Clinical evaluation of a patient with single maxillary central incisor

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Hypodontia in permanent dentition is the most common developmental anomaly and frequently found in the second premolar and maxillary lateral incisor. In the primary dentition, however, hypodontia appears to be less frequent, with the exception of cases such as ectodermal dysplasia and cleft lip and palate. We report a child with one primary maxillary central incisor at midline. The presence of a single permanent maxillary central incisor was also confirmed by radiological examination. Other intraoral abnormalities were detected including absence of upper labial frenulum and abnormal palatal structure, but no other facial or brain anomalies.

Although the condition is exceedingly rare, a thorough examination for more serious anomalies should be conducted since it is suggested to be the mildest feature of holoprosencephaly.
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INTRODUCTION

Anatomic abnormalities of teeth development include the presence of a single maxillary central incisor at midline instead of two central incisors. The coronal form of this tooth is symmetrical. The single maxillary central incisor is the only tooth present in both deciduous and permanent dentitions. The incidence of cases with a single maxillary central incisor is approximately 1 in every 50,000 live births.[1]

The exact mechanism of this abnormality is unknown, but several studies have postulated that the appearance of the single maxillary central incisor is related to the fusion of two neighboring tooth buds or to agenesis of a tooth germ.[2-5] However, cases of a single maxillary central incisor with short stature have been reported, and growth hormone deficiency was confirmed in several of those cases.[6]

Moreover, other studies reported that the condition correlated with congenital nasal stenosis such as choanal atresia, nasal pyriform aperture,[1-7] various congenital anomalies and chromosomal defects.[8-12] These findings suggest that the single maxillary central incisor is not an isolated dental anomaly and consideration for the physical status is necessary when examing a patient with this condition.

Besides, the clinical features of individuals with a single maxillary central incisor resemble the mild manifestations of holoprosencephaly. The latter is a congenital anomaly characterized by median facial anomalies and defective cleavage of prosencephalon.[13] It is suggested that there is a close relationship between the two abnormalities.[14,15] In this regard, previous studies have confirmed the presence of mutations in the human sonic hedgehog (SLIH) gene in chromosomal region 7q36.1 of patients with holoprosencephaly type III.[13-16] In Japan, two cases of a single maxillary central incisor with chromosomal deletion of 7q36.1 have been reported.[9,17]

The purpose of the present report is to describe midline anomalies in a child with a single maxillary central incisor. Several studies that have reported a variety of clinical features of the single maxillary central incisor suggested that careful systematic examination was necessary in patients with the single maxillary central incisor for proper dental management.

CASE REPORT

A 5-year 10-month-old boy visited the Pediatric Dental Clinic of Nagasaki University in June 1998. The mother

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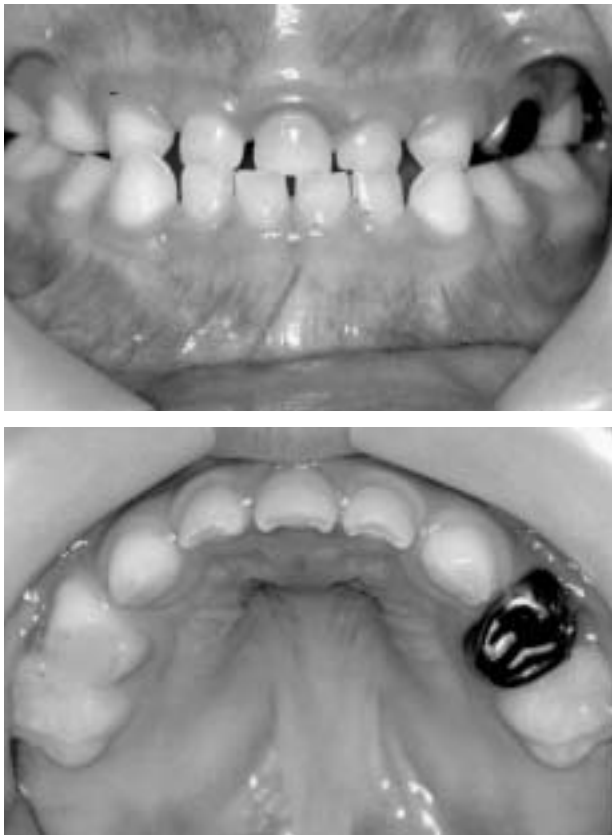

Figures 1a, 1b. Intraoral photograph taken at the age of 5Y 11M. Note the presence of the single maxillary central primary incisor at midline. Note also the absence of upper labial frenulum and abnormality of palatal structure.

