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# Managing Soft Tissue Defects in Parry-Romberg Syndrome: An Individualized Approach

Alona D. Salita, DO*
Matthew K. Shaheen, MD*†
Arvind U. Gowda, MD†
Konstantinos Kouzounis, MS3‡
Suphalerk Lohasammakul, MD†§
Kongkrit Chaiyasate, MD†

**Background:** Parry-Romberg syndrome (PRS) is a rare condition characterized by a progressive shrinkage and degeneration of the tissues beneath the skin, usually on one side of the face. Managing this disease presents numerous challenges due to its heterogeneity and unpredictable outcomes. The existing literature is primarily composed of case reports and series, leading to a lack of comprehensive guidelines on surgical intervention for the various manifestations of PRS. We propose an approach to address these challenges and optimize surgical outcomes.

**Methods:** We conducted a retrospective review of all patients who underwent surgical correction for PRS between 2012 and 2022. Surgical interventions were determined based on the location and severity of the facial defect. The revision procedures were tailored to each patient until they were satisfied with the results.

**Results:** Eleven patients underwent surgical correction, with an average of 3.2 procedures per patient. Fat grafting or dermal fat grafting was sufficiently effective for mild deficits in all areas and for upper-third deficits regardless of severity. For moderate to severe defects in the mid and lower face, a combination of buried free flaps and fat grafting yielded satisfactory results. Upon final revision, all patients rated their results as satisfactory or excellent.

**Conclusions:** We propose an approach to surgical management that takes into account the specific deficits of each patient. Our approach has proven to yield aesthetically pleasing and reliable results, aligning with findings in the existing literature. This method could provide a foundation for standardized guidelines and improve the prognosis for individuals with PRS. *(Plast Reconstr Surg Glob Open 2024; 12:e6043; doi: 10.1097/GOX.0000000000006043; Published online 8 August 2024.)*

## INTRODUCTION

Parry-Romberg syndrome (PRS), also known as progressive hemifacial atrophy, is an idiopathic inflammatory disease affecting facial soft tissues, with occasional involvement of underlying bone and cartilage structures.[1] PRS typically manifests during the first two decades of life and exhibits a higher prevalence in women. Some cases involve multiple branches of the trigeminal nerve,[2–4] and although it primarily affects soft tissues, reports of bony changes exist.[5] The disease often leads to pronounced aesthetic deformities, accompanied by psychosocial challenges. The etiology of PRS remains a subject of debate, with proposed theories ranging from trophic fiber dysfunction and immune responses to trauma and infectious causes.[6,7] Skin biopsy findings typically reveal lymphocytic dermal infiltration with signs of fat atrophy, sclerosis, and perivascular plasma cells.[2] The variability in clinical presentation and disease progression necessitates diverse reconstructive strategies. Current treatment options include soft tissue fillers, autologous fat grafting, dermal fat grafts, synthetic implants, and combinations of these modalities.[4,8–18] However, much of the existing literature consists of case studies, leaving a gap in systematic treatment recommendations. Treatment decisions must consider volumetric, aesthetic, and functional deficits.[9] This article outlines a treatment approach developed by the

*From *Department of Plastic and Reconstructive Surgery, Corewell Health East Farmington Hills Hospital, Farmington Hills, Mich.; †Department of Plastic and Reconstructive Surgery, Corewell Health East William Beaumont University Hospital, Royal Oak, Mich.; ‡School of Medicine, St. George's University, Grenada, West Indies; and §Department of Anatomy, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok, Thailand.*

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senior author, supported by case examples illustrating the complexities of PRS management.

## METHODS

In adherence to the Declaration of Helsinki guidelines, we obtained institutional review board approval for this study. We conducted a retrospective analysis of clinical records of patients who underwent surgical management for PRS between 2012 and 2022 under the care of the senior author. We categorized facial defects as upper, middle, or lower face and classified volume deficits as mild, moderate, or severe. Postoperative assessments included evaluations of patient and surgeon satisfaction, taking into account symmetry and aesthetic outcomes. Patients were informed preoperatively about the potential need for follow-up surgery. Regardless of the surgeon's satisfaction, revision procedures were offered until the patient achieved contentment.

