Clinical Update Developmental Anomalies and Oral Pathology in Children
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OVERVIEW

I. PRINCIPLES OF DENTAL DEVELOPMENT
II. ANOMALIES OF NUMBER
III. ANOMALIES OF SHAPE, FORM AND SIZE
IV. ANOMALIES OF COLOR AND TEXTURE
V. ANOMALIES OF ERUPTION
VI. SOFT TISSUE PATHOLOGY
VII. EARLY LOSS OF TEETH

I. PRINCIPLES OF DENTAL DEVELOPMENT

Calcification of the primary dentition begins around 14.5 weeks in utero
Calcification of the permanent dentition begins around birth
Development proceeds from cusp tip to root apex
Stages are simultaneous, not sequential
Symmetry
Sequence
Mandibular arch before maxillary arch
Females before males
There is a range of normal

II. ANOMALIES OF NUMBER

-Anodontia - refers to a total lack of tooth development
-Hypodontia - refers to a lack of development of one or more teeth
-Oligodontia - refers to the lack of development of six or more teeth
-Hyperdontia - refers to the development of an increased number of teeth; teeth of which are referred to as supernumerary

Supernumerary teeth

- Etiology -
-genetic factors implicated
-associated with numerous hereditary syndromes/conditions
-variable expression and penetrance due to environmental factors
-result from defects in the initiation period of the life cycle of the tooth
- Prevalence -
- 0.3-1.8% in primary dentition cited in literature, but in actuality very rare
- 1.4 -3.1% in permanent dentition
- 75% in maxillary anterior fail to erupt
- 2:1 male predominance
- positive correlation with macrodontia
- most develop during first 2 decades of life
- most are conical or irregular in shape

-single tooth supernumerary -
- permanent dentition > primary dentition
- 90% occur in maxilla anterior region
- Most common site = max. incisors > max. 4th molars > mand. 4th molars > premolars, canines, and laterals
- multiple supernumeraries (nonsyndromal)
- mostly in mandible
- premolar > molar > anterior regions
- Clinical presentation -
- extra tooth in primary or permanent dentition
- failure of normal eruption pattern
- Mesiodens - supernumerary tooth in maxillary anterior region
- incidence is 1:100; usually single, but may be paired
- Paramolar - posterior supernumerary tooth lingual or buccal to molar
- Distomolar or distodens - accessory fourth molar
- Radiographic features -
-overlapping of teeth or too many teeth
- Treatment concerns -
- avoid damage to adjacent tooth roots
- treatment timing - if adjacent tooth is within 6 months of erupting, wait for adjacent tooth to erupt
- if adjacent tooth eruption is blocked by supernumerary tooth, supernumerary must be removed before adjacent tooth loses eruption potential
- Treatment and prognosis -
- early identification is critical
- early removal of supernumerary teeth (dentition self corrects in 75% of cases)
- fail to intervene - delayed eruption of adjacent teeth, displaced teeth, malocclusion, cysts

Congenitally Missing Teeth
- Etiology -
- genetic factors implicated (specific genes identified in some cases e.g. *MSX1, PAX9*)
- associated with numerous hereditary syndromes/conditions
- most cases autosomal dominant with incomplete penetrance
- variable expression and penetrance due to environmental factors
- result from defects in the initiation or proliferation periods of the life cycle of the tooth
- may also be caused by tumors and irradiation
- Prevalence -
- anodontia (total lack of tooth development) is rare - most cases related to hereditary hypohydrotic ectodermal dysplasia
- hypodontia (lack of development of one or more teeth) is rare in primary dentition (0.09-0.4% of population) - in primary dentition most often seen in mand incisor region
- most commonly missing permanent tooth – mandibular second premolar (not including 3rd molars)
- hypodontia in permanent dentition seen in 1.7 - 10 % of the population
- seen more frequently in females than males (1.5:1)
- hypodontia more frequently seen in Native American and Asian populations
- frequently seen in patients with cleft palate (lateral incisor)
- positive correlation with microdontia
- Clinical presentation -
- if primary tooth is congenitally missing, permanent successor will also be missing
- spacing between teeth and abnormally low number of teeth
- Radiographic features -
- missing teeth and excess spacing

- Treatment concerns -

-esthetics, bone support, treatment often depends on cause
- Treatment and prognosis -
- depends on severity of case - no treatment or prosthetic replacement may be indicated

III. ANOMALIES OF SHAPE, FORM AND SIZE

Gemination/Fusion/Concrescence

- Gemination = two clinical crowns that share a single root and single root canal - geminated teeth arise from attempted division of a tooth germ by invagination
- Fusion = joining together of two clinical crowns which each have their own root and root canal - fused teeth arise from the joining together of two normal tooth germs
- Concrescence = joining together of two fully formed teeth along the root surfaces by cementum
- gemination, fusion, and concrescence occur in the primary and permanent dentitions with a higher frequency in the anterior and maxillary regions
- count the teeth and take radiographs to determine whether gemination, fusion or concresence

- Prevalence -
- 0.5% prevalence in primary dentition; 0.1% in permanent dentition

- Clinical presentation -
- bilateral cases are rare (0.02% prevalence) in both dentitions
- appear similar and may be differentiated by assessing the number of teeth
- presence in primary dentition may result in delayed or ectopic eruption of permanent teeth
- found more frequently in anterior and maxillary regions
- often involves a second molar in which its roots closely approximate an impacted third molar

