Venezuelan Protocol for Management of Patients with Syndromic Craniosynostosis and Morphometric Craniofacial Changes with Frontofacial Monobloc Advancement Procedure Using Internal Osteogenic Distraction

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Abstract

Purpose: To describe the management of patients with syndromic craniosynostosis using the monobloc frontofacial advancement technique currently used in Venezuela.

To analyze the morphometric changes in patients with syndromic craniosynostosis, who underwent monobloc with facial bipartition advancement with internal osteogenic distraction devices and quantification of the vectors of movement by pre- and post-surgery CT scan and radiograph analysis.

Materials and Methods: A retrospective study of cases diagnosed with syndromic craniosynostosis who underwent for surgery correction at the Dr. Ángel Larralde University Hospital in Valencia, Carabobo, Venezuela from 2018 to the present year, without distinction of gender, age, type of syndrome, was carried out.

Describing the protocol currently used in our country and in the same way, evaluating the morphometric changes in patients with syndromic craniosynostosis, who underwent monobloc and/or facial bipartition advancement with internal osteogenic distraction devices by pre and post-surgery cephalometric analysis.

Results: A total of 3 patients were studied, where the combination of monobloc osteotomy, Le Fort III, bipartition and fasciotomy was performed with the use of 4 internal distractors, with an angulation of 35º between two parieto-frontal distractors and two temporozygomatic distractors, these patients were between the ages of between 8 and 11 years old, 2 female and 1 male, all diagnosed with Crouzon Syndrome. In these cases, cephalometry was performed on preoperative (T1) and same postoperative analysis at the end of the consolidation phase of distraction osteogenesis (T2), where each craniometric point was quantified and compared to establish the resulting movement vector in CT scan and radiograph analysis. Other changes can be seen in a sagittal plane, which reflect advancement of the upper and middle third, 17.33 mm ± 1.26 for the middle third and 13.33 mm ± 1.15 in the upper third. Similarly, a decrease of up to 3.00 mm ± 0.50 in the infraorbital region can be seen in the middle third, however, in the upper third there is a rise in the Gb point of 0.5 mm ± 1.50 and a decrease in the N point of just 0.50 mm ± 0.50. In relation to these values, divergent distraction vectors are expressed, with a linear advancing distraction vector in the upper third and a simultaneously advancing and descending distraction vector in the middle third. Distraction vectors indicate an average of 13.83 mm advance for the upper third and 17.67 mm forward and downward for the middle third. In a coronal plane, transversal dimension changes were obtained with a decrease in the interorbital distance, which is evidenced with a mean of -7.50 mm ± 0.50 between the frontozygomatic sutures (Z) and -6.67mm ± 0.76 between both. Medial walls at Dacyron’s point level (Dc). However, at the level of the zygomatic arches, an increase of transverse width is shown with a mean of 1.33 mm ± 0.76 and expansion in the maxillary region at the level of the point (J) of 3.50 mm ± 0.50. Thus, obtaining in this study a transversal decrease in the orbital region and slight increase in the most inferior area of the middle third.
Respect to orbital advancement, rotation counterclockwise of the orbital cone for diopter correction is crucial for visual field improvement in patients with syndromic craniosynostosis.

A mayor objective is to eliminate the dependence of a tracheostome in these patients and to correct the obstructive apnea syndrome due to middle third deficiency, by anteroposterior advance of the airway.

Whenever possible, the use of endoscopic surgery is a gold standard to correct craniosynostosis from 6 months of age. Allowing rapid brain expansion and a recovery time of 2 days, without the need for pediatric Intensive Care Unit.

Frontofacial advancement through osteogenic distraction allows us to make great anteroposterior advancement, superior to rigid internal fixation technique and in turn allow a correct adaptation of the soft tissue, improving function and aesthetics.

Conclusions: The fundamental keys to success are reflected in the experience and preparation of the surgical team.

