

## Complete anodontia of the permanent dentition: case report

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### Abstract

*A disturbance during the embryology of tooth formation of the seven-year-old girl presented in this case resulted in her having the unusual combination of nearly a complete complement of primary teeth but anodontia of the permanent dentition. It could not be determined if this child had any of the many syndromes or conditions characterized by missing teeth. Clinical characteristics and management of this case are described.*

### Introduction

About four weeks after fertilization, the cells of the future toothbearing area proliferate and form the epithelial band called the dental lamina. Ten swellings appear along the dental lamina and develop into the primary teeth. The dental lamina continues to grow and by the end of the fourth month *in utero*, the tooth germs for the permanent first molars have formed. This process continues until the third molars are initiated in four-year-old children. Agenesis of the teeth is the result of a disturbance in this process, and the ectodermal tissues do not proliferate and differentiate into highly specialized cells which make up the tooth buds (Ooe 1981; Provenza 1988). The reason for this deficiency is unknown. Such an interference is relatively common, and the prevalence of congenitally absent teeth is reported to be as high as 8% (Pito 1987). The permanent teeth most frequently missing congenitally are the third molars, second premolars, and the maxillary lateral incisors (Tal 1981). Many syndromes have hypodontia as a feature, and therefore congenitally missing teeth may be a significant diagnostic feature of a syndrome or an associated finding. Fifty-six syndromes have hypodontia as a clinical feature (Hall 1983). Anodontia, the complete absence of the primary and permanent teeth, is a rare condition usually associated with a systemic disorder.

There is no single etiology for anodontia. It has been proposed that partial or complete anodontia is an evolutionary stage which will result in mankind not having teeth (Herbst and Apfelstaedt 1930; Sperber 1963). Three syndromes characterized by partial or complete anodontia are oculomandibulodyscephaly, mesoectodermal dysplasia, and ectodermal dysplasia (Gorlin and Pindborg 1964).

Oculomandibulodyscephaly is characterized by microphthalmia, blue scleras, and microcephaly. In

addition, permanent teeth are absent, but the primary teeth endure. Symptoms of mesoectodermal dysgenesis include a wide face, eye deformity, muscle dystrophy, underdeveloped premaxilla, and sometimes complete hypodontia (Gorlin and Sedano 1971).

Ectodermal dysplasia is suspected when a patient has dystrophic fingernails and toenails, a malady of the eccrine and sebaceous glands, and oligodontia. Types of ectodermal dysplasia include: hypohidrotic or anhidrotic, hydrotic or Clouston, ectrodactyly, Rapp-Hodgkin, and Robinson (Behrman and Vaughn 1983).

Hypohidrotic ectodermal dysplasia usually is sex-linked recessive with full expression in males. In some families (particularly if there is consanguinity), transmission is autosomal recessive with expression in both sexes. Symptoms include hypohidrosis, anomalous dentition, hypotrichosis, episodes of fever, frontal bossing, malar hypoplasia, a flattened nasal bridge, everted lips, hyperpigmented periorbital skin, and low-set ears. The skin over the body is thin and hypopigmented, the hair is sparse, multiple teeth are congenitally absent, and the anterior teeth are conical and spaced. There are other less common symptoms. Gorlin and Sedano (1971) described a 4 1/2 year old girl with hypohidrotic ectodermal dysplasia and the complete absence of primary and permanent teeth.

The hydrotic form of ectodermal dysplasia is the autosomal dominant form. Aplasia of the sweat glands, absence of sebaceous glands, dry skin, blond, fine, stiff, scant hair, as well as missing eyelashes and eyebrows are common. Few and conical teeth are typical. The ectrodactyly, Rapp-Hodgkin and Robinson types of ectodermal dysplasia are autosomal dominant. The ectrodactyly type is characterized by cleft lip and palate and ectodermal anomalies such as thin, dry skin with glandular deficiency, and anomalies of the hands, feet, eyes, and urinary tract. The dentition also is defective. The Rapp-Hodgkin type of ectodermal dysplasia has symptoms which include hypohidrosis, sparse fine hair, oral clefts, and retarded growth. Finally, the Robinson type has symptoms which include deafness, nail involvement, and partial anodontia (Behrman 1983).

