# Upper Labial Deficiency in Möbius Syndrome: A Previously Unreported Feature and Its Correction

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Bilateral facial palsy in Möbius syndrome remains one of the greatest challenges in reconstructive plastic surgery. Facial reanimation is an invaluable aid to such patients because it allows for greater social interaction by means of the ability to smile. In performing facial reanimation surgery on patients with Möbius syndrome, it is the observation of the senior author (Harrison) that upper labial deficiency is a consistent and previously unreported feature of the syndrome. It has been the practice of the senior author to perform upper labial augmentation on Möbius syndrome patients by insertion of a lipodermal autograft, in addition to facial reanimation. Nine patients with Möbius syndrome who presented to the Department of Plastic Surgery during an 8-year period were reviewed. All nine possessed bilateral facial palsy and upper labial deficiency in addition to other abnormalities consistent with Möbius syndrome. Six patients underwent bilateral facial reanimation and upper labial augmentation alone. One patient refused facial reanimation surgery but consented to upper labial augmentation. One patient, with concomitant micrognathia, underwent bilateral facial reanimation, upper labial augmentation, and insertion of a Silastic chin implant. In one patient, a child who also exhibited micrognathia, bilateral facial reanimation alone was carried out, with further procedures for upper labial and chin cosmesis being postponed until adulthood. The indication for performing upper labial augmentation was cosmetic. The procedure improved upper labial appearance and restored balance to the mouth. Patients also expressed higher satisfaction with eating and drinking, which they related to the improved fullness of the upper lip. This was before the facial reanimation had become functional. Upper labial deficiency warrants addition to the list of facial features of Möbius syndrome and is something that must be assessed in the context of facial reanimation surgery. (*Plast. Reconstr. Surg.* 112: 1762, 2003.)

In 1888, Möbius described combined sixth and seventh cranial nerve palsy separate from various congenital cranial nerve palsies.[1] The syndrome, which came to bear his name, has also come to include further abnormalities of the face, cranial nerves, upper limb, lower limb, and trunk.[2,3]

Abramson et al.[4] proposed a comprehensive classification and grading system for the Möbius syndrome. This system, CLUFT, is based on the five anatomical sites that may become affected, namely, cranial nerves, lower limb, upper limb, face, and thorax. The minimum clinical sign most commonly and classically associated with Möbius syndrome is bilateral involvement of cranial nerves VI and VII manifesting as either partial or complete weakness of each. Patients who present to reconstructive surgeons are usually seeking corrective surgery for bilateral facial nerve paralysis. Möbius syndrome patients have characteristic mask-like faces with adducted eyes and downturned mouth angles, and usually no voluntary facial movements are possible. Oral incontinence and drooling can occur in some patients.[5] Children with Möbius syndrome often report low self-esteem, which they relate to their appearance. These problems pose major limitations to social integration and interpersonal communication.[6] The prime aim of reconstructive surgery is to restore some degree of facial movement, particularly in response to emotion. This has been addressed by using innervated vascularized muscle flaps.[7] In Möbius syndrome, where paralysis is often bilateral, the masseteric motor branches of the

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fifth nerve are commonly used to provide innervation of the flap. This does mean that the patient has to smile by simultaneously clenching the teeth. It is, however, sometimes possible for patients to separate these actions by extensively retraining themselves.

It has been observed by the senior author (Harrison) that all patients with Möbius syndrome referred for facial reanimation surgery have a distinctive appearance characterized by upper labial deficiency, a previously unreported feature. Clinically, there is hypoplasia of the vermilion resulting in a reduction in its bulk. The labial height (nasal floor to the vermilion edge), definition of the vermilion edge, and Cupid's bow appear to be normal. In comparison, the lower lip seems to be of normal dimensions, which exaggerates the appearance of the abnormally deficient upper lip. Correction of this abnormality improves the appearance of the mouth and restores balance to the face in the static and smiling phases. In addition, the teeth become less prominent during smiling, and to some extent, this masks the fact that the teeth are clenched and produces a more natural appearance.

