Osseointegrated Implants in a Patient With Hermansky-Pudlak Syndrome: A Case Report

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Partial edentulism in a 16-year-old female with Hermansky-Pudlak syndrome was treated using the Brånemark osseointegration technique. Two 15 × 3.75 mm implants were placed in the mandibular anterior area. The two implants were tested for mobility using a Periotest instrument at the time of the implant placement, at abutment connection, and at 3 months following delivery of the final prosthesis. The results from this study showed that the Brånemark osseointegration procedure can be used to treat patients with platelet disorders affecting the clotting mechanism.
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Key words: Hermansky-Pudlak Syndrome, implants, osseointegration, Periotest, platelet disorders

Hermansky-Pudlak Syndrome (HPS) is an extremely rare blood disorder exhibiting a triad of clinical signs. The characteristic features of HPS include albinism, mild hemorrhagic problems resulting from platelet dysfunction, and accumulation of ceroid-like pigment in the bone marrow macrophages. Although Hermansky and Pudlak originally thought that the syndrome was related to the absence of platelets,[1] the bleeding disorder associated with HPS has been ascribed to a platelet defect[2–4] that includes a reduced concentration of serotonin, decreased number of platelet-dense granules, and concomitant diminution in the reaction essential for initiation and propagation of the second wave of irreversible platelet aggregation.

Longitudinal studies have shown that osseointegrated implants function successfully in healthy bone for the treatment of complete[5,6] and partial edentulism.[7–9] This article describes the hematologic considerations and clinical management of a partially edentulous patient with Hermansky-Pudlak syndrome using Brånemark implants to restore oral function and comfort.

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Case Report

A 16-year-old white female with Hermansky-Pudlak syndrome was admitted to the hospital for the treatment of traumatic injuries sustained in a motor vehicle accident. The preoperative diagnosis included the following: *(1)* open complete fracture of the symphysis of the mandible, *(2)* bilateral condyle fractures, *(3)* multiple fractures of the maxilla, *(4)* multiple fractured teeth, and *(5)* avulsed mandibular right central and lateral incisors. Three months after emergency treatment, the patient presented for prosthodontic rehabilitation. Clinical and radiographic evaluation revealed both the loss of the mandibular right central and lateral incisors and alveolar ridge resorption (Fig 1). The decision was made to place two Brånemark titanium implants (Nobelpharma USA, Chicago, IL) rather than a conventional fixed partial prosthesis that would result in excessive loss of healthy tooth substance. In consideration of her medical history, the patient was referred for additional hematologic consultation. A cooperative treatment plan was prepared by the authors, to coordinate therapy on an outpatient basis.

Stage 1 Osseointegration Surgery. The patient was scheduled for stage 1 osseointegration surgery, which was to follow the presurgical transfusion of platelets (six units). Thirty minutes after the platelet transfu-

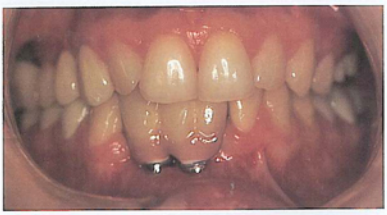

Fig 8 Final ceramometal tissue-integrated prosthesis.

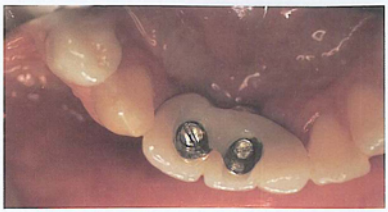

Fig 9 Incisal-lingual view showing screw access for the prosthesis through the cingulum areas.

