



## Aesthetic treatment of Romberg's disease: experience with 35 cases

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**SUMMARY.** Our experience with dermis-fat free flaps and conventional reconstructive procedures for 35 patients with Romberg's disease over a 7-year period is presented. The groin flap was used for increasing volume in 33 cases, the scapular flap in 3 cases, and 2 were treated with the latissimus dorsi free flap.

To achieve better contour, secondary procedures such as defatting of the flap, pedicled temporal fascial flaps, cartilage and bone grafts, orthognathic surgery, and other conventional aesthetic procedures were performed in 32 patients.

Excellent results were achieved in 9 mildly deformed patients, in 12 out of 16 moderately, and in 3 out of 10 severely affected patients. Four moderate and 5 severe cases were classified as good results. Poor results were achieved in 2 severe cases.

Romberg's disease, also known as progressive hemifacial atrophy, is a poorly understood entity<sup>1-4</sup>. Previously healthy patients are usually affected in the first or early second decade of life with progressive atrophy of the skin, fat, muscle, cartilage and even bone, producing a severe facial deformity.<sup>5-9</sup> The disease is more frequent in females and usually affects one side of the face. The initial sign may be a slow wasting of subcutaneous tissue on the lateral aspect of the chin, on the malar area or a subcutaneous cleft in the eyebrow or paramedian forehead area ("Coup de sabre"). One of the earliest diagnostic signs is the spread of a pigmented atrophic area.

The atrophy of skin and subcutaneous tissue progresses over a period of 2-10 years, usually involving one side of the face.<sup>5-9,17</sup>

If the disease appears during the first decade of life, it may interfere with the development of the skeletal structures on one side of the face. In contrast, when the disease begins during the second decade of life, it is less likely to interfere with the overall growth of the facial bony skeleton.<sup>5,6,8,9</sup>

Histological studies of the involved skin tissue frequently show chronic inflammatory changes which lead to scar formation. The microscopic differentiation of Romberg's disease from localised scleroderma depends on the preservation of elastic tissue in Romberg's disease.<sup>10,11</sup> No specific pathological factor has been demonstrated as a causal agent in this disorder<sup>12-19</sup> and it may be associated with a great number of pathological features.<sup>12,20-25</sup>

The aim of surgical treatment is a cosmetic amelioration of the defect. Several conventional reconstructive procedures have been used for correcting the facial asymmetry.<sup>26</sup> They include dermal fat grafts,<sup>14,26</sup> silicone fluid injections,<sup>27</sup> cartilage and bone grafts,<sup>14,26,28</sup> pedicled cutaneous, muscular or musculocutaneous flaps,<sup>29,30</sup> injection of lipoaspirated fat<sup>31</sup> and free flaps.<sup>32-34</sup> Wells and Edgerton used the

first free groin flap;<sup>35</sup> other authors have used omental, scapular and latissimus dorsi free flaps.<sup>30,36-39</sup>

The purpose of this paper is to present our experience in treating Romberg's disease with dermis fat free flaps and conventional aesthetic procedures.

### Materials and methods

We reviewed all the cases of Romberg's disease over a period of 7 years (1984-1990) at the General Hospital "Dr. Manuel Gea Gonzalez" in Mexico City. Twenty-three females and 12 males with Romberg's disease underwent transfer of free flaps and several reconstructive procedures.

Our 35 patients were classified as mild, moderate or severe according to the severity of the disease, based upon the atrophy of skin and subcutaneous tissue as well as bone involvement in the territories of the sensory branches of the trigeminal nerve, either frontal, maxillary or mandibular. The cases were classified as mild when the atrophy of skin and subcutaneous tissue affected the territory of only one of the sensory branches of the trigeminal nerve, and there was no bone involvement. If the disease affected two trigeminal territories without involvement of the osseous orbital region or maxillary and mandibular development, the case was considered as moderate. Finally, when the disease involved all three trigeminal territories or there was bone involvement, the cases were classified as severe.

Surgery was undertaken 1 year after photographic monitoring recorded no further progression of loss of volume.

