Prosthetic management of Albers-Schönberg disease with osteomyelitis: A 10-year follow-up case report

Catherine Millet, DDS, PhD
Department of Prosthodontics, Faculty of Dentistry, Lyon 1 University, Lyon, France; Oral Manifestations of Rare Diseases Center, Hospices Civils de Lyon, Lyon, France.

Maxime Ducret, DDS, PhD
Department of Prosthodontics, Faculty of Dentistry, Lyon 1 University, Lyon, France; Oral Manifestations of Rare Diseases Center, Hospices Civils de Lyon, Lyon, France.

Arnaud Lafon, DDS, PhD
Department of Oral Surgery, Faculty of Dentistry, Lyon 1 University, Lyon, France; Oral Manifestations of Rare Diseases Center, Hospices Civils de Lyon, Lyon, France.

Correspondence to: Dr Catherine Millet, Department of Prosthodontics, Lyon 1 University, Faculty of Dentistry, 11 Rue Guillaume Paradin, 69372 Lyon Cedex 08, France. Phone: +33 4 72 41 39 77, Fax: +33 4 72 41 39 99, Email: catherine.millet@univ-lyon1.fr

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ABSTRACT

Albers-Schönberg disease is a rare bone syndrome characterized by increased bone density and infectious complications after dental extraction or minor surgery. The prosthodontic management of such edentulous patients with osteomyelitis is very challenging and requires special strategies due to a high risk of failure and worsening of the condition. This clinical report describes the
rehabilitation of a 31-year-old edentulous woman presenting with Albers-Schönberg disease and secondary chronic osteomyelitis, maxillary hypoplasia, compromised oral conditions, temporomandibular disorders, and psychologic distress. The treatment included mandibulectomy and removable prostheses. A crucial element for the successful long-term treatment and quality of life improvement observed in this patient was the 1-year transitional phase with interim dentures and frequent follow-up appointments. The complications and management proposed during a 10-year follow-up are presented. *Int J Prosthodont* 2022. doi: 10.11607/ijp.8255

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Osteopetrosis encompasses a clinically heterogeneous group of rare diseases characterized by increased bone mineral density due to impaired osteoclast activity. To date, mutations in at least 10 genes have been identified as responsible. The disorder can lead to skeletal deformation, dental abnormalities, and delayed wound healing that can affect the quality of life. The adult type, also called Albers-Schönberg disease, is the most common form of osteopetrosis with an incidence of 1:20,000, and affects men and women equally. This autosomal dominant type, mostly due to mutations in the chloride channel 7 (CLCN7) gene, presents with variable symptomatology. It is usually characterized by osteosclerosis with marrow spaces filled with necrotic calcified cartilage, associated with empty lacunae and plugged Haversian canals. These alterations in bone microstructure can explain the general complications observed such as bone pain, optic and auditory nerve palsies, slightly altered long bones, growth retardation, and high prevalence of fractures after minor shocks. The encroachment of bone marrow, which leads to reduced
hemopoiesis, increases the susceptibility to hemorrhage and infectious complications. The head manifestations are characterized by a high susceptibility to jaw fractures and an increased risk of mandibular osteomyelitis, which concerns about 10% of the patients after dental infection, tooth extraction, and oral procedures. This complication, due to a poor vascularization and an impairment of white blood cell function, often requires surgical debridement and resection. Different dental manifestations such as agenesis, delayed eruption, enamel hypoplasia, and dentin defects, are common. Additionally, jaw deformities, mandibular prognathism, and severe malocclusion may occur. Diagnosis is generally made after accidental fractures, radiological examinations revealing osteosclerosis with the appearance of endobones, delayed healing procedures, anemia, and hepato-splenomegaly due to extramedullary hematopoiesis. Genetic studies and bone biopsies are not essential to confirm the diagnosis. The dentist is often the first practitioner to suspect the disease during routine imaging or oral examination. Such a suspicion should encourage the practitioner to refer the patient to a specialized medical team.

