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ABSTRACT

Apert syndrome is a rare craniosynostosis syndrome characterized by irregular craniosynostosis, midface hypoplasia, and syndactyly of the hands and feet. Previous studies analyzed individuals with Apert syndrome and reported some facial and intraoral features caused by severe maxillary hypoplasia. However, these studies were performed by analyzing both individuals who had and those had not received a palate repair surgery, which had a high impact on the maxillary growth and occlusion. To highlight the intrinsic facial and intraoral features of Apert syndrome, 5 Japanese individuals with Apert syndrome from 5 years and 2 months to 9 years and 10 months without cleft palate were analyzed in this study. A concave profile and a skeletal Class Ⅲ jaw-base relationship caused by severe maxillary hypoplasia were seen in all patients. The patients exhibited anterior and posterior crossbites possibly due to a small dental arch of Maxilla.

Key words: Apert syndrome, skeletal Class Ⅲ jaw-base relationship, maxillary hypoplasia

INTRODUCTION

Apert syndrome (MIM#101200) is a rare craniosynostosis syndrome with an estimated incidence of 1 in every 160,000 live births; it accounts for 4% to 5% of all craniosynostosis syndromes (Cohen & Kreiborg, 1992, Cohen & Kreiborg, 1993, Cohen & Sulik, 1992). The syndrome is characterized by irregular craniosynostosis, midface hypoplasia, and syndactyly of the hands and feet. The disorder is associated with a mutation in the fibroblast growth factor 2 receptor gene (FGFR2) that maps to chromosome 10q25-10q26 and follows an autosomal dominant inheritance pattern (Ciurea & Toader, 2009, Wilkie, 1996). The recurrence risk for an unaffected parent of a child with Apert syndrome is minor, but an affected person has a 50% risk of having a baby with the syndrome.

Early synostosis of calvarial coronal, sagittal and metopic sutures coupled with synostosis of the cranial base result in midface hypoplasia and vertically progressive craniofacial complex (Cohen & Kreiborg, 1993). Consistent with midface hypoplasia, the maxilla also exhibits sagittal and transverse hypoplasia. Intraoral manifestations include open bite, anterior crossbite and crowding. A narrow and high palatal arch can also be seen. Bulbous palatal swellings, mostly consisting of mucopolysaccharides, can give the appearance of pseudocleft. A 30% incidence of soft-palate clefting has been reported (Ferraro, 1991).

Patients with Apert syndrome often require craniofacial team care and dental, orthodontic and orthognathic surgical management because of their esthetic and functional problems such as Class Ⅲ malocclusion and midface hypoplasia.

Previous studies analyzed individuals with Apert syndrome and reported some facial and intraoral features caused by severe maxillary hypoplasia. (Kreiborg et al., 1999, Dalben Gda et al., 2006, Ferraro, 1991, Letra et al., 2007, Nurko & Quinones, 2004) However, these studies were performed by analyzing both individuals who had and those had not received a palate repair surgery, which has a high impact on the maxillary growth and occlusion. The systematic orthodontic features of patients with Apert syndrome without the effect of palate repair surgery have not been described and characterized in detail. The purpose of this study was to evaluate the complications in a series of 5 cases of Apert syndrome without cleft palate and to clarify the specific facial and intraoral features.

MATERIALS AND METHODS

Five Japanese children with Apert syndrome who came to the Clinical Department of Orthodontics, Hokkaido University Hospital were evaluated in this study. All 5 children had craniosynostosis and syndactyly of the hands and feet, and they were diagnosed as Apert syndrome at birth. Their ages at the first visit to our department ranged from 5 years and 2 months to 9 years and 10 months. Records on systemic complications and procedures, including forehead advancement, shunt placement, hand surgery, ear, nose and throat (ENT) surgery such as placement of pressure equalization tubes, tonsillectomy and adenoidectomy, were obtained by inspection of individual medical records.

Frontal and lateral cephalograms and orthopantomograms were obtained for all patients at the first visit, and skeletal and dental characteristics were evaluated. The Sassouni method was used to determine the facial midline (Krogman & Sassouni, 1957), and deviation from the midline was measured by using the following parameters: occlusal plane cant, deviation of the anterior nasal spine (ANS), Menton, and upper and lower incisors. Traditional cephalometric landmarks and measurements were used in this study (Takeuchi et al., 1978, Nakamura et al., 1979). Reference points, planes and lines used for lateral cephalometric analysis are shown in Fig. 1. The sex- and age-matched Japanese norms reported by Nakamura and Masaki were used.N-S, N-S-Ar, SNA, ANS-PNS, N. Pog-A, SNB, SN-Pog, Gonial angle, GZN, SN-Mp, Go-Pog, Ar-Go, Ar-Me, NA-Pog, ANB, L1 to Mp, N.Pog-L1, U1 to SN, U1 to NF, N.Pog-U1, Interincisal, SN-FH, SN-Occ values were reported by Nakamura (Nakamura et al., 1979); S-Ba, N-S-Ba values were reported by Masaki (Masaki, 1980).

Cast models were also taken in all patients at the first visit, and dental arch width and length were measured by a caliper (Mitutoyo, Tokyo, Japan), which had a ±0.05-mm error. Dental arch width, the distance between the imus of lingual cervical margins of the right and left teeth (C: deciduous canine, E: deciduous second molars and 6: permanent first molars), was measured in both arches, and the dental arch length, the distance from the contact point of deciduous or permanent central incisors to the line connecting the distal surfaces of the right and left deciduous second molars (cases 1, 2 and 3) or permanent first molars (case 5), was also measured in both arches (Sakai et al., 1974) and compared with the norms of Japanese dental arch width and length reported by Sakai (Sakai et al., 1979). Overbite and overjet were measured from cast models.

To minimize error, each measurement was repeated at least twice by one experienced orthodontist (H.K.). Random error in lateral cepharometric measurements estimated by the Dahlberg formula was 0.59° (Baumrind S.＆Robert C., 1971, Maria Christina de Souza Galvao, 2012).

