Oligodontia: Case Report and Review of Literature

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ABSTRACT

Congenital absence of primary teeth is relatively rare. The prevalence, possible etiological factors and management of multiple missing primary teeth was briefly reviewed. This paper reports a rare case of multiple missing teeth in a 9-year-old female child.

Keywords: Oligodontia, Management.

INTRODUCTION

Congenital tooth agenesis of one or more teeth, which may be either hypodontia (agenesis of fewer than six teeth) or oligodontia (agenesis of six or more teeth), also known as selective tooth agenesis (STHAG), is the most common abnormality of human dentition. Tooth agenesis may be caused by local trauma, chemotherapy, or radiotherapy, or it may occur as part of a systemic genetic syndrome or as an independent oral trait shown to be associated with mutations in several genes. Oligodontia is caused by mutation in LTBP3, the gene encoding latent TGF-β binding protein 3, an extracellular matrix protein believed to be required for osteoclast function. In the permanent dentition, hypodontia has a prevalence of 1.6 to 9.6%, excluding agenesis of the third molars. Oligodontia has a population prevalence of 0.3% in the permanent dentition. It occurs more frequently in girls at a ratio of 3:2.

Oligodontia may occur alone, as part of syndrome, or in more serious systemic disturbances, such as ectodermal dysplasia. If oligodontia is a part of a syndrome there are usually changes on the skin, nails, eyes, ears or skeleton. It is an integral part of more than 120 syndromes. A rare case of nonsyndromic missing primary and permanent teeth with the management and review of literature is discussed.

CASE REPORT

A 9-year-old female patient reported to the department of oral medicine and radiology with a complaint of missing teeth and difficulty in mastication (Fig. 1). Medical history was non-contributory. Patient’s history revealed that missing teeth were not extracted, were absent since childhood and there was no history of trauma. The family history regarding the absence of teeth was not significant. An intraoral examination revealed the presence of primary second molars and permanent first molars in all quadrants but there was no teeth in upper and lower anterior region (Figs 1 and 2). The alveolus present was also very thin. The teeth present were of normal size, shape and color. On OPG, the teeth clinically present were confirmed and there was an erupting crown of lower right second permanent molar was seen.

There was a congenital absence of the maxillary and mandibular right and left primary central incisors, lateral incisors, canines and first molars. After examination, the diagnosis of oligodontia was made and in the management part upper and lower removable partial denture replacing all missing teeth were fabricated.

DISCUSSION

This case is interesting for several reasons. First, there are very few cases reported in the literature regarding congenital absence of primary and permanent teeth, and, in this case, there were only eight teeth present, which in itself is very rare. At the age...
of 9 years, usually the permanent maxillary central and lateral incisor and mandibular central and lateral incisors tooth should be present. But in this case, only the permanent first molars were present and the other tooth was absent. They could be formed later or they can be congenitally missing.

The most common developmental problem with this type of clinical picture is ectodermal dysplasia. Ectodermal dysplasia can be classified mainly as: (1) Hair defect, (2) tooth defect, (3) nail defect and (4) sweating defect.

This can be classified again into many types based on the characteristic features and few conditions associated with the absence of teeth are as follows:

1. **Hypohidrotic ectodermal dysplasia/Anhidrotic ectodermal dysplasia:** It is characterized by partial/complete absence of sweat glands, hypotrichosis and hypodontia. In this condition, primary/permanent teeth may be entirely absent/few may be present. Incisors and canines are conical and pointed. The conical, pointed teeth are the key feature of the syndrome and may be the only obvious abnormality.

2. **Trichoonychodental dysplasia:** A rare syndrome characterized by taurodontic molars, defective enamel and dentin dysplasia. There are few teeth and widely spaced and deciduous teeth tend to persist. Nails are thin with longitudinal striations and cracks.

3. **Fried’s tooth and nail syndrome:** In this condition, hair is fine and short, the teeth are few and peg shaped and the nails are thin and dystrophic.

4. **Hypodontia and nail dysgenesis:** It is characterized by the presence of few teeth which are conical and widely shaped. Nails are small, dystrophic/ spoon shaped.

5. **Odontomicronychial ectodermal dysplasia:** This condition is characterized by precocious eruption and shedding of deciduous dentition, precocious eruption of secondary dentition with short, rhomboid roots and short, thin, slow-growing nails.

6. **Odontotrhomelic syndrome:** Characterized by severe hypotrichosis, few, small, conical teeth and hypoplastic or absent aerolae. There is cleft lip and extensive tetramelic dysplasia. Growth is retarded. However, this patient does not have peg-shaped teeth; dry, scaly skin, straw-like hair, frontal bossing, wide-spaced eyes or any problem with body-heat regulation. She appears to be a perfectly normal young child, with no known family history of missing teeth. The features in this patient also are not consistent with any specific forms of ectodermal dysplasia.

### CONCLUSION

The consequences of missing teeth are numerous and depend on the number and type of teeth that are missing. Most frequently speech and masticatory functional disorders occur and esthetic problems caused by disturbed growth and development of the orofacial area, which can manifest outside the mouth. In the case of patients with oligodontia, prompt, accurate diagnosis is necessary and careful planning of treatment, with a preconception of the final solution. This can only be achieved by multidisciplinary cooperation, which usually includes the following specialists: Orthodontist, pedodontist, oral surgeon and prosthodontist.

Surgical therapy for patients with marked aplasia can include various aspects, such as autotransplantation, insertion of osteointegrated implants. Autotransplantation is applied most frequently in those cases where aplasia is localized mainly in one jaw and the teeth from the other jaw can be utilized, e.g. in persons with asymmetric aplasia. Osteointegrated implants can be a suitable solution for patients with multiple aplasia. However, experience is limited with regard to long-term results in children and adolescents. Because of its irreversible character, the different aspects of their application should be considered, particularly for younger patients. It is well known that implants react like ankylosed teeth, when inserted before the end of growth of the alveolar ridge. It is therefore considered that implant therapy can be applied at the earliest when growth and development has nearly finished. In rare cases of complete aplasia, such as in ectodermal dysplasia, implant therapy can be recommended during childhood.

### REFERENCES


