Wake Forest™
School of Medicine

Otologic Care of the Down Syndrome Patient

Dan Kirse, MD, FAAP
Professor
Medical Director, Pediatric Otolaryngology

1

***brief discussion about…

- ‘Down Syndrome 101’
- Recent Down Syndrome care recommendations
- Hearing loss
- Otitis media / ear tubes
- Tympanoplasty/cholesteatoma surgery
- Paucity of medical literature (variable quality)
- A little perspective from personal experience

2

Acknowledgment:

Tamison Jewett, MD

Professor, Pediatric Genetics

Down syndrome

- Occurs in ~1:650 live births
- “most common” chromosomal aberration in children
- The underlying causes:
  - 94% due to nondisjunction (unequal cell division) resulting in 47 chromosomes with an extra #21
  - 3.3% due to an unbalanced translocation
  - ~2.4% are mosaic (some cells have the typical 46 chromosomes, and some have 47)
  - <1% have a duplication of a portion of chromosome 21

Abnormal cell division leading to abnormal chromosome distribution can occur in EITHER PARENT. As women age, our risk for this to occur increases, as follows:

Maternal Age	Incidence of DS at delivery
15-29	1 in 1500
30-34	1 in 800
35-39	1 in 270
40-44	1 in 100
45 and over	1 in 50

Why do kids with DS have ear problems?

- Shape of skull base
- Abnormal cartilage in eustachian tube
- Muscular hypotonia

Health Supervision for Children and Adolescents With Down Syndrome

Marilyn J. Bull, MD, FAAP,[a] Tracy Trotter, MD, FAAP,[b] Stephanie L. Santoro, MD, FAAP,[c] Celanie Christensen, MD, MS, FAAP,[a] Randall W. Grout, MD, MS, FAAP,[d] THE COUNCIL ON GENETICS

PEDIATRICS Volume 149, number 5, May 2022:e2022057010

American Academy of Pediatrics

DEDICATED TO THE HEALTH OF ALL CHILDREN™

(previous versions in 2001 and 2011)

7

HEALTH SUPERVISION FROM BIRTH TO 1 MONTH: NEWBORN INFANTS

Congenital hearing loss, with objective testing, such as brainstem auditory evoked response or otoacoustic emission. If the infant did not pass newborn screening studies, refer to an otolaryngologist who is experienced in examining infants with stenotic external canals to determine whether a middle-ear abnormality is present. Tympanometry may be necessary if the tympanic membrane is poorly visualized.[42,43] Refer to early intervention within 48 hours of confirmation that the infant is deaf or hard of hearing.[43,44]

8

HEALTH SUPERVISION FROM 1 MONTH TO 1 YEAR: INFANCY

- ***Rescreen*** hearing at 6 months (OAE vs screening ABR?)
- Risk of OME is 50-75% is identified
- Diagnostic ABR ***should*** be obtained (clear ME spaces)
- Refer to ENT if pt has stenotic ear canals → → →
- Ear exams by ENT every 3 to 6 months
- Try behavioral audiogram at 1 year → → →
- Get ABR if not able to get behavioral audiogram

9

HEALTH SUPERVISION FROM 1 TO 5 YEARS: EARLY CHILDHOOD

- Tymps and behavioral audiogram every 6 months **until…**
- Normal hearing established bilaterally, **then…**
- Annual hearing tests
- IF cannot establish normal hearing, then OAE or ABR (age?)
- IF hearing loss per above, **then** refer to ENT

10

HEALTH SUPERVISION FROM 1 TO 5 YEARS: EARLY CHILDHOOD

- Tymps and behavioral audiogram every 6 months **until…**
- Normal hearing established bilaterally, **then…**
- Annual hearing tests
- IF cannot establish normal hearing, then OAE or ABR (age?)
- IF hearing loss per above, **then** refer to ENT

I would argue that most PCP's will not be able to navigate all of these hearing tests. IMO, recommendation for ENT referral is too late in the algorithm!

11

HEALTH SUPERVISION FROM 5 TO 12 YEARS: LATE CHILDHOOD

- Annual hearing test

12

HEALTH SUPERVISION FROM 12 TO 21 YEARS OR OLDER: ADOLESCENCE TO EARLY ADULTHOOD

- Annual hearing test

13

What about adults with Down Syndrome?