Optimization of intraoperative time to minimize excessive blood loss is a crucial factor in the outcome.

Correction of intracranial hypertension, obstructive sleep apnea syndrome, the visual field in these patients.

Aesthetics changes to allow their rapid psychological and social integration at the school and the society as soon as possible to avoid bullying.

The preparation of the surgical team is essential to avoid intraoperative complications, both a trained anesthesiology team and a team of surgeons with surgical training in craniofacial surgery.

Introduction

Craniofacial surgeons use a variety of surgical techniques for the correction of craniofacial deformities of syndromic etiology, such as Crouzon, Pfeiffer, Apert, Saethre Chotzen syndrome, and non-syndromic etiology such as midline clefts or isolated orbital hypertelorism. Tessier was a pioneer in describing techniques in which the fronto-orbital band is advanced independently in conjunction with a Le Fort III osteotomy, to produce frontofacial advancement [1]. Later, in 1978, Ortiz Monasterios et al. advanced the orbital component, the middle facial third, and the replacement of the frontal bone as a single unit, which he called Monobloc, for the correction of Crouzon syndrome [2,3]. One year later, Van der Meulen et al. performed the correction of facial clefts in the midline with a procedure that he called “median fasciotomy” by vertically splitting the monobloc in a half and removing the ethmoid bone in order to correct orbital hypertelorism [4]. Tessier later changes the shape of this vertical osteotomy, for correction of the deformity in three planes, to what he describes as facial bipartition [1]. A variety of techniques for correction of craniosynostosis syndromes has been described, however the main objective is to correct the intracranial hypertension, ocular proptosis and sleep obstructive apnea syndrome. For children and young adults there are special considerations in the protocol. The purpose of this study was to describe the protocols currently used at the Dr. Ángel Larralde University Hospital in Valencia, Carabobo, Venezuela. And to clinically evaluate the improvement of function, aesthetics and psychosocial development of patients with syndromic craniosynostosis operated on at the institution.

Materials and Methods

A retrospective study of cases diagnosed with syndromic craniosynostosis who underwent for surgery correction at the Dr. Angel Larralde University Hospital in Valencia, Carabobo, Venezuela from 2018 to the present year. Describing the protocol currently used in our country and in the same way, to analyze morphometric changes in patients who underwent monobloc frontofacial advancement with osteogenic distraction, through the use of internal distractors with facial. Within the inclusion criteria of this research, the following were taken into account: Patients of any age, with syndromic craniosynostosis, of any ethnic group, operated on at the Oral and Maxillofacial Surgery Service of the Dr. Angel Larralde University Hospital in a period from 2018 to present day. Within the exclusion criteria: Craniosynostosis of non-syndromic etiology, monobloc frontofacial advance with internal rigid fixation. The data collection method was a retrospective revision of the medical clinical archives and individual clinical cases for each patient that underwent for surgery, as well as pre- and post-surgical cephalometric analysis of plain radiographs, at Dr. Angel Larralde hospital adscript to Carabobo University. Ethical approval by the Ethical committee of the institution was given for this investigation, We declare that we had read the Helsinki Declaration and followed its guidelines in this investigation as well.

Body

Surgical correction of syndromic craniosynostoses can be achieved by a Le Fort III osteotomy, an advance of a monobloc, either by rigid fixation or with the use of osteogenic distraction. It can also be achieved by early endoscopic surgery. At 6 months of birth, it is possible to make osteotomies of the affected cranial sutures using endoscopic surgery and thus allowing the brain to expand quickly. This technique improves the recovery time at hospital, maintaining at minimum (2 days) without the need of a pediatric intensive care unit, also Improving psychosocial and neurological aspects in the child as soon as possible, making endoscopic surgery the Gold Standard in syndromic craniosynostosis [5].

