

Surgical and Prosthodontic Rehabilitation in a Patient With Freeman-Sheldon Syndrome

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Abstract: Dental and prosthetic rehabilitation possess significant challenges in patients who have Freeman-Sheldon syndrome. Microsomia is one of the main diagnostic criteria for Freeman-Sheldon syndrome, and it creates difficulty in working in the intraoral cavity. Most patients with small orifice often have difficulties in oral hygiene maintenance, and it gives rise to loss of some of the teeth. It incurs the need for dental and oral treatment.

In the presented study, the patient with limited mouth opening that disabled the dentists to perform dental treatment was given prosthodontic therapy after having commissuroplasty and implant placement simultaneously.

Key Words: Microsomia, Freeman-Sheldon syndrome, commissuroplasty, dental implants, implantology, small orifice

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Freeman-Sheldon syndrome (FSS) is an uncommon clinical condition. Freeman-Sheldon syndrome was first described by E. A. Freeman and J. H. Sheldon in 1938.¹ Freeman-Sheldon syndrome is a heterogeneous condition with neither sex nor ethnic preference. The syndrome is encountered in the musculoskeletal systems. Stevenson et al² reported strict diagnostic criteria for FSS. These included 2 or more major manifestations plus the following: microsomia, whistling face, nasolabial creases, and “H-shaped” chin dimple. Major manifestations involve ulnar deviation in wrists and fingers, camptodactyly, talipes equinovarus, calcaneovalgus, scoliosis, vertical talus, and/or metatarsus varus.² A distinctive facial appearance of microsomia, flatness of the middle part of the face, deeply sunken eyes, epicanthal folds, long philtrum, H-shaped chin dimple, hypoplastic alae nasi, and mask-like immobility.^{2–4} Mental development of patients with FSS is usually normal. Freeman-Sheldon syndrome is a heterogeneous condition, and both sexes are equally affected. Early diagnosis of FSS is usually made based on the clinical appearance at birth; however, it has been diagnosed prenatally using ultrasound.⁵

An abnormality of the small orifice is defined as microsomia. Microsomia may be caused by surgical treatment of orofacial neo-

plasm, maxillofacial trauma, burns, cleft lips, radiotherapy, connective tissue diseases, for example, scleroderma, or some craniofacial syndromes, including FSS. This clinical aspect introduces significant challenges for food intake and oral hygiene maintenance. After all, microsomia creates difficulty during dental treatment.^{6,7} Patients with microsomia who need dental prostheses often face the difficulty of being unable to insert or remove impression trays and the prosthesis because of the constricted opening of the oral cavity. The prosthodontist needs to create special impression trays for the patient. Oral and/or dental surgeries such as alveoloplasty or placement of dental implants are the other challenging procedures for patients who have microsomia.

The treatment of microsomia presents particular difficulties. Conservative management of microsomia includes the use of microsomia orthoses to expand oral opening. The surgical correction of microsomia will be helpful for the prosthodontist. To release the lips at the commissures, a procedure known as commissuroplasty or commissurotomy provides free access to the oral cavity.

The aim of this article was to describe the clinical management of a patient with microsomia induced by FSS using implant-supported prostheses.

CLINICAL REPORT

A 26-year-old man was referred to the Faculty of Dentistry for dental and prosthetic rehabilitation. Clinical examination revealed poor oral hygiene. The extent of mouth opening was limited and restricted. The patient had the symptoms of FSS in his face.

The patient had immobile face, sunken eyes, high-arched palate, small tongue, microsomia with a characteristic whistling mouth, and H-shaped dimpling on the chin (Figs. 1–3). Radiographic skeletal survey showed that scoliosis was present (Fig. 4). He had ptosis on both sides. Despite his nasal speech, he communicated easily and seemed to be of normal intelligence. The left foot of the patient showed a cutaneous syndactyly (Fig. 5).

Intraoral examination revealed evidence of poor oral hygiene with plaque deposits. His left maxillary first premolar and second molar, right maxillary canine and left canine, first premolar, first and second molar, and right mandibular second molar were present in the panoramic radiograph (Fig. 6). He was partially edentulous, and some maxillary and mandibular teeth were missing. Commissuroplasty was planned to improve the mouth opening and place endo-osseous dental implants for prosthetic rehabilitation.

After nasotracheal intubation, commissuroplasty was carried out using Converse's technique.⁸ Incisions were made with no. 15 blades at the perioral skin and fibrotic bands to the underlying mucosa (Fig. 7). Then, the buccal mucosa was released, rotated toward lower-upper lips and vermillion to cover created defects, and was sutured. Immediately after commissuroplasty, 5 implants (Implant Diffusion International, Montreuil, France) were placed in the maxilla. Two implants were also placed in the mandible (Fig. 8). The intraoral and perioral sutures were removed on the 10th postoperative day.

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FIGURE 1. Typical facial features of FSS.

