Atlas of Pediatric Oral and Dental Developmental Anomalies

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Preface

The problem of disturbed enamel and dentine formation leading to structural defects (anomalies) is important for dental surgeons who are dealing with the clinical features of such anomalies in their daily practice. The degree of defect may vary from minor to extensive, affect one or more dental structures, and will be affected by both the severity and length of the causative insult. The correct causal diagnosis obtained from the individual clinical manifestations will allow the clinician to select the most appropriate management for each patient. Of equal importance, this knowledge will allow the clinician to reassure the parent/guardian and explain to them why the teeth appear different.

This handbook has been prepared in a format that we hope will encourage readers to correlate their personal experiences with that of the authors. This will then allow them to self-test their personal diagnostic skills of the more common conditions.

The structure of this book has been organized with an initial brief review of the normal dento-facial structure, progressing to discussions on dental and oral anomalies, and then finally considering the more frequent syndromes associated with tooth disturbances. We hope this will give the reader a better understanding of anomalies, along with an appreciation of how clinical presentations can differ during the long course of development of the teeth and jaws.

Ghassem Ansari, Mojtaba Vahid Golpayegani, and Richard Welbury

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Oral and Dental Anatomy

The oral cavity consists of soft and hard tissues. The lips, cheeks, tongue, gingivae, palate, and tonsils are the former, while the teeth are the latter. The oral cavity is bounded by the lips anteriorly, the nasopharynx posteriorly, the cheeks laterally, the tongue and sublingual tissues inferiorly, and the soft and hard palate superiorly. Various muscles, nerves, and vascular systems contribute to these surrounding structures. The muscles of the oral cavity include mylohyoid, geniohyoid, stylohyoid, hyoglossus, glossopharyngeal, thyroglossus, buccinator, masseter, medial and lateral pterygoid, orbicularis oris, and temporalis. These muscles, together with their tendons, nerves, and blood vessels, keep the oral cavity functional.

1.1 The Lips: Macro Anatomy

The lips are composed of the muscular layer of orbicularis oris, connective tissue, dermis, and mucosa (Figure 1.1). The red vermilion border and its junction with the skin and mucosa at its outer and inner borders may vary in width between races and genders. Lips may have a different posture at rest, including: (i) sealed or competent, and (ii) not sealed or incompetent. Lip position may affect the alignment and profile view of teeth and occlusion (Figures 1.2–1.4). In certain circumstances, the lips appear shorter than normal, or the jaws are not in normal skeletal relationship, with a large part of the maxillary labial gingiva being visible during speech and smile. This condition is often referred to as "gummy" smile (Figure 1.5). Alternatively, there are cases where a longer-than-normal lip length or lost vertical height is noted due to lost or missing teeth – for example, ectodermal dysplasia. This in turn could cause the lips to overlap heavily, producing the appearance of an edentulous individual (Figure 1.4).

1.2 The Palate

The palate is divided into two major parts – *soft* and *hard* palate, with each of them having specific characteristics related directly to the role they play in different oral functions. The hard palate is supported by a hard, bony structure in the roof of the mouth, while the soft palate is mainly supported by fibrous tissue. The hard palate is covered with keratinized membrane, with a prominent eminence at the anterior mid-line located on top of the incisive foramen of maxillary bone, called the "incisive papilla." The nasopalatine nerve and blood supply pass through this foramen. "Rugae" are the anterior rough mucosal folds of the palate located on either side of the incisive papilla and midline raphe (Figures 1.6 and 1.7).

The soft palate, in contrast, consists of muscles, salivary glands, and neurovascular components. The uvula is a soft, small,

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Figure 1.1 Normal intraoral tissue and teeth.

Figure 1.2 Lips of a newborn.

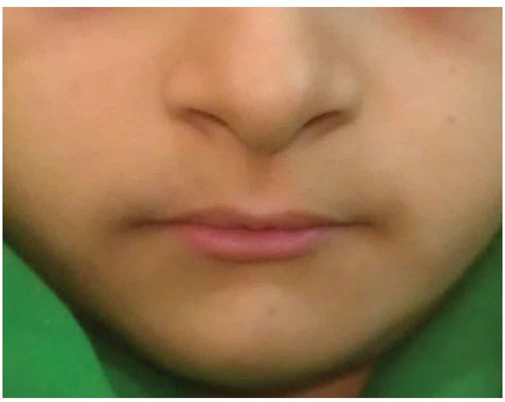

Figure 1.3 Lips of an 8-year-old.

Figure 1.4 Lateral lip pattern in ectodermal dysplasia.

Figure 1.5 High upper lip resulting in a "gummy" smile.

