Glossary of terms

1. **Malformation** is a defect due to localized error of morphogenesis resulting in an abnormal shape or structure, with interference in function. E.g. cleft lip.

2. **Deformation** is an alteration in shape and/or structure of a previously normally formed part. E.g. torticollis (wry neck).

3. **Anomaly** is a deviation from normal, same as malformation but without interference in function. E.g. peg shaped laterals, enamel hypoplasia.

4. **Anomalad** is a malformation and subsequently derived structural changes. E.g. Pierre-Robin anomalad (1 degree defect in hypoplastic mandible resulting in abnormal tongue development and positioning, cleft palate & respiratory difficulty).

5. **Syndrome** is a group of symptoms & signs of a disordered function related to one another by means of same anatomic, physiological or biochemical peculiarity presumably of same aetiology but not consequent of a single localized error in morphogenesis. E.g. Parry Romberg syndrome, Pierre-Robin syndrome.

6. **Congenital** is a term used for a condition that occurs at birth or noticed at birth. E.g. congenital syphilis.

7. **Hereditary** refers to a genetic character transmitted from parent to the offspring. E.g. Hereditary ectodermal dysplasia.

8. **Familial** is a term used for disease occurring more frequently in a family than would be expected by chance. E.g. familial fibrous dysplasia, familial hypophostasia.

9. **Teratogens** are environmental aetiological factors responsible for developmental disorders independent of any known inheritance pattern. E.g. radiations.

10. **Chromosome** is a linear thread like structure in the nucleus of a cell containing DNA which transmits genetic information.

11. **Gene** is the basic unit of heredity present on a chromosome.

12. **Locus** is the position occupied by a gene on a chromosome.

13. **Allele** is one of 2 or more different genes containing specific heritable characteristics that occupy loci on paired chromosomes.

14. **Homozygous** means identical alleles (2 dominant or 2 recessive).

15. **Heterozygous** means dissimilar alleles (1 dominant & 1 recessive).

16. **Penetrance** is the capacity to express abnormal genes clinically in a carrier. Frequency with which a heritable characteristic /trait is manifested by individuals carrying the principle gene or genes conditioning it. It could be reduced penetrance or complete penetrance.

17. **Expressivity** is the degree of variation in expression of severity of abnormal genes, given that a gene is penetrant.

18. **Mutation** is the change of genetic constitution of an individual.

19. **Genotype** is the genetic constitution of an individual.

20. **Phenotype** is the expression of the genetic constitution of an individual. Final product of a combination of genetic & environmental influences.

**AUTOSOMAL DOMINANT TRAIT**
- Almost always heterozygous.
- Passed from one generation to the other without break (provided the gene does not affect fertility.
- ½ children are abnormal ½ are normal.
- Affected children pass it down but unaffected cannot.
- Sometimes a dominant disorder is found in a child of 2 apparently normal parents-(not a true biological offspring)

**AUTOSOMAL RECESSIVE TRAIT**
- Majority of individuals have unaffected parents.
- Almost never passed on from one generation to next.
- On an average ¼th of the offspring affected (if both parents are affected heterozygous).
- Parents are likely to be related.

**SEX-LINKED INHERITANCE**
- X-LINKED RECESSIVE: Usually males are affected because they are in a hemizygous state whereas females are heterozygous.
- X-LINKED DOMINANT: Heterozygous females and hemizygous males are affected Females are affected and not males.
CAUSES OF DEVELOPMENTAL DISORDERS IN GENERAL
1. Genetic
2. Environmental
3. Multifactorial

DEVELOPMENTAL DISTURBANCES AFFECTING THE TEETH NUMBER
1. Anodontia
2. Hypodontia
3. Oligodontia
4. Hyperdontia (supernumerary teeth)
5. Predeciduous dentition
6. Post-permanent dentition

1. ANODONTIA
Anodontia means absence of all teeth. True Anodontia is congenital absence of all teeth due to absence of tooth germs. Pseudoanodontia is when teeth are present within the jaw but do not erupt or prevented from erupting. False anodontia is a term which is used when all teeth have been extracted – a case of edentulism.

Types
Total and partial
Causes
1) True Anodontia
   a) Absence of tooth germs (AR)
   b) Aplasia of dental lamina
   c) Environmental
      i) Rubella
      ii) Thalidomide
      iii) Irradiation
      iv) Osteomyelitis
      v) Neoplasms
      vi) Trauma
      vii) Extraction of deciduous teeth along with permanent tooth germs
   d) Associated Syndromes
      i) Ectodermal dysplasia
      ii) EEC Syndrome
2) Pseudoanodontia
   a) Rutherford Syndrome
   b) Lack of eruptive force
3) False Anodontia
   a) Extraction of teeth affected by caries, trauma or periodontal disease

Clinical Significance
Not aesthetic, masticatory functions affected.

2. HYPODONTIA & OLIGODONTIA
If few teeth (six or less) are missing, the term hypodontia is employed; if many teeth (more than six), but not all teeth, are missing, then the condition is called oligodontia (also called partial anodontia).