## RESULTS

During the study period, 11 patients underwent surgical treatment, with their specific procedures detailed in Table 1. Among these patients, four are highlighted below. Preoperative assessments categorized two patients as having mild deficits, five with moderate deficits, and four with severe deficits. An additional four patients with mild deficits either declined surgery or are awaiting fat grafting. Six patients, including all four with severe deficits and two with moderate, underwent free tissue transfer. Patient 4 required maxillomandibular deficiency correction before addressing soft tissue deficits. Except for patient 6, all patients received fat grafting. On average, patients underwent 3.2 surgical procedures, including those performed by other

### Takeaways

**Question:** Parry-Romberg Syndrome has a remarkably wide range of presentations. How can we organize the management of this disease in a systematic way that addresses its various presentations?

**Findings:** We present the results of the senior surgeon using an approach to organize patient deficits and guide the choice in surgical management.

**Meaning:** Surgical management of Parry-Romberg Syndrome can be daunting; however, we have demonstrated consistently satisfactory results using a simple approach that we hope can guide other surgeons.

**Table 1. Patient Characteristics and Operative Details**

| | Age* | Sex | Facial Thirds | Severity | Procedures Performed |
|---|---|---|---|---|---|
| 1 | 9 | M | Upper/mid | Severe | 1. Right circumflex scapular free flap<br>2. Suture removal under anesthesia, revision of incision<br>3. Flap debulking, fat grafting, dermal fat graft to upper third<br>4. Flap debulking, periosteal release of brow and forehead, fat grafting<br>(5. Le Fort 1 osteotomy, iliac crest bone graft to maxilla) |
| 2 | 14 | M | Upper/mid | Mild (linear scleroderma) | 1. Dermal fat grafts to nose and forehead<br>2. Allograft dermal matrix graft to nose and forehead<br>3. Fat grafting to nose and forehead |
| 3 | 47 | F | Mid/lower | Moderate | 1. Left circumflex scapular free flap, full-thickness skin graft<br>2. Fat grafting, flap readvancement, excision of skin graft<br>3. Fat grafting |
| 4 | 16 | F | Mid/lower | Moderate | (1. Le Fort 1 and sagittal split ramus osteotomies, iliac crest bone graft to left maxilla)<br>2. Dermal fat graft and fat grafting left mid face and upper lip<br>3. Dermal fat graft and fat grafting left mid face and upper lip |
| 5 | 24 | F | Mid/lower | Severe | 1. Right circumflex scapular free flap<br>2. Flap debulking, dermal fat graft to chin<br>3. Flap debulking, flap readvancement, dermal fat graft midface and chin<br>4. Flap debulking, fat graft midface<br>5. Flap readvancement, fat grafting cheek, and chin |
| 6 | 65 | F | Mid/lower | Moderate | 1. Excision silicone granulomas right lower face and neck, Z-plasty contracture release neck<br>2. Revision local tissue rearrangement for neck contracture<br>3. Excision silicone granulomas neck, revision local tissue rearrangement neck contracture |
| 7 | 54 | F | Mid | Moderate | 1. Left mid face fat grafting, excision and local flap right face wound |
| 8 | 11 | F | Upper/mid/lower | Severe | (1. Right Scapular Free flap)<br>(2. Unknown revision)<br>(3. Unknown revision)<br>(4. Unknown revision)<br>5. Debulking of right scapular flap and fat grafting |
| 9 | 40 | M | Mid/lower | Moderate | 1. Right circumflex scapular free flap<br>2. Flap debulking, readvancement, dermal fat graft to chin and lower lid<br>3. Flap debulking, readvancement, fat graft to mid face and upper lip |
| 10 | 37 | F | Upper/mid | Mild | 1. Fat grafting forehead, nose, cheek, upper lip<br>2. Rib cartilage graft to nose, fat grafting forehead, cheek, and upper lip |
| 11 | 21 | F | Mid/lower | Severe | 1. Left circumflex scapular free flap<br>2. Readvancement of flap and debulking, fat grafting to forehead, midface, and chin |