JAMA | Special Communication

Medical Care of Adults With Down Syndrome
A Clinical Guideline

Amy Y. Tsou, MD, MSc; Peter Bulova, MD; George Capone, MD; Brian Chicoine, MD; Bryn Gelaro, MA, LSW; Terry Odell Harville, MD, PhD, D(ABMLI), D(ABHI); Barry A. Martin, MD; Dennis E. McGuire, PhD, LCSW; Kent D. McKelvey, MD; Moya Peterson, PhD, APRN, FNP-BC; Carl Tyler, MD, MSc; Michael Wells, BS; Michelle Sie Whitten, MA; for the Global Down Syndrome Foundation Medical Care Guidelines for Adults with Down Syndrome Workgroup

JAMA. 2020;324(15):1543-1556. doi:10.1001/jama.2020.17024

- No mention of need for hearing surveillance

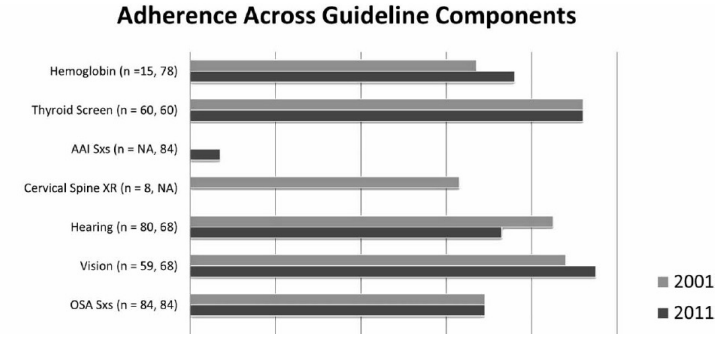
14

Evaluation of Pediatrician Adherence to the American Academy of Pediatrics Health Supervision Guidelines for Down Syndrome

Meghan E. O'Neill, Alexandra Ryan, Soyang Kwon, and Helen J. Binns

AMERICAN JOURNAL ON INTELLECTUAL AND DEVELOPMENTAL DISABILITIES
2018, Vol. 123, No. 5, 387–398

- Retrospective review looking at 4 years of care on 37 pts
- Analyzed "adherence" with 2001 and 2011 guidelines
- Finer details on "adherence'" not given
- "attending level care" in specialized care clinic at academic center



15

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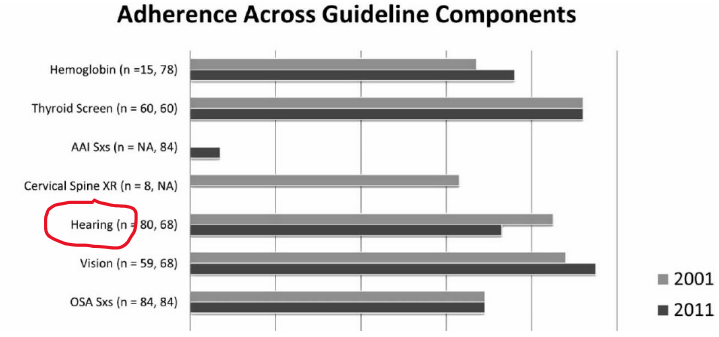

16

Hearing loss- identification

17

18

Identification of Hearing Loss in Pediatric Patients with Down Syndrome

Albert H. Park, Matt A. Wilson, Paul T. Stevens, Richard Harward and Nancy Hohler

Otolaryngology -- Head and Neck Surgery 2012 146: 135 originally published online 10 October 2011
DOI: 10.1177/0194599811425156

- 332 DS patients screened with NBHT over 5 year period
- 87 (26%) failed NBHT
  - 33 (38%) had CHL due to middle ear effusion
  - 5 had SNHL
  - 3 had mixed HL
- Average time to definitive diagnosis of hearing loss was 485 days (!!!)
- Of those who originally passed NBHT, 43% eventually needed BMT

19

Enlarged vestibular aqueducts and other inner-ear abnormalities in patients with Down syndrome