Types: TRUE / FALSE
Causes:
- Hereditary – AD with point mutations with incomplete penetrance and variable expressivity.
- Evolutionary
- Retention of deciduous teeth
- X-ray irradiation at early age
- Lack of eruptive force

Clinical Features:
More common in girls, equal frequency in maxilla and mandible,
Commonly affected teeth are
- Third molars
- Maxillary and mandibular lateral incisor (permanent and deciduous)
- Mandibular central incisor
- Mandibular second premolar
DEVELOPMENTAL DISORDERS

- Mandibular deciduous canine

Associated Syndromes
- Hypohidrotic Ectodermal Dysplasia
- Chondro Ectodermal Dysplasia
- Crouzon’s Syndrome
- EEC Syndrome
- Down’s Syndrome

Clinical Significance
- Functional problems
- Aesthetic
- Increased freeway space

3. SUPERNUMERARY TEETH
Definition: Teeth in addition to normal series. Also called hyperdontia.
Types:
According to shape
1) Supplemental
2) Conical
3) Tuberculate
4) Odontome

According to position
1) Mesiodens
2) Paramolars
3) Distomolars
4) Ectopic

Causes
- Excessive growth of dental lamina
- Atavistic tendency
- Dichotomy
- Hereditary

Clinical features
- 0.1-3.6%
- Common site – Anterior maxilla, Maxillary molar, Mandibular premolar
- If supernumerary teeth have erupted detection is uncomplicated
- If unerupted – presence of unexplained bulging of oral mucosa or mobility of normal teeth due to root resorption induced by a supernumerary tooth.
- Can be detected by radiographs.

Associated Syndromes
- Gardner’s Syndrome
- Cleidocranial Dysplasia
- Achromodroplasia
- Ehlers Danlos Syndrome

Clinical Significance
- Causes crowding and malpositioning of adjacent teeth
- Prevent eruption of adjacent teeth
- Caries and periodontal disease

4. PREDECIDUOUS DENTITION
These are not teeth but hornified epithelial structures without roots occurring on the gingival over the crest of the ridge.
Causes
- Accessory bud of dental lamina ahead of the deciduous buds
- Bud of accessory dental lamina

① The term atavism (derived from the Latin atavus, a great-grandfather's grandfather, more generally, an ancestor) denotes the tendency to revert to ancestral type. An atavism is an evolutionary throwback, such as traits reappearing which had disappeared generations ago. Atavisms occur because genes for previously existing phenotypical features are often preserved in DNA, even though the genes are not expressed in some or most of the organisms possessing them. For example, babies have been born with a vestigial tail, called “coccygeal process.”
More information at:
http://en.wikipedia.org/wiki/Atavism
5. POST-PERMANENT DENTITION

3rd set of dentition
Retained or unerupted teeth
Multiple unerupted supernumerary teeth

DEVELOPMENTAL DISTURBANCES AFFECTING THE TEETH SIZE

1. Microdontia

Definition: Teeth smaller in size than normal.

Types & Clinical features
- True generalised where all teeth smaller.
  - Pituitary dwarfism
  - Downs syndrome
  - Atrial septal defect
  - Johnson & Blizzard syndrome
  - Dentinogenesis imperfecta
- Relative generalised where normal or small teeth are present in large jaws.
- Localised (focal) where only few teeth are affected
  - E.g.: Peg shaped laterals

Clinical significance
Cosmetic, spacing, and periodontal problems.

2. Macrodontia

Definition: Teeth larger in size than normal

Types & Clinical features
- True generalised, where all teeth are larger as seen in pituitary gigantism.
- Relative generalised, where there are normal or large teeth in small jaws.
- Localised (focal) where only few teeth are affected.
  - Unilateral hypertrophy (facial hemihypertrophy)
  - Crouzon’s syndrome.

Clinical significance: Cosmetic problems, crowding of teeth and abnormal eruption pattern

Differential diagnosis: Fusion of teeth.

3. RhizomegalY

Teeth have larger roots than normal.
E.g.: Maxillary canine may have root length up to 43 mm and may project into maxillary sinus which could be a problem during extraction or a periapical lesion may give rise to problems of maxillary sinus.

4. Rhizomicry.

Teeth have smaller roots than normal.
E.g.: Premolars, 3rd molars
Especially seen in Japanese, where central incisors are commonly affected.
Cause: Heavy occlusal load sustained by teeth during formation.

DEVELOPMENTAL DISORDERS AFFECTING SHAPE & FORM OF TEETH

1) Gemination and Twinning
2) Fusion
3) Concrescence
4) Taurodontism
5) Dilaceration
6) Dens-in-dente
7) Dens evaginatus
8) Supernumerary cusps
9) Supernumerary roots
10) Talon cusp
11) Molarization of bicuspids
12) Enamel pearls
13) Peg shaped teeth
14) Shovel shaped incisors
15) Accentuated palato-gingival groove

1. GEMINATION & TWINSING
Definition: Partial cleavage of tooth bud is termed germination. Complete cleavage of tooth bud into two teeth is called twinning (1 apparently normal and the other supernumerary).
Causes: Trauma
Clinical features:
- Gemination: large clinical crown
- Twinning: 2 separate teeth. More common in deciduous teeth.
Differential diagnosis: Twinning must be differentiated from supernumerary teeth, which are more likely to occur in permanent teeth. In case of germination, one should consider macrodontia, concrescence, and fusion of teeth.
Radiographic features: Enlarged pulp chamber which is partially divided. Only one root is seen in germination.
Clinical significance: Cosmetic problems and crowding of teeth.
Treatment: Shaping the affected teeth to normal appearance or extraction.

2. FUSION (Physiologic fusion)
Definition: Coalescence of 2 tooth buds, which could be normal + normal or normal + supernumerary.
Causes: Trauma or hereditary factors.
Clinical features:
- Known to occur in 0.5% of all patients.
- More in deciduous dentition.
- More common in anterior teeth.
- A groove (bucco-lingual or cervico-incisal) gives a bifid appearance.
- DENTIN ALWAYS CONFLUENT IN CASES OF FUSION.
Radiographic features: Single common pulp chamber or 2 separate ones. Common area of dentin.
Clinical features: Cosmetic, periodontal problems, spacing (if fusion is between 2 normal teeth), crowding (if fusion is between a normal & supernumerary tooth)
Differential diagnosis: Fusion, gemination, hypercementosis, and bony ankylosis.