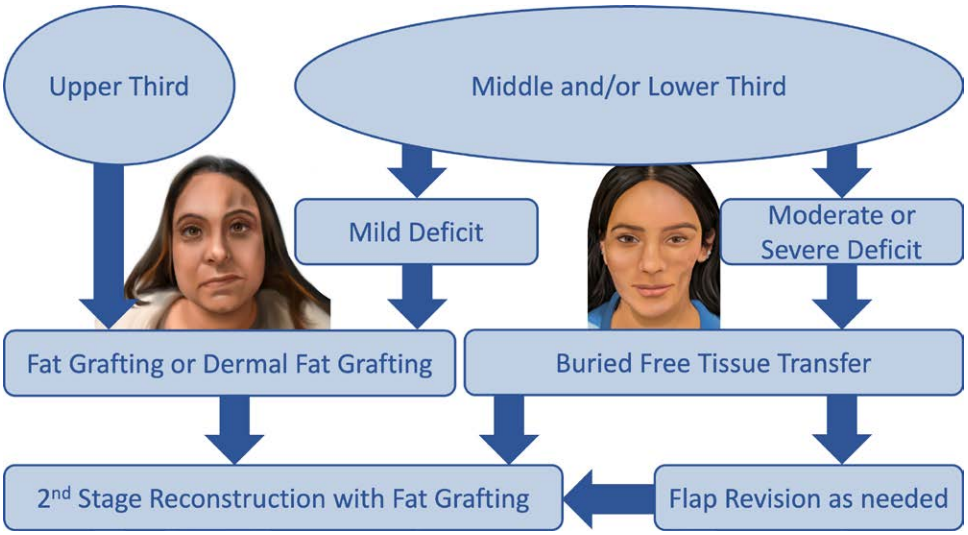


**Fig. 1.** Approach for reconstruction based on severity and involvement of the facial regions.

surgeons. Following final revisions, both surgeons and patients rated all outcomes as satisfactory or excellent. However, patients 5 and 11, initially content with their outcomes, sought further surgical revisions during their latest checkups. No significant complications were noted at surgical or donor sites.

## DISCUSSION

Our proposed approach divides the face into thirds and employs the severity of soft tissue deficits to guide the initial surgical plan, categorizing defects as mild, moderate, or severe. Surgeons must exercise their aesthetic judgment by considering both individual facial subunits and overall facial harmony when assessing defect severity. This assessment inherently involves subjectivity. Figure 1 provides an overview of our recommendations for the initial surgery.

For mild volume deficits across the facial areas, fat grafting or dermal fat grafting usually yield favorable results. Although the overcorrection principle has been successful in some cases, its challenges lie in the inability to precisely control overcorrection and the unpredictable extent of graft atrophy postoperatively.[18] These factors hinder consistent replication of favorable outcomes, particularly in cases with extensive involvement. Therefore, we recommend restricting the application of the overcorrection principle to mild volume deficits. In the upper facial third, deficits of any severity seem amenable to these interventions, as elaborated in case 1 below. The decision is influenced by the variation in soft tissue thickness across facial regions, with the upper face being the thinnest and less likely to require substantial tissue reconstruction. Dermal fat grafting offers distinct advantages, especially when addressing conditions like linear scleroderma.

Dermal fat grafting involves creating a dermal site through a small incision along the hairline or within a preexisting scar. A subcutaneous tunnel is meticulously formed in the target area. The dermal fat graft is harvested from the hip region, resulting in a scar located within the underwear area. This harvested skin comprises both the dermal layer and a thin layer of superficial fat tissue, making it suitable for transplantation to the recipient site. The dermal fat graft is then inserted through the tunnel, and the skin incision is closed using standard techniques. Regarding fat grafting, we use a 17-gauge blunt cannula connected to a 60-mL Luer-Lok syringe with a syringe snap lock for fat graft harvesting. The harvested fat tissue is subsequently transferred into multiple 1-mL syringes for precise injection into the defect site.