- Treatment and prognosis -
- treatment determined by patient's preference - restoration/contouring or extraction
- no treatment required unless the union interferes with eruption; then surgical removal

Mulberry Molars/Hutchinson's Incisors

- Etiology -
- congenital syphilis - caused by chronic infection of *Treponema pallidum*
- modes of transmission are venereal or from mother to fetus
- primary infection is concentrated in young adults (mainly males)

- Prevalence -
- rare
- general incidence declining since 1860's but sharp increases during WWII and 1970's

- Clinical presentation -
- anterior and posterior teeth are affected
- Hutchinson's Incisors - greatest mesio-distal width in mid 1/3 of crown, incisal 1/3 tapers to incisal edge, resulting in a straight-edge screwdriver appearance; incisal edge often resembles a hypoplastic notch.
- Mulberry Molars - taper toward the occlusal surface with a constricted grinding surface; occlusal anatomy is abnormal with numerous disorganized globular projections

- Radiographic features -
- none

- Laboratory findings -
- spirochete organisms (*T. pallidum*) may or may not be present

- Treatment concerns -
- Overall health?

- Treatment and prognosis -
- treat patient with penicillin (drug of choice) (Tetracycline if allergic)
- cosmetic reconstruction of incisors; tx of molars not always indicated

Macrodontia/Microdontia

- tooth size is variable among races and between genders
- Microdontia - the presence of unusually small teeth
- Macrodontia - the presence of unusually large teeth

- Etiology -
  - genetic and environmental factors influence the size of teeth
  - deciduous teeth appear to be affected more by maternal intrauterine influences whereas the permanent teeth appear to be more influenced by the environment

- Prevalence -
  - females have a higher frequency of microdontia and hypodontia
  - males have a higher tendency for macrodontia and hyperdontia
  - isolated microdontia is common - maxillary lateral incisor most common (peg lateral) with a prevalence of 0.8-8.4%
  - may also affect third molars (note that these teeth are also the most frequent teeth to be congenitally missing)

- Clinical presentation -
  - alterations in the size of teeth
  - usually have symmetry of the two sides of the jaws
  - entire dentition is rarely affected.
  - true microdontia is rare (Down Syndrome, pituitary dwarfism and other hereditary disorders)
  - true macrodontia is also rare (pituitary gigantism)

- Radiographic features -
  - abnormally small or large teeth otherwise normal

- Treatment concerns -
  - malocclusions, esthetics?

- Treatment and prognosis -
  - no treatment necessary unless desired for esthetics - maxillary peg laterals are often restored

Taurodontism -

- is an enlargement of the body and pulp chamber of a multi-rooted tooth with apical displacement of the pulpal floor and bifurcation of the roots

- this pattern of molar formation has been found in ancient Neanderthals
- diagnosis is made from radiographic appearance
- affected teeth tend to be rectangular and pulps with increased apico-occlusal height
- classified according to the degree of apical displacement of the pulpal floor (mild = hypotaurondontism; moderate = mesotaurondontism; severe = hypertaurondotism)
- may be unilateral or bilateral
- affects primary dentition < permanent dentition
- no sex predilection
- prevalence varies; in U.S. 2.5-3.2% of population
- may occur as isolated trait or a component of a specific syndrome
- no therapy required; however, are poor abutment teeth due to less strength and stability

Talon cusp-

- is a well delineated additional cusp that is located on the surface of an anterior tooth and extends at least 1/2 the distance from the CEJ to the incisal edge
- predominantly occur on perm max incisors (less on the mand incisors and max canines)
- rarely found on primary maxillary incisors
- in nearly all cases, accessory cusp projects from lingual and forms a three-pronged pattern (Eagle's talon), rare to extend from facial surface
- most talon cusps contain a pulpal extension (not all)

- Radiographic features -
- the cusp overlies the central part of the crown and includes enamel and dentin
- frequency in population ranges from 0.17 to 5.2% (extensive prevalence studies not performed)
- both sexes may be affected
- may occur unilaterally or bilaterally
- no treatment necessary in most cases - must be cautious of pulp if intervention is necessary
- high prevalence in Rubenstein - Taybi syndrome

IV. ANOMALIES OF COLOR AND TEXTURE

Apposition – Process of enamel and dentin matrix formation
Calcification – hardening of previously formed matrix by precipitation of mineral salts

Disturbances during Apposition/Calcification:
- metabolic disturbances
- severe illness/fever
- primary tooth trauma
- primary tooth infection