3. CONCRESCENCE (Pathologic fusion)
Definition: Defined as a condition where roots of two or more teeth have been united by cementum after the formation of crowns
Types:
- True, where there is cemental union of teeth formed by separate tooth buds during tooth formation
- Acquired, where fusion takes place after tooth formation due to hypercementosis
Causes: Crowding of teeth and trauma
Clinical features:
- May occur before or after eruption of teeth.
- Commonly affects maxillary molars.
- Clinical crowns normal in size.
- True type commonly occurs between 2nd & 3rd maxillary molars.
- Fusion of roots may be observed.
- May occur between 2 normal teeth, or between 2 supernumeraries or between a normal tooth and a supernumerary tooth.
Clinical significance: Problem during extraction
Differential diagnosis: Fusion, gemination, hypercementosis and bony ankylosis.

4. TAURODONTISM
This condition was first described by Sir Arthur Keith in 1913.
Definition: Taurodontism is a dental anomaly in which the body of the tooth is enlarged at the expense of roots. Floor of the pulp chamber is lower than normal and there is an increase in the root trunk area.
Causes:
1. Genetic
2. Primitive pattern
3. Atavistic tendency
4. A specialised or retrograde character
5. Mutation

Pathogenesis:
Failure of invagination of Hertwig’s epithelial root sheath to form the root aperture in the normal position during the root development

Types:
A Hypotaurodont Below (apical to) middle third & apical third junction
B Mesotaurodont Middle of apical third
C Hypertaurodont Near apex of apical third

Clinical features:
- No cervical constriction.
- Stocky teeth.
- Affected teeth are parallel-sided which resemble teeth of ungulate animals (bulld-like and hence the term taurodont).
- Both deciduous & permanent teeth may be affected.
- Unilateral or bilateral; usually molars.
- Especially common in American Indians and Eskimos. Also seen in hominids (Neandral man).

Radiographic features:
- Floor of the pulp chamber lower than normal.
- Rectangular shaped teeth.
- Large pulp chamber.
- Lack of cervical constriction.
- Bifurcation or trifurcation more apically located.

Associated syndromes:
- OFDI
- Ackerman syndrome
- Trichodento-osseous syndrome
- Kleinfelter’s syndrome
- Down’s syndrome

Clinical significance:
- Patient should be examined closely to rule out any syndrome.

5. DILACERATION

Definition: Angulation or a sharp bend or curve in the root or crown of a formed tooth.

Aetiology and pathogenesis: Trauma to a partially erupted tooth causes change in position of the calcified portion with an angle to the remainder of the tooth. Injury to overlying deciduous tooth can result in a similar effect to the underlying permanent tooth germ.

Clinical features:
- No sex predilection.
- May not be visible clinically.
- Crown may be bent abnormally may give an indication.
- Seen in both dentitions.

Radiographic features: Dilaceration can be seen on a radiograph, but should be differentiated from a bent radiograph.

Clinical significance:
- Difficulty during extraction or endodontic therapy etc.
- Delay in eruption

6. DENS IN DENTE (DENS INVAGINATUS) (DILATED & GESTANT ODONTOME)

Definition: Malformation with invagination of enamel into the tooth- either in coronal portion or radicular portion.

Pathogenesis:
1. Fusion of two tooth germs.
2. Dislocation of enamel organ with relation to dental papilla.
3. Defects in enamel organ occupied by connective tissue.
4. Local pressure on tooth germs during odontogenesis.
DEVELOPMENTAL DISORDERS

5. Focal growth retardation of inner enamel epithelium.
6. Focal growth stimulation of inner dental epithelium which grows (invaginates) into dental papilla.

Causes:
- Trauma to deciduous tooth germs.
- Pressure on the growing teeth by disproportionately small jaws.

Clinical types (Hallet’s classification):
1. Type 1: Cleft in palatal enamel cervicaly. No expansion or dilatation.
2. Type 2: Definite pit in the cingulum; invagination toward pulp chamber.
3. Type 3: Invagination deep into the pulp chamber. Tooth dilated.
4. Type 4: Invagination occludes whole of coronal pulp chamber & extends beyond DEJ.

Pathology:
(A) CORONAL DENS IN DENTE
- Tooth may appear normal or a deep pit may be seen sometimes with wide entrance.
- Extensive (severe forms), Crown wide & bulbous (ballooned).
- Roots normal or incompletely developed with wide apical foramen.
- Cavity may contain bone, cementum or debris.
- Cavity may be lined with enamel, partially or completely.
- Pulp normal, small or grossly encroached.

(B) RADICULAR DENS IN DENTE
Due to invagination of Hertwig’s epithelial root sheath into dental papilla, which ultimately gets lined with enamel.

Radiographic features: Pear shaped invagination of enamel and dentin with a narrow constriction at the opening on the surface of the tooth. Wide periapical foramen.

Clinical significance: Caries occurs due to thin dentin between deep pit and pulp cavity resulting rapidly in pulpitis followed by periapical abscess formation.

7. DENS EVAGINATUS: (LEONG’S PREMOLAR)

Definition: Developmental malformation that appears clinically as an accessory cusp or a globule of enamel on the occlusal surface between buccal and lingual cusps of premolars and molars.

Causes: Extrusion of dental papilla into inner enamel epithelium with resultant evagination of enamel organ.


