Different Types of Dysplasia in Pediatric Dental Practice

8

This chapter contains examples of different types of dysplasias on radiographic images taken with different radiographic techniques and their radiographic differential diagnoses. The aim of this chapter is to familiarize the reader with the different results of the techniques explained in the previous chapters. This is a grasp of what general pediatric dental practice could provide so at least the reader will get a better feel about how the images turn out and what information can be retrieved from them.

Names of distinguished colleagues who supplied the images for this chapter are mentioned with the radiographs. If there is no name mentioned with the radiographs, the radiographs were taken by the author of this book or collected from the different university clinics he has worked in (Ghent University in Belgium, University of Washington in Seattle, USA, and University of Western Australia in Perth, Australia).

8.1 Cleidocranial Dysplasia

Dysostosis cleidocranialis, or cleidocranial dysplasia as it is called today, is a syndrome associated with hyperdontia and in some occasions also with cleft lip and/or palate. These supernumerary teeth cause serious issues, as many don't even erupt and teeth stay embedded in the jaws, giving rise to dentigerous cysts. This rare familial disorder is characterized by defective formation of the clavicles (patient can bring shoulders extremely forward and almost touching if the clavicles are missing), delayed closure of the fontanelles (causing bulging of frontal, parietal, and occipital bones), and sometimes retrusion of the maxilla (a hypoplastic midfacial development). Figures 8.1 through 8.3 are three examples of patients with cleidocranial dysplasia.

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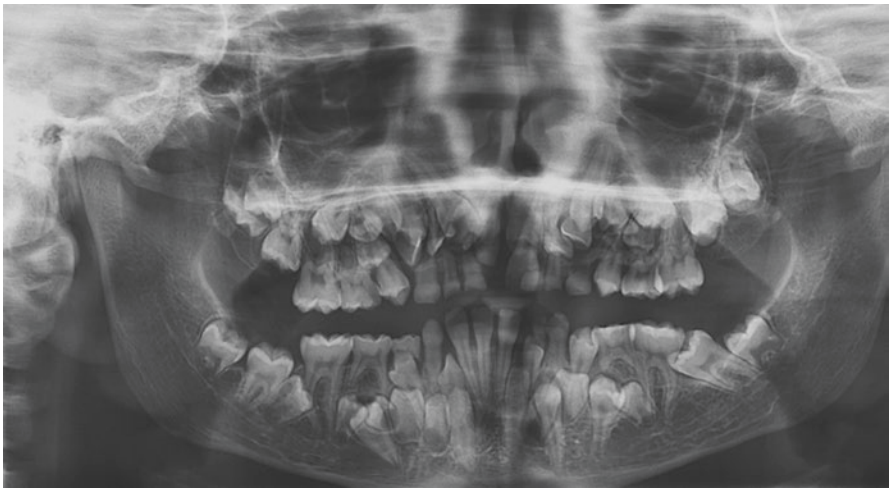

Fig. 8.1 This panoramic radiograph of a 10-year-old patient with cleidocranial dysplasia shows the haphazard organization of the teeth and the supernumerary tooth germs in both jaws (courtesy of Dr. Thierry Boulanger, Belgium)

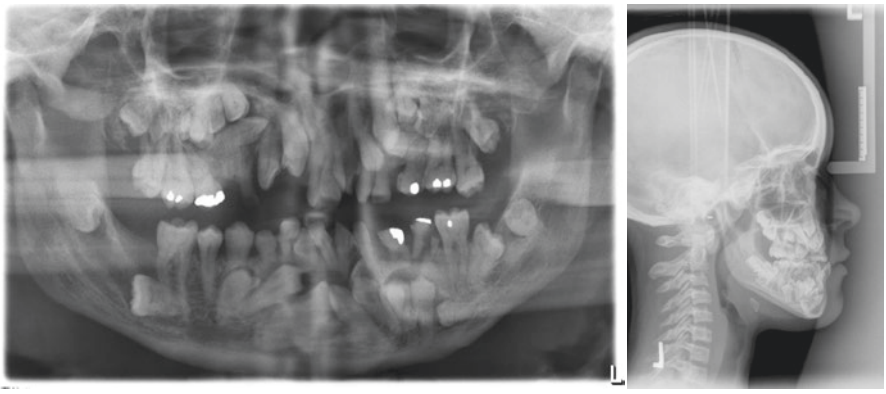

Fig. 8.2 This panoramic radiograph of a 15-year-old girl with cleidocranial dysplasia shows the impacted supernumerary teeth and also the superiorly displaced mandibular third molars. One can also appreciate the position of the supernumerary teeth in the maxilla obliterating the maxillary sinuses. The latter can also be seen on the cephalometric radiograph that was taken