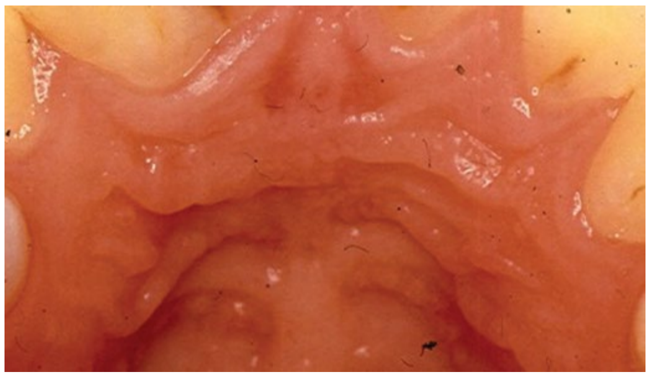

Figure 1.6 Normal palatal appearance, 11-year-old.

Figure 1.7 Normal palatal appearance, 8-year-old.

double-sided eminence of soft tissue located at the postero-inferior aspect of the soft palate. During swallowing, the soft palate and the uvula move together to close off the nasopharynx, preventing food from entering the nasal cavity.

1.3 The Tongue

The tongue is a muscular structure attached to the floor of the mouth at it's posterior. The ventral part of the tongue is covered with a thinly keratinized mucosal membrane firmly attached to the underlying muscles. The lingual frenulum is a thin layer of membranous tissue that attaches the anterior half of the tongue at its midline to the muscular structures in the floor of the mouth. The dorsal surface of the tongue is divided into two parts; the anterior two-third, and the posterior one-third (also known as the "pharyngeal" part). The border between these two segments is a shallow "V"-shaped groove, with the apex of the "V" lying posteriorly. Occasionally, there is a pit located at this apex, known as the "foramen caecum."

The dorsal part of the tongue contains several types of papillae that function as taste organs: filiform; fungiform; foliate (Figures 1.8–1.10); and circumvallate forms.

1.4 The Cheek and Floor of the Mouth

The mucosal structure forming the interior surface of the cheek is non-keratinized, such as the floor of the mouth. These parts of the mouth contain numerous blood vessels and

Figure 1.8 Multiple filiform papillae on dorsum of tongue.

Figure 1.9 Foliate papillae on lateral tongue border.

Figure 1.10 Larger red fungiform papillae on dorsum of tongue.

nerve bundles. The main salivary glands and their ducts are located mainly in the cheeks (parotid glands and parotid ducts) and floor of the mouth (submandibular and sublingual glands and their ducts). The parotid is the largest salivary gland, with the parotid duct exiting from the frontal portion of the gland, passing over the masseter muscle and buccal pad of fat, then penetrating the body of buccinator muscle, before opening into the oral cavity via the parotid papilla close to the second maxillary molar. The parotid glands secrete mainly serous saliva. The submandibular and sublingual are mixed serous/mucous glands located in the floor of the mouth on either side of the lingual frenum, close to the lingual surfaces of the lower incisors. They look different in clinical view when in rest or in tension (Figure 1.11a and b).

The mylohyoid and digastric muscles help to form the floor of the mouth alongside the base of the tongue.

1.5 The Periodontium

The periodontium includes ligament tissue bundles that control the teeth within their bony alveolar sockets and attaches them to the alveolar bone in one side while attaching to the cementum of the teeth on the other side. Vascular and nerve bundles are also present within the collagenous ligament layers. The outer layer of cementum is covered by a cellular layer, while the inner layer adjacent to dentine is mainly acellular. The gradual and continuous formation of cementum is responsible for the “compensation” of tooth structure loss due to attrition throughout life, as well as for the production of new connections and bonds between the root surfaces and the periodontal ligaments (Figures 1.12 and 1.13).

1.6 The Periodontal Ligament (PL)

The PL is a relatively firm fibrotic connective tissue that is located in the space between the root surface and alveolar bone surface.

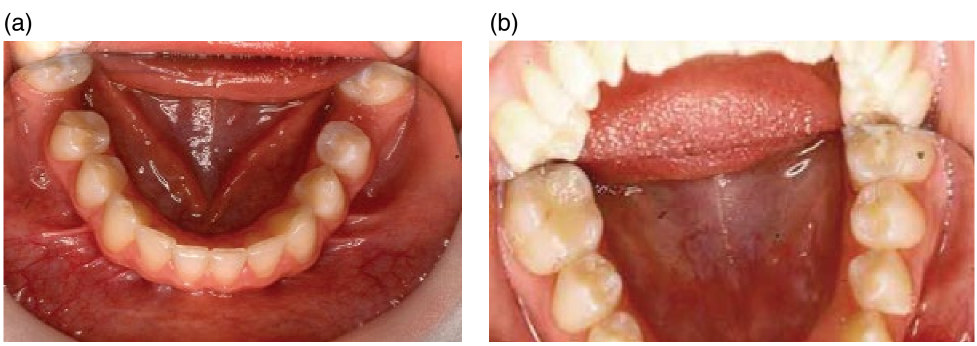

Figure 1.11 Floor of the mouth: (a) sublingual papilla in rest, (b) sublingual papilla in tension.