Clinical Significance:
- Thin enamel of the extra cusp breaks easily allowing organisms to enter into pulp to cause pulpitis followed by periapical abscess or even osteomyelitis.
- This extra cusp may contribute to incomplete eruption and displacement of teeth.

8. SUPERNUMERARY CUSPS AND ROOTS.

Definition: Supernumerary cusps is an extra cusp than the normal number. Supernumerary root is an extra root than the usual number.

Carabelli’s Cusp: Described by Carabelli in 1842. Supernumerary cusp on lingual surface of molars of maxilla especially common among Caucasians,. Particularly seen nent 1st ,molars. Expression of the character varies from pits and grooves to protubverances and free cusps.


Accessory roots: Most frequently seen on maxillary teeth in Caucasians. But also seen in premolars and canines. 3-rooted lower 1st permanent molars are common in people of mongoloid origin.

9. TALON CUSP

Definition: A cusp projecting lingually from the cingulum area of a maxillary or mandibular permanent incisor that resembles an eagle’s talon.

Features: Also called “T” shaped incisor because of its shape from an incisal view. Cusp may be separated form sloping lingual tooth surface by a deep developmental groove. Cusp is composed of enamel, dentin and pulp.
Clinical significance: Aesthetic problems, caries & problem with occlusal accommodation.
Associated syndromes: Rubinstein-Taybi syndrome.

10. MOLARISATION OF PREMOLARS
Supernumerary cusps and/or increased groove pattern in premolars may result in molar like appearance of premolars. This phenomenon is called molarisation of premolars.

11. SHOVEL-SHAPED INCISORS
Morphological anomaly of crowns of incisor teeth, manifested by prominence of mesial & distal margins which enclose a central fossa. More common in maxillary incisors, more in Mongoloids.

12. ACCENTUATED PALATO-GINGIVAL GROOVE
Palato-gingival groove, usually seen in maxillary lateral incisors, is accentuated increasing the risk of caries development.

13. ENAMEL PEARLS.
Definition: small drop or nodule of enamel situated in an abnormal location of a tooth.
Classified as follows:

Extra dental Enamel pearls: Externally on the tooth.
Pathogenesis:
1. Proliferation or budding occurring at the margin of Enamel organ.
2. Local activity of Hertwig’s epithelial root sheath before it forms cell rests of Malassez.

Intradental Enamel pearls: Within dentin of a tooth
Pathogenesis: Invagination of ameloblasts at a certain period of tooth formation, into the developing dentin resulting in enamel formation surrounded by dentin.
Features: A small core of enamel within the tooth –coronal dentin, radicular dentin, or at the junction of the two (cervical)-surrounded by dentin.
Pathology of Enamel pearls: May consists of enamel, enamel and dentin and/or pulp. Cuticle covering the extradental enamel pearl may be continuous with the sulcular epithelium.
Clinical significance: May create a stagnation area that leads to gingivitis; aesthetic problem.

14. PEG SHAPED LATERALS
See under microdontia.

DEVELOPMENTAL DISORDERS AFFECTING POSITION OF TEETH
1. Ectopia
2. Transmigration
3. Transposition
4. Rotation
5. Inversion
All these developmental disorders are due to displacement of tooth germs.

1. ECTOPIA: Tooth developing or present in a location remote from the area where the tooth is supposed to be located.
DEVELOPMENTAL DISORDERS

2. **TRANSMIGRATION**: Tooth present within the dental arch but in an incorrect location. Usually crosses the midline of the jaws. Also called translocation.

3. **TRANSPOSITION**: Interchanging of positions of a tooth with another tooth. E.g.: canine in a premolar position and vice versa.

4. **ROTATION**: Change in facio-lingual axis of a tooth.

5. **INVERSION**: Tooth is upside down—that is, root is in the oral cavity and crown within the bone.

DEVELOPMENTAL DISORDERS AFFECTING ERUPTION OF TEETH

1. Premature eruption
2. Delayed eruption
3. Impacted and embedded teeth
4. Premature loss of deciduous teeth
5. Delayed shedding of deciduous teeth
6. Eruption sequestrum
7. Ankylosed deciduous (submerged) teeth

1. PREMATURE ERUPTION: NATAL AND NEONATAL TEETH

Definitions:
- **Natal teeth**: teeth present at birth (Dentes connatalis)
- **Neonatal teeth**: Teeth that erupt within first thirty days of life (Dentes neonatalis)

Causes:
- Hereditary
- Superficial placement of tooth germs
- Hormonal: Hyperpituitarism, hyperthyroidism, hypergonadism
- Congenital syphilis, Exanthematous diseases, Hypovitaminoses

Clinical features: Teeth maybe freely mobile and loose because of lack of root formation. Teeth are of normal size and shape, but poorly mineralised. Enamel thin, rough and yellowish—readily chipped off. Greyish brown discoloration.

Pathology: Hypomineralized enamel, large pulp with many vessels, many areas of interglobular dentin, cervical dentin irregular with many cellular inclusions, formation of cellular cementum rare, remnants of enamel matrix seen in decalcified sections.

Associated syndromes:
- Chondroectodermal dysplasia
- Pachonychia congenita
- Hallerman-Streiff syndrome

Complications: Ulceration of ventral surface of tongue (Riga-Fedé syndrome) injury to mother’s nipple during feeding.

Treatment: Removed if they interfere with feeding or if they are very loose because of danger of inhalation. Extraction should be done only after 8 days of birth because of risk of extensive haemorrhage.

