Early detection and aggressive management of otologic pathologies in children with Down syndrome (DS)

Jessica Hoffner Leventhal, MSN, FNP-BC, CLC

Disclosures

- None

Learning Objectives

- To be able to proactively detect and manage otologic pathologies in children with Down syndrome (DS)

Children with Down syndrome (DS)

- Most common chromosomal abnormality among live born infants (U.S.)
- Many otolaryngological problems:
  - Increased incidence of ear infections and potential for hearing loss
  - Obstructive sleep apnea and subglottic stenosis
  - Chronic rhinorrhea and sinusitis
- Highly susceptible to otitis media with effusion (OME) and present particular challenges:
  - Younger age of onset
  - Prolonged course
  - Greater risk of complications
  - Potential diagnostic difficulties
- Lack of evidence-based literature related to medical and surgical care

Terms, Abbreviations, Definitions

<table>
<thead>
<tr>
<th>Middle ear effusion</th>
<th>MEE</th>
<th>Fluid in middle ear from any cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Otitis media with effusion</td>
<td>OME</td>
<td>Fluid in middle ear</td>
</tr>
<tr>
<td>Chronic otitis media with effusion</td>
<td>Chronic OME</td>
<td>OME for 2-3 months from: date of onset (if known) or date of diagnosis (if onset is unknown)</td>
</tr>
<tr>
<td>Acute otitis media</td>
<td>Acute OME</td>
<td>Rapid onset symptoms of middle ear inflammation</td>
</tr>
<tr>
<td>Conductive hearing loss</td>
<td>CHL</td>
<td>Abnormal/reduced sound transmission to inner ear. Often assoc. with MEE/middle ear abnormalities</td>
</tr>
<tr>
<td>Sensorineural hearing loss</td>
<td>SNHL</td>
<td>Abnormal sound transmission from sensory cells of inner ear to brain</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Otitis media with effusion</th>
<th>OME</th>
<th>Fluid in middle ear</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic otitis media with effusion</td>
<td>Chronic OME</td>
<td>OME for 2-3 months from: date of onset (if known) or date of diagnosis (if onset is unknown)</td>
</tr>
<tr>
<td>Acute otitis media</td>
<td>Acute OME</td>
<td>Rapid onset symptoms of middle ear inflammation</td>
</tr>
<tr>
<td>Conductive hearing loss</td>
<td>CHL</td>
<td>Abnormal/reduced sound transmission to inner ear. Often assoc. with MEE/middle ear abnormalities</td>
</tr>
<tr>
<td>Sensorineural hearing loss</td>
<td>SNHL</td>
<td>Abnormal sound transmission from sensory cells of inner ear to brain</td>
</tr>
</tbody>
</table>
Predisposition to ENT disease in DS

- Dysmorphic facial features:
  - Midface hypoplasia
  - Narrow nasopharynx
  - Adenoids close to ET opening
  - Abnormal insertion of ET in nasopharynx
- Eustachian tube (ET) malformation:
  - Increased cartilage cell density within ET
- External ear stenosis (cerumen impactions)
- Muscular hypotonia (ET collapse)
- Decreased immune status
- Role of the PCP: recommend preventive measures for ET dysfunction and otitis media
  - Encourage breastfeeding
  - Vaccinate (pneumococcal and flu)
  - Counsel regarding tobacco smoke exposure

Speech and Language

- Compared with other children, children with DS have a significantly greater incidence of speech and language deficits
  - Cognitive delay alone cannot fully explain this; good hearing is essential
- Frequent/persistent MEE diminishes tympanic membrane (TM) mobility and can impede sound conduction
  - Complicates speech recognition, processing, noise perception and sound localization
- MEE can cause a shifting, mild to moderate hearing loss
  - Inconsistent hearing difficult to learn how sounds and actions are related
- Role of the PCP: screen for communication difficulties

Screening for OME and hearing loss

- When a child is diagnosed as being at-risk and between the ages of 12 and 18 months:
  - Ideal surveillance: every 3 to 6 months
- Role of the PCP: discuss with caregivers that the following should prompt evaluation for OME and hearing loss:
  - Behavior changes
  - Worsening balance/coordination
  - Decreased attention spans