a: Front view
b: Intraoral view of maxillary arch

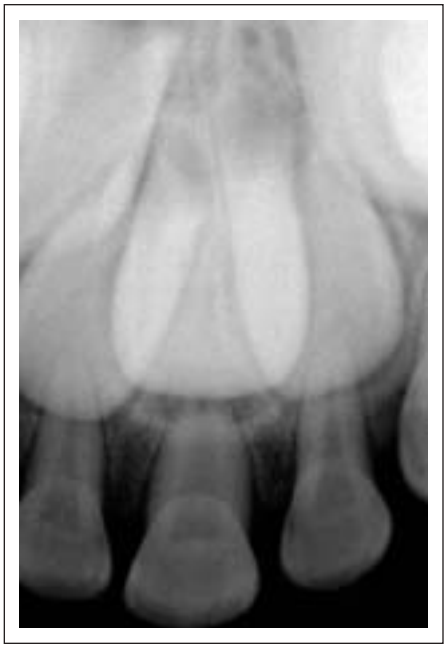

Figure 2. Radiograph of the maxillary incisor region at the age of 5 years. Note the presence of a single maxillary primary central incisor and a single root and root canal. Note also the single maxillary permanent central incisor on the midline of the maxilla.

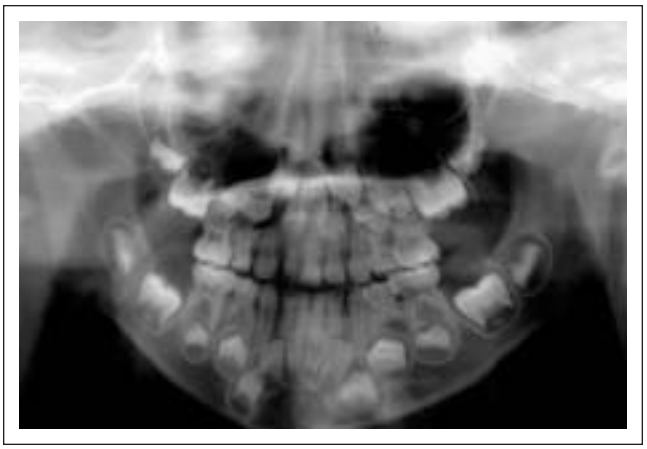

Figure 3. Panoramic radiograph at the age of 5 years. Note the normal developmental status for age, except for the presence of a maxillary primary central incisor and one permanent central incisor.

was concerned about the non-eruption of one maxillary central primary incisor (Figure 1). The patient was born at full gestation and delivery was uneventful. The medical history was uneventful apart from 1-month hospitalization soon after birth for enlargement of his thymus gland, but no treatment was provided. The child was reported to have persistent nasal airway obstruction. The family history was negative. There was no history of previous trauma involving the maxillary incisor region.

Physical examination showed body height of 115cm (±0.8S.D.) and weight of 20kg (about average) on admission. Examination of the face showed an interpupillary distance of 46.5mm, which was smaller than the average 2S.D. of Japanese boy,[18] and the child was therefore, diagnosed to have hypotelorism. Intraoral findings included a single maxillary central primary incisor, absence of upper labial frenulum and incisal papilla (Figure 1). The single maxillary central primary incisor was located at midline, and the coronal form was symmetric and did not show the angle symbol and the curve symbol. The palatal structure was abnormal with bilateral paramedian anteroposterior furrows and midline ridge over the raph6 anteroposteriorly. (Figure 1-b)

A dental radiograph showed a single maxillary primary central incisor with a single root at midline. Furthermore, the succedaneous permanent tooth was also single with a single root at midline (Figure 2).

A panoramic radiograph showed a normal dental developmental status for a child, aged five years, with the exception of the absence of one maxillary primary and permanent central incisor (Figure 3).

As shown in Figure 4, the terminal plane of the vertical type, and edge-to-edge bite of incisors and class III

Table 1. Mesiodistal (M-D) crown width (in mm) of primary tooth.

	Patient	Control
Maxilla		
Cental incisor	6.1	6.8±0.4
Lateral incisor	5.4	5.5±0.4
Canine	6.3	6.8±0.4
First molar	6.5	7.4±0.4
Second molar	9.1	9.2±0.4
Mandibular		
Cental incisor	4.0	4.3±0.3
Lateral incisor	4.6	4.8±0.3
Canine	5.9	6.1±0.3
First molar	7.9	8.4±0.5
Second molar	10.4	10.4±0.5

Data are mean ± SD of normal Japanese boys established at the Department of Pediodontics, Tokyo Dental College.