PREMATURE ERUPTION OF PERMANENT TEETH

Due to early shedding of deciduous teeth: in Sturge-Weber syndrome due to high degree of vascularity; in hyperthyroidism, hypergonadism, hyperpituitarism. Teeth that are commonly affected are incisors, 2nd premolars and mandibular canine.

DELAYED ERUPTION OF TEETH

Definition: Also called *dentitia tarde* where teeth do not erupt at their normal eruption time. More common in deciduous dentition than permanent dentition.

Causes:

Delayed eruption of single tooth
- Abnormally large crowns
- Early loss of deciduous teeth
- Displacement of tooth germs: ectopia
- Transmigration
- Eruption cyst
- Thick overlying soft tissue (operculum)
- Normal biologic variation

Delayed eruption of multiple teeth
- Hyperpituitarism, hypothyroidism
Infantile rickets
Syndromes: Down’s syndrome, Crouzon’s syndrome, Rutherford syndrome, Goltz syndrome
Hemifacial atrophy
Ectodermal dysplasia, Cleido cranial dysostosis

3. IMPACTED AND EMBEDDED TEETH

Impacted teeth: Teeth that are not in a position to erupt, either because they are not oriented in a proper way or because they are prevented from erupting.
Embedded teeth (unerupted teeth): Teeth that are in correct position and orientation but do not erupt because they lack the eruptive force. Should not be confused with submerged teeth.

4. PREMATURE LOSS OF DECIDUOUS TEETH

Can be due to premature exfoliation (true loss) or due to extraction of deciduous teeth (false loss).

Causes:
Generalized loss
1. Papillion-Lefevre syndrome
2. Juvenile diabetes
3. Cyclic neutropenia
4. Hypophosphatasia

Localized loss
- Caries & sequelae – necessitates extraction
- Periodontal disease

Complications:
If deciduous tooth has to be extracted, permanent tooth germ may be removed along with it or there is a risk of dilaceration of the underlying permanent tooth.

5. DELAYED SHEDDING OF DECIDUOUS TEETH.

Deciduous teeth that do not exfoliate at proper time; teeth undergo attrition faster than permanent teeth

Causes of retention of individual deciduous teeth
1. Absence of particular permanent tooth bud or late development
2. Malpositioned underlying permanent tooth bud
3. Periapical infection of deciduous tooth
4. Submerged deciduous teeth

Commonly affected teeth

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Causes of retention of all or many deciduous teeth
1. Hereditary
2. Oligodontia or anodontia of permanent teeth
3. Delayed eruption of permanent teeth

5. ANKYLOSED DECIDUOUS TEETH (SUBMERGED TEETH)

Definition: This is reimpaction of deciduous tooth, where a previously erupted tooth becomes submerged in the tissues

Etiology:
1. Trauma
2. Infection
3. Disturbed local metabolism
4. Genetic influences
5. Defect in periodontal ligament
6. Premature eruption of permanent 1st molar

Pathogenesis: Ankylosis of deciduous tooth alveolar process growth in the affected area Causes: Neighbouring teeth continue to erupt occlusally & tilt over ankylosed tooth. Affected deciduous tooth becomes SUBMERGED.

Features:
More common in mandible than maxilla.
Nearly always mandibular deciduous 2nd molar is affected.
Tooth may be covered by oral mucosa or occlusal surface may be seen.
Tooth lacks mobility on percussion the ankylosed tooth exhibits a SOLID SOUND IN contrast to the dull sound of a normal tooth.

**Radiographic features:**
- Partial absence of periodontal ligament space around involved tooth.
- Presence of Root resorption (independent of underlying perm. tooth).

### 6. ERUPTION SEQUESTRUM

**Definition:** Tiny irregular spicule of bone overlying the crown of an erupting permanent molar, found just prior to or immediately following the emergence of tips of cusps thro oral mucosa.

**Pathogenesis:** A small osseous fragment is caught in the cuspal fossae of the erupting tooth, which is not resorbed.

**Features:** The central occlusal fossa of the erupting tooth consists of a spicule of bone, which later gets resorbed.

**Clinical significance:** Soreness of the area or pain during mastication due to irritation of soft tissues where it is present.

### DEVELOPMENTAL DISORDERS AFFECTING THE STRUCTURE OF TEETH

1. Enamel hypoplasia / hypocalcification
   a. Amelogenesis imperfecta (hereditary / genetic)
   b. Environmental Enamel hypoplasia
2. Dentinogenesis imperfecta
3. Dentin dysplasia (“Rootless teeth”)
4. Regional Odontodysplasia (“Ghost Teeth”)
5. Dentin hypocalcification
6. Acementogenesis

**Enamel hypoplasia** is a defect in which the enamel of the teeth is hard but thin and deficient in amount as a result of defective enamel matrix formation. The defect can be a small pit or dent in the tooth or can be so widespread that the entire tooth is small and/or mis-shaped. It is characterized by lack of contact between teeth, rapid breakdown of occlusal surfaces, and a yellowish-brown stain that appears where the dentin is exposed. The condition may affect both primary and secondary dentition. It is transmitted genetically or caused by environmental factors.

**Enamel hypocalcification** is a defect in which the enamel of the teeth is soft and undercalcified and opaque in appearance. It is caused by defective mineralization of enamel matrix which has formed normal in quantity. The teeth are chalky in consistency, their surfaces wear down rapidly and are more susceptible to caries, and a yellowish-brown stain appears on the teeth as the underlying dentin is exposed. The condition affects both primary and secondary teeth. It is transmitted genetically or caused by environmental factors. Sometimes the two terms are used interchangeably.

