Original Article

Therapeutic appro Therapeutic Approach To The Parry-Romberg Syndrome Based On A Severity Grading System

Abordagem terapêutica da síndrome de parry-romberg baseada em um sistema de classificação de gravidade

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ABSTRACT

Introduction: The Parry-Romberg Syndrome (PRS) is characterized by progressive hemifacial atrophy that often leads to severe esthetic and functional difficulties. Although there are systems for grading disease severity, none have proven ideal in optimizing the therapeutic approach to these patients. This study aimed to establish the surgical strategies for the treatment of PRS based on a new system for severity grading of the disease. **Methods:** This retrospective study included PRS patients undergoing surgery between 2005 and 2011. The surgical strategies were adapted for each patient according to a clinical severity grading system based on disease progression: type I, affecting the epidermis, dermis, and subcutaneous tissue; type II, type I + muscle involvement; and type III, Types I+ II + bone involvement. The sample included four patients (28.57%) with PRS type I, six patients (42.85%) with PRS type II, and four patients (28.57%) with PRS type III. **Results:** Forty-seven procedures were performed. Free-fat grafts were used in all patients. Dermal fat grafts were used in all type II patients and one type III patient (25%). Bone grafts with temporoparietal fascia flaps were performed for the treatment of all type III patients. One type III patient (25%) underwent orthognathic surgery. All patients were improved in their overall facial appearance and there were no procedure-related complications. **Conclusion:** Our proposed system for grading PRS severity can facilitate the choice of therapeutic approaches and with a combination of surgical techniques based on the severity of the disease partially satisfactory outcomes can be attained.

Keywords: Parry-Romberg syndrome; Progressive hemifacial atrophy; Surgical treatment; Grading system.

RESUMO

Introdução: Síndrome de Parry-Romberg (SPR) é caracterizada pela atrofia hemifacial progressiva que, muitas vezes, resulta em graves distúrbios estéticos e fun-

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cionais. Embora existam escalas de gravidade, nenhuma delas é completamente ideal para auxiliar na abordagem terapêutica destes pacientes. O objetivo deste estudo foi delinear as estratégias cirúrgicas para o tratamento da SPR baseado em um novo sistema de classificação de gravidade da doença. **Método:** Trata-se de uma análise retrospectiva dos pacientes com SPR operados em 2005-2011. As abordagens cirúrgicas foram individualizadas de acordo com a escala de gravidade clínica baseada na evolução da doença: tipos I (envolvimento da epiderme, derme e tecido subcutâneo); II (tipo I + envolvimento muscular); e III (tipo I + II + envolvimento ósseo). Quatro (28,57%) pacientes com SPR tipo I, 6 (42,85%) tipo II e 4 (28,57%) tipo III foram incluídos. **Resultado:** Um total de 47 procedimentos foi realizado. Gordura livre foi enxertada em todos os pacientes. Todos os pacientes do tipo II e 1 (25%) do tipo III foram submetidos a enxertos dermogordurosos. Enxertos ósseos com retalhos de fáscia têmporo-parietal foram aplicados no tratamento de todos os pacientes do tipo III. Um (25%) paciente do tipo III foi submetido à cirurgia ortognática. Houve melhora global na aparência facial em todos os pacientes, sem complicações relacionadas aos procedimentos. **Conclusão:** O sistema de classificação de gravidade proposto para a SPR pode facilitar a decisão terapêutica e resultados parcialmente satisfatórios podem ser alcançados com a combinação de técnicas cirúrgicas de acordo com a gravidade da doença.

Descritores: Síndrome de Parry-Romberg; Atrofia facial progressiva; Tratamento cirúrgico; Sistema de classificação.

INTRODUCTION

The Parry-Romberg syndrome (PRS), also known as progressive hemifacial atrophy, is a rare craniofacial condition of unknown etiology. The disease is primarily characterized by progressive hemifacial atrophy, affecting the skin, subcutaneous tissues, muscles, nerves, cartilage, and, less frequently, the bones[1,2]. These alterations often lead to tridimensional asymmetry in the faces of these patients and are associated with severe functional and psychological disturbances[1,2]. As such, craniofacial surgeries, aimed at restoring facial symmetry, are a crucial part of the strategy leading to the full rehabilitation of these patients[1,3]. However, and in spite of the three previously reported grading systems for PRS severity,[4,6] there is no consensus regarding the appropriate surgical procedures to be employed for each degree of severity[1,3-6].