C M CLARK[1], H H PATEL[1], S G KANEKAR[2], H ISILDAK[1]

Departments of [1]Surgery, Division of Otolaryngology – Head and Neck Surgery, and [2]Radiology, Pennsylvania State University College of Medicine, Hershey, PA, USA

*The Journal of Laryngology & Otology (2017), **131**, 298–302.*

- Retrospective study of 35 pts with CT scans
- No mention of indication of need for imaging
- No correlation with audiometry data
- Not all were dedicated temporal bone CT's
- Likely selection bias (most pts likely had HL)
- 53% had inner ear abnormalities

20

TABLE I
PROPORTION OF EARS EXHIBITING EACH ANOMALY ON CT IN DOWN SYNDROME PARTICIPANTS

Anomaly	Ears affected* (*n* (%))
Inner-ear anomalies	
– Enlarged vestibular aqueduct†	7 (9.3)
– Dilated IAC‡	4 (5.3)
– Cystic malformations of LSCC	10 (13.3)
– Dilated vestibule	11 (14.7)
Mastoid air cell anomalies	
– Under-pneumatisation	14 (18.7)
– Opacification	12 (16.0)
– Sclerosis	1 (1.3)
– Fluid present	2 (2.7)
Middle-ear anomalies	
– Decreased MEC size	5 (6.7)
– MEC opacification	9 (12.0)

*Total $n = 75$. †Width of more than 1.5 mm; ‡width of more than 7.8 mm on right and 7.7 mm on left. CT = computed tomography; IAC = internal auditory canal; LSCC = lateral semicircular canal; MEC = middle-ear cavity

Clark

21

Inner ear anomalies seen on CT images in people with Down syndrome

Jarunee Intrapiromkul · Nafi Aygun · David E. Tunkel · Marco Carone · David M. Yousem

Pediatr Radiol (2012) 42:1449–1455

- 51 patients with dedicated temporal bone CT's
- 75% had inner ear abnormalities
- Malformed bone island of LSCC was most common finding
- IAC stenosis had highest correlation with SNHL

22

Table 1 Percentage and number of ears exhibiting each CT finding in the group with an SNHL component (SNHL and mixed HL) and in the group without an SNHL component (CHL and normal hearing)

	Small LSCC bone island	Cochlear nerve canal stenosis	Enlarged endolymphatic sac	Semicircular canal dehiscence	Narrow IAC
SNHL+MHL (14)	57.1% (8/14)	21.4% (3/14)	0% (0/14)	7.1% (1/14)	57.1% (8/14)
CHL+normal (28)	57.1% (16/28)	25% (7/28)	3.6% (1/28)	7.1% (2/28)	17.9% (5/28)

CHL conductive hearing loss, *MHL* mixed hearing loss, *SNHL* sensorineural hearing loss, *LSCC* lateral semicircular canal, *IAC* internal auditory canal

- Narrow IAC defined as <3.3mm at mid-canal
- Narrow IAC was the only finding that differentiated between SNHL and CHL

Tunkel

23

The Prevalence of Congenital Hearing Loss in Neonates with Down Syndrome

Adrienne S. Tedeschi, MD[1], Nancy J. Roizen, MD[2], H. Gerry Taylor, PhD[2], Gail Murray, PhD[3], Christine A. Curtis, PhD[4], and Aditi Shah Parikh, MD[5]

(J Pediatr 2015;166:168-71)

- 15 year retrospective study from Case Western in OH
- 109 patients- 28/109 (25%) failed NBHT (AABR)
- 19/28 completed follow up evaluation (***32% lost to f/u)
- 15/19 had confirmed hearing loss (4 unilateral, 11 bilateral)
- No mention of tympanometry data
- No attempt to discern SNHL from CHL
- No information regarding severity of hearing loss
- No information regarding treatment/resolution

24

Table. Associations between risk factors for hearing loss and presence of hearing loss

