Electrocardiogram Screening in Children with Congenital Sensorineural Hearing Loss: Prevalence and Follow-up of Abnormalities

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Abstract
Objective. The purpose is to determine the prevalence of electrocardiogram (ECG) abnormalities, including borderline and prolonged QT, among screened children with sensorineural hearing loss (SNHL) and to analyze their subsequent medical workup.


Setting. Tertiary academic center.

Subjects and Methods. Cases from 1996 to 2014 involving pediatric patients (N = 1994) with SNHL were analyzed. Abnormal ECGs were categorized as borderline/prolonged QT or other. A board-certified pediatric cardiologist retrospectively determined the clinical significance of ECG changes. For follow-up analysis, children with heart disease, known syndromes, or inaccessible records were excluded.

Results. Among 772 children who had ECGs, 215 (27.8%) had abnormal results: 35 (4.5%) with QT abnormalities and 180 (23.3%) with other abnormalities. For children with QT abnormalities meeting inclusion criteria (n = 30), follow-up measures included cardiology referral (46.6%), repeat ECG by ear, nose, and throat (ENT) specialist (20%), clearance by ENT specialist with clinical correlation and/or comparison with old ECGs (20%), and pediatrician follow-up (6.7%). Documentation of further workup by ENT or referral was absent for 6.7%. For children with other ECG changes meeting inclusion criteria (n = 136), abnormalities were documented for 57 (41.9%); normal QT without other abnormality was documented for 18 (13.2%). The most common follow-up referrals were to pediatricians (16.9%) and cardiologists (10.3%). Among patients with clinically significant non-QT abnormalities mandating further evaluation (n = 122), 38 (31.1%) had documented follow-up in medical records.

Conclusion. There is a high prevalence of ECG abnormalities among children with congenital SNHL. If findings are confirmed by future studies, screening should be considered for congenital unilateral or bilateral SNHL, regardless of severity. We describe a standardized protocol for ECG screening/follow-up.

Keywords
sensorineural hearing loss, congenital, hearing loss, audiology, screening, Jervell and Lange-Nielsen syndrome, electrocardiogram

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The prevalence of hearing loss in the United States is 1 to 2 per 1000 children at birth, rising to 3.5 per 1000 by adolescence.¹ Among the known causes, over half the cases of pediatric prelingual hearing loss are genetic.² Hereditary hearing loss is primarily nonsyndromic and attributed to mutations, such as connexin-related hearing loss.₂,₃ Early identification and intervention of children with hearing loss have favorable developmental outcomes,⁴,⁵ and thorough workup provides direction for management. Syndromic hearing loss requires evaluation for

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associated comorbidities to appropriately treat these conditions.

Many providers use electrocardiograms (ECGs) to screen children with hearing loss for Jervell and Lange-Nielsen syndrome (JLNS). This form of long QT syndrome (LQTS) places patients at risk for arrhythmias, syncopal episodes, and sudden death.6-12 Although percentages vary by demographics, the incidence of JLNS/LQTS is very low, with historical numbers up to 4% found in a cohort of 707 children with sensorineural hearing loss (SNHL).13 While the possibly lethal nature of JLNS has galvanized research highlighting QT changes in children with severe to profound bilateral hearing loss, no studies have reported on the prevalence and follow-up measures taken among screened patients with other significant ECG abnormalities. The purpose of this study was to determine the prevalence of ECG abnormalities, including borderline and prolonged QT, in children with mild to profound sensorineural hearing loss and to analyze the subsequent medical workup of these patients.

Materials and Methods

This study received Institutional Review Board approval (13-3883) from the University of North Carolina. It was based on a case series with chart review of pediatric patients (n = 2603) at the Carolina Children’s Communicative Disorders Program from 1996 to 2014. Children (ages, 1 month to 18 years) with hearing loss who were seen for routine clinical care at the program or University of North Carolina Hospitals were included in the study. A total of 609 patients were excluded for reasons including missing data, unspecified hearing loss type, or conductive hearing loss. Of the 1994 remaining patients with SNHL, 772 had ECGs and composed our study population (Figure 1). Of note, the patients had varying degrees of hearing loss, ranging from mild to profound.