The indication for upper labial augmentation in Möbius syndrome patients is primarily cosmetic. Various functional problems that relate to the mouth have been described in Möbius syndrome, including drooling, oral incompetence, and speech difficulties. Facial reanimation has been shown to have positive effects in all of these areas.[5] None of these functional impairments were present in our group of patients. Therefore, we are unable to comment on whether the procedure has a functional benefit to the mouth. The patients, however, did report greater satisfaction with eating and drinking, which they related to the increased upper labial volume. In this series, upper labial augmentation with a lipodermal graft was performed on eight patients with Möbius syndrome; the extra improvement made to their overall appearance is reported.

## PATIENTS AND METHODS

A retrospective review of the 8-year period from 1994 to 2001 yielded nine patients with Möbius syndrome who were referred to the senior author for facial reanimation surgery. The group was composed of six male patients and three female patients with a mean age at first operation of 15.6 years (range, 6 to 39 years). Mean follow-up was 3.5 years (range, 4 months to 7 years). Table I lists the variety of clinical features expressed and the operations that were performed. None of the patients

TABLE I
Patient Data

| Patient | Age (yr) | Sex | Features | Operation(s) |
|---|---|---|---|---|
| 1 | 29 | M | Bilateral complete CN VI and VII palsy, hand anomalies, upper labial deficiency | Staged bilateral free LD flaps to CN V and upper labial lipodermal graft |
| 2 | 8 | M | Bilateral complete CN VII palsy, upper labial deficiency | Staged bilateral free LD flaps to CN V, and upper labial lipodermal graft |
| 3 | 6 | M | Bilateral complete CN VI and VII palsy, bilateral partial CN III palsy, upper labial deficiency | Lipodermal graft only |
| 4 | 18 | F | Bilateral complete CN VI and VII palsy, upper labial deficiency | Staged bilateral free LD flaps to CN V and upper labial lipodermal graft |
| 5 | 7 | M | Bilateral complete CN VII palsy, upper labial deficiency | Staged bilateral free LD flaps to CN V and upper labial lipodermal graft |
| 6 | 13 | M | Bilateral complete CN VI and VII palsy, absent left pectoralis major and nipple, syndactyly left hand, hypoplasia right hand, micrognathia, upper labial deficiency | Staged bilateral free LD flaps to CN V No upper labial augmentation |
| 7 | 14 | M | Bilateral complete CN VI and VII palsy, upper labial deficiency | Staged bilateral free LD flaps to CN V and upper labial lipodermal graft |
| 8 | 6 | F | Bilateral complete CN VI and VII palsy, conductive hearing deficiency, upper labial deficiency | Staged bilateral free LD flaps to transposed CN XI and upper labial lipodermal graft |
| 9 | 39 | F | Bilateral complete CN VII palsy, micrognathia, bulbar palsy, upper labial deficiency | Staged bilateral free LD flaps to CN V, upper labial lipodermal graft, and Silastic chin implant |

CN, cranial nerve; LD, latissimus dorsi.

complained of a functional eating problem, such as drooling or oral incompetence. Their primary concern was the lack of facial expressions. All but one patient underwent facial reanimation with staged bilateral free latissimus dorsi flaps. Eight patients underwent upper labial augmentation with a lipodermal graft. The graft was obtained at the site of harvesting of the latissimus dorsi flap, except in the one patient undergoing upper labial augmentation alone, in whom the graft was taken from the groin. The graft was inserted into the upper lip by means of two small lateral mucosal incisions at the submucosal level.

Three plastic surgeons (a senior resident, a consultant, and the senior author) evaluated preoperative and postoperative photographs. Upper labial fullness was assessed, as was the balance between the upper and lower lips. In addition, the overall appearance of the mouth in static and dynamic phases was also assessed.