Thus, the aim of this study was to establish craniofacial surgical strategies for the treatment of PRS based on a new grading system for the severity of the Parry-Romberg syndrome.

METHODS

This is a retrospective study of all PRS patients surgically treated in the Sobrapar Hospital between 2005 and 2011. All aspects concerning surgical interventions were verified by analyzing the medical records, photographs, and clinical interviews. Patients with facial atrophy of known origin (trauma, burns, or craniofacial tumors) and patients with incomplete medical files and/or with incomplete postsurgical follow-up were excluded.

Fourteen patients with a diagnosis of PRS met the inclusion criteria. The average patient age was 19.4 years. Eleven patients (78.57%) were women and three (21.43%) were men. Eleven patients (78.57) had atrophy on the right side of the face, two (14.29%) on the left side, and one (7.14%) had bilateral facial atrophy.

Surgical interventions

The four surgical procedures (free-fat grafts,[7,8] dermal fat grafts,[9] cranial bone grafts with temporoparietal fascia flaps (TPFF),[10,11] and orthognathic surgery[12]) used to correct the craniofacial defects of the PRS patients have been previously described.

Free-fat grafts:[7,8] The collection, preparation, and injection of the free-fat grafts were based on the technique described by Coleman et al[7]. A 2–3mm diameter cannula connected to a 10 mL manual syringe (negative pressure) was used to collect the fat tissue preferentially from the lower abdomen or less frequently from the medial thigh. The aspirated tissue was centrifuged for 2 minutes at 2000 rpm. Following the removal of the supernatant, the fat was transferred to 1 mL syringes and injected into the affected facial regions. Multiple access points, multiple tunnels, and multiple layers were used to transfer small aliquots of fat, at different depths, to the hypoplastic facial regions. Approximately 0.1ml of fat was deposited with each cannula insertion.

Dermal fat grafts[9]*:* The longest horizontal axis of lower abdominal region was determined. This area was deepithelized (total thickness) with the aponeurosis of the rectus abdominis muscle serving as the lower limit for the dissection. The dermal fat graft was cut in the shape of a triangle and then introduced in the affected facial region; meticulous technique ensured minimal facial undermining.

Cranial bone grafts with TPFF[10,11]*:* The superficial tem-

poral artery was carefully marked along its entire course. The initial incision of the scalp was performed in the most distal position in relation to the origin of the superficial temporal artery. The scalp was elevated in the subcutaneous plane towards the ear. Following the complete elevation of the scalp skin graft, the graft's axial pedicle was exposed. The thin superficial temporoparietal fascia was then elevated in its entirety, followed by dissection in the subgaleal plane. The periosteal flap was delineated and elevated to expose the frontal bone and the orbital cone. A craniotomy was performed in the parietal area. The parietal bone, in its entire thickness, was divided. One subciliary incision with zygomaticomaxillary exposure allowed the correct fixation of segments of the bone flap to the recipient hypoplastic region with 1.5mm screws. The external bone flap was returned to the donor parietal region. The TPFF was then rotated towards the region of the facial deformity to cover the bone flap.

Scale for grading clinical severity

All surgical approaches were individually considered according to the grading scale for the severity of the disease developed by our group. The distinction between the three degrees of severity was based on photographs, and on clinical, radiographic, and tomographic examinations, The three grades were: mild (type I), with epidermal, dermal, and subcutaneous tissue involvement; moderate (type II), with epidermal, dermal, and subcutaneous tissue involvement, as well as muscle involvement; and severe (type III), with epidermal, dermal, and subcutaneous tissue involvement, as well as muscle and bone involvement (Table 1). Based on this scale, four (28.57%) patients were classified as type I, six (42.85%) patients as type II, and four (28.57%) patients as type III.