Two surgical procedures were performed for aesthetic enhancement: first, a transfer of a dermis-fat free flap for volume; and second, adjunctive conventional reconstructive procedures.

The groin flap was our first option because the vessel diameters (superficial circumflex iliac artery and vein)



Fig. 1

**Figure 1**—The dimensions of the defect were measured with a paper model and transferred to the donor area for designing the flap.

corresponded to the recipient facial artery and vein. The donor area was also ideal for leaving an inconspicuous scar and furthermore, two teams can work simultaneously. If the groin flap was not suitable for free transplantation or a longer pedicle was needed, a scapular dermis-fat free flap was chosen. The musculocutaneous free flap (latissimus dorsi) was used only when more volume was required, especially in very thin patients.

A paper model of the exact dimensions of the facial defect was transferred to the donor area for flap designing (Fig. 1). The flap was first de-epithelialised, then dissection proceeded from lateral to medial, until the pedicle was visualised. The donor area was closed primarily in all cases.

We always placed the proximal portion of the flap in the lower part of the face. To create the recipient site, a pocket was dissected between the SMAS and the overlying skin and subcutaneous tissue through a preauricular incision. A small separate incision exposed the recipient vessels. The flaps were always placed with the dermis on the superficial aspect. In order to minimise shifting or displacement of the transferred flap, it was fixed to the periosteum of the zygomatic arch and inferior orbital rim. An end-to-end anastomosis was performed either to the facial artery and vein or to the superior thyroid artery and external jugular vein. External stitches were always used in order to avoid the tendency for the flap to contract. Hence, the exact dimensions of the defect were covered (Fig. 2).

The treatment of each case was selected based upon the severity of the disease. Mild cases usually required primarily a dermis-fat free flap. In a second stage the flap was defatted. Liposuction was not used in any of the cases. Secondary procedures such as cartilage grafts in the piriform area, V-Y plasties for labial mucosa, rhinoplasty and hemirhytidectomy were performed.

Similarly, moderate cases required the same procedures as the mild cases. In addition they also required a pedicled ipsilateral temporal fascial flap for the frontal region and "coup de sabre", bone grafts for depressed areas of the chin and mandible, and sliding osteotomies for the chin in some patients.



Fig. 2

**Figure 2**—The recipient area showing the preauricular incision and a pocket between the SMAS and the overlying skin and subcutaneous tissue, and the separate small incision to expose the recipient vessels.

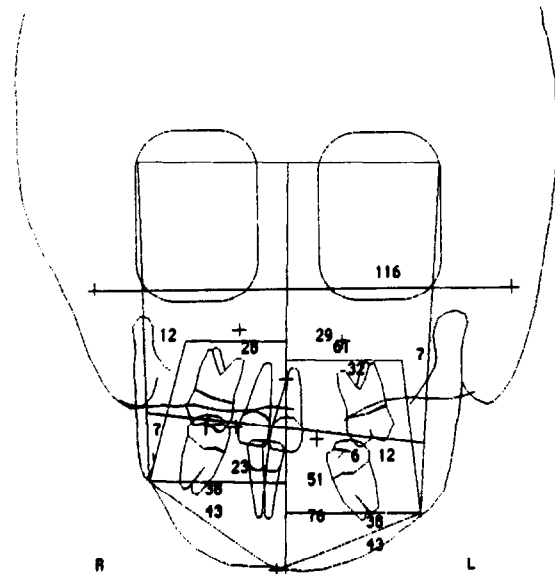


Fig. 3

**Figure 3**—A-P cephalometry of a 15-year-old patient with maxillary, mandibular and orbital involvement on the right side. In this severe case of Romberg's disease orthognathic surgery was performed.

Severe cases with bone involvement required all the procedures mentioned herein for mild and moderate cases, and in addition orthognathic surgery. The maxilla was lowered by a LeFort I osteotomy to improve the vertical dimension of the face on the affected side. A sagittal split osteotomy accomplished the required rotation and projection of the mandible (Fig. 3).

