IMPLANT REHABILITATION FOR A PATIENT WITH HALLERMAN-STREIFF SYNDROME: A CASE REPORT

Behruz J. Abadi, DDS; Joseph E. Van Sickels, DDS; Thomas A. McConnell, DDS, MSEd; G. Thomas Kluemper, DMD, MS

Hallermann-Streiff syndrome is a rare genetic disorder characterized by craniofacial malformations, sparse hair, eye abnormalities, dental defects, degenerative skin changes, and short stature. The syndrome has many implications for dental treatment. Patients typically present with multiple missing and poorly formed teeth. The purpose of this case report is to discuss the overall management of a patient with Hallermann-Streiff syndrome by oral maxillofacial surgery, orthodontic treatment, and prosthodontic reconstruction.

Key Words: dental implants, prosthodontics, Hallermann-Streiff syndrome

INTRODUCTION

Hallermann-Streiff syndrome, which is also known as the François dyscephalic syndrome, is a rare disorder that affects body stature and facial structure. Patients with this disorder have sparse hair, ocular abnormalities (specifically cataracts), and degenerative skin changes. Of interest to maxillofacial surgeons, orthodontists, prosthodontists, and restorative dentists are the deficient mandible with a narrow, highly arched palate and multiple missing and sometimes poorly formed teeth. The syndrome appears to be a sporadic mutation with an unknown inheritance pattern. Because of the very deficient mandible, there may be risk for obstructive sleep apnea. In a review by David et al, 11 of 15 patients with this disorder had significant intermittent respiratory difficulties, which required tracheostomy in 3 of them. Surgical correction of cataracts is often done early in life to protect vision. Despite the fact that patients with Hallermann-Streiff syndrome have multiple issues of interest to the dental profession, there is little information in the dental literature regarding their care and management. Limeres et al reported the management of a 29-year-old with dental extractions, scaling, and restorations performed under local anesthesia, followed by orthodontics. Many studies endorse the clinical success of implant retained fixed partial dentures. Due to the young age of the patient, as well as the partial anodontia, ankylosed primary teeth, and few permanent teeth, it was decided that prosthodontic rehabilitation with implant retained fixed partial dentures would be the best treatment option.

CASE REPORT

Initial examination revealed a severely retrognathic 19-year-old man with multiple missing permanent teeth and retained primary teeth (Figures 1 through 3). The patient had all the characteristics of Hallerman-Streiff syndrome, and a permanent tracheostomy for airway management was in place. From a skeletal assessment, his mandible was severely deficient, and he had posterior maxillary hyperplasia with a transverse deficiency. A multiple-phase plan was developed to address both his skeletal and dental discrepancies. The first phase was a mandibular advancement of 11 mm by placement of intraoral distractors (Figures 4 and 5). The second phase of treatment involved orthodontic therapy. Because of the extremely narrow palate,
Figures 1-5. Figure 1. Facial 3-dimensional reconstruction. Figure 2. Profile 3-dimensional reconstruction. Figure 3. Preoperative intraoral image. Figure 4. Intraoral distractors in place. Figure 5. Intraoral distractors in place.
surgically assisted, rapid palatal expansion (SARPE) was performed. A hyrax expander was used with bands cemented on the first molars; a soldered lingual bar extended forward to the ankylosed deciduous first molars to increase anchorage (Figures 6 and 7). Expansion of 11 mm was achieved and retained with the passive expander for 6 months. Although the SARPE corrected the patient’s transverse discrepancy, it did not address the excess vertical development of the maxilla. This was corrected by a Le Fort I maxillary impaction.

After surgical procedures, the third phase of treatment was started. Impressions (Jeltrate, Dents-
for diagnostic casts (Silky Rock, WhipMix Corporation, Louisville, Ky) were obtained for prosthetic rehabilitation. Using the diagnostic cast, the ideal position for implants was selected for the prosthetic restoration. The 12 remaining maxillary teeth, half of which were deciduous, were extracted and six $4.1 \times 12$ mm implants (Straumann USA, Andover, Mass) were placed. Two months later the 5 deciduous mandibular teeth were extracted and 5 implants were placed (three $4.1 \times 10$ mm and two $4.1 \times 10$ mm). After 6 months of integration (Figure 8), preliminary impressions (Jeltrate, Dentsply/Caulk)
were taken for provisional complete dentures. The vertical dimension was established, and the provisional complete dentures were inserted (Figures 9 and 10). After a month of wearing the provisional complete dentures, the patient was comfortable with the vertical dimension and occlusion (Figure 11). Eleven 4-mm abutments (Straumann USA) were torqued to 35 Ncm (Figure 12), and impression caps and positioning cylinders were placed (Figure 13). The implants were impressed with polyvinyl siloxane impression material (Extrude, Kerr Dental, Orange, Calif). Maxillary and mandibular frameworks for fixed partial dentures were tried in, the vertical dimension was evaluated, and the occlusion was adjusted (Figure 14). The patient returned for a final try-in of the maxillary and mandibular fixed partial dentures with porcelain. After patient approval, the maxillary and mandibular fixed partial dentures were cemented with a glass ionomer cement (Ketac-Cem, 3M Espe, St Paul, Minn) (Figure 15 and 16). The entire treatment took place over 3 years, and the patient has been satisfied with treatment for 1 year.

**DISCUSSION**

Patients with Hallermann-Streiff may present with a number of different medical, surgical, and dental challenges. Treatment therefore dictates an individual approach to each patient and frequently involves multiple disciplines. David et al \(^1\) and Limeres et al \(^3\) discussed a variety of different dental treatments and focused on the dental care needed for these persons. Most patients with this disorder have missing teeth and need some type of reconstruction. The patient in this case study had such a compromise of his dentition that it was decided that he would be best served by total implant reconstruction. His underlying skeletal deformities mandated that they be addressed first before the implants and fixed appliances could be placed. With distraction, SARPE, and surgical maxillary impaction, the final position of the maxilla and mandible in space is not as predictable as that seen with fixed restorations. Hence, once the skeletal surgery was completed, diagnostic models were obtained to achieve the best functional and esthetic results.

**REFERENCES**