Figure 1.12 Relatively normal gingivae (note absent central incisors).

Figure 1.13 Alveolar bone covering both dentitions in the skull of a child.

The PL:

a) Holds the tooth to the alveolar bone and prevents damage to the tooth.
b) Is involved in the maintenance and repair of the alveolar bone and tooth cementum.
c) Is actively involved in neurogenic mastication control via its mechanoreceptors.

The connective tissue fibers of the PL are mostly type 1 collagen; however, a small portion of fibers are oxytalan and reticulin, and elastin has been found in some parts. Fibroblasts are the most frequent cells found in the connective tissues of the PL. They cover the surface of the cementum and alveolar bone, and are considered to be part of the periodontal ligament. They include: cementoblasts, cementoclasts, osteoblasts, and osteoclasts. In addition, there are also undifferentiated mesenchymal cells, defense cells, and remnants of the epithelial cells of Malassez.

1.7 The Alveolar Bone

The bony part protecting the teeth is called the "alveolar bone" of the maxilla and mandible. The alveolar bone is dependent on the presence of teeth to remain. In cases of congenital absence of all teeth (anodontia), the alveolar bone is negligible or absent. After individual tooth extraction, the alveolar bone that used to encase the root of the tooth will atrophy. Bone is a mineralized connective tissue with almost 60% mineral content, together with 25% organic materials and 15% water. By volume, bone is 36% mineral, 36% organic, and 28% water. The mineral phase consists of hydroxyapatite, and 90% of the organic part is type I collagen. In addition, it has a small amount of other proteins, such as osteocalcin, osteonectin, osteopontin, and proteoglycan.

1.8 The Teeth: Dental Anatomy

Human dental systems consist of two dentitions: a *primary* (deciduous, milk) dentition and a *permanent* (secondary) dentition. The eruption process of the primary dentition starts at and around 6 months of age, and is usually complete by 24–30 months. The eruption process for the permanent dentition starts at 6 years of age (±6 months), and continues into the late teens until the third molar (if present) erupts. The number of primary teeth is 20 (10 in each jaw), and the number of permanent teeth is 32 (16 in each jaw) (Figures 1.14 and 1.15).

The correct terminology of the parts of the tooth is essential:

a) *Crown*: The *clinical crown* is the part of the tooth that is visible on oral examination. The *anatomical crown*, on the other hand, is the part covered with enamel (Figure 1.16).
b) *Root*: The *clinical root* is the part of the tooth covered by alveolar bone. The *anatomical root* is the part covered with cementum. The *furcation area* (bifurcation or trifurcation) is the area of the root in multi-rooted teeth where the roots start to develop away from the crown (Figure 1.16).

Figure 1.14 Normal primary dentition.

Figure 1.15 Normal permanent dentition.

Figure 1.16 A human maxillary molar with typical landmarks.

Figure 1.18 Pulp in a traumatically separated crown with the remaining radicular pulp tissue intact.

c) *Cervical area*: The part of the tooth where the root and crown join; also known as the *tooth neck* (Figure 1.16).
d) *Dental pulp*: The central space in the teeth occupied by blood vessels, lymph vessels, and connective tissue (Figures 1.17 and 1.18). Odontoblasts are located at the outer surface of the pulp adjacent to the dentine.
e) *Anatomic landmarks of the tooth crown* (Figures 1.19 and 1.20):
   Occlusal: the biting surface on posterior teeth (molars and premolars).
   Incisal: the cutting edge on the anterior teeth (incisors and canines).
   Cusp: eminences on the occlusal surface of the posterior teeth.
   Tubercle: small projection on some coronal tooth parts due to excess enamel formation at the developmental stage that could be considered as a deviation from normal structure and shape.
   Cingulum: a bulbous convexity close to the cervical part of the lingual surface of anterior teeth.
   Ridge: a linear eminence on the occlusal surface of the crown seen in three parts and shapes: marginal, oblique, and triangular. The first two are mainly seen

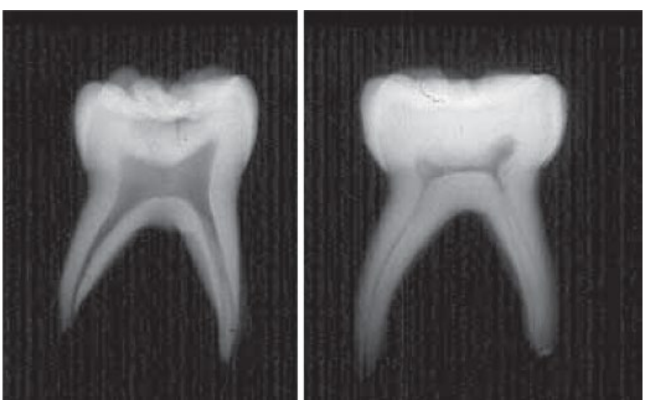

Figure 1.17 Radiographic view of primary (left) and permanent (right) molar pulp outlines.