All the patients were followed up for a period of 2–7 years. All cases were evaluated at least 2 years after the



Fig. 4

**Figure 4**—(A) Preoperative frontal view of a 25-year-old mild case. (B) Postoperative frontal view of the same patient 2 years after a groin dermis free flap was performed. Secondary defatting of the flap, coronal lift, rhinoplasty, V-Y plasty for the inferior lip, and a cartilage graft in the piriform fossa were performed in a second stage 8 months after the microsurgical procedure. Her result was considered excellent.

secondary procedures were performed. According to this evaluation, the results were considered excellent when: a) very good symmetry was obtained, b) the volume was maintained for 2 years or more and c) no shift of the flap was evident. The results were classified as good if: a) acceptable symmetry was obtained, b) only a slight sag of the flap was present, and c) a stigma of the disease persisted. Results were considered poor when a) symmetry was not achieved, b) the flap sagged and placed traction on the lower eyelid, and c) the bone problem was not adequately resolved, specially in the orbital region.

## Results

Since 1984, 35 patients—23 females and 12 males—with Romberg's disease underwent transfer of free flaps. Ages ranged from 12–34 years old, with an average age at the time of surgery of 18 years.

In all patients, the disease had been active for a period of 1–9 years (mean 4 years). All patients presented with soft tissue atrophy. 70% of the cases began with progressive soft tissue atrophy. The remaining 30% began with a pigmented atrophic area in the trigeminal nerve distribution. We found 8 “coup de sabre” deformities, alterations of facial bone development in 8 cases and one patient with initial retinal alterations. Other associated disorders were found, such as one patient with a haemangioma of leptomeninges, another with blindness in one eye due to retinal atrophy, one with bilateral manifestations, and another involving one half of the body. Three patients had undergone previous attempts to correct the deformity with procedures other than free flaps.

Nine cases were considered as mild, 16 as moderate and 10 as severe.

Of the 38 free flaps transferred, one case was bilateral and two required a secondary free flap after initial flap failure due to venous thrombosis.

A groin flap was used in 33 patients, a scapular free flap in 3 cases, and a latissimus dorsi musculocutaneous free flap was selected in 2 cases.

Ancillary procedures were performed in 32 cases in order to achieve better aesthetic results. These surgical procedures were performed at least 6 months after the dermis-fat free flap transplantation. In all cases the flap was defatted and repositioned. 28 patients required a cartilage graft in the piriform area. Seventeen V-Y plasties for labial mucosa, 14 rhinoplasties and 16 hemirhytidectomies were also performed.

In moderate and severe cases, 8 pedicled ipsilateral temporal fascial flaps for the frontal region, 6 bone grafts for depressed areas of the chin and mandible, and 7 sliding osteotomies for the chin were performed.

Three severe cases required orthognathic surgery—Le Fort I and a sagittal split osteotomy. The procedure was performed before the free flap transplantation. Thus, these patients underwent three different surgical procedures.

All the postoperative photographs were taken at least 1 year after the secondary procedures for refining contour were performed.

The results of the nine mild cases were considered as excellent; adequate symmetry was found in all nine cases (Fig. 4). No sag of the flap was evident. The ancillary procedures left patients with no stigma of the disease. None of the patients presented only frontal involvement.

Out of 16 moderate cases, 4 patients had slight shift



Fig. 5

Figure 5—(A) Preoperative frontal view of a 25-year-old moderate case of Romberg's disease. (B) Postoperative frontal view one year after defatting of a groin dermis free flap, performed 6 months after the free flap. The result was classified as excellent.



Fig. 6

Figure 6—(A) Preoperative  $\frac{3}{4}$  view of a severe case of Romberg's disease with bone involvement. (B) Postoperative view of the same patient after orthognathic surgery, a groin dermis free flap 6 months later and subsequent defatting, cartilage grafting in the piriform fossa, and temporal fascial flap for frontal region.

of the flap. Therefore, adequate symmetry could not be achieved. However, the results were considered as good. The remaining 12 patients had good volume and symmetry, and they were considered as excellent results (Fig. 5).