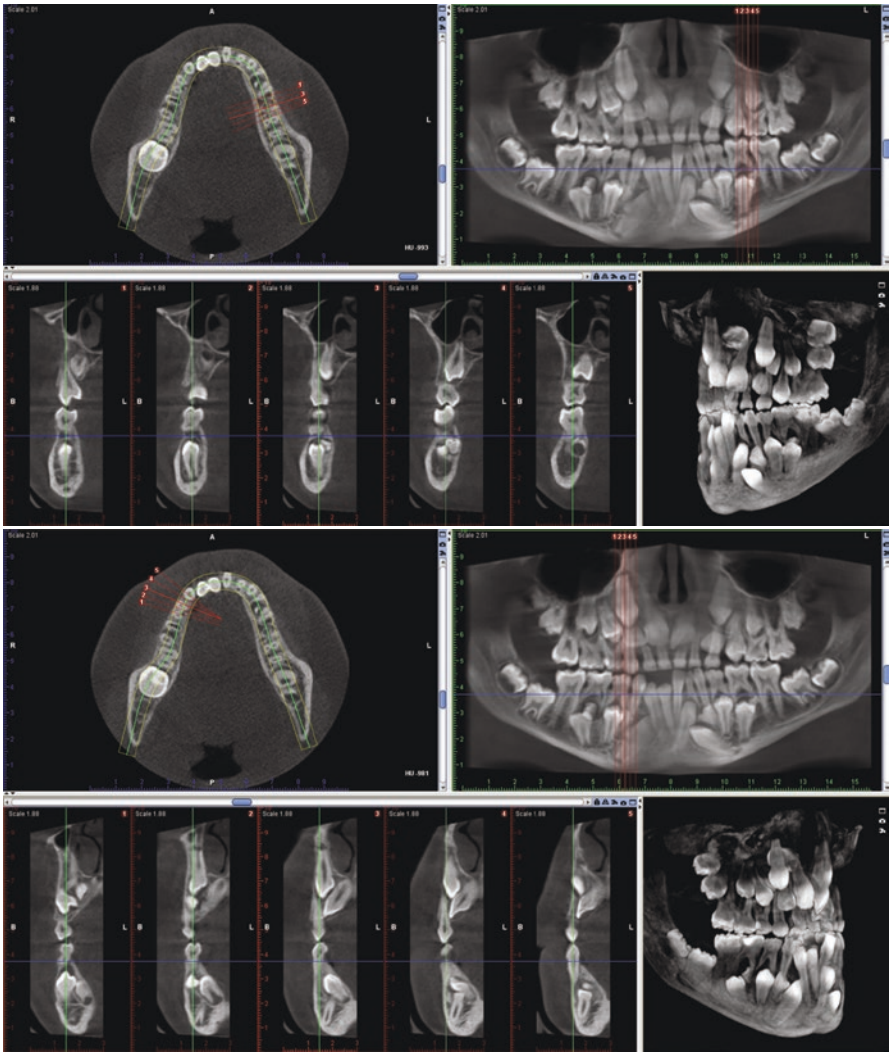

Fig. 8.3 This is a cone beam computed tomography of an 8-year-old girl with cleidocranial dysplasia, taken for surgical planning purposes to determine which supernumerary teeth were going to be sacrificed for extraction. The top image shows a couple of slices through the left-hand side of the patient's jaws, while the bottom images show the patient's right-hand side. Notice the supernumerary teeth on the lingual side in the mandible which can easily be missed on a traditional panoramic radiograph. The three-dimensional reconstructed radiographs can help the surgeon in this case to visualize the impacted supernumerary teeth better in relation to the erupted teeth

Cleidocranial dysplasia is a rare skeletal dysplasia with autosomal dominant inheritance (chromosome 6p21, OMIM 119600). A great number of different mutations are identified and studied, but what is interesting is the variable phenotype of the syndrome. Patients usually present with short stature, aplastic or hypoplastic clavicles, and delayed ossification of the calvaria, with accompanied delayed ossification of the sutures, leading to Wormian bones, wide forehead with broad midline furrow, midface hypoplasia, persistent mandibular symphysis, and several dental anomalies. The latter include lack of exfoliation of primary teeth, presence of multiple supernumerary teeth, abnormal anatomy especially of the roots of teeth, and not erupting permanent teeth.