Figure 1.19 Maxillary primary and permanent molars with cusps and fissures.


Figure 1.20 Maxillary central and lateral incisors.

on maxillary molars, while the latter is seen in both maxillary and mandibular molars and premolars.

Fissures: the groove between cusps and ridges.

Fossa: irregular intrusions and concavities on the tooth surface, examples of which include the lingual fossa on the lingual surface of incisors and the central fossa on the occlusal surface of molars.

Pit: pinpoint inversions of the surface enamel at the junction of grooves or at their ends. An example would be the occlusal pit on the occlusal central fossa of molars where the fissures meet.

Buccal: the crown surface touching /adjacent to the cheek (posterior teeth).

Labial: the crown surface touching/adjacent to the lips (anterior teeth).

Palatal: crown surface adjacent to the palate (maxillary teeth).

Lingual: the crown surface touching/adjacent to the tongue (mandibular teeth).

Mesial: the crown surface facing the midline.

Distal: the crown surface facing away from the midline.

1.9 Normal Occlusion

The skeletal jaw relationship dictates the inter-cuspal position of the teeth (occlusion). *Occlusion* can be defined as the relationship of the teeth in both jaws when they are in contact. A normal occlusion (Class I) is the commonest occlusion in the population:

a) The maximum number of teeth are in contact, and the mastication forces are at the physiological limit and along the long axis of the crowns.
b) Lateral movements of the jaws are carried out without interferences.
c) The space between teeth at rest is known as the *freeway space*.
d) Teeth alignment is acceptable esthetically.

1.10 Classification of the Occlusion

Normal occlusion: All teeth are in appropriate occlusion, with correct molar and incisor relationships (Figure 1.21a and b).

a) *Class I malocclusion*: One or more teeth are not in the normal position (malposition), while the molar relationship is normal (Figure 1.22a and b)
b) *Class II malocclusion*: The maxillary first molar is located mesially by a minimum of half a cusp from its normal Class I position. There are two sub-groups of Class II: Division I (maxillary anterior teeth are proclined, as shown in Figure 1.23) and Division II (maxillary anterior teeth are retroclined, as shown in Figure 1.24). In certain cases, only the centrals are retroclined, and laterals are proclined.
c) *Class III malocclusion*: The maxillary first molar is at least a half cusp more distal than a Class I. The incisor relationship changes from a normal maxillary over-jet to a reverse over-jet (Figure 1.25a and b).

(a) (b)

Figure 1.21 Normal occlusion, bite relationship: (a) over-jet, (b) over-bite.

(a) (b)

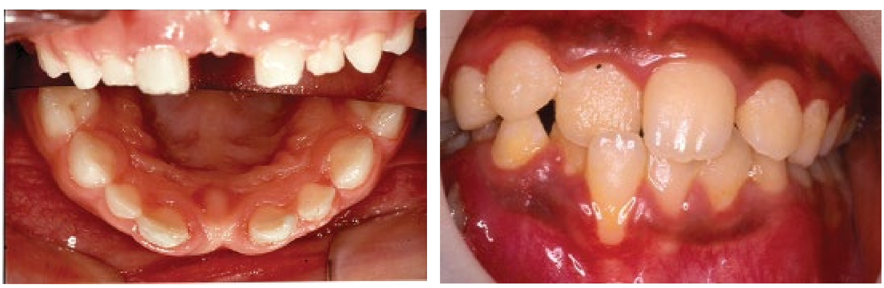

Figure 1.22 Class I malocclusion: (a) wide diastema, (b) severe crowding and cross-bite.

Figure 1.23 Class II Division I: anterior open bite.

(a) (b)

Figure 1.24 Class II Division II: (a) deep over-bite, (b) increased anterior over-jet and over-bite (occlusal view).

(a) (b)

Figure 1.25 Class III malocclusion: (a) reverse over-jet, (b) edge-to-edge and posterior cross-bite.

2

Histology and Embryology of the Teeth and Periodontium

2.1 Tooth Histology

2.1.1 Enamel

Enamel is the strongest structure in both humans and animals, and consists of minerals (96%) and water (4%). Microscopic views reveal that it consists of "enamel rods" that are formed of bundles of hydroxyapatite crystals. The area of the crystals adjacent to other crystals is known as the "rod sheath." In longitudinal sections of enamel, the crystals closer to the enamel surface are more divergent from the long axis of the rod, and get close to 90°. The external border of the two orientations appears in the shape of a key hole.

2.1.1.1 Striae of Retzius

These are a series of lines representing the stages of growth in cross-sectional dimensions, and appearing as dark bundles. These lines are representative of the intermittent production of enamel while the dentine is being formed. They are more common in permanent teeth than in primary, and are least common in natal and neonatal teeth. Incremental lines also appear as the result of disturbances in the development cycle – for example, owing to fever, which directly affects amelogenesis.