Evaluation of craniofacial surgical results

All surgical results were evaluated by the same plastic surgeon who had no previous contact with the patients. Pre-operative frontal, oblique, and profile facial photographs, taken days or weeks before the first surgical procedure, were compared to post-surgery photographs taken 12–14 months after the last surgical procedure. Photographs were classified according to a scale for grading the degree of improvement of the facial symmetry previously used in PRS[8]: (a) satisfactory result, symmetrical face with no need for additional interventions; (b) partially satisfactory result, overall improvement of the facial appearance but facial asymmetry can still be observed after careful examination; and (c) unsatisfactory result, lack of evident improvement in facial symmetry after the surgical interventions.

RESULTS

The indications for surgical procedures were asymmetry/facial malformation (100%), hypoplasia of the soft tissues (71.43%), and hypoplasia of the soft tissues and bones (28.57%). In total, 47 craniofacial surgical interventions were performed based on the clinical severity of each patient's disease and also on the basis of the level of soft tissue altera-

Table 1 – Surgical approach according to the grading scale for severity based on the clinical findings in the Parry-Romberg syndrome proposed in this study.

	Involvement			
Type (Degree)	Epidermis, dermis, and subcutaneous cellular tissue	Muscle tissue	Bone structures	Surgical approach*
I (Mild)	+	–	–	Free-fat graft
II (Moderate)	+	+	–	Free-fat graft or dermal fat graft Cranial bone graft with TPFF
III (Severe)	+	+	+	+Increase in soft tissues ** + Combined orthognathic surgery #

+, Present; –, Absent; TPFF, temporoparietal fascia flaps;*, The individual number of surgical interventions should be defined according to the requirements of each patient; **, Because the cranial bones cannot be divided before seven years of age, only free-fat grafts can be performed in these cases; #, In case of altered occlusion and cessation of growth of the craniofacial bones.

tions detected by the pre-surgical evaluations. The number of surgeries performed in each patient was variable (1–6 procedures/patient). The average number of interventions per patient was 2.5 procedures/patient in type I PRS patients, 3.33 procedures/patient in type II patients, and 4.5 procedures/patient in type III patients. All patients received free-fat grafts at some point during the treatment period (type I: 1–4 free-fat grafts/patient; type II: 1–6 free-fat grafts/patient; and type III: 2–5 free-fat grafts/patient). Five (35.71%) patients received dermal fat grafts (type I: none; type II: 1–3 dermal fat grafts/patient; and type III: 1 dermal fat graft/patient). Four patients (28.57%) had cranial bone grafts with TPFF (types I and II: none; and type III: 1 cranial bone graft/patient) and one patient (7.14%) underwent orthognathic surgery (types I and II: none; and type III: 0.25 TPFF/patient) (Table 2).

Utilizing the different combinations of surgeries we attained an overall improvement in the facial appearance (partially satisfactory result) in all patients (Figures 1–5). In this series, there was no satisfactory or unsatisfactory result according to the scale introduced by Xie et al.[8] A variable degree of fat absorption was detected following free-fat grafting. There was no infection or necrosis in any of the dermal fat grafts or cranial bone grafts with TPFF. In addition, there were no complications in the donor areas.

Table 2 – Distribution of the surgical procedures performed in patients with Parry-Romberg syndrome (n=14) according to the clinical severity of the disease.

Clinical severity	Free-fat graft	Dermal fat graft	Cranial bone graft with TPFF	Combined orthognathic surgery	Total
Type I					
Patient A	++++	–	–	–	4
Patient B	+	–	–	–	1
Patient C	+++	–	–	–	3
Patient D	+	–	–	–	1
Total patients A-D	9	–	–	–	9
Type II					
Patient E	++	+++	–	–	5
Patient F	+	+	–	–	2
Patient G	+	+	–	–	2
Patient H	++++++	–	–	–	6
Patient I	+	+	–	–	2
Patient J	+++	–	–	–	3
Total patients E-J	14	6	–	–	20
Type III					
Patient K	++	–	+	–	3
Patient L	+++++	+	+	–	7
Patient M	+++	–	+	–	4
Patient N	++	–	+	+	4
Total patients K-N	12	1	4	1	18
Total all patients	35	7	4	1	47

