Prevalence of Childhood Permanent Hearing Loss after Early Complex Cardiac Surgery

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Objectives To estimate the prevalence of childhood permanent hearing loss (PHL) after early cardiac surgery.

Study design This prospective observational (1996-2015) study after complex cardiac surgery with cardiopulmonary bypass at ≤6 weeks of life reports audiology follow-up by registered pediatric-experienced audiologists at 6-8 months postsurgery, age 2 years, and as required throughout and thereafter to complete diagnoses. PHL at any frequency (500-4000 Hz) is defined as responses of >25-decibel hearing level in either ear. PHL was evaluated by type (conductive or sensorineural), pattern (flat or sloping), and severity (mild to profound).

Results Survival rate was 83.4% (706 of 841 children) with a 97.9% follow-up rate (691 children); 41 children had PHL, 5.9% (95% CI 4.3%, 8.0%). By cardiac defect, prevalence was biventricular, 4.0% (95%CI 2.5%, 6.1%); single ventricle, 10.8% (95%CI 6.8%, 16.1%). Eighty-seven (12.6%) of 691 had syndromes/genetic abnormalities with known association with PHL; of these, 17 (41.5%) had PHL. Of 41 children, 4 had permanent conductive, moderate to severe loss (1 bilateral); 37 had moderate to profound sensorineural loss (29 bilateral with 20 sloping and 9 flat), 6 with cochlear implant done or recommended.

Conclusions Infants surviving complex cardiac surgery are at high risk for PHL. Over 40% with PHL have known syndromes/genetic abnormalities, but others do not have easily identifiable risk indicators. Early cardiac surgery should be considered a risk indicator for PHL. *(J Pediatr 2018;198:104-9).*

"Targeted hearing loss" was defined by the American Academy of Pediatrics in the 2007 Position Statement on Principles and Guidelines for Early Hearing Detection and Intervention programs as including congenital permanent bilateral sensory, unilateral sensory, permanent conductive hearing loss, and auditory neuropathy in infants admitted to the neonatal intensive care unit (NICU).[1] For newborns requiring intensive care, the usual well-infant universal screening is expanded and, if not passed, an early direct referral to an audiologist is recommended.[1] For those children passing the newborn screen but with 1 or more risk indicators for hearing loss, at least 1 diagnostic audiology assessment by age 24-30 months of age is recommended.[1] Risk indicators needing more frequent and complete assessments include a NICU stay of more than 5 days, assisted ventilation, extracorporeal membrane oxygenation (ECMO), or exposure to ototoxic medications.[1,2]

As part of the developmental assessment of infants with congenital heart disease, the American Heart Association recommends an audiology examination if there is a suspicion of hearing loss, if the infant has undergone surgery since the neonatal audiology examination, or if there is no record of a neonatal examination.[3] Little has been reported on the prevalence of permanent hearing loss (PHL) after early cardiac surgery until recently. Grasty et al have shown 6.9% of 4-year-old survivors at assessment have sensorineural and 2.3% indeterminate hearing loss.[4] In a retrospective study, Bourdages et al found a prevalence of 8.3% compared with a reference population of 0.14%.[5]

CCS	Complex cardiac surgery
dB HL	Decibel hearing level
ECMO	Extracorporeal membrane oxygenation
NICU	Neonatal intensive care unit
PHL	Permanent hearing loss
PTA	Pure tone average
SV	Single ventricle

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The objectives of this prospective study were to determine the prevalence estimate, types, patterns, and degree of severity of PHL among survivors after complex cardiac surgery (CCS) with cardiopulmonary bypass done at ≤6 weeks of life.

Methods

This was a prospective, observational study with enrollment of infants in the multidisciplinary developmental follow-up program at the time of their first CCS with ongoing follow-up of this inception cohort as has been described.[6-8]

The Stollery Children's Hospital in Edmonton, Alberta has been a referral hospital in western Canada for neonatal and pediatric CCS since 1996. The associated interprovincial neurodevelopmental longitudinal follow-up program for children identified at ≤6 weeks of age at the time of their CCS completes assessments of survivors, including audiological examinations, at 6 tertiary referral sites in western Canada.[6,7] Beginning in 1989, Edmonton hospitals also hosted a referral program for neonatal and pediatric ECMO with the follow-up program reporting the prevalence of sensorineural PHL among survivors.[9-13]