In a review of the literature, Beierle and Jorgenson (1978) found 14 cases of complete anodontia of the primary and permanent teeth. They also described three cases in which primary teeth were present but the

permanent teeth were missing. Such cases that have been radiographically documented are summarized in the Table.

### Case Report

A seven-year, seven-month-old Caucasian female reported to the Pediatric Dentistry Clinic at LSU School of Dentistry with a chief complaint of carious primary teeth and missing permanent teeth.

Her medical history revealed a period of jaundice shortly after birth but no heart, respiratory, bleeding problems, or allergies. She took no medications. Her height was in the 25th percentile and weight in the 50th percentile. The child was evaluated by an endocrinologist who concluded that even though her symptoms included onychodysplasia, hyperkeratosis of the palms of the hands and soles of the feet, frontal bossing, and saddle nose, the diagnosis of ectodermal dysplasia could not be confirmed. The hereditary background included no known consanguinity in the family or history of anodontia or ectodermal dysplasia in either the maternal or paternal lineages. The grandfather had conical mandibular central and lateral incisors. The mother and father had complete primary and permanent dentitions. The one sibling, a sister, nine years old, had a normal complement of primary and permanent teeth.

Extraoral examination revealed a well-developed, well-nourished child who was cooperative for examination. The face was symmetrical, the profile convex, the lower lip everted, and the chin prominent (Figs 1 and 2). Facial proportions were nearly normal, although the lower facial height was almost equal to the upper facial height because the vertical dimension was reduced. The soft tissue drape appeared normal in fullness, was nonwrinkled, and had normal pigmentation. Lacrimation was observed, and the child had normal visual, olfactory, and gustatory sensations.

Oral examination revealed normal mucosal, palatal, and periodontal tissues. The tongue was normal in size, and no tongue thrust was exhibited. Linguoalveolar, bilabial and sibilant speech sounds were articulated



Fig 1. Facial profile shows depressed nasal bridge and everted lower lip.

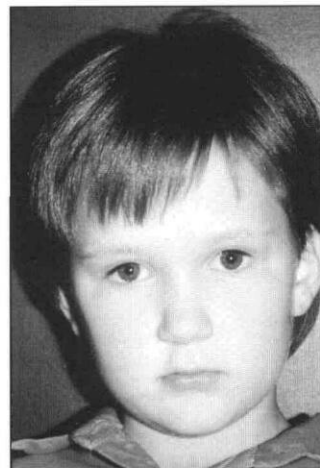


Fig 2. Full face shows normal hair, eyes, and facial appearance.

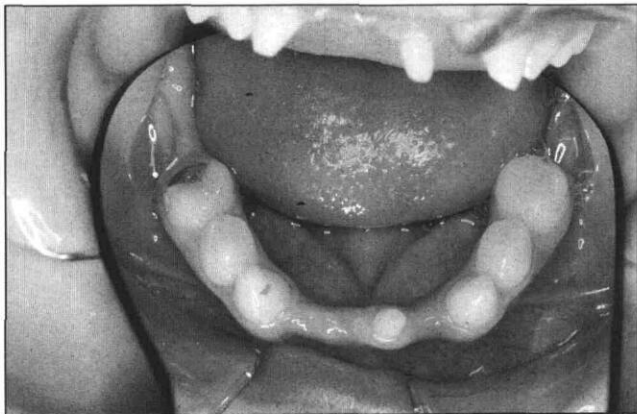
correctly. The primary maxillary lateral incisors were congenitally absent, and both the mandibular primary lateral and the right central incisors had exfoliated (Figs 3 and 4, see next page). In centric occlusion, the terminal plane was flush bilaterally, the midline was coincident, the incisors had no overjet or overbite, and the posterior teeth were in bilateral crossbite. There were six carious teeth including the maxillary right and mandibular right second primary molars. The primary teeth were slightly delayed in eruption. Neither the parent nor the child were aware of a bruxism habit, and the loss of the enamel on the incisal edges of the incisors was probably the result of attrition due to the end-on occlusion.