For moderate to severe deficits in the midface, lower face, or continuous defects spanning all three regions, we recommend buried free tissue transfer to provide the necessary bulk. This approach has demonstrated greater reliability than relying solely on fat grafting. However, it should not be pursued for patients with an unknown final soft tissue deficit (cases 4 and 6 in Table 1) or for those who have declined free flap surgery (case 7 in Table 1). When formulating surgical strategies, it is essential to align with patient expectations, keeping in mind the evolving nature of results often necessitating multiple procedures. The senior author advocates for a buried free flap based on the circumflex scapular artery, which harnesses both scapular and parascapular flaps via a single pedicle. This methodology, including flap insetting, aligns with previously documented techniques.[10] The circumflex scapular flap is considered the preferred option for facial augmentation due to its ample vascular pedicle, vessel diameter, significant soft tissue volume, and minimal donor site complications.[3,5,8,10,12] In comparison with other fasciocutaneous flap options such as anterolateral thigh, groin, and deep inferior epigastric artery perforator (DIEP) free flaps, previous reports have discussed their respective applications.[19] Considerations for choosing between anterolateral and groin flaps are based on defects with significant depth

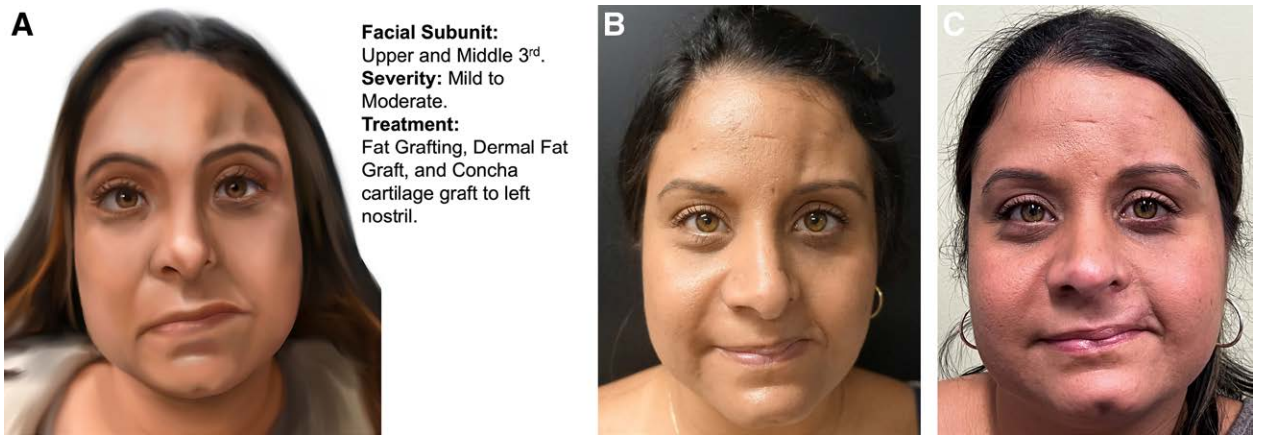


**Fig. 2.** Case 1. A, Illustration of defect analysis and surgical treatments. B, Preoperative picture. C, Postoperative result at 1 year.

and relatively small dimensions, whereas the DIEP flap is preferred for defects requiring a larger volume. The primary criterion for evaluating these flap options is their ability to effectively conceal donor site scars, making groin and DIEP free flaps the two most viable choices. Given the limitations of the groin flap, including a short pedicle and small diameter, the senior author's perspective favors the DIEP free flap as an alternative. Notably, the circumflex scapular artery remains the preferred free flap choice due to its consistent anatomical features, including transverse and descending branches, which enable versatile customization of the flap to accommodate various facial regions, and, if necessary, bone harvesting. It is also used in reconstructing head and neck tumors and managing hemifacial microsomia.[13,14] However, like other free flap-based reconstructions, potential postoperative issues can arise, such as partial flap loss, excessive bulk, atrophy, seromas, hematomas, and dehiscence.[11] Regardless of the primary surgical procedure, we recommend prescheduled follow-up procedures, predominantly involving fat grafting, to address potential contour irregularities. This proactive approach aids in managing patient concerns regarding early postoperative appearance. For those who have undergone free flap procedures, adjustments can be made concurrently as needed. Common reasons for flap revisions include excessive flap volume, resorption, or the need to readjust flap placement.[15]

It is important to acknowledge that PRS is a rare condition, and the approach presented in this study is derived from a retrospective review and the collective experience of the senior author. While this approach can serve as a preliminary guide for patient selection, its validation requires further investigation involving a larger and more diverse patient population in future studies.

