Mouth and dental problems:

Experience in Chile

Dra. Susanne Krämer - Debra Chile
Mechelen, 25 October 2008
Population: 16,763,470
GDP per capita: 14,400 USD

Debra Chile:
• Provide treatment
• Education
• Research
EB in Chile

162 Diagnosed cases in Chile

- 85 EBS
- 9 JEB
- 68 DEB

Dental treatment in Chile

- Dental Health Care provided by the national health system:
  - 6 year old
  - 60 years old
  - Cleft lip and palate
  - Pregnant women
  - Emergency dental treatment
Mouth and dental problems

1. Oral Manifestations
2. Oral Health related Quality of Life
3. Today: Treatment protocol
4. Research
1. Oral Manifestations
May present with episodes of oral ulceration 2 – 3 times monthly. Heal without scarring.

One patients with EBS (K5:A22G) and with Generalized Hypoplasia, main feature of JEB.
Limited Mouth Opening

Hypergranulation tissue
Amelogenesis Imperfecta
(Generalized Hypoplasia)

100% patients with JEB present Amelogenesis Imperfecta, which can differ between hypoplastic, hypomature or hypocalsified type.
Bullae:

Number of affected sites inside the mouth

<table>
<thead>
<tr>
<th>Group</th>
<th>EBS</th>
<th>EBJ</th>
<th>EBD-HS</th>
<th>EBD-no-HS</th>
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Affected sites:
Microstomia (Limited Mouth Opening)

Progressive

0-6 years (n=6) : 24.2mm
7-12 years (n=6): 22.1mm
> 13 years (n=6): 20.2mm

Normal mouth opening
(45 - 55mm)
Ankyloglosia

Milímetros en que la lengua sobrepasa el borde incisal

<table>
<thead>
<tr>
<th></th>
<th>EBS</th>
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<th>EBD-no HS</th>
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Loss of the vestibular depth
Enamel abnormalities
Caries: DMF index

<table>
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<th>Age</th>
<th>P</th>
<th>O</th>
<th>C</th>
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<td>0-6 años</td>
<td>1.01</td>
<td>1.18</td>
<td>9.01</td>
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<tr>
<td>7-12 años</td>
<td>0.16</td>
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<td>&gt;13 años</td>
<td>3.33</td>
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<td>18.50</td>
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</table>

DMF Index in DEB

- M (Blue)
- F (Red)
- D (Green)
2. Oral Health related Quality of Life
Main oral health problems?
Oral Manifestations

Quality of Life

Today

**EBS:**
- They were “Sometimes” concerned about problems with teeth and gums.
- “Seldom” they had troubles biting or chewing.

**JEB:**
- They were “always” concerned about problems with their teeth and gums.
- “Sometimes” they had troubles biting or chewing.

**Severe generalized RDEB :**
- “Often” they had troubles biting or chewing.
- “Seldom” they were able to swallow comfortably
- “Seldom” they were able to eat without feeling discomfort.

**Other DEB subtypes:**
- “Often” they had troubles biting or chewing.
- “Sometimes” they were concerned about problems with teeth and gums.
3. Today:
Dental Treatment Protocol
DebRA Chile

Goal:
Prevention

2. Dental Clinic
   ➢ Emergency.
   ➢ All treatment starting with anterior segment.

3. General Anaesthesia
   ➢ Extractions
   ➢ More complex treatment

4. Dental Clinic
   ➢ Fillings and crowns, according to the treatment provided under GA.

5. Review
   ➢ Review every 3 months.
Early Referral!

1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review

Education
Education - Prevention

- **Tooth brush:**
  - Small head.
  - Short and soft bristle.
  - Handle easy to grab.

More preventive measures:

- High dose fluoride tooth paste.
- Fluoride mouth wash.
- Chlorhexidine mouth wash.
Exercises:

Mouth opening and tongue movement.
Some aspects dentists, patients and parents need to be aware of:

Start working on the anterior segment, to give time to increase mouth opening.

Explain to the patient and carer that during the dental treatment some ulcers can appear.

Suction tip should be leaned against tooth surface. If the suction tip touched the mucosa it can produce an ulcer.
Prevention: Sealants and Fluoride

1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review
1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review

Endodontics
Rehabilitation
Fillings
Surgery
Crowns

Periodontal treatment

1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review
Pulpotomies y multiple extractions

1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review
Oral Rehabilitation

1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review
1. Oral Examination
   Education
   Treatment plan

2. Dental Clinic

3. General Anaesthesia

4. Dental Clinic

5. Review

Review every 3 months
4. Research - Dentistry
Scientific literature today

• Only 2 case series published
  (1991 [NEBR], 2003 [C. Serrano])
• Most literature available:
  case reports
• Actual interest:

Future

• Therapeutic Approaches for Increasing Mouth opening in severe generalized RDEB.
Therapeutic Approaches for Increasing Mouth opening in severe generalized RDEB

Randomized controlled trial in clinical phase.
Results expected for September 2009.