Dental anomalies in patients carrying the Apert syndrome and the Crouzon syndrome

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ABSTRACT

The present study aimed at analyzing the prevalence of dental anomalies in patients carrying Apert and Crouzon syndromes. The sample comprised 20 orthopantomograms of patients with Apert syndrome and 9 orthopantomograms of patients with Crouzon syndrome, who were regular patients of the Hospital for Rehabilitation of Craniofacial Anomalies – USP – Bauru/SP (HRAC/USP). The radiographs were selected from the files of the Dental Radiology sector of the Hospital. The recordings of the patients with syndromes allowed evaluation of the presence or absence of some type of cleft lip and palate. The most frequent dental anomaly observed was the eruption disturbance (retained and impacted teeth), probably due to the remarkable maxillary hypoplasia displayed by such patients.

KEY WORDS: Apert syndrome; Crouzon syndrome; orthopantomogram; acrocephalosyndactyly; tooth abnormalities

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INTRODUCTION

According to Alvares and Tavano (2000), growth or develpomental anomalies in the buccal anatomic structures that results in abnormality is called anomaly. These anomalies can be due to local condition linked to an inherent hereditary trend or can be a manifestation of a systemic disturbance. These anomalies, furthermore, can be associated to systemic disease or other developmental disturbances. According to Pasler (1992) both endogenous and exogenous factors could induce, directly or indirectly, alterations in the dental arch. For Castro et al. (2000), during odontogenesis, the dental anomalies can occur due to nutritional deficiencies (hypovitaminosis A, C and D, hypocalcemy), endocrine factors (hypothimia, hypo and hypertireoidism), infectious and inflammatory processes (syphilis, exanthematic fever), excessive intake of drugs (fluoride, antibiotics), hereditary diseases (meso and ectomesenchimal dysplasias), congenital diseases (congenital syphilis), local trauma and ionizing radiations. Alvares and Tavano (1998a) noted that the participation of the dentist is important in the search for probable ethyological causes of developmental disturbances. The dental anomalies as well as those of the maxillomandibular complex, can be restricted to the buccal cavity. However, they can also be result of a systemic alteration or be part of a general syndrome.

According to Kreiborg (1981) the syndrome of Crouzon or craniofacial dysostosis is a rare syndrome of developmental disturbance characterized by craniosynostosis (early fusion of the intracranial sutures), maxillar hypoplasia and exophthalmia. Peterson-Falzone (1996) stressed that the syndrome of Crouzon, also known as Disease of Crouzon, and the Apert syndrome have similar features in what regards structural anomalies, simptomatology and, perhaps, ethiology. Curiously, many symptoms are less severe in patients with Crouzon than in Apert (FIGURES 1, 2 and 3).

Apert syndrome is among the almost 600 described genetic syndromes and is classified as a type I acrocephalosyndactyly. The syndrome is due to a genetic mutation that interferes in the fibroblast growth factors (FGFR2), which is produced during the formation of gametes. Collin et al. (1995), Kreiborg et al. (1999) and Fernandes (2002) reported the main characteristics of Apert syndrome: craniosynostosis, retrusion of the midface and syndactyly in hands and feet. Smith (1985) described, among other features, cleft palate, heart defects, pulmonary atresia, tracheo-esophageal fistula, pyloric stenosis, polycystic kidneys, bifid uterus, hydrocephalia, repetition otitis, sleep apnea, severe acne, eye problems and macroglossia.

According to Peterson-Falzone (1996), Apert syndrome is autosomic dominant, although many cases are sporadic, meaning that they are a result of new mutation (FIGURES 4, 5, 6 and 7).