8.2 Fibrous Dysplasia

Fibrous dysplasia (Figs. 8.4 through 8.7) is characterized by a localized change in the normal bone metabolism, resulting in replacement of cancellous bone with fibrous tissue. This translates in a typical radiographic appearance of fingerprint or orange peel appearance of the affected spongy bone. Expansion of the jaws is not uncommon and is usually the reason why radiographs are taken and how the pathology is found. There are mono- and poly-ostotic forms of fibrous dysplasia. The mono-ostotic form is more prevalent in the jaws; however the most common sites are ribs, femur, and tibia. The poly-ostotic form is usually found in children under the age of 10 years old.

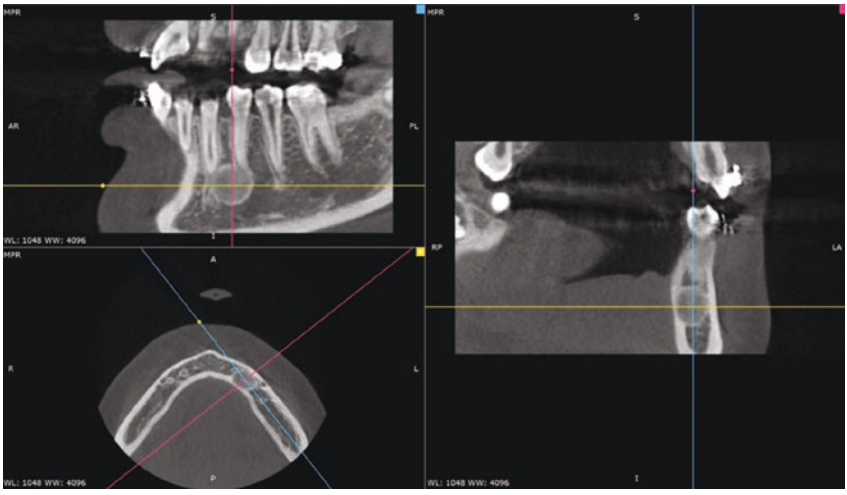

Fig. 8.4 This cone beam computed tomography shows a well-defined, corticated, uniform-appearing lesion, located between the apices of the left-hand-side mandibular canine and first premolar. The radiopaque border is typical for young lesions, whereas older lesions tend to have ill-defined borders. In this case the lingual cortical plate of the mandible is affected slightly and further expansion of the lesion can be expected. Since the lesion is not associated with the apex of a tooth, the differential diagnosis can exclude periapical cemento-osseous dysplasia, which by the way occurs in an older patient age group. The radiopaque border would be unusual for ossifying fibroma, which cannot be excluded from the differential diagnosis

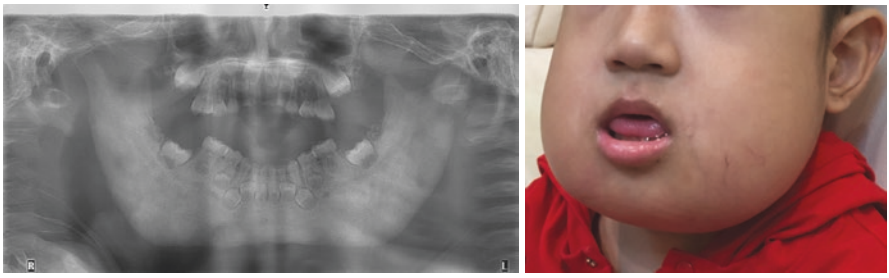

Fig. 8.5 The panoramic radiograph of this 4-year-old boy shows massive changes in the circumference of the mandible, as well as in the radiographic appearance of the cancellous bone which is typical for fibrous dysplasia. As can be seen from the cropped clinical photograph, the disfigurement is substantial (courtesy of Dr. Nik Kantaputra, Thailand)

