the craniofacial area and a knowledge about malformations.
Surgical treatment of craniofacial malformations can only be conceived through collaboration between neurosurgery and plastic-maxillo-facial surgery, guided by the knowledge of the malformation, under the close supervision of anaesthetists and paediatricians. This surgery is not limited to remodel morphology but should integrate also the recovery of the functions that are maintained by rehabilitation treatment.

Riassunto

Le malformazioni complesse del distretto cefalico comprendono tutta una serie di anomalie di sviluppo che oltre a presentare una notevole difficoltà classificativa e di inquadramento, comprendono forme che sono talora di stretta pertinenza specialistica.
La correzione delle malformazioni craniofacciali fa ricorso ad una disciplina chirurgica particolarmente esigente, che deve associare una formazione chirurgica di base con un apprendimento delle tecniche specifiche per il territorio craniofacciale e una conoscenza della patologia malformativa. L'eziologia malformativa pone il chirurgo davanti a problemi specifici, dominati dallo sviluppo della tecnica chirurgica. Si stabilisce dunque un protocollo terapeutico dopo aver identificato la malformazione, stimato la gravità delle alterazioni tissutali e valutato la capacità di crescita dei territori interessati. E' fondamentale, oltre che il rimodellamento della morfologia, il recupero delle funzioni ed il trattamento rieducativo. E' quindi evidente che si tratta di una chirurgia di equipe che può portare risultati positivi solo se realizzata con fiducia reciproca tra i vari operatori.

References

1. Thanapaisal C, Chowchuen B, Chowchuen P: *Craniofacial surgery for craniosynostosis: Challenges in diagnosis, management and long-term outcome*. J Med Assoc Thai, 2010; 93 Suppl 4:S24-33.

2. Posnick JC, Ruiz RL, Tiwana PS: *Craniofacial dysostosis syndromes: Stages of reconstruction*. Oral Maxillofac Surg Clin North Am, 2004; 16(4):475-91.

3. Miller JJ, Schendel, SA: *Invited discussion: Surgical treatment of treacher collins syndrome*. Ann Plast Surg, 2006; 56(5).

4. Tanna N, Wan DC, Kawamoto HK, Bradley JP: *Craniofacial microsomia soft-tissue reconstruction comparison: inframammary extended circumflex scapular flap versus serial fat grafting*. Plast Reconstruct Surg, 2011; 12(2).

5. Kim J-T, Siew-Weng, Youn-Hwan Ki: *Application of various com-*

positions of thoracodorsal perforator flap for craniofacial contour deformities. Journ Plast Reconstruct Aesth Surgery, 2011; 64:902-10.

6. Bradley JP, Hurwitz DJ, Carstens MH: *Embryology, classification and descriptions of craniofacial clefts*. In: Mathes SJ(ed): *Plastic Surgery.* Vol. 4, 2nd ed. Philadelphia: Elsevier; 2006;15-43.

7. Iannetti G, Torroni A, Chiummariello S, Cavallotti C: *Clinical and morphological characteristics of head-facial haemangiomas*. Head Face Med, 2007; 3:12.

8. Alfano C, Chiummariello S, De Gado F, Fioramonti P, Bistoni G, Scuderi N: *Divided nevus of the eyelids: Three case studies*. In Vivo, 2007; 21(1):137-39.

9. Chiummariello S, Angelisanti M, Arleo S, Alfano C: *Surgical treatment of upper eyelid coloboma: Our experience*. Ann Ital Chir, 2012; 83(5):379-83.

10. Scuderi N, Ribuffo D, Chiummariello S: *Total and subtotal upper eyelid reconstruction with the nasal chondromucosal flap: A 10-year experience*. Plast Reconstr Surg, 2005; 115(5):1259-265.

11. Gokalp S, Gulden A, Mithat Akan, Gaye Taylan, Ilhan Elmaci, Tayfun Akoz: *The surgical treatment of plagiocephaly*. Turkish Neurosurgery. 2011; 21(3): 304-14.

12. McCarthy JG, Glasberg SB, Cutting CB, Epstein FJ, Grayson BH, Ruff G, Thorne CH, Wisoff J, Zide BM: *Twenty-year experience with early surgery for craniosynostosis: The craniofacial synostosis syndromes and pansynostosis. Results and unsolved problems*. Plast Reconstr Surg, 1995; 96(2):284-95; discussion 296-98.

13. Washington KM, Zanoun RR, Cadogan KA, Afrooz PN. Losee JE: *Composite Tissue Allotransplantation for the Reconstruction of Congenital Craniofacial Defects*. Transplantation Proceedings, 2009; 41, 523-27.