Cleft Lip & Palate
ENT & Phoniatriy Evaluation

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Plan

• Velopharynx
  – Physiology
  – Anatomy
• Cleft palate
• ENT and Phoniatry Evaluation
• Conclusion
ANATOMY & PHYSIOLOGY REVIEW
Velopharyngeal closure

1. Raising +
2. Stretching of soft palate

3. Concentrical contraction of pharyngeal wall
Velopharynx - Functions

- **Suction**
  - Negative intra-oral pressure
    - Efficacity of orbicular muscles
    - velopharyngeal closure

- **Swallowing**
  - Propulsion
  - velopharyngeal closure

- **Phonation**
  - velopharyngeal closure

- **Speech**
  - Articulation of consonants

- **Audition**
  - Opening of Eustachian tube
ANATOMY
Tensor Veli Palatini

- **Superficial layer:**
  - Eustachian Tube → median raphe
  - ✓ Tension

- **Deep layer:**
  - Eustachian Tube → pteryboidal hamulus
  - ✓ Opening of Eustachian Tube

- **Innervation:** V3
Levator veli palatini

• Eustachian Tube → median raphe
  ✓ Elevation
  ✓ Opening of Eustachian tube
M. Uvulae

- Elevation and shortening of Uvula
- Pharyngeal Lubrification (*Back*, 2004)
- Immunity
Palatoglossus

- Soft palate aponevrosis
  → tongue = anterior pilar

✓ Elevation of the base the tongue
→ pharyngeal narrowing
Palatopharyngeus

1. Soft palate
2. Pterygoidal hamulus
3. Eustachian Tube

→

1. Thyroid cartilage
2. pharynx (meet contolateral fibers)
= posterior pilar
→ pharyngeal narrowing
Superior Constrictor

- Pharyngeal narrowing
- → Passavant’s pad
  - 30% normal subjects
  - Velopharyngeal compensation
Passavant’s pad
CLEFT LIP & PALATE
Embryology - Lip

Images adapted from "The developing Human" 2nd Ed. – W.B. Saunders.
Embryology - Lip

Images adapted from "The developing Human" 2nd Ed. – W.B. Saunders.
Embryologie – Palate

6 semaines

9 semaines

12 semaines

Images adapted from ”The developing Human” 2nd Ed. – W.B. Saunders.
Embryology - Palate

6 semaines

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12 semaines

Images adapted from “The developing Human” 2nd Ed. – W.B. Saunders.
Cleft Lip and Palate

Figure 3:
A. Normal
B. Cleft lip and alveolus
C. Cleft lip and primary palate
D. Unilateral cleft lip and palate
E. Bilateral cleft lip and primary palate
F. Bilateral cleft lip and palate
G. Cleft palate only

^3
Submucosal Cleft

Type I - classical
- Uvula bifida
- Midline pit
- Paramedial pad
- Rare VPI
- Cave: adenoidectomy

Type II - occult
- Normal aspect of soft palate
- Muscle hypoplasia,
- Abnormal muscle insertion or anastomosis
- Uvulae m. may be absent
- Short soft palate
- $\rightarrow$ frequent VPI
Epidemiology

- 1 / 700
- M:F = 2:1
- G:D = 3:1
- 50% lip and palate
- 50% soft palate → M:F = 1:2
Etiology

• 90% non syndromic
  – Genetic
  – Environmental
    • Teratogens (Thalidomide, Roaccutane, Phenytoine,...)
    • Cigarette: risk x 2
    • Alcohol
    • Folate?

• 10% syndromic
Chromosomal
   Trisomy 13
   Trisomy 18
   Velocardiofacial syndrome (22q11 deletion)

Non-Mendelian
   Pierre Robin sequence
   CHARGE association
   Goldenhar syndrome

Mendelian disorders
   Ectrodactyly-ectodermal dysplasia-clefting syndrome (AD)
   Gorlin syndrome (AD)
   Oto-palato-digital syndrome (XL)
   Oral-facial-digital syndrome (XL)
   Smith-Lemli-Opitz syndrome (AR)
   Stickler syndrome (AD)
   Treacher Collins syndrome (AD)
   Van der Woude syndrome (AD)

Unknown
   de Lange syndrome
   Kabuki syndrome

Teratogenic
   Fetal alcohol syndrome
   Fetal phenytoin syndrome
   Fetal valproate syndrome

AD = autosomal dominant.
AR = autosomal recessive.
XL = X-linked inheritance

Hodgkinson et al. 2005
Velocardial Facial Syndrome

- 22q11 deletion
- 1/3-4’000
- Cleft palate
  - “real” (38%)
  - Submucousal (62%)
- Cardiac malformation (85%)
- Thymus aplasia (31%)
- Multiple malformations (medial carotids)
- Delayed development, speech
- Schizophrenia (30%)
Management

• Multidisciplinary
  – Pediatric Surgery
  – Orthodontist
  – ENT - Phoniatry
  – Logopedy
  – Pediatrics
  – Audiologist
  – Psychologist
  – Genetician
ENT PHONIATRY EVALUATION
Why?