2.1.1.2 Hunter-Schreger Bands

These are a lighting phenomenon formed by changes in the direction of enamel rods. These bands are best seen following light reflection on longitudinal dried sections.

2.1.1.3 Gnarled Enamel (Spiral Enamel)

These consist of wavy rods and are mostly seen at the cusp areas.

2.1.1.4 Enamel Tufts and Lamella

These originate from the dentine enamel junction and extend a short way into the enamel. These highly mineralized structures are divided into branches at their enamel ends, and they contain higher protein levels as compared to other parts of the enamel. Lamellae and the proteins between the tufts are formed from the tufts.

2.1.1.5 Enamel Surface

Striae of Retzius lines are continued to the external surface, ending in deep surface fissures known as "perikymata." The surface of an unerupted tooth is covered with layers of 0.5–1.5 μm cuticles without a specific structure. Small, poorly bonded crystalline pieces are formed immediately beneath the cuticles. The surface layer and cuticles will vanish due to attrition and abrasion as soon as the tooth erupts. Figure 2.1 a and b shows the ground and demineralized sections of a sound maxillary canine tooth as a representation of enamel, dentin, and pulpal space.

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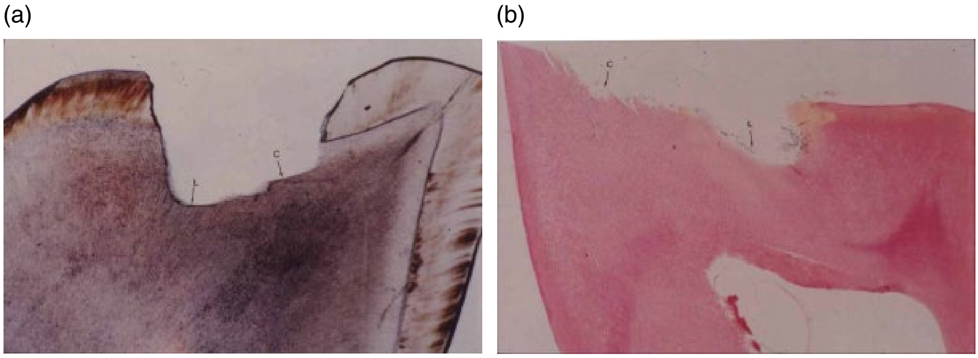

Figure 2.1 (a) Ground section of the crown of the maxillary canine enamel; (b) demineralized section of the same tooth, where almost all the enamel tissue has disappeared leaving the remaining collagenous structure.

2.1.2 Dentine

The bulk of the tooth structure is known as *dentine*. It consists of more water (10%) and collagen (20%) as compared to enamel, and the level of mineral drops to almost 70%. The orientation of the dentine components is different in coronal and radicular dentine (Figure 2.2a and b). Both areas contain tubular odontoblastic processes that extend from the pulp into the surrounding dentine structure providing sensitivity, thereby protecting against stimuli and hazards. The odontoblasts located at the pulpo-dentinal junction are the source of dentine secretion and production.

2.1.2.1 Dentinal Tubules

Dentinal tubules are small spaces within the dentine structure filled with interstitial fluid and the odontoblastic process. They are mainly present in the coronal segment of dentine, and follow an "S" shape, extending from the outer surface of dentine (*dentin–enamel junction*, or DEJ) to the periphery of the pulp (*pulp dentine junction*, or PDJ).

Various proteins, including glycosaminoglycans, proteoglycan, and glycoproteins, are present within dentine, as well as mineral hydroxyapatite. Dentine has an elastic structure, and this is critical for the teeth as it allows some degree of flexibility, protecting enamel from easy fracturing. The dentine

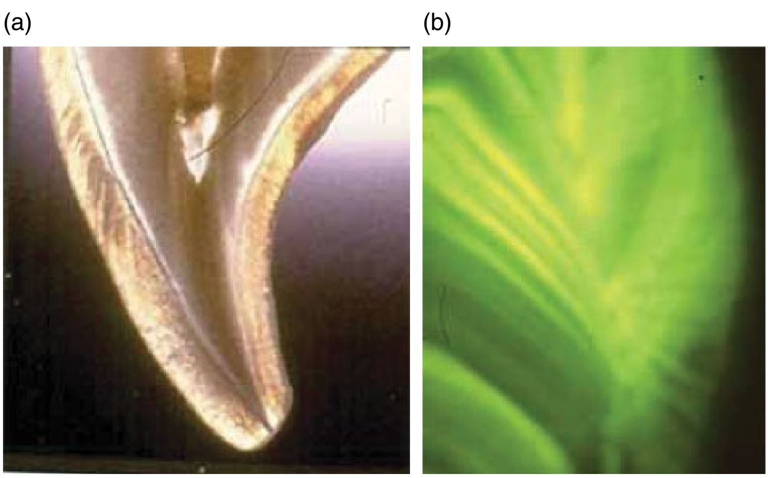

Figure 2.2 (a) Ground section showing dentin close to the enamel and pulp; (b) illustration of tubular dentin.

tubular structure is a unique characteristic feature of dentine, and, in addition to dentine tubules, there are three other elements within the tubular structure: intertubular dentine, intratubular dentine, and interglobular dentine.