RESULTS

Systemic condition

Records on systemic complications and procedures including forehead advancement, shunt placement, hand surgery and ear, nose and throat (ENT) surgery are summarized in Table1. Forehead advancement was performed by a team of craniofacial and neurosurgeons in all 5 cases at 1 month to 4 years and 8 months of age to correct intercranial hypertension. In case 3, forehead advancement was performed at 1 year and 5 months and at 4 years and 8 months of age. In case 5, forehead advancement was performed twice at 1 month and 4 months of age. Placement of pressure equalization tubes was performed at 2 years and 6 months in case2. Adenoidectomy and placement of pressure equalization tubes was performed at 4 years in case3. Because of severe respiratory distress and perioperative airway management, case 4 required tracheal intubation at birth and underwent tracheostomy in the neonatal period.

Facial and oral characteristics

Facial and oral photos of the 5 patients are shown in Fig. 2. All patients had a concave profile with midfacial hypoplasia. Moderate exorbitism and a small retrodisplaced nose were also noted. All 5 patients had anterior open bite. The terminal plane was a mesial-step type on both sides in cases 1 to 4. Case 5 had Class Ⅰ molar relationships. None of the patients in this study had cleft palate. Narrowing in the upper arch was marked in case 1, 2, 4 and 5, and a pseudo cleft palate with a Byzantine-arch shape was noted in all cases. Hellman dental ages were IIC (case 1, 2, 3 and 4) and IIIA (case 5). Overbites and overjets measured from cast models are summarized in Table 1. Anterior tongue position during speech and swallowing, commonly called tongue thrusting, was seen in all cases.

Skeletal characteristics

Frontal cephalograms, lateral cephalograms and orthopantomograms are shown in Fig. 3. Frontal cephalometric measurements are shown in Table 2. Case 2 had left-side deviations of the mandible from the facial midline with the frontal view of the occlusal plane inclined toward the contralateral side. Upper and lower incisors were deviated to the left side. Case 3 had minimal right-side deviation of the mandible with the occlusal plane inclined toward the ipsilateral side. Lower incisors were deviated to the left side. Case 4 had minimal right-side deviation of the mandible. Upper and lower incisors were also deviated to the right side. Case 5 had left-side deviations of the mandible and right-side deviations of the maxilla with the frontal occlusal plane inclined toward the ipsilateral side. Upper incisors were deviated to the right side.

Lateral cephalometric measurements of the 5 patients are summarized in Table 3, and representative scatter diagrams are shown in Fig. 4. For the cranial base, z scores of the anterior cranial base (N-S) and the saddle angle (N-S-Ar) were less than -1points lower than the norm in case 2, 3, 4 and 5. N-S and N-S-Ar in case 1 could not be evaluated because of the lack of an age-matched Japanese norm.

The posterior cranial base (S-Ba) was relatively low and z scores of N-S-Ba were less than -2 points lower than the norm in case 5. S-Ba and N-S-Ba in case 1, 2, 3 and 4 could not be evaluated because of the lack of an age-matched Japanese norm. For the maxilla, SNA ranged from 66.2° to 76.3° and z scores of ANS-PNS were less than -2 points lower than the norm in all 5 cases. For the mandible, SNB varied from 75.8° to 81.6° among cases. The mandibular plane angle (SN-Mp) was larger than the norm. ANB ranged from -11.0° to -4.5° and z scores was less than -5 points lower than the norm in all 5 cases. Results of cephalometric analysis, when compared to the Japanese norm, revealed a skeletal Class III jaw-base relationship due to severe maxillary deficiency. Labial inclination of the upper incisors (U1 to SN) and/or lingual inclination of the lower incisors (L1 to Mp) were found as the result of dental compensation to skeletal maxilla-mandibular disharmony.

Profilograms of the 5 cases are shown in Fig. 5. The Japanese age- and sex- matched norm is denoted by a dotted line. Apparent maxillary hypoplasia was seen in all cases.

The state of development and eruption of permanent teeth were evaluated from each orthopantomogram (Fig. 3). Permanent mandibular bilateral second premolars were not detected in case 1. It was suspected that these teeth were congenitally missing or that there was developmental delay, follow-up assessment was needed. Agenesis of the permanent maxillary lateral incisors and congenital absence of the permanent mandibular left lateral incisor were seen in case 2. Abnormal development and delayed eruption of permanent teeth were not observed in case 3 and case 4. In case 5, two impacted anterior maxillary supernumerary teeth were observed, but severely delayed eruption of permanent teeth was not detected.

Dental arch characteristics

The dental arch was measured in each patient (Table 4). The maxillary intercanine and intermolar widths were overall smaller than the Japanese norms in cases 1 to 4. In these patients, the mandibular dental arch widths were also smaller. Although the upper and lower dental arch widths were both greatly reduced, the upper arch was relatively small in cases 1 and 2, which showed bilateral posterior crossbites.

The maxillary arch lengths were overall smaller than the norm, but the mandibular arch lengths showed variation. The maxillary and mandibular arch lengths in case 3 could not be evaluated because of the lack of an age-matched Japanese norm.

DISCUSSION

A concave profile and a skeletal Class III jaw-base relationship caused by severe maxillary hypoplasia were seen in all of the patients with Apert syndrome without cleft palate.

Previous studies showed that suture fusion is not limited to the skull but may also involve facial sutures and cartilages in patients with Apert syndrome. Ousterhout et al. examined the cranial base of a 38-months-old boy histologically and described significant microanatomic changes including premature bony fusion of the spheno-occipital synchondrosis and fusion of the vomer to the sphenoid bone and maxillae (Ousterhout & Melsen, 1982). Their results suggested that premature fusion of several bones constituting the cranial base eventually reduces growth potential of the maxillofacial structure. Sutures between the maxillae and adjacent craniofacial bones are normally present during development until the teenage periods; however, premature synostosis may inhibit original structural growth. Despite of decreased growth activity of the naso-maxillary complex, mandibular growth will occur during the peak growth period. As a result, severe midface hypoplasia might deteriorate the patient's profile and their skeletal Class III disharmony will become worse. Because of serious structural and functional disorders, the systematic management from infancy to adulthood including orthodontic procedures and orthognathic surgical interventions will be required.