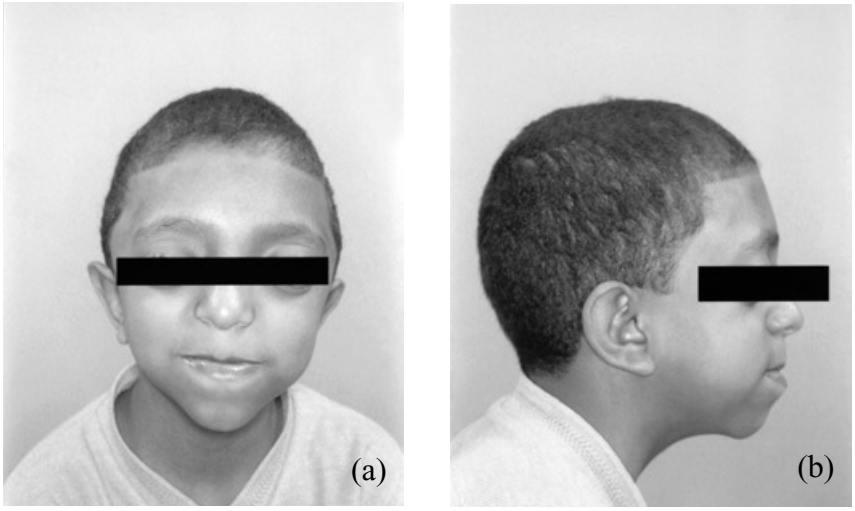

FIGURE 1 – Crouzon syndrome in a male. (a) Frontal view of the face. (b) Side view showing maxillar athresia.

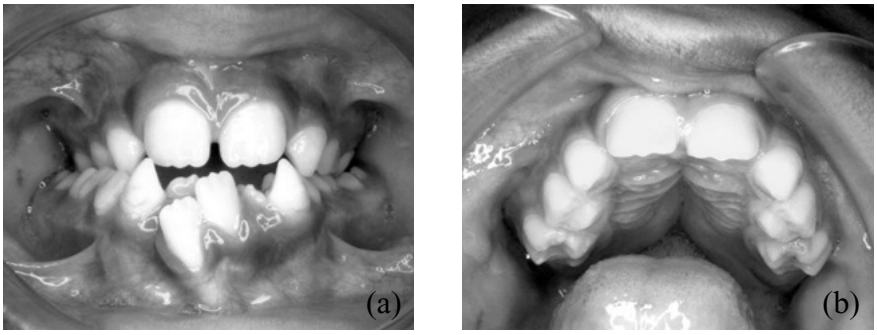

FIGURE 2 – Crouzon syndrome in male. (a) Buccal view (upper and lower arch), maxillary atresia and dental crowding. (b) Maxilla and the absence of clef lip and palate.

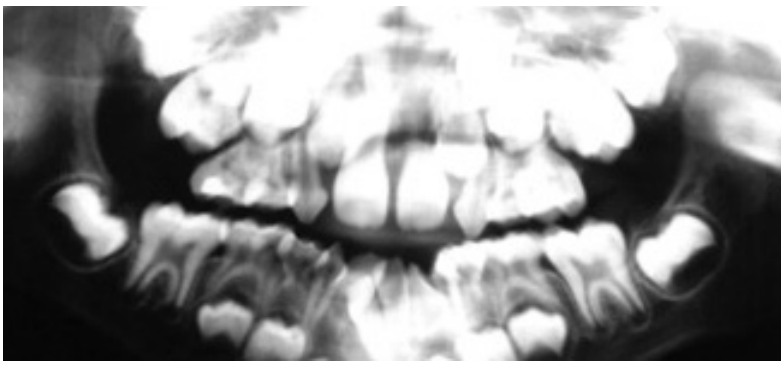

FIGURE 3 – Panoramic radiography in a male case of Crouzon syndrome.