Table 2. Measurement of dental arch (in mm).

	Patient	Control
Dental Arch Width		
Upper jaw		
C-C	19.8	25.1±1.7
D-D	24.5	28.0±1.8
E-E	27.9	30.2±1.8
Lower jaw		
C-C	19.7	19.5±1.4
D-D	26.2	24.9±1.6
E-E	30.3	29.2±1.4
Dental Arch Length		
Upper jaw		
Anterior	10.9	12.1±0.8
Posterior	15.1	16.7±0.7
Total	26.1	28.8±1.2
Lower jaw		
Anterior	7.8	8.7±0.6
Posterior	17.5	17.6±0.9
Total	25.4	26.2±1.3

Data are mean ± SD of normal Japanese boys established at the Department of Pediodontics, Tokyo Dental College.

Table 3. Cephalometric analysis.

	Patient	Control
Facial angle	84.9	82.9±5.0
convexity	11.0	11.5±3.0
A-B plane angle	-5.7	-5.8±1.6
Mandibular plane angle	37.0	31.1±5.2
Y-axis	62.0	63.8±3.3
Occlusal plan	13.5	14.3±4.3
Interincisional	159.0	147.8±7.4
L-1 to mandible	78.5	84.4±6.2
FH t78o SN plane	9.5	7.8±3.1
SNA	80.5	81.4±2.8
SNB	78.0	76.4±2.1
NF to FH plane	4.5	-0.1±4.4
U-1 to FH plane	78.0	96.4±4.7
U-1 to SN plane	69.0	88.8±4.4
GZN	90.5	88.5±3.8
NSM	72.5	71.4±3.2
Gonial angle	135.0	130.5±4.3
Ramus angle	7.5	9.2±4.4

Data are mean ± SD of normal Japanese boys standardized by Iizuka (Japanese boyes age 6.3±1.8Y).

of canine to canine relationship were noticed. Using study models, we measured the following parameters; mesiodistal (M-D) crown width of primary teeth, and the width and length of dental arch (Table 1 and 2). The M-D width of the single maxillary central primary incisor was smaller than the average -1 S.D. (Table 1). The other maxillary primary teeth tended to be smaller than the average size, but mandibular teeth were within the normal range (Table 1). Both the width and length of the maxillary dental arch were smaller than the average size, particularly the intercanine width, which was smaller than the average -3 S.D. However, the corresponding dimensions in the lower jaw were within the normal range (Table 2).

After routine treatment of carious teeth, a lateral cephalometric roentgenogram was taken at a periodical dental examination at age 7 years 5 months. The results of cephalometric analysis are showed in Table 3 and Figure 5. Interincisal angle, mandibular plane angle and Gonial angle were larger than the average ±1 SD. of Japanese children. Furthermore, L-1 to mandibular plane angle, U-1 to FH plane angle and U-1 to SN plane angle were smaller (average -1S.D., -4S.D.,<-4S.D.). The line analysis of the cranial base and measurement of the pituitary fossa were performed in accordance with the report of Saito[19]. There was no significant maldevelopment of the cranial base and the pituitary fossa showed a normal radiographic outline.

DISCUSSION

There is a general agreement that the incisor is not a mere disorder of tooth clinical findings include the presence of a incisor at midline in both dentitions, absence and abnormality of the palate structure.[1,17] In additional, hypotelorism, choanal atresia, stenosis of nasal pyriform aperature and midnasal stenosis have been reported in subjects with a single maxillary central incisor.[1-7] The above defects are anomalies that appear coincidentally at midline of the craniofacial region, which is thought to be due to embryonic interaction defect during the 5 to 9 weeks of gestation (embryonic developmental period from the frontnasal process to the lateral and medial nasal process). The underlying mechanism of the defective embryonic development is still unknown.

