En Coup de Sabre Scleroderma and Parry-Romberg Syndrome in Adolescents: Surgical Options and Patient-related Outcomes

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ABSTRACT. ***Objective.*** There is little information regarding surgical options and outcomes in patients with facial localized scleroderma. We evaluated the surgical outcomes of procedures performed for linear scleroderma of the face in the pediatric age group; and assessed psychosocial effects of surgical interventions on the affected children.

Methods. A retrospective chart review was performed of children with en coup de sabre scleroderma (ECDS) and Parry-Romberg syndrome (PRS) who underwent surgical intervention; this included demographic data, clinical features, and type of surgical interventions. A questionnaire of 13 questions covering 4 domains (physical, emotional, social, and symptoms) was sent to patients who consented to take part in the survey. Surgical treatments and outcomes were analyzed retrospectively.

Results. Seventeen patients underwent surgical intervention (autologous fat injections, Medpor implants, bone paste cranioplasty, and free groin flap) to correct facial asymmetry. Ten patients answered the questionnaire (58.8% response rate). Unhappiness with their appearance, loss of confidence, and bullying were cited as reasons for surgery. The appearance subscale of the survey demonstrated the lowest standardized scores and greatest negative effect on the patients' quality of life compared to the 3 other subscales. All subjects would consider another surgery and would recommend surgery to other patients with ECDS and PRS.

Conclusion. Surgical treatment is a potential useful intervention in children with facial disfigurement. Prospective data are needed. (J Rheumatol First Release Sept 15 2010; doi:10.3899/jrheum.100062)

Key Indexing Terms:
MORPHEA LOCALIZED SCLERODERMA EN COUP DE SABRE SCLERODERMA
PARRY-ROMBERG SYNDROME AUTOLOGOUS FAT INJECTION MEDPOR IMPLANT
BONE PASTE CRANIOPLASTY FREE GROIN FLAP

En coup de sabre scleroderma (ECDS) and Parry-Romberg syndrome (PRS; also known as progressive hemifacial atrophy) are 2 uncommon disorders in the morphea group of diseases. These 2 variants of facial morphea may lead to significant facial deformity. ECDS is generally located in the paramedian forehead with extension into the frontoparietal scalp and is associated with significant cutaneous sclerosis and alopecia. PRS, on the other hand, is characterized by progressive subcutaneous atrophy without skin or scalp involvement. Given that they affect the face primarily, these disorders may have a profound psychosocial impact. However, there are no current studies investigating the influence of these 2 disfiguring disorders on the patient's quality of life and the benefit that surgical treatment may afford.

Studies in the past explored different surgical options for facial morphea[1,2,3,4,5,6,7,8,9]. The primary objective of these reconstructive techniques is volume augmentation. The determination of treatment success was based on physicians' clinical judgment and qualitative measurement of patient satisfaction. However, to date there has been no quantitative measure by which to assess the surgical outcomes of facial reconstructive intervention in terms of patient satisfaction, symptomatic relief, complications, and longterm effects.

The objectives of our study were to provide insight into the benefit of surgical interventions in patients with facial

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morphea and to evaluate the psychosocial effects of surgical interventions on pediatric patients with facial morphea.

MATERIALS AND METHODS

This study was conducted at The Hospital for Sick Children, Toronto, Canada, and was approved by the Institutional Research Ethics Board. Patients were recruited from the specialized Morphea Clinic, a multidisciplinary clinic consisting of a pediatric dermatologist, pediatric rheumatologist, pediatric nurse and occupational therapist, and a pediatric plastic surgeon. All patients followed in the Morphea Clinic who underwent facial plastic surgery for facial morphea from 2000 to 2008 were retrospectively identified. The extracted data consisted of sex, age at diagnosis, type of morphea, age at surgical intervention, type of intervention, surgical outcome, need for repeat surgery, and interval between surgeries. Pre and post digital photographs were compared to corroborate the patient's opinion of improvement.

A questionnaire (Table 1) was adapted for use in this patient population that was patterned after 3 validated instruments: the Skin Cancer Index[10], the Facial Plastic Surgery Outcomes Evaluation[11], and the Nasal Obstructive Symptoms Evaluation scale[12]. It consisted of 13 questions covering 4 domains (appearance, social, emotional, and symptoms). The physical or appearance subscale focuses on the issues of scarring, disfigurement, and self-image perception. The social and emotional subscales address issues related to the psychological and interpersonal effects on patients. The symptoms subscale addresses physical and functional deficits and perceived complications of surgery. The questionnaire was sent to patients who consented to participate in the study. A 5-point response format was used to measure patients' responses, 1 representing "very much" to 5 "not at all." Subscale scores were obtained by adding the individual items and dividing by the number of items on the subscale. The total score was computed by summing the subscale scores and dividing by 4. Higher scores indicate patient satisfaction after surgery with less impairment of the quality of life.

