OPEN ACCESS
Saudi Journal of Oral and Dental Research
Abbreviated Key Title: Saudi J Oral Dent Res
ISSN 2518-1300 (Print) |ISSN 2518-1297 (Online)
Scholars Middle East Publishers, Dubai, United Arab Emirates
Journal homepage: http://scholarsmepub.com/sjodr/

Case Report

Oral Management of a Patient with Juvenile Hypophosphatasia: Ten-Year Follow-Up

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| **Received:** 01.01.2019 | **Accepted:** 08.01.2019 | **Published:** 19.01.2019
DOI:10.21276/sjodr.2019.4.1.3

Abstract

Hypophosphatasia is a rare inherited metabolic disorder characterized by a deficiency of alkaline phosphatase activity. It is characterized by a defect in bone and tooth mineralization. There are remarkable dental manifestations, mainly, the premature loss of fully-rooted deciduous teeth and delayed eruption of permanent teeth. This clinical report describes the oral management of a patient with juvenile hypophosphatasia, followed 10 years in the department of dentistry at the Rabta Hospital-Tunis-Tunisia. Removable prostheses were made to establish an acceptable masticatory function, speech, occlusion and aesthetics for the patient.

Keywords: Hypophosphatasia, Alkaline phosphatase, mineralization defect, Cementum, Early tooth loss, deciduous teeth, delayed eruption, dental prosthesis.

INTRODUCTION

Hypophosphatasia is a rare inherited metabolic disorder characterized by a deficiency of alkaline phosphatase activity, a ubiquitous enzyme found particularly in the liver, kidney and bone [1-3]. It is characterized by a defect in bone and tooth mineralization [4]. There are six clinical forms according to the age of onset: neonatal lethal, neonatal benign, infantile, juvenile, adult and odonto-hypophosphatasia [1, 4, 5, 6]. The prevalence of severe forms is estimated at 1/300000 newborns in Europe with inter-population variation [3]. The diagnosis of this condition is essentially based on biology [1, 6].

Patients with hypophosphatasia generally have bone hypomineralization problems with the development of deformities like scoliosis and rickets [1, 3, 6]. There are also remarkable dental manifestations, mainly, the premature loss of fully-rooted deciduous teeth and delayed eruption of permanent teeth [4, 5, 7].

This clinical report describes a case of juvenile hypophosphatasia followed 10 years in the department of dentistry at the Rabta Hospital-Tunis-Tunisia.

OBSERVATIONS

In 2005, a four-year-old child consulted with his parents the service of Pediatric Odontology at the Rabta University Hospital -Tunis-Tunisia. He complained of the premature loss of temporary teeth with difficulty eating and talking. The medical history revealed that the child was born from a first-degree consanguineous marriage, and that pregnancy and delivery were normal, without any special incidents. At the age of one year, the child was hospitalized for lower limb deformities and scoliosis, as well as difficulty walking (Figure-1). X-rays of the limbs showed bone deformities and mineralization's anomaly (Figure-2). The determination of alkaline phosphatase and serum calcium confirmed the diagnosis of juvenile hypophosphatasia.