Enamel Hypoplasia -
- Etiology -
- developmental defect due to an injury to the ameloblasts during enamel formation
- *systemic causes*: birth related trauma, ingestion of chemicals (e.g. lead, fluoride, etc.), chromosomal abnormalities, infections, fevers, inherited diseases, metabolic disorders, nutritional deficiencies
- *local causes*: acute mechanical trauma, electric burn, irradiation, local infection (Turner's Tooth e.g. infection in primary tooth)
- extent of defect depends on:
1) intensity of etiologic factor or insult
2) duration of insult
3) timing of insult during tooth development
- Prevalence -
- very common (no reliable prevalence reported in literature)
- maxillary incisors most affected
- Clinical presentation -
- hypoplasia - defined as quantitatively defective enamel of normal hardness
- hypocalcification - defined as qualitatively defective enamel in which normal amounts of enamel are produced but are hypomineralized - enamel is softer than normal
- teeth may have areas of coronal discoloration, pits, or surface irregularities
- most commonly seen in permanent teeth
- most systemic insults occur between birth and 6 years of age
- anterior teeth and first permanent molars most likely to be affected (from systemic involvement that occurs between 0-18 months)
- Radiographic features -
- fuzzy, unclear border of enamel of erupting teeth
- Treatment concerns -
- risk of future fracture?
- Treatment and prognosis -
often no treatment; esthetic solutions include full crowns, bonded restorations, veneers

Fluorosis -
- Etiology -
    - excessive ingestion of fluoride ion leads to permanent hypomaturation of the enamel in which there is an increased surface and subsurface porosity of the enamel
    - developmental defect due to excess fluoride during matrix deposition
    - most problems associated with fluorosis are esthetic
    - the critical period for clinically significant dental fluorosis is during the 2nd and 3rd years of life (when permanent teeth are forming and the fluoride levels are greater than 1 ppm)
- Prevalence -
    - increasing among children due to the increased ingestion from multiple sources (soft drinks, toothpastes, infant formulas, juices, etc.)
    - some studies suggest 23% of population affected by mild-moderate fluorosis
- Clinical presentation -
    - severity is dose-dependent, increased severity with higher intakes during critical periods
    - mild (white opaque flecks near the incisal edge, enamel remains smooth); moderate (varying degrees of pitting and brownish discoloration); severe (enamel is softer and weaker prone to wear and fracture)
    - affected teeth are caries-resistant, tooth structure presents as areas of lusterless white opaque enamel, which may have zones of yellow to dark brown discoloration
    - often not seen unless teeth are dried
    - often found in a bilaterally symmetric distribution
    - differential diagnosis includes enamel hypoplasia, caries, hypoplasia secondary to trauma
- Radiographic features -
    - none
- Laboratory findings -
    - drinking source fluoride levels in excess of 2 ppm often lead to fluorosis
- Treatment concerns -
    - esthetics, sensitivity? (due to porosity)
- Treatment and prognosis -
    - often no treatment is indicated, consider enamel microabrasion, also consider esthetic restorations (crowns or veneers); bleaching is temporary

Tetracycline Staining -
- Etiology -
    - Tetracycline becomes incorporated into the developing tooth structure resulting in clinical discoloration; can occur in permanent and primary dentitions; occurs in primary teeth because drug crosses placental barrier
- Prevalence -
    - prevalence is decreasing since FDA issued warning in 1963
    - mainly found in youngsters but may affect adults over long-term (due to secondary dentin)
- Clinical presentation -
    - severity of the alteration is dependent upon the time of administration, the dose, and duration
    - produces discoloration that varies from bright yellow to dark brown (discoloration may show a bright yellow fluorescence when visualized with ultraviolet light)
- Radiographic features - none
- Laboratory findings - none
- Treatment concerns - esthetics
- Treatment and prognosis - esthetic solutions include full crowns, bonded restorations, veneers

Turner's Tooth -
- refers to a permanent tooth with a pattern of enamel defects secondary to periapical inflammatory disease or trauma associated with deciduous tooth
- developmental defect due to an injury to the ameloblasts during enamel formation

- appearance varies widely due to the timing and severity of the insult
- enamel defects may vary from focal areas of white, yellow, or brown discolorations to extensive hypoplasia involving the entire crown
- Turner's Teeth resulting from caries most frequently involves the premolars due to relationship with overlying primary molars (anterior teeth are less involved because crown formation is completed before caries develops)
- Turner's Teeth resulting from trauma are most likely maxillary anterior teeth (facial surface)
- studies have shown that 23% of the corresponding teeth develop disturbances following traumatic injury to the deciduous tooth (max central incisor > max. lateral incisor)
- trauma may also lead to dilaceration
- treatment may include restorations and esthetic improvements

Amelogenesis Imperfecta

- encompasses a complicated group of conditions that demonstrate developmental alterations in the structure of enamel in the absence of a systemic disorder.
- developmental defect due to an injury to the ameloblasts during enamel formation
- at least 14 different hereditary subtypes exist with numerous types of inheritance and a variety of clinical manifestations.
- several classification schemes exist - Witkop's is the most widely accepted
- overall prevalence is 1:14,000
- clinical, histologic, and genetic criteria are needed to accurately diagnose/classify AI
- treatment = full coverage crowns (dentin and roots are usually normal); orthodontics?