## ILLUSTRATIVE CASES BASED ON THE APPROACH

**Case 1: Mild Upper and Midface Deficit (Case 10 in Table 1)**

A 37-year-old woman presented with mild left upper and midface deficits. Given the minimal soft tissue atrophy, we opted for fat grafting. Fat grafting outcomes can vary, with estimated take rates between 30% and 70%.[16,17] As recommended, we overfilled by approximately 25%.[18] Research suggests that most PRS patients require a volume of just 5–17 mL in the upper third.[16] Synthetic implants were not considered due to the lesser volume needed, unless a significant bony deficiency was noted. We emphasized the potential need for multiple fat grafting sessions. Figure 2 displays her pre- and postoperative appearances.

**Case 2: Severe Mid and Lower Face Deficit (Case 11 in Table 1)**

A 21-year-old woman presented with pronounced deficits in her left mid and lower face. We used a circumflex scapular free flap to address the extensive soft tissue deficiency. After designing the flap size using a template of the defect, a fasciocutaneous flap was formed and de-epithelialized. This flap was tucked under her native skin using a rhytidectomy flap and a parachuting method with transcutaneous sutures. Subsequent procedures included flap debulking, fat grafting, and a scheduled third surgery. The technique, including preoperative, intraoperative, and postoperative images, is depicted in Figures 3 and 4.

**Case 3: Postoperative Flap Ptosis Revision (Case 9 in Table 1)**

A 40-year-old man with moderate left mid and lower face deficits underwent a flap procedure similar to that in case 2. However, his flap later exhibited significant ptosis due to tissue weight, leading to asymmetry and mild scleral show. The patient subsequently received a flap revision, flap resuspension, and direct excision. Regarding the resuspension, the flap was entirely devoid of its epithelial layer and carefully inserted alongside the adjacent periosteum or deep fascia. Figure 5 illustrates how a substantial flap may evolve, with Figure 5B suggesting undercorrection during the initial procedure. A second revision yielded a satisfactory outcome. It should be noted that flap ptosis is a common issue that can arise over the long term. In our initial procedure, we use a technique where the flap is securely anchored to either the SMAS layer or periosteum using polydioxanone. Despite our best efforts with


**Fig. 3.** Case 2. Surgical techniques of circumflex scapular free flap. A–B, Preoperative defect marking and flap design. C–D, Intraoperative flap insetting and flap harvest.

this method, some degree of flap ptosis may still occur. The decision to undergo revision surgery is ultimately a patient's choice because they must determine whether the degree of ptosis warrants further intervention.

**Case 4: Addressing Pediatric Patient Growth (Case 1 in Table 1)**

PRS manifestation in adolescence presents challenges, requiring anticipation of facial growth changes. Such patients often necessitate monitoring until adulthood and potential multiple revisions. A notable case in our series involved a boy who exhibited severe hemifacial atrophy symptoms at age 5. To address school teasing, he underwent a procedure at age 9. The insetting of the free flap involved both deficits of the upper face and midface. As he matured, he displayed asymmetrical bone growth, leading to further surgery 6 years postoperative. His most recent checkup revealed satisfaction despite minor persistent