Evaluation for OME

- Use pneumatic otoscopy or obtain tympanometry if OME is suspected but the dx is uncertain after pneumatic otoscopy
  - Pneumatic Otoscopy: otoscope with an attached rubber bulb that changes pressure in ear canal to observe how eardrum reacts
    - Normal eardrum: moves briskly with applied pressure
    - Fluid in the middle ear: minimal or sluggish movement
  - Tympanometry: small probe is placed in ear canal; assesses TM mobility and middle ear function by measuring the reflected sound energy

Evaluation for hearing loss

- Newborn hearing screen by 1-month of age
  - For those who fail comprehensive audiological evaluation by 3 months
  - Every 6 months up to 3 years of age
  - Annual after 3 years of age
- Other situations:
  - OME of any duration
  - Any parent/enthusiasm concern about hearing loss
- Abnormal screen or unable to obtain accurate results refer to ENT and audiology

Management

- Treatment of underlying infection with antibiotics
- Multidisciplinary care
  - PCPs, otolaryngologists, audiologists, speech therapists, and specialized teachers
- Tympanostomy tubes
- Hearing aids
- Early intervention (EI)
Management: Tympanostomy tubes

- In children with DS:
  - Significantly improve hearing
  - Reduce effusion prevalence
  - May reduce the incidence of recurrent AOM
  - Allow for drainage and topical antibiotic administration
- Risks are related to general anesthesia and the effect of the tube on the TM and middle ear
- Role of the PCP:
  - Be familiar with the evidence:
    - Limited evidence regarding the impact on at-risk children with OME
    - Variable results: hearing outcomes, surgical complications and need for reoperation
  - Know the recommendations:
    - Can use tubes in at-risk children with unilateral/bilateral OME that is unlikely to resolve quickly
      
      [AAO-HNSF](https://www.entnet.org)  

Management: Hearing aids

- May be preferred to tympanostomy tubes if:
  - Eardrum damage from persistent OME and repeated tube insertion
  - Ear canals are too narrow for tube insertion
- Should be considered even in mild hearing loss to avoid educational, emotional, and language delays
- In-school amplification devices such as frequency modulation (FM) sound field systems in classrooms
- Role of the PCP:
  - Discuss preferential seating: move the child to the front of the class, with the better-hearing ear directed toward the instructor

Management: Early Intervention (EI)

- A program of therapy, exercises, and activities designed to address developmental delay
- Should begin shortly after birth and continue until age 3
- Federal law requires that states provide EI services for all children who qualify
- Role of the PCP:
  - Refer to EI
  - Newborn visit: discuss the efficacy of EI and availability of services in the community
  - 30-month visit: discuss the transition from EI to preschool, which occurs at 36 months of age

Challenges

- Diagnostic Challenges:
  - Difficult otoscopic examination if ear canals are stenotic
  - Poor reliability of behavioral hearing testing
  - Lack of child cooperation for cerumen removal and tympanometry
  - Uncertain validity of tympanometry in the setting of middle ear disorders
- Provider Burden:
  - Remembering to screen and evaluate at-risk children
  - Proficiency with pneumatic otoscopy & tympanometry
  - Ensuring compliance with follow-up

Challenges

- Lack of Self-Reporting:
  - Children with DS may lack the communication skills or sensory perception to reliably express discomfort associated with acute otitis media (AOM) or complain of diminished hearing
- Parent/Caregiver Challenges:
  - Access barriers to specialists and appropriate services
  - Poor compliance related to a lack of understanding
  - Frustration:
    - Multiple office visits, tests, and referrals to a number of specialists
    - The condition may be refractory to multiple treatment attempts

Conclusion

- Children with DS:
  - Have many ENT problems that may prevent them from reaching their full developmental potential
  - Present distinct challenges of assessment and management
- Early detection and aggressive treatment is essential for physical, emotional, social, language, and educational development
- Multidisciplinary care will help ensure the best possible outcomes
To Clip or Not to Clip: An Update on Pediatric Tongue Tie

Laurie Newton DNP, RN, CPNP

Objectives

- Describe oral restrictions that may lead to feeding and speech difficulties
- Demonstrate procedure for assessing oral restrictions
- Discuss other factors that contribute to feeding and speech difficulties
- Understand patient selection for frenotomy and the technique of frenotomy

Background

In 2017:

- No high-quality evidence on best practices
- Global problem, not just local, regional, or national
- Endless sources of information
- Peer circles
- Social media
- Varying professional opinions and preferences
- How to “navigate the fog” of information to reach best choice for a particular child?