Hypocalcified type

- most common type of Amelogenesis Imperfecta (1:20,000)
- autosomal dominant mode of transmission
- enamel of newly erupted teeth is of normal thickness and has a dull, lusterless opaque-white, honey-colored, yellow-brown-orange appearance
- enamel is soft and is lost soon after eruption - leaves crown of dentin only
- enamel is of cheesy consistency - can be scraped off with an explorer
- the enamel at the DEJ is better calcified
- patients often experience sensitivity following loss of enamel and delayed eruption
- 60% of patients have anterior open bite
- teeth tend to form calculus rapidly
- may see resorption of unerupted teeth
- loss of enamel is less at the cervical region of the teeth - and is uneven
- radiographic - dentin is often more radiodense than the enamel; enamel appears moth-eaten

Hypoplastic type -

- alteration due to the inadequate deposition of enamel matrix (not full thickness)
- matrix that is present is appropriately mineralized
- autosomal dominant (AD) or autosomal recessive (AR) mode of inheritance
- five hypoplastic subtypes -
    1) Pitted hypoplastic Amelogenesis Imperfecta - AD
        - affects primary and permanent dentition
        - pinpoint pits randomly distributed over surface
        - enamel on newly erupted teeth hard with a normal yellow/white color
        - staining occurs after exposure to oral environment
    2) Local hypoplastic Amelogenesis Imperfecta - AD
        - horizontal rows of pits/linear depressions or one large hypoplastic area with hypocalcification adjacent to and below the hypoplastic area (mid-buccal)
    3) Smooth hypoplastic Amelogenesis Imperfecta - AD
        - enamel is thin, hard, and glossy with smooth surface
        - may be yellow-opaque-white to brown in color

- 1/4 to 1/8 of normal thickness, enamel may abrade and not contact proximally
- some enamel may be missing upon eruption
- delay or failure to erupt may occur with resorption of teeth in alveolus (impactions or partially resorbed teeth may be present)
- radiographic findings - enamel appears missing; pulpal calcifications seen

4) Rough hypoplastic Amelogenesis Imperfecta - AD
- enamel is hard with a rough, granular surface
- primary and permanent teeth affected
- enamel may chip away
- white or yellow/white when newly erupted
- enamel 1/4 to 1/8 of normal thickness
- appear to have a more normal tooth outline
- radiographic findings - thin line of enamel covering dentin

5) Rough Amelogenesis Imperfecta (Enamel agenesis) - AR
- yellow color (like dentin)
- enamel surface is rough and granular (like ground glass)
- teeth widely spaced apart - open proximal contacts
- all patients have anterior open bite
- unerupted teeth often undergo resorption
- primary and permanent dentitions affected
- radiograph findings - no evidence of enamel

Hypomaturation type -
- enamel mottled (brown-yellow-white in color)
- enamel normal thickness and teeth have proximal contact points
- teeth are soft; tooth structure chips away from dentin
- may be autosomal dominant (AD) or autosomal recessive (AR)
- three hypomaturation subtypes

1) Hypomaturation - hypoplastic Amelogenesis Imperfecta with taurodontism - AD
- distinguishable from Trichodentosseous (TDO) syndrome
- most striking defect found in males
- permanent teeth are mottled yellow-white
- enamel approaches normal thickness (slightly thinner)
- soft enamel - leads to lost enamel but not as fast as hypocalcified forms
- primary teeth of affected males have a ground glass opaque white appearance
  - color of primary teeth is opaque-white with translucent white mottling
  - surface is moderately smooth (not as smooth as hypocalcified forms)
  - in females, primary and permanent teeth show alternating vertical bands of opaque-white and normal translucent enamel which vary in width

2) Pigmented hypomaturation Amelogenesis Imperfecta - AR
- primary and permanent dentitions affected
- enamel has milky appearance upon eruption but becomes stained following contact with oral cavity
- enamel is normal thickness but chips away from dentin
- resorption may occur in alveolus before eruption of teeth
- some areas are more severely affected resembling hypocalcified types
- radiographic findings - enamel is less radiodense, no contrast with dentin

3) Snow capped teeth
- white-opaque hypomaturated enamel seen on incisal or occlusal surface
- fairly common disorder
- opaqueness may be solid or flecked
- maxillary teeth more likely to be affected than mandibular teeth
- both primary and permanent teeth are affected

Dentinogenesis Imperfecta -

- developmental defect of the matrix common to both bone and teeth
- specific genetic defect = mutation in type I collagen
- one of the most common disorders inherited in an autosomal dominant manner (prevalence 1 in 8,000)

Type I Dentinogenesis Imperfecta (Shields Type I)

- one of several clinical manifestations of osteogenesis imperfecta (multiple bone fractures, hyperextensible joints, blue sclera, progressive deafness)

- Etiology -
  - genetic defect
- Prevalence -
  - autosomal dominant and autosomal recessive forms with variable expressivity
  - autosomal recessive form (osteogenesis congenita) is severe and lethal
- Clinical presentation -
  - clinical presentation of Type I appears to be more varied than Type II
  - both dentitions affected (primary teeth more affected than permanent teeth)
  - tooth color varies from blue to brown (with amber translucency)
  - enamel tends to break away (permits more rapid attrition of exposed softer dentin)
  - crowns are bulbous with pronounced cervical constrictions
- Radiographic features -
  - initially pulp chambers are abnormally wide but then obliterate after eruption
  - short and constricted roots (both dentitions), root canals absent or thread-like
  - cervical constriction gives teeth bulbous appearance
  - differential diagnosis includes Dentin Dysplasia
- Laboratory findings -
  - dentin adjacent to the DEJ appears normal, but the remaining dentin is abnormal
  - atypical odontoblasts line the pulp surface
  - dentinal tubules are larger and irregular, but are reduced in number
  - pulpal chamber is completely replaced by irregular dentin over time
  - enamel appears normal, but DEJ is smooth rather than scalloped
  - 1/3 patients may also have hypoplastic or hypocalcified enamel defects
- Treatment concerns -
  - occlusal vertical dimension
  - esthetics
  - sensitivity
- Treatment and prognosis -
  - in the past, most cases were untreated until adulthood, then extraction and dentures were performed, now treatment involves two phases: 1) increase vertical dimension to compensate for attrition and preserve integrity of dentition, maintain esthetics and function until adulthood (transitional phase); 2) permanent restorations i.e. fixed or removable prosthodontics