2.1.2.2 Intratubular Dentine

The circular, hyper-mineralized dentine inside the tubules is called *intratubular dentine*. A ground section with a cutting degree of 90° illustrates these hyper-mineralized rings of dentine, which are formed within the tubules, causing a narrowing of the tubules. These rings are more easily detected when a cross-section includes parts of mineralized dentine in the butt joints of the tubules.

2.1.2.3 Intertubular Dentine

The part of dentine surrounding the tubules is called *intertubular dentine*. It contains type I collagen fibrils, which are in the form of an interwoven network providing the appropriate structure for apposition of the apatite crystals.

2.1.2.4 Interglobular Dentine

The term *interglobular dentine* refers to the areas of mature dentine in which mineralization is poor or absent. The tubular pattern remains intact and visible within the interglobular dentine.

2.1.2.5 Incremental Lines

Dentine formation follows a series of intervals of active and rest phases. These intervals leave developmental lines that cross the tubules and indicate the step-by-step formation of dentine.

2.1.2.6 Granular Layer of Tomes

This is a specific layer of dentine laying within the radicular dentine.

2.1.3 Cementum

The cementum is a covering structure for the root of the teeth that plays important roles in nourishing the tooth as well as in stabilizing the tooth via the attachment to the periodontal ligament. This enables the tooth to maintain its relationship to adjacent and opposing teeth. It is an essential part of the periodontium (Figure 2.3).

2.1.3.1 Cementum Connective Tissue

This is a hard, bonelike structure covering the entire root surface and receiving the periodontal ligament fibers. Cementum forms from an organic matrix that is made mainly of collagen and stem cells. Unlike bone, it does not contain blood vessels, and therefore cannot remodel. The majority of cementum is connective tissue, with differing amounts of cells, fibers, and ground substance. There are two types of cementum: cellular and acellular. Cementoblasts and cementocytes are the two types of cells found in cementum.

2.1.3.2 Fibrous Matrix

The fibrous matrix consists of two types of collagen fibers: internal and periodontal ligament. There are three variations of the cementoenamel junction (CEJ): (i) enamel is covered with cementum, (ii) edge-to-edge cementum/enamel, and (iii) a gap between cementum and enamel, with exposed dentine.

Figure 2.3 Cementum covering the root of a freshly extracted permanent incisor with abnormal coronal form.

2.1.4 Dental Pulp

The dental pulp is the central, highly vascular, and innervated segment of the tooth responsible for nutrient supply and sensation. The pulp tissue originates from the periapical region, and the blood supply reaches the tooth through the apical foramen. The pulp tissue extends to the pulp horns within the tooth crown in order to nourish the dentine and enamel cells.

2.1.5 Periodontium

The periodontium is a complex of cementum and bone with blood vessels, nerves, and bundles of fibers throughout the whole length of the socket, providing nutrition and sensibility, and allowing the tooth to remain within its socket during mastication and speech. The periodontium has the potential for regeneration and remodeling throughout life, which allows the primary dentition to be transient and to be replaced by the permanent dentition.

2.2 Embryology of Teeth: Life Cycle of the Tooth

The development of a tooth germ from embryonic cells is called the *life cycle* of the tooth. This cycle includes several steps that are very sensitive to threats that cause potential defects. Any damage to the tooth germ cells could interfere with the formation or calcification, or both. Developmental defects are described depending on the severity and stage of interruption of tooth development. The life cycle of the tooth includes the following stages:

2.2.1 Initiation (Bud) Stage

This is a stage at which the initial signs of the formation of a tooth bud are seen, with the resultant ameloblastic activity laying down the enamel substance in a designated area dictated by the genetic instruction of cells.

2.2.2 Proliferation (Cap) Stage

This is the stage reached at 9–11 weeks of gestation. The shape of the initial structure is determined by the genetic order of cell and organ formation. The initial shape looks like a cap or hat, and is known as the *cap stage*. The tooth germ in this stage is formed of three segments: (i) dental organ, (ii) dental papilla, and (iii) dental sac. The dental organ is responsible for the production of enamel. The dental papilla will produce dentine and pulp in the future. The dental sac is the origin of the cementum and periodontal ligament.

2.2.3 Histodifferentiation and Morphodifferentiation (Bell) Stage

This step happens during 14–18 weeks of gestation, when the cap continues its proliferation until it reaches a bell shape. Cells are aligned in a format and shape close to the final tooth shape. The inner epithelial cells are converted to ameloblasts, which in turn produce the enamel matrix. Parallel to ameloblast formation, the dental papilla forms adjacent to the basal membrane and starts to differentiate into odontoblasts. These newly formed structures start to differentiate into early enamel and dentine.