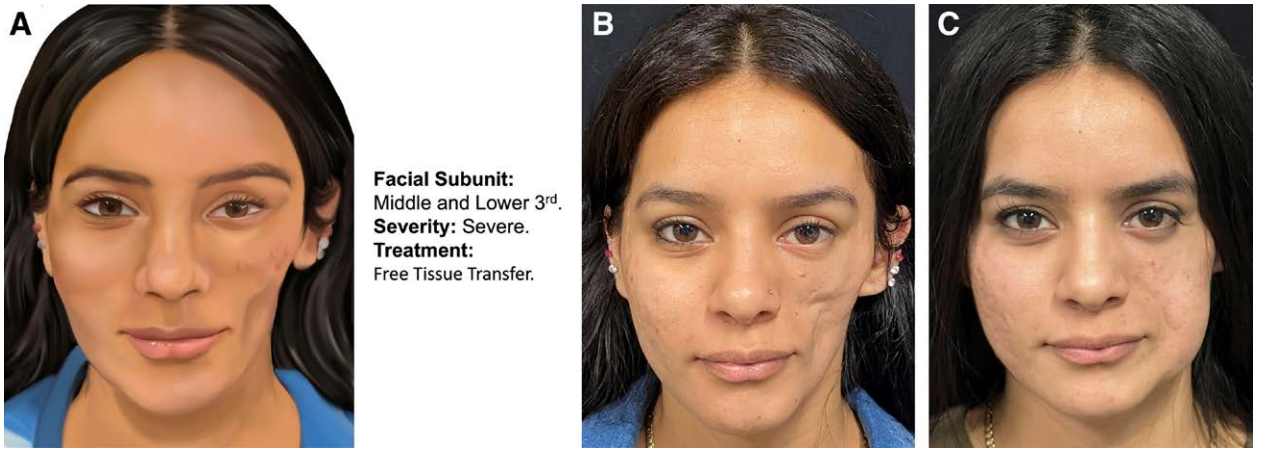


**Fig. 4.** Case 2. A, Illustration of defect analysis and surgical treatments. B, Preoperative picture. C, Postoperative result at 1 year.


**Fig. 5.** Case 3. A, Preoperative picture. A possible evolvement of a free flap from initially undercorrected appearance (B) to ptotic look required revision surgery (C).

irregularities. Figure 6 showcases his preoperative and final postoperative appearances.

## CONCLUSIONS

This article aims to provide surgeons with a roadmap for addressing PRS. Recognizing the diverse nature of PRS, it is crucial to tailor treatments to each patient. Our outlined approach offers a foundational approach to clinical decision-making, aligning with existing literature on PRS treatment. Patient outcomes reinforce the aesthetic success of these recommendations. Notably, this study's limitation lies in the absence of objective metrics to evaluate the severity of volume deficits and progress extent. Subsequent research may delve into these facets more extensively. Nonetheless, gauging patient satisfaction remains pivotal in facial aesthetic contexts.

***Kongkrit Chaiyasate, MD***
3555 W 13 Mile Road, Suite N120
Royal Oak, MI
E-mail: kongkrit.chaiyasate@corewellhealth.org

## DISCLOSURE

*The authors have no financial interest to declare in relation to the content of this article.*

## PATIENT CONSENT

*The patients provided written consent for the use of their images.*

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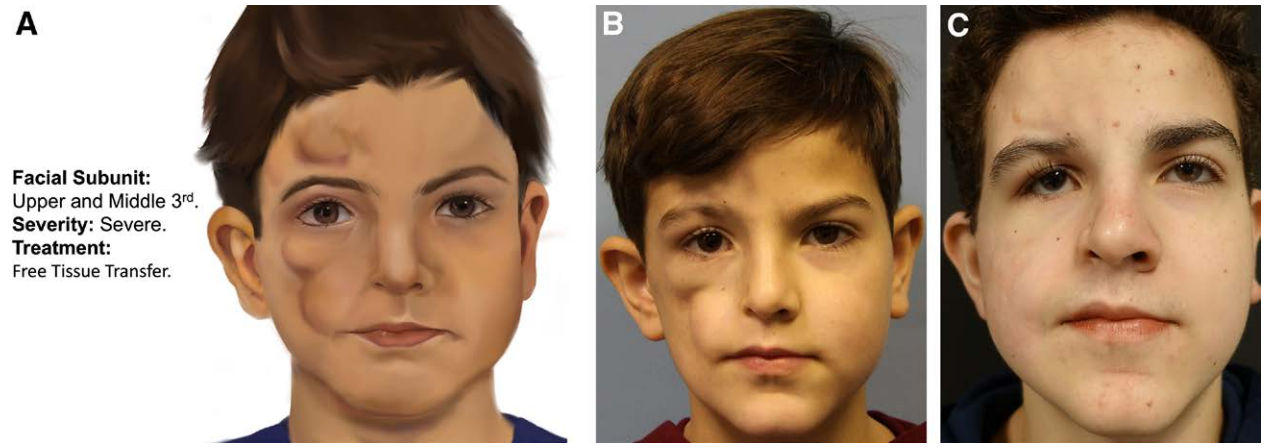


**Fig. 6.** Case 4. A, Illustration of defect analysis and surgical treatments. B, Preoperative picture. C, postoperative result at 9 years.


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