## RESULTS

Upper labial deficiency was observed in all the patients referred for facial reanimation. This deficiency is best described as a decrease in labial bulk with a reduction in the vermilion. It is not possible to say to what degree there is hypoplasia of the submucosa and orbicularis oris. The labial height, the white roll, and the Cupid's bow, however, appeared to be normal. The parents of one patient (patient 3) did not consent to facial reanimation but did agree to upper labial augmentation. For patient 6, no upper labial augmentation was performed because of the concomitant micrognathia. There were no complications in any of the upper labial augmentations. Assessment of the preoperative and postoperative photographs revealed a definite improvement in the cosmesis of the patients. In addition, all the patients felt that upper labial augmentation had improved their overall appearance. All also expressed greater satisfaction with eating and drinking that was noted within several weeks after the operation but before facial reanimation had become functional.

The preoperative and postoperative photographs for patients 1 through 4 are shown in Figures 1 through 4, respectively. In Figures 1 and 2, it can be seen that, in addition to the considerable improvement in appearance and function brought about by staged bilateral facial reanimation, upper labial augmentation has also added to the overall improvement in both static and smiling phases. Figure 3 demonstrates the improvement achieved in the static appearance of the mouth by upper labial augmentation alone in the one patient whose parents did not consent to facial reanimation. Figure 4 demonstrates preoperative static and smiling appearances in patient 4. Patient 4 shows a functioning platysma, as evidenced by the descent of the lower lip in the preoperative

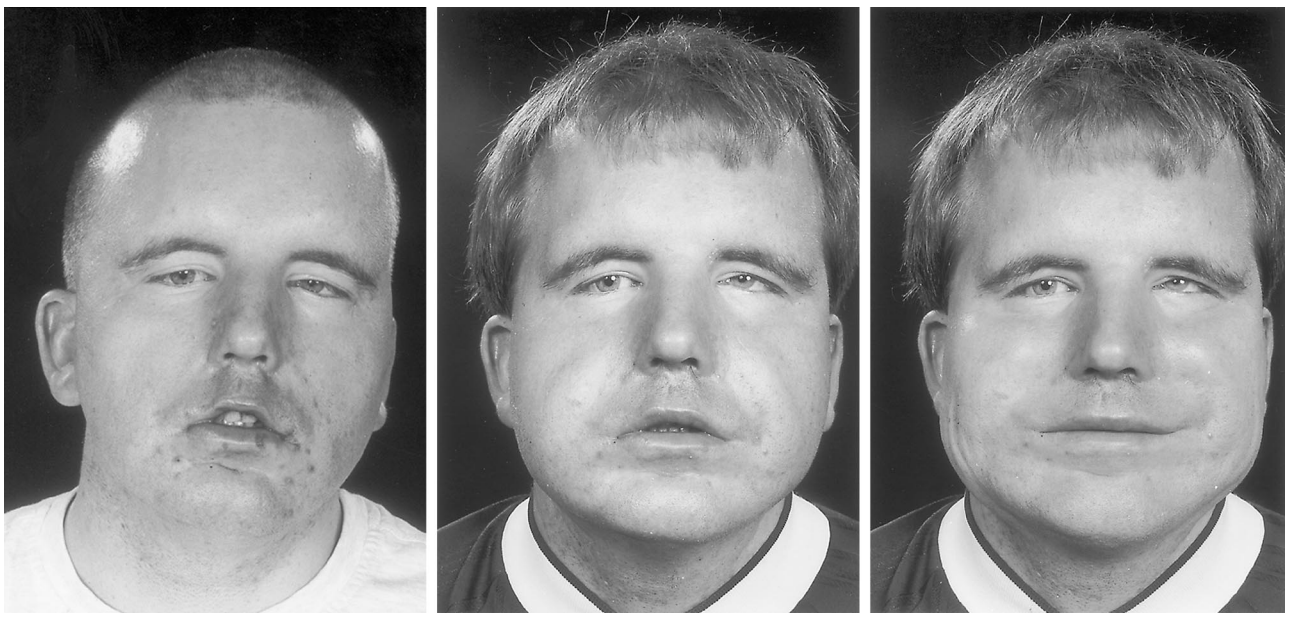

FIG. 1. Static view preoperatively (*left*) and views 9 months after staged bilateral facial reanimation and upper labial augmentation in patient 1: static (*center*) and smiling (*right*).