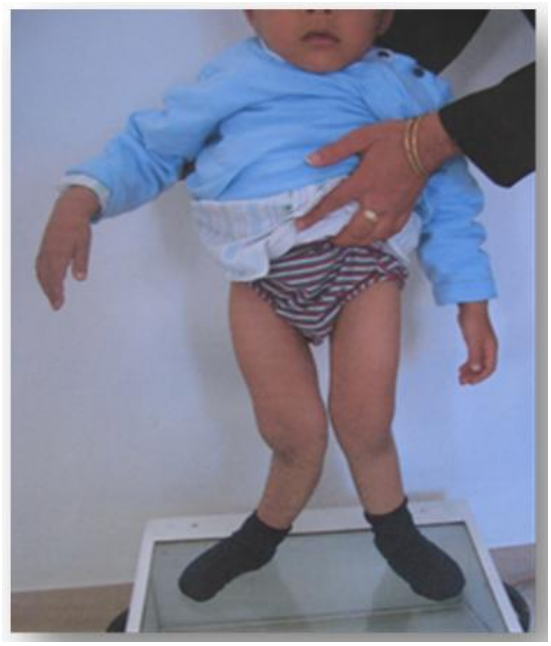

Fig-1: Lower limb deformities

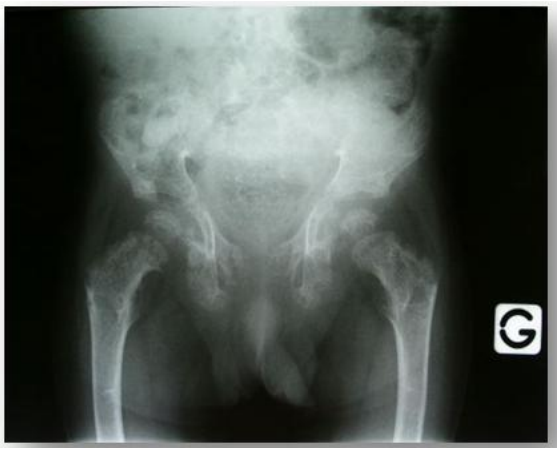

Fig-2: Bone hypomineralization

Fig-3: Water therapy

The child underwent corrective surgery for scoliosis, in France, followed by water therapy (Figure-3).

Family history showed that grandparents, parents and siblings were in good general health and did not show any signs of premature tooth loss.

Clinical examination, the day of the consultation, revealed a complete mandibular and subtotal maxillary edentulism with the persistence of the second deciduous molars (figure 4 a et b). The alveolar ridges were well formed with a normal-looking overlay mucosa.

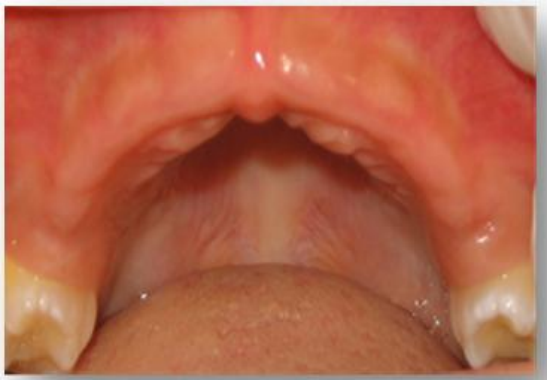

Fig-4(a): Subtotal maxillary edentulism

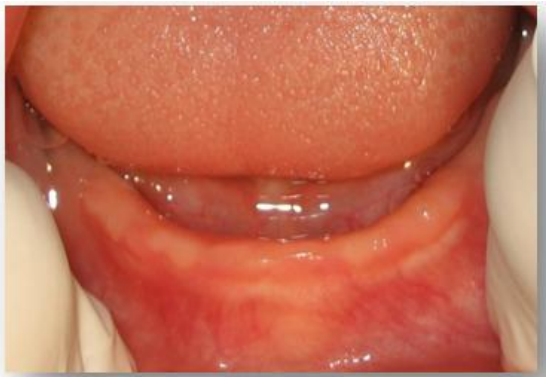

Fig-4(b): Total mandibular edentulism

The treatment planning for prosthetic rehabilitation was made after considering the age of the patient, the growth potential and the number of remaining teeth. Partial removable maxillary and total removable mandibular prostheses were made (Figure 5 (a)(b)(c))