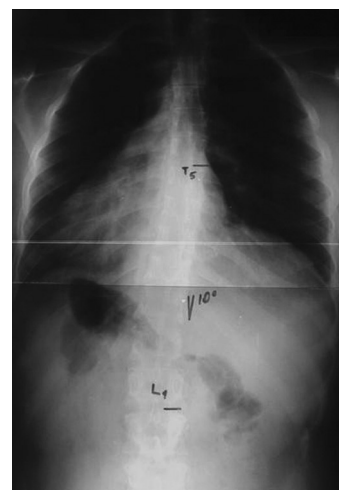


FIGURE 4. Scoliosis at level T5, L1 = 10 degrees.

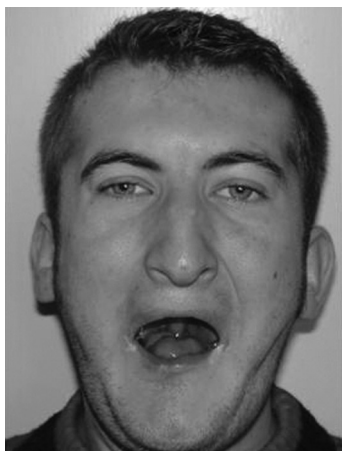


FIGURE 2. Profound microstomia.



FIGURE 5. Cutaneous syndactyly in the left foot.



FIGURE 3. Highly arched palate.



FIGURE 6. Preoperative panoramic view.



FIGURE 7. Intraoperative view of commissuroplasty.

After providing free access to the oral cavity, root canal therapy was applied to the left maxillary canine, second molar, and right mandibular second molar approximately 1 month after commissuroplasty.

The implants were exposed approximately 5 months after implant placement. Implant exposure and abutment screw connection were carried out, and definitive crom cobalt/ceramic bridges were fixed. A panoramic radiograph was obtained just before implant loading and 24 months after implant insertion (Fig. 9). The patient was satisfied with the aesthetics and function of the prosthesis (Fig. 10).

DISCUSSION

Freeman-Sheldon syndrome can follow either a dominant or a recessive inheritance pattern; however, some sporadic cases have been reported.⁹ The first FSS case described by Freeman and Sheldon had pinched lips (whistling-like lips), microstomia, prominent nasolabial folds, and H-shaped dimpling of the chin. The patients with FSS had characteristic face but lacked contractures of the hands and feet.² Alternatively, the expressivity of FSS may be so variable that some affected individuals show clearly only facial contractures. The patient in this study presented microstomia, high-vaulted palate and microglossia, pinched lips, H-shaped dimpling of the chin, sunken eyes, as described in the diagnostic criteria of the FSS.

The presence of scoliosis in an individual with facial contractures, like in the presented case, might be predictive of FSS.

In the differential diagnosis, other congenital disorders associated with microstomia such as Hutchinson-Gilford progeria, Hallerman-Streif syndrome, Burton skeletal dysplasia, Fine-Lubinsky



FIGURE 9. Panoramic radiography at the end of follow-up.

syndrome, Leopard syndrome, and Schwartz-Jampel syndrome should be considered.⁴

Other conditions that may cause microstomia are collagen group of disease and submucous fibrosis. Acquired microstomia is often a consequence of burns and surgery to the lips or a result of scarring after radiotherapy.

In patients with microstomia, complex dental applications such as dental implant placement, which require general anesthesia, may become complicated due to the difficulties in intubation. Muscular rigidity after halothane anesthesia has been reported.¹⁰

The presence of microstomia obstructs dental treatment procedures. Treatments of microstomia are based mainly on surgical techniques, nonsurgical approaches, or on a combination of both methods.^{8,11}

Ohyama et al¹² presented treatment of microstomia in the case with FSS. They used a mouth expander for 2 to 3 h/d before active orthodontic treatment. The authors claimed that this therapy produced an increase in mouth width. It is debatable whether this change was induced by mouth expander or whether it was a result of normal facial growth. However, widening obtained by nonsurgical stretching devices is limited and unsatisfactory for a prosthetic treatment. Therefore, surgical methods should be preferred to achieve maximal widening. Treatment period is short in surgical methods. Surgical interventions to improve microstomia have been reported in FSS by several authors.^{13,14} Ferreira et al¹³ reconstructed the vermillion deficit of the upper and lower lip using mucosal advancement flaps. Neumann and Coetzee¹⁴ reported that the upper lip is an aesthetically more important part of the mouth and used lower lip vermillion in the correction of the upper lip vermillion deficit. We observed stable results with complete satisfaction by using Converse's



FIGURE 8. Panoramic radiography just before implant loading (2 mo after placement of implants).



FIGURE 10. Intraoral view of definitive prosthesis.

technique.⁸ In a later follow-up, there was no recurrence or contracture of the orbicularis oris muscle.

In the treatment of the presented patient, commissuroplasty and implant placement have been applied simultaneously. Surgical treatment of microsomia and simultaneous dental implant placement have never been reported in patients with FSS so far.

The functional and aesthetic satisfaction of patients also depends on the success of prosthetic treatment after surgical managements. The shape of prostheses is determined by the limitation of narrow mouth opening in microsomia. The presented study showed that surgical reconstruction of the lips provides better access to the intraoral cavity, and simultaneously placed endosseous dental implants provide better rehabilitation in patients who need prosthetic treatment and who have small orifice.

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