The technique to be used to surgically correct craniosynostosis and all conditions that it generates, (i.e., intracranial hypertension,
ocular conditions: Corneal keratitis, proptosis, amaurosis, alterations in the visual field, airway conditions: Obstructive sleep apnea syndrome, central sleep apnea, lifelong dependence on the use of a tracheostome. Neurological alterations), which prevents a normal adaptation of the patient to the society, will depend on the specific deformity of each patient [6,7]. The shape and position of the supraorbital rim and the lateral portion of the orbit are key pieces in the aesthetics of the upper third. In a profile view, the frontonasal angle normally varies between 90 and 110°. The frontal bone should have a smooth convexity, a curvature of the orbital rim which arches posteriorly at an angle of approximately 60°, and forming an angle with respect to the infratemporal fossa of 90°. In Crouzon syndrome, as well as in syndromic craniosynostosis, there is involvement of the base of the skull, which generates a deficiency in the naso-orbitozygomatic complex, the product of a skull base that is short in the anteroposterior axis and wide in the transverse axis, which gives us a true deficiency of the middle third, with a poor anteroposterior projection, a short upper third, a reduced maxillary width in the dentoalveolar area, and a high arched palate, exhibiting the classic symptoms of these patients [7]. However, the selection of a monobloc (with or without facial bipartition) or a correction by a Le Fort III osteotomy will depend on several factors, such as the projection of the orbital rim, seen from the plane sagittal, the arc of facial rotation, which depend on both the frontal projection and the middle third, and the presence or absence of hypertelorism. If any alteration affects all of these structures, then their correction is indicated by a monobloc advance. In addition to that, if there is a presence of hypertelorism, then this monobloc must be accompanied by a facial bipartition to correct the interorbital distance. In cases where there is an absence of the nasal dorsum, it must be reconstructed at the same time as the facial bipartition is completed. When there are deficiencies of the middle third that involve the lower orbital rim, that is, a true deficiency of the middle third, this can be corrected without the need for a monobloc, using a Le Fort III osteotomy. However, in patients with syndromic craniosynostosis who also exhibit a dysmorphia of the upper third of the face accompanied by a hypoplasia of the middle third of the face, typical features of some syndromes, the correction of the deformity with the use of a Le Fort III osteotomy alone is not effective, as confirms a study carried out by McCarthy et al. [8], becoming necessary to perform a monobloc advance in order to correct these alterations.

The ideal age to surgically correct these patients as long as intracranial hypertension is controlled is between 7 to 10 years of age, since at this age the cranial vault is 90% the size of an adult and preferably when the first molars have erupted. The objective of surgery is firstly to relieve intracranial hypertension, then to allow rapid brain expansion, as well as to achieve an adult facial morphology, stable over time once healing has occurred [9]. In the same way, it seeks to normalize the orbits, zygomatic projections and the cranial vault.

**Results**

A total of 3 patients were studied, where the combination of monobloc osteotomy, Le Fort III, bipartition and fasciotomy was performed with the use of 4 internal distractors, with an angulation of 35° between two parieto-frontal distractors and two temporozygomatic distractors, these patients were between the ages of between 8 and 11 years old, 2 female and 1 male, all diagnosed with Crouzon syndrome. In these cases, cephalometry was performed on preoperative (T1) and same postoperative analysis at the end of the consolidation phase of distraction osteogenesis (T2), where each craniometric point was quantified and compared to establish the resulting movement vector in CT scan and radiograph analysis. Other changes can be seen in a sagittal plane, which reflect advancement of the upper and middle third, 17.33 mm ± 1.26 for the middle third and 13.33 mm ± 1.15 in the upper third. Similarly, a decrease of up to 3.00 mm ± 0.50 in the infraorbital region can be seen in the middle third, however, in the upper third there is a rise in the Gb point of 0.5 mm ± 1.50 and a decrease in the N point of just 0.50 mm ± 0.50. In relation to these values, different distraction vectors are expressed, with a linear advancing distraction vector in the upper third and a simultaneously advancing and descending distraction vector in the middle third. Distraction vectors indicate an average of 13.83 mm advance for the upper third and 17.67 mm forward and downward for the middle third. In a coronal plane, transversal dimension changes were obtained with a decrease in the interorbital distance, which is evidenced with a mean of -7.50 mm ± 0.50 between the frontozygomatic sutures (Z) and -6.67 mm ± 0.76 between both. Medial walls at Dacrion’s point level (Dc). However, at the level of the zygomatic arches, an increase of transverse width are shown with a mean of 1.33 mm ± 0.76 and expansion in the maxillary region at the level of the point (J) of 3.50 mm ± 0.50. Thus, obtaining in this study a transversal decrease in the orbital region and slight increase in the most inferior area of the middle third (Table 1).

