

Surgical correction of Tessier number 10 cleft

Correção cirúrgica de fenda número 10 de Tessier

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RESUMO

Objetivo: Revisar as diferentes técnicas cirúrgicas utilizadas no tratamento de pacientes com fissura 10 de Tessier e deformidades associadas. **Método:** Estudo retrospectivo com avaliação de pacientes com fissura 10, entre 1996 a 2009, realizado no Hospital de Clínicas da Universidade Federal do Paraná e Centro de Atendimento Integral ao Fissurado Lábio-Palatal (CAIF) – Curitiba, PR. **Resultados:** Foram avaliados dez pacientes com fissura 10, cinco apresentavam a deformidade na forma isolada, três com associação das fissuras 10 e 7, e dois pacientes com fissura 10 e 2. As principais deformidades craniofaciais encontradas foram: coloboma de pálpebra superior (7), encefalocele frontal (4), ambliopia (4), macroblefaro (3), hipertelorismo e distopia orbital (3), e alteração da linha de implantação capilar frontal (4). Dos sete pacientes com coloboma de pálpebra superior, seis foram tratados com reavivamento de bordos palpebrais e aproximação de retalhos, e um caso com retalho de Cutler-Beard. Nos casos de encefalocele frontal, após a correção do tecido cerebral herniado, foram submetidos à reconstrução óssea frontal. **Conclusões:** A fissura 10 possui duas formas de apresentação (coloboma e encefalocele frontal). Este é o primeiro trabalho exclusivo desta malformação rara.

Descritores: Fissura facial. Anormalidades craniofaciais. Encefalocele. Coloboma.

SUMMARY

Purpose: To review different surgical techniques used to manage patients presenting Tessier cleft #10 and associated deformities. **Method:** A retrospective study including Tessier #10 cleft patients treated between 1996 and 2009, in Hospital de Clínicas of Federal University of Paraná and Assistance Center for Cleft Lip and Palate (CAIF) – Curitiba, PR. **Results:** There were ten patients with Tessier #10 cleft, five presented as isolated deformity, three had association between cleft #10 and 7, and two cases of Tessier #10 and 2 cleft. The main craniofacial deformities diagnosed were: upper eyelid coloboma (7), frontal encephalocele (4), amblyopia (4), macroblefaron (3), hypertelorism and orbital dystopia (3), and frontal hairline alteration (4). Six cases of upper eyelid coloboma were treated with primary closure, and one case with Cutler-Beard flap. Patients with frontal encephalocele were submitted to the resection of the herniated cerebral tissue and calvarium reconstruction. **Conclusions:** The Tessier # 10 cleft has two presentation forms (eyelid coloboma and frontal encephalocele). It is the first exclusive study on this rare malformation.

Descriptors: Facial cleft. Craniofacial abnormalities. Encephalocele. Coloboma.

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INTRODUCTION

Craniofacial clefts are rare deformities of unknown cause. The incidence range from 1.4 to 4.9 per 100,000 live births¹. The exact cause of craniofacial clefts is unknown and those theories of cleft formation that apply to typical cases of cleft lip and palate may also be applicable to the craniofacial clefts. It is probably caused by bone incomplete fusion or primary ossification centers failure during the embryonic development. Tessier classification system is based on anatomical parameters and is universally accepted to describe rare craniofacial clefts².

The bony defect of Tessier #10 cleft patients is located on the upper central orbit, lateral to supraorbital nerve, with extension through orbital roof and frontal bone, it may be associated to frontal encephalocele (Figure 1)³. The upper eyelid coloboma is located on medial third, and it may be the form of commitment of soft tissues. The eyebrow may be divided into two regions: lateral with vertical positioning, sometimes together with the hairline and the medial region which is atrophic and occasionally absent⁴. Tessier described that cleft #10 represents the northbound of cleft #4.

Few studies on rare craniofacial clefts were published. Specifically on cleft #10, we did not find in the English literature studies or series on this subject. Tessier et al.⁴, in their book, presented methods of repair of some of these deformities. Our study consists in a revision of different surgical techniques used to manage patients presenting cleft #10 and associated deformities.

METHODS

This is a retrospective study including Tessier #10 cleft patients treated between 1996 and 2009, in Hospital de Clínicas of Federal University of Paraná and Assistance Center for Cleft Lip and Palate (CAIF) – Curitiba, PR.

Data were collected including soft tissues and osseous commitment.

