

## Outcome and Long-Term Follow-Up of a Phenylketonuria Patient After Prosthetic Treatment: a Case Report

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### Abstract

Phenylketonuria is the most common inborn error of amino acid resulting from hereditary factors. It occurs on average at a frequency of 1 in 3000 or 4000 in Turkey. If the treatment isn't achieved, this aminoacid will be toxic to the developing brain, and mental retardation may progressively develop in the untreated individuals. Also Phenylketonuria effects the skeletal and dental development.

A 10-year-old boy with phenylketonuria is presented in this case. This patient was referred to our clinic two years ago because of inefficient mastication resulting from absence of posterior teeth. At clinical examination, it was determined that maxillary and mandibular anterior teeth were present but posterior teeth were absent. The patient was treated with removable partial dentures that had been modified during eruption of permanent posterior teeth.

The aim of the treatment was to reinforce the function of the chewing system and to accelerate eruption of the teeth. After the dentures were placed, they were controlled every month. The posterior parts of the dentures were renewed two times because of eruption of the premolar teeth. The patient has been controlled since 2005.

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### Introduction

Phenylketonuria (PKU) is an inborn error of phenylalanine metabolism resulting from deficient activity of L-phenylalanine-4-monooxygenase, the enzyme that catalyzes the synthesis of tyrosine from phenylalanine.<sup>1</sup> Phenylalanine accumulation in plasma and tissues with decreased tyrosine biosynthesis seems to be involved in the PKU pathogenesis. Treatment of those with PKU consists of restriction of phenylalanine intake, which means a natural protein restricted diet supplemented with a phenylalanine free amino acid mixture enriched with some essential micronutrients, such as vitamins, minerals, and trace elements.<sup>2,3</sup>

Studies show that effective management of many children with inborn errors of PKU requires a highly cariogenic diet. Where protein intake must be restricted, for example, carbohydrate may be needed to supply up to 80% of energy intake per day. The role of sugars of all types is stresses in dietary management, including not only sucrose but also glucose polymers and cornstarch. Although they carry a risk to dental health, sugars have the advantage of being palatable and encouraging energy intake in children with poor appetite.

Glucose polymers and cornstarch may also carry cariogenic potential. Recently studies have focused on caries, but other aspects of oral health may also be affected.<sup>4,5</sup>

There are few reports in the literature concerning with PKU after prosthetic treatment. This clinical study describes the long term follow-up and management of PKU in point of oral health.

### CASE REPORT

A 10-year-old boy was referred to the prosthetics clinic at Dentistry Faculty of Erciyes University as his parents noted that his milestones were becoming delayed. He was diagnosed in Medicine Hospital, as having PKU and placed on a

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low-phenylalanine diet. However, the diet was hard to implement because of inefficient mastication resulting from absence of posterior teeth. His care was then transferred to our clinic where he was reviewed regularly during the following years.

Firstly, the intraoral examination revealed bone atrophy of the alveolar ridges at posterior, on both the maxilla and the mandibula. Except central and lateral teeth, nearly no teeth were present. Orthopantomograph: displaying having less than half the normal number of the dentition (oligodontia), delayed eruption, and missed teeth.

An acceptable solution to this complex case was sought, given the young age of the boy, and the emotional state of his parents, worried by the absence of numerous teeth in such esthetically compromised areas.

Under these conditions, it was considered appropriate to make a lower and upper removable partial prosthesis that had been modified during eruption of permanent posterior teeth (Figure 1a,1b).

This design allows modifications to be made as and when necessary (Figure 2a,2b,2c,2d), as well as providing an acceptable masticatory function on complementing the lack of dentition, at the same time as attempting to maintain the alveolar ridges free of atrophy, and allowing an almost normal social life, which is so important at this stage in a child's personal and psychological development.



2a



2b



2c



1a



Fig. 1a,1b Initial removable prostheses applied to the patient 1a and intraoral view of the prosthesis 1b.



Fig. 2a, 2b, 2c, 2d Adjustment of prostheses during eruption of maxillary right first premolar and view of prostheses after trimming.

