

## Audiology Assessment & Monitoring of patients with Cleft Palate, Craniofacial and Syndromic Conditions

### Site Applicability

All VCH Community Audiology sites (Vancouver, Richmond, North Shore, Squamish, and Powell River)

### Practice Level

Clinical Audiologists – basic skill

### Need to Know

#### Guidelines for Audiology Assessment and Monitoring

In March 2012 BC Children's Hospital (BCCH) Audiology developed a Clinical Practice Guideline for Cleft palate / Craniofacial and Syndromic (CP/CF/S) patients to support consistent audiological care of infants and young children in BC with cleft palate and craniofacial anomalies or syndromes associated with hearing loss. Principles of this guideline include evidence-based practice, family centered care, integrated and coordinated interprofessional care, and coordination with BC Early Hearing Program (EHP). VCH has adapted the BCCH guideline to detail the type, timing, frequency, and location of testing for CP/CF/S patients within VCH. Please note that this population is at risk for unusual configurations of hearing loss, as well as high frequency hearing loss, so testing at mid (1000 Hz) and high frequencies (6000-8000 Hz) is recommended when possible.

### Equipment & Supplies

The following equipment is necessary to meet this CPD:

- Clinical audiometer
- Immittance bridge
- Otoscope
- Diagnostic otoacoustic emissions
- Disposable, single-use insert earphones, ear tips, and probes for above equipment

### Practice Guideline

#### All patients with CP / CF / S:

Patient Population	Service Description	Service Location	Minimal Clinical Requirements
All infants with CP/CF/S (except Downs' Syndrome)	<b>ABR Assessment</b> by 3 months of age (to inform surgical decisions)	VCH infants will be seen at/in: <ul style="list-style-type: none"> <li>• BCCH if PASS EHP screen result</li> <li>• VCH (Vancouver, Richmond, North Shore) if REFER EHP screen result</li> </ul>	As per BC EHP ABR assessment protocol:* <ul style="list-style-type: none"> <li>• Ear specific A/C thresholds or appropriate MRL at 500 and 2000 Hz</li> <li>• Ear specific B/C thresholds at 500 and 2000 Hz if A/C thresholds elevated.</li> </ul>
All infants with CP/CF/S	<b>9 month Audiology Assessment</b>	Primarily BCCH Audiology, occasionally VCH Community Audiology on referral in BEST from BCCH Audiology	As per BC EHP 9 month protocol: <ol style="list-style-type: none"> <li>1. Behavioural testing: <ul style="list-style-type: none"> <li>• Ear specific MRL of less than or equal to 25 dB HL at 500 and 2000 Hz, and EITHER (1) ear specific MRL of less than or equal to 25 dB HL at 4000 Hz OR (2) present DPOAEs at</li> </ul> </li> </ol>

**Note:** This is a **controlled** document for VCH internal use. Any documents appearing in paper form should always be checked against the electronic version prior to use. The electronic version is always the current version.

Patient Population	Service Description	Service Location	Minimal Clinical Requirements
			4000 Hz bilaterally; OR <ul style="list-style-type: none"> <li>Soundfield MRL of less than or equal to 25 dB HL at 500 and 2000 Hz, AND DPOAEs present at 2000, 3000, 4000 kHz;</li> </ul> 2. Link with necessary community resources, including schools

\*VCH audiologist must enter 9 month monitor in BEST (To: BCCH Audiology; Title: 9 months follow up)

Patient Population	Service Description	Service Location	Target Clinical Results
Patients with documented unaided permanent hearing loss	<b>Audiology assessment</b> every 6 months for at least a year or until stable by Audiologist judgment; ongoing monitoring as per Audiologist.	VCH Community Audiology	1. Establish middle ear status 2. Behavioural testing: <ul style="list-style-type: none"> <li>Ear-specific A/C thresholds at 500, 1000, 2000, and 4000 Hz bilaterally</li> <li>B/C testing at applicable frequencies if elevated A/C thresholds;</li> </ul> 3. DPOAEs, if indicated by middle ear status; 4. Link with necessary community resources, including schools

#### Patients requiring sedated ABR assessment at BCCH Audiology:

Patient Population	Service Description	Service Location	Referral Guidelines
All patients with CP/CF/S	<b>Sedated ABR</b>	BCCH Audiology	1. EHP guidelines recommend consideration of sedated ABR testing after a maximum of 2 attempts with unknown hearing status. 2. Sedated ABR referrals will be considered at any time on clinician request (i.e. family concern, rejection of hearing aid, speech/language change or concern). 3. Please contact BCCH Audiology Program Lead directly if you would like to discuss referral.

