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Prosthodontic Treatment of a Class IV ACP PDI Patient, with Langerhans Cell Histiocytosis (LCH)

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Abstract

Langerhans cell histiocytosis (LCH) is a disease of unknown etiology with a frustrating and unpredictable course. Surviving adult patients suffering from the multisystem type of the disease present with problems in most organs. This article presents the oral rehabilitation of a 28-year-old patient, with multisystem sequelae that included the oral cavity, classifying him as a Class IV American College of Prosthodontists Prosthodontic Diagnostic Index patient. A 5-year course of treatment is analyzed, starting from merely replacing missing teeth with a removable partial denture. The second stage of prosthetic rehabilitation included replacement of the removable prosthesis with fixed partial dentures. The final and most important aspect of treatment was the 2-year follow-up, when the patient presented with no problems or adverse effects. The purpose of this presentation is to offer an insight to prosthodontic treatment possibilities for patients suffering from multisystem LCH and to show the value of a "team approach" to achieving a positive outcome.

"I am glad you are here and wish you all a happy stay. We are here to try and destroy this disease that children around the world have. If we can't beat it nothing else can but we have to try. And try."[1]

That was the message Nikolas, then age 12 (1993), wrote for the participants of the fifth meeting of the symposium named after him, focusing on the fight against Langerhans cell histiocytostis (LCH).[2,3] Any patient with such strength and ability to defy his medical disability deserved the best effort from the rehabilitating team as well.

Background

The patient presented for initial treatment at age 23 (2005). His dental problems were minor, including fractures on polymer filling material on anterior teeth and a recurrent fracture of an artificial denture tooth from his removable partial denture (RPD), used for the rehabilitation of the upper jaw. The patient's medical history became a major concern[1] and is summarized in Table 1. From 2005 to 2008 the patient presented in a relatively stable condition as an outgoing, semiindependent adult with cerebral ataxia, well-controlled hypothalamic-pituitary-adrenal dysfunction and epilepsy, and was on therapy for osteoporosis. He was taking medications to control epilepsy with very good results (no incidents of seizures for the past 10 years). He was also on medication to control a minor bladder problem. Osteoporosis medication was the most important medication to be considered, as it could possibly affect treatment planning for prosthetic rehabilitation. The patient was on oral biphosphonates (Fosamax, Merck & Co., Whitehouse Station, NJ) from 1994 to 2005. The dose was gradually reduced as the patient's condition showed signs of improvement. Since 2005 he has been taking a slow-release biphosphonate (Boniva 150 mg/month, GlaxoSmithKline & Roche, Basel, Switzerland). His latest bone mass density counts showed constant improvement (DEXA test results: Lumbar spine: T score $= -1.7$; Z score $= -0.9$. Total hip: T score $= -2.4$; Z score $= -1.9$).

The patient's history reflects the frustrating and unpredictable course of LCH.[4,5] The cause of the disease is unknown, although scientists and clinicians are trying to clarify its pathogenesis.[6-8] Initially, a syndrome was described by and named after Hand,[9] Schuller,[10] and Christian.[11] A few years later, a similar syndrome was described by and named after Letterer[12] and Siwe.[13] Lichtenstein[14] showed that both syndromes were expressions of a greater disease entity. It was named Histiocytosis X, because it involves the histiocyte, and its etiology is unknown.

Table 1 The patient's course of disease from birth to adulthood

Age	Condition	Treatment
2 months	Letterer-Siwe disease	
2 months	Histiocytosis X	Surgery to the mouth
1 to 2 years	Skin and lung Langerhans cell histiocytosis in regression	Short dose of corticosteroids
2 years	LCH scalp disease, Lytic bone lesions	Topical nitrogen mustard and prednisone for 6 months
6 to 7 years	Cerebellar dysfunction	Six courses of oral etoposide for 6 months
10 years	Raised intracranial pressure/ stenosis of the aqueduct of Sylvius	Fistula between third and fourth ventricles
11 to 16 years	Early puberty, psychological difficulties, growth-hormone deficiency	Growth-hormone therapy
16 years	Diabetes insipidus, osteoporosis, epilepsy	Therapy for osteoporosis and epilepsy

LCH affects most organs in the human body,[15,16] including bone, skin, lymph nodes, the thymus, ears, bone marrow, peripheral blood, liver, spleen, lungs, the endocrine system, and the gastrointestinal tract. Oral cavity involvement is common, presenting with ulceration of the mucosa. The posterior part of the palatal ridges in the upper molar region is often broadened. The mucosa in these areas appears granular if infiltration by the disease is present.[15] The prognosis differs between patients having single-system and multisystem disease. The former suffer minimal long-term sequelae.[17] The latter present long-term problems[18,19] including, but not limited to, growth hormone deficiency, diabetes insipidus, cerebral ataxia, loss of dentition, orthopedic problems, and pulmonary fibrosis. Many of those manifestations were present in the patient (Nikolas) described in this report.