Risk factors	Fisher exact test	Infants with risk factor, n (%)
NICU stay >5 days	0.159	77 (70.6)
Maternal illness	1.000	2 (1.8)
Family history of hearing loss	1.000	1 (0.9)
Low Apgar score	0.279	17 (15.6)
Low birth weight	0.097	8 (7.3)
Bilirubin >20 mg/dL	1.000	1 (0.9)
Meningitis	0.150	1 (0.9)
Defects of face/head	1.000	5 (4.6)
Ototoxic drugs (prenatal and postnatal)	0.101	53 (48)
Mechanical ventilation	0.008	19 (17.4)

- Mechanical ventilation was only variable associated with presence of hearing loss (p=0.008)

Tedeschi

25

A comparison of tympanometry with 226 Hz and 1000 Hz probe tones in children with Down syndrome

Meagan P. Lewis [a,*], Elizabeth Bradford Bell [b,1], Adele K. Evans [c,2]

International Journal of Pediatric Otorhinolaryngology 75 (2011) 1492–1495

- Comparison of 226Hz vs 1000Hz probe for accuracy of screening ears
- 1000Hz probe is considered standard for all infants 0-6 months of age
- This pilot study compared use of these two probes in pediatric DS patients
- Compared visual inspection by Peds ENT with tympanograms
- 26 ears- patient age 1-11 years
- 226Hz probe → 71% specificity
- 1000Hz probe → 100% specificity
- Fewer false positives with 1000Hz probe (flat tymps in presence of clear ME)

26

OME – ear tubes

27

28

Hearing loss in children with Down syndrome☆

Sally R. Shott [a,*], Aileen Joseph [b], Dorsey Heithaus [c]

International Journal of Pediatric Otorhinolaryngology
61 (2001) 199–205

- 5 year longitudinal study- 48 DS pts enrolled less than 2 years of age
- "state of the art" treatment paradigm
- All kids have ENT exam and hearing assessment every 6 months
- Kids with stenotic EAC's (40%) examined by ENT every 3 months
- 90% of kids need ear cleaning using microscope
- Bi-weekly phone calls to screen for trips to PCP for treatment of AOM
- 40 of 48 (83%) received ear tubes for treatment of OME
- 98% of kids had normal hearing test at conclusion of study
- Success rate attributed to aggressive surveillance and management of ME

29

Hearing loss in Down Syndrome revisited – 15 years later

Vairavan Manickam [a], Gordon S. Shott [b], Dorsey Heithaus [c], Sally R. Shott [d, e, *]

[a] *Department of Otolaryngology–Head & Neck Surgery, Geisinger Medical Center, Danville, PA, USA*
[b] *University of Cincinnati College of Medicine, Cincinnati, OH, USA*
[c] *Division of Audiology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA*
[d] *Department of Otolaryngology-Head & Neck Surgery, University of Cincinnati, Cincinnati, OH, USA*
[e] *Department of Pediatric Otolaryngology, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA*

International Journal of Pediatric Otorhinolaryngology 88 (2016) 203–207

- Retrospective review of 54 pts from previous study
- Age range 14-18 years
- 89% had at least 1 BTT- mean of 3.5 BTT's
- 30% of ear tubes placed after age 6
- 17% had chronic perforation- 0 cholesteatomas
- "overall decrease in hearing levels" as pt aged
- No mention of frequency of testing done after original study
- Average age of completion of ear specific testing was 4.5 years (range 1.75-11.5 years)- 4 patients never achieved this ability

30

Incidence and Frequency of PET placement in study population.

Number of PET	Frequency	Percent of children
0	4	7.41%
1	9	16.67%
2	10	18.52%
3	4	7.41%
4	12	22.22%
5	5	9.26%
6	5	9.26%
7	1	1.85%
8	1	1.85%
≥9	3	5.56%

Functional hearing at last audiology visit.