A previous study showed that subjects with maxillary constriction have increased airway resistance and resultant mouth breathing (Langford et al., 2003). Furthermore, Reitsma et al. reported that a low tongue posture, seen in patients with Apert syndrome, might contribute to the underdevelopment of the maxillary arch dimensions (Reitsma et al., 2013). In this study, tongue thrusting was detected in all cases. Functional activity of the tongue and lip muscles is closely related to dentofacial morphology (Hanson, 1988). Correction of this abnormal muscular balance combined with orthodontic and orthognathic procedures should be necessary for effective tooth movement and stability after treatment in our 5 cases.

None of the patients in this study had cleft palate, and we therefore evaluated maxillary structure without the effect of palate repair surgery. In Apert syndrome patients complicated with cleft palate who have received palate surgery, maxillary hypoplasia would be more severe. We observed highly arched and constricted palates with lateral gingival swelling in our patients. Previous studies revealed that palate constriction and lateral swelling increased with aging and caused oral hygiene and periodontal problems (Peterson & Pruzansky, 1974)(Kreiborg & Cohen, 1992). Moreover, difficulty in brushing the teeth because of fused shoulder and elbow joints, hand anomalies and lack of motivation partly due to the mental condition of the patient makes it difficult to maintain adequate oral hygiene (Ferraro, 1991, Nurko & Quinones, 2004). These patients need a plaque control program including professional tooth cleaning and careful oral hygiene instructions on proper tooth cleaning methods, especially during orthodontic treatment.

Apert syndrome has been shown to be the result of mutation of the FGFR2 gene (Ciurea & Toader, 2009, Wilkie 1996). The FGFR2 gene is not only essential for sutural development but is also required for epithelial-mesenchymal interaction during tooth development. Mutation of the FGFR2 gene therefore may affect tooth morphogenesis and development (De Coster et al., 2007, Thesleff & Sharpe, 1997). Delayed dental maturation and tooth agenesis were suspected in 2 of the 5 cases in this study. In such cases, congenital absence and abnormal shape of teeth must be considered when planning orthodontic tooth movement. Previous studies showed that there was a significant delay in dental development in patients with Apert syndrome compared to the control group (Kaloust et al., 1997, Reitsma et al., 2014a, Reitsma et al., 2014b); however, another study showed that there was no difference (Woods et al., 2015). The difference in results might be due to differences in sample size or population, and a study with more subjects is needed.

CONFLICTS OF INTEREST

The authors declare that there is no conflict of interest to disclose.

REFERENCES

Ciurea AV, Toader C (2009) Genetics of craniosynostosis: review of the literature. *Journal of medicine and life***2**: 5-17.

Cohen MM, Jr., Kreiborg S (1992) New indirect method for estimating the birth prevalence of the Apert syndrome. *International journal of oral and maxillofacial surgery***21**: 107-9.

Cohen MM, Jr., Kreiborg S (1993) An updated pediatric perspective on the Apert syndrome. *American journal of diseases of children* **147**: 989-93.

Cohen MM, Jr., Sulik KK (1992) Perspectives on holoprosencephaly: Part II. Central nervous system, craniofacial anatomy, syndrome commentary, diagnostic approach, and experimental studies. *Journal of craniofacial genetics and developmental biology***12**: 196-244.

Cohen MM, Jr., Kreiborg S (1993) Growth pattern in the Apert syndrome. *American journal of medical genetics***47**: 617-23

Dalben Gda S, Costa B, Gomide MR (2006) Oral health status of children with syndromic craniosynostosis. *Oral health & preventive dentistry***4**: 173-9.

De Coster PJ, Mortier G, Marks LA, Martens LC (2007) Cranial suture biology and dental development: genetic and clinical perspectives. *Journal of oral pathology & medicine: official publication of the International Association of Oral Pathologists and the American Academy of Oral Pathology***36**: 447-55.

Ferraro NF (1991) Dental, orthodontic, and oral/maxillofacial evaluation and treatment in Apert syndrome. *Clinics in plastic surgery***18**: 291-307.

Hanson ML (1988) Orofacial myofunctional therapy: historical and philosophical considerations. *The International journal of orofacial myology: official publication of the International Association of Orofacial Myology***14**: 3-10.

Kaloust S, Ishii K, Vargervik K (1997) Dental development in Apert syndrome. *The Cleft palate-craniofacial journal: official publication of the American Cleft Palate-Craniofacial Association***34**: 117-21.

Kreiborg S, Aduss H, Cohen MM, Jr. (1999) Cephalometric study of the Apert syndrome in adolescence and adulthood. *Journal of craniofacial genetics and developmental biology***19**: 1-11.Kreiborg S, Cohen MM, Jr. (1992) The oral manifestations of Apert syndrome. *Journal of craniofacial genetics and developmental biology***12**: 41-8.Krogman WM, Sassouni V (1957) *Syllabus in roentgenographic cephalometry* Philadelphia center for research in child growth.

Langford RJ, Sgouros S, Natarajan K, Nishikawa H, Dover MS, Hockley AD (2003) Maxillary volume growth in craniosynostosis. *Plastic and reconstructive surgery***111**: 1598-604.

Letra A, de Almeida AL, Kaizer R, Esper LA, Sgarbosa S, Granjeiro JM (2007) Intraoral features of Apert's syndrome. *Oral surgery, oral medicine, oral pathology, oral radiology, and endodontics***103**: e38-41.