Dental condition

Intraorally, Nikolas presented a normal mandible with well-developed dentition. On the maxillary arch and dentition (Fig 1) the signs of previous surgical intervention (at 2 months old) were obvious, with extensive scarring of the soft tissues, bone loss at the premolar region, missing teeth (maxillary right canine and second premolar and maxillary left premolars) in the same region and abnormal position of the molars. There were a few minor teeth lesions (maxillary right central incisor and left first molar), a missing filling on the maxillary right first premolar and a retained primary canine, ground to gum level but vital, in the position of the maxillary right canine.

The periodontal condition of the patient was very good (Fig 2), with pocket depths less than 3 mm and bleeding on probing (BOP) of 40%. These results were considered excellent, especially since he was wearing an RPD and because he was dependent on his family for oral hygiene procedures. The dedication shown by his family members on this matter throughout the treatment period (close to 3 years) played a major role on the decision process for the final rehabilitation.

Occlusal analysis revealed marginal anterior Class III position (Fig 2) with the incisors at an edge-to-edge position holding a repeatable occlusal vertical dimension. Due to malpositioned maxillary molars, the patient had a bilateral posterior open bite (Figs 3 and 4), excessive among the first and fourth quadrants. The posterior teeth, although not occluding, were at a cross-bite position. The patient was wearing a maxillary RPD mainly for esthetic reasons (replacement of missing teeth), as it was leaving the posterior open bite untreated. The acquired maxillary bone defects, the occlusal destruction, and the patient's medical condition classify him as a Class IV patient according to the American College of Prosthodontists Prosthodontic Diagnostic Index (ACP PDI).

Treatment considerations

Many important limiting factors were considered during treatment planning:

- The patient's "enigmatic" course of disease, although stable for some years;
- The presence of osteoporosis;
- The hard tissue and occlusal destruction due to surgery at infancy; and
- The absence of prior knowledge due to the rareness of the disease, especially in surviving adults.

Two positive and decisive factors were the family's attention, which had led to a very well socially educated patient, including their attention to oral hygiene and the patient's cooperation with dental treatment.

The bare necessities

Considering the limiting factors, including the patient's medical condition primarily, but also the long-term use of biphosphonates (although per os), treatment with osseointegrated implants was excluded as a treatment option. The presence of healthy abutment teeth at good "strategic" positions (Fig 5) led to the consideration of fixed prostheses, a plan that was aborted due to the patient's family's hesitation as a result of the absence of prior knowledge for long-term results. The final plan was a removable partial overdenture (Fig 6) to address the main problems of missing teeth, lack of posterior occlusion, and esthetics. To leave bone undisturbed, no endodontic treatment was planned, mainly because of the lack of knowledge on what causes the disease to regress, or even what can cause a new outburst. The framework of the RPD included metal overlays that provided the needed occlusal contacts with the lower teeth. Care was taken to create a sturdy framework (Fig 7) to support the artificial teeth at proper esthetic positions, without fracturing. Finally, the maxillary anterior teeth were treated with

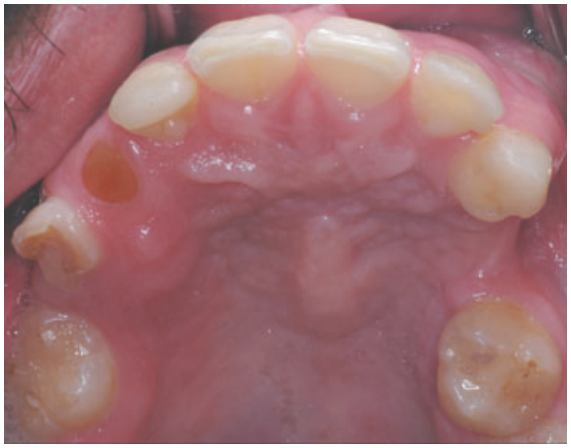

Figure 1 Occlusal view of the patient's maxillary arch and dentition.