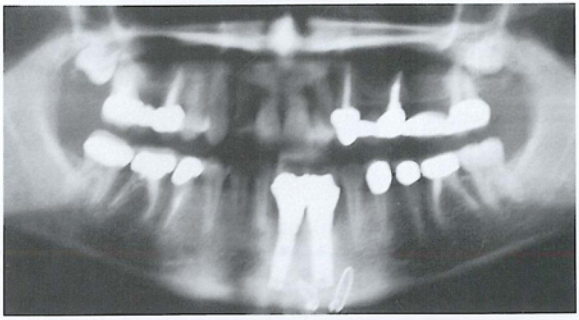

Fig 10 Panoramic radiograph illustrates the final prosthesis supported by two osseointegrated Brånemark implants.

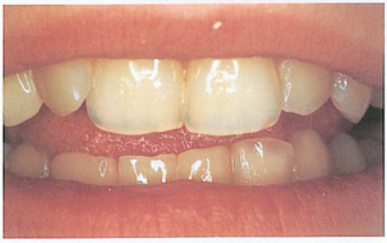

Fig 11 Postoperative view shows the esthetic rehabilitation provided using the fixed tissue-integrated prosthesis.

Healing occurred uneventfully, with no postoperative bleeding. The incisions were closed with Vicryl sutures. Three weeks later, the patient returned for suture removal and delivery of the final ceramometal tissue-integrated prosthesis (Figs 8 to 11).

Three months following placement of the final prosthesis, the patient returned for reevaluation. The tissue-integrated prosthesis was removed and Periotest values (Table 1) were obtained again for both implants (Fig 12). Results of all three Periotest evaluations are summarized in Table 1.

Plaque accumulations are known to produce gingival inflammation and, in advanced stages of periodontal disease, bleeding is common. Comprehensive oral hygiene instructions were given the patient, including an instructional videotape for home review.[13]

Discussion

Patients afflicted with HPS can be successfully managed on an outpatient basis for osseointegration treatment. Because no other patient with HPS has been reported to have had Brånemark implants placed, the osseous response to the titanium implants is of significant interest. Since Hermansky and

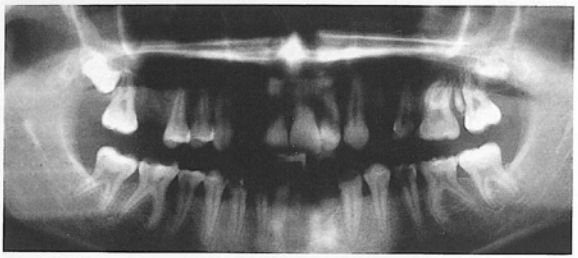

Fig 1 Panoramic radiograph of the healed mandible 3 months following emergency treatment.

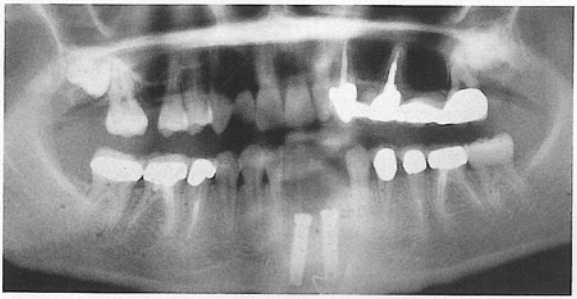

Fig 2 Panoramic radiograph illustrating the position of two Brånemark implants immediately following placement. Note that the mandibular-fracture wires are still in place.

sion, the patient was draped for surgery in the standard manner. Following the Brånemark protocol for osseointegration, two 15 × 3.75 mm implants were placed in the edentulous anterior area of the mandible (Fig 2). Because of a large mesial osseous defect, the left central incisor was extracted at the time of the implant placement. At the time of implant placement, Periotest values (PTVs) were recorded for each implant. Six individual readings were made for each implant (Table 1). The Periotest instrument (Siemens, Bensheim, Germany), originally designed to record the stability of teeth, has been used to record implant stabilization in comparative studies analyzing differences between stage 1 and stage 2 and follow-up in patients treated with Brånemark implants.[10–12] The mucosa was closed with Vicryl (Johnson & Johnson, New Brunswick, NJ) sutures. The patient clotted normally and had no postoperative bleeding.