Maria Christina de Souza Galvao, Joao Ricardo Sato, Edvaldo Capobiango Coelho (2012) Dahlberg formula -a novel approach for its evaluation *Dental press journal of orthodontics***17**: 115-24

Masaki F (1980) The longitudinal study of morphological differences in the cranial base and facial structure between Japanese and American whites. *NipponKyouseisika Gakkai Zasshi***39**: 436-56. (In Japanese.)
Nakamura S, Takeuchi Y, Suzuki A et al. (1979) An atlas of growth analyses on craniofacial structures and dentitions using longitudinal materials collected at Nanporo−cho. *Hokkaido Kyouseisika Gakkai Zasshi***7**: 45-71.(In Japanese.)
Nurko C, Quinones R (2004) Dental and orthodontic management of patients with Apert and Crouzon syndromes. *Oral and maxillofacial surgery clinics of North America***16**: 541-53.
Ousterhout DK, Melsen B (1982) Cranial base deformity in Apert's syndrome. *Plastic and reconstructive surgery***69**: 254-63.
Peterson SJ, Pruzansky S (1974) Palatal anomalies in the syndromes of Apert and Crouzon. *The Cleft palate journal***11**: 394-403.
Reitsma JH, Balk-Leurs IH, Ongkosuwito EM, Wattel E, Prahl-Andersen B (2014a) Dental maturation in children with the syndrome of crouzon and apert. *The Cleft palate-craniofacial journal: official publication of the American Cleft Palate-Craniofacial Association***51**: 639-44.
Reitsma JH, Elmi P, Ongkosuwito EM, Buschang PH, Prahl-Andersen B (2013) A longitudinal study of dental arch morphology in children with the syndrome of Crouzon or Apert. *European journal of oral sciences***121**: 319-27.
Reitsma JH, Ongkosuwito EM, van Wijk AJ, Prahl-Andersen B (2014b) Patterns of tooth agenesis in patients with crouzon or apert syndrome. *The Cleft palate-craniofacial journal: official publication of the American Cleft Palate-Craniofacial Association***51**: 178-83.
Sheldon Baumrind, Robert C. Frantz (1971) The reliability of head film measurements. 1. Landmark identification.*American journal of orthodontics***60**: 111-127.
Sakai M (1979) Relationship of dental arch and basal arch with growth changes.-A study based on the average growth from 3 years to 14 years- *Shigaku***67**: 481-489. (In Japanese.)
Sakai M (1974) Study on eruption locality of the posterior permanent teeth with chronological casts. *Shigaku***61**: 1120-1139.(In Japanese.)
Takeuchi Y (1978) The cephalometric analyses with a function correcting thereference plane-Review and improvement based on the six years clinical application- *NipponKyouseisika Gakkai Zasshi***37**: 353-363. (In Japanese.)
Thesleff I, Sharpe P (1997) Signalling networks regulating dental development. *Mechanisms of development***67**: 111-23.
Wilkie AO (1996) Fibroblast growth factor receptor mutations and craniosynostosis: three receptors, five syndromes. *Indian journal of pediatrics***63**: 351-6.
Woods E, Parekh S, Evans R, Moles DR, Gill D (2015) The dental development in patients with Aperts syndrome. *International journal of paediatric dentistry / the British Paedodontic Society [and] the International Association of Dentistry for Children***25**: 136-43.

TABLES

Table 1 Summary of subject characteristics and interventions

	case1	case2	case3	case4	case5
Sex	F	M	M	F	M
Present age	5y2m	5y10m	6y6m	6y10m	9y10m
Forehead advancement	0y6m	1y1m	1y5m, 4y8m	2y5m	0y1m, 0y4m
Shunt placement	-	-	-	-	+
Hand surgery	+	+	+	+	+
ENT surgery PE tubes / T&A	- / -	+ / -	+ / +	- / -	- / -
other medical interventions	-	-	-	-	tracheostomy
Facial type	concave	concave	concave	concave	concave
Hellman dental age	IIC	IIC	IIC	IIC	IIIA
Overbite (mm)	-1.7	-5.3	-6.5	-5.0	-7.1
Overjet (mm)	-5.5	-1.0	-9.5	-5.0	-7.3

M, male

F, female

ENT, ear, nose and throat

PE tubes, placement of pressure equalization tubes

T&A, tonsillectomy and adenoidectomy

+, received treatment

-, did not receive treatment

Overbite, the vertical distance between upper and lower deciduous incisor edges (case 1 and 2), the vertical distance between upper deciduous incisor edge and lower permanent incisor edge (case 3), the vertical distance between upper and lower permanent incisor edges (case 4 and 5)

Overjet, the horizontal distance between upper and lower deciduous incisor edges (case 1 and 2), the horizontal distance between upper deciduous incisor edge and lower permanent incisor edge (case 3), the horizontal distance between upper and lower permanent incisor edges (case 4 and 5)

Table 2 Frontal cephalometric measurements

	case1	case2	case3	case4	case5
Lateral shift of ANS (mm)	0	0	0	0	1.5 (Right)
Lateral shift of U1 (mm)	0	2.5 (Left)	0	1.5 (Right)	1.5 (Right)
Lateral shift of L1 (mm)	0	3 (Left)	2 (Left)	2 (Right)	0
Lateral shift of Menton (mm)	0	5 (Left)	0.5 (Right)	0.5 (Right)	2.5 (Left)
Occlusal plane cant (°)	0	-0.5	1	0	2

Lateral shift of ANS, Midline-ANS distance

Lateral shift of U1, Midline-midpoint of bilateral upper incisors deviated distance

Lateral shift of L1, Midline-midpoint of bilateral lower incisors deviated distance

Lateral shift of Menton, Midline-Menton distance

Occlusal plane angle, angle between occlusal plane, the line connecting the right and left deciduous second molars or permanent first molars, and the perpendicular of Midline. Positive values indicate inclination of the occlusal plane toward the mandibular deviation side