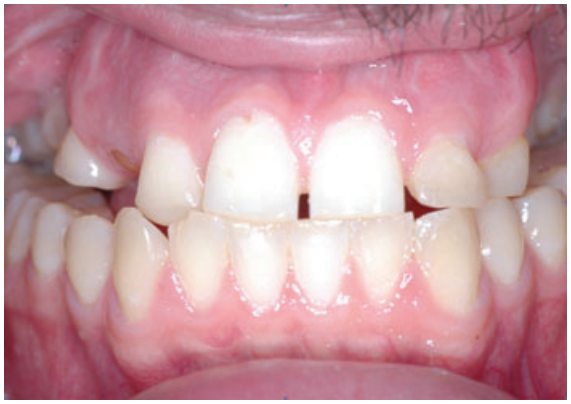

Figure 2 Facial view of maximum intercuspation position.

polymer fillings mainly for esthetic reasons. The patient was given fluoride treatment to reduce the risk for decay. He was able to insert and remove the RPD himself, but his oral hygiene was supervised by his family members. He was placed on a 4-month recall schedule, which was followed for the next 1.5 years.

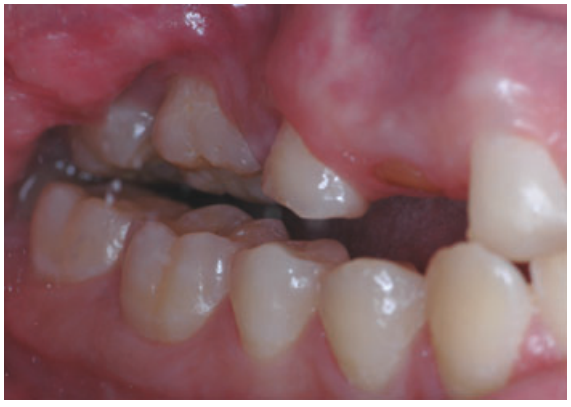

Figure 3 Right lateral view showing posterior "open bite."

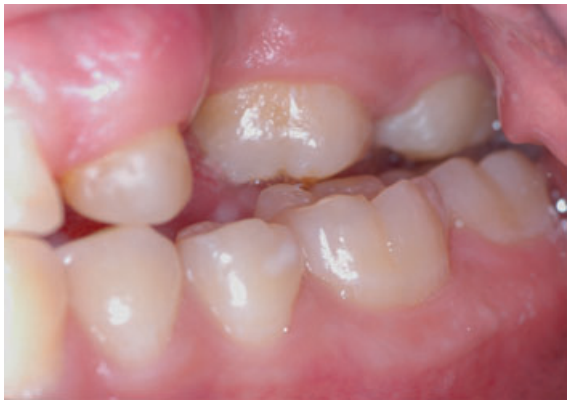

Figure 4 Left lateral view showing posterior "open bite."

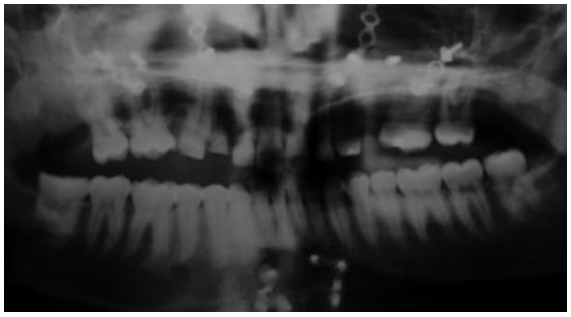

Figure 5 The patient's initial panoramic radiograph.

Figure 8 shows the patient's condition at the 1.5-year recall. There was a minor fracture at the acrylic over the artificial teeth, and two of the fillings were replaced once. No caries or tooth mobility was detected, and the periodontal condition was even improved through the years. The patient's coordination was very good—he could place and remove the RPD by himself. He was able to have an ordinary diet and experienced no discomfort.

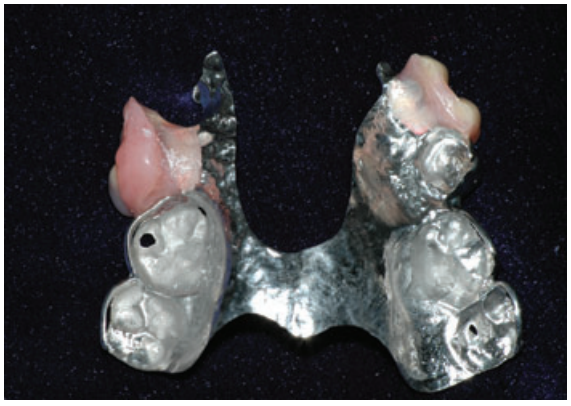

Figure 6 The tissue side of the patient's RPD, showing the metal occlusal coverage.