RESULTS

Seventeen patients underwent surgical intervention to correct facial deformity resulting from facial morphea, 12 (71%) with ECDS and 5 (29%) with PRS. Of these 17 patients, 11 were female and 6 were male (F:M ratio 1.8:1). The mean age at onset of symptoms was 7.38 (SD 3.54) years and mean age at diagnosis was 8.86 (SD 3.01) years. The mean time from onset to diagnosis was 1.48 years. The mean age at first surgery was 15 (SD 2.22) years, with a 6.14 year mean time from diagnosis to first surgery.

Six patients (35%) had a single surgical procedure, while 11 (65%) had multiple surgical interventions (Table 2, Figure 1 and Figure 2). Only one patient sustained an adverse event related to a surgical procedure, consisting of fat necrosis 2 weeks after undergoing autologous fat injection, requiring incision and drainage. One patient (5.88%) underwent debulking of free groin flap due to volume overcorrection.

Ten of the 17 patients completed the survey (59% response rate). All responders (10/10) had been unhappy with their appearance prior to surgery, prompting them to undergo surgical intervention. Eight subjects (80%) stated loss of confidence, one (10%) was unhappy with facial discoloration, while one (10%) stated bullying as an indication for surgery. Table 3 summarizes the survey results. The appearance subscale demonstrated the lowest standardized scores and possibly the greatest negative effect on the patients' quality of life compared to the 3 other subscales. The symptoms subscale demonstrated the highest standardized scores.

One subject (10%) was "extremely satisfied" with surgery, 4/10 (40%) were "very satisfied" with surgical results, 3/10 (30%) were "somewhat satisfied," 1/10 (10%) was "not too satisfied," and 1/10 (10%) was "not at all satisfied" with the results. However, given the same indications and known outcomes, all subjects stated that they would consider another surgical intervention and 9/10 (90%) would recommend surgery to other patients with ECDS and PRS.

DISCUSSION

En coup de sabre scleroderma and Parry-Romberg syndrome are 2 relatively rare forms of facial morphea that result in loss of facial contour and symmetry, requiring surgical intervention. Our study is the first to describe the surgical options and the outcomes of surgical interventions in patients with facial morphea.

Volume restoration was the main objective of the reconstructive techniques, with the majority of patients undergoing multiple procedures to correct facial asymmetry. The most common volume-augmenting techniques in our cohort were autologous fat injections, Medpor implant, bone paste cranioplasty, and free groin flap. Autologous fat injections and biomaterials are generally used for small to moderate defects. When large-volume correction is necessary, free groin flap and bone paste cranioplasty are the surgical options.

Lipoaspirated fat injection was most commonly employed because it is a relatively safe and easy procedure, with minimal complications. One of our patients had fat necrosis requiring incision and drainage. Because of its temporary nature due to fat resorption, most patients had to undergo multiple procedures.

The second most common augmenting technique used an alloplastic material, the Medpor implant. The advantage of a biomaterial like Medpor is that it allows correction of facial contour defects with a one-stage minimally invasive procedure. Medpor implants are fixed with titanium miniscrews, and thus migration is not a problem. Its potential disadvantages relate to the fact that there is a foreign material, with potential for rejection, instability over time, and migration[1,2].

The free groin flap was used for volume augmentation in 2/17 of our patients. The advantages of free flaps in general are their good structural strength, permitting adequate adjustment of the defect[3] and their longterm functional and morphological result[1]. The specific advantage of the groin flap for facial recontouring is its less noticeable donor-site defect and lower morbidity[4]. The major disadvantage is the effect of gravity on the position of the flap, resulting in sag-

Table 1. A questionnaire was adapted for use in this patient population, patterned after 3 validated instruments: the Skin Cancer Index, the Facial Plastic Surgery Outcomes Evaluation, and the Nasal Obstructive Symptoms Evaluation scale.

Patient's initials: _____
Sex: _____
Age: _____
Clinical Diagnosis: _____

Which of the following are your reasons for undergoing surgery?