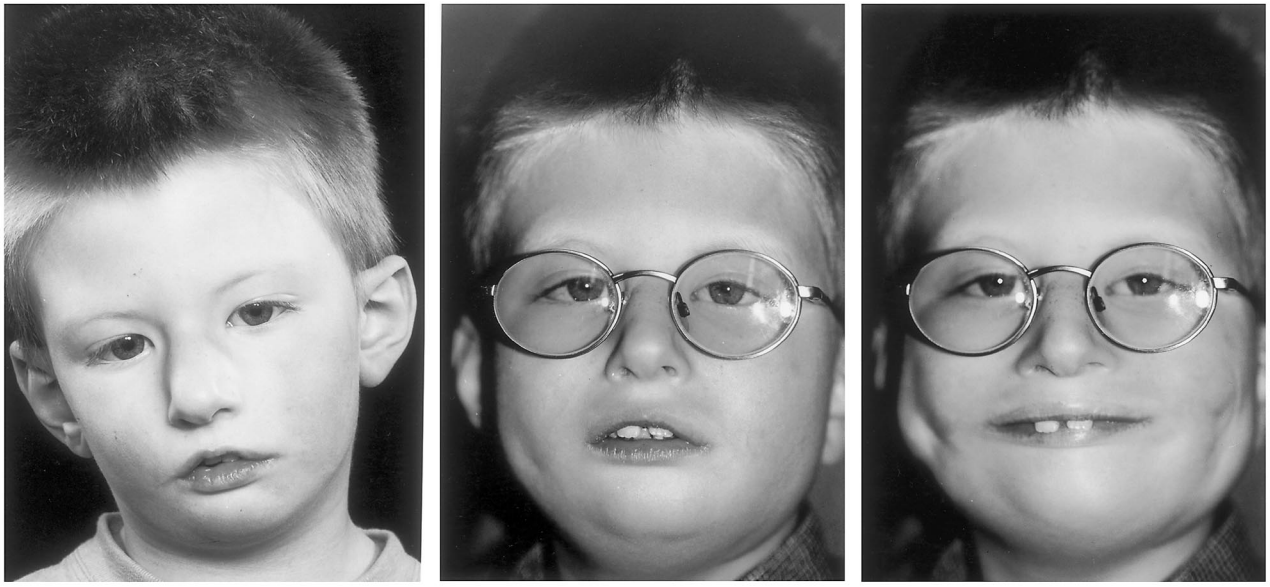

FIG. 2. Static view preoperatively (*left*) and views 5 months after staged bilateral facial reanimation and upper labial augmentation in patient 2: static (*center*) and smiling (*right*).

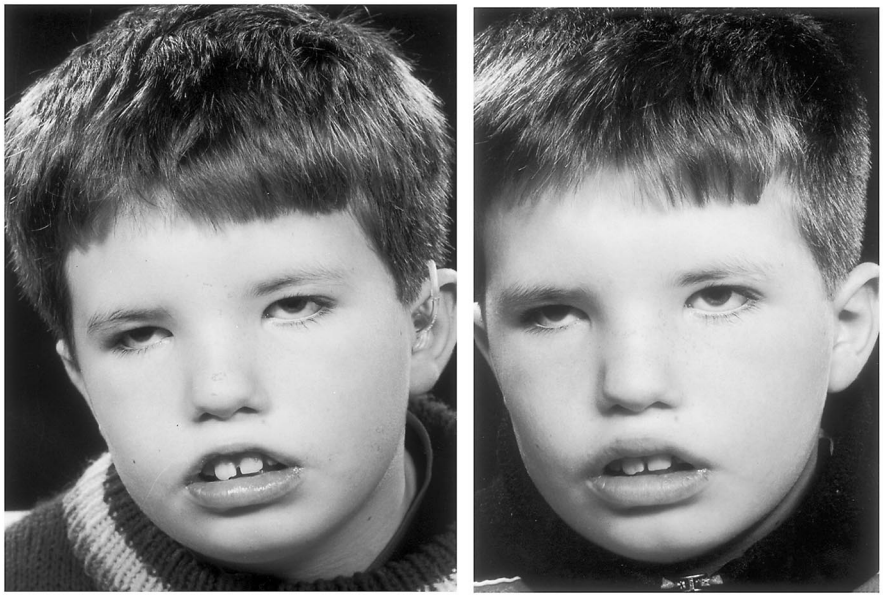

FIG. 3. Static views preoperatively (*left*) and 3 months after upper labial augmentation only in patient 3 (*right*).