Type II Dentinogenesis Imperfecta (Hereditary Opalescent Dentin)

- Etiology -
  - no skeletal involvement
  - very similar to Type I but classified as a separate entity because: 1) phenotypic variability is less in Type II; 2) many cases reported which had DI but no OI involvement; 3) in Type II DI, both dentitions equally affected (in Type I primary > permanent)
- Prevalence -
  - one of the most common disorders inherited in an autosomal dominant manner (prevalence 1 in 8,000)
- Clinical presentation -
  - very similar to DI Type I except all teeth affected equally
- Radiographic features -
  - same as in DI Type I
- Treatment concerns -

- same as in DI Type I
- Treatment and prognosis -
- same as in DI Type I

Type III Dentinogenesis Imperfecta (Brandywine Variant)
- described in a tri-racial isolate in Brandywine, Maryland
- clinical features closely resemble those of type II Dentinogenesis Imperfecta
- major radiographic finding is the diminished width of mineralized dentin giving teeth a "shell-like" appearance
- pulps eventually become obliterated with age (like DI type I and II)
- multiple pulp exposures and periapical radiolucencies are also evident

Dentin Dysplasia -
Dentin Dysplasia Type I (Radicular Dentin Dysplasia or "Rootless Teeth")
- Etiology -
- autosomal dominant pattern of inheritance
- caused by the loss of organization of the root dentin leading to a short root
- enamel and coronal dentin are well formed but radicular dentin is not
- wide variation in root formation is produced due to dentinal disorganization during different stages of tooth development
- variability is most pronounced in permanent teeth (may vary from tooth to tooth)
- Prevalence -
- rare - 1:100,000 (only 30 cases in the literature)
- Clinical presentation -
- malalignment and malpositioning of teeth
- clinical crowns are normal shape size and consistency - may be amber in color
- occurs in permanent teeth before eruption (crescent shaped pulp remnants)
- teeth have sharp conical apical constrictions
- Radiographic features -
- deciduous teeth are severely affected with little or no pulp (pulp obliteration)
- if any pulp present, chevron or crescent shaped
- markedly short or absent roots.
- calcified masses in pulp chamber
- multiple periapical radiolucencies
- Laboratory findings -
- enamel and dentin adjacent to DEJ appear normal, deeper dentin layers show irregular dentinal tubules in both primary and permanent dentitions
- globular masses of abnormal dentin seen
- histologic appearance of "streams flowing around boulders"
- Treatment concerns -
- excessive wear and overclosure; esthetics
- Treatment and prognosis -
- preventive care is crucial
- protect tooth from wear and improve esthetics; full coverage crowns at an early age
- because pulp vascular channels run close to DEJ, even shallow restorations can lead to pulp necrosis
- short roots compromise prosthodontic options

Dentin Dysplasia Type II (Coronal Dentin Dysplasia)
- Etiology -
- autosomal dominant mode of transmission (100% penetrance)
- Prevalence -
- very rare - only 400 documented cases in the literature
- Clinical presentation -
- amber translucency of primary teeth (closely resembles DI Type II)
- permanent teeth normal color

- cervical constriction leads to bulbous appearance of crowns

- Radiographic features -
  - primary teeth have obliterated pulp chambers by age 5-6 years
  - permanent teeth have evidence of pulp chambers (thistle-tubed appearance of teeth due to radicular extension of the pulp chamber)
  - nearly all teeth reveal pulp stones in an unusual shaped pulp chamber
  - periapical radiolucencies not as common as seen in Dentin Dysplasia Type I
- Treatment concerns -
  - same as in Type I
- Treatment and prognosis -
  - same as in Type I, except root length is not compromised

V. ANOMALIES OF ERUPTION

Ectopic Eruption

-any eruption that deviates from a normal pathway (some definitions include resorption)

- Etiology -
  - thought to be related to malposition of tooth, lack of space
- Prevalence -
  - fairly common, approximately 5% of the population
- Clinical presentation -
  - most commonly seen in molar and incisor regions
  - delayed eruption of permanent tooth
  - may occur bilaterally or unilaterally
  - mainly occurs with permanent canines and 1st molars
- Radiographic features -
  - partial resorption of tooth
- Treatment concerns -
  - premature space loss
  - oral hygiene
  - complications - abscess, discomfort
- Treatment and prognosis -
  - ectopically erupting maxillary first permanent molars spontaneously correct (66%)
  - may need to intervene - brass wire, separator, split saddle appl., headgear to distalize first permanent molar, extraction of primary tooth

Delayed Eruption Related to:
Ankylosis
Premature loss of primary teeth
Supernumerary teeth
Malposition/impaction/ectopic eruption
Lack of adequate space
Cleidocranial Dysplasia
Cherubism
Trisomy 21
Hypothyroidism
Hypopituitarism
Unknown Etiology

Early Eruption Related to:
Natal/Neonatal Teeth
Premature loss of primary teeth
Increased Nutritional Intake???