___ Unhappy with appearance
___ Unhappy with color change
___ Loss of self-confidence
___ Bullying
___ Headaches
___ Eye problems
___ Others: _____

During the past year, how much have you….	Very much	Quite a bit	Moderately	A little bit	Not at all
1. Worried about the appearance of your face	o	o	o	o	o
2. Been concerned about how LS affects your attractiveness	o	o	o	o	o
3. Thought about how your appearance can still be better	o	o	o	o	o
4. Considered surgery to alter the appearance of your face	o	o	o	o	o
5. Felt uncomfortable meeting new people	o	o	o	o	o
6. Been bothered by people's questions or stares	o	o	o	o	o
7. Avoided social interactions	o	o	o	o	o
8. Felt anxious about your linear LS	o	o	o	o	o
9. Felt frustrated about your linear LS	o	o	o	o	o
10. Worried about facial asymmetry progressing in the future	o	o	o	o	o
11. Been concerned about skin changes (hyperpigmentation, sclerosis, atrophy)	o	o	o	o	o
12. Been bothered by headaches or eye problems	o	o	o	o	o
13. Been bothered by any complication of surgery	o	o	o	o	o

Overall, how satisfied are you with your surgery?

___ Extremely satisfied
___ Very satisfied
___ Somewhat satisfied
___ Not too satisfied
___ Not at all satisfied

Would you consider another surgery in the future?

___ Yes
___ No

Would you recommend this surgery to other patients with facial linear scleroderma?

___ Yes
___ No

ging. Another disadvantage is the macroscopic increase in volume during growth and with weight gain[1]. One of our patients in fact had to undergo debulking of the graft due to volume overcorrection.

Bone paste cranioplasty was performed in 4/17 patients. This procedure is performed for patients with severe facial tissue depression in which the soft tissues are so atrophied that the skin is practically next to the bone[5], or for patients

Table 2. Patient characteristics, diagnosis, and type of surgical intervention.

Patient	Sex	Age at Onset, yrs	Age at Diagnosis, yrs	Age at Surgery, yrs	Present Age, yrs	Diagnosis	Medical Intervention	Surgical intervention	Repeat Surgery
1	M	14	15	17	18	ECDS	Calcipotriol	Fat injection × 1	No
2	F	10	12	16	19	ECDS	MTX, pulse steroids	Fat injection × 3	Yes
3	F		7	13	17	ECDS	Calcipotriol	Bone paste cranioplasty, Medpor implant, fat injection 3×, I & D of fat necrosis	Yes
4	F		5	15	20	ECDS	MTX, oral steroids	Free groin flap, debulking of flap, facelift, Medpor implant ×2, fat injection ×1	Yes
5	F		10	16	19	ECDS	Topical steroids	Free groin flap, scar revision, fat injection ×4, bone paste cranioplasty, facelift	Yes
6	M			19	22	PRS		Fat injection ×1	No
7	M	4	4	13	17	ECDS		Medpor implant ×2, fat injection ×2	Yes
8	F		9	19	22	ECDS	Calcipotriol	Medpor implant, scar revision, fat injection ×1	Yes
9	F		10	14	20	ECDS	MTX	Fat injection ×3	Yes
10	M				21	PRS		Bone paste, canthoplasty, Medpor implant, fat injection ×1, bimaxillary surgery, Le Fort 1	Yes
11	M	8	8	13	15	PRS		Fat injection ×2	Yes
12	F	5	7	14	16	ECDS	MTX, oral steroids	Medpor implant	No
13	F	5	8	16	18	ECDS	Vit A cream, MTX	Medpor implant, fat injection ×1	No
14	F		7	15	19	PRS		Medpor implant, fat injection ×3, rhinoplasty	Yes
15	F	4	9	13	14	ECDS	MTX, oral steroid, MMF, IVIG	Rhinoplasty	No
16	M	9	13	16	20	ECDS	Vit A cream	Scar revision	No
17	F			11	20	PRS		Fat injection ×2, canthoplasty, bone paste cranioplasty, rhinoplasty ×3, scar revision, brow suspension	Yes
Mean (SD)		7.38 (3.54)	8.86 (3.01)	15 (2.22)	18.65 (2.29)				

ECDS: en coup de sabre scleroderma; PRS: Parry-Romberg syndrome; MTX: methotrexate; Vit: vitamin; MMF: mycophenolate mofetil; IVIG: intravenous immunoglobulins; I & D: incision and drainage.

with far-advanced facial morphea characterized by bony atrophy.