Prosthodontic management of these patients using dental implants presents a high risk of failure due to the effects of the disease on the healing process. For this reason, very few clinical studies present implant rehabilitation in such patients. In the absence of scientific evidence relating to dental implants, the management of these patients is challenging due to the lack of standardized strategies. In the same way, orthognathic surgery is usually avoided to minimize complications. Thus, malocclusions and skeletal deformities, associated with the risk of osteomyelitis, discourage many practitioners who refuse to treat affected patients, leaving them in great psychological distress. In the absence of therapeutic guidelines, the purpose of this clinical report was to share our clinical experience related to the prosthetic management and follow-up of a young edentulous patient with Albers-Schönberg disease, chronic osteomyelitis, and severely compromised oral conditions. The successful prosthetic outcome was based on surgical treatments,
including mandibulectomy, combined with a long transitional phase during which interim treatment dentures, that were fully polished to prevent soft tissue irritation, were placed.

**Case history**

A 31-year-old woman suffering from esthetics and chewing problems was referred to the author's department after prosthetic treatment refusal by several clinicians due to a complex situation. The patient had a short stature, frontal bossing, and a deficient midface. She had previously been diagnosed with Albers-Schönberg disease based on hepato-splenomegaly, elevated serum levels of acid phosphatase, radiographic features of generalized osteosclerosis, and multiple fractures. Her past medical history revealed a cranial fracture that required a tracheostomy procedure at the age of 2 years, and fractures of the right femur, the right radius, and the tibias during adolescence. She claimed that her father and a cousin also carry this condition, but genetic examinations were lacking. She underwent septoplasty surgery and was under medical care for visual and hearing impairments. The patient presented with low self-esteem, poor quality of life, insomnia, and anxiety. She was living alone for the last 8 years and had a prescription of antidepressant drug. She had no known allergies, and was a moderate smoker. She reported multiple dental agenesis and several extractions performed over the last twenty years. The last extractions were performed the previous year and concerned all maxillary anterior teeth and the right mandibular second molar. The extractions occurred under local anesthesia and were associated with secondary chronic mandibular osteomyelitis despite several debridements and antimicrobial therapy. A sequestrectomy of the right mandible was performed 6 months earlier in the oral and maxillofacial surgery department under general anesthesia. The patient did not wear any previous dentures.
The clinical examination revealed a mandibular deviation to the right side, prognathic aspect, limited mouth opening amplitude (20 mm), altered mandibular movement, left temporomandibular joint (TMJ) clicking with mild pain during opening, stiffness in the periauricular region, and tenderness of the masticatory muscles. The patient presented with full maxillary edentulism and subtotal mandibular edentulism with the persistence of the 2 canines, which were affected by carious lesions near the pulp. An enlarged tongue, a small, narrow, and retrognathic maxilla (measuring 3.5 cm in length and 2.5 cm in width), with a class III skeletal relationship were noted. In the mandible, a lateral bone exposure area was observed on the left posterior side. Panoramic radiographic analysis and computed tomography (CT) images showed a severe maxillary hypoplasia, mandibular asymmetry, and osteolytic lesions in the mandibular body. Figure 1 illustrates the clinical and radiological states at the first visit. The proposed treatment was the placement of 2 crowns on the canines, and removable prostheses due to chronic osteomyelitis and the increased risk of implant failure. An alternative prosthetic option, consisting of a mandibular complete overdenture, was refused by the patient. The treatment plan comprised a transitional phase with interim treatment dentures permitting a neuro-musculo-articular conditioning and the follow-up of the osteomyelitis, prior to a second phase of fabrication of permanent prostheses.

After endodontic treatment, the canines were transitionally restored with resin composite. Despite the limited mouth opening, preliminary impressions were obtained using pediatric stock trays with irreversible hydrocolloid (Jeltrate; Dentsply Caulk). Due to the palatal anatomy, the use of mini custom tray for a high-quality impression was essential to increase the stability of the maxillary prosthesis. Border molding was done using polyether material (Permadyne; 3M) and the final impressions were made using polysulfide (Permlastic; Kerr Corp). The casts were articulated onto a semi-adjustable articulator (Quick Master; Fag-Dentaire) with a facebow and a centric
relation (CR) occlusal record using bimanual manipulation. Because of jaw disproportion and mandibular deviation, it was decided to realize only an anterior teeth arrangement with flat occlusion rims (Fig 2) rather than occluding teeth. After esthetic approval, the protheses without posterior teeth were polymerized and remounted on the articulator to eliminate interferences. The cameo and intaglio surfaces were polished to minimize trauma on the supporting tissues, and the dentures were delivered with instructions concerning their maintenance and the diet (Fig 3). The patient was asked to come back the following week for control, and then recalled every 3 weeks.