All the tomographic studies and surgical procedures were re-evaluated.

RESULTS

There were ten patients with Tessier #10 cleft, five presented as isolated deformity, three had association between cleft #10 and 7, and two cases of Tessier #10 and 2 cleft (Table 1). The craniofacial deformities diagnosed were: upper eyelid coloboma (7), frontal encephalocele (4), amblyopia (4), macroblefaron (3), hypertelorism and orbital dystopia (3), and frontal hairline alteration (4). There was also association with other malformation such as: preauricular appendix (3), microtia (1), cleft lip and palate (2), macrostomia (3), mandibular branch agenesis (1), external ear auditive conduct atresia (1), lacrimal duct bifid (1), hemifacial microsomia (1).

Three patients were only evaluated in our service, but they were not operated. There were seven patients with upper eyelid coloboma (Figure 2), six were treated with primary closure, and one case was treated in another service with Cutler-Beard flap. There were 2 cases of leukoma, prior to our intervention. The displaced eyebrow was treated with the remaining eyebrow flap rotation, hair transplantation and tattooing. The loss of frontal hairline was corrected with the resection of the committed area. The number of surgical procedures per patient ranged from two to ten, according to the deformity degree.

Patients with frontal encephalocele needed the resection of the herniated cerebral tissue, associated to calvarium reconstruction (Figure 3). Two patients were grafted with inner layer of parietal bone, after splitting. One patient, who received previously split costal graft in another hospital, was submitted to alloplastic implant of porous polyethylene, to solve the contour irregularity. The fourth case of encephalocele was only evaluated in our service, and was not submitted to surgical procedure.

DISCUSSION

Basically, Tessier #10 cleft has two presentation forms: upper eyelid coloboma and/or frontal encephalocele (Figure 1).

Figure 1 – Tessier #10 cleft. **A:** Upper eyelid coloboma. **B:** Large frontal encephalocele.



Figure 2 – Patient with Tessier # 7 – 10 cleft. **A:** Male, one year of age, with upper eyelid coloboma, macrostomia, eartags and microtia. **B:** Post-operative picture. Patient was undergone to repair of eyelid in the first year.

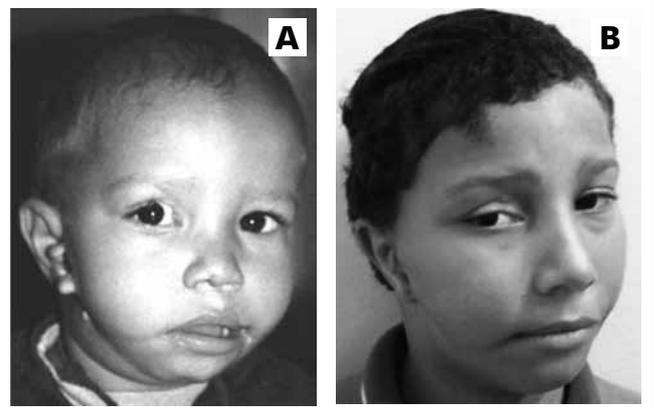


Figure 3 – Patient had his encephalocele operated earlier.

A: He presented orbital dystopia, macroblepharia, and absence of eyebrow at 4 years of age. **B:** It was performed orbital reposition, medial canthus rotation, eyebrow reconstruction. **C:** Picture of 6 years of follow-up.



Corneal protection is the primary goal in the medical treatment of eyelid colobomas. The eyelid deformity may be most commonly triangular in shape, with the base at the eyelid margin. It is usually located on the medial half of the upper eyelid and may vary in size from a small indentation of the eyelid margin to near absence of the entire eyelid (ablepharia). The treatment depends directly on the defect's size⁵. The rule of 25% for eyelid reconstruction is imperative to choose the method of repair. Defects until 25% of eyelid extension may be closed primarily; between 25-50%, a lateral canthotomy may be performed; and cases with commitment larger than 50% is necessary to associate local and regional flaps. Most of our patients presented with small upper eyelid colobomas, which were closed with local flaps. According to Tessier et al.⁴, the upper eyelid colobomas are symmetrical, the cleft is frequently incomplete and does not involve completely the height of tarsus. The surgical treatment is quite simple and is based on freshening of the coloboma borders. The resection should include all the vertical extension of the tarsus to achieve good continuity of tarsal plates. A small Z-plasty should be performed avoiding skin retraction.