### Patients with Cleft Palate only:

Patient Population	Service Description	Service Location	Target Clinical Results
Patients with CP post-myringotomy and/or ventilation tube insertion	<b>Audiology Assessment **</b>	VCH Community Audiology will see patients referred to them in BEST by BCCH Audiology.	1. Establish middle ear status 2. Behavioural testing: <ul style="list-style-type: none"> <li>• Ear-specific A/C thresholds at 500, 1000, 2000, and 4000 Hz bilaterally,</li> <li>• B/C testing at applicable frequencies if elevated A/C thresholds</li> </ul> 3. DPOAEs, if indicated by middle ear status 4. Link with necessary community resources, including schools
Patients with CP with normal hearing at 9 month or post-tube assessment	<b>Audiology Assessment</b> at 3, 4, 5, and 6 years of age**	VCH Community Audiology	

\*\*please fax results to BCCH CP/CF team at 604-875-2743

### Patients with Craniofacial and Syndromic conditions only:

Patient Population	Service Description	Service Location	Target Clinical Results
Patients with CF/S conditions with normal hearing at 9 months and <b>limited</b> <sup>^</sup> evidence of risk of permanent late onset hearing loss	<b>Audiology Assessment</b> at 3 years of age	VCH Community Audiology	1. Establish middle ear status 2. Behavioural testing: <ul style="list-style-type: none"> <li>• Ear-specific A/C thresholds at 500, 1000, 2000, and 4000 Hz bilaterally,</li> <li>• B/C testing at applicable frequencies if elevated A/C thresholds</li> </ul> 3. DPOAEs, if indicated by middle ear status 4. Link with necessary community resources, including schools
Patients with CF/S conditions with normal hearing at 9 months and <b>lower</b> <sup>^</sup> risk of permanent late onset hearing loss	<b>Audiology Assessment</b> at 3, 5, and 10 years of age**		
Patients with CF/S conditions with normal hearing at 9 months and with <b>higher</b> <sup>^</sup> risk of permanent late onset hearing loss	<b>Audiology Assessment</b> at 3, 3½, 4, 5, 6, 7, 8 and 10 years of age**		

\*\*please fax results to BCCH CP/CF team at 604-875-2743

<sup>^</sup>please see table (Level of risk for late onset hearing loss by syndrome) below

### Level of risk for late onset hearing loss by syndrome:

Limited		Lower		Higher	
Aperts	Klippel-Feil	22Q11 (VCF DiGeorge)	Noonan	Alport	Norrie
Chondro dysplasias	Nager	Branchio-oto-renal	Refsum	Alstrom	Osteo Imperfecta
Cornelia deLange	Ohdo	Charcot-Marie-Tooth	Saethre-Chotzen	Friedreich Ataxia	Osteopetrosis
Crouzon	Pierre Robin	CHARGE	Sticklers	Hunter	Pendred
Downs	Pfieffer	Goldenhars		Hurler	Turners
Ehler-Danlos	Treacher-Collins	Hemifacial Microsomia		Long QT	Ushers
Jervell-Lange	Waardenburg	Kabuki		Large Vestibular Aquaduct	
Klinefelter		Muenke		Neurofibromatosis type 2	

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### Legend:

Acronym	Full name
ABR	Auditory brainstem response test
A/C	Air conduction
ANSD	Auditory neuropathy spectrum disorder
BC EHP	BC Early Hearing Program
BCCH	BC Children's Hospital
BEST	<b>BC Early Hearing Surveillance Tool</b>

Acronym	Full name
B/C	Bone conduction
CP/CF/S	Cleft palate/craniofacial and syndromic
CP/CF team	Cleft palate / craniofacial team
DPOAE	Distortion product otoacoustic emissions
MRL	Minimum response level
PARIS	Primary Access Regional Information System

### Expected Patient/Client/Resident Outcomes

Consistent audiological care and follow up across VCH for patients with cleft palate / craniofacial and syndromic conditions, as well as better service coordination with BCCH Audiology / Cleft Palate team.

### Documentation

All attempted and/or completed audiology assessment in VCH must be documented as an Audiology Assessment in PARIS. Patients enrolled in the BC EHP must have their EHP number entered into the PARIS Audiology Assessment. PARIS automatically sends EHP assessments to BEST daily.

### References

Adapted from BCCH:

[Audiology Clinical Practice Guideline Cleft Palate / Craniofacial and Syndromic Patients](#) (March 2012).

### Developed/Revised by

**CPD Developer Lead:** Regional Coordinator, BCEHP, Community Audiology Centre

**Other members:**

*Practice Leads*

Clinical Supervisor, Community Audiology Centre  
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Manager, Community & Family Health, RH  
Manager, Public Health & Prevention Early Years, West Vancouver Community Health Centre, Coastal  
Manager, Home and Community Care, STS  
Manager Public Health & Prevention Services, Powell River/Sunshine Coast, Coastal

### Endorsed by

VCH: *(Regional SharePoint 2<sup>nd</sup> Reading)*

Health Authority Profession Specific Advisory Council Chairs (HAPSAC)  
Health Authority & Area Specific Interprofessional Advisory Council Chairs (HAIAC)  
Operations Directors  
Professional Practice Directors

### Final Sign-off & Approved for Posting by

Professional Practice Director on behalf of Chief Nursing Officer & Executive Lead Professional Practice , VCH

### Date of Approval/Review/Revision

Approved: November 15, 2012

Posted: November 22, 2012