Examples of Congenitally Acquired Pathology in Pediatric Dental Practice

10

This chapter contains examples of congenitally acquired pathology. Obviously not all types of pathology were captured in this textbook chapter, but 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).

10.1 Cleft Palate Patients

The etiology of cleft lip or palate has a genetic component in 40% of cases. If one or both parents are affected, the chance of having a child with a cleft lip and/or palate is higher than if none of the parents are affected. Cleft lip, with (58%) or without cleft palate (22%), is more seen in males, whereas a cleft palate alone (20%) is twice as often observed in girls. The prevalence of isolated cleft lip is 1 in every 1000 live births, and the prevalence of isolated cleft palate is 1 in every 2000 live births. It is obvious that in terms of radiation dose, cone beam computed tomography can be beneficial in planning and following up of the surgeries these children with palatal clefts require, compared to medical CT. Figures 10.1 and 10.2 illustrate the use and benefits (image quality and low dose compared to medical CT) of cone beam computed tomography in patients with cleft palate.

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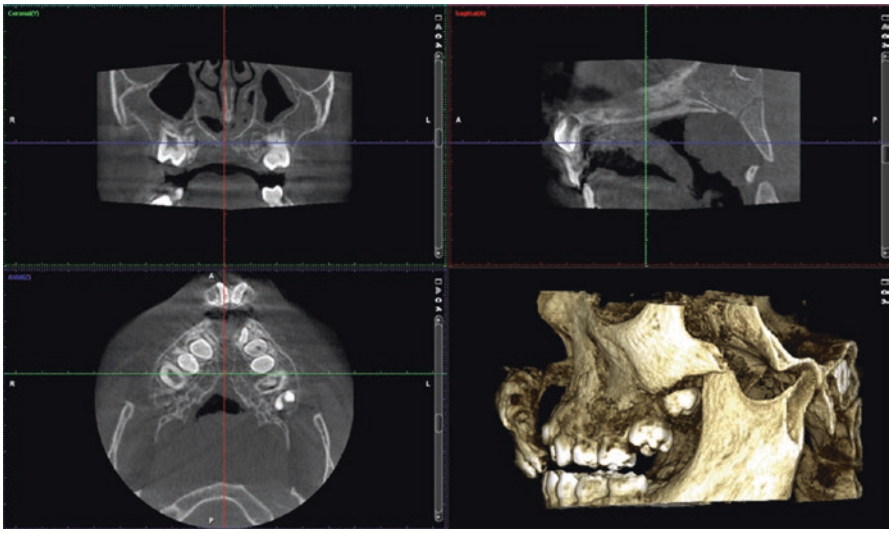

Fig. 10.1 This is a cone beam computed tomography of the maxilla of a 10-year-old patient with a cleft lip and palate. The defect in the bony palate can be clearly observed and assessed and the surgeon can plan their surgery to fix the premaxilla. Also notice the mucosal hypertrophy in the left-hand-side maxillary sinus and the spheno-occipital synchondrosis