**Causes of Enamel Hypoplasia / Hypocalcification**

1) **LOCAL CAUSES**
   a) Periapical infection of a deciduous tooth
   b) Trauma to the deciduous tooth
   c) Direct injury to the permanent tooth bud

   **Turner’s Hypoplasia**
   When a permanent tooth erupts into the oral cavity, it may have a roughened, irregular, or pitted area in the enamel, sometimes with white or yellow discoloration. This is sometimes referred to as a "Turner's tooth", or "Turner's hypoplasia", and is most commonly observed in
   - Permanent canines or bicuspid teeth
     Secondary to decayed and infected primary molars with periapical inflammation affecting the apically situated developing permanent tooth; the appearance of the abnormality will depend on the severity and longevity of the infection
   - Permanent central incisors
     Secondary to injury to the primary incisors; the deciduous incisor is pushed into the developing tooth underneath it and consequently affects the formation of enamel especially on the labial aspect

2) **GENERAL CAUSES**
   a) **HEREDITARY**
      i) Amelogenesis imperfecta
      ii) Ectodermal dysplasia
iii) EEC syndrome
iv) Chondroectodermal dysplasia
v) Ehlers-Danlos syndrome

b) NEONATAL DISTURBANCES
i) Physical injury at birth
ii) Hypoxia at birth
iii) Haemolytic disease of the new born
iv) Premature birth
v) Cerebral palsy with kernicterus
vi) Infantile tetany
vii) Congenital allergies

c) INFECTIONS
i) Congenital syphilis
ii) Exanthematous fevers: Measles, Scarlet fever, Chicken pox

d) NUTRITIONAL DEFICIENCIES
i) Vitamins A & C deficiencies
ii) Vitamin B deficiency (Rickets)
iii) Mineral salt deficiency: Ca, P, Mg

e) GIT DISTURBANCES
i) Non-Specific diarrhoea & Coeliac syndrome; Incorrect feeding & GIT infections of the infant

f) INTOXICATIONS
i) Fluorosis; Lathyrism; Drugs like Tetracycline

g) ENDOCRINOPATHIES
i) Hypoparathyroidism; Hypothyroidism; Diabetic mothers & Hypogonadism

h) MISCELLANEOUS
i) Calcinosi s; Nephrotic syndrome; congenital heart disease.

AMELOGENESIS IMPERFECTA.

Definition: Hereditary defect of enamel that affects amelogenesis resulting in qualitative and/or quantitative defects of enamel.

Types & Subtypes:
1) HYPOPLASTIC - Quantitative defect
   - Smooth AD
   - Pitted AD
   - Rough AD
   - Smooth X-linked (Lyonizing hypoplasia)
   - Rough AR
   - Localized Heterogeneous AD

2) HYPOCALCIFIED - Qualitative defect
   - Autosomal Dominant
   - Autosomal recessive

3) HYPOMATURATION - Qualitative defect
   - Pigmented AR
   - X-linked
   - Snow-capped

4) WITH OTHER DEFECTS
   - With Epilepsy & mental retardation (AR)
   - With Nephrocalcinosis (AR)
   - Taurodontism+Curly hair+ Sclerotic bone (AD)
   - Onchyolysis+Seborrheic dermatitis+hypohydrosis (AD)

HYPOPLASTIC AMELOGENESIS IMPERFECTA

Pathogenesis:
- Less amount of enamel matrix formation → decreased thickness
- Both primary & permanent teeth are affected
- Mineralization is normal.

Features of different types:
- Smooth AD: Enamel thin, hard & glossy. Teeth small. Associated delay or failure in teeth eruption. Pulp stones seen in unerupted teeth
- Rough AD: Thin Enamel, rough surface.
• **Pitted AD:** Enamel approaches normal thickness. Pinpoint to pinhead-sized pits distributed over the surface of enamel.
• **Rough AR:** Nearly complete lack of Enamel formation. Failure of eruption common. Unerupted teeth frequently undergo resorption.
• **Smooth X-linked:**
  ▪ Males: Thin, hard, smooth, yellowish-brown enamel.
• **Localized:** Horizontal row of pits, linear depressions, large hypoplastic areas – usually in middle 3rd of facial surface.

**Radiographic features:** Normal radiopacity; decreased thickness; pulp stones may be seen.

**HYPOCALCIFIED AMELOGENESIS IMPERFECTA**

**Pathogenesis:**
• Matrix formation is normal
• Mineralization of the matrix is affected.

**Features:**
• Newly erupted teeth with hypocalcification.
• Enamel is of normal thickness.
• But later enamel is lost because it is soft.
• There may delay or failure in eruption.
• Teeth are prone to develop calculus, especially mandibular teeth.

**Radiographic features:**
Radiopacity of Enamel & Dentin nearly the same; difficult to differentiate them on radiographs.

**HYPOMATURATION AMELOGENESIS IMPERFECTA**

**Pathogenesis:**
• Maturation of enamel affected.
• Crystals do not enlarge.
• Defect in Rod sheaths and inter-rod layers.

**Features:** Enamel is
• of normal thickness
• opaque mottled
• brownish yellow to white appearance
• soft and tends to chip off

**Pigmented AR**
• Milky white to shiny brown.
• In ground sections, brown pigment midway between enamel surface & DEJ seen that shows red violet fluorescence.
• These patients often prone to calculus deposition.