All abnormal ECGs were recorded and divided into 2 groups: borderline and prolonged QT and other abnormalities. At the time of database review, a board-certified pediatric cardiologist retrospectively determined the clinical significance of ECG changes and need for follow-up. Based on these determinations, the appropriateness of follow-up measures was analyzed. In both groups, we studied (1) how often patients were referred to their primary care physicians/pediatricians or cardiologists for further workup and (2) what follow-up investigations were undertaken, including repeat ECGs, echocardiograms, and Holter monitoring. Forty-nine patients (n = 5 with QT changes and n = 44 with non-QT changes) with congenital heart disease, CHARGE syndrome, and inaccessible medical records and those who received an ECG for reasons other than SNHL screening were excluded in the analysis of follow-up measures (Figure 1). Of note, part of the database was created prior to the implementation of a new medical record system at the academic center. The nomenclature “inaccessible medical records” is utilized to indicate partial loss of patient data in the transition that was not already present in the database. Since this study primarily analyzed prevalence in

Figure 1. Exclusion of patients from database. ECG, electrocardiogram; SNHL, sensorineural hearing loss.
a retrospective case series, detailed statistical analysis was not performed.

**Results**

Of 1994 children with SNHL (1062 males, 932 females), 150 (7.5%) had unilateral SNHL. Of the 1994, 772 (38.7%) had ECGs, 215 (27.8%) of whom had abnormal results. Of the 772 whose ECGs, 48 (6.2%) had unilateral SNHL, and of the 215 subjects with abnormal ECGs, 19 (8.8%) had unilateral SNHL.

On initial ECG screening, the prevalence of borderline or prolonged QT and other abnormal changes was 35 (4.5%) and 180 (23.3%) of 772, respectively. Two of 35 (5.7%) subjects with QT changes and 17 of 180 (9.4%) with other abnormal changes had unilateral SNHL. Of note, no children met the criteria of Schwartz et al for LQTS. Briefly, Schwartz et al suggested diagnostic criteria in 1993, divided into 3 main categories—ECG findings, clinical history, and family history—with higher scores indicating higher probability of LQTS. The average age at the time of ECG was 4.4 years (SD, 4.6). The average age at the time of ECG in the subset with abnormal results was 4.0 years (SD, 4.8). Among the patients with QT abnormalities who had complete audiologic data with recorded severity of degree of hearing loss in the database (28 of 35), mild, moderate, and severe-profound hearing loss was present in 3 (11%), 4 (14%), and 21 (75%), respectively. Among patients with other ECG abnormalities, 103 of 180 (57.2%) had documented severity of SNHL with complete audiologic data: mild, 5 (4.9%); moderate, 17 (16.5%); and severe-profound, 81 (78.6%).

Among those with QT changes who met inclusion criteria for follow-up analysis (30 of 35), follow-up measures included cardiology referral for 14 (46.7%), repeat ECG by ear, nose, and throat (ENT) specialist for 6 (20%), clearance by ENT specialist via clinical correlation and/or comparison with old ECGs for 6 (20%), and pediatrician follow-up for 2 (6.7%). Documentation of further workup by ENT specialist or referral was absent for 2 (6.7%) children.

Of children with other ECG changes who met inclusion criteria for follow-up analysis (136 of 180), ENT documentation of ECG abnormalities in medical records was present for 57 (41.9%). Of these 57, there was documentation of normal QT but no documentation of other abnormalities for 18 (13.3% of 136). When further evaluation was undertaken, the most common follow-up included pediatrician follow-up for 23 (16.9%) and cardiology for 14 (10.3%). Clearance from anesthesiology prior to cochlear implantation and ENT clinical evaluation and comparison with old ECG were done for 1 patient each (1.5%). Among the clinically significant non-QT abnormalities determined by pediatric cardiologist review to recommend further evaluation Table 1, 30.1% (38 of 122) were documented to have received follow-up. All patients with ECGs were living at time of database and chart review.