To enhance the results of these volume-augmenting surgeries, our patients underwent various procedures to address functional deficits and to improve cosmesis. These additional procedures include excision of alopecic or scar tissue,

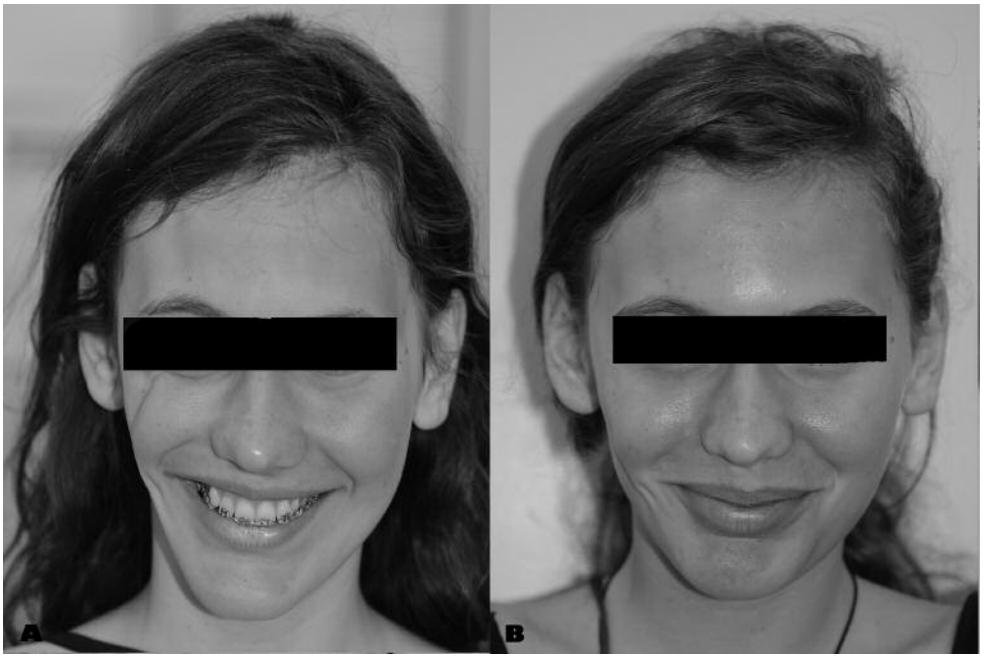

Figure 1. A patient who underwent Medpor right zygoma, fat injection cheek and forehead (75 cc, 5-month interval), before (A) and after (B) the procedure.

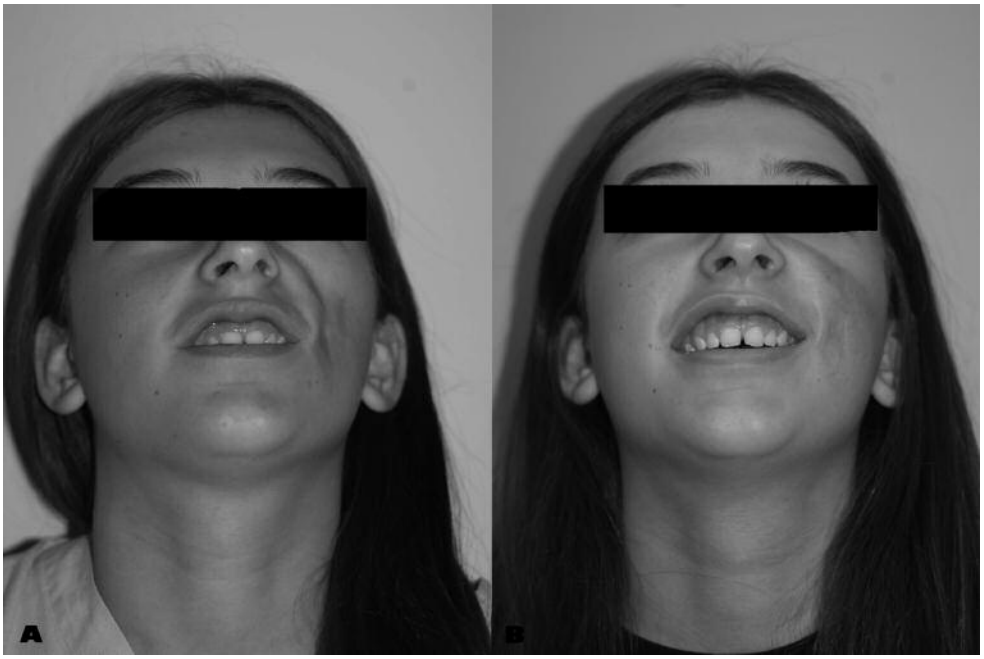

Figure 2. A patient who underwent Medpor left zygoma and fat injection cheek (25 cc, 5-month interval) before (A) and after (B) the procedure.

nasal reconstruction or rhinoplasty, lateral canthoplasty, facelift, bimaxillary surgery, Le Fort 1 osteotomy, and brow suspension.