Table 3 Lateral cephalometric measurements

	case1		case2		case3		case4		case5	
	norm [SD]	patient [z score]	norm [SD]	patient [z score]	norm [SD]	patient [z score]	norm [SD]	patient [z score]	norm [SD]	patient [z score]
Skeltal pattern										
Cranial base										
N-S(mm)	-	53.5	59.5	49.0	60.1	56.5	58.8	54.0	61.6	57.0
	[-]	[ne]	[2.6]	[-4.0]	[2.5]	[-1.4]	[2.5]	[-1.9]	[2.6]	[-1.8]
S-Ba(mm)	-	46.0	-	42.0	-	53.0	-	46.0	50.5	49.5
	[-]	[ne]	[-]	[ne]	[-]	[ne]	[-]	[ne]	[2.5]	[-0.4]
N-S-Ar(°)	-	128.0	129.5	122.0	129.4	119.0	129.3	118.5	128.3	127.5
	[-]	[ne]	[4.9]	[-1.5]	[5.3]	[-2.0]	[5.5]	[-2.0]	[5.4]	[-0.1]
N-S-Ba(°)	-	134.0	-	131.0	-	125.5	-	125.5	143.7	130.0
	[-]	[ne]	[-]	[ne]	[-]	[ne]	[-]	[ne]	[5.0]	[-2.7]
Maxilla										
SNA(°)	83.1	67.7	81.9	66.2	81.9	76.3	83.1	76.0	82.6	68.5
	[3.8]	[-4.1]	[3.0]	[-5.2]	[3.0]	[-1.9]	[3.8]	[-1.9]	[3.5]	[-4.0]
ANS-PNS(mm)	45.7	35.2	47.3	37.2	47.3	42.8	45.7	41.1	48.2	36.0
	[2.2]	[-4.8]	[2.2]	[-4.7]	[2.2]	[-2.1]	[2.2]	[-2.1]	[2.1]	[-5.8]
N.Pog-A(mm)	5.4	-5.0	5.3	-6.3	5.3	-2.0	5.4	-3.7	5.5	-8.5
	[2.0]	[-5.1]	[2.1]	[-5.5]	[2.1]	[-3.5]	[2.0]	[-4.5]	[2.4]	[-5.9]
Mandible										
SNB(°)	78.4	76.3	77.3	75.8	77.3	80.9	78.4	81.6	77.6	79.5
	[3.7]	[-0.6]	[2.8]	[-0.5]	[2.8]	[1.3]	[3.7]	[0.9]	[3.1]	[0.6]
SN-Pog(°)	77.4	73.7	76.4	74.6	76.4	78.5	77.4	80.8	77.0	79.2
	[3.4]	[-1.1]	[2.8]	[-0.6]	[2.8]	[0.8]	[3.4]	[1.0]	[3.1]	[0.7]
Gonial angle(°)	132.6	141.6	133.1	132.2	133.1	129.2	132.6	132.0	132.2	135.6
	[4.8]	[1.9]	[4.2]	[-0.2]	[4.2]	[-0.9]	[4.8]	[-0.1]	[4.8]	[0.7]
GZN(°)	84.9	77.1	84.9	89.5	84.9	89.3	84.9	86.3	85.4	86.2
	[4.0]	[-2.0]	[4.3]	[1.1]	[4.3]	[1.0]	[4.0]	[0.3]	[4.0]	[0.2]
SN-Mp(°)	37.5	38.7	38.0	41.6	38.0	38.5	37.5	38.2	37.6	41.8
	[4.5]	[0.3]	[4.0]	[0.9]	[4.0]	[0.1]	[4.5]	[0.2]	[4.0]	[1.0]
Go-Pog(mm)	63.9	50.0	63.0	59.1	65.3	62.0	64.6	58.0	68.7	68.7
	[2.5]	[-5.6]	[4.1]	[-1.0]	[3.1]	[-0.2]	[2.2]	[-3.1]	[3.2]	[0]
Ar-Go(mm)	38.9	40.0	41.3	35.8	41.8	39.0	39.8	41.0	42.5	48.0
	[2.5]	[0.5]	[1.8]	[-3.0]	[2.8]	[-1.2]	[2.7]	[0.5]	[3.3]	[1.7]
Ar-Me(mm)	88.3	77.0	89.6	77.5	91.9	82.0	89.5	84.5	95.9	100.0
	[3.0]	[-3.8]	[6.7]	[-1.8]	[3.4]	[-1.1]	[3.5]	[-1.4]	[4.5]	[0.9]
Maxilla-Mandible										
NA-Pog(°)	12.1	-11.6	11.8	-14.5	11.8	-4.2	12.1	-8.3	11.7	-17.1
	[4.2]	[-5.7]	[4.6]	[-5.8]	[4.6]	[-3.5]	[4.2]	[-4.9]	[5.0]	[-5.8]
ANB(°)	4.7	-8.6	4.6	-9.6	4.6	-4.5	4.7	-5.6	5.1	-11.0
	[1.6]	[-8.4]	[1.8]	[-7.8]	[1.8]	[-5.0]	[1.6]	[-6.4]	[2.1]	[-7.6]

Denture pattern

L1										
L1 to Mp(°)	84.9	77.9	84.6	59.4	84.6	88.7	84.9	75.0	89.4	86.9
	[6.3]	[-1.1]	[5.8]	[-4.4]	[5.8]	[0.7]	[6.3]	[-1.6]	[7.0]	[-0.4]
N.Pog-L1(mm)	4.9	5.8	4.9	-2.7	10.7	10.7	4.9	4.6	5.9	4.2
	[2.3]	[0.4]	[2.5]	[-3.1]	[2.3]	[2.3]	[2.3]	[-0.1]	[3.1]	[-0.5]
U1										
U1 to SN(°)	92.2	103.0	93.9	92.5	93.9	102.0	92.2	102.5	102.8	110.5
	[6.3]	[1.7]	[7.6]	[-0.2]	[7.6]	[1.1]	[6.3]	[1.6]	[6.6]	[1.2]
U1 to Nf(°)	99.4	114.5	101.9	103.6	101.9	107.1	99.4	100.8	111.1	104.6
	[6.3]	[2.4]	[7.1]	[0.2]	[7.1]	[0.7]	[6.3]	[0.2]	[6.6]	[-1.0]
N.Pog-U1(mm)	6.7	0.2	6.8	-3.7	6.8	0.8	6.7	-1.9	9.0	-1.6
	[2.3]	[-2.8]	[2.6]	[-4.1]	[2.5]	[-2.3]	[2.3]	[-3.7]	[3.3]	[-3.2]
U1-L1										
Interincisal(°)	145.4	140.5	143.6	166.5	143.6	130.9	145.4	144.2	130.2	120.9
	[9.7]	[-0.5]	[10.8]	[2.1]	[10.8]	[-1.2]	[9.7]	[-0.1]	[11.4]	[-0.8]
Angle between two planes										
SN-FH(°)	3.5	-4.8	4.7	-11.7	4.7	-0.5	3.5	-1.8	3.7	4.7
	[3.9]	[-2.1]	[3.6]	[-4.6]	[3.6]	[-1.5]	[3.9]	[-1.4]	[3.6]	[0.3]
SN-Occ(°)	23.8	14.9	22.7	14.42	22.7	20.3	23.8	15.2	19.4	21.9
	[3.6]	[-2.5]	[4.9]	[-1.7]	[4.9]	[-0.5]	[3.6]	[-2.4]	[3.9]	[0.6]