Hypophosphatasia -

- Etiology -
    - rare metabolic bone disease - most severe forms are transmitted in an autosomal recessive manner; milder forms can be transmitted in an autosomal dominant manner
    - severity not related to serum alkaline phosphatase levels
    - disease manifests in the first few years of life
    - is one of the main causes of premature loss of the primary dentition
- Prevalence -
    - severe forms occur in 1 in 100,000 live births
    - one source estimates that 1 in 300 in US is a carrier for hypophosphatasia
- Clinical presentation -
    - enlarged pulp chambers of primary teeth
    - loosening and premature loss of the primary teeth (especially the incisors)
    - alveolar bone loss with a predisposition for the anterior segments of the mandible or maxilla
    - hypoplasia or true aplasia of cementum along the root surface
    - root deficiency (especially towards the apex)
    - hypoplastic enamel defects
    - blue schlera during infancy and childhood (similar to Osteogenesis Imperfecta)
    - depending on severity, may see deformity of arms, legs, and chest
    - recurrent fractures, recurrent bouts of pneumonia
- Radiographic features -
    - enlarged pulp chambers and canals
    - cemental hypoplasia
    - short roots
    - young children: irregular defects in long bones (irregular/streaky ossification)
    - older children: multiple radiolucent areas (beaten copper appearance)
    - adults: general decrease in bone density, lucency of maxilla and mandible, thin cortical bone and lamina dura, deficient alveolar bone, decrease in enamel thickness
- Laboratory findings -
    - reduced alkaline phosphatase in blood or tissue
    - increased levels of phosphoethanolamine in the blood and urine
- Treatment concerns -
    - space loss leading to space deficiency when permanent teeth erupt
- Treatment and prognosis -
    - dental prostheses often compromised due to poor alveolar bone support
    - therapeutic measures usually unsuccessful
    - vitamin D in high doses results in partial improvement but may lead to deposition of calcium
    - prognosis depends upon age on onset - infantile onset = poor, adult onset = fair to good

Regional Odontodysplasia (Ghost teeth)
- Etiology -
    - a localized, non-hereditary developmental anomaly of teeth characterized by a local arrest in tooth development
    - most cases are idiopathic, but some are related to syndromes, growth abnormalities, neural disorders and vascular malformations
    - may be related to vascular nevi of the head and neck
- Prevalence -
    - uncommon finding that occurs in both dentitions
    - no gender predilection
    - occurs in bimodal peaks that correlate with the normal eruption times of both dentitions (2-4 years and 7-11 years)
    - typically involves a focal area of the dentition affecting several contiguous teeth
    - maxilla is favored (2.5:1) with a predilection towards the anterior teeth
    - involvement of > 2 quadrants is rare
    - involvement of the primary dentition is followed by similarly affected permanent teeth

- Clinical presentation -
    - many affected teeth fail to erupt
    - when erupted, teeth have small irregular crowns that are yellow to brown and rough
    - caries and periapical lesions are common
- Radiographic features -
    - altered teeth have thin enamel and dentin surrounding an enlarged radiolucent pulp, resulting in a pale, wispy image of a tooth
    - lack of contrast between enamel and dentin
    - short roots and open apices may be seen
    - pulp stones are prominent
- Treatment concerns -
    - esthetics, sensitivity that may impact normal feeding habits
- Treatment and prognosis -
    - treatment is directed towards retention of the altered teeth whenever possible
    - most affected teeth are removed
    - preparation of teeth is contraindicated due to the nature of the coronal hard tissue and ease of pulp exposure

Ankylosis (see Messer L. Ankylosed Primary Molars. (1980) Ped Dent 2:37-47.)
- Etiology -
    - biological mechanism not known but several hypotheses exist: 1) deficient eruptive force 2) disturbed metabolism of PDL 3) trauma 4) deficient local vertical bone growth 5) local inflammation 6) hereditary role 7) disturbance in interaction between normal resorption and hard tissue repair
- Prevalence -
    - 1.3-38.5% documented in literature (most likely 1 - 9 %)
    - most commonly seen in primary dentition
    - condition is likely to recur
- Clinical presentation -
    - crown is located below the occlusal plane
    - tooth is immobile to manual movement
    - dull thump is heard upon percussion
    - may cause delayed eruption of permanent teeth
    - may occur bilaterally or unilaterally
    - mandibular teeth more affected than maxillary teeth
    - maxillary molars undergo ankylosis earlier and more severely than mandibular molars
    - can occur in primary and permanent dentitions
- Radiographic features -
    - best indicated in series of films - marginal ridges of teeth at different heights
    - difficult to differentiate between PDL and bone
- Laboratory findings -
    - fusion of cementum and alveolar bone
- Treatment concerns -
    - an ankylosed tooth allowed to remain in place may permit the adjacent tooth to tip or may allow opposing tooth to supraerupt
    - delayed eruption of permanent teeth???
    - malocclusion
    - sometimes difficult to extract
- Treatment and prognosis -
    - monitor exfoliation of primary tooth
    - if become too submerged may cause space loss
    - must evaluate succedaneous tooth
    - extraction indicated if too submerged, causing malalignment or discomfort