smiling view. The postoperative photographs show marked improvements in both the static phase following upper labial augmentation and the smiling phase following an excellent result from the facial reanimation.

## DISCUSSION

For the purposes of this study, a sample size of nine patients may be deemed sufficient when the rare incidence of Möbius syndrome is taken into account. Indeed, previously unreported features of the syndrome are still put forward in the literature on the basis of individual case reports alone. The finding of upper labial deficiency in these patients does appear to be a consistent one within the group studied, and to our knowledge, this feature has not been previously reported in the literature. Clinically, the primary abnormality appears to be a vermilion that is hypoplastic and deficient

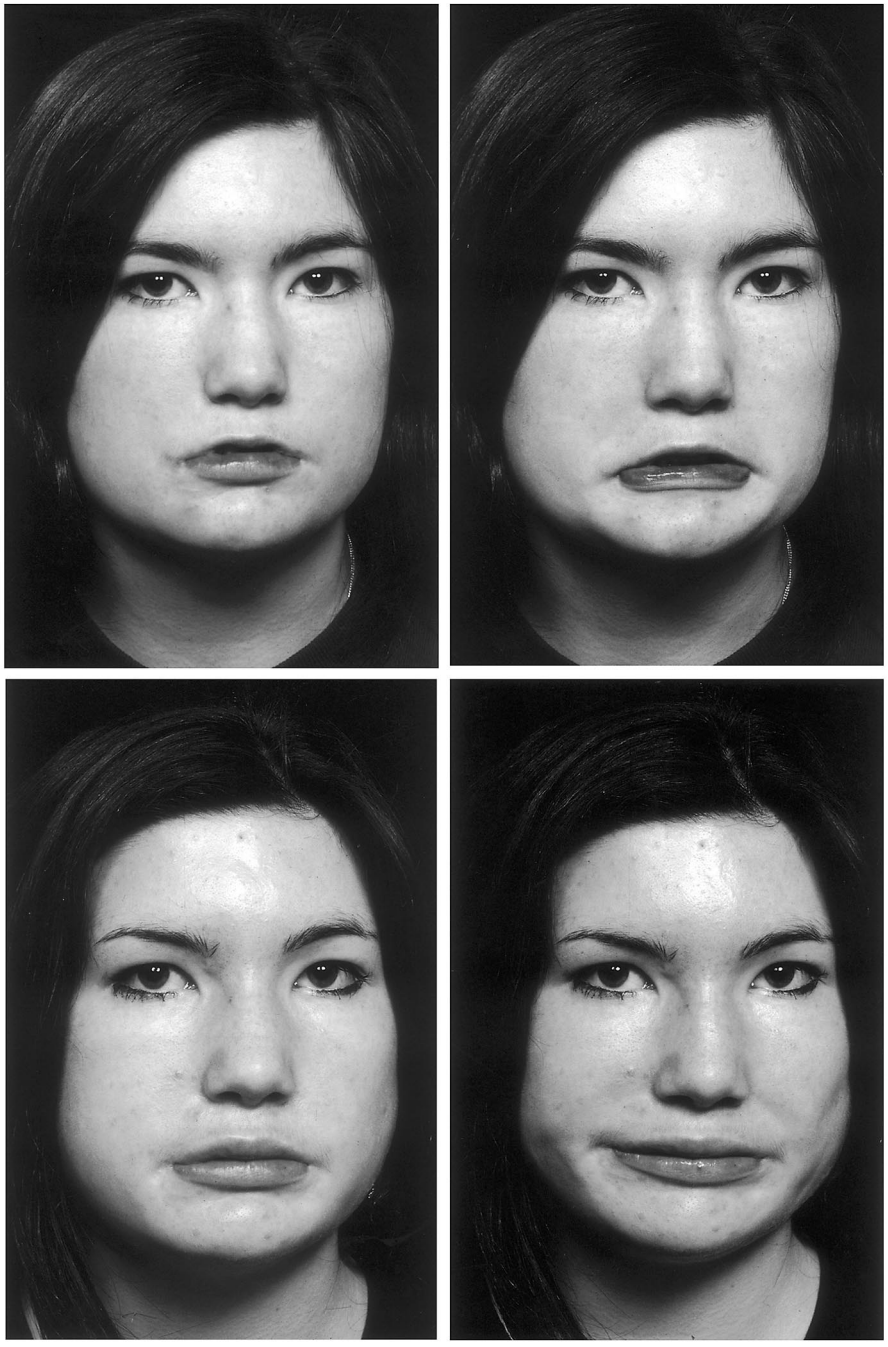

FIG. 4. Preoperative static view (*above, left*) and smiling view (*above, right*) in patient 4. Six months after staged bilateral facial reanimation and upper labial augmentation: static (*below, left*) and smiling (*below, right*).

in bulk. The labial height appears to be normal, as does the definition of the white roll and Cupid's bow. In contrast, the lower lip appears to be of normal size and shape. This disparity between the lips results in a distinctive appearance in the Möbius syndrome patient. It is the opinion of the senior author that it is possible to identify these patients by this characteristic appearance. The most likely explanation is that atrophy of the facial musculature, including the orbicularis oris muscle, secondary to facial nerve paralysis, contributes to upper labial deficiency. It is puzzling, however, why the lower lip does not seem to be equally deficient. It may be that platysma muscle fibers, which tend to be functional in Möbius syndrome patients,

contribute toward the lower lip bulk. If muscle atrophy were the only cause, one would expect to see asymmetrical lip hypoplasia in unilateral facial palsy. There are no reports in the literature to support this. In our experience with unilateral facial palsy, we have not observed an obvious size asymmetry between the paralyzed and normal sides. It is possible that other factors contribute to upper labial deficiency in the Möbius syndrome patient, such as deficiency in fat or connective tissue deposition. The cause of this abnormality can only be accurately established by histologic means to ascertain the deficient structures. This would require a full cross-section of the lip, as a punch biopsy would not be sufficient. However, it would be difficult to justify such a procedure.