After 3 months, the proposed occlusal vertical dimension (OVD) was well accepted. The patient was pleased with esthetics, comfort, psychological well-being, and the mastication of soft food. An improvement in maximal mouth opening of 5 mm without pain was also noted. Stiffness in the periauricular region, TMJ clicking, and functional limitation were significantly reduced. The bone exposure was decreased but a large right jugal abscess with suppuration was observed. A panoramic radiograph showed an irregular radiolucency indicating a sequestrum formation in the right mandible, which was confirmed by CT images. The patient was treated with antibiotic therapy using amoxicillin/clavulanic acid and was referred to the department of maxillofacial surgery for debridement and resection of the affected bone. The operative procedure under general anesthesia consisted of a right segmental mandibulectomy with the placement of a reconstructive titanium plate (Fig 4). The healing was uneventful and the patient remained asymptomatic at the 1-month follow-up. Some adjustments on the acrylic occlusal rims were made due to further mandibular rotation toward the right side after the resective surgery. Six months after the mandibulectomy, no TMJ problem or muscle tenderness was detected clinically. The patient felt comfortable with the interim prostheses and her oral hygiene was satisfactory. Moreover, there were no further recurrences, the osteomyelitis appeared to be resolved. Thus, the final phase of prosthodontic rehabilitation could be conducted.
The mandibular canines were restored with metal-ceramic crowns. Due to severe mandibular deviation and a persistent instability of the supporting mandibular bone and mucosa, the fabrication of an acrylic resin-based removable partial denture (RPD) was preferred to a metal alloy-based RPD. This choice was made because acrylic resin-based RPDs are more easily altered in case of changes in the mandibular oral tissue. The border molding and final impression procedures were the same as the previous ones. The casts were mounted on articulator, at the same OVD as the interim prostheses, using facebow and CR occlusal record. Semi-anatomic acrylic resin teeth (SR Orthotyp DCL; Ivoclar Vivadent AG) were selected to minimize trauma. Because of the severe underdeveloped maxilla and mandibulectomy, the second molars were excluded from the denture’s designs. In order to compensate the mandibular deviation toward the defect side and to reduce the leverage forces, unilateral posterior cross-bite arrangement was tried. However, the patient perceived cheek biting and restricted tongue movement. Hence, the maxillary right posterior teeth were arranged buccally in relation to the crest and the mandibular teeth were positioned over the center of the crest’s ridge. The dentures with a bilateral balanced occlusion were delivered 7 months after the partial mandibulectomy (Fig 5). The adaptation of the denture bases was controlled using pressure-indicating media and the appropriate adjustments were carefully made. During periodic recalls, the patient adapted to the prostheses easily. She reported improved appearance, quality of live, confidence, and resocialization. Eight months later, the patient presented with a right submandibular abscess, which was treated with a surgical drainage under general anesthesia and adequate antibiotherapy. Subsequently, her condition deteriorated and a sero-purulent discharge was noted. X-ray examination revealed a fracture in a screw retaining the titanium plate (Fig 6). The patient was admitted to hospital for an ablation of the osteosynthesis devices and did not undergo further bone reconstruction. Postoperative healing was uneventful with
minimal pain. At the 6-month follow-up assessment, the patient had no complaints and the mandibular RPD was relined in the laboratory.

The patient was followed every 6 months for 2 years, and then on a yearly basis for maintenance visits. During the 10 years following the delivery of the dentures, no sign of relapse or infectious problems were observed (Fig 7). At recall visits, the patient had good oral hygiene, and remained satisfied with esthetics, function, and comfort. She reported no disturbance in the joint, no arthralgia during opening, and local myalgia and tenderness were absent. No TMJ clicking nor limitation in mouth opening were observed. Only a midline fracture of the mandibular RPD, related to an accidental dropping during its cleaning, required a repair 10 years after its insertion. A timeline of the patient’s care is shown in Fig 8.