N-S, distance between N and S; S-Ba, distance between S and Ba; N-S-Ar, angle between SN plane and S-Ar line; N-S-Ba, angle between SN plane and S-Ba line; SNA, angle between SN plane and N-A line; ANS-PNS, distance between ANS and PNS; N. Pog-A, distance between N-Pog line and A; SNB, angle between SN plane and N-B line; SN-Pog, angle between SN plane and N-Pog line; Gonial angle, angle between Ramus plane and Mp; GZN, angle between SN plane and ramus plane; SN-Mp, angle between SN plane and Mp; Go-Pog, distance between Go and Pog; Ar-Go, distance between Ar and Go; Ar-Me, distance between Ar and Me; NA-Pog, angle between N-A line and A-Pog line; ANB, difference between SNA and SNB; L1 to Mp, angle between long axis of L1 and Mp; N.Pog-L1, distance between N-Pog line and L1 edge; U1 to SN, angle between long axis of U1 and SN plane; U1 to Nf, angle between long axis of U1 and Nf; N.Pog-U1, distance between N-Pog line and U1 edge; Interincisal, angle between long axes of U1 and L1; SN-FH, angle between SN plane and FH plane; SN-Occ, angle between SN plane and Occ plane.

Brackets in the columns of norm represent SD values of normal samples. Brackets in the columns of patient represent z scores [(measurement－norm)/SD]. Each z score was estimated by the sex- and age- matched norm reported by Nakamura.

ne, not evaluated because of the lack of an age-matched norm

Table 4 Dental arch measurements

		case1	case2	case3	case4	case5
Dental arch width						
Maxilla						
	C-C	16.5	15.5	26.0	14.5	nm
		[-4.9]	[-5.4]	[-0.3]	[-5.8]	
	E-E	24	25	30.6	26.0	nm
		[-3]	[-2.7]	[-0.9]	[-3.0]	
	6-6	nm	nm	nm	nm	38.0
						[0.8]
Mandible						
	C-C	16.2	17.0	19.1	14.5	nm
		[-2.7]	[-2.1]	[-1.5]	[-4.8]	
	E-E	26.3	27.2	25.2	22.0	nm
		[-1.9]	[-1.4]	[-2.3]	[-4.2]	
	6-6	nm	nm	nm	27.0	33.0
					[-4.1]	[1.0]
Dental arch length						
Maxilla						
	A-E	28.4	26.1	27.8	nm	nm
		[-0.4]	[-1.9]	[ne]		
	1-6	nm	nm	nm	nm	31.5
						[-2.2]
Mandible						
	A-E	26.5	21.0	28.0	nm	nm
		[0.1]	[-3.6]	[ne]		
	1-6	nm	nm	nm	31.5	35.0
					[-0.6]	[1.6]

Dental arch width, distance between the imus of lingual cervical margins of the right and left teeth (C: deciduous canine, E: deciduous second molars and 6: permanent first molars), was measured.

Dental arch length, distance from the contact point of deciduous central incisors to the line connecting the distal surfaces of the right and left deciduous second molars or permanent first molars, was measured. Each number in parenthesis represents the z score[(measurement－norm)/SD]. Each z score was estimated by the sex- and age-matched norm reported by Sakai.

nm, not measured because of the condition of no eruption of permanent first molar or loss of deciduous second molar

ne, not evaluated because of the lack of an age-matched norm

FIGURE LEGENDS

Fig. 1. Reference points, planes and lines used for lateral cephalometric analysis.

Points: A, A point; ANS, anterior nasal spine; Ar, articulare; B, B point; Ba, basion; Go, gonion; L1, deciduous or permanent lower central incisor; Me, menton; Mo, molar point; N, nasion; Or, orbitale; PNS, posterior nasal spine; Po, porion; Pog, pogonion; S, sella turcica; U1, deciduous or permanent upper central incisor.

Planes and lines: A-Pog line, line passing through A and Pog; FH plane, Frankfort horizontal plane, plane passing through Po and Or; Mp, mandibular plane, plane passing through Go and Me; N-A line, line passing through N and A; N-B line, line passing through N and B; Nf, nasal floor, plane passing through ANS and PNS; N-Pog line, line passing through N and Pog; Occ plane, plane passing through Mo and midpoint of U1 edge and L1 edge; Ramus plane, plane passing through Ar and Go; S-Ar line, line passing through S and Ar; S-Ba line, line passing through S and Ba; SN plane, plane passing through S and N.