**X-linked & “snow-capped” teeth:**

a) X-linked males:
  ▪ Permanent teeth – mottled yellow white that darken later.
  ▪ Deciduous teeth – Opaque white with translucent white mottling.

b) Females:
  Vertical opaque bands on enamel surface
  **Snow-capped** teeth: only the cusp tips affected, which are opaque white appearing like “snow-caps” on mountain peaks (cusps)

**ENVIRONMENTAL ENAMEL HYPOPLASIA**

Enamel hypoplasia / hypocalcification due to environmental factors. *(Please refer to page 12 for causes).* Only few types of environmental enamel hypoplasia will be discussed here.

**ENDEMIC & DENTAL FLUOROSIS**

**Definition:** Endemic fluorosis is disturbance associated with ingestion of excess fluoride. Dental fluorosis is specific disturbance of tooth formation caused by excessive fluoride intake.

**Pathogenesis:** Excessive Fluoride
1. Affects matrix synthesis by ameloblasts **HYPOPLASIA**
2. Later; affects mineralisation **HYPOCALCIFICATION**
The observations that the fluorosed enamel retains a relatively high proportion of immature matrix proteins (amelogenins), support the notion that incomplete removal of amelogenin proteins under excessive fluoride ingestion during development could be responsible for dental fluorosis.

Clinical features:
- Lustreless opaque white patches-mottled, striated or pitted.
- Yellow or brown stain.
- Accentuation of perikymata with multiple pits.
- Loss of tooth form in severe cases.

Dean’s Index

<table>
<thead>
<tr>
<th>Score</th>
<th>Type</th>
<th>Features</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>Smooth, glossy, pale creamy-white translucent surface</td>
</tr>
<tr>
<td></td>
<td></td>
<td>No white patches pits or other abnormalities</td>
</tr>
<tr>
<td>0.5</td>
<td>Questionable</td>
<td>When factors other than fluorosis are suspected to have caused enamel hypoplasia.</td>
</tr>
<tr>
<td>1</td>
<td>Very mild</td>
<td>Small opaque, paper white spots or flecks on enamel covering less than 25% of the tooth surface</td>
</tr>
<tr>
<td>2</td>
<td>Mild</td>
<td>White patches on enamel covering less than 50% of the tooth surface</td>
</tr>
<tr>
<td>3</td>
<td>Moderate</td>
<td>Pitting + brownish staining of enamel</td>
</tr>
<tr>
<td></td>
<td></td>
<td>marked wear on biting surfaces</td>
</tr>
<tr>
<td></td>
<td></td>
<td>All tooth surfaces affected</td>
</tr>
<tr>
<td>4</td>
<td>Severe</td>
<td>Corroded enamel surface, with brownish staining and greater wear rate</td>
</tr>
<tr>
<td></td>
<td></td>
<td>All tooth surfaces affected</td>
</tr>
</tbody>
</table>

Clinical Criteria and Scoring for the TF (Thylstrup-Fejerskov) Index


<table>
<thead>
<tr>
<th>Score</th>
<th>Criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal translucency of enamel remains after prolonged air-drying.</td>
</tr>
<tr>
<td>1</td>
<td>Narrow white lines corresponding to the perikymata.</td>
</tr>
<tr>
<td>2</td>
<td>Smooth surfaces: More pronounced lines of opacity that follow the perikymata. Occasionally confluence of adjacent lines. Occlusal surfaces: Scattered areas of opacity &lt;2 mm in diameter and pronounced opacity of cuspal ridges.</td>
</tr>
<tr>
<td>3</td>
<td>Smooth surfaces: Merging and irregular cloudy areas of opacity. Accentuated drawing of perikymata often visible between opacities. Occlusal surfaces: Confluent areas of marked opacity. Worn areas appear almost normal but usually circumscribed by a rim of opaque enamel.</td>
</tr>
<tr>
<td>4</td>
<td>Smooth surfaces: The entire surface exhibits marked opacity or appears chalky white. Parts of surface exposed to attrition appear less affected. Occlusal surfaces: Entire surface exhibits marked opacity. Attrition is often pronounced shortly after eruption.</td>
</tr>
<tr>
<td>5</td>
<td>Smooth surfaces and occlusal surfaces: Entire surface displays marked opacity with focal loss of outermost enamel (pits) &lt;2 mm in diameter.</td>
</tr>
<tr>
<td>6</td>
<td>Smooth surfaces: Pits are regularly arranged in horizontal bands &lt;2 mm in vertical extension. Occlusal surfaces: Confluent areas &lt;3 mm in diameter exhibit loss of enamel. Marked attrition.</td>
</tr>
<tr>
<td>7</td>
<td>Smooth surfaces: Loss of outermost enamel in irregular areas involving &lt;1/2 of entire surface. Occlusal surfaces: Changes in the morphology caused by merging pits and marked attrition.</td>
</tr>
<tr>
<td>8</td>
<td>Smooth and occlusal surfaces: Loss of outermost enamel involving &gt;1/2 of surface.</td>
</tr>
<tr>
<td>9</td>
<td>Smooth and occlusal surfaces: Loss of main part of enamel with change in anatomic appearance of surface. Cervical rim of almost unaffected enamel is often noted.</td>
</tr>
</tbody>
</table>
DEVELOPMENTAL DISORDERS

HISTOLOGICAL AND CHEMICAL FEATURES

Enamel: Extensive porosity, hypomineralisation, decreased thickness, prominent striae of Retzius.

Dentine: Prominent Von Ebner lines, hypoplastic area, increased interglobular dentine.

Chemical: Presence of fluorapatite, greater protein and nitrogen content.

Other features of endemic fluorosis:
Skeletal Fluorosis: Sclerotic brittle bone, Bone more resistant to resorption.

Other changes: Arthritis, Cataract, Urinary tract calculi, gall stones, hearing problems, GIT problems.