Other changes can be seen in a sagittal plane, which reflect

**Table 1: Procedure.**

<table>
<thead>
<tr>
<th>Gender</th>
<th>Age</th>
<th>Syndrome</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>F 11</td>
<td>Sx Crouzon</td>
<td>Monobloc, Le fort III, Biparticion + DO interna</td>
<td></td>
</tr>
<tr>
<td>F 10</td>
<td>Sx Crouzon</td>
<td>Monobloc, Le fort III, Biparticion + DO interna</td>
<td></td>
</tr>
<tr>
<td>F 8</td>
<td>Sx Crouzon</td>
<td>Monobloc, Le fort III, Biparticion + DO interna</td>
<td></td>
</tr>
</tbody>
</table>

**Table 2: Comparison and quantification respect to “y” axis in the lateral cephalographic and CT scan.**

<table>
<thead>
<tr>
<th>Osteotomy</th>
<th>Point</th>
<th>T1 mm</th>
<th>T2 mm</th>
<th>Difference</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean</td>
<td>ED</td>
<td>Mean</td>
</tr>
<tr>
<td>Monobloc + Distraction</td>
<td>Gb</td>
<td>89.00</td>
<td>6.56</td>
<td>102.33</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>86.33</td>
<td>6.51</td>
<td>98.00</td>
</tr>
<tr>
<td></td>
<td>Or</td>
<td>74.00</td>
<td>3.61</td>
<td>91.33</td>
</tr>
<tr>
<td>Bipartition with OD</td>
<td>Ena</td>
<td>84.50</td>
<td>3.28</td>
<td>99.83</td>
</tr>
<tr>
<td></td>
<td>A</td>
<td>81.00</td>
<td>5.00</td>
<td>94.83</td>
</tr>
<tr>
<td></td>
<td>Is</td>
<td>84.33</td>
<td>6.11</td>
<td>97.00</td>
</tr>
</tbody>
</table>

Gb: Glabella; N: Nasion; Or: Infracristal rim; Ena: Anterior Nasal Espine; A: point A; Is: superior central incisor edge; T1: Preoperatory radiograph; T2: Postsurgical Radiograph (posterior to consolidation phase); OD: Osteogenic Distraction; ED: Standard Deviation

**Table 3: Vectors movement quantification analysis in CT scan and radiograph.**

<table>
<thead>
<tr>
<th>Osteotomy</th>
<th>Point</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean (mm)</td>
</tr>
<tr>
<td>Monobloc + Distraction</td>
<td>Gb</td>
<td>13.83</td>
</tr>
<tr>
<td></td>
<td>N</td>
<td>12.17</td>
</tr>
<tr>
<td></td>
<td>Or</td>
<td>17.67</td>
</tr>
<tr>
<td>Bipartition with OD</td>
<td>Ena</td>
<td>16.00</td>
</tr>
<tr>
<td></td>
<td>A</td>
<td>14.00</td>
</tr>
<tr>
<td></td>
<td>Is</td>
<td>11.67</td>
</tr>
</tbody>
</table>

Gb: Glabella; N: Nasion; Or: Infracristal rim; Ena: Anterior Nasal Espine; A: point A, Is: superior central incisor edge
advancement of the upper and middle third, 17.33 mm ± 1.26 for the middle third, and 13.33 mm ± 1.15 in the upper third (Table 2). Similarly, a decrease of up to 3.00 mm ± 0.50 in the infraorbital region can be seen in the middle third, however, in the upper third there is a rise in the Gb point of 0.5 mm ± 1.50 and a descent of the N point of 0.50 mm ± 0.50 (Table 3 and Figure 1).

