Developmental Enamel Defects

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Enamel defects

Stages of enamel development

- Matrix formation: protein laid down
- Mineralisation: mineral deposition, majority of original proteins removed
  - diffuse, opaque white, soft enamel
- Maturation: final mineralization
  - translucent, hard enamel

• COLOUR

• STRUCTURE
Life cycle of ameloblast

Morphogenic stage
Organizing stage
Formative (secretory) stage
Maturative stage
Protective stage
Desmolytic stage
Incremental deposition

**Figure 5-26.** Summary of the stages of enamel mineralization. Initial enamel is formed in A and becomes more mature (more calcified) in B as further matrix is formed. (C). Further increments are formed. (D). Mineralization and matrix deposition increases. (E). Enamel matrix is formed on the side of the cusps. (F). Final matrix is formed and progresses cervically.
Objectives of diagnosis and management

- Identify anomaly
- Reassure child and parents
- Provide information
  - Expectations – aesthetics and function
  - Treatment options
  - Optimal age
- Interdisciplinary management
- Genetic counselling
- Prevention
Definitions

• Opacity
  – Alteration in the translucency of enamel which has a smooth surface and is of normal thickness

• Hypoplasia
  – Defect involving the surface of enamel associated with reduced thickness of enamel (pits, linear defects, areas of missing enamel, grooves)

• Hypominalisation
  – Defective calcification of enamel
• Principal category
  – Demarcated
  – Diffuse
  – Hypoplastic
  – Combination of these

• Subtype
  – combinations

Any defect
Principal category of DDE
1. Demarcated opacities
2. Diffuse opacities
3. Hypoplasia
4. Combination of three main types

Subtype of DDE
1.1 Demarcated opacities (white/cream)
1.2 Demarcated opacities (yellow/brown)
2.1 Diffuse opacities – lines/patchy
2.2 Diffuse opacities – confluent
2.3 Confluent/patchy + staining + loss of enamel
3.1 Hypoplasia – pits
3.2 Hypoplasia – missing enamel
4.1 Demarcated opacities + diffuse opacities
4.2 Demarcated opacities + hypoplasia
4.3 Diffuse opacities + hypoplasia
4.4 All three main types
Developmental defects of enamel (DDE Index)

- Area affected
- Shape of lesion
- Demarcation
- Color
- Teeth affected
  Symmetrical/asymmetrical
  Localised / generalised
  Demarcated / diffuse
Enamel defects in primary dentition

- Protective in-utero effect
- Thinner layer of enamel
- Whiter colour of teeth
- Less expression of some genetic diseases in primary dentition
- Undetected
Differential Diagnosis

Chronological Enamel Hypoplasia

- Quantitative defect
- Thin enamel
- Normal quality
Chronological Hypoplasia
Fluorosis

- Diffuse opacities
- Chronological distribution
Defects of Enamel

- **Acquired**
  - Fluorosis
  - Low birthweight
  - **Nutrition:** Vitamin A, C, D, Ca, P
  - **Infections:** Rubella, CMV, syphilis
  - ** Syndromes:** Down syndrome, VDRR, Epidermolysis bullosa, Hunter/Hurler, Hypoparathyroidism
  - Allergies/Asthma
  - Radiation
- **Inherited**
  - Amelogenesis imperfecta
  - Associated with syndromes
Diagnosis

- Does any one else affected in the family?
- Are all teeth affected in similar manner?
- Is there chronological distribution?
- Is there anything in the past medical history e.g. low birth weight, metabolic disturbances?