Natal and Neonatal Teeth- (see Zhu J (1995) Natal and Neonatal Teeth. J Dent Child 62:123.)
Natal teeth = refers to teeth present at birth

Neonatal teeth = refers to teeth appearing within the first 30 days of life

- Etiology -
  - cause thought to be related to superficial position of the developing tooth germ
  - may be a familial or hereditary factor (8-62%)
  - higher prevalence associated with syndromes e.g. CL(P)
- Prevalence -
  - varies considerably from 1 in 700 to 1 in 30,000 births
  - natal teeth are more common than neonatal teeth (3:1)
  - rarely will a child present with both natal and neonatal teeth
  - 38-75% of natal and neonatal teeth occur in pairs
  - 85% of natal teeth are mand incisors, 11% are max. incisors and 4% are posterior
  - Natal and Neonatal teeth usually represent normal primary teeth and only 1-10% are supernumerary teeth
- Clinical presentation -
  - often mobile
  - may resemble normal primary teeth but are often poorly developed, small, conical, yellowish, with hypoplastic enamel, poorly formed dentin, and partial or total failure of root development
- Radiographic features -
  - hypoplastic enamel
  - no cementum present
- Treatment concerns -
  - possibility of aspiration by child - although this has never been reported
  - tooth will likely disturb Mother if child is breast feeding
  - sharp incisal edges may cause ulceration of the ventral tongue - Riga-Fede Disease
- Treatment and prognosis -
  - if tooth is not causing any difficulty to Mother or infant - leave alone
  - teeth that survive beyond 4 months have good prognosis
  - Riga-Fede disease is not an absolute indication for removal - can smooth sharp edges
  - extraction of these teeth should be followed by curettage of the socket to prevent continued development of the dental papilla
  - no need to delay removal/treatment until child is older

Hereditary Hypohydrotic Ectodermal Dysplasia -

- Etiology -
  - congenital dysplasia of primary ectodermal structures (may affect any ectodermally derived tissue (eye, hair, teeth, etc.)
  - often X-linked recessive (may be autosomal dominant or recessive)
  - males > females
- Clinical presentation -
  - exhibit soft, smooth, thin dry skin with complete or partial absence of sweat glands
  - patients cannot perspire thus cannot tolerate warm temperatures without cooling jackets
  - hair follicles and sebaceous glands are absent or defective
  - hair tends to be fine, blond, and scant
  - depressed bridge of nose, pronounced supraorbital ridges, frontal bossing, protruded lips, straight profile, ears may be involved
  - oral findings: anodontia or oligodontia with malformed cone-shaped teeth, jaw growth is normal, reduced vertical dimension due to no alveolar bone growth, high palatal vault - cleft palate is sometimes seen, hypoplastic salivary glands - xerostomia, more prone to caries
- Treatment and prognosis -
  - none/dentures or overdentures/ implant human trials currently underway at NIH

Cleidocranial Dysplasia -

- Etiology -
  - unknown etiology
  - affects male and females equally

- Clinical presentation -
    - flattened skull
    - brachycephalic head - short and wide
    - Oral findings:
    - high narrow palate, cleft palate common
    - small underdeveloped maxilla and large mandibles
    - prolonged retention of primary teeth and delayed eruption of permanent teeth
- Radiographic features -
    - defective or absent clavicles
    - multiple unerupted teeth; supernumerary teeth
    - fontanelles and sutures remain open;
    - absence/decrease in the amount of cellular cementum on permanent tooth roots
    - anodontia seen in rare cases
- Treatment and prognosis -
    - surgical removal of unerupted teeth
    - maintain primary teeth if there is no chance for permanent teeth to erupt

Cherubism -
- Etiology -
    - uncommon disease involving the jaws
    - autosomal dominant with variable expression (100% in males, 70% in females)
- Prevalence - males > females (2:1)
- Clinical presentation -
    - manifests in early childhood (<3 yrs)
    - progressive, painless swelling of the jaws (mand or max) causing a chubby face
    - mandible > maxilla
    - jaws are firm and hard
    - enlarged palate
    - regional lymphadenopathy
    - no associated systemic manifestations
    - spontaneous shedding of primary dentition
    - defective permanent dentition with absent teeth
- Radiographic features -
    - bilateral destruction of bone with expansion and severe thinning of cortical plates
    - multilocular appearance
    - numerous unerupted and displaced teeth "floating in cyst-like spaces"
-Laboratory findings - all within normal limits
-Treatment and prognosis -
    - entity becomes static at puberty
    - surgical correction for cosmetics after puberty
    - often normal by age 30

Histiocytosis X -
    - inflammatory reticuloendothelial condition, most likely a single disease with three forms (Hand Schuller-Christian disease, eosinophilic granuloma of the bone, and Letterer-Siwe disease)
    - caused from a proliferative reticuloendothelial disturbance (the histiocyte)
    - eosinophils common early but disappear in older individuals

1) Eosinophilic Granuloma of Bone - (unifocal eosinophilic granuloma)
    - solitary or multiple bone lesions without visceral involvement
    - occurs primarily in older children and young adults
    - males > females (2:1)
- Clinical presentation -
    - may exhibit with no physical signs or symptoms
    - may present with local pain, swelling, and/or tenderness, general malaise and fever