- Suction
- Swallowing
- Phonation
- Articulation
- Language
- Hearing
Suction-Swollowing

- Shorter suctions, less effective, increased intra-oral pressure *(Reid et al, 2007)*
- Evaluation: at birth
- Breast / Bottle feeding observation
  - *Fausses routes*
  - *Jetage nasal*
  - Drooling
  - Feeding duration
Suction/Swallowing

- Breast feeding is not a contra-indication (on the contrary)
- Habermann bottle / larger teat
- Vertical
- Palatal plate not mandatory
- Thicken up
Rhinolalia

**Hypernasality**

Open Rhinolalia

- Velopharyngeal insufficiency during phonation
- → oral phonems

**Hyponasality**

Closed Rhinolalia

- Nasal / Nasopharyngeal obstruction during phonation
- → nasal phonems

**Mixed Rhinolalia**

- Open + Closed
- VPI + nasal/nasopharyngeal obstruction during phonation
- → all phonems
Hypernasality

• Nasal puff
  – Audible nasal turbulence
  – Before occlusive

• Nasal snoring
  – Mucusal or adenoids vibration during phonation
Hypernasality - Evaluation

• Gutzmann test
  – /a:/-/i:/ with and without closing the nose
    • Inchanged: normal
    • Changed: hypernasality

• Glatzel’s mirror test
  – Vizualisation of the nasal air deperdition
    • /a:, papapapapa

• Stethoscop’s test
Perceptive evaluation

- Spontaneous speech
- Denomination: from 3 y
- Standardized text
- Scale? No consensus
  - 5 grade scale *(Hirschberg, 1997)*
  - 1-10
  - Borel-Maisonny scale
• http://www.acpap-cpf.org/education/educational_resources/speech_samples/
Borel-Maisonny scale

- **Phonation I**
  - Normal

- **Phonation II**
  - Hypernasality
    - PhIIb: normal intelligibility
    - PhIIIm: lack of intelligibility

- **Phonation III**
  - Phonemic substitution, compensation (*coup de glotte, souffle rauque*)

- **Phonation I/II**
  - almost normal, rare hypernasality

- **Phonation II/I**
  - Constant nasal deperdition
Instrumental - Nasometry

- Nasalance

\[
\frac{(\text{nasal acoustic energy dB})}{\text{(oral acoustic energy dB)}} \times 100
\]

- Vowels, logatoms, words, sentences, text
- Follow-up
- Reeducation
- No normative values in French but in Bärndütcsh
Aerophonometry

• Nasal flow measure even not audible
Nasofibroscopy

- Submucosal cleft
- Adenoids
- Velopharyngeal closure
## Velopharyngeal closure

<table>
<thead>
<tr>
<th>Repos</th>
<th>Fermeture partielle</th>
<th>Fermeture complète</th>
<th>Type de fermeture</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image1.png" alt="Image" /></td>
<td><img src="image2.png" alt="Image" /></td>
<td><img src="image3.png" alt="Image" /></td>
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<tr>
<td><img src="image4.png" alt="Image" /></td>
<td><img src="image5.png" alt="Image" /></td>
<td><img src="image6.png" alt="Image" /></td>
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<td><img src="image7.png" alt="Image" /></td>
<td><img src="image8.png" alt="Image" /></td>
<td><img src="image9.png" alt="Image" /></td>
<td>Circulaire</td>
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<td><img src="image10.png" alt="Image" /></td>
<td><img src="image11.png" alt="Image" /></td>
<td><img src="image12.png" alt="Image" /></td>
<td>Circulaire avec Passavant</td>
</tr>
<tr>
<td><img src="image13.png" alt="Image" /></td>
<td><img src="image14.png" alt="Image" /></td>
<td><img src="image15.png" alt="Image" /></td>
<td>Sagittal</td>
</tr>
</tbody>
</table>

The sphincteric mechanism of velopharyngeal closure. **Skolnick ML, McCall GN, Barnes M. Cleft Palate J. 1973 Jul;10:286-305**
Articulation

• Phonological troubles
  – /p/, /b/, /m/ → lip occlusion
  – /p/, /t/, /k/ → velopharyngeal closure
  – /f/, /v/ → lip + VP closure

• Compensation mechanisms

• coup de glotte, souffle rauque

• Syncinesy
Articulation

• Logopedic evaluation
  – Spontaneous speech
  – Articulation of all phonems
  – Image denomination

• Validated test and scores
Speech Therapy

• 2 air passage
• Breath exercises
  – Balloon, candle
• Oro-facial praxies
• Articulation exercises
Speech

• Delayed canonical babbling (n=5-10 m)
• Babbling less complex
• Delayed first words (Chapman, 2001; Chapman, Hardin-Jones, 2003; Scherer, 2008)

• Evaluation
  – Spontaneous speech
  – Simple orders
  – Parental questionnaire
  – Valided test
Radiology

- Videofluoroscopy
- MRI
- CT-scan

- Cave 22q11: carotid ectopy
Otology & Audiometry

- Increased risk of glue ear
- Perceptive deafness in syndroms
- → otoscopy
- →±tympanometry
- → OAE ± PEA
- Audiogramm from 3
Surgery

• T-Tube
• Adénoidectomy
  – CAVE: risk of VPI
  – précoce
• Tonsillectomy
  – No VPI
  – CAVE: scars in posterior pilar
VPI Surgery

- Fat autologous graft in nasopharynx
VPI Surgery

- Posterior pharyngeal flap
- Pharyngeal sphincteroplasty
- Palatoplasty
Calendar

• Neonatal
  – Suction + Swallowing
  – Audiometry (OAE, EAP)

• 12 mois
  – Speech & Language: babillage
  – Otoscopy ± Audiometry

• 18 mois
  – Nasality: perceptive evaluation
  – Speech & Language: 1-2 words
  – Otoscopie ± Audiometry

• 3, 5, 7, 12, 15, 19 ans
  – Nasality: perceptive evaluation + nasometry
  – Speech & Language: intelligibility
  – Otoscopie ± Audiometry

• Nasofibroscopy