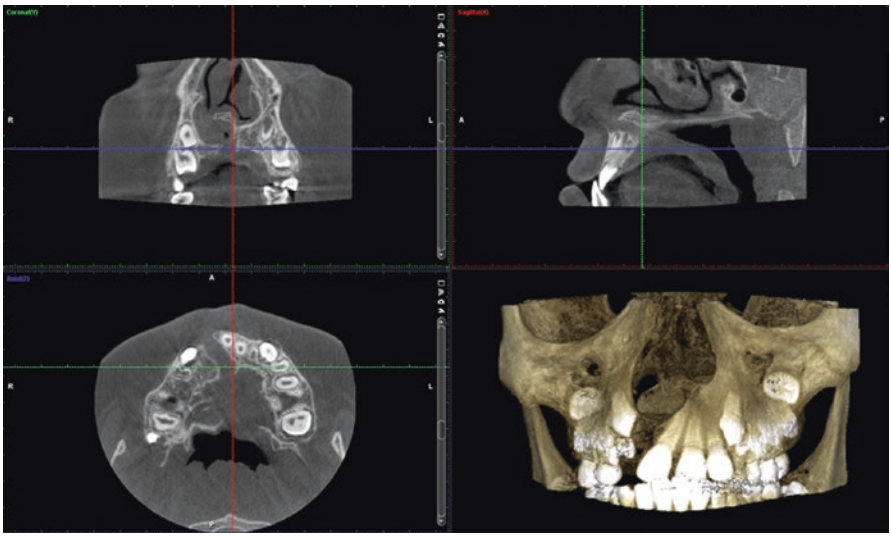

Fig. 10.2 This is a cone beam computed tomography taken in an 8-year-old boy with a unilateral cleft lip and palate on his right-hand side. Typical is the missing tooth (right-hand side maxillary lateral incisor) in the cleft and the deviation of the nasal septum as is clear from these images. Also notice the presence of the spheno-occipital synchondrosis and the rather large foramen of the sphenoid sinus into the nasal cavity (sagittal view)

A synchondrosis is a fusion between two bones to form one bone. The sphenooccipital synchondrosis consists of the basilar part of the occipital bone and the body of the sphenoid bone, hence the synonym called sphenobasilar synchondrosis. They both form the clivus, which is the slope from the posterior clinoid process towards the foramen magnum. The union between the two bones signifies adulthood and although there is not 100% consensus in the literature when the synchondrosis is ossified, the age span is between 18 and 25 years of age, with females being 2 years ahead of males, in general. Ossification starts from the clivus towards the base of the skull, so from intracranial to extracranial.

10.2 Hemifacial Microsomia

Hemifacial microsomia or *hemifacial hypoplasia, craniofacial microsomia,* and *lateral facial dysplasia* are all synonyms and syndromes associated with it are Goldenhar syndrome and oculo-auriculo-vertebral dysplasia. It is the second most common developmental craniofacial anomaly after cleft lip and palate. The patients show reduced growth, and development of the affected side of the face, rendering their appearance asymmetrical. This is due to the fact that the anomaly is related to the first and second branchial arches. If bilateral, it is called craniofacial microsomia.

If an entire side of the cranial anatomy is affected, the following structures will be involved and hypoplastic: mandible, maxilla, zygoma, external and middle ear, hyoid bone, parotid gland, vertebrae, cranial nerves V (trigeminal nerve) and VII (facial nerve), and all associated soft tissues (e.g., microtia, musculature). Hypodontia and/or failure of teeth to erupt on the affected side are common features, as is malocclusion obviously. There is a progressive form which is known under the name of Parry-Romberg syndrome. The latter patients do not have any features at birth and their ears are normal. Figures 10.3 and 10.4 illustrate a case of hemifacial microsomia where cone beam computed tomography was used. It is obvious that collaboration with a dentomaxillofacial or medical radiologist is essential as this large field of view CBCT scan goes beyond the field of expertise of a paediatric dentist and additional information from either radiology specialist may affect the comprehensive treatment plan of the patient.