In relation to these values, divergent distraction vectors are expressed, with a linear advancing distraction vector in the upper

Table 4: Quantification and comparison in the posteroanterior radiograph analysis and CT scan.

<table>
<thead>
<tr>
<th>Osteotomy</th>
<th>Point</th>
<th>T1 mm</th>
<th>T2 mm</th>
<th>Difference</th>
<th>Transverse total width</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Mean right side</td>
<td>Mean left side</td>
<td>Total</td>
<td>Mean right side</td>
</tr>
<tr>
<td>Monobloc, Le Fort III, Bipartition with OD</td>
<td>Z</td>
<td>52.50</td>
<td>52.50</td>
<td>105.00</td>
<td>48.67</td>
</tr>
<tr>
<td></td>
<td>Dc</td>
<td>15.33</td>
<td>15.17</td>
<td>30.50</td>
<td>11.83</td>
</tr>
<tr>
<td></td>
<td>Za</td>
<td>60.00</td>
<td>59.67</td>
<td>119.67</td>
<td>60.33</td>
</tr>
<tr>
<td></td>
<td>J</td>
<td>31.67</td>
<td>31.50</td>
<td>63.17</td>
<td>34.33</td>
</tr>
</tbody>
</table>

Z: Medial aspect of the frontozygomatic suture; Dc: Dacron's; Za: Most lateral aspect of the Zygomatic arch; J: Jugal process

Figure 1: Quantification and comparison in the lateral cephalic radiograph analysis and CT scan. A) Preoperatory cephalometry and CT scan, B) T1 post quirurgica cephalometry and CT Scan analysis posterior to consolidation phase, C) Comparison of T1 and T2.

Figure 2: Quantification and comparison of the movement of the vectors. A) Preoperatory cephalometry and CT scan, B) T1 post-quirurgica cephalometry and CT Scan analysis posterior to consolidation phase, C) Comparing the vectors of movement in T1 and T2.

Figure 3: Quantification and comparison in the posteroanterior radiograph analysis. A) Preoperatory cephalometry and CT scan, B) T1 post-quirurgica cephalometry and CT scan analysis posterior to consolidation phase, C) Comparation of T1 and T2.
third and a simultaneously advancing and descending distraction vector in the middle third. Distraction vectors indicate an average of 13.83 mm advance for the upper third and 17.67 mm forward and downward for the middle third (Table 4 and Figure 2,3).

Otherwise, in a coronal plane, transversal dimension changes were obtained with a decrease in the interorbital distance, which is evidenced with a mean of -7.50 mm ± 0.50 between the frontozygomatic sutures (Z) and -6.67 mm ± 0.76 between both. Medial walls at Dacron’s point level (Dc). However, at the level of the zygomatic arches, an increase of transverse width is shown with a mean of 1.33 mm ± 0.76 and expansion in the maxillary region at the level of the point (J) of 3.50 mm ± 0.50 (Table 4). Thus, obtaining in this study a transversal decrease in the orbital region and slight increase in the most inferior area of the middle third.