Anatomy

Lingual frenulum
- Small fold of mucous membrane connecting underside of tongue to floor or mouth
- Effects movement of the tongue

Labial frenulum
- Collagenous fibrous tissue fold of mucous membrane connecting lip to alveolar process

Feeding Difficulties of Healthy Term Infants
Ankyloglossia (tongue-tie)

- Short, thickened, altered attachment of lingual frenulum (Messner & Lakakea, 2002; Yoon et al, 2016)
- Varying degrees of reduced tongue mobility
- Functional limitations
  - sucking, atypical swallowing, speech articulation, mechanical problems during oral phase, psychosocial stress
- Types:
  - Anterior
  - Posterior

Maxillary labial lip-tie

- Vertical band of lip tissue extending from inside portion of upper lip attaching to alveolar mucosa of maxillary arch (Kotlow, 2011)
- Varying degrees of reduced upper lip mobility
- Functional limitations of
  - proper lip flange for seal,
  - maternal pain,
  - milk retention leading to dental caries

Epidemiology

- Widely varied incidence: 0.1 to 12.8% (Martinelli at al, 2013; Shay et al., 2016)
- ~3:1 male:female ratio (Steehler et al., 2012)
- Race and ethnicity not predisposing factors
- Positive family history in 10-53% of patients (TBX22 gene)
- 3.2 times higher among children exposed to cocaine in utero

Trends in Ankyloglossia

- 834% increase in reported diagnosis
- 866% increase in reported frenotomies
- Pediatric discharges and births remained relatively stable

- Potential explanations:
  - National and global efforts to support breastfeeding
  - Increased awareness of the diagnosis of ankyloglossia and its role in successful breastfeeding
  - Earlier diagnosis and treatment (before first discharge?)
  - Higher incidence in “Private Insurance” patients and patients from higher median income zip codes
  - Increased access to breastfeeding support?
ENT Practice Patterns

ENT referral: infants
- Infants with feeding difficulties
- Painful latch
- Poor weight gain

ENT referral: toddlers and older
- Articulation difficulties: l, r, t, d, n, th, sh, and z
- Length of lingual frenulum
- Amount of tongue movement
  - Difficulty lifting tongue to upper dental alveolus
  - Inability to protrude tongue more than 1-2 mm past lower central incisors
  - Impaired lateral movement of tongue
- Heart-shaped appearance to the tongue tip
- Thick, fibrous cord palpated on physical examination

ENT: Assessment
- Tongue range of motion ratio
  - Grade 1: >80%
  - Grade 2: 50-80%
  - Grade 3: 50%
  - Grade 4: <25%

 Lingual frenotomy technique
- Office: Patient selection: lingual frenotomy up to age 6 months in the office
  - Anesthesia
  - Topical viscous lidocaine on a swab
  - Sweet-ease
- Operating room: Patient selection: thick frenulum, posterior tongue tie, older children
  - Procedure
    - Injected lidocaine
    - Bovie cautery/sharp dissection
    - Suture placement
Issues around tongue tie

- Timing of procedure
- Post-procedure intervention
- Reattachment/Revision
- "Bubble" palate (high-arched) and changes in palate post-op
- Paced feeding
- Poor tongue mobility post-op
  - Learned or different concern

Considerations for posterior tongue tie intervention

- Consequence of skin or mucosal incision:
  - Collagen deposition, granulation tissue formation, epithelialization, wound contraction
  - Potential for less flexible healed connective tissue
  - Increased pain and post-procedure intervention resulting in subtle oral defensiveness

ENT referral

Maxillary labial restriction

- Referral considerations in infants:
  - Infants with feeding difficulties
  - Painful latch
  - Poor weight gain
- Referral consideration in toddlers and older:
  - Impact on dentition, causing splaying of the upper central incisors