	Left ear	Right ear
Functional hearing	45 (83.3%)	45 (83.3%)
Nonfunctional hearing	7 (12.9%)	7 (12.9%)
Inconclusive	2 (3.7%)	2 (3.7%)

- 'functional' hearing defined as ≤30dB

Shott f/u

31

Otitis media with effusion in children with in Down syndrome

Marit Erna Austeng [a,b,*], Harriet Akre [c,d,e], Britt Øverland [c], Michael Abdelnoor [f], Eva-Signe Falkenberg [g], Kari Jorunn Kværner [b,h]

International Journal of Pediatric Otorhinolaryngology 77 (2013) 1329–1332

- Population based study from Norway including all 8 year-old children with Down Syndrome
- All pts had ENT exam and audiologic testing
- 20/52 (38%) had OME
- Pts with OME had worse hearing compared to non-OME
- No mention of incidence of SNHL
- No mention of interventions made for OME and results of follow up testing
- ***stress need for ongoing surveillance of hearing

32

Outcomes of tympanostomy tube placement in children with Down syndrome—A retrospective review

Lorien M. Paulson [a,*], Tyler S. Weaver [b], Carol J. Macarthur [a]

[a] Department of Otolaryngology, Oregon Health Sciences University, Portland, OR, United States
[b] Oregon Health Sciences University School of Medicine, Portland, OR, United States

International Journal of Pediatric Otorhinolaryngology 78 (2014) 223–226

- 10 year retrospective study from Oregon
- 102 patients who received BTT (no info on prevalence of BTT)
- 100/102 had tubes placed due to COME
- Less than half of pts had failed NBHT
- 64% required more than one set of BTT
- 71% had normal hearing bilaterally after surgery
- 86% had normal hearing in at least one ear after surgery
- Kids needing 3 or more sets of tubes had more TM problems: atrophy, retraction, atelectasis, perforation, cholesteatoma
- Adenoidectomy did not prevent chronic ear complications

33

Chronic ear surgery

34

35

Outcomes of tympanoplasty in children with down syndrome

Saied Ghadersohi[b], Jonathan B. Ida[a,b], Bharat Bhushan[a,b], Kathleen R. Billings[a,b,*]

[a] *Ann & Robert H. Lurie Children's Hospital of Chicago, Division of Otolaryngology – Head and Neck Surgery, Chicago, IL, USA*
[b] *Northwestern University, Feinberg School of Medicine, Department of Otolaryngology – Head and Neck Surgery, Chicago, IL, USA*

International Journal of Pediatric Otorhinolaryngology 103 (2017) 36–40

- Retrospective study of 91 ears on 69 patients (2008-2017)
- 60 ears observed only- 33% had spontaneous closure (avg time 3.3 yrs)
- 31 ears had tympanoplasty
- 55% closure rate after initial surgery- 71% after secondary surgery
- Average age for surgery was 11.7 years (77% fascia, 10% cartilage, 5 pts had mastoidectomy)
- Failed surgery correlated with ongoing ETD/OME
- HA usage: 53% in observation group, 48% in surgical group
- Although hearing improved in surgical group, HA usage was still common

36

Outcome features for the Observation and Tympanoplasty Groups are shown. Significant *p*-values are shown in bold.

	Observation Group	Tympanoplasty Group	*p*-value
Follow-up time since Perforation (years)	4.6 ± 3.1 (range 0.53–11.9)	4.8 ± 2.9 (range 0.32–11.3)	0.82
Closure rate	20 (33%)	22 (70.9%)	< **0.001**
Time to Spontaneous Closure (years)	3.3 ± 2.5 (range 0.46–8.14)		
Needed TT after Perforation Closure	4 (6.7%)	3 (9.7%)	0.56
Hearing aids utilized	32 (53.3%)	15 (48.4%)	0.41

- Only measured outcome improved by surgical intervention was closure rate
- ~10% required replacement of ear tube after TM repair
- High rate of HA usage in both groups

Billings

37

Challenges and outcomes of cholesteatoma management in children with Down syndrome☆

Saied Ghadersohi[b], Bharat Bhushan[a,b], Kathleen R. Billings[a,b,*]

[a] *Ann & Robert H. Lurie Children's Hospital of Chicago, Division of Pediatric Otolaryngology-Head and Neck Surgery, Chicago, IL, United States*
[b] *Northwestern University Feinberg School of Medicine, Department of Otolaryngology-Head and Neck Surgery, Chicago, IL, United States*