- may present with loose teeth
- skull and mandible are commonly affected sites
- destructive and often well demarcated lesions (cyst-like)

- Radiographic features -
- irregular radiolucent lesions (single or multiple) involving superficial alveolar bone
- cortex is often destroyed, pathologic fractures may occur
- if lesions are located in jaw, usually appear well circumscribed resembling cysts, periapical granulomas, or even periodontal disease

- Treatment and prognosis -
- excellent prognosis
- cured by curettage and radiation therapy
- symptoms usually subside within 2 weeks

2) Hand-Schuller-Christian Disease (Multifocal Eosinophilic Granuloma)
- occurs primarily early in life (< 5 years of age) and has a chronic clinical course
- males > females (2:1)
- soft tissue and skeleton may be involved

- Clinical presentation - triad in 25% of cases
- triad consists of:
1) single or multiple areas of "punched out" bone destruction in skull;
2) unilateral or bilateral exophthalmos;
3) diabetes insipidus with or without symptoms of dyspituitarism (dwarfism)
- other clinical features include: facial asymmetry due to swelling of facial bones, otitis media is common, nodular or papular skin lesions
- oral findings may be the earliest sign of disease: precocious loss of teeth, failure of tooth sockets to heal, loss of supporting alveolar bone, sore mouth (with or without ulcers), bad taste, halitosis, gingivitis

- Radiographic features -
- individual skull lesions are often sharply outlined
- lesions within jaws may be diffuse (may simply manifest as alveolar bone destruction with tooth displacement)

-Laboratory findings -
- anemia, leukopenia, thrombocytopenia

-Treatment and prognosis -
- good prognosis - 50% undergo spontaneous remission
- if treatment is indicated, curettage or excision of lesions
- also consider radiation therapy and chemotherapy
- prognosis is dependent upon extent of disease at initial diagnosis

3) Letterer-Siwe Disease -
- acute fulminating histiocytic disorder in infants less than 3 years of age
- both skeletal and extraskeletal tissues, including the skin, involved

- Clinical presentation -
- usually begins as a skin rash of scalp, trunk and extremities
- persistent low-grade fever with malaise and irritability
- splenomegaly, hepatomegaly, lymphadenopathy seen early
- skeletal involvement seen later
- oral findings: oral ulcerative lesions and gingival hyperplasia, diffuse destruction of maxillary and mandibular bone causing loosening and premature loss of teeth
- sometimes the disease is so rapid, oral involvement not seen

- Laboratory findings -
- progressive anemia, leukopenia, thrombocytopenia

- Treatment and prognosis -
- extremely poor prognosis
- rapid and fatal disease in short period of time
- low percentage of cases show response to chemotherapy

VI. SOFT TISSUE PATHOLOGY

See:

Flaitz CM, Coleman GC
Differential diagnosis of oral enlargements in children.
Pediatr Dent (1995 Jul-Aug) 17(4):294-300

Sciubba JJ
Oral soft tissue pathology in children.
N Y State Dent J (1992 Feb) 58(2):30-4

Hebert AA, Lopez MD.
Oral lesions in pediatric patients.
Advances in Dermatology. 12:169-93; discussion 194, 1997.

VII. EARLY LOSS OF TEETH

DIFFERENTIAL DIAGNOSIS OF EARLY TOOTH LOSS IN CHILDREN

1. Trauma
2. Caries
3. hypophosphatasia - primary and perm dentitions, enlarged pulp spaces, no ging inflamm, decr serum alkaline phosphatase, incr urinary phosphoethanolamine, cemental dysplasia, do not really see alveolar bone loss, skeletal lesions, bone pain, bone fractures, bowing of legs, wide osteoid seams, hereditary
4. Papillon-LeFevre
5. Letterer Siwe (Histiocytosis X) “non-lipid reticuloendotheliosis” age of onset less than 3 years, lymphomatous proliferation of poorly differentiated histiocytes
6. Hand-Schuller-Christian (Histiocytosis X) “non-lipid reticuloendotheliosis” non-neoplastic reactions of well differentiated histiocytes to an unknown stimulus
7. Eosinophillic Granuloma (Histiocytosis X) “non-lipid reticuloendotheliosis”, soliatry or unifocal lesion in bone - only bone affected
8. Cyclic Neutropenia
9. Chronic Neutropenia (Agranulocytosis)
10. Familial Fibrous Dysplasia (Cherubism)
11. Localized Prepubertal Periodontitis
12. Generalized Prepubertal Periodontitis (accd to Page, this is the oral manifestation of leukocyte adhesion deficiency)
13. Chediak-Higashi
14. Acatalasia
15. Crohn’s Disease
16. Acrodynia - Mercury toxicity - hands, feet, nose, and cheeks are red/pink, increased irritability and sweating
17. Pseudohypophosphatasia - normal serum alkaline phosphatase levels
18. Leukemia
19. Facial hemihypertrophy
20. Bruxism (seen in severe burn patients)
21. Down Syndrome (Trisomy 21)

22. Burkitt's Lymphoma
23. Regional Odontodysplasia
24. Natal/Neonatal Teeth
25. Sickle Cell Anemia
26. Juvenile diabetes
27. Acquired Immune Deficiency
28. Nutritional deficiencies (i.e., scurvy)
29. Rickett's disease
30. Dentin dysplasia