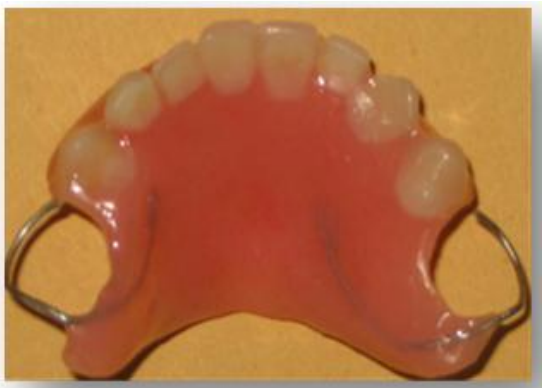

Fig-5(a): Maxillary prosthesis

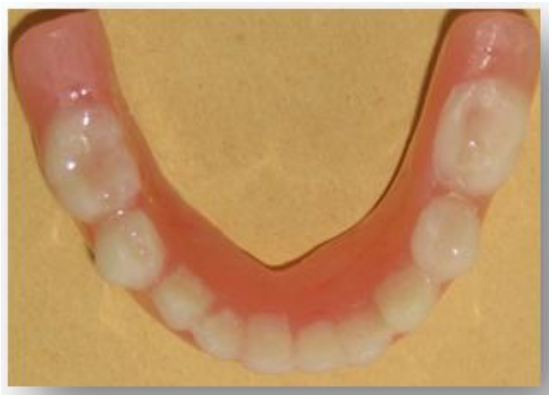

Fig-5(b): Mandibular prosthesis

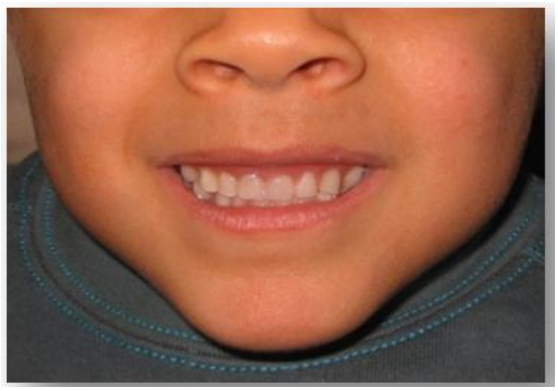

Fig-5(c): Patient new smile

The follow-up appointments were scheduled every six months but were not all respected. *A recess* in the fitting surface of the *denture* base was made whenever a tooth made its eruption, in order to maintain stability (Figure-6). These prostheses were also renewed following the growth of the patient.

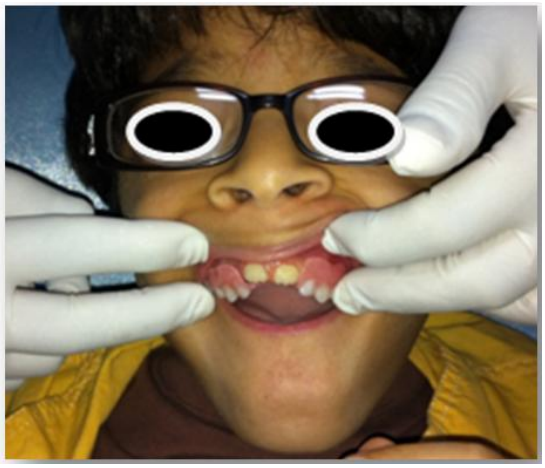

Fig-6: Prosthesis adjustments

Over the years, we noticed that the chronology of eruption of permanent teeth was not respected, and the delay of eruption was very obvious. On the panoramic radios, the alveolar bone was reduced. The dental age of the child did not correspond to his age. And finally, the pulp chambers and root canals were large (Figure-7(a) (b).