+, Performed; –, Not performed; TPFF, temporoparietal fascia flaps

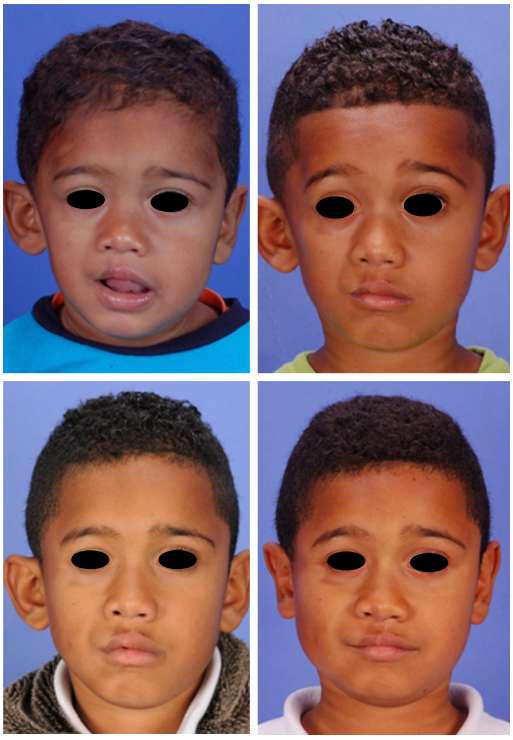

Figure 1 – *(Above, left) Pre-surgical frontal photographs of a three-year-old type I Parry-Romberg syndrome patient showing early manifestations of the disease (Table 2, patient D). (Above, right) Frontal photograph of the same patient, two years later, showing the slow progression of the disease in this case. (Below, left) At seven years of age, the patient underwent hemifacial free-fat inclusion and otoplasty for correction of his prominent ears. (Below, right) Two years after the surgery, the patient had a satisfactory esthetic result, with maintenance of the volume in the right hemiface and without any signs of recurrent ear deformity.*

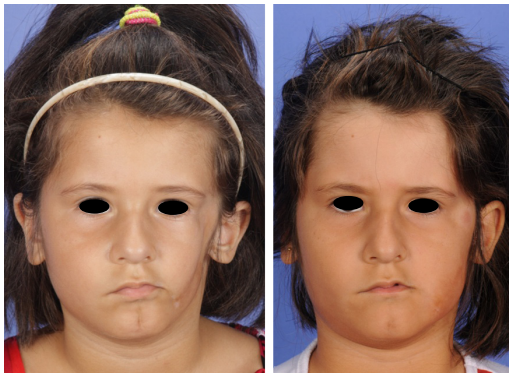

Figure 2 – *(Left) Pre-surgical frontal photographs of a 6-year-old type II Parry-Romberg syndrome patient (Table 2, Patient J). (Right) Post-surgical frontal photograph of the same patient, three years after initial hemifacial (left) free-fat inclusion surgery.*

Figure 3 – *(Left) Pre-surgical oblique photographs of the same patient as in Figure 2. A hyperchromic stain in the middle third of the face can be observed. (Right) Post free-fat inclusion surgery oblique photographs of the same patient showing a significant improvement in the facial atrophy and hyperchromic stain that covered the entire middle third of the child's face.*

Figure 4 – *(Left) Pre-surgical frontal photograph of a type III Parry-Romberg syndrome patient (Table 2, patient N). (Right) Post-surgical frontal photograph of the same patient after the following series of surgeries: free-fat graft, parietal bone grafts with rotation of the superficial temporal fascia flaps over the bone grafts, and combined orthognathic surgery and free-fat graft.*

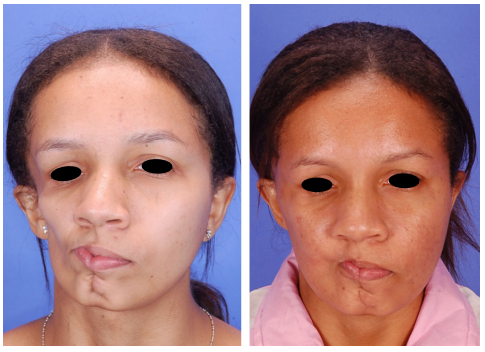

Figure 5 – *(Left) Pre-surgical oblique photograph of the same patient as in Figure 4. The severe facial atrophy can be observed. (Right) Post-surgical oblique photograph of the same patient showing significant improvement of the facial atrophy.*

DISCUSSION

PRS is a craniofacial deformity of unknown etiology, with a higher prevalence in women. It affects the face unilaterally in 95% of cases and usually has its onset during the first or second decades of life, with an active phase spanning 2–10 years before clinical stabilization[1,2].

