Anodontia with hypohidrotic ectodermal dysplasia in a young female: a case report

Kohachiro Ohno, DDS, PhD Ikuo Ohmori, DDS, PhD

Dr. Ohno is an associate professor; and Dr. Ohmori is a professor and chairman, and they are both at the Tsurumi University School of Dental Medicine, Department of Pediatric Dentistry, Yokohama, Japan.

Abstract

A five-year-old Japanese girl was referred to our clinic for non-eruption of the teeth. Panoramic radiographs revealed absence of all teeth of both primary and permanent dentitions. She showed hypotricosis, hypohidrosis, and anodontia. In this article, the reported cases of anodontia were reviewed and the fabricating procedure of full dentures for a young child was described. (Pediatr Dent 22:49-52, 2000)

Anodontia, which represents the congenital absence of all teeth in the primary dentition and/or the permanent dentition, is a rare condition. The characteristic feature of the patient with anodontia is a senile facial appearance due to lack of all teeth and underdevelopment of the alveolar ridges. Tanner[1] states in the case of ectodermal dysplasia that such abnormal appearance may affect normal social and psychologic development in young patients. Therefore, the dental care for a patient with anodontia is important.

The purpose of this article is to describe a literature review of reported cases of anodontia and their dental care, and to demonstrate the clinical manifestations and the fabricating procedure of prosthesis in this rare case.

Literature review

Reported cases of anodontia

There were many reported cases containing the term of anodontia in the title. Most cases were not true anodontia, but nearly anodontia or oligodontia. The cases of true anodontia were extremely limited to rare conditions. During the last 50 years, some cases of anodontia have been reported, as listed in Table 1. All cases of anodontia in both dentitions were associated with ectodermal dysplasia and occurred predominantly in males, but some cases of anodontia of permanent dentition only occurred as an isolated abnormality and in both sexes.

Table 1. List of Reported Cases of Anodontia

Authors	Year Reported	Gender	Age	Involved Dentition	HED	Remark
Sarnat et al.[2]	1953	M	6y~16y	primary & permanent	accompanied	
Eisenson[3]	1956	M	5y	primary & permanent	accompanied	
Ochiai et al.[4]	1961	M	4y 8 mo	primary & permanent	accompanied	
Issa[5]	1965	M	9y	primary & permanent	accompanied	
Elfenbaum[6]	1966	M	4y 6 mo	primary & permanent	accompanied	
Gorlin et al.[7]	1970	F	4y 9 mo	primary & permanent	accompanied	
Galeone[8]	1972	M	8y	primary & permanent	accompanied	
Shaw[9]	1990	M	10y	primary & permanent	accompanied	
Nomura et al.[10]	1993	F	6y 1 mo	primary & permanent	accompanied	
Franchi et al.[11]	1998	F	4y 2 mo	primary & permanent	accompanied	
Everett et al.[12]	1952	M	8y 6 mo	permanent	accompanied	
Hutchinson[13]	1953	M	8y	permanent	not accompanied	
Swallow[14]	1959	M	11y	permanent	not accompanied	
Sperber[15]	1963	M	5y	permanent	accompanied	
Herman et al.[16]	1977	F	14y	permanent	accompanied	
Gorlin et al.[17]	1980	F	14y	permanent	accompanied	same case with Herman
Yamashita et al.[18]	1992	F	8y	permanent	not accompanied	

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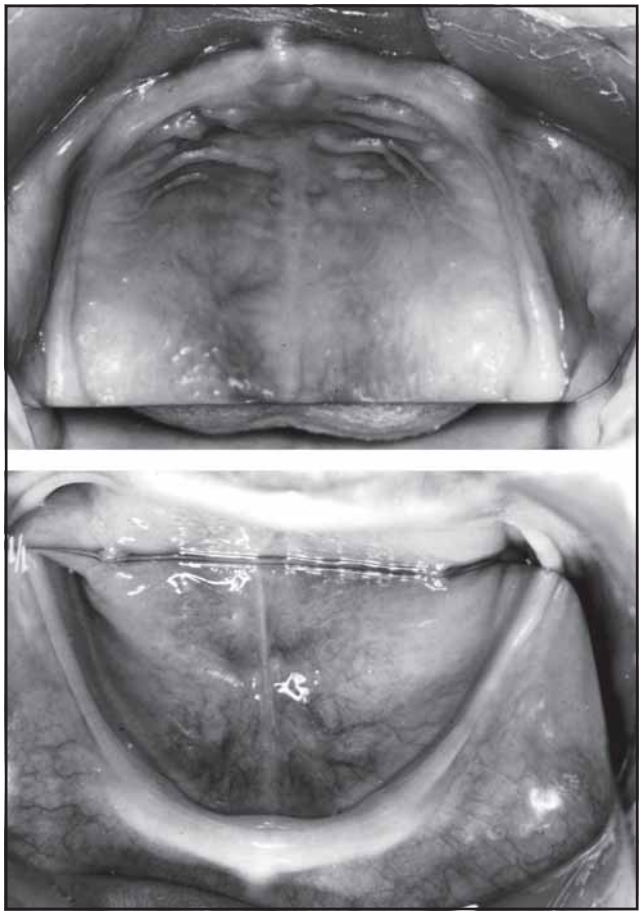