In some cases, the single maxillary central incisor is associated with short stature due to deficiency of growth hormone,[6] which is released from the anterior pituitary gland. It is thought that the pituitary gland originated

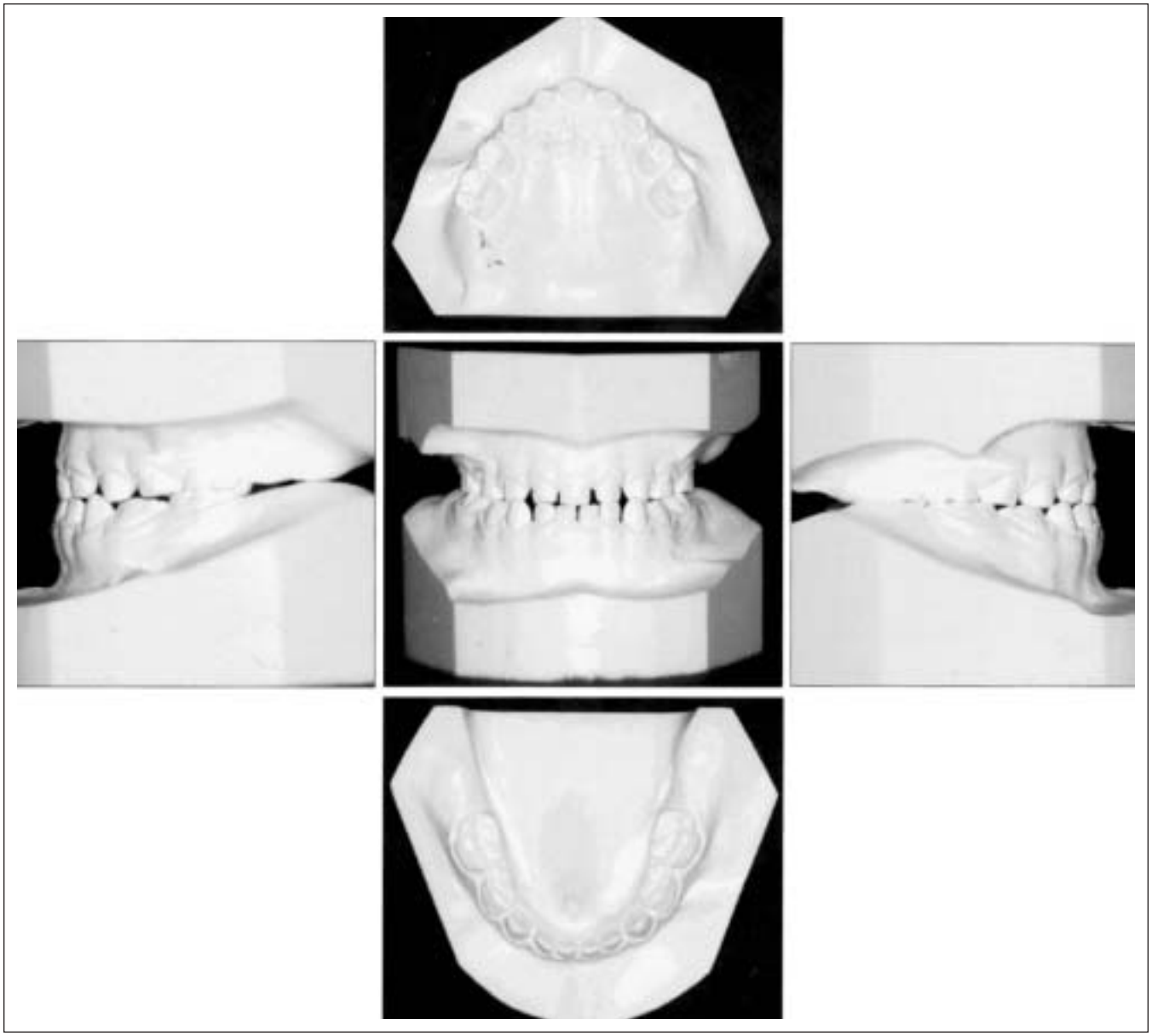

Figure 4. Frontal, lateral and occlusal views of a study model of the patient at 5 years of age.

Occlusal relationship: edge-to-edge bite of incisors
Type of terminal plane: vertical type
Canine to canine relationship: class III

from the ectoderm as well as the tooth has been affected in some cases. Previous studies have suggested that the function of the pituitary gland relates to the size and form of pituitary fossa. Therefore, radiological examination of sella turcica is important for clinical diagnosis. Hypopituitarism is associated with reduced secretion of growth hormone, and can cause mandibular deficiency, delayed eruption of teeth and small size of the pituitary fossa.[20,22] In hyperpituitarism, gigantism, overgrowth of mandible, and expansion of the pituitary fossa may be noted.[20,22]

In our case, measurement of the pituitary fossa on the cephalometric roentgenogram did not show any abnormality in the form or size of pituitary fossa, and the child did not present with short stature. It should be noted, however, that anatomic changes in the pituitary fossa do not always represent dyspituitarism; nor does dysfunction of the hypophysis present as abnormal form or size of sella turcica. Defects of the hypophyseal-portal circulation, which is involved in transportation of growth hormone releasing hormones from the hypothalamus to the pituitary gland, may influence the synthesis and/or release of growth hormone. Formation of the pituitary gland from diencephalon and Rathke's pouch commences earlier than that of tooth during the