Two Mexican[4,5] and one Chinese[6] classification systems have previously been used to grade the clinical severity of the disease. The grading system by Iñigo et al[4]. is based on the involvement of tissues in the region of the trigeminal nerve (dermatomes), whereas the classifications by Guerrerosantos et al.[5] and Hu et al.[6] are based uniquely on tissue involvement. In this study, we propose a severity scale related to the clinical progression of the disease, associated with anatomical involvement, independent of the affected dermatome.

The onset of PRS appears to be associated with atrophy of the epidermis, dermis, and subcutaneous tissue, with progressive involvement of the facial muscles followed by bony involvement[1,2]. Thus far, there have been no reports in the literature of bony involvement without disease in the surrounding soft tissues. Actually, a close relation between bone involvement and the severity of facial atrophy has been reported. Additionally, the restriction imposed by the deformed soft tissues leads to compromised bone growth[13]. Therefore, type III disease represents the progression of types I and II disease, and type II is the result of the progression of type I disease. These degrees of disease severity reflect the progression of the disease itself, which suggests that this classification is both logical and functional. In addition, this classification can be used to guide the therapeutic approach to patients with PRS.

It should be emphasized that this grading system was based solely on the analysis of 14 patients with PRS and is supported by other studies[4–6] with limited numbers of patients. As such, multicenter studies, including a larger number of patients, should be done so that the clinical progression of PRS can be better characterized, and potentially corroborate our findings.

Because PRS is usually associated with tridimensional craniofacial deformities, the complete restoration of facial symmetry is often difficult to achieve[3–6]. Furthermore, there is no consensus regarding the ideal surgical treatment for the wide spectrum of soft tissue and bone hypoplasia found in PRS, though several therapeutic options (free-fat grafts, dermal fat grafts, cartilage and bone grafts, muscle flaps, and alloplastic implants) have been described with the aim of increasing the facial volume lost to progressive atrophy[1,3–6,14–19].

The clinical phase, whether active or stable, is highly relevant in determining the optimal surgical treatment of PRS. There is no consensus in the literature regarding the ideal time for correcting the patient's deformities[8,18]. Traditionally, surgical procedures have only been performed after the stabilization of facial atrophy.[6,8,18] However, it should be noted that the time required for stabilization of the disease is variable and unpredictable[14,16], and children with PRS can develop psychosocial disturbances while awaiting surgery[14]. This is particularly true with regard to peer interactions in school for example, and may directly impact the patient's quality of life. In light of these findings, our group and others[14,16] have supported the view that craniofacial surgical interventions should be performed earlier, especially in those children with craniofacial deformities that, if left untreated, might lead to psychosocial impairments and learning disabilities. Early intervention may potentially improve or maintain physical capacities, levels of independence, and social relations at a critical time in the development of the patient[14].

In craniofacial surgery clinics, the protocols for treatment of craniofacial hypoplasias are guided primarily by facial growth and the functional needs of the patients[20]. In addition, a successful therapeutic approach requires individualized treatments employing reconstructive procedures at the soft tissue and bone level, performed at the appropriate time.[20] Taking these principles into consideration, the current therapeutic approach for PRS consists, in general, of either only one procedure or combined surgical procedures.[1,2] Our group and others[4–6,14,15,17] have treated each patient according to the degree of tissue atrophy[4–6,14,16] and also according to the age of the patient[14,15].