Medications used postoperatively included acetaminophen with codeine (Tylenol 3, McNeil Consumer Products, Fort Washington, PA), as well as erythromycin 250 mg four times daily for 10 days. Peridex (Proctor & Gamble, Cincinnati, OH) was also prescribed twice daily. Family circumstances delayed the uncovering procedure for 8 months. The patient was again transfused with platelets prior to the uncovering as a precaution to assure hemostasis.

Stage 2 Osseointegration Surgery. Crestal and labial releasing incisions were made and flaps were raised to uncover the edentulous area directly above the implants. Bone overgrowth was excessive in the area of the right central incisor and covered a portion of the cover screw for the implant in the right lateral incisor area (Fig 3). Careful removal of the excess bone allowed the cover screws to be removed (Fig 4). Two 3-mm titanium EsthetiCone abutments (Nobelpharma USA, Chicago, IL) were fastened to the implants using a torque force of 20 Ncm. At that time, Periotest readings were made on the abutments. The PTVs at stage 2 surgery were generally lower than those recorded at stage 1 surgery, indicating increased stability of the implant, presumably the

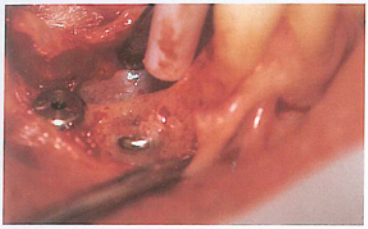

Fig 3 Eight months following implant placement, stage 2 surgery reveals significant bone growth over the cover screw of the implant in the position of the mandibular right central and lateral incisors.

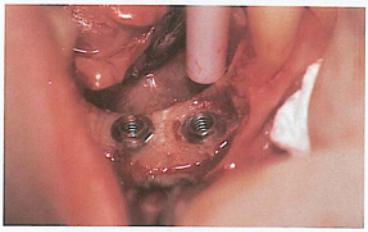

Fig 4 Careful removal of the overlying bone permits clear access to the implants.

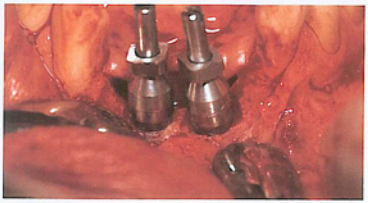

Fig 5 Reduced impression copings become the substructure for the conversion prosthesis.

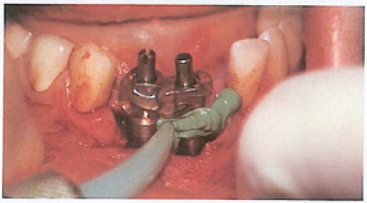

Fig 6 Light-cured Triad gel is used to connect impression copings prior to the master impression using a vinyl polysiloxane impression material.

Table 1 Periotest Values for Mandibular Implants

Implant location	Time of measurement: Stage 1 surgery	Stage 2 surgery	3 months
Right central incisor	−4.8	−5.1	−5.3
Right lateral incisor	−2.2	−5.4	−5.5

result of additional deposition of cortical bone (Table 1). Impression copings were cut in half and the lower half was secured on the abutments with a guidepin to 10 Ncm (Fig 5). Using the teeth from the provisional removable partial denture, a conversion prosthesis was fabricated. Full-size impression copings were connected using light-cured Triad gel (Dentsply/York Division, York, PA) (Fig 6). A master impression, using elastic impression material, was made while the flaps were open. Following removal of the impression, the surgical site was cleansed to assure that no impression material remained. The conversion prosthesis was fastened to the titanium abutments with gold flathead screws tightened to 10 Ncm (Fig 7) and the flap sutured against the temporary prosthetic restoration.