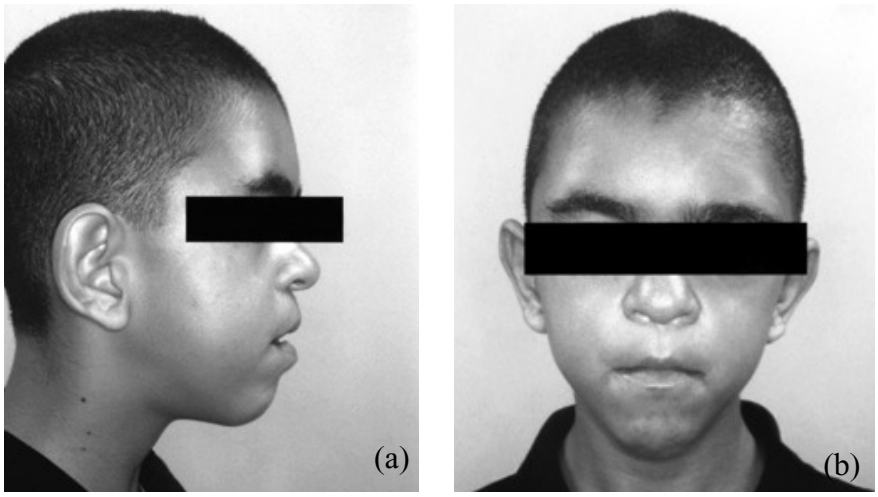

FIGURE 4 – Apert syndrome, male. (a) Frontal view. (b) Side view with maxillar athresia.

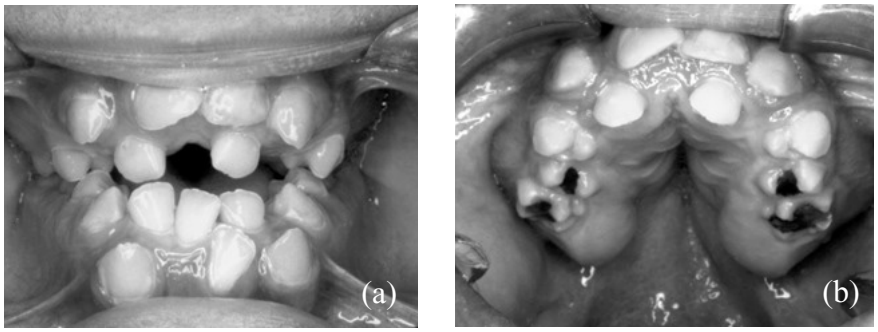

FIGURE 5 – Apert syndrome, male. (a) Buccal view (Upper and lower arch), maxillar atresia and dental crowding. (b) View of the maxilla and presence of cleft-lip and palate.

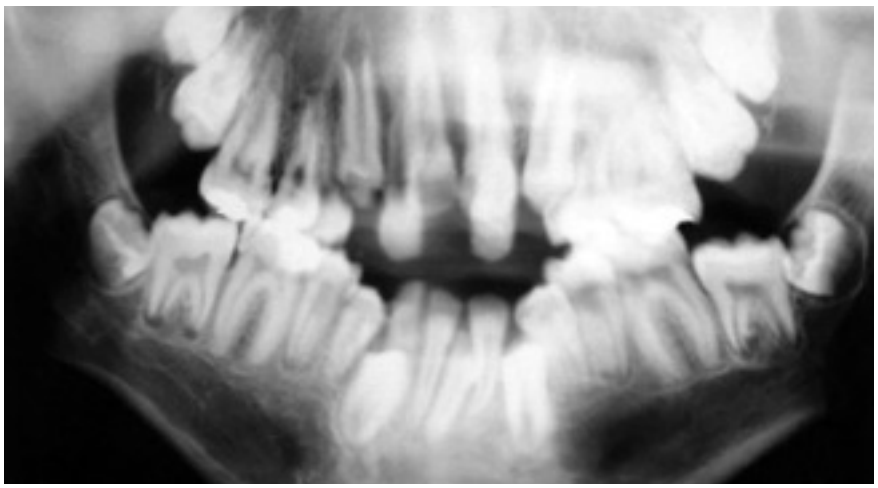

FIGURE 6 – Panoramic radiography of a male case of Crouzon syndrome.