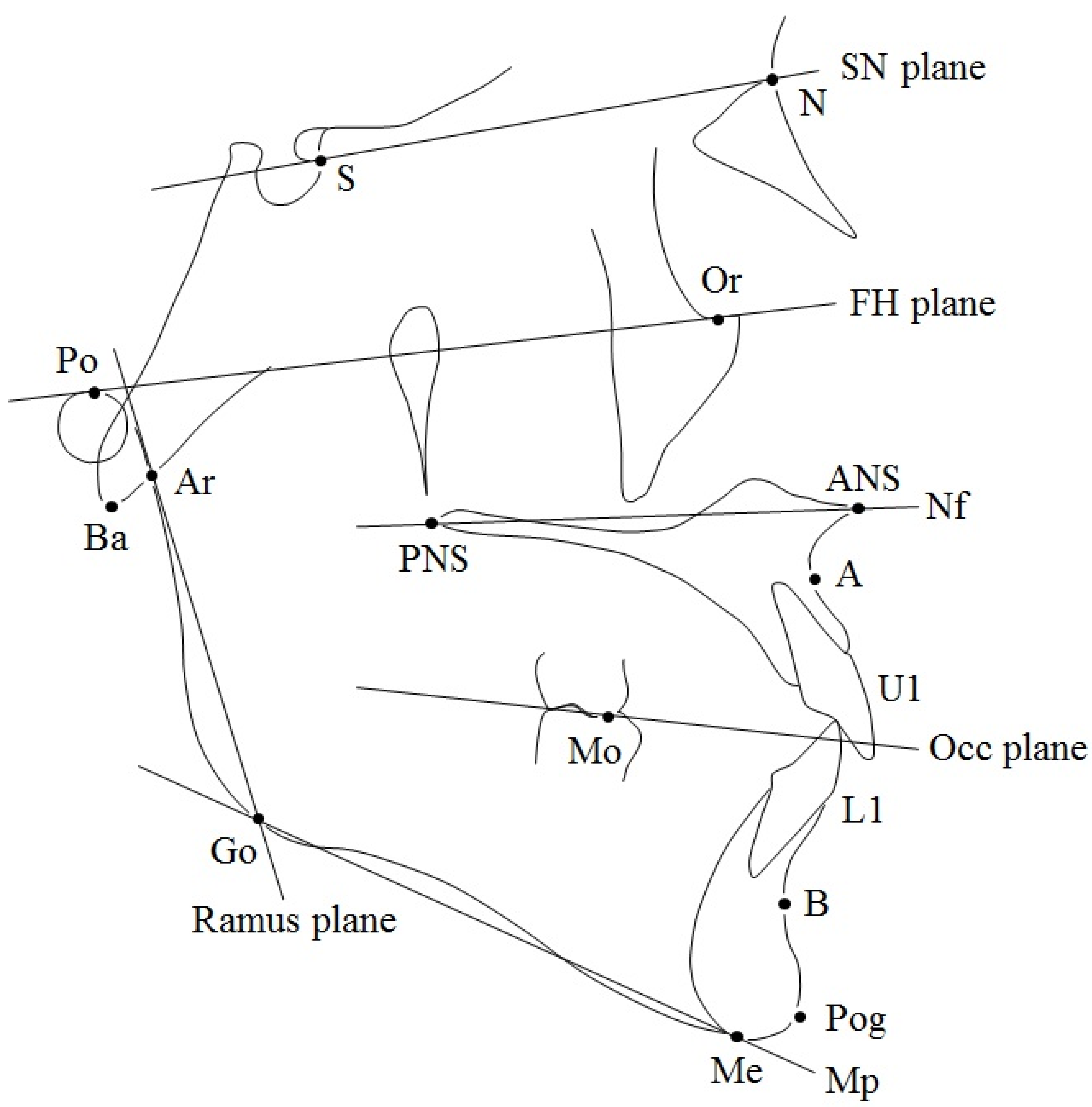
Fig. 2. Facial and oral photos.

Fig. 3. Frontal cephalograms, lateral cephalograms and orthopantomograms.

Fig. 4. Diagrams of lateral cephalometric measurements.

Fig. 5. Profilograms.

Fig.1



SN plane
N
S
Or
FH plane
Po
Ar
Ba
ANS
Nf
PNS
A
U1
Mo
Occ plane
L1
Go
Ramus plane
B
Pog
Me
Mp

