Surgical Correction of the Mandible with Genioplasty Using a Porous Hydroxyapatite Block Graft for Hallermann-Streiff Syndrome: A case report

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We present the case of a 20-year-old female with micrognathia associated with Hallermann-Streiff syndrome. Successful surgical extension of the mandible was achieved by sagittal splitting osteotomy and genioplasty using a porous hydroxyapatite block graft. Titanium dental implants were used to correct malocclusion due to congenital partial adontia. The clinical course has been satisfactory with no functional or cosmetic abnormalities as of 15 years postoperatively.

Key words: Hallermann-Streiff syndrome, sagittal splitting, hydroxyapatite, genioplasty, dental implant

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Introduction
We performed surgical extension of the mandible by sagittal splitting of the mandibular rami and genioplasty using a porous hydroxyapatite block graft for micrognathia associated with Hallermann-Streiff syndrome. In addition, titanium dental implants were used to correct malocclusion due to congenital partial adontia. We review a case of Hallermann-Streiff syndrome in which functional and cosmetic improvement was achieved by this method.

Case report
Clinical findings
A 20-year-old woman who had been diagnosed shortly after birth with bilateral congenital cataracts and Hallermann-Streiff syndrome was referred to our department in March 1988 by a pediatrician from our hospital, for both functional and cosmetic oral and maxillofacial improvement. The chief complaints were mandibular deformity and malocclusion. The patient presented with proportionate dwarfism and was moderately-nourished. Height was 144 cm; body weight was 37 kg. She was born following a normal pregnancy and delivery. Gestation was 40 weeks and birth weight was 3080 g. No current illnesses were present and her family history was unremarkable. The patient presented with micrognathia and associated double cutaneous chin, thin pointed nose, ptosis of the cheek fat, and hypoplasia of the middle and lower face. Scalp hair and eyebrows were sparse and the palpebral fissures were small. Uncorrected vision was 0.07 and corrected visual acuity was 0.1 in both eyes. Her nose appeared symmetrical and beaked (Figs.1a, 2a). Psychoneurological examination was unremarkable. The oral cavity revealed congenital partial adontia and prolonged retention of upper and lower anterior deciduous teeth. Open bite and malocclusion were also revealed.

Surgical treatment
In March 1988, surgical extension of the mandible was undertaken by sagittal splitting of the mandibular rami and genioplasty using a porous hydroxyapatite block graft and extraction of the bilateral buccal fat pad under general anesthesia. Genioplasty was conducted as follows: a middle incision was made at the gingivolabial fold of the anterior tooth and a porous hydroxyapatite block was inserted to recontour the chin. The block was fixed using stainless steel wire (Fig.3). Titanium dental implants (OGA Implant, Japan) were substituted for missing teeth.
Dental implant surgery was performed under local anesthesia on two occasions in September 1988 (Fig. 4a).

Postoperatively, facial appearance and occlusion of the patient were significantly improved. At present, 15 years after surgery, her condition remains satisfactory (Figs. 1b, 1c, 2b, 4b, 5).

Discussion

Following the original case reports by Aubry in 1893, Hallermann and Streiff described the syndrome in 1948 and 1950, respectively. Known as the Ullrich-Frenmery-Donah syndrome by 1953, it was further described by Falls and Schull in 1960 (1-5). Characteristic features include dwarfism, micrognathia, pinched nose, congenital cataracts, a high palatal vault, malocclusion, and hypoplasia of the dental enamel. The cause of this malformation syndrome is unknown. Few cases of the syndrome have been reported in the oral surgery literature, as the syndrome has traditionally been investigated within the field of ophthalmology due to the occurrence of concomitant congenital cataracts. On the other hand, oral surgery for Hallermann and Streiff syndrome has been reported by Patterson (6) and Sclaroff (7), who undertook osteotomy and bone collecting with autogenous bone of the jaw for treatment of jaw deformity. In this patient's case, all fundamental symptoms according to Francois's classification were present except atrophy of the skin. Although the patient presented with micrognathia, congenital partial adontia, microdontia, and hypoplasia of the mandible, jaw movement was normal. Porous hydroxyapatite block graft was selected because...
porous hydroxyapatite offers numerous advantages. First, the material is in plentiful supply. Second, the size and shape can be freely adjusted due to the excellent processing properties of the material. Third, hydroxyapatite is not resorbed unpredictably as is often the case with bone graft. There was a great need for autogenous bone in transplantation although the patient showed well-proportioned dwarfism. However, we did not expect to take her autogenous bone, and porous hydroxyapatite block already had been shown to be useful for genioplasty (8). Artificial bone such as the transplanted porous hydroxyapatite block allows reconstruction and supplementary function without the risk of absorption, and is thus useful for creating a stable, long-term facial appearance. Furthermore, organization continuity with the covering tissue and circumference can be recovered by porous bodies with the invasion of surrounding organizational structures. The surface area of the material is enlarged, including as it does the walls of the pores. This facilitates osteoconduction, by which bone formation progresses along a surface. In other words, this material can expect biocompatibility with the bone by the covering which makes bone organization connect.

No absorption was observed even in radiographic images taken 6 years after the operation, and a favorable outcome was achieved. Bone stability can therefore be achieved using porous hydroxyapatite block. The porous hydroxyapatite block proved extremely useful as the transplant material in the present case.

There is, however, a report of pathological reaction
of hydroxyapatite plate and hydroxyapatite granules used to repair a craniotomy defect, which was removed after 2 years and 9 months of use (9). In that report, neovascularization and osteogenesis were observed in tissues surrounding the bone, at the same time with the new bone containing a Haversian system in the porous hydroxyapatite block body.

Mechanical strength was reportedly achieved following bone conduction from the surrounding bone to the implanted material (9). In the present case, unification of the porous hydroxyapatite block and mandible were observed postoperatively under radiography, as well. In addition, titanium dental implant restoration to the anterior teeth enabled mastication, providing both functional and cosmetic outcome improvement. As of 6 years postoperatively, bone absorption was not apparent in either the top of the radiograph or the dental implant, and occlusion also remains good.

Summary

We have described successful surgical correction of a mandible with genioplasty using porous hydroxyapatite block graft and titanium dental implants to treat Hallermann-Streiff syndrome. We conclude that porous hydroxyapatite block is useful in genioplasty.

References


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