Barron et al., 2008
**Fluorosis**

*Questionable to very mild:* loss of translucency of enamel at the incisal and proximal edges of the labial surface

*Mild:* fine white lines
Fluorosis

**Moderate:**
very chalky, opaque enamel which fractures soon after tooth eruption

**Severe:**
mottling and loss of portions of the outer enamel, staining of the enamel
<table>
<thead>
<tr>
<th>Stage of enamel formation</th>
<th>Effect of fluoride exposure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Presecretory stage (cell proliferation and differentiation)</td>
<td>No effect at physiologically relevant fluoride doses</td>
</tr>
<tr>
<td>Secretory stage (protein synthesis and secretion, early mineralization)</td>
<td>No effect on protein synthesis</td>
</tr>
<tr>
<td></td>
<td>Inhibited protein secretion at high F levels</td>
</tr>
<tr>
<td></td>
<td>Increased fluoride in matrix</td>
</tr>
<tr>
<td>Maturation (matrix removal and mineral deposition)</td>
<td>Altered rate of ameloblast modulation</td>
</tr>
<tr>
<td></td>
<td>Fewer bands of modulating ameloblasts</td>
</tr>
<tr>
<td></td>
<td>Decreased height of enamel organ</td>
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<tr>
<td></td>
<td>Slower removal of amelogenin proteins</td>
</tr>
<tr>
<td></td>
<td>Increased fluoride and magnesium in enamel</td>
</tr>
</tbody>
</table>
### Enamel opacities

Percentage of 8 and 15 yr old children with labial surfaces of maxillary incisors affected by enamel opacities/hypoplasia (DDE Index)

<table>
<thead>
<tr>
<th></th>
<th>8 year olds</th>
<th>15 year olds</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Nil</td>
<td>Opacities</td>
</tr>
<tr>
<td>No Fluoride</td>
<td>71%</td>
<td>28%</td>
</tr>
<tr>
<td>Full Fluoride</td>
<td>67%</td>
<td>31%</td>
</tr>
</tbody>
</table>

*Childrens Dental Health in Ireland 1984*
### Table 2. Planned clinical examinations for each age group.

<table>
<thead>
<tr>
<th></th>
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<th></th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Height and Weight</td>
<td>Height and Weight</td>
<td>Height and Weight</td>
<td>DDE</td>
</tr>
<tr>
<td>2</td>
<td>Dental caries</td>
<td>DDE</td>
<td>Self-perception of enamel opacities</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td></td>
<td>Dean's Index</td>
<td>DDE</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>MIH</td>
<td>Dean's Index</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Oral photographs</td>
<td>Oral photographs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>Dental caries</td>
<td>IOTN—aesthetic component</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td></td>
<td>Dental caries</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### Table 3. Questions to be asked for the measurement of self-perceived enamel opacities in 12 year old participants.

<table>
<thead>
<tr>
<th>Question</th>
<th>Response (entered into the clinical record)</th>
</tr>
</thead>
<tbody>
<tr>
<td>“Do you have any white marks on your front teeth that won’t brush off?”</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Don’t know</td>
</tr>
<tr>
<td>For those who say “Yes”, the examiner then asks: “Does the appearance of these white marks bother you?”</td>
<td>Yes</td>
</tr>
<tr>
<td></td>
<td>No</td>
</tr>
<tr>
<td></td>
<td>Don’t know</td>
</tr>
<tr>
<td>The examiner shows three sets of photographs showing groups of teeth with varying types of appearance.</td>
<td>Photograph set N</td>
</tr>
<tr>
<td></td>
<td>Photograph set S</td>
</tr>
<tr>
<td></td>
<td>Photograph set A</td>
</tr>
<tr>
<td>All children are then asked “Thinking about white marks on teeth, do you think your front teeth look more like those in this group, or the ones in this group, or this group?”</td>
<td>Don’t know</td>
</tr>
</tbody>
</table>
Amelogenesis Imperfecta

- A hereditary developmental disorder which affect the structure and clinical appearance of enamel in both dentitions, affecting all or nearly all the teeth in a more or less equal manner (Crawford et al, 2007).

- Prevalence rates: 1.4:1000 to 1:16 000 depending on the population studied (Seow et al. 2014).