**Discussion**

For children with SNHL, an ECG to rule out JLNS is often utilized. When pediatric otolaryngologists in the United States were surveyed in 2011 regarding evaluation of pediatric SNHL, one-third of responders reported ordering ECGs as part of their initial assessment. ECGs were more often obtained when patients had bilateral hearing loss, particularly in the moderate to profound range. This is in line with a diagnostic pathway for SNHL proposed by Preciado et al, in which an ECG is recommended subsequent to a negative GJB2 screen for children with severe to profound SNHL. A study in the otolaryngology literature and a Triological Society Best Practice paper from 2013 recommended ECG screening for children with bilateral sensorineural hearing loss with a history of loss of consciousness and arrhythmia and a family history of sudden death. However, one might argue that waiting for symptom manifestation for cardiology workup may be dangerous. An early diagnosis of JLNS leads to prompt lifesaving intervention—namely, optimization on beta-blockers. This is particularly important prior to cochlear implant surgery, since patients may be at risk for torsade de pointes perioperatively or with implant activation. However, the prior recommendations for ECG screening of children with hearing loss focused solely on evaluating for JLNS/LQTS, which admittedly is a very rare entity. We propose that this may bias

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<th>Electrocardiogram Changes</th>
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<th>Not Necessitating Follow-up</th>
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<tr>
<td>Left anterior fascicular block</td>
<td>Incomplete right bundle branch block</td>
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<td>Deep Q waves</td>
<td>Early repolarization</td>
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<td>Abnormal QRS</td>
<td>Nonspecific T-wave flattening</td>
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<td>Complete right bundle branch block</td>
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<td>Sinus bradycardia*</td>
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<td>Premature supraventricular or ventricular beats</td>
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<td>Prominent voltage in lateral or inferior leads</td>
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*Denotes electrocardiogram changes that may necessitate follow-up if patient is clinically symptomatic (see Figure 2).
the practitioners to overlook other significant ECG abnormalities, as they may focus primarily on QT findings, and it may limit ECG evaluation to bilateral profound hearing loss as well.

Despite the widespread use of ECGs to evaluate children with SNHL, cardiology referrals are rarely obtained in the United States.12 Patients with non-QT abnormalities on ECG screen are likely being cleared by otolaryngologists for JLNS but may receive no further evaluation for the other ECG abnormalities. This may be due to the intended use of ECG for diagnosis of JLNS as the SNHL etiology, instead of screening for other cardiac abnormalities. Less than one-third of ECG abnormalities that required further cardiology workup actually received additional follow-up in this study. Given the high number of ECG abnormalities discovered in the current study and other previously reported series18,19 and given the disparity of follow-up in the QT group versus the other ECG abnormality group, we believe that cardiology consultation and follow-up for ECG abnormalities may be underutilized in this group of patients. This lack of follow-up (or, at least, documentation of follow-up) at our institution and the similar findings in the pediatric otolaryngology survey12 may be attributed to the complex model of tertiary referral medical centers in which follow-up external to the center is difficult to determine. Less attributable factors observed in our study nonetheless included instances when (1) the next visit after screening ECG was with other health care professionals, (2) the next ENT visit was scheduled several months later, and (3) the ECG was performed in the preoperative visit prior to cochlear implantation.

Importantly, close follow-up was maintained between otolaryngologists and patients who had borderline or prolonged QT changes on ECG. Of note, 40% of patients were cleared by otolaryngology due to the absence of clinical history and signs of JLNS and/or comparison with prior or repeat normal ECGs. However, we think that cardiology referral should still be obtained in cases of abnormal QT, other concerning symptoms, or a family history of JLNS. Exercise testing, additional monitoring, or genetic testing to exclude JLNS/LQTS can then be obtained. The importance of referral is highlighted in that the diagnosis of LQTS is not always straightforward, and some potential difficulties include asymptomatic patients, negative family histories, and the observation that 12% to 30% of resting ECGs may be normal in patients with LQTS.20 Additionally, it has been reported that 10% of patients with JLNS are asymptomatic during the first year of life.7

**Figure 2.** Protocol for cardiac screening in children with congenital SNHL. BVH, bilateral ventricular hypertrophy; ECG, electrocardiogram; JLNS, Jervell and Lange-Nielsen syndrome; LAD, left axis deviation; LBBB, left bundle branch block; LVH, left ventricular hypertrophy; RAD, right axis deviation; RBBB, right bundle branch block; RVH, right ventricular hypertrophy; SNHL, sensorineural hearing loss.
There is a paucity of literature highlighting clinical pathways for further evaluation when an ECG screen in patients with SNHL is abnormal, particularly beyond QT abnormalities. A prior series of 132 patients with congenital SNHL by Ilhan et al. demonstrated other abnormalities found on screening ECGs in 15% of children (20 of 132) but did not elucidate further workup. Mehta et al. reviewed 420 children with SNHL who had ECGs, reporting a 12.9% rate of abnