

# [Developmental defects of teeth essay](https://assignbuster.com/developmental-defects-of-teeth-essay/)

developmental defects of teeth with recent genetic concepts INTRODUCTION: Disorders of development of teeth may be prenatal or postnatal in origin and may be inherited or acquired. Their recognition and evaluation requires a thorough knowledge and evaluation of the normal chronology of the human dentition and of the normal development and structure of the teeth.

Disorders of development of teeth of teeth may be due to abnormalities in the differentiation of the dental lamina and the tooth germs, causing anomalies in the number, size and form of teeth (abnormalities of morphodifferentiation) or to abnormalities in the formation of the dental hard tissues resulting in disturbances in tooth structure (abnormalities of histodifferentiation). Abnormalities of histodifferentiation occur at a later stage in development than abnormalities of morphodifferentiation; in some disorders both stages are abnormal. Histophysiology of tooth development:

A number of physiologic growth processes participate in the progressive development of the teeth. These are – 1. Initiation The dental lamina and associated tooth buds represent those parts of the oral epithelium that have the potential for tooth development. Different teeth are initiated at different times. Initiation induction requires ectomesenchymal epithelial interaction. A lack of initiation results in the absence of either a single or multiple teeth or complete lack of teeth. Abnormal initiation may result in the development of single or multiple supernumerary teeth. 2. Proliferation

Enhanced proliferative activity after initiation results successively in the bud, cap and bell stages of the odontogenic organ. Proliferative growth causes regular changes in the size and proportions of the growing tooth germ. A disturbance has entirely different effects according to the time of occurrence and the stage of development that it affects. Tooth anomalies may include disturbances in the size, proportion or number of teeth 3. Histodifferentiation It succeeds the proliferative stage. The formative cells of the tooth germ developing during the proliferative stage undergo definite morphologic as well as functional assignment.

Differentiation of odontoblasts and ameloblasts takes place resulting the formation and apposition of enamel and dentin. Disturbance is the stage results in defects in the structure of tooth like amelogensis imperfecta type 1 and 4. 4. Morphodifferentiation The morphologic pattern or basic form and relative size of the future tooth are established by morphodifferentiation that is by differential growth. Dentinoenamel junction and dentinocemental junctions are established. Disturbances in morphodifferentiation may affect the form and size of the tooth without impairing the function of the ameloblasts or odontoblasts. 5. Appposition

It is the deposition of the matrix of the hard dental structures. Defects in apposition results in anomalies like amelogenesis imperfecta type 2, dentin dysplasia. 6. Mineralization and Maturation After matrix formation full mineralization and dental hard tissue maturation. Anomalies like amelogensis imperfecta type 3, fluorosis and interglobular dentin. CLASSIFICATION OF DEVELOPMENTAL DEFECTS OF TEETH 1. Anomalies of initiation and proliferation – anomalies of number hypodontia and anodontia hyperdontia – anomalies of size Microdontia Macrodontia – anomalies of shape Germination Fusion Concrescence 2. Anomalies of morphodifferentiation anomalies of size and shape Dilacerations Dens invaginatus Dens evaginatus Taurodontism Talon cusp 3. Anomalies of histodifferentiation – enamel defect Amelogensis imperfecta type I – dentin defect Dentinogensis impefecta type I, II, III 4. Anomalies of apposition – enamel defects Amelogensis imperfecta type II, IV Enamel hypoplasia Enamel pearl – dentin defects Dentin dysplasia Regional odontodysplasia – cementum defects Hypercementosis Hypophophatasia 5. Anomalies of mineralization – enamel defects Amelogenesis imperfecta type III Enamel fluorosis Anomalies of initiation and proliferation Development disturbances in number a. ypodontia and anodontia True anodontia or congential absence of teeth may be of two types Total anodontia – in which all teeth are missing. It may involve both deciduous and permanent teeth. It is a rare condition when it occurs it is frequently associated with generalized disturbances, hereditary ectodermal dysplasia. Partial anodontia (hypodontia or oligodontia) Hypodontia when there is lack of development of one or more teeth Oligodontia is a subdivision of hypodontia indicating the lack of development of six or more teeth. An incidence of 1. 5 – 10% is seen excluding 3rd molars in permanent dentition and . 09 – . % for primary dentition. Most frequently affected teeth are 3rd molars, mandibular 2nd premolar, maxillary lateral incisor and maxillary 2nd premolar. In case of deciduous dentition usually maxillary lateral incisor are missing. According to Grahen and Granath there is a close correlation between congenitally missing deciduous teeth and their permanent successors suggesting a genetic factor. A familial tendency for this defect is noted. Graber reported enough evidence that it is actually the result of one or more point mutations most often transmitted in an autosomal dominant pattern with incomplete penetrance and variable expressivity.

Some investigators implied that hypodontia is a normal variant suggesting that humans are in an intermediate stage of dentitional evolution. A proposed future dentition would include one incisor, one canine, one premolar and two molars per quadrant. Hypodontia correlates with the absence of appropriate dental lamina. Mostly this is due to genetic alteration. However, environmental factors like trauma, infection, radiation and endocrine disturbance may also result in loss of developing tooth bud. Hypodontia is associated positively with microdontia, reduced alveolar development, increase free way space and retained primary teeth.

Pax 9 gene maps to chromosome 14, it encodes a transcription function that is important in the development of pharyngeal pouches. Its mutation may lead to congenitally missing teeth. Syndromes associated with hypodontia: – cleft lip/palate – crouzon syndrome – down syndrome – hyphidrotic ectodermal dysplasia – ellis van crevald syndrome – oro facial digital syndrome b. hyperdontia it is the development of an increase number of teeth. Genetics of hyperdontia suggests an autosomal dominant pattern of inheritance with incomplete penetrance. The variable expression and penetrance of the gene may be affected by the environmental factors.

The development of hyperdontia is development of excess dental lamina which leads to formation of additional teeth. An incidence of . 3-3% and a male to female ratio of 2: 1 is noted. It occurs most frequently in permanent dentition and approximately 90% of cases present in maxilla with a strong predilection for the anterior region. Terms used to describe supernumerary tooth depending on their location i. Mesiodens is located in the maxillary incisor resion. It is the most common supernumerary. ii. Distodens/ distomloar is the accessory fourth molar iii.

Paramolar is a posterior supernumerary tooth situated lingually or buccally to a molar tooth. Classification based on morphology In the primary dentition morphology is usually normal or conical. Greater variety is seen in case of permanent dentition Conical is a small peg shaped conical tooth and is the most common supernumerary. It develops with root formation ahead or at an equivalent stage to that of permanent incisor. It can result in rotation or displacement of the permanent incisor but rarely delays eruption. Tuberculate possess more than one cusp or tubercle and is barrel shaped.

Root formation is delayed as compared to permanent incisors. They are often paired and are commonly located on the palatal aspect of the central incisors. It results in delayed eruption of the incisors. Supplemental refers to a duplication of teeth on the normal series and is usually found at the end of the tooth series. Most commonly permanent maxillary lateral incisor is the supplemental supernumerary. Majority of the deciduous supernumeraries are of this type. Odontome is listed as the fourth category by Howard. It is a hamartomoatous malformation nd is composed of more than one type of tissue called composite odontoma. Mainly two types are there complex and compound. Syndromes associated with hyperdontia 1. cleft lip/palate 2. cleidocranial dysplasia 3. gardner syndrome 4. oro facial digital syndrome Developmental disturbances in size of tooth a. microdontia – teeth which are smaller than normal. Females demonstrates a higher a frequency of microdontia and hypodontia. Three types are – True generalized microdontia in this all the teeth are smaller than normal. It is seen in cases of piutatry dwarfism, Down syndrome, congenital heart diseases. Relative generalized microdontia normal slightly smaller teeth present in the jaws larger than normal producing an illusion of microdontia. Role of hereditary as jaws are inherited from one aren’t and tooth size from other parent. – Microdontia involving single tooth is common condition affects maxillary lateral incisors and 3rd molar most commonly. b. macrodontia – teeth that are larger than normal. Three types similar to that microdontia – True generalized macrodontia all teeth are large. Seen in cases of pituitary gigantism and pineal hyperplasia with hyperinulinism. Relative generalized macrodontia presence of normal teeth in similar jaw giving an illusion of macrodontia – Macrodontia of single tooth relatively uncommon. Occasionally seen in cases of hemihypertrophy of the face. Should not be confused with fusion of teeth. Development disturbances of shape of teeth a. germination – anomalies which arise from an attempt at division of single tooth germ by an invagination with resultant incomplete or complete formation of teeth. Incidence of . 5% is seen and it more common in primary dentition. Higher frequency in maxillary anterior region.

It may retard eruption of permanent successor. Usually appear as one with two completely or incompletely separated crowns that have a single root and root canals. Twining production of equivalent structure by divisions resulting in one normal and one supernumerary tooth. Etiology is unknown although a genetic basis has been suggested. b. fusion – union of two normally separated tooth germs. Depending upon the stage of development of the teeth at the time of the union. More common in the deciduous than in the permanent dentition. Union by dentin, separate pulp chambers/ canals. May retard eruption of permanent successor. . concrescence – form of fusion which occurs after root formation. Teeth are united by cementum only. As a result of traumatic injury or crownding of teeth with resorption of the interdental bone. More frequently in the posterior and maxillary region. Anomalies of morphodiferentiation Anomalies of size and shape a. dilaceration: An angulation or a sharp bend or curve in the root or crown of a formed tooth. Due to trauma during the period in which the tooth is forming, with the result that the position of the calcified portion of the tooth is changed and the remainder of the tooth is formed at an angle.

Syndrome associated with dilaceration. Lamellar ichthyosis. Present difficulty at the time of extraction. b. dens invaginatus: dens in dente It is a deep surface invagination of the crown or root that is lined enamel. Results due to invagination in the surface of tooth crown before calcification has occurred. This can be due to increased localized external pressure focal growth retardation and focal growth stimulation in certain areas of the tooth buds. The permanent maxillary lateral incisors are the teeth most frequently involved. Two forms are usually seen as Coronal dens in dente – more frequent prevalence varies from . 4% to 10%. Depth of iinvagination varies from a slight enlargement of the cingulum pit to deep infolding that extends to the apex. Radicular dens in dente – rare and thought to arise secondary to proliferation of hertwig’s root sheath with the formation of a strip of enamel that extends along the surface of the root. Radiographiclaly the affected tooth demonstrates an enlargement of the tooth. It chances of caries and pulp infection. c. dens evaginatus – Leong’s premolar it is a developmental condition that appears clinically as an accessory cusp or a globule of enamel on the occlusal surface between the buccal and lingual cusps.

Unilaterally or bilaterally with an increased mandibular predominance. Pathogenesis is the proliferation and evagination of an area of the inner enamel epithelium and subjacent odontogenic mesenchyme in to the dental organ during early tooth development. If seen in association with another varaiton of coronal anatomy. Shovel shaped incisors affected incisors demonstrate prominent lateral margins, creating a hollowed lingual surface. Maxillary incisor is the most commonly affected. This extra cusp may contribute to incomplete eruption, displacement of teeth and/or pulp exposure with subsequent infection following occlusal wear or fracture. . taurodontism – it is an enlargement of the body and pulp chamber of multirooted tooth with apical displacement of the pulpal floor and bifurcation of the roots. Overall shape of the taurodont resembles that of the molar teeth of cud chewing animals “ bull like” Shaw classified taurodont teeth into 3 division according to the degree of apical displacement of pulpal floor – hypotaurodont (mild) – mesotaurodont (moderate) – hypertaurodont (severe) may occur as an isolated trait or as a component of specific conditions like 1. klinfilter’s syndrome . amelogenesis imperfecta 3. orofacial digital syndrome 4. hypohydrotic ectodermal dysplasia 5. tricho dento osseous syndrome 6. down syndrome It is seen that taurodontism may develop in presence of any one of a large number of different genetic alterations. These findings suggest that chromosomal abnormalities may disrupt the development of the tooth form and that taurodontism is not the result of any specific genetic abnormality e. talon’s cusp – Is also known as dens evginatus of the anterior teeth. It is a well delineated cusp that is located on the surface of an anterior tooth.

Predominantly affects permanent dentition esp maxillary lateral incisors. It projects from the lingual surface of the affected tooth and forms a three pronged pattern that resembles an eagle’s talon. The cusp blends smoothly with the tooth except that there is a deep development groove where the cusp blends with the sloping lingual tooth surface. It is associated with other dental abnormalities like supernumearary teeth, odontomas, impacted teeth; peg shaped lateral incisors and dens invaginatus. It is also associated with other syndromes like Rubinstein Tyabi syndrome & Sturge Weber syndrome.

Complications like interference with occlusion, comprised esthetics, displacement of teeth, caries, periodontal problems and irritation of the adjacent soft tissue may occur. ANOMALIES OF HISTODIFFERENTIATION Enamel defects a. Amelogenesis imperfecta type I AI is a heritable enamel defect in the absence of any systemic disease. Reported incidence varies from 1: 14000 to 1: 4000. Multiple inheritance patterns are noted. It may differentiated into 4 main groups depending on the clinical presentation and the likely stage of enamel formation that is primarily affectes. – hypoplatic hypocalcified – hypomature – hypomature hypoplastic with taurodontism Each group is further subdivided into several subgroups depending on the mode of inheritance as well as the clinical appearance of the defective enamel. Genetic studies – have shown that etiology of AI is related to the alteration of genes involved in the process of formation and maturation of the enamel. X linked AI has shown the defective gene is closely linked to the locus DXS85 at Xp22. this site has been identified as the general location of the human gene amelogenin the principal protein in the developing enamel.

In the case of autosomal dominant type of AI the locus of the defective gene is on the ch 4q21 to which enamel protein enamelin maps. AI type I Hypoplatic The basic alteration centers an inadequate deposition of enamel matrix. Any matrix present is mineralized appropriately. Subgroups (Witkop 1989) I A – hypoplastic, pitted AD I B – hypoplastic, local AD I C – hypoplastic, local AR I D – hypoplastic, smooth AD I E – hypoplastic, smooth XR I F – hypoplastic, rough Ad I G – enamel agenesis, AR Clinical feature: – In the generalized pattern pinpoint to pinhead sized pits are scattered across the surface of the teeth. Buccal surfaces of the teeth are affected and staining of these pits may occur – In the localized pattern, the affected demonstrate horizontal rows of pits a linear depression. The altered area is located in the middle third of the buccal surface of teeth. – The type IC is more severe and typically demonstrate involvement of all teeth in both dentitions – In type ID the enamel of all teeth exhibits a smooth surface and is thin hard and glossy – Anterior open bite is seen and teeth exhibit a thin peripheral outline of radiopaque enamel on radiograph – In type IE this is a lesion showing lionization effect. In females the teeth would exhibit alternating zones of normal and abnormal enamel. Males exhibit diffuse thin, smooth and shiny enamel in both dentitions. The teeth have the shape of crown formation with open contacts. – In the rough pattern, the enamel is thin, hard and rough surfaces. An anterior open bite is common. – In enamel agenesis demonstrate total lack of enamel. The teeth are the shape and color of the dentin. The surface is rough. Dentin defects b. Dentinogenesis imprfecta A hereditary developmental disturbance of the denitn in the absence of any systemic disorder.

This is an autosomal dominant condition affecting both deciduous and permanent teeth. Affected teeth are gray to yellowish brown and are tulip shaped. Genetic studies show that the gene defective maps to chromosome 4 in case of type II. It encodes a protein called dentin sialophosphoprotein and constitutes about 50% of the noncolllagenous component of dentin matrix. DI type I is due to mutational changes in gene coding pro alpha 1, pro alpha 2 chains of type I collagen, major protein of organic matrix in bone and dentin.

Recent studies report that type II and type III arise from a single mutation of DSPP gene and that these are not separate disease but rather phenotypic variation of a single disease. Acc. to Sheild’s classification DI type I – osteogenesis imperfecta with opalescent teeth DI type II – isolated opalescent teeth DI type III – isolated opalescent teeth Brandywine Extensive studies have proven that DI is a disorder distinct from Osteogenesis imperfects so a revised classification is proposed DI type I corresponds to type II of Sheild’s Type II corresponds to type III of Sheild’s

There is no substitute for type I of Sheild’s classification Clinical and radiographic features – deciduous teeth are affected most severely followed by the permanent incisors and first molars – enamel is normal but separates easily from dentin because of defective dentinoenamel junction – once exposed the dentin often demonstrates significantly accelerated attrition – radiographically, the teeth have bulbous crowns cervical constriction thin roots and early obliteration of the root canals and pulp chambers. Type III has shell teeth demonstrating normal thickness enamel in association with extremely thin dentin and dramatically enlarged pulps Histologic features – The dentin adjacent to the enamel junction appears similar to normal dentin but the remainder is distinctly abnormal – Short tubules course through an atypical granular dentin matrix which often demonstrates interglobular calcificaiton. – Cells can be seen entrapped within the defective dentin Anomalies of apposition Enamel defects a. amelogenesis imperfecta type II & IV

AI type II – hypomaturation the enamel matrix is laid down appropriately and begins to mineralize however there is a defect in the maturation of the enamel crystal structure Classified into Type IIA – hypomaturation, pigmented AR Type IIB – hypomaturation, X linked recessive Type IIC – snow capped teeth, AD Genetic studies show that multiple mutations in the AMELX gene in the type IIA AI whereas IIA & C the gene defect is unknown. Clinical features – Affected teeth are normal in shape but exhibit a mottled, opaque white brown yellow discoloration.

The enamel is softer than normal and tends to chip from the underlying dentin. Radiographically the affected enamel exhibits a radiodensity that is similar to that of dentin. – Type IIA the surface enamel is mottles and agar brown. Enamel is soft enough to be punctures by an explorer – Type IIB shows lionization that is male exhibit differebti pattern in deciduous and permanent teeth. The deciduous teeth are opaque white with a translucent mottling; the permanent teeth are opaque and may darken with age.

Female patients exhibit a similar pattern in both dentitions. The teeth demonstrate vertical bands of white opaque enamel and normal translucent enamel. Radiographically the bands are not perceptible. – Type IIC exhibits a zone of white opaque enamel on the incisal or occlusal third of the crown. AI type IV – hypomaturation hypoplastic with taurodontism This type of Ai exhibit enamel hypoplasia in combination with hypomaturation. Classified into Type IV A hypomaturation hypoplastic Type IVB hypoplastic hypomaturation Genetic defects in both groups is unknown

Type IV A the predominant defect is one of enamel hypomaturation in ehich the enamel appears mottled yellowish white to yellow brown. Pits are seen frequently on the buccal surface of the teeth. Radiographically the enamel is similar to dentin in density and large pulp chambers may be seen in addition to varying degree of taurodontism Type IVB the predominant defect is one of enamel hypoplasia in which the enamel is thin; the enamel that is present demonstrates hypomaturation. Radiographically there is decrease in the thickness of the enamel.

Both these types are seen in the systemic disorders, trich dento osseous syndrome. Other features seen are kinky hair, osteosclerosis and brittle nails. b. enamel pearl Refers to the presence of enamel in unusual locations mainly the tooth root. These are hemispheric structures that may consist entirely of enamel or contain underlying dentin and pulp tissue. These are thought to arise from a localized bulging of the odontoblastic layer. This bulge may provide prolonged contact between Hertwig’s root sheath and the developing dentin, triggering induction of enamel formation. t is found most frequently on the roots of maxillary molars. The enamel pearls precludes normal PDL attachment with CT. Dentin defects c. dentin dysplasia It is a rare disturbance of dentin formation characterized by normal enamel but atypical dentin formation with abnormal pulpal pathology. It is an abnormality that maps to the same site on ch 4 as does DI type II & III. It too may be an allelic variant. Classified into Type I radicular dentin dysplasia Type II coronal dentin dysplasia Both these patterns appear to be hereditary diseases, transmitted as an autosomal dominant characteristic.

Clinical features – Type I both dentitions are affected although the teeth are clinically normal. The teeth are extremely mobile and exfoliated prematurely – Type II both dentitions are affected. Deciduous teeth have the same color as that in DI but the permanent dentition is normal. – Type I both dentitions roots are short blunt and conical. In deciduous pulp chambers and root canals are completely obliterated. Permanent teeth crescent shaped pulpal remanant remains. Periapical radiolucencies involving intact teeth are present. Type II deciduous dentition presents with completely obliterated pulp chambers. Permanent teeth exhibit an abnormal large pulp chamber in the coronal portion. Histologic features – Type I a portion of coronal dentin is normal. Most of the pulp is obliterated by calcified tubular dentin, osteodentin and fused denticles. New dentin forms around these obstacles giving the characteristic appearance described as Lava flowing around the boulders. – Type II deciduous teeth exhibit amorphous and atubular dentin in the radicular portion while coronal dentin is normal.

Permanent teeth same but the pulp has multiple pulp stones. d. regional odontodysplasia It is a localized non hereditary development abnormality of teeth with extensive adverse effects on the formation of enamel, dentin and pulp. Proposed causes – abnormal migration of neural crest cells – latent virus – local circulatory deficiency – local trauma or infection – hyperpyrexia – malnutrition – radiation therapy – somatic mutation pathosis associated with RO – ectodermal dysplasia – epidermal nevi – hypophosphatasia – hydrocephalus neurofibromatosis – Rh incompatibility – Vascular nevi Clinical features – maxillary teeth esp anterior are most commonly affected – female predilection with a ratio of 1. 4: 1 – the affected teeth fail to erupt and their shape is markedly altered – the erupted teeth show small irregular crown that is yellow to brown with rough surface – on x ray they have a very thin enamel and dentin surrounding an enlarged radiolucent pulp – There is lack of contrast between enamel and dentin.

Histologic features – marked reduction in the amount of dentin and widening of predentin layer – presence of large areas of interglobular dentin and an irregular pattern if dentin – the reduced enamel epithelium around nonerupted teeth show many irregular calcified bodies known as enameloid conglomerates (also seen in AI) e. hypercementosis It is a nonneoplastic deposition of excessive cementum that is continous with normal radicular cementum Clinical and radiographic features in x ray affected teeth demonstrate a thickening or blunting od the root – the enlarged root is surrounded by the radiolucent PDL space and intact dental lamina – may be isolated or involve many teeth – premolars are involved more frequently – predominantly in adults and the frequency increases with age – factors associated with hypercemntosis i. local 1. abnormal occlusal trauma 2. adjacent inflammation 3. unopposed teeth ii. systemic 1. acromegaly 2. arthritis 3. alcinosis 4. paget’s disease 5. rheumatic fever Histologic features – periphery of root exhibits deposition of an excessive amount of cementum – this may be hypocellular or ostecementum – the material is arranged in concentric layers Anomalies of mineralization a. amelogenesis imperfecta type III hypocalcifeid in this type the enamel matrix is laid down appropriately but no significant mineralization occurs. Classified into Type III A – autosomal dominant Type IIIB – autosomal recessive

In both gene defect is unknown. Most common type of AI. Teeth are norma on eruption but the enamel is very soft and easily lost. Enamel is yellow brown on eruption and turns to brown black due to staining. Autosomal recessive are more severe than the dominant type. Radiographically the density is similar for both enamel and dentin. CONCLUSION An understanding of the many disturbances of development of teeth is predicated upon s thorough understanding of the embryology of these structures.

Genetic factors are undoubtedly of importance in the development of these defects. REFERENCE: ? Oral pathology by Shafer ? Oral pathology by Somes and Southam ? Oral & maxillofacial pathology by Naville ? Developmental defects of teeth by www. UNC DENTISTRY. com ? Oral histology by Orban’s ? Oral histology by Tencates ? Normal and abnormal dental development- by Isabelle Miletich & Paul T. Sharpe Human Molecular Genetics, 2003. ? Mouse models of tooth abnormalities Eur J Oral Sci 2008; 116: 1–10