Fig 1. Intraoral photograph of the patient. The complete absence of teeth is noted in both jaws. The alveolar ridges are rather atrophic. The alveolar and other oral mucosa seems normal.

Dental care for the patient with anodontia

Nowak[19] suggests that the pediatric dentist is best suited to manage the pediatric patient's special needs, although there may be dentists in the community who possess the special ability to manage pediatric patients as well as the skills for fabricating dentures. As some authors indicate, the major goal for the pediatric dentist is to provide the patient with an oral apparatus with optimal esthetics and function, and to allow the patient to develop physically, emotionally, and socially like his/her siblings or peers[19-21]. The dental literature describes some conventional prosthetic approaches to the clinical management of these patients[9-11, 22]. The new approach of using implant-supported overdentures, instead of conventional removable dentures, has recently been reported[23-25].

Case report

A girl of 5 years and 3 months of age was referred to the pediatric dental clinic of Tsurumi University Dental Hospital for non-eruption of the teeth. She was born at 40 weeks gestation by a normal delivery. At birth, the patient's weight was 2665 g and her length was 48.5 cm. During two months after birth, she had experienced recurrent attacks of high fever and crying. She was admitted to Tokyo Metropolitan Toshima Hospital to examine the physical condition, and was diagnosed as hypohidrotic ectodermal dysplasia, because she showed hypohidrosis, hypotrichosis and dental anomalies by oral radiographs. She attended the hospital once a year for a regular checkup. She had an episode of otitis media, but did not have a history of bronchitis, pharyngitis, asthma and allergies. The patient's father and mother were normal and not consanguineous. Her elder sister was also normal and none of the parent's relatives was known to have a similar condition.

At first visit, she showed a senile facial appearance and fine sparse hair on the scalp, especially on the temporal. Her eyebrows and eyelashes were also sparse. Additionally, she showed a prominent forehead, a saddle nose and everted lips in the profile. The skin of the body was dry and atrophic. Sweat tests by the iodine-starch reaction revealed that she was positive partially on the head and neck and a little positive in the armpit, but negative on the leg and foot. However, the shape of the nails of the fingers and toes seemed to be normal.

Intraoral examination revealed the edentulous condition (Fig 1). The alveolar ridges were rather atrophic. The color of alveolar mucosa and other oral mucosa was normal. The flow of saliva was observed in the floor of oral cavity. Panoramic radiographs and cephalometric radiographs revealed complete absence of teeth or tooth buds and aplasia of the alveolar processes of the maxilla and the mandible (Figs 2, 3). From these manifestations, we diagnosed this case as anodontia with hypohidrotic ectodermal dysplasia.

In order to restore the oral function, the full dentures were constructed by a modified conventional approach. The procedure of fabricating the dentures was as follows:

1. Impression taking for the study cast was performed with alginate material by using selected stock tray for children.
2. The individual trays were made with tray resin.
3. Final impression taking was performed with silicon rubber material (Exaflex, G-C. Co., Tokyo, Japan) by using the trays, without border moulding with compound material.
4. The wax rims were made on the working casts.
5. At the occlusion record, the horizontal jaw relation was determined without using the face-bow and the proper vertical dimension and free way space were established. The vertical dimension of occlusion was first determined by measuring face height in respiratory rest position and checked by means of cephalometric criteria.
6. Wax dentures were constructed with available acrylic primary teeth (Nisshin dental product, Kyoto, Japan). The articulater used was Gysi Simplex OU-type (Onuki Dental Co., Tokyo, Japan).
7. Wax dentures were adjusted in the mouth.
8. The dentures were made of heat-cured acrylic resin.