- Mutations in genes encoding enamel matrix proteins

- Can be part of a syndrome e.g. Tricho-dento osseous syndrome

- Variable expression can result in substantial or subtle differences between different affected individuals in the same family
## Witkop Classification (1988)

<table>
<thead>
<tr>
<th>Type 1</th>
<th>Type 2 Hypomaturation</th>
<th>Type 3 Hypocalcified</th>
<th>Type 4 Hypomutation-Hypoplastic with Taurodontism</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypoplastic</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>IA: Hypoplastic, pitted AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IB: Hypoplastic, local, AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IC: Hypoplastic, local, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ID: Hypoplastic, smooth, AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IE: Hypoplastic, smooth, X-linked D.</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IF: Hypoplastic rough, AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IG: Enamel agenesis, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>IG: Generalised thin hypoplastic, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2A: Hypomutation, pigmented, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2A: Pigmented hypomutation, AR</td>
<td></td>
<td></td>
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<tr>
<td>2B: Hypomutation, X-linked recessive.</td>
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<tr>
<td>2C: Hypomutation, snow capped teeth, X-linked</td>
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<tr>
<td>2D: Hypomutation, snow-capped teeth, AD</td>
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<tr>
<td>3A, AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3B, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3B Hypocalcified, AR</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4A: Hypomutation-hypoplastic with taurodontism, AD</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4B: Hypoplastic-hypomutation with Taurodontism, AD</td>
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</tbody>
</table>

Modified by Nusier et al. 2004 (underlined)
Amelogenesis imperfecta

- Hypoplasia
  - quantitative defect, reduced thickness
  - pitting, surface loss
Hypoplastic (Secretory defect)

Radiographically enamel contrasts normally to dentin
Hypocalcified (Mineralization Defect)
Hypomaturation
(Protein processing and crystallite maturation defect)
Psychosocial implications

- Ideal smile
- Neglect
- Lazy
- Dissatisfaction
- Age dependent
What about the future?......
Parental Perception of Children Affected by AI and DI: A Qualitative Study

Purpose: To investigate and explore parental attitudes and values regarding aesthetics and treatment need of children in primary dentition affected by Amelogenesis and Dentinogenesis Imperfecta.

6 main themes:
- the impact on affected children,
- the impact on parents,
- the life course of the disease,
- coping mechanisms,
- treatment need
- experience of treatment.

Parents expressed 4 key emotions about having an affected child;
- sadness,
- fear of bullying and negative social reactions,
- worry about associated impact on the child
- feelings of guilt.

• Conclusions: Parents of children affected with dental anomalies experience significant emotional and psychological challenges that must be acknowledged by the treating dentist.
Affected parents own experience with bullying

Childs experience with bullying/teasing in school

Childs self consciousness

Societal perception

Childs character and personality

- Fear of bullying.
- Fears of child feeling different.
Parents rely on their trust in and expertise of the dentist when making decisions for their children.

AlQuadi and O’Connell
General Management Principles

• Driven by patient needs
  • Age dependent
  • Good communication

• Maintain existing dentition
  • Primary care very important
  • Monitor development

• Long term (flexible) plan
  • Keep options open
Associated Dental Anomalies

- Pulp calcification.
- Taurodontism.
- Delayed /accelerated eruption
- Gingival overgrowth
- Skeletal anterior open bite (50% in hypoplastic AI, 31% in hypomaturation AI, and 60% of hypocalcified AI.)
- Increased susceptibility to calculus deposition.
- Root and crown resorption (PEB after or before eruption)
- Enlarged follicles, impacted permanent teeth, ectopic eruption, congenitally missing teeth.

Bailleul-Forestier et al., 2008
Crawford, 2007
Management

- Early diagnosis and preventive care
- Careful treatment planning
  - Aesthetics
  - Function
  - Sensitivity
  - Stabilisation
- Life long extensive restorative care
Enamel defects

Problems:
- Diagnosis
- Sensitivity
- Caries risk
- Poor aesthetics
- Poor OH
- Delayed eruption
- Anterior open bite
Treatment options

• Vital Bleaching
• Macroabrasion
• Microabrasion
• Composite resin veneers  direct/indirect
• Full coverage
  – Stainless steel crown / Cast crown
  – Lab processed composite
  – All ceramic restorations
  – Metal ceramic crowns
No randomised controlled trials of restorative treatments for children and adolescents with AI, and therefore there is no evidence as to which is the best restoration.

THANK YOU

Anne O’Connell