ENT: Assessment

Length of maxillary frenulum, site of insertion, tethering of the upper lip

<table>
<thead>
<tr>
<th>Maxillary Frenulum</th>
<th>Definition (Lottke, 2010)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class 1</td>
<td>Little or no tissue attachment of the lip to the gingival tissue</td>
</tr>
<tr>
<td>Class 2</td>
<td>Frenulum attaches above or at the border between free and attached gingival tissue</td>
</tr>
<tr>
<td>Class 3*</td>
<td>Frenulum attaches at or into the interproximal area between the upper central incisor</td>
</tr>
<tr>
<td>Class 4*</td>
<td>Frenulum attaches into the palatal tissue</td>
</tr>
</tbody>
</table>

*May seriously compromise breastfeeding
Maxillary labial frenotomy

OPERATING ROOM:
Local infiltration with lidocaine
Reflect lip upwards with gauze
Incise frenulum with needle tip bovie.
Approximate mucosa with suture to prevent stricture.

Treatment of Ankyloglossia and Breastfeeding Outcomes: A Systematic Review

Pediatrics 2015
• Review of 29 studies of impact of frenotomy and breastfeeding
• Limited evidence for benefit of frenotomy
• Small studies
• Short-term follow-up
• No studies addressed nonsurgical interventions

Other Considerations

Team Approach:
• Collaborative approach between Speech Pathology, Lactation Consultant, and ENT
• Simultaneous evaluation when possible
• Referral is bi-directional
• Recognition that anatomy may not be only contributing factor in infants with feeding disorders

References

Expanding Access to Oral Health Care through an Interprofessional Educational and Clinical Training Program

Karen G. Duderstadt, PhD, RN, CPNP, FAAN
Abbey Alkon, PhD, RN, CPNP, FAAN
UCSF School of Nursing
Brent Lin DDS
JungSoo Kim, Dental Fellow
UCSF School of Dentistry

Learning Objectives
• Describe an Interprofessional oral health educational and clinical training program
• Increase participants knowledge and training strategies to improve oral health outcomes in underserved children living in low-income families

Dental Caries
• Multifactorial

Burden of Pediatric Dental Disease
• Dental caries are the most prevalent chronic disease in childhood in U.S.
• Children suffer from dental caries at a rate 5 times greater than asthma in the U.S.
• 23% of children from two to five years of age have dental caries in their primary teeth
• Children living in low-income families are least likely to receive dental care (32.9%)

Acknowledgement
This clinical training program is supported by Health Resources & Services Administration (HRSA) grant #D85HP28498


Check for Early Signs of Decay: White Spots
Disparities in Oral Health Care Access

One of the primary contributing factors to disparities in children’s oral health is the lack of access to pediatric dentists

− Low number of dentists in U.S. willing to provide services to patients with Medicaid due to low reimbursement rates (42% of dentists)
− Low number of dentists reluctant to treat children with extensive dental disease

• California has more dental health professional shortage areas than any other state (29% dentists)

Interdisciplinary Practice & Education (IPE) Course

Study Aims

• Phase 1: Design and implement an interdisciplinary elective course on children’s oral health for students in dentistry, nursing, and medicine
  - Evaluate the trainees’ improvement in oral health knowledge, attitude, confidence and interest after the training.

• Phase 2: Evaluate the trainees’ employment, practice setting and time spent in oral health assessment after course completion.

Interdisciplinary Oral Health Course

Course Content:

<table>
<thead>
<tr>
<th>Class</th>
<th>Didactic Class Topics</th>
</tr>
</thead>
<tbody>
<tr>
<td>Class 1</td>
<td>Introduction to Children’s Oral Health and Community Dentistry</td>
</tr>
<tr>
<td>Class 2</td>
<td>Physical Assessment of Oral Cavity and Recognition of Abnormalities</td>
</tr>
<tr>
<td>Class 3</td>
<td>Caries Risk Assessment and Disease Prevention</td>
</tr>
<tr>
<td>Class 4</td>
<td>Infant Oral Health Care, Dental Home, and Referral</td>
</tr>
<tr>
<td>Class 5</td>
<td>Anticipatory Guidance in Pediatric Dentistry</td>
</tr>
<tr>
<td>Class 6</td>
<td>Relationship between Children’s Oral Health and the Overall Systematic Health</td>
</tr>
<tr>
<td>Class 7</td>
<td>Effect of Cultural and Linguistic Competency and Health Literacy on Access to Oral Health Care</td>
</tr>
<tr>
<td>Class 8</td>
<td>Oral Health in Special Needs and Vulnerable Children</td>
</tr>
<tr>
<td>Class 9</td>
<td>Management of Orofacial Trauma and Acute Dental Care</td>
</tr>
<tr>
<td>Class 10</td>
<td>Case Presentations</td>
</tr>
</tbody>
</table>