Monasterio & Taylor⁶ presented a large series of 495 patients with major craniofacial clefts, and a total of 998 clefts. Tessier #10 cleft represented 2% of the malformations. The incidence of isolated Tessier #10 cleft was of three cases. Seventeen

patients had cleft #10 associated to other clefts. Amniotic band syndrome, particularly of the limbs, was associated with rare facial clefts. Coady et al.⁷ reported 24 patients, nine cases of Tessier #10 cleft associated with limb ring constriction and 15 cases with no correlation.

Encephaloceles are neural tube closure defects, a persistent ectodermal connection between the skin and the meninges, or even with the cerebral parenchyma. The prognosis for encephalocele is largely dictated by three variables: anatomical site, volume of neural contents and the presence of coexisting malformations (cerebral and extracerebral)⁸. It is necessary to perform the resection of neural tissue, in order to correct this defect. Most part of the encephalocele content is composed by glial tissue and a few neurons. The donor area for bone graft may be the parietal region, using the bipartition technique, but it is difficult to be performed because of the small thickness and absence of dysplasia of the cranial bones. This maneuver avoids remaining defects on the donor area. In one of our four cases, it was possible to perform this procedure in spite of the low age (6 months). The use of rib graft (bipartition) was indicated in another service, because it leads to frontal contour deformities, we don't think it is a good option, only in extreme cases. The use of alloplastic material is not indicated to young patients. The use of porous polyethylene was performed in one adult patient, to correct a depression located in the frontal area. Our three patients with frontal encephalocele had a good evolution after cranioplasty.

The most challenging deformity of Tessier #10 cleft is the macroblepharon. The eyelid lengthening is elongated, with medial and lateral canthus laterally displaced. So it is very difficult to be completely corrected. In our three cases, after the orbital dystopia correction, medial canthopexia was associated with bony fixation. The lateral canthus was medially positioned to reduce the width of the eyelid. We could not achieve the perfect correction of the macroblepharon in our cases. We did not find either a surgical technique description to correct this malformation.

Ablepharia of the upper eyelid is defined where the remnants represent less than a third of the eyelid. Tessier referred that a large dermatocele may be covered by cutaneous epithelium reaches the upper half of the cornea, and the superior rectus muscle may divided and be unable to turn the globe upwards. Acquired blindness may occur, and it is necessary early approach to patients with ablepharia⁴.

Tessier described that the # 10 cleft represents the continuation of cleft #4⁴. In our personal experience of 310 cases of rare craniofacial clefts^{9,10}, we did not observe the association between these two clefts. Cleft # 10 was associated to clefts # 2 and 7 in this series. Tessier # 4 clefts did not present any cranial commitment.

CONCLUSION

Children born with facial cleft represents a surgical challenge and their treatment requires both aesthetic sense and technical skills to repair form and function. The treatment of rare craniofacial cleft patients represents a great challenge to the craniofacial surgeon, it should be managed according to the complexity, localization, variability and extension of the

Table 1 – Craniofacial Cleft #10. List of patients.

Patients	Age	Gender	Cleft	Side of #10	Upper eyelid	Eyebrow	Hairline	Frontal bone	Orbita	Others
1	1	M	10	R	medial coloboma	normal	clefted	normal	normal	ear tags, macrostomia, microtia
2	0	F	10 and 7	L	medial coloboma	normal	normal	normal	normal	ear tags, macrostomia
3	0	M	10 and 7	R	medial coloboma	normal	normal	normal	normal	ear tags, cleft lip and palate
4	4	M	10	L	macroblepharia	clefted	normal	encephalocele	dystopia	lacrima duct bifid
5	25	M	10	R	medial coloboma	absent	clefted	normal	normal	leukoma
6	0	F	10 and 7	L	ablepharia	normal	normal	normal	normal	hemifacial microsomia (macrostomia, mandible hypoplasia)
7	0	M	10	R	medial coloboma	clefted	normal	encephalocele	dystopia	absence of orbital roof, cleft lip and palate
8	8	M	10	L	medial coloboma	clefted	clefted	normal	normal	leukoma
9	20	M	10	R	medial canthus dystopia	clefted	normal	encephalocele	normal	no
10	6	F	10	R	medial coloboma	clefted	normal	encephalocele	normal	no

deformities. The Tessier # 10 cleft has two presentation forms (eyelid coloboma and frontal encephalocele). It is the first exclusive study on this rare malformation).

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