Our study also assessed the psychosocial status of these patients after surgical treatment. A previous study from this institution revealed that, despite the potentially disfiguring effects of morphea, patients had normal self-perception[13]. It did not, however, investigate differences occurring between disease subtypes, and only 14 patients (mean age 9.6 yrs) with facial morphea were included. In our current study,

Table 3. Summary of the quality of life survey data.

Question	Quality of Life Factor	Unstandardized Raw Score*, mean (SD)	Standardized Score** (0–100), mean (SD)
1	Worried about appearance	2.8 (1.03)	45 (25.82)
2	Concerned about attractiveness	2.8 (1.55)	45 (38.73)
3	Concerned about how appearance can improve	2.6 (1.35)	40 (33.75)
4	Considered surgery to alter appearance	2.5 (1.43)	37.5 (35.84)
Appearance subscale		2.68 (1.31)	41.88 (32.71)
5	Uncomfortable meeting new people	3.6 (1.07)	65 (26.87)
6	Bothered by people's stares	3.2 (1.32)	55 (32.91)
7	Avoided social activity	4.5 (0.71)	87.5 (17.68)
Social subscale		3.77 (1.17)	69.17 (29.13)
8	Felt anxious about condition	3.6 (0.97)	65 (24.15)
9	Felt frustrated about condition	3.2 (1.32)	55 (32.91)
10	Worried about progression of asymmetry	3.2 (1.14)	55 (28.38)
Emotional subscale		3.33 (1.12)	58.33 (28.11)
11	Concerned about skin changes	3.2 (1.48)	55 (36.89)
12	Bothered by headaches or eye problems	4.2 (0.92)	80 (22.97)
13	Bothered by surgical complications	3.9 (1.10)	72.5 (27.51)
Symptoms subscale		3.77 (1.22)	69.17 (30.57)
Total score		3.39	59.64

* A 5-point response format was used to assess the extent each item described the patients's feelings, 1 (very much) to 5 (not at all). ** Each item standardized as follows: (raw score 1) × 100/4. Subscale total scores were obtained by summing individual standardized scores and dividing by number of items on the subscale. Total score was computed by summing subscale total scores and dividing by 4.

mean age at surgery was 15 years. It may be reasonable to hypothesize that children with facial morphea may be more emotionally distressed than patients with more limited lesions, such as plaque morphea or linear lesions, that are not nearly as evident as facial lesions. In addition, the perceived physical appearance may assume greater importance in late adolescence and adulthood. In our small cohort, all patients underwent surgery because of dissatisfaction with their appearance. As expected, the appearance subscale demonstrated the lowest standardized scores and possibly the greatest negative effect on patients' quality of life. The emotional subscale was the second lowest, with patients expressing worry about progression of asymmetry and frustration with their appearance despite surgery. Relatively higher scores were obtained for the social and symptoms subscale. The majority of patients continued to engage in social activity despite their disfigurement. Patients were also not concerned about complications of surgery, skin changes, headaches, and eye symptoms. The results of the questionnaire clearly establish the benefits of surgical interventions. While 9/10 patients reported various degrees of satisfaction with surgical intervention, all of them would recommend surgical intervention for other patients with ECDS and PRS, confirming that it is a useful intervention in children with facial disfigurement.

This study has several limitations: i.e., the small number of subjects and survey respondents, the retrospective design of the study with lack of preoperative assessment of patient's quality of life, and absence of prior reliability and validity of the questionnaire. However, despite these limitations, our study adds important data to the limited literature.

The 2 potentially disfiguring disorders ECDS and PRS have a considerable influence on the patients' quality of life. Our data prove that the psychological distress of patients brought on by these 2 processes is significant, prompting them to seek surgery for facial recontouring. Surgical interventions are available to these patients for volume augmentation and cosmesis, with the majority of patients requiring multiple treatments. Additional prospective studies on surgical outcomes, ideal timing, and type of intervention as well as questionnaire validation are recommended.

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