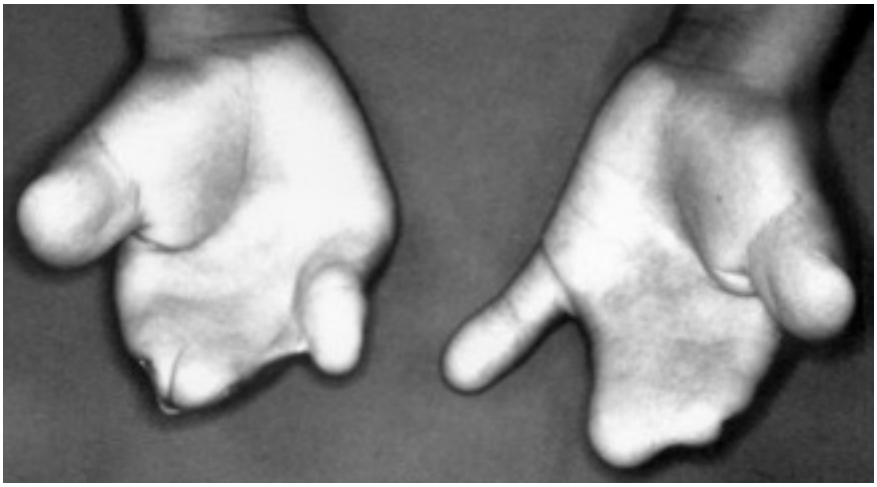

FIGURE 7 – Syndactyly in hands of a male case of Apert syndrome.

Taking these aspects into consideration the objective of the present study is to analyze the occurrence of clef lip and palate and of dental anomalies in individuals carrying Apert and Crouzon syndrome registered at the HRAC–USP, Bauru/SP.

MATERIALS AND METHODS

Twenty panoramic radiographies of patients with Apert syndrome and 9 radiographies of cases with Crouzon syndrome from the Hospital for Rehabilitation of Cranio-facial Anomalies from the University of São Paulo (HRCA–USP) were studied. According to Alvares and Tavano (1998b) and Freitas and Torres (2000), the panoramic radiography allow ample visualization of the structures of the third mean of the face, including all the maxillomandibular complex with a reduced exposition of the patient to X-ray radiation. This is the choice radiography for an initial evaluation of any new case.

The interpretation of the selected radiographies was done at the sector of radiology of the HRAC–USP with a cool light negatoscope and a magnifying glass. For each radiography it was made a report with all dental anomalies detected in a form designed specifically to this study (FIGURE 8).

Patient:	
ID:	Date:
Age:	Syndrome:
1. Shape alteration	
1.1. Macrodontia	
1.2. Microdontia	
1.3. Germination	
1.4. Fusion	
1.5. Dens in dente	
1.6. Enamel hypoplasia	
1.7. Imperfect dentinogenesis	
1.8. Hutchinson teeth	
1.9. Dilaceration	
1.10. Taurodontism	
2. Alteration in number	
2.1. Anodontia: total or partial	
2.2. Supranumerary teeth	
2.3. Predeciduous dentition	
2.4. Postdeciduous dentition	
2.5. Suplementary roots	
3. Alteration in eruption	
3.1. Retained teeth or impacted	
3.2. Zelayed eruption	
3.3. Concrescency	
3.4. Supraeruption eruption	

FIGURE 8 – Model of the report used in the study.

RESULTS

Only 20 out of the 47 cases of Apert syndrome registered at the HRAC, had a panoramic radiography, in which shape alteration (Hutchinson teeth, macrodontia, microdontia, dilacerations and fusion), number alteration (anodontia, supranumerary tooth and supranumerary cuspid), eruption alteration (retained teeth and delayed eruption) were observed (TABLE 1).

TABLE 1 – Dental anomalies found in panoramic radiographies of 20 cases of Apert syndrome from HRAC–USP of 20 cases of Apert syndrome from HRAC–USP.

Dental anomalies	n	%
Hutchinson teeth	2	10
Macrodontia	1	5
Microdontia	2	10
Dilaceration	3	15
Fusion	1	5
Anodontia	7	35
Supranumerary teeth	5	25
Supranumerary cuspid	2	10
Retained/impacted teeth	13	65
Delayed eruption	5	25

Thirty-one out of the 47 cases with Apert, did not have cleft lip and palate, 12 had incomplete post-foramen cleft, 2 had complete post-foramen cleft and 2 had submucous cleft palate, according to the Spina (1972) classification of clef lip and palate.