Fig.2

Fig.3

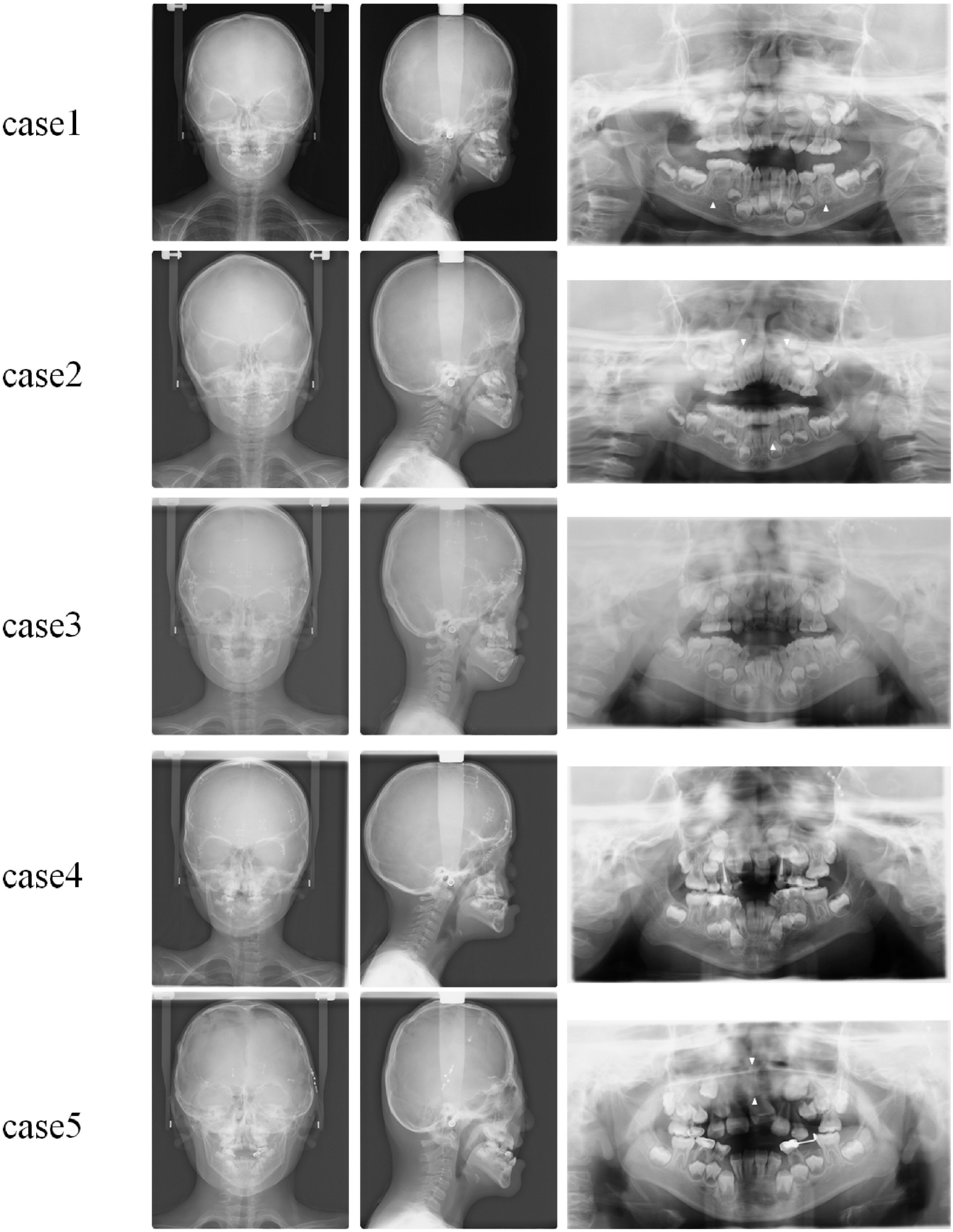


Fig.4

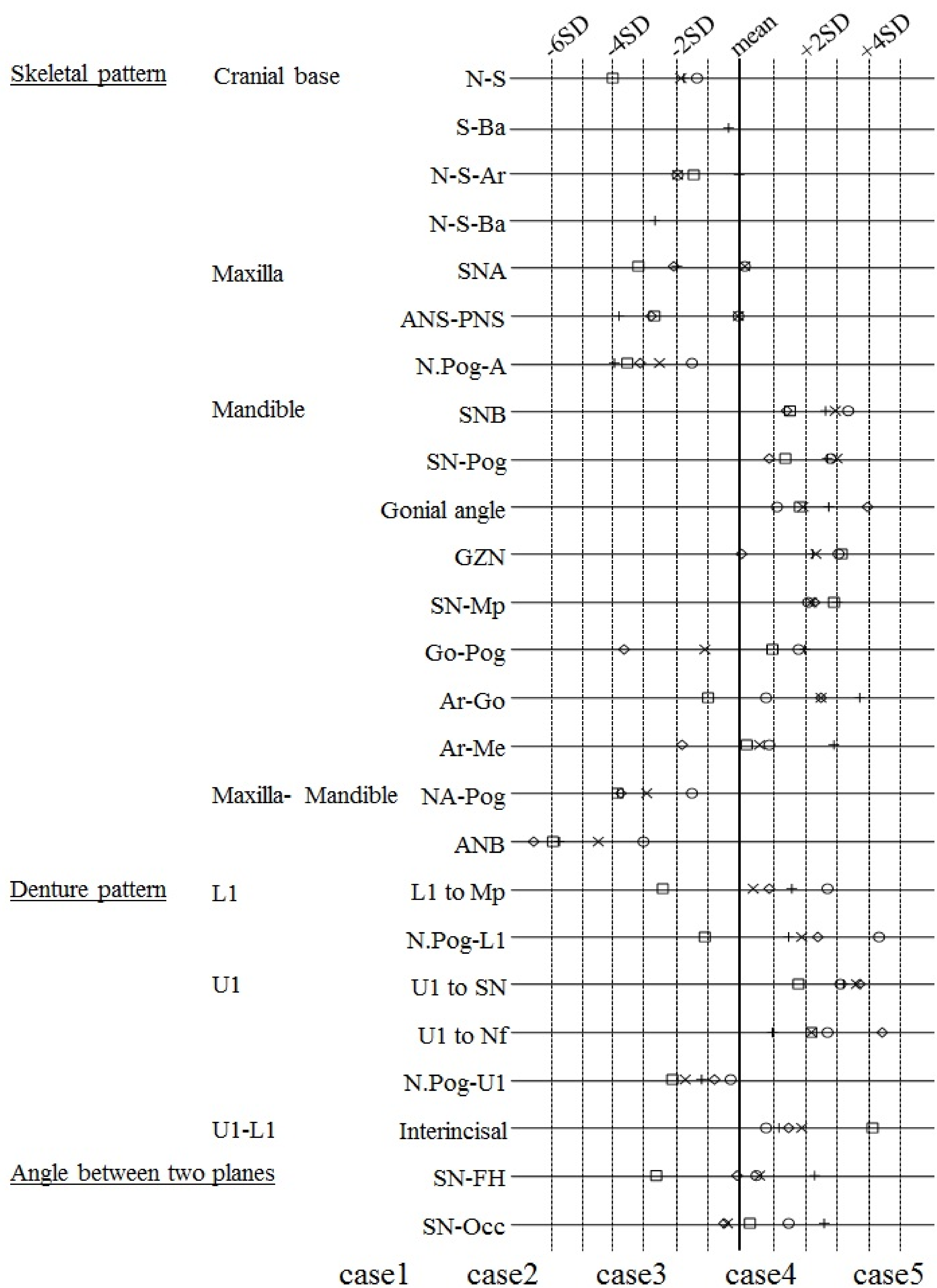

-6SD
-4SD
-2SD
mean
+2SD
+4SD
Skeletal pattern
Cranial base
N-S
S-Ba
N-S-Ar
N-S-Ba
Maxilla
SNA
ANS-PNS
N.Pog-A
Mandible
SNB
SN-Pog
Gonial angle
GZN
SN-Mp
Go-Pog
Ar-Go
Ar-Me
Maxilla- Mandible
NA-Pog
ANB
Denture pattern
L1
L1 to Mp
N.Pog-L1
U1
U1 to SN
U1 to Nf
N.Pog-U1
U1-L1
Interincisal
Angle between two planes
SN-FH
SN-Occ
case1
case2
case3
case4
case5

Fig.5