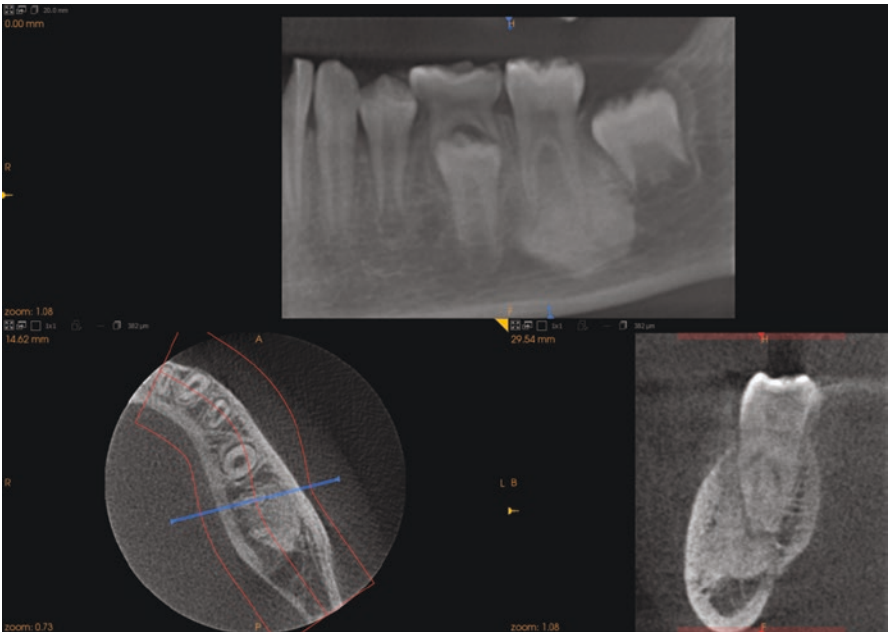

Fig. 8.6 This is a small-field-of-view cone beam computed tomography of a patient with fibrous dysplasia around the roots of the left-hand-side permanent first mandibular molar. In the coronal section one can appreciate the difference in radiopaque character between the lesion, the dentin of the tooth involved, and the cortical bone. One can also differentiate the periodontal ligament space between the tooth's root and the lesion, which helps identifying it from a dense bony island, hypercementosis, and cementoblastoma

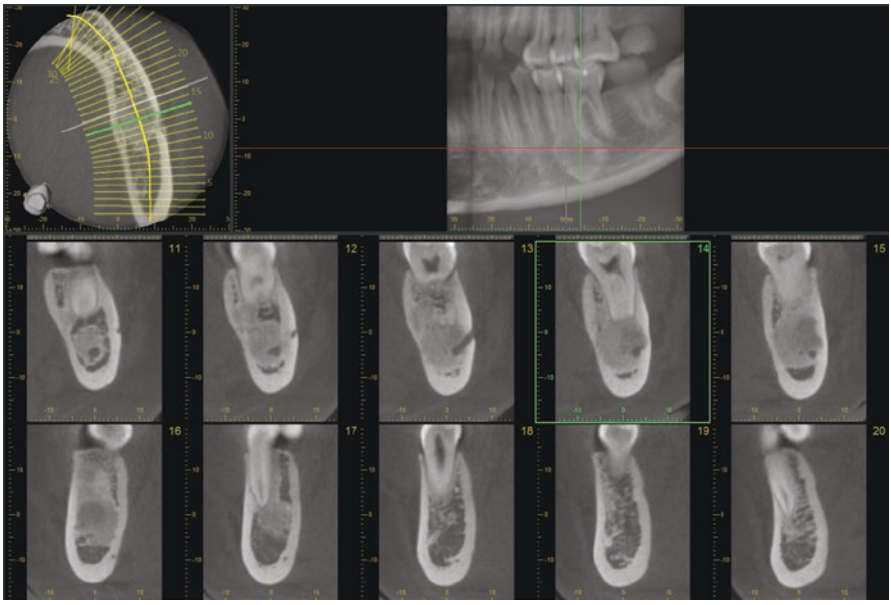

Fig. 8.7 This is another case of fibrous dysplasia which is located between the left-hand-side mandibular second premolar and first molar. One can appreciate the lesion forming around the inferior alveolar canal and expanding into the buccal and lingual cortical plates of the mandible. The integrity of the teeth is not affected, which differentiates this lesion from hypercementosis, cementoblastoma, and dense bony island