Only 9 out of the 19 cases with Crouzon syndrome registered at the HRAC, had panoramic radiography, in which it was observed shape alteration (Hutchinson teeth and macrodontia), alteration in number (anodontia and supranumerary teeth), eruption alteration (retained teeth and delayed eruption) (TABLE 3).

TABLE 3 – Dental anomalies found in 9 panoramic radiography of cases with Crouzon at the HRAC–USP.

Dental anomalies	Occurrence	%
Hutchinson teeth	1	11
Macrodontia	2	22
Anodontia	2	22
Supranumerary teeth	1	11
Retained/impacted teeth	3	33
Delayed eruption	1	11

None of the 19 cases with Crouzon syndrome at the HRCA showed clef lip and palate.

Ectopic eruption was not analyzed in this study since it cannot be evaluated by radiography. However, by studying the clinical photographs of these cases it could be seen that ectopic eruption was a common finding in both Apert and Crouzon syndrome cases.

DISCUSSION

According to Kreiborg and Cohen (1998) the syndrome of Apert and the syndrome of Crouzon are different disorders with different craniofacial developments. The craniofacial fenotypes are distinct in all ages. The differences between these two syndromes are well observed in childhood, but can be less marked with age. In general, the craniofacial morphological anomalies are more severe in the Apert syndrome than in Crouzon's (CASTRO et al., 2000; PETERSON-FALZONE, 1996).

In the present study it was observed that anodonty was more frequent in Apert (35%) than in Crouzon syndrome (22%). Dental anomalies were more frequent in Apert and tends to reflect the greater complexity of the clinical manifestation of this syndrome, as Kreiborg and Cohen (1998) and Peterson-Falzone (1996) remarked.

It was observed that 65% (13) of cases of Apert syndrome had dental impactation, 62% of them had impactation in the mandible and 38% in the maxilla. These data differ from the ones reported by Gorlin et al. (1990) and Kreibrog and Cohen (1992) that mention the maxilla as the place for impaction (96%).

In the present study, teeth more commonly affected by impaction were the upper and lower canines and the lower third molars. Supranumerary teeth were present in 25% of the Apert cases evaluated. Despite the absence of other data, the present results are supported by the description of the buccal manifestation in cases of Apert syndrome reported by Gorlin et al. (1990).

Data form the present study show that 16 cases, out of the 66 cases studied, showed some type of post-foramen cleft. All of them were cases of Apert syndrome. The finding at the HRAC–USP confirm the assertion by Tessier (1971) that cleft palate seems to be specific for Apert syndrome. The present findings are also in accordance with those of Solomon et al. (1973) for whom all cleft lip and palate in cases of Apert syndrome were limited to the posterior part of the hard palate and to the soft palate.

Kraus et al. (1967) and Cohen and Cohen Jr. (1971) observed some clinical buccal manifestation in patients with Apert syndrome.

They reported anomalies in the shape of teeth of 56 patients. In a third of this group the upper permanent incisives had a shovel shape and the second and third molars (lower and upper) showed reduced mesiodistal diameter. These authors also observed that the ectopic eruption in permanent teeth was present in half of the cases studied, more commonly in the region of the upper incisive and molars, which can be confirmed by the clinical photographs of the cases of Apert and Crouzon syndrome evaluated in the present study.

CONCLUSION

According to the results it is possible to conclude that:

– The most common dental anomaly observed in both syndromes was the alteration in dental eruption (retained/impacted).

– In Apert syndrome the teeth more commonly involved were the superior and inferior canines and the inferior 3th molars, whereas in the Crouzon syndrome superior canines were more affected.

– Anodontia was observed in 35% of cases with Apert, being the upper 2nd pré-mollars the more affected teeth.

– 16 (34%) out of 47 cases with Apert syndrome, had some sort of cleft palate

– None of the cases with Crouzon syndrome had clef lip and palate.

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