Recently, a therapeutic algorithm addressing hypoplasia of the soft tissues in PRS has been described. However, bone defects in these patients were not considered in the grading system[15]. In patients with type III disease an increase in soft tissue alone is not sufficient[18]. Therefore, we have favored a therapeutic strategy that combines the reconstruction of both soft tissue and bone: free-fat grafts for those patients with type I PRS, free-fat grafts or dermal fat grafts for type II PRS patients, and bone grafts with TPFF for type III PRS patients (Figure 6).

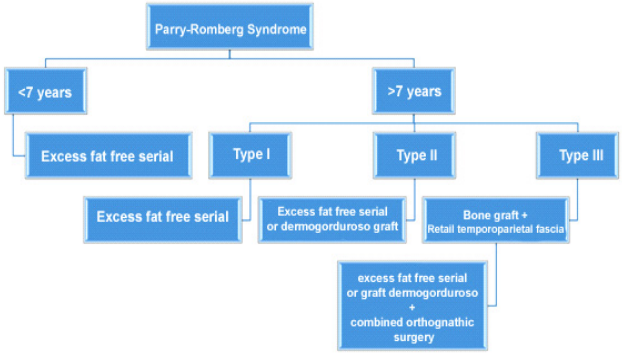

Figure 6. *Therapeutic algorithm for the surgical treatment of hypoplasia of the soft tissues and craniofacial bones in patients with Parry-Romberg syndrome based on clinical severity.*

In our hospital, as well as at the UCLA Craniofacial Clinic[14–16,20], the serial free-fat graft has been the first-choice method for the reconstruction of soft tissues in several craniofacial abnormalities, including PRS. Along with other authors[8,14,15,20], we prefer the free-fat graft instead of the injection of synthetic materials that more commonly lead to infection, seroma, exposure, and migration[14,15,20]. With regard to free-fat

grafts, it should be noted that adipose tissue is among the tissues with highest angiogenic capacity[21]. A series of studies have shown that free-fat grafts are associated with local neo-angiogenesis, which indicates that this intervention contributes to an improved vascularization of the receiving area. In PRS, compromised vascularization in the affected area may be the cause of soft tissue atrophy, and consequently, atrophy of the underlying bony structure[18]. Therefore, we believe that free-fat grafting can alter the prognosis of patients with type I PRS, as it may slow disease progression through improvement of local vascularization. Other investigators have also reported that the vascularized free flap can potentially act as a barrier against PRS progression[18]. Future studies should focus on the angiogenesis associated with free-fat grafts, as well as on their impact on PRS progression, perhaps employing Doppler scans to measure disease progression.

The variability in the survival rate of free-fat grafts has been reported as one of the main concerns with regard to this therapeutic approach[15,24]. We, along with others[8], have shown subjectively that patients with PRS absorb a certain degree of the grafted free-fat. However, no specific research on the level of free-fat absorption, or on the factors that may decrease absorption, have been done. Any information regarding the existence of fat absorption should be interpreted cautiously. One published study[16] investigated the rate of retention of the free-fat graft in the post-surgical period (1 year) and claimed that the "stickiness" of the free-fat graft is lower in PRS patients than in healthy patients without PRS. This diminished incorporation of the free-fat graft can be a consequence of both the poor vascularization at the receptor site, as well as other intrinsic characteristics of the PRS-affected region[16].

In addition, the age of the PRS patient can influence the survival of the free-fat grafts[16]. Laboratory studies[26,27] have shown that free-fat grafts are more effective in younger patients. Besides age, other factors have been reported to influence the absorption rate of free-fat grafts[26-28] including sedimentation rate, the nutritional status of the patient, and body mass index. However, there is no consensus on the actual impact of these variables on the absorption of free-fat grafts[22-28]. Hence, future research should focus on defining the roles of these different factors on the rate of incorporation of free-fat grafts. Meanwhile, the use of free-fat grafts, in particular in PRS patients, should be based on objective data, such as the volumetric tridimensional photogrammetric analysis of the faces of these patients. This type of analysis has demonstrated the improvement in symmetry and in the facial volume of patients one year after the free-fat injections, when compared with facial symmetry and facial volume prior to the intervention[16]. Furthermore, the impact of surgical interventions in the psychosocial context and quality of life of these patients should also be considered when choosing a therapeutic approach[14].