At 5 years and 10 months of age, the full dentures were completed and inserted (Fig 4). Figures 5 and 6 show her frontal views before and after insertion of the dentures. After

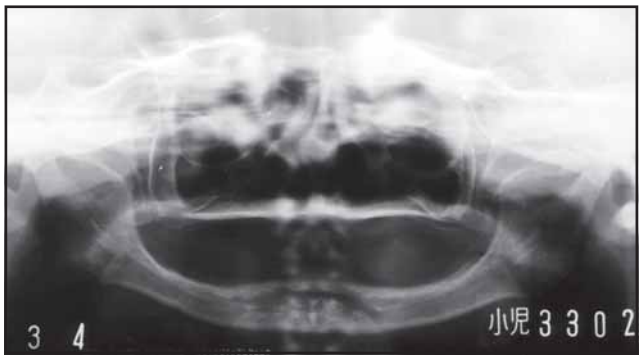

Fig 2. Panoramic radiograph revealed complete absence of both the primary and permanent teeth.

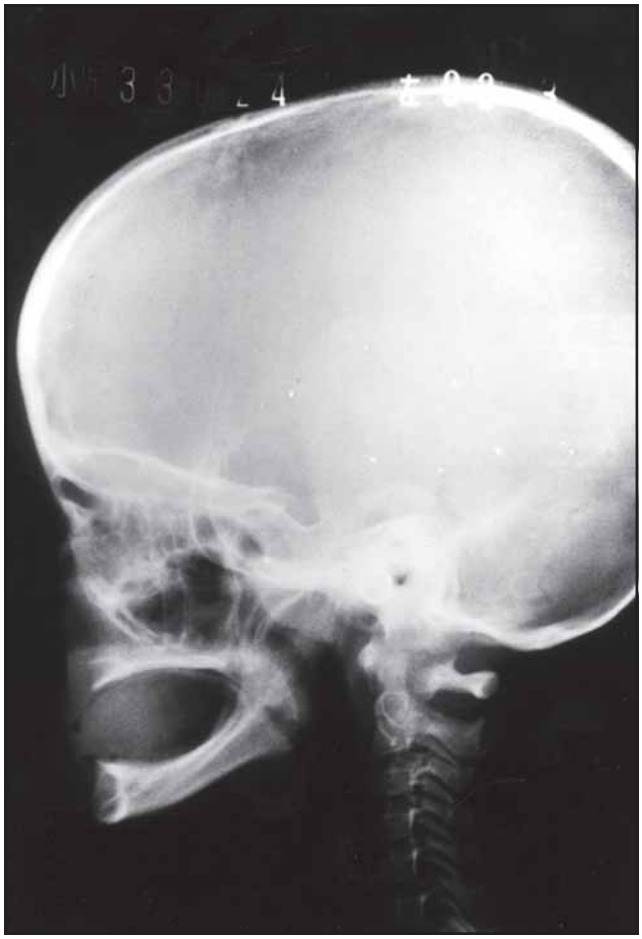

Fig 3. Cephalometric radiograph taken at 5 years and 3 months of age.

insertion of the dentures, her facial appearance was improved by alteration of the lower facial portion. By the cephalometric analysis, it was confirmed that the length of ANS-Me after insertion of the dentures was 2.5 mm larger.

The patient and her mother were instructed that the dentures should be handled with care, worn at all times except while brushing and sleeping, and washed with tepid water while brushing. The patient adapted well to the dentures. After a month, she complained of pain near the oral mucosa under the upper denture. By checking the fitness with white silicone rubbers (Fit checker, G-C Co. Tokyo, Japan), the denture was

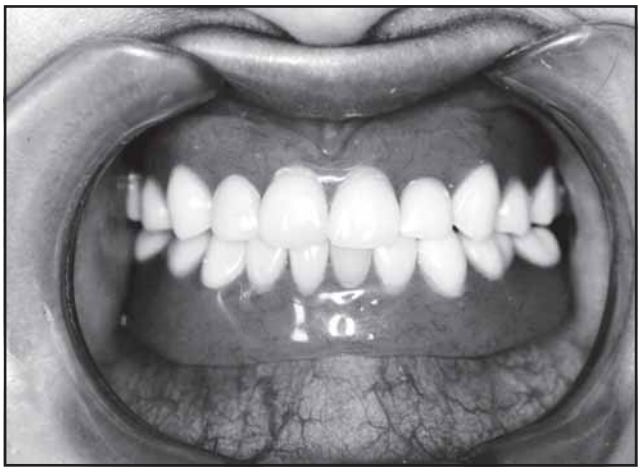

Fig 4. Intraoral photograph with insertion of the acrylic full dentures.