2.2.4 Apposition and Calcification

The apposition stage starts following the formation of the matrix of the tooth. This stage is responsible for enamel and dentine formation by ameloblasts and odontoblasts from a single center of growth, starting from the Dentino Enamel Junction (DEJ) and extending to the DEJ. Calcification occurs by the entry of organic salts into the mature tissue matrix. Calcification of enamel starts by the opposition of mature enamel at the cusp tips and incisal edges of the incisor teeth, and continues from these points toward the cervical margins. The older (mature) enamel is, therefore, located at the cusps, and the newly formed enamel is at the cervical region. Interference in any of these stages is potentially hazardous and could cause developmental defects of the tooth. Damage to the tooth germ in each stage will cause different clinical effects.

3

Epidemiology and Diagnosis of Teeth Developmental Disturbances

3.1 Prevalence and Incidence

Various defects (anomalies/malformations) can develop in the tooth structures depending on the causative agent and its relationship to the stage of the tooth formation and calcification. The incidence of defects differs based on race, geographic region, and sex, and these variations have been reported on different tooth parts as well as tooth supporting structures. In one study, enamel defects were reported in 33% of the population (Masumo et al. 2013).

The frequency of defects is common, so it is important for the dentist to have the knowledge to inform the parents appropriately. Accordingly, this topic should be adequately covered in the dental undergraduate teaching curriculum.

3.2 Diagnosis and Classification of Defects in Teeth

The classification of defects in teeth varies and is based on the different structures involved. When each part of the tooth is affected, it is classified as a "structural defect." This may affect the quantity of the tooth substance produced, causing shape and size variation, and, in certain circumstances, the absence of teeth or the formation of extra teeth. Tooth color is also affected in certain defects, causing abnormal appearance. Apart from these variations in classification, the causative source of the defect may also be considered as a tool for the classification of dental defects. The following sections discuss the most common classifications used by dental professionals and scientists.

3.2.1 Cause of Disturbance

a) *Genetic*: Tooth shape and structural content can be influenced by genome dictation. There may be a wide range of disturbances as part of a much larger clinical manifestation, such as a syndrome involving several body organs as well as the teeth, including those in ectodermal dysplasia; or, alternatively, only teeth may be involved, such as the case in amelogenesis imperfecta.
b) *Congenital*: There are instances when a child's tooth development is affected by events during pregnancy or at birth. This is known as *congenital*, and there is no faulty gene involved, an example of which is enamel hypoplasia caused by maternal dehydration or viral infection during pregnancy.
c) *Acquired*: These cases are affected by environmental factors – for example, fluorosis defects caused by water fluoride content or isolated enamel, and dentine hypoplasias in permanent teeth caused by infections involving primary antecedent teeth, such as in Turner's hypoplasia.

Atlas of Pediatric Oral and Dental Developmental Anomalies, First Edition. Ghassem Ansari, Mojtaba Vahid Golpayegani, and Richard Welbury.

Companion website: www.wiley.com/go/ansari/pediatric_oral_dental_anomalies

3.2.2 Extent of Involvement of the Dentition

Defects can either involve part or all of the teeth. They may also affect one tooth, a few teeth, or the full dentition. It is important to classify them on the level of involvement of the teeth:

a) Generalized
b) Localized

3.2.3 The Structure Involved

3.2.3.1 Enamel Defects

a) *Enamel hypoplasia*: This condition accounts for 15.8% of all enamel defects – for example, molar incisor hypoplasia (MIH) (Gurrusquieta et al. 2017). Localized cases are more frequent than those with more generalized involvement.
b) *Amelogenesis imperfecta*: In this condition, defects occur on parts or all of the enamel of teeth. Several types of this condition have been described. Morphologically, four clinical types are recognized:
   1) Hypoplastic
   2) Hypomature
   3) Hypocalcified
   4) Combined

The *hypocalcified* type is the most frequent, and the *combined* type is the least frequent.

3.2.3.2 Dentine Defects

a) *Dentine dysplasia*: This is a rare familial condition reported in different parts of the world. Dentine formation is affected in such a way that the volume of dentine production is reduced, resulting in shortening of the roots. It has two types:
   1) Radicular
   2) Coronal

b) *Dentinogenesis imperfecta*: In this case, the dentine structure has abnormal collagen content and is weaker than normal dentine. The condition was originally mainly seen and reported in America, India, and Africa. Three types are recognized:
   1) *Type I*, associated with osteogenesis imperfecta
   2) *Type II*, affecting only the teeth
   3) *Type III*, Brandywine isolate in Maryland (USA), also called "shell" teeth

Type II is the most frequent type among all three types.