Discussion

The prosthodontic management of an edentulous patient with Albers-Schönberg disease complicated by chronic osteomyelitis, instability of the oral bone-support, and impaired posterior guidance is particularly rare and challenging due to the lack of standardized procedure. To our knowledge, no treatment guideline has been reported for such situations. The sharing of experiences, particularly when they are complex, are thus very valuable. In this case, implant placement and surgical correction for maxillary hypoplasia were excluded to prevent the complications commonly reported.\textsuperscript{8,13,18} Indeed, chronic osteomyelitis contraindicated the implant option, leaving removable prostheses as the only solution. After interrupting bone resection, the preservation of both mandibular canines as abutment teeth proved to be of great benefit for the success of the prosthetic rehabilitation. In fact, the canines minimized the off-vertical forces transmitted to the underlying mucosa ensuring minimal trauma on the affected tissues. Moreover, meticulous attention was paid to each step of the prosthetic phases, including impressions, teeth
arrangement, occlusal equilibration, and polishing, to prevent soft tissue irritation, a known aggravating factor for the development of osteomyelitis. This is in accordance with the recommendations in head and neck cancer irradiated patients to ensure minimal trauma to the mucosa, thus decreasing the risk of osteonecrosis. 19,20

In this report, the two-stage prosthodontic management was essential. During the transitional phase, the use of interim treatment dentures allowed to progressively follow the evolution of the osteomucosal tissues subjected to chronic osteomyelitis and improve the postsurgical management. The adaptations were made by intaglio surface adjustments and/or the placement of tissue conditioner in relation to the bone exposure areas. The interim dentures also allowed a long temporization, which is absolutely necessary for the long and difficult management of osteomyelitis. 5,13 Relapses are common because the bacteria remain hidden in the bone and the decreased marrow vascularity reduces the effectiveness of antibiotics. 5 In this case, a mandibulectomy was necessary and resulted in mandibular deviation and rotation of occlusal plane which are not uncommon after the loss of mandibular continuity. 17 Also, a second advantage of the placement of stable interim treatment dentures with posterior rims was the easy management of the occlusion after mandibulectomy. In the context of temporomandibular disorders, another advantage of interim dentures without posterior teeth is a gradual adaptation and coordination of the neuro-musculo-articular system due to the absence of occlusal instability and interference during mandibular movements. 21,22 This strategy is similar to that used for the rehabilitation of patients lacking condylar guidance. 16

There are some limitations to the proposed rehabilitation. The transitional phase was chairside time-consuming, since many appointments were complicated or postponed because the patient suffered from localized bone pain with sometimes jugal abscesses and suppuration. However, the final prosthetic rehabilitation was easy to manage. Furthermore, the second molars
were excluded from the definitive prostheses. Nevertheless, studies have shown good masticatory efficiency and oral health-related quality of life in complete or partial denture wearers with reduced dental arches. Fortunately, the patient herein was able to eat a wide range of foods despite the lack of mandibular reconstruction after osteosynthesis system removal.

**Conclusion**

In the absence of treatment guidelines for Albers-Schönberg disease with chronic osteomyelitis and complex conditions, this case report highlights that a favorable treatment outcome can be achieved by combining existing procedures to adapt to the situation. In this particular clinical context, an adaptation period with interim treatment dentures in conjunction with mandibulectomy was a crucial element for the successful long-term treatment.

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**References**


**Figure Legends**

**Fig 1** Initial situation. (a) Maxillary hypoplasia. (b) Mandibular left bone exposure area. (c) Panoramic radiograph. (d) Computed tomography (CT) images. Note the mandibular osteolytic lesions and severe class III jaw relationship.

**Fig 2** Procedure for interim treatment dentures. (a) Mini maxillary master cast and mandibular deviation toward the right side. (b) Anterior teeth arrangement and posterior occlusal rims.
Fig 3 Interim treatment dentures. (a) Intraoral view. (b) Smile appearance.

Fig 4 Panoramic radiograph showing titanium plate after mandibulectomy.

Fig 5 Definitive prostheses. (a) Occlusal view. b) Atypical arrangement on the right side due to mandibulectomy. (c) Frontal view post-treatment with two ceramic crowns cemented and the prostheses in occlusion.
Fig 6 Panoramic radiograph showing fractured anterior screw before ablation of osteosynthesis device.

Fig 7 Panoramic radiograph 10 years post-prostheses insertion. The continuity of the mandible was lost 1 year after denture delivery.

Fig 8 Representative timeline of the prosthodontic treatment and follow-up. TMD: temporomandibular disorders; RPD: removable partial denture.