Interdisciplinary Oral Health Clinical Training

<table>
<thead>
<tr>
<th>Criteria for Clinical Experience Sessions</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Assessment of Oral Cavity</td>
</tr>
<tr>
<td>- Proper position of examination</td>
</tr>
<tr>
<td>- Clinical examination of oral mucosa and soft tissue, teeth, gingiva, timing and sequence of eruption, crowding/spacing, and oral hygiene</td>
</tr>
<tr>
<td>- Recognition of dental caries</td>
</tr>
<tr>
<td>2. Caries Risk Assessment and Anticipatory Guidance</td>
</tr>
<tr>
<td>- Identify dental chief complaint</td>
</tr>
<tr>
<td>- Categorize caries risk level and rationalize it</td>
</tr>
<tr>
<td>- Provide guidance to parent/child at appropriate level (incl. oral hygiene instruction and basic brushing and flossing techniques)</td>
</tr>
<tr>
<td>3. Topical Fluoride Application</td>
</tr>
<tr>
<td>- Dry teeth and apply fluoride varnish to all teeth</td>
</tr>
<tr>
<td>- Post-application instructions</td>
</tr>
<tr>
<td>4. Follow-Up Plan</td>
</tr>
<tr>
<td>- Refer for urgent dental care</td>
</tr>
<tr>
<td>- Refer to dental home for further evaluation</td>
</tr>
<tr>
<td>- No issue, low risk, and evaluate at next well-child visit</td>
</tr>
</tbody>
</table>
### Interdisciplinary Oral Health Course

- **Student demographics**
  - 25 students (80%) from UCSF School of Dentistry
  - 3 students (10%) from the UCSF School of Nursing
  - 3 students (10%) from the Touro University College of Osteopathic Medicine
  - 20 and 21 years of age (78%), female (73%), Asian (61%) and first generation college students (51%)
  - 27% of students from underrepresented groups
  - 29% were from disadvantaged backgrounds

### IPE Oral Health Course Evaluation

- **Evaluation of both didactic and clinical component**
  - Survey questions on content knowledge including oral health assessment, oral health anticipatory guidance, care of underserved populations and access to oral health care
  - Student confidence and attitude were evaluated pre and post course experience
  - Clinical practicum requirement with skill observation and checklist evaluation on skill performance and completion

### IPE Oral Health Course Outcomes

- **Oral Health knowledge showed overall improvement with significance**
  - Use of fluoride toothpaste in children ($p=0.008$)
- **Confidence showed overall improvement with significance**
  - Consulting on fluoride supplements ($p=0.004$)
  - Oral health and assessment of infants and toddlers ($p=0.004$)
  - Identification of tooth decay in early childhood ($p=0.004$)
  - Anticipatory guidance on dental visit in early childhood ($p=0.004$)

### IPE Oral Health Course Outcomes

- **Attitude showed overall improvement with significance**
  - Prescription of fluoride supplements ($p=0.028$)
- **Clinical practice showed overall improvement with significance**
  - Number of dental exams performed in past 3 months ($p=0.001$)
  - Application of fluoride varnish during routine examination ($p=0.001$)
  - Counseling children and families on routine dental visits ($p=0.001$)
  - Referral in early childhood of high-risk infants and children to dental care ($p=0.011$)

### IPE & Improving Oral Health Outcomes

#### Study Findings
- Improvement in the overall knowledge of children's oral health topics
- Confidence in their ability to provide oral health services and clinical practice application

#### Course case studies
- Demonstrated difference in approach between dental and nurse practitioner- holistic vs. clinical
- Discussions showed impact on addressing the overall disparity in children's oral health

### IPE & Improving Oral Health Outcomes

- **IPE clinical practicums provided valuable service-learning experiences for PNP students to address the national shortage of dental providers**
- **Phase II**
  - Transitioning oral health course to hybrid course format to increase IP participation
  - Increase the diversity of disciplines including medical students & increasing participation of PNP students
  - Ongoing evaluation of students and graduates integration of oral health assessment and application of fluoride varnish in practice settings
References