Aside from the major abnormalities associated with Möbius syndrome, which are the focus of ongoing research into the condition, upper labial deficiency is perhaps of greatest importance to the reconstructive surgeon dealing with these patients. In contrast to the technically demanding procedure of facial reanimation, the far simpler procedure of upper labial augmentation in this study using a lipodermal autograft is easy to perform. It adds to the overall cosmetic improvement achieved in both the static and smiling phases. Although the indication for labial augmentation was cosmetic, patients did express greater satisfaction with eating and drinking following the procedure. One patient (patient 3) in the group who underwent upper labial augmentation as a sole procedure gained an appreciable improvement in the appearance of the mouth. This is an important point, because of the psychological impact of Möbius syndrome on patients caused by the restriction in social interaction. All procedures aimed at normalizing their appearance should be considered worthwhile, especially one so straightforward as upper labial augmentation.

There was no significant shrinkage of the lipodermal graft in this series, and none of the patients required regrafting procedures. The follow-up, however, was short (4 months to 7 years). With one exception, all the patients were followed up for less than 2 years. This is because we operate on patients referred from all over the United Kingdom and Ireland, and for geographical reasons, patients are discharged to their referring source. The movement in these patients, even after facial reanimation, is less than that seen in the healthy population, and this may account for the reduction in graft resorption.

## CONCLUSION

These patients may be offered this simple procedure, which can improve the appearance of the mouth and restore improved balance to the face, even if they choose not to undergo facial reanimation itself.

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