Of 706 2-year-old survivors of CCS at ≤42 days of life at Stollery Children's Hospital, Edmonton, Alberta from September 1996 until May 2015, 691 (97.9%) participated in audiology follow-up as part of full multidisciplinary assessments. There were no exclusions. Of 691 assessed children, 194 (28.1%) had single ventricle (SV) cardiac defects, including those with hypoplastic left heart syndrome. The remainder, 497, had biventricular cardiac defects. Those children requiring staged palliative surgery had the first stage Norwood with the Blalock-Taussig shunt until August 2002 when the Norwood-Sano shunt was introduced. Survival for children with SV defects improved after this change.[14]

Preoperative and postoperative acute care variables were collected for each child as has been described.[7,8] Socioeconomic status was determined using the Blishen Index, a measure based on the education and employment of the main wage earner of the family.[15] For the purpose of this study, the following variables are reported from the first CCS for each child. The preoperative variables include—sex, birth weight and gestation, presence of genetic or syndrome diagnoses known to be associated with PHL, SV defect, family socioeconomic index; operative variables—year of first CCS, weight at surgery, duration of cardiopulmonary bypass, deep hypothermic circulatory arrest used; overall hospitalization—dialysis used, presence of convulsions, cardiopulmonary resuscitation needed, ECMO used, and duration of ventilation and hospitalization. All children received various ototoxic medications that may have included antibiotics, diuretics, and neuromuscular blockers; the details and dosages of these for all children were not recorded.

Audiological examinations in sound booths were administered by registered pediatric-experienced audiologists at a minimum of 6-8 months postsurgery, at age 2 years, and as required throughout and thereafter to complete diagnoses and monitor thresholds.[6,13] Developmentally appropriate and clinically standardized hearing testing procedures were used.[16] Behavioral testing was done at the first visit using visually reinforced audiometry if the child was able to complete the test; if not, then diagnostic auditory brainstem response testing was done.[16] For any child not found to have responses within normal limits on any behavioral test, a diagnostic auditory brainstem response test was done. Serial visually reinforced audiometry or play audiometry was used as needed.[6,13] For older children, conditioned play audiometry included bilateral pure-tone responses at 500-4000 Hz with earphones or inserts.[17] If a hearing loss was found, then bone-conduction was completed to differentiate between types. Temporary conductive hearing loss as with, eg, otitis media, was ruled out or medically treated before a PHL diagnosis was given. All children with PHL were seen by an otolaryngologist and investigated for cause of hearing loss; this included a referral to a geneticist as indicated.[6]

Terms defining PHL types include, permanent conductive hearing loss resulting from abnormalities of the external ear and/or the ossicles of the middle ear, or middle ear space, and sensorineural hearing loss resulting from malfunction of the inner ear structure (ie, cochlea).[17,18] Patterns were defined as flat or sloping.[18,19] Hearing loss at any one of the frequencies of 500-4000 Hz was defined as responses of more than 25-decibel hearing level (dB HL) in either ear.[6] Results for flat losses determined in this manner were compared with pure tone averages (PTAs) of mean air-conduction thresholds at 500, 1000, and 2000 Hz[19] and no differences were found. By convention, the degree of loss of these averages is recorded in dB HL as mild, 26-40; moderate, 41-55; moderately severe, 56-70; severe, 71-90; and profound, more than 90.[18] For sloping losses, PTA may be misleading,[19] therefore, we considered the configuration of the loss and applied the terms for degree of loss to the greatest loss at any one of the frequencies tested as recommended.[19]

Ethics board approvals were obtained at all sites (institutional review board approval: Health Research Ethics Board, University of Alberta, Edmonton, Alberta, Canada, ID# Pro00001030; last renewal date, November 17, 2017). All parents or guardians signed consent.

Statistical Analyses

Continuous variables are presented as means (SD); categorical variables as counts and percentages. Prevalence estimates of PHL are given as percentages of assessed survivors with 95% CIs. Comparisons of the descriptive variables were done using the Student *t* test for continuous variables and the Fisher exact test (2-sided) for noncontinuous variables. Statistical significance was defined after bidirectional Bonferroni corrected comparisons. Analysis was completed with SPSS v 23 (SPSS Inc, Chicago, Illinois).

Results

From September 1996 to May 2015, 841 infants of 6 weeks of age or less had CCS at this center. Survival to age 2 years was 706 (83.4%) with audiology assessment results available for 691 children at age 2 years or after (97.9% of survivors). **Table I**