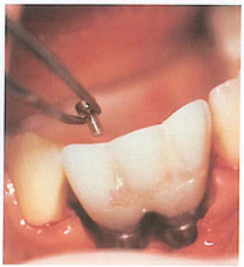

Fig 7 Gold flathead screws are used to fasten the conversion prosthesis.

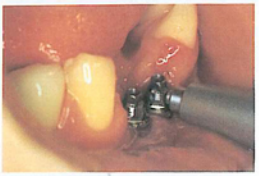

Fig 12 Periotest instrument used to record values 3 months after placement of the tissue-integrated prosthesis.

Pudlak described pigmented macrophages in the bone marrow and the hemorrhagic diathesis associated with prolonged bleeding time, the use of osseointegrated implants penetrating the bone marrow is of clinical and scientific value. Clinical evidence obtained from this treatment procedure demonstrates that bone, even with patients having Hermansky-Pudlak syndrome, can heal with a normal clinical appearance, as evidenced by the bone growth over the cover screws during the 8-month healing period.

Clinical management of patients with HPS must be a coordinated effort between the hematologist and osseointegration team in using platelet transfusions prior to stage 1 or stage 2 osseointegration surgery. With this bleeding disorder, the use of a platelet transfusion minimizes the risk of prolonged postoperative bleeding.

Conclusion

The long-term results of osseointegration for HPS are unknown. However, this early clinical evidence appears to suggest that osseointegration will remain stable for the long term. Using clinical measurements, such as provided by the Periotest values, continued clinical observation of this particular patient is required to confirm the hypothesis that osseointegration can continue to dynamically improve the stability of the bone-implant interface. □

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Résumé

Implants ostéointégrés chez une patiente atteinte du syndrome de Hermansky-Pudlak: rapport d'un cas

L'édentation partielle chez une patiente de 16 ans atteinte du syndrome de Hermansky-Pudlak fut traitée à l'aide de la technique d'ostéointégration selon Branemark. Deux implants de 15 × 3.75 mm furent placés à la région antérieure de la mandibule. La mobilité des deux implants fut testée à l'aide du Periotest lors du placement des implants, lors des connection des piliers et trois mois après la pose de la prothèse. Les résultats de cette étude démontrèrent que le procédé d'ostéointegration selon Branemark peut etre utilisé dans le traitement prothétique de patients atteints de problèmes de plaquettes affectant la coagulation.

Zusammenfassung

Osseointegrierte Implantate in Verbindung mit Hermansky-Pudlak-Syndrom: Ein Fallbericht

Eine teilbezahnte 16 Jahre alte Patientin mit Hermansky-Pudlak-Syndrom wurde mit Branemark-Implantaten behandelt. Zwei 15 × 3,75 mm Implantate wurden in der Unterkieferfront eingebracht. Mit Hilfe des Periotests wurde die Beweglichkeit der beiden Implantate intraoperativ, zum Zeitpunkt der Implantateröffnung und 3 Monate nach Eingliederung der Suprakonstruktion bestimmt. Die Ergebnisse der vorliegenden Studie zeigen, daß Patienten mit einer die Blutgerinnung beeinträchtigenden Thrombozytenfunktionsstörung mit Branemark-Implantaten versorgt werden können.

Resumen

Implantes oseointegrados en un paciente con Síndrome de Hermansky-Pudlak: Reporte de caso

Se trato el edentulismo parcial de una paciente de 16 años afectada por el Síndrome de Hermansky-Pudlak, por medio de implantes oseointegrados tipo Brånemark. Se colocaron dos implantes de 15 × 3,75 mm en la región anterior de la mandíbula. La movilidad de los implantes fue determinada por medio del Periotest en el momento de colocar los implantes, al conectar el pilar (componente transepitelial), y a los 3 meses luego de colocar la prótesis final. Los resultados de este estudio demuestran que el procedimiento de oseointegración Brånemark puede ser utilizado para trafar pacientes con desórdenes plaquetarios que afectan el mecanismo de coagulación.