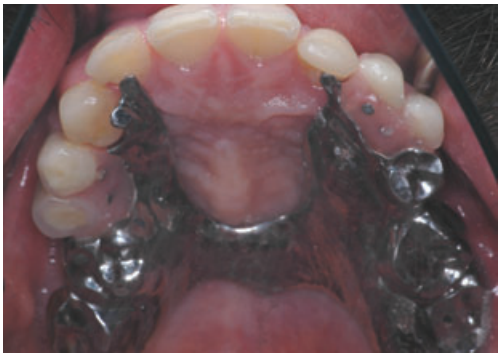

Figure 7 Occlusal view of the maxillary arch with the RPD in place.

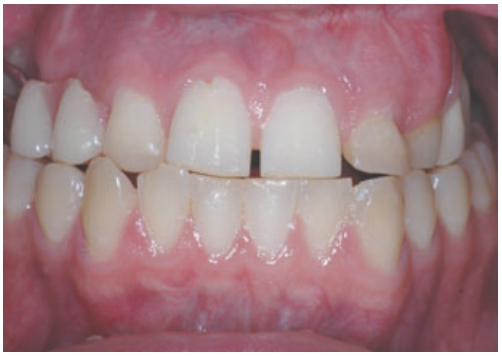

Figure 8 Facial view of maximum intercuspation position, with the RPD in place, at the 1.5-year recall appointment.

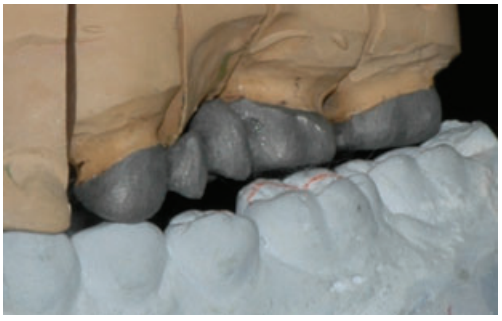

Figure 9 Left lateral view showing the metal framework's design.

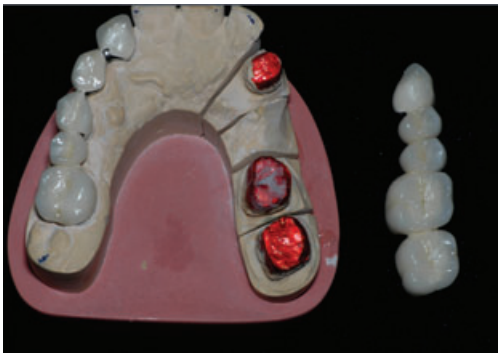

Figure 10 The occlusal tables of the posterior fixed prostheses were enlarged to achieve occlusal contacts.

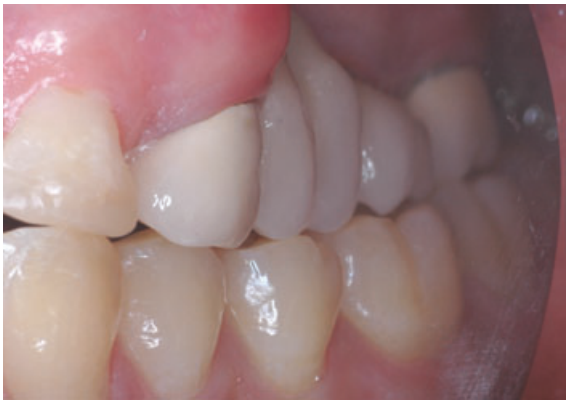

Figure 11 Left lateral view with the prosthesis in place. Only the buccal cusps of the FPD occlude with the lower teeth.