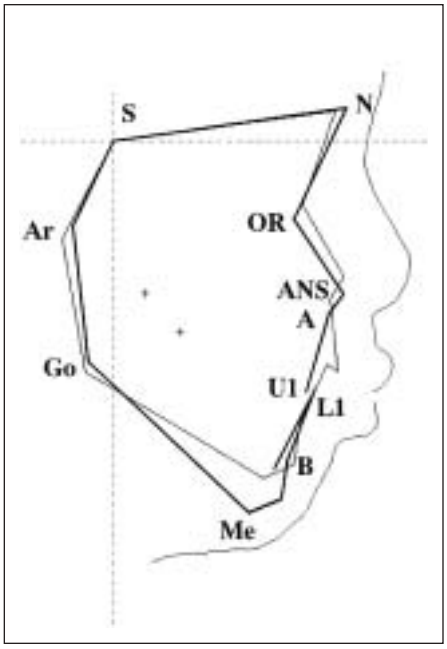

Figure 5. Cephalometric analysis using a profilogram. The control profile is standardized by Sakamoto.

Narrow line: control (Japanese boys aged 6Y2M-8Y11M),
Broad line: patient of this study at 7Y5M

Occlusal relationship of incisors had shifted to a crossbite during the eruption of the first mandibular molar tooth. (dental age of IIC).

third week of gestation. It may be thought that the developmental periods occurred anomalies would be related with the severity of the clinical condition. Anyway, radiological examination of sella turcica alone is not sufficient for the evaluation of the function of pituitary gland; appropriate examination of the hormonal function of hypophysis should be performed, and we have to observe the overall development up to the adolescent growth spurt.

Previous studies have indicated that the single maxillary central incisor may be the mildest feature of holoprosencephaly,[14,15] which is a severe malformation of the brain involving complete separation of the central nervous system into left and right halves.[13] The condition results from impaired midline cleavage of the embryonic forebrain, following basic defect in embryonic - development and differentiation of notochordal plate occurring before 21-25 days of gestation.[13,16]

Holoprosencephaly is associated with cyclopia, ethmocephalic or cebocephalic face in severe case, while mild holoprosencephaly could be associated with a single maxillary central incisor, microcephaly, hypotelorism and other craniofacial findings or normal face with or without brain malformation[1315,7]. In this case, the feature of palate (showing in Figure 1-b) resembles that of holoprosencephaly in the previous report.[23] It is important to take notice of mild clinical conditions in the dental management in order to identify the presence of holoprosencephaly.

Holoprosencephaly is thought to be an autosomal dominant or recessive hereditary disorder of isolated abnormality.[13] According to Camera *et al.*[24], the single maxillary central incisor may be considered as an indicator of potential holoprosencephaly in the next generation.

Furthermore, type III holoprosencephaly has been confirmed to be associated with a disorder of human sonic hedgehog (SIHIH) gene on chromosome 7q36.1. [13,16] In this regard, few cases of single maxillary central incisor have been reported to have 7q terminal deletion of the same breakpoint at 7q36.[1,9,12,17] Sonic hedgehog (SHH) is expressed in the developing tooth germ and plays an important role in tooth development.[25] Considered together, our case and the above chromosomal/developmental anomalies suggest that this gene is important for the normal development of the midline maxillary region. Mutation analysis of this gene is currently under investigation in our laboratory.

In conclusion, pediatric dentists should be aware of the characteristic clinical signs such as single maxillary central incisor, absence of upper labial frenulum and abnormal palatal structure. According to the severity of these anomalies, one should suspect malformation of the brain, and potential presence of holoprosencephaly in the proband offspring and/or members of the proband's family.

These patients should be managed carefully including genetic counseling. While it is not uncommon that parents of otherwise normal looking children may refuse examination of pituitary function and chromosomal analysis, such tests are necessary and should be performed.

Our patient had a considerably small intercanine width of the maxillary dental arch, relative to the mandible, and the relationship between upper and lower incisors had shifted to cross bite during the eruption of the first mandibular molar. It seemed that the maxilla tended to be less prognathic in this patient. It was also thought that approach using such a maxillary protractive appliance and/or maxillary expansion plate may be needed. It should be stressed that before any treatment is contemplated, it is important to perform radiographic examination of mid-palatal structures.

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