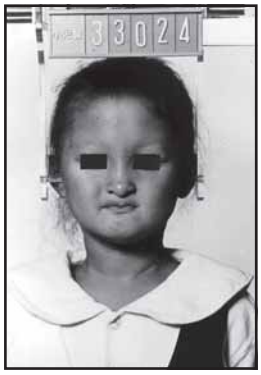

Fig 5. Frontal view before inserting the full dentures.

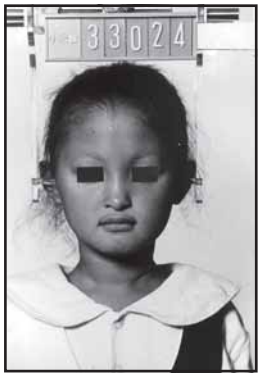

Fig 6. Frontal view after inserting the full dentures. Her facial appearance was improved due to alteration of the lower facial patterns.

adjusted. She was now able to masticate meats and rice crackers by using the dentures. When speaking, her "s" sound was audible normally, but her "t" sound, especially the "tsu" sound in Japanese, was audible as the "chu" sound. She was able to communicate better and was more engaging and interactive with others, probably due to the improvement of her appearance. She attended our clinic about once a month for a regular checkup and the dentures were adjusted 4 times, although she did not complain of any pain.

To our regret, she did not return to our clinic after 6 months due to changes in her family situation.

Discussion

Anodontia is an extremely rare condition, especially in females. It has been reported as a manifestation of one of the most severe forms of ectodermal dysplasia, of which more than 120 different variations are known.[26] Among them, X-linked hypohidrotic ectodermal dysplasia is most common. In our case, a triad of symptoms of hypohidrotic ectodemal dysplasia was observed. The other disorders requiring differential diagnosis were congenital insensitivity to pain with anhidrosis, incontinentia pigmenti, ectrodactyly-ectodermal dysplasia-clefting syndrome, Rapp-Hodgkin syndrome, and Ellis-van Creveld syndrome. They could be ruled out because of sensitivity to pain, regular color of skin, normal fingers, normal palate and normal limbs, respectively.

The gene responsible for the X-linked type has been recently reported by Monreal et al.[27] , indicating that direct molecular diagnosis of the disorder is feasible and will allow for the identification of female carriers and help to distinguish between the X-linked form and recessive form through genetic testing. Since we did not perform the genetic testing by their identification method, the etiology in our case is not known. However, Gorlin et al.[7,17] have stated that an autosomal recessive mode of inheritance may occur in most cases of the affected females with unaffected parents, which may suggest the possibility of this case.

Anodontia can be diagnosed by taking a panoramic radiograph at about 4 years of age, because teeth, if any, can be expected to be visible on radiographs at this age[28]. Primary teeth can also be visible on radiograph at birth.

The dental literature describes the dental treatment and management for the patient with anodontia or severe oligodontia. There is not a definitive time to begin treatment, but Pigno et al.[22] suggest that an initial prosthesis should be delivered

before school age of the patient. In this case, the full dentures were inserted at 5 years and 10 months of age. Consequently, she will probably have time to adapt to the dentures and her appearance will seem normal when she starts school. According to Kupietzky and Houpt[20], it is feasible to fabricate a denture for a patient as young as 3 years of age. As the child grows, the denture will have to be modified and replaced, because it is indicated in the longitudinal studies of anodontia that the growth of jaws is independent of existence of teeth[2,4,10]. Replacements will be needed at least 3 times between the period of early and late mixed-dentition and permanent-dentition. When the patient is in the last stage, the removable prosthesis may be replaced by a fixed type restoration using osseointegrated implants. The implant-supported dentures reported are excellent in stability and retention,[23-25] but the placement of dental implants in young children generates several problems. Guckes et al.[23] recommend that this approach should be postponed until age 13 because of possible implant movement caused by jaw growth. In addition, Guckes et al.[29] suggest that their use in the anterior mandible may be routinely recommended in young patients if implant-supported prostheses were shown to have positive effects on craniofacial growth, self-image and food choice.

Regarding the vertical dimension record for full denture of an edentulous adult, Silverman[30,31] has described that the components of the recording procedure include morphologic and physiologic phenomena, functional activity, and psychologic and social criteria, particularly in relation to esthetic decisions. As mentioned above in our case, the modified recording procedure was used and the acceptable vertical dimension was obtained. The fabricating procedure of dentures for the young patient was not so complicated, although the treatment requires knowledge of growth and development, behavioral management and the skills for fabricating dentures[20]. The effect of prosthetic management of the patient should be evaluated in long-term followup. Unfortunately, the followup was limited in this case, so the evaluation of the treatment result could not be mentioned.

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