Assuming that some degree of fat absorption will occur in the post-surgical period,[8,15,16] we and others[8,14,16,27] prefer to over-correct the facial defect when transferring autogenic tissue. Xie et al[8]. have over-corrected by 20–30% of the total injected volume, as it has been reported that about 70–80% of the free-fat can survive following graft transplantation in PRS patients[8]. Along with others, we have used grafts with approximately 10% more volume than that required to achieve symmetry in the patient's face with the aim of attaining a higher survival rate of the fat and decreasing the degree of fat necrosis and the formation of palpable nodules[27]. In addition, the free-fat graft procedure can be performed several times without worsening the disease, based on the individual patient's requirements during the follow-up period[8,14-16].

Decreased blood circulation can be demonstrated in the atrophic facial tissues, specifically in type III patients. This may decrease the viability of the graft following surgery[6]. In these cases, we prefer TPFF, as this approach provides additional blood flow to the bone graft, as well as to fat grafts that may be placed in the future. In this series we did not observe clinically relevant reabsorption of any of the bone grafts associated with TPFF, which contrasts with the findings of other studies[14] that have used layering of bone grafts without additional blood flow. Our findings are in line with a study[18] that previously reported improved results by combining rib bone grafts with free dermal fascia flaps of the lateral intercostal artery perforator.

We prefer to use the TPFF procedure as this flap has several advantages such as a flexible outline, high vascularization with a wide pedicle rotation arch, anatomic proximity to structures in the face, and minimal morbidity of the donor site, among others[11,29], that facilitate and contribute to the symmetrical and harmonious reconstruction of the face. This procedure also assures adequate blood flow to the associated cranial graft without the need for microsurgical procedures that require additional skills[15].

There is no consensus in the literature concerning the age at which PRS patients should undergo surgical therapy. Many authors contend that correction of hypoplasia of the soft tissues should be performed only after treatment of the bony defects[14,16]. Meanwhile, we agree with others who have treated patients as early as possible, independent of the clinical severity of the disease[16]. Satisfaction associated with the results of surgery has been higher for the youngest PRS patients, even if more surgeries were required during the follow-up period[14]. It should be emphasized that before reaching seven years of age, patients have insufficient donor regions and the parietal bone is not thick enough to be divided[30]. Therefore, patients younger than seven years old are limited to serial free-fat grafting.

The maturity of the craniofacial bones is also relevant in the decision-making process using a different therapeutic algorithm[15], since, as previously mentioned, only the reconstruction of soft tissue defects has been graded[15]. As the bones of patients with type III disease become involved, malocclusion can occur as the craniofacial bones stop growing. These patients may have skeletal open-bite malocclusion as a consequence of progressive unilateral atrophy of the jaw. Therefore, coordinated planning of these procedures by the plastic surgeon and the orthodontic surgeon is paramount. According to our experience and that of others[6,14-16], such patients can be considered candidates for combined orthognathic surgeries with or without osteodistraction, aimed at decreasing facial

asymmetry, and may be followed by augmentation of the soft tissues with the use of serial free-fat grafts.

Our study, similar to previous studies[8], evaluated the results of craniofacial surgical procedures subjectively with the use of a previously described scale in PRS patients[8]. All patients had an overall improvement in facial appearance. However, a carefully conducted evaluation showed residual facial asymmetry and additional interventions were likely required, as showed in Figures 4 and 5. In addition, and despite the partially satisfactory results achieved, the analysis method[8] used has limitations, as it is based on a static characterization of a dynamic disease and therapeutic interventions that change over time (i.e., the absorption of grafts or flaps). Follow-up continues for all of the PRS patients reported here and new surgical approaches will be adopted in the future depending on the individual needs of each patient. Future studies using objective analytic methods, such as photogrammetric volumetric quantification or tomography, are expected to help elucidate the nature of PRS and the optimal therapy for this condition.

CONCLUSION

In this retrospective study, the therapeutic approach to patients with PRS was guided by the graded severity of the craniofacial deformation. The scale for grading severity proposed here describes the clinical stages of the disease and contributes to the decision-making process when considering the different therapeutic approaches in PRS patients.

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