The importance of oral hygiene as a fundamental aspect of dental prognosis for PKU patients was stressed to the parents, in the sense that they monitor brushing, provide daily fluoride drops according to body weight, and control the eating of sweets and snacks between meals as far as possible. Periodic check ups were prescribed, approximately every months (Figure 3a,3b), at which topical applications of fluoride gel were to be applied as well as making small adjustments and modifications to the prosthesis, especially the upper one (Figure 4a,4b).



3a



Fig. 3a, 3b During eruption of maxillary right first

premolar and left first premolar, intraoral mucosa was marked for determining trimming areas.



4a



Figure 4a, 4b Adjustment of the maxillary prosthesis during maxillary right first, second and left second premolar erupted 4a, readjustment prostheses 4b.

Now days, we are following the eruption of a still unerupted tooth, as well as insisting on good hygienic measures of daily brushing and mouth rinse, topical fluoride applications every six months, periodic checkups etc (Figure 5a,5b,5c,5d).

### Discussion

Dietary treatment is a primary choice avoids severe neurological damage in PKU. Good communication between the pediatrician, a specialist dietitian and, particularly, pediatric dentist is essential for effective management.<sup>6</sup>

According to Cleary at al.<sup>7</sup> restorative treatment may be required for these cases. The main aim of restorative treatment should be to

reduce the risk of further infection. Glass ionomer cements can be used temporarily and may provide useful fluoride release during the initial stages of caries stabilization. However, to date these materials have insufficient durability to be recommended for the long term.<sup>8</sup> So, prosthetic treatments are fundamental in these patients, attempting to provide a functional and esthetic solution that will allow the child as normal a life-style as possible, without damaging psychological development.<sup>7,8</sup>

common disorders in pediatric practice. The long term survival has improved as a result of early diagnosis. In addition, provision of dental treatment may be complicated by their metabolic disorders, and PKU requires careful multidisciplinary management of pediatric.



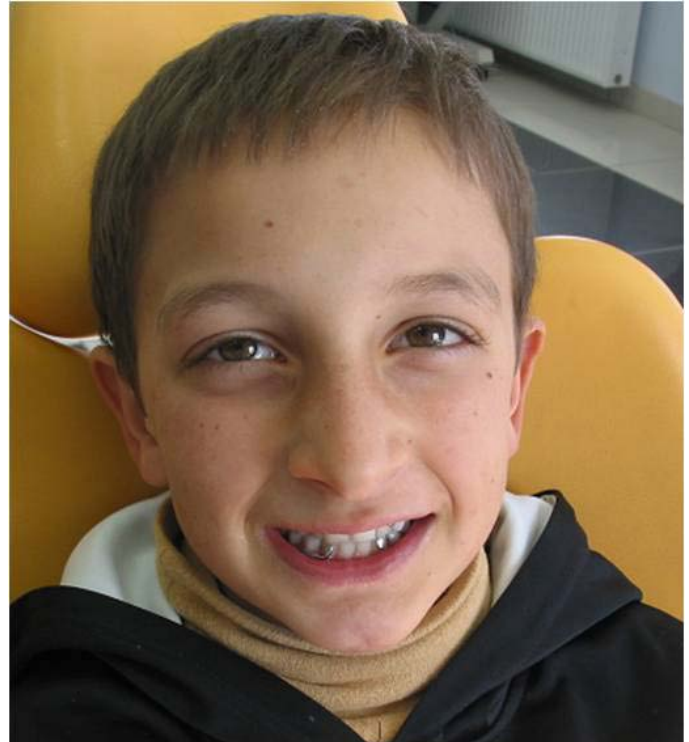
5a



Figure 5a, 5b After maxillary right first, second and left second premolar partially erupted, a second maxillary prostheses constructed.

### Conclusion

In conclusion, the inborn errors of PKU are a group of individually rare but collectively relatively



5c



Fig. 5c, 5d extraoral view of the second maxillary prostheses.

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