International Journal of Pediatric Otorhinolaryngology 106 (2018) 80–84

- 9 year retrospective study
- 17 ears in 14 kids (15 acquired, 2 congenital)
- 76% had at least 2 sets of tubes previously
- 60% diagnosed on office exam, 40% at time of planned BTT placement
- Average age for first cholesteatoma surgery was 9.8 years
- 87% had ossicular erosion at time of diagnosis
- ***likely late diagnosis of cholesteatoma due to challenges of adequate ear exams in office and reliable audiologic assessments
- Pts with cholesteatoma generally present with advanced disease

38

- ***40% ended up with CWD procedure
- 7 ears had 1 surgery, 7 ears had 2 surgeries, 1 ear had 5 surgeries
- No information on hearing results
- No information regarding challenges with mgmt of CWD cavities
- 7 of 11 patients had residual/recurrent disease at time of 2nd look:

Table 4
Findings associated with second-look or secondary procedures performed in 7 patients (8 ears) with acquired cholesteatomas. Patient 12 had 4 additional procedures. Years after 1st surgery done as a planned second look are shown in bold.

Ear	Years after 1st surgery	Approach	Cholesteatoma Location	OCR	Residual Disease	Recurrent Disease
3L	**0.4**	TC Endo	–	PORP	-	-
4	**2.8**	CWD	MT, MS	-	-	+
8	**0.5**	CWD	granulation	-	-	-
9R	**0.5**	CWU	Stapes	TORP	+	-
9L	**0.3**	CWU	FR, HT	TORP	-	+
10	**0.8**	CWU	RW	–	+	-
12a	**0.4**	CWU	Stapes	TORP	+	-
12b	**0.9**	CWU	Stapes	TORP[a]	-	-
12c	**2.0**	CWU	ET, MS	-	-	+
12d	**4.0**	CWD	MT, MS	-	-	+
13	**3.3**	CWD	MT, MS	-	-	+

Billings

39

Surgical Treatment of Middle Ear Cholesteatoma in Children with Down Syndrome

Andrea Bacciu, Enrico Pasanisi, Vincenzo Vincenti, Davide Giordano, *Antonio Caruso, Lorenzo Lauda, and Salvatore Bacciu

*Department of Otolaryngology, University of Parma, Italy and the *Gruppo Otologico, Piacenza, Rome, Italy*

Otology & Neurotology
26:1007–1010 © 2005, Otology & Neurotology, Inc.

- 7 year retrospective study
- 11 ears on 9 pts
- All had primary acquired cholesteatomas
- All had hx of COME but only 2 pts had previous BTT (???)
- 8 ears had CWD as primary procedure
- 2 ears had CWU as primary and then converted to CWD later
- No information on use of ossicular reconstruction
- No information regarding challenges of surveillance of CWD cavities

40

TABLE 2. *Treatment and surgical outcomes*

Patient/Ear	First surgery	Surgical outcomes	Second surgery
1/Left	CWD	No recurrence	No
1/Right	CWU	Recurrent cholesteatoma after 8 months	Canal wall-down mastoidectomy
2/Right	CWD	No recurrence Perforation of the neotympanic membrane	Myringoplasty
3/left	CWD	No recurrence	No
4/Left	CWD	No recurrence	No
4/Right	Modified Bondy mastoidectomy	No recurrence	No
5/left	CWU	Resdual cholesteatoma	Preplanned second stage operation
6/left	CWD	No recurrence	No
7/Right	CWD	No recurrence	No
8/Left	CWD	No recurrence	No
9/Right	CWD	No recurrence	No

TABLE 3. *Pre- and postoperative pure-tone averages for air and bone conduction, and corresponding air-bone gaps with respect to each ear*

Patient/Ear	Pre-op PTA BC (db)	Post-op PTA BC (db)	Pre-op PTA AC (db)	Post-op PTA AC (db)	Pre-op ABG (db)	Post-op ABG (db)
1/Left	25	25	70	45	45	20
1/Right	20	20	50	55	30	25
2/Right	20	20	65	40	45	20
3/Left	50	50	75	70	25	20
4/Left	20	30	50	55	30	30
4/Right	20	30	40	50	20	20
5/Left	10	10	30	15	20	5
6/Left	10	15	50	50	40	35
7/Right	15	15	50	30	35	15
8/Left	40	20	60	40	20	20
9/Right	1	15	60	35	45	20

AC, air conduction; ABG, air-bone gap; BC, bone conduction; PTA, pure tone average.