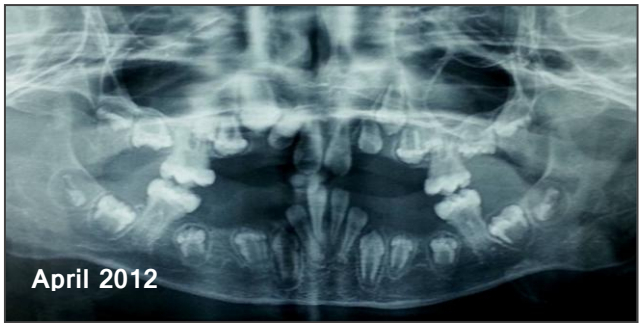

Fig-7(a): The patient was 11 years old

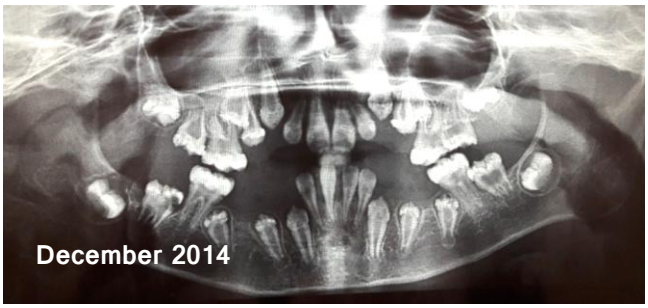

Fig-7(b): The patient was 13 years old

In 2015, the patient was fourteen years old and still followed in our service. He unfortunately had a poor oral hygiene, a deplorable periodontal condition (Figure-8(a) and he lost his old mandibular prosthesis. Added to that, the relationship with his new dentist was very difficult to establish given his critical age. Therefore, a scaling and polishing were carried out regularly, and a new non clasp removable prosthesis was made (Figure 8(b).

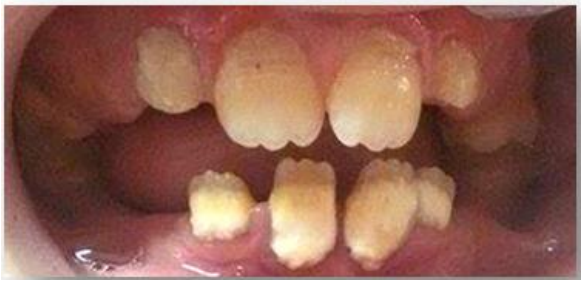

Fig-8(a): Poor oral hygiene

Fig-8(b): After scaling and polishing

DISCUSSION

Juvenile hypophosphatasia is a form of hypophosphatasia that develops after the age of six months. In this case, there are several clinical and radiological manifestations such as the appearance of rachitic deformities with hypomineralization, failure to thrive, early loss of temporary teeth without physiological root resorption, and delayed eruption [3, 5, 6, 7]. This spontaneous loss is the pathognomonic sign of hypophosphatasia [4]. It is due to an alteration of the cellular and acellular structure of the cementum [4, 5]. All these aberrations explain the multidisciplinary management of these patients but also they explain, in part, the reasons why patients consult the dentist late [3]. This is the case of our patient who consulted after having lost almost all teeth, not because

of negligence but only because there are other general problems that take precedence like respiratory failure.

For patients with hypophosphatasia, long term follow-up and reassessment are also required. Indeed, the altered surface of cement constitutes a favorable environment for bacteria. It promotes the retention of food and the accumulation of dental plaque. Therefore, there teeth are periodontally *compromised [4, 7, 8]*.

The patient's edentulism at the age of 4 had impacts on different levels: Functional, by the alteration of the masticatory coefficient and maxillofacial growth. Aesthetic, by the alteration of the smile as well as the vertical dimension of occlusion. And finally, heavy consequences on the child and his parent's psychology.

The initial edentulism guided the choice of prosthesis. The maxillary and mandibular ridges were well formed which increased the retention of removable prostheses. After the eruption of permanent teeth, the fragility of the periodontal attachment dictated the type of prosthesis. The literature suggests non-clasp prosthesis especially at the level of single rooted teeth since they are poorly anchored to the bone [7, 8].

Thanks to these conventional prostheses, a significant improvement in appearance, speech and mastication was noticed. Our patient and his parent's expectations were successfully met.

CONCLUSION

The dental practitioner play a key role in the initial diagnosis of hypophosphatasia, since the early loss of deciduous fully-rooted teeth is an almost constant-sign.

Patients with hypophosphatasia need a multidisciplinary management. Indeed, dental follow-up starts from an early age and interim removable prosthesis seems to be the best transitional solution until reaching the age of the definitive prosthesis

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