3.2.3.3 Cementum Defects

These are rare, with each type having different racial incidences. An increase or decrease in cementum substrate influences its role as part of the periodontium. Excess cementum production is described more frequently, since reduced or absent cementum leads to early tooth loss. There are three types:

a) *Hypercementosis*: Cementum secretion increases, causing accumulation of large areas of cementum around the root apex, which appears radiographically as "clubbing" of the apical area.
b) *Hypocementosis*: In hypocementosis, the amount of cementum formed is reduced, in contrast to hypercementosis. A thin layer of cementum results in failure of the periodontal ligaments to attach properly, resulting in loosening of the teeth and early tooth loss.
c) *Acementosis*: This is a very rare condition in which the roots of teeth have no cementum cover, leading to early tooth loss. Areas of acementosis can occur in hypophosphatasia.

3.2.3.4 Entire Tooth Structures Involved

a) *Aplasia (Anodontia)*: No teeth develop in either primary or permanent dentitions. This could also happen in one or a few teeth, where the term "congenital missing" is used to describe the condition.
b) *Regional odontodysplasia/odontogenesis imperfecta*: This is a developmental disturbance involving one or more neighboring teeth. Deficiency extends from

enamel and dentine to the pulp, making an abnormally large space provided for the pulp chamber to fill. It has a unique radiographic appearance, giving it the name "ghost tooth," as a ghostlike radiographic appearance is detected. Delayed eruption of affected teeth is a common finding associated with the condition.

c) *Segmental odontodysplasia*: There are cases where expansion of the condition has reached the entire quadrant, but it is more commonly seen in the maxillary alveolus. It is interestingly reported to occur only on one side of the jaw, and is associated with spacing between the involved teeth. The permanent premolars are usually absent on the affected side, and there is a lack of pneumatization of the maxillary antrum in the region.

3.2.4 Teeth Morphology

The incidences and prevalence of malformations differ between races and ethnic groups. Many malformed teeth are associated with syndromes. Malformations can be classified as:

a) *Invagination (dens in dente)*: In-growth of enamel, with or without dentine, from the occlusal surface toward the pulp space.
b) *Evagination (talon cusp)*: Outgrowth of enamel, with or without dentine, ranging from a small eminence to a full cusp.
c) *Gemination*: When the tooth crown has a cleft, giving the appearance of an extra crown. It may start with a small groove, but could reach as deep as the pulp at the cervical part of the crown. In extreme cases, the tooth bud may be divided into two pieces, one of which becomes a supernumerary tooth.
d) *Fusion*: When two adjacent teeth are fused together, making the two look like one single large tooth. In true fusion, the roots remain separated.
e) *Peg lateral*: The rounded and more conical formation of a lateral incisor.
f) *Hutchinson incisors*: Notched appearance of permanent central incisors associated with congenital syphilis.
g) *Mulberry molar*: Formation of multiple cusps instead of, or in addition to, each normal cusp on molar teeth.
h) *Supplemental tooth*: An extra tooth of similar size and morphology as a normal tooth
i) *Odontome*: Any mass of malpositioned and deformed tooth tissue occurring in the alveolus in the position of normal teeth. Based on their composition, odontomes can be classified into (a) compound, and (b) complex.

3.2.5 Teeth Size

The "normal" size of teeth has been standardized, and a certain range is accepted among populations. When proportions are reduced or increased as compared to "normal," the condition can be classified as follows:

a) *Macrodontia*: When tooth size is larger than the normal range
b) *Microdontia*: When tooth size is smaller than the normal range
c) *Short roots*: When root growth does not reach the normal limit

3.2.6 Teeth Count

Hyperdontia and hypodontia are reported frequently among many races and populations, especially when associated with syndromes.

3.2.6.1 Hypodontia

a) Hypodont: one to four missing teeth
b) Oligodont: more than four missing teeth
c) Anodont: total missing teeth

3.2.6.2 Hyperdontia

a) Mesiodens: Maxillary Midline supernumerary
b) Paramolar: supernumerary of molar region
c) Multiple extra teeth

3.2.7 Color of the Teeth

Changes in color of the teeth have many presentations, depending on their origin and causative factors – for example, high fluoride intake, tetracycline use, liver disease, etc. Since the condition was first reported in children with cystic fibrosis, tetracycline discoloration has now significantly reduced, thanks to educational efforts. The frequency of fluorosis ranges between 20 and 40%, and tetracycline was at approximately 23% during the 1970s when it was widely prescribed, with very few cases and their managements being reported in recent years (Stewart 1973; Koleoso et al. 2004; Kuzekanani and Walsh 2009; Wiener et al. 2018). Discoloration can be classified based on the color changes in the normal tooth color. These may include: chalky white, snowcap white, gray, black, brown, blue, yellow, and red. Discoloration can also be classified based on the cause, as follows:

a) Food and diet
b) Vitamins and minerals
c) Excess ions of fluoride
d) Systemic disease
e) Cystic fibrosis
f) High fever
g) Jaundice
h) Dehydration
i) Medications
j) Trauma and teeth infection
k) Congenital enamel and dentine defects