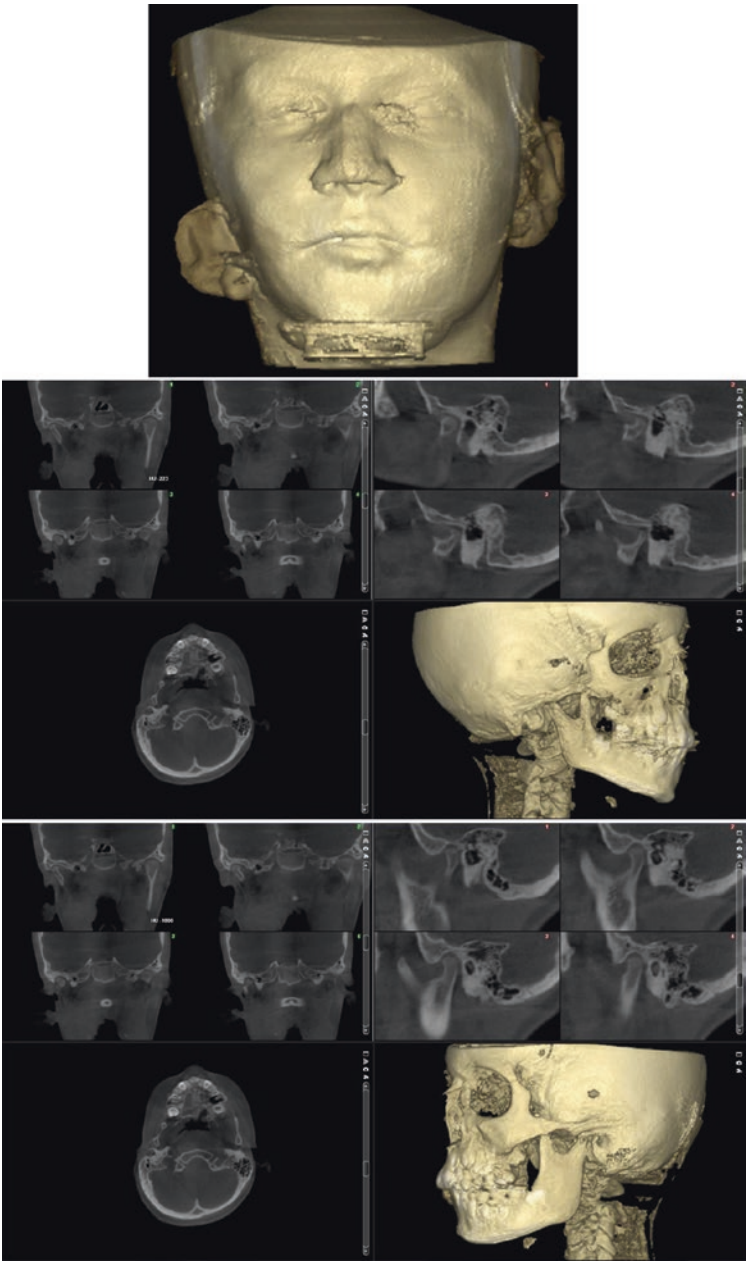

Fig. 10.3 This is a large-field-of-view cone beam computed tomography of a girl with hemifacial microsomia on the right-hand side of her face. At the top the three-dimensional reconstructed image of the soft tissues is shown to illustrate the shape of the right-hand-side auricle, which is obviously hypoplastic and more inferior than the auricle on the patient's left-hand side. The other images illustrate the difference in the mandibular condyles (seen in both lateral and anterior views—top image being the patient's right-hand side), rami, and mandibular notches