Bacciu

41

Canal wall reconstruction and preservation in the surgical management of cholesteatoma in children with Down's syndrome

Robert Nash *, Victoria Possamai, Scott Maskell, Martin Bailey, David Albert

Department of Paediatric Otolaryngology, Great Ormond Street Hospital for Children, London WC1N 3JH, UK

International Journal of Pediatric Otorhinolaryngology 78 (2014) 1747–1751

- 12 year retrospective study
- 12 ears in 9 patients
- Premise- can CWU procedure be successful?
- 7 ears had CWD as primary procedure
- 5 ears had CWU as primary procedure (3 canal wall reconstructions)
- 1 of 5 eventually converted to CWD
- Almost all had type 3 or 4 ossicular reconstruction
- Almost all kids had at least 40dB CHL post op (no signif. change compared to pre-op)
- Mention of challenge of CWD cavities- one pt required 4 general anesthetics over the course of 2 years to manage bowl infx

42

Prevalence and surgical management of cholesteatoma in Down Syndrome children

Anna Poliner [a], Chenge Mahomva [b], Chelsea Williams [c], Kristan Alfonso [d,e], Samantha Anne [b], Mary Musso [f,g], Yi-Chun Carol Liu [f,g,*]

International Journal of Pediatric Otorhinolaryngology 157 (2022) 111126

- 9 year retrospective review from 3 large tertiary Ped hospitals
- 16 (out of 2266) kids with cholesteatomas (~1% of DS cohort)
- 4 pts treated with tympanoplasty alone
- Persistent significant hearing loss is common

Surgical approach and pre- and postoperative pure tone averages (PTA).

Patient	N of tympano- plasty	N of tympano-mastoidectomy	CWU vs. CWD
1	0	4	CWU x 4
2	0	2	CWU x 2
3	0	1	CWD
4	0	1	CWU
5	0	4	CWU x 4
6	1	1	CWU
7	1	0	
8	2	1	CWU
9	1	0	
10	1	1	CWD
11	0	1	CWU
12	1	0	
13	1	0	
14	1	1	CWU
15	7	5	CWU x 5

43

Prevalence and surgical management of cholesteatoma in Down Syndrome children

Anna Poliner [a], Chenge Mahomva [b], Chelsea Williams [c], Kristan Alfonso [d,e], Samantha Anne [b], Mary Musso [f,g], Yi-Chun Carol Liu [f,g,*]

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2	0	2	CWU x 2
3	0	1	CWD
4	0	1	CWU
5	0	4	CWU x 4
6	1	1	CWU
7	1	0	
8	2	1	CWU
9	1	0	
10	1	1	CWD
11	0	1	CWU
12	1	0	
13	1	0	
14	1	1	CWU
15	7	5	CWU x 5

Multiple procedures common!

Only one CWD.

44

Monitoring these kids is hard…

- Small ear canals- partial view of TM's is the rule
- Not very cooperative
  - Tread lightly on the little ones….
- Frequently need to rely on tympanograms, partial audiologic testing, and parental concerns regarding hearing to make clinical decisions
- Adjust frequency of visits to accommodate needs/concerns

Personal experience with DS patients

- EPIC search- ~175 DS patients
- Challenging/rewarding patient population
- Strongest physician-family relationships
- Prototypical long term Otolaryngic care paradigm
- Vigilant monitoring for OME in especially early years
- Annual audiograms through high school (at least)
- SNHL tends to manifest by middle school (***my experience)
- Don't be in a rush to repair perforations- higher failures/poor hearing improvement
- Close monitoring after any major ear surgery (ongoing ETD)
- Avoid CWD cavities if at all possible

When to refer to Pediatric ENT?

- We believe that **ALL** kids with DS can benefit from ENT
  - **This is NOT specified in recent DS care guidelines
  - Specialized equipment for cleaning ears
  - Liaison to audiologists to coordinate testing
  - Coordinated care with other specialists
  - These patients need continuity of care by ENT
- If pt fails NBHT → refer immediately
- If pt passes NBHT → refer around 4 months of age