The pathogenesis of fibrous dysplasia is known to involve a postzygotic mutation of the GNAS1 gene. This results in an altered biochemical pathway, which leads to constitutive activation of G proteins and proliferation of undifferentiated mesenchymal cells. Mutations occur sporadically and postzygotically, which results in mosaicism and highly variable presentations. Abnormal bone matrix and growth of abnormal tissue are the consequences. It can occur at any age, both as monostotic and poly-ostotic or as part of McCune-Albright syndrome. The poly-ostotic variant affects the skull often, with 50–100% of cases having this involvement, with a preference for the frontal bone. The monostotic variant may arrest after puberty, whereas the poly-ostotic variant may continue through adulthood. Patient can have symptoms, like pain and mass effect on adjacent structures. This condition is benign, but in less than 1% can turn malignant. If asymptomatic, fibrous dysplasia does not require treatment.

8.3 Ectodermal Dysplasia

Ectodermal dysplasia is a large group of autosomal dominant, autosomal recessive, and X-linked inherited conditions in which two or more ectodermal derived tissues fail to develop properly, resulting in hypoplasia or aplasia of skin, hair, nails, teeth, salivary glands, or sweat glands. Teeth can be missing and/or shapes can be aberrant (usually conical) and both dentitions are involved. In general these patients will show hypodontia (Figs. 8.8 and 8.9) or even anodontia.

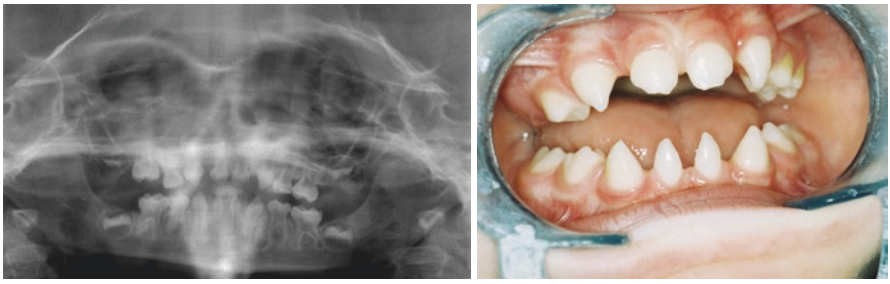

Fig. 8.8 This is a panoramic radiograph of a 4-year-old boy with hypohidrotic ectodermal dysplasia. Notice the conical shaped anterior teeth on both the panoramic radiographs as the clinical picture. Unfortunately there is no follow-up panoramic radiograph of the boy at an older age to verify hypodontia in the permanent dentition

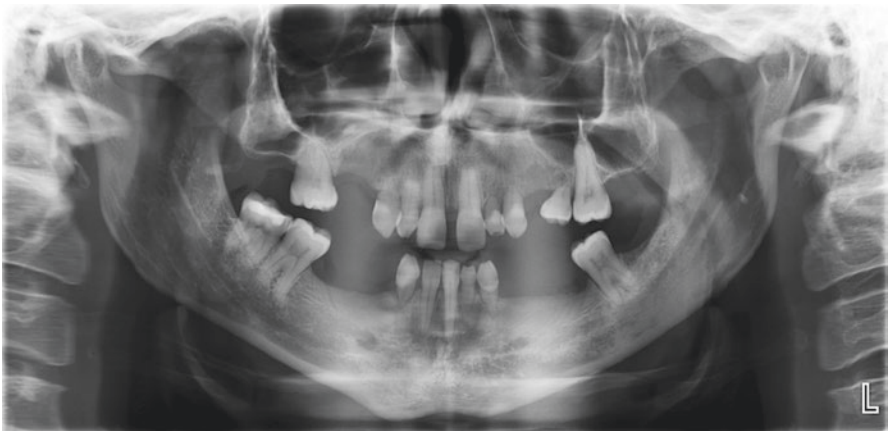

Fig. 8.9 This is a panoramic radiograph of an adult patient with ectodermal dysplasia. Notice the severe hypodontia, and most of the permanent molars appear to have a normal anatomy, while the opposite can be observed for the incisors and canines, which appear rather conical and short in root length

Human WNT10A mutations are associated with developmental dental abnormalities and adolescent onset of a broad range of ectodermal defects. Hair follicles, sebaceous glands, taste buds, nails, and sweat ducts are also affected. The WNT10A gene is the most commonly mutated gene in human non-syndromic selected agenesis of permanent teeth and these mutations are also associated with ectodermal dysplasia syndromes odonto-onycho-dermal dysplasia and Schöpf-Schulz-Passarge syndrome. Patients with these mutations exhibit variable developmental dental defects, which include microdontia in the deciduous dentition, defective root and molar cusp formation, and even anodontia of the permanent dentition. Besides these dental associations, other tissues are affected as well, resulting in palmoplantar keratoderma, thinning of hair, sweating abnormalities, smooth tongue surface, and defective nail growth appearing in adolescence or even at later age.