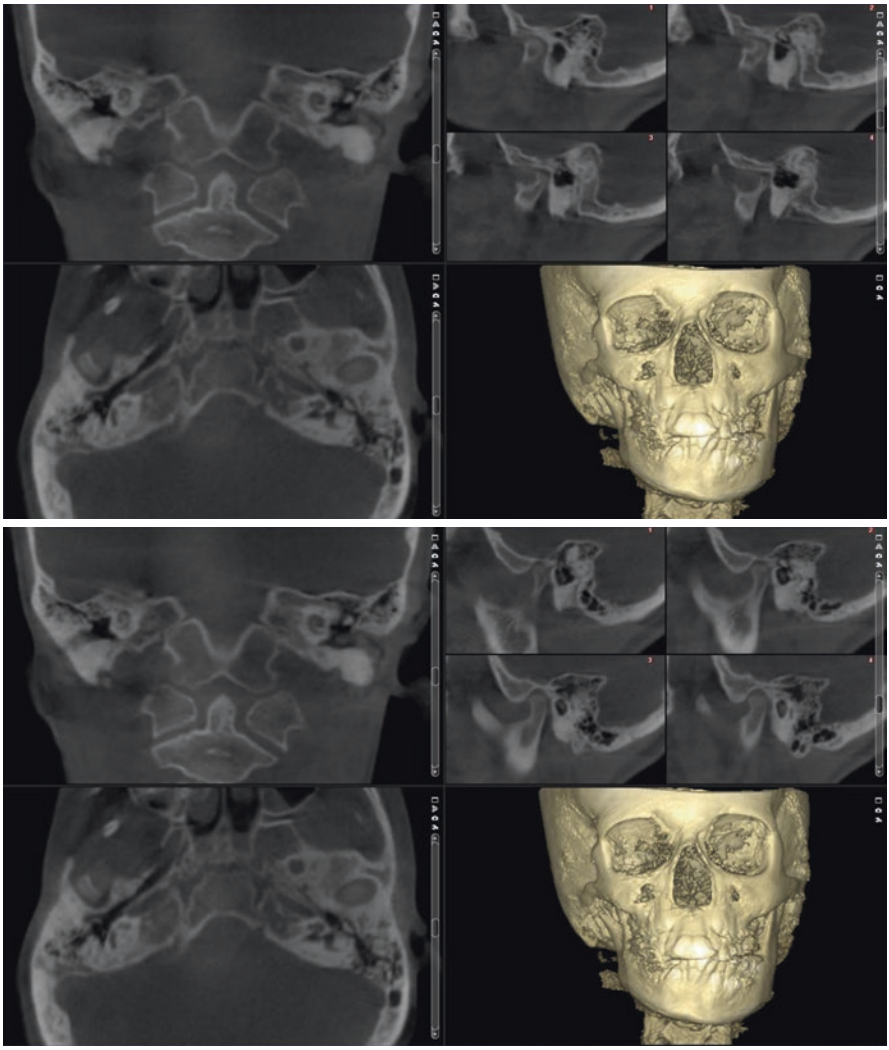

Fig. 10.4 This is the same patient as in Fig. 10.3. These images illustrate the differences in the ossicles between the right-hand side (top) and left-hand side (bottom). The aberrant anatomy of the inner right ear explains why the patient is deaf on that side. From the three-dimensional reconstructed images one can appreciate the malocclusion in this young girl's dentition due to her hemifacial microsomia

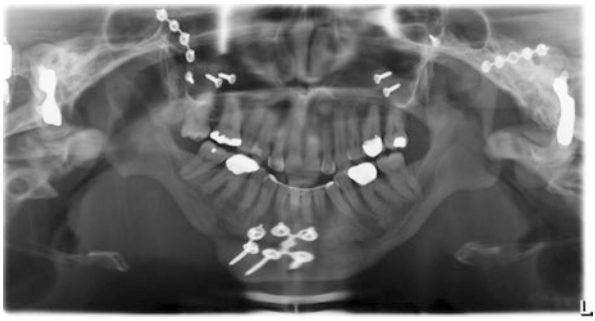

Fig. 10.5 This is a panoramic radiograph of a patient with Treacher Collins syndrome who had orthognathic surgery done. Despite the surgical paraphernalia in place to reconstruct the craniofacial skeleton, one can distinguish the following features: hypoplasia of the zygomatic arches, steep mandibular angles with distinct antegonial notches, auricular implants placed to establish a better aesthetical appearance, and a genioplasty to reconstruct the anterior mandible and to create a more prominent chin

10.3 Treacher Collins Syndrome

Treacher Collins syndrome and *mandibulofacial dysostosis* are synonyms and it is the most common type of mandibulofacial dysostosis. These patients can have a wide range of anomalies, but the most prominent one is the absence or hypoplasia of the zygomatic bones, hypoplasia of the mandible with a steep angle, (Fig. 10.5) and malformation of the auricles and often also the ossicles, all which give these patients their typical facial appearance: small narrow face with a high palate, downward inclination of the palpebral fissures, downturned wide mouth, and sometimes complete absence of auricles and external acoustic meatus. Occasionally patients also have cleft lip and/or palate. If the ossicles are affected, deafness is also a feature.