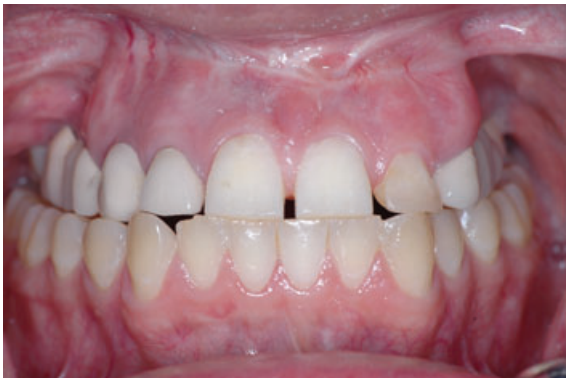

Figure 12 Facial view of maximum intercuspation position, at the 2.5-year recall appointment.

Rehabilitation

The treatment with fixed prostheses was addressed again as the patient's teeth proved to be able to withstand occlusal loading. Their periodontal environment would improve through the absence of the extensive RPD framework, and the ongoing relationship with the patient and his family, powered by mutual

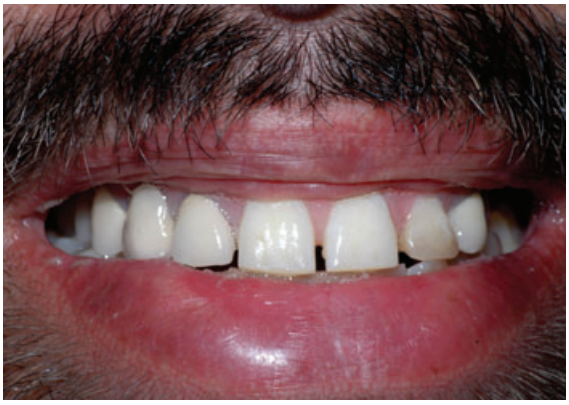

Figure 13 The patient's smile after FPDs were delivered.

respect, allowed us to perform the less conservative treatment. The aims of this treatment were to reduce the long-term risk for periodontal disease, to improve the patient's masticatory efficiency, to reduce the amount of care needed for the prosthesis (inherent to removable devices), and most importantly to improve the patient's self-esteem and mental outlook.

A diagnostic wax-up led to the visualization of the final treatment and the decision on the minimal number of abutment teeth necessary. No extractions or endodontic treatments were to be performed so the primary tooth at position 13 was going to be left untreated under the pontic. Preparation of the abutment teeth was done under local anesthesia, counting on the patient's cooperation. Margins were placed at the gingival level on anterior teeth for esthetic reasons, and supragingival for the posterior teeth to facilitate cleaning and for conservation of tooth structure. The occlusal surfaces of the posterior teeth were minimally prepared in most areas because the open bite offered adequate space. Acrylic resin interim prostheses replaced the patient's RPD.

The metal frameworks (Fig 9) were made to support the "awkward" morphology of some of the teeth, in order to achieve occlusal contacts, even at a crossbite position. The arch shape correction was evident when the definitive restorations were removed from the cast (Fig 10). Following occlusal adjustment, the restoration was finalized and definitively luted on the abutment teeth with resin-modified glass ionomer cement (Fig 11). The patient was again given regular fluoride treatment to reduce the risk for decay and continued on a 4-month recall following his initial 30-day recall. Supervised oral hygiene included brushing three times a day, and use of interproximal brushes twice a day.

Discussion

Finally, Nikolas' usual question, "Do I get my teeth today?," was answered with a "yes." Two and a half years after permanent rehabilitation (Fig 12) both the rehabilitating team and his family were cautiously optimistic. His periodontal condition was as good as always (BOP 25%), there was no mobility or other symptoms from the teeth, and there were no problems from the prostheses. More importantly, the patient's mental outlook regarding his oral condition had greatly improved and was expressed by his warm and radiant smile (Fig 13).

One successful patient by no means allows for concrete clinical decisions for patients with LCH. It shows there is potential for treatment with fixed prostheses under careful planning. Mandatory prerequisites for this, apart from early detection,[20] are:

1. Careful patient selection;
2. General health condition allowing dental treatment;
3. Good oral hygiene and home care; and
4. Short recall periods (3 to 4 months).

The decisive factor for this and similar patients[21] is a "team approach," including the patient, the family, and the rehabilitating team. It seems there are no written reports for prosthetic rehabilitation of patients with LCH, although oral manifestations are often serious, affecting the patient's medical condition and everyday life. Research regarding the oral cavity is focused on early detection of signs for the onset of LCH intraorally,[22] and prediction of oral manifestations long term.[23] An additional aim of this article was to share information possibly leading to the creation of a "case series" that will allow clinicians to draw conclusions on the best treatment modalities to be provided to patients like Nikolas, improving their quality of life.[24]

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