8.4 Segmental Odontomaxillary Dysplasia

Hemimaxillofacial dysplasia (Fig. 8.10) is a synonym for this anomaly, which is developmental and of unknown etiology. The process affects the maxilla unilaterally and also affects the teeth and the attached gingiva in that section. The alveolar process is enlarged, with or without hypertrophy of the gingiva, and with dental anomalies. The latter involve missing teeth, hypoplastic teeth, and/or non-erupting teeth. In some cases, the following features have been observed as well: ipsilateral hypertrichosis, closely packed sebaceous glands in the upper lip, hyper- or hypopigmentation of the skin, Becker's nevus (pigmented hairy epidermal nevus), cleft lip, and/or palate. Depending on the hyperplasia of the alveolar crest, facial enlargement can occur, with facial asymmetry resulting. Radiographically the orientation of the trabeculae in the affected side of the maxilla is vertical. The roots and crowns of the affected primary teeth are larger than on the unaffected side. Absence of normal resorption of these teeth and a hypoplastic maxillary sinus are also common features. Permanent teeth eruption is delayed often. Differential diagnosis with fibrous dysplasia might be challenging.

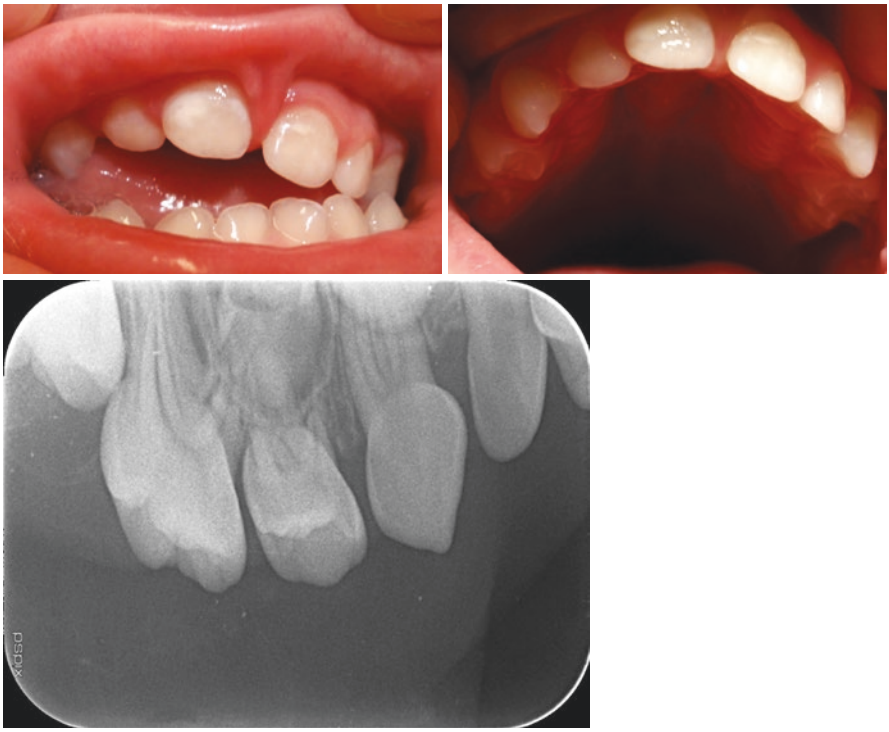

Fig. 8.10 The clinical pictures of this 5-year-old girl with segmental odontomaxillary dysplasia show the changes in the position of the teeth on the affected side of the maxilla. This case did not have significant mucosal hypertrophy, though the position of the teeth worried the parents of the girl and the dentist. On the radiograph one can depict the double-rooted right-hand-side maxillary deciduous canine and the large crown of the second deciduous molar (courtesy of Dr. Anouk Eloot, Belgium)

Further Reading

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