10.4 Mucopolysaccharidosis

Mucopolysaccharidosis (MPS) is caused by deficiency of iduronate-2-sulfatase which is characterized by excretion of dermatan sulfate and heparin sulfate in urine. Hurler-Hunter syndrome (Fig. 10.6) is another terminology often used; however, Hurler and Hunter are two separate entities or syndrome types of MPS, the former being autosomal recessive inherited and the gravest and the latter being X-linked recessive. Typical orofacial features of MPS include heavy brow ridges, stiff temporomandibular joints, cloudy degeneration of the corneas, macroglossia, gingival hyperplasia, thin enamel, pointed cusps sometimes (Morquio-B type), and impacted teeth.

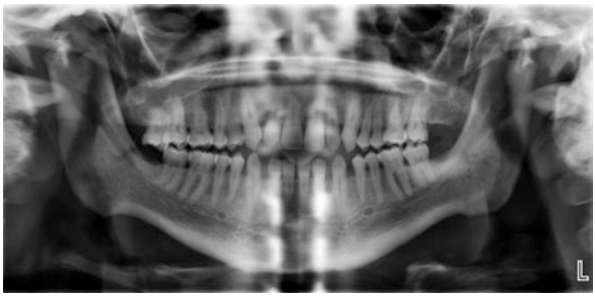

Fig. 10.6 This patient, with Hunter-Hurler syndrome diagnosis, was seen for a dental checkup. From this panoramic radiograph one can appreciate the unusual spacing in the anterior mandible, due to gingival hyperplasia. The canines show rather conically shaped crowns, but what is most prominent is the aberrant anatomy of the mandibular rami and their condyles, with posterior concavities and absence of mandibular notches and prominent coronoid processes. The latter can explain the restricted mouth opening and jaw movements. Distinct antegonial notches are also present, which can be related to the forces applied to the masseteric muscles

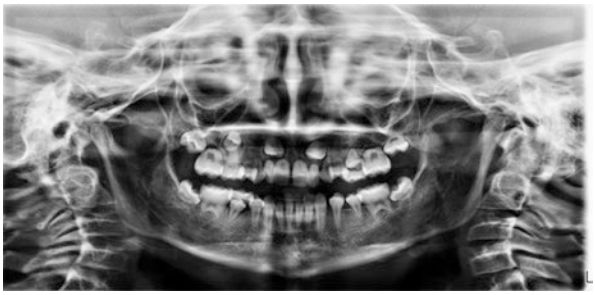

Fig. 10.7 This 11-year-old patient had an alveolar rhabdomyosarcoma of the hard palate, diagnosed at age 3. The panoramic radiograph shows generalized short roots. The latter is caused by chemotherapy and probably partially by the radiation therapy the patient received earlier. One can appreciate the longer root lengths on the mandibular incisors, compared to the maxillary, which can be linked to the therapy and the period in the patient's life the therapy was delivered (courtesy of Dr. Maite Demeester, Belgium)

10.5 Alveolar Rhabdomyosarcoma of the Palate

Rhabdomyosarcoma is a malignant neoplasm of skeletal muscle origin and it is the most common soft-tissue sarcoma in children. Approximately 40% occur in the head and neck region and the genitourinary tract is the second most common location. In the head and neck region, the orbit is most often affected, followed by nasal cavity and nasopharynx. The palate is the most common site if this neoplasm occurs intraorally (Fig. 10.7). In the latter case, they usually have a polypoid clinical appearance, sometimes resembling a cluster of grapes (botryoid). Treatment consists of surgical removal, chemotherapy, and radiation therapy. If no metastasis is present, 5-year survival rates are around 60–70%.

Further Reading

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