In the severe cases, excellent results were achieved in 3 patients (Fig. 6). Five cases were considered as good results, and in 2 cases the results were classified as poor, due especially to the orbital involvement.

Figures 7 and 8 show further results.



Fig. 7

**Figure 7**—(A) Preoperative basal view of an 18-year-old severe case. (B) Postoperative basal view 2 years after defatting of a groin dermis-fat free flap, coronal lift, and lateral canthopexy. This was considered a poor result as although good symmetry was achieved, the orbital problem was still present due to significant three-dimensional underdevelopment of this area, including the eye and soft tissues.



Fig. 8

**Figure 8**—(A) Preoperative frontal view of a 20-year-old moderate case. (B) Postoperative frontal view 7 years after defatting of a groin dermis-fat free flap, coronal lift and alar reconstruction. The flap remained in a good position and shape. A continuous slow thinning of the skin was noted which did not compromise the flap volume.

### Discussion

Several procedures have been proposed for amelioration of the deformities of Romberg's disease.

Cartilage and bone grafts provide limited results because they can only increase volume with a bony base. These grafts were used in this study group only to enhance such areas as the orbits, piriform fossa and

chin. Fat grafts and rapidly reabsorbed. Hence, this procedure has been abandoned. Lipoaspirated fat injection as a surgical procedure is a temporary resource, with no control over the final result due to reabsorption. Muscular atrophy in pedicled or free muscular flaps presents a problem for calculating and accommodating the final volume. Since free omental flaps tend to sag, some authors have suggested placement in subcutaneous tunnels for preventing this complication, but this may lead to a cobblestone appearance. Due to the characteristics of the skin in patients with Romberg's disease, alloplastic materials have a high rate of potential complications and should be considered only in special conditions, such as increasing volume on the chin or the malar regions.

Based upon the results in this study, we consider that dermis-fat free flaps may be the treatment of choice for increasing volume in patients with Romberg's disease. They have good structural strength which allows an adequate adjustment to the defect and it is easy to refine the distribution of fat with secondary procedures. The free transfer of groin or scapular flaps with revascularisation to facial or neck vessels has a high rate of success.

Cosmetic improvement in mild and moderate cases is easier because the main surgical objective is only to increase volume and this may be achieved with dermis-fat free flaps. Secondary procedures such as defatting the flap provide a reliable and predictable result.

The major concern of dermis-fat free flaps is long term stability. For this reason surgical treatment was carefully divided in two stages, first, transfer of the free flap, and second, defatting and reposition. This latter stage was done at least 6 months after the first procedure. This period of time allows for evaluation of shape and position of the initially bulky flap.

Temporal fascia flaps gave good results, especially in the frontal area. Cartilage grafts in the piriform fossa produced good results and maintained volume in all cases. We used only autologous grafts. The chin was improved by sliding osteotomies for projections of more than 1 cm and by alloplastic implants for projections of less than 1 cm.

In the orbital region, poor results (Fig. 7) were obtained since it was difficult to achieve good overall symmetry with three dimensional osseous involvement in this area. In some patients orbital expansion was considered. However, in the presence of an underdeveloped eye and orbital soft tissues, expansion may increase the deformity. Furthermore, when the volume in the cheek and malar area is augmented, any orbital deformity may be enhanced.

We believe that in severe cases it is extremely important to achieve skeletal balance before a dermis-fat free flap is attempted.

Twenty-three patients with long term follow-up for at least 4 years demonstrated good position and volume of the flaps, especially in those cases evaluated as excellent and good results.

In none of the 9 patients with follow-up periods greater than 6 years has a new free flap been indicated (Fig. 8). In 6 of our 35 patients, the active phase of the disease persisted. The main problem in these patients is a continuous thinning of the skin and overlying soft

tissues. Nonetheless, no compromise of any flap was seen in any of these patients. Furthermore none of them showed significant reduction of the flap volume. Thus a new flap was never indicated.

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Paper received 26 May 1992.

Accepted 18 September 1992, after revision.