Discussion

After performing the monobloc advance, it is very important to manage the extradural dead space that is produced when advancing, which produces communication between the cranial vault and the nasal cavity, which facilitates the passage of air and bacteria between the nasal cavity and the nasal cavity [5-7,10]. Therefore, when it is not properly sealed, it can lead to the formation of cerebrospinal fluid leak by a formation of cranionasal fistulas, which can be verified for the presence of rhinorrhea by using a glucometer, halo test, or by means of the presence of the B2 transferase isoenzyme by western blot. As well as a bacterial gateway which can end in a potentially lethal meningitis. To seal it, the same peri-cranial tissue can be used, as well as flaps, autologous grafts, fibrin sealants, gel foam [7]. The function of this sealing is to provide a separation for a period of time to allow re-epithelialization of the nasal mucosa. However, a study published by Posnick et al. suggests that this space is occupied and sealed by the expanding brain itself after surgery at 6 to 8 weeks (Figures 5-9) [11-13]. An excellent option to avoid the formation of this extradural space is by advancing the monobloc by means of distraction osteogenesis. Through this technique, osteogenic distractors are positioned in the body of the zygoma, prior to the
osteotomy, as well as in the frontotemporal region, to produce forward and downward bidirectional counterclockwise advancement [14,15]. After the placement of the osteogenic distractor, a latency period of 7 days must be waited and then the distraction can be started at 1 mm per day (0.5 mm twice a day) for 2 to 4 weeks [1,6,7,10]. Allowing a progressive expansion of the brain as the distractor activation is carried out, especially in adults and older infants, and subsequently the progressive advance and finally a consolidation phase of 3 months and subsequent removal of the distractors on the operating table.

We prefer the latter, despite the fact that a second surgical procedure is required to remove the distractors, the results are excellent. It is important to emphasize that the maximum immediate advancement that can be achieved by immediate advancement is 15 mm, without causing amaurosis in the patient, due to optic nerve elongation, and the formation of the extradural dead space, especially in adults and infants, which generates this predisposition to meningeal infections and fistulas, which can be avoided in this procedure, being performed at an early age (<3 years of age) because the brain reaches the size of an adult at 3 years of age [5]. An advantage of distraction osteogenesis is that it minimizes this dead space, by advancing progressively and also allowing a progressive
expansion of the brain tissue and an adaptation of the upper soft tissue and muscle tissue, compare to the immediate advance and rigid fixation [10]. In the same way, it is necessary to accompany the movement of the monobloc with the use of intermaxillary fixation with class III elastics, in favor of the movement, to maintain occlusal stability [6]. In those patients that depend of a tracheostome, or with a considerably reduced airway, we perform, in addition to monobloc advancement, osteogenic distraction of the mandibular body in a forward unidirectional vector, allowing total facial advancement through osteogenic distraction whose objective is to increase the airway and eliminate, once the distractors are removed, the need for the tracheostome [10].

**Conclusion**

To achieve the most favorable facial harmony and head and neck function in the individual with a craniosynostosis syndrome, the surgeon needs both an esthetic sense and the technical expertise to execute effective upper and midface surgical procedures. Several key aspects of such expertise are:

1. The ability to remove, segment, reshape, and then stabilize the cranial vault.
2. The ability to separate the orbits and midface as a unit (monobloc) from the skull base.
3. The ability to segment and reshape the upper orbits of the monobloc, including interposing bone grafts as needed to reconstruct each orbital esthetic unit during a single operative setting.
4. The ability to separate the MB into halves (facial bipartition) and then three dimensionally reposition and stabilize the two facial halves (plate and screw fixation) to achieve the most favorable morphology in all three planes (i.e., pitch, roll, and yaw orientation). This often requires an increase in the maxillary width (i.e., arch expansion) and a decrease in the upper face width (i.e., correction of hypertelorism of the orbits, zygomas, and bitemporal regions). Facial bipartition also provides the ability to correct the transverse facial arc of rotation, such as changing the concave facial arc of rotation (i.e., yaw orientation) in Apert syndrome.
5. The Pryor management of intracranial hypertension and compromised airway.
6. Masticatory function and diopter corrections and visual field improvement, and psychosocial positive changes to avoid bullying at school and a correct individual integration to society as soon as possible.
7. The preparation of the surgical team is essential to avoid intraoperative complications, both a trained anesthesiology team and a team of surgeons with surgical training in craniofacial surgery.

**References**