Confirmation of complete anodontia of the permanent teeth was made by panoramic radiograph (Fig 5, see next page), maxillary, and mandibular anterior occlusal, and bite-wing radiographs of the eight-year-old child. Although the potential for the formation of permanent teeth still may be present at age eight years, there was no radiographic evidence of follicular sacs, calcification of cusps, or other indications of tooth formation. The mandibular primary molars were taurodonts and the incisors; canines and primary molars were conical.

The first phase of dental treatment included restoring the anterior carious teeth with resin restorations and the posterior teeth with amalgam or stainless steel crowns. The enamel over the functioning occlusal surfaces of the noncarious posterior teeth showed no evidence of attrition, and protective restorations were not indicated. To improve esthetics, a modified lingual arch was fabricated to replace the three missing mandibular incisors. The child demonstrated excellent home-care habits, meticulously brushed with a soft

TABLE. Summary of Radiographically Confirmed Cases of Children with Primary Teeth and Anodontia of the Permanent Dentition.

Author	Age	Sex	Number of Primary Teeth	Treatment
Clark 1922	5	Female	5	overdentures
Ware 1938	12	Male	18	not described
Hutchinson 1953	8	Male	20	not described
Swallow	11	Male	20	maxillary overdenture
Sperber 1963	5	Male	5	overdentures
Herman 1977	14	Female	17	overdentures



**Fig 3.** Mandibular intraoral occlusal view demonstrating arch form and exfoliated primary incisors.

toothbrush, and consumed a healthy diet. Professionally applied fluorides and vigilant monitoring of the dentition were performed at six-month intervals.

The second phase of treatment involved construction of overdentures. The parent elected to delay this care until the child was eleven years old. The overdentures were designed to increase the vertical dimension by 14 mm, improve esthetics and function, preserve the alveolus, and prepare the patient for dentures, if needed in later life. A daily fluoride mouthrinse was prescribed, and the added importance of frequent daily toothbrushing and flossing were emphasized to the child and parent to minimize the possibility of caries and periodontal disease. It has been shown that the amount of attached gingiva, sulcular depth, mobility of abutment teeth, and gingival inflammation are not significantly affected by an overdenture if the oral hygiene is good (Tolson et al. 1982).

## Discussion

Lifetime preservation of the primary teeth and alveolar ridges is a problem rarely encountered by the clinician. Based on the number of reported cases, complete anodontia is more frequent than anodontia of the permanent teeth only. The objective of treatment is to maintain the primary teeth by protecting the thin enamel cap and dentin from occlusal abrasion and trauma. This protection may be achieved by restoring the posterior teeth with full crowns and prescribing a mouthguard. In addition, esthetic facings may be placed on the anterior teeth to increase the mesial-distal diameters and to close diastemas. In this case, this problem was managed with an overdenture. Preservation of the teeth is particularly important to preserve alveolar bone because implants and fixed restorations may not be indicated.

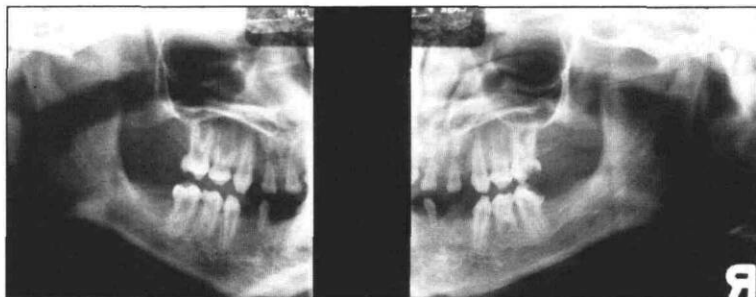
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**Fig 4.** Maxillary intraoral occlusal view of the congenitally absent lateral and conical central incisors.

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**Fig 5.** Panoramic radiograph shows complete anodontia of the permanent teeth.