Treatment
Dental fluorosis can be cosmetically treated with procedures like tooth bleaching, microabrasion, and conservative composite restorations or porcelain veneers.

CONGENITAL SYPHILIS

- Congenital syphilis occurs when an infected woman becomes pregnant or when a pregnant woman becomes infected.
- The longer the mother has had the disease at the time of pregnancy, the chance of foetal infection decreases, <40% in early latency & 10% in late latency.
- Foetal transmission
  - Haematogenous spread
  - Direct contact with infected genital lesions (uncommon)

- Pitting type of Enamel hypoplasia
- Hutchinson’s incisors
- Moon’s molars or Fournier’s molars
- Notched incisal edge (absence of central tubercle or calcification centres)
- Peg- shaped laterals
- Screw-driver shaped incisors

ONLY PERMANENT 1st molars and/or central incisors are affected.

Deciduous teeth and other permanent teeth are not affected.

ENAMEL HYPOPLASIA - TREATMENT OPTIONS

Treatment for posterior teeth
1) For sensitive teeth with minimal wear
   a) Desensitizing agent (such as potassium nitrate)
2) For mildly hypoplastic molars
   a) Pit and fissure sealant on the occlusal surface
   b) Periodic re-evaluation (every 6 months)
3) Removal of demineralised enamel and restoration with composite
4) Minimal reduction of tooth and cementation of a stainless steel crown
   a) Periodic re-evaluation clinically and radiographically
5) For permanent molars, stainless steel crowns are intended for temporary use only. These teeth should be restored with a permanent cast crown in the late teen years or early adulthood.
6) In cases where the first permanent molars are unrestorable or marginally restorable, extraction prior to the eruption of the second molars may be a reasonable alternative.

Treatment for anterior teeth:
1) For sensitive teeth with minimal wear
   a) Desensitizing agent (such as potassium nitrate)
2) If there are aesthetic concerns, direct or indirect composite veneers may be bonded to the affected tooth.
3) For permanent anterior teeth, composite or porcelain veneers or porcelain crowns may be used.

DENTINOGENESIS IMPERFECTA

Definition: Hereditary defective dentine formation with opalescent teeth in absence of systemic disease.

Pathogenesis: Due to failure in transformation of foetal collagen (type III collagen) to mature collagen (type I collagen)

Types
- Shields Type I: Assoc with Osteogenesis imperfecta
  - Inherited defect in collagen formation
  - More common in primary teeth
  - Amber translucent tooth colour; bulbous crowns with cervical constriction
  - Periapical lesions, obliteration of pulp chambers, root fractures
• **Shields Type II**: Hereditary Opalescent Dentine  
  o Primary and permanent dentition equally affected  
  o Features same as Type I but no Osteogenesis imperfecta

• **Shields Type III**: Brandywine type/ Maryland type/ Shell teeth  
  o Bell-shaped crowns  
  o Multiple pulp exposures

**Clinical / Radiological features**

- Bluish-gray teeth, Amber-coloured teeth, Bulbous teeth crowns, Absent tooth roots, Absent root canals, Absent pulp chambers, Too small tooth roots, Too small root canals, Too small pulp chambers, Malaligned teeth, and/or Recurring dental abscesses
- Due to the lack of support of the poorly mineralized underlying dentin, the enamel frequently fractures from the teeth leading to rapid wear and attrition of the teeth.
- Teeth are also weaker than normal, making them prone to rapid wear, breakage, and loss. These problems can affect both primary teeth and permanent teeth.
- Brittle bones and blue sclerae (if associated with Osteogenesis imperfecta)

**DENTINE DYSPLASIA**

**Definition**: Rare autosomal-dominant disorder that affects dentine formation. Prevalence about 1:100,000.

**Types and features**

- **Shields Type I: Radicular dentine dysplasia (Rootless teeth)**  
  - Normal crown morphology  
  - multiple periapical radiolucencies  
  - short roots; pulp chambers absent (obliterated)  
  - Wide variation in root formation because dentinal disorganization may occur at different stages of tooth development  
  - Deciduous teeth more severely affected

- **Shields Type II: Coronal dentine dysplasia**  
  - Root length normal in both dentitions  
  - Primary teeth  
    - Clinically resemble dentinogenesis imperfecta - Amber coloured  
    - Radiographically have similar appearance to Type I  
  - Permanent teeth  
    - Normal colour  
    - Pulp chambers enlarged with apical extension-thistle tube - shaped or flame-shaped

**REGIONAL ODONTODYSPLASIA**

**Definition**: Regional odontodysplasia is a localized, non-hereditary developmental abnormality of teeth, characterized by extensive adverse effects on formation of enamel, dentin and pulp – with resultant “Ghost-like” appearance on the radiographs

**Features**

- Occurs in a region or a quadrant  
- Aetiology unknown - Localized arrest in tooth development due to regional vascular developmental anomaly  
- Occurs in both dentitions  
- If present in primary dentition, permanent teeth in area usually affected  
- More in Maxilla 2.5:1  
- Many affected teeth fail to erupt  
- Erupted teeth have small irregular yellow-brown crowns  
- Short roots, enlarged pulp & shell-like crowns “egg-shell” appearance  
- Large diffusely calcified pulp chamber  
- Open apical foramina

**ACEMENTOGENESIS**

In hypophosphatasia, there is generally a lack of acellular cementum formation and severely disrupted cellular cementum. Absence of cementum prevents insertion of periodontal ligament fibres, leading to lack of attachment and thereby resulting in loss of teeth by exfoliation.

*In case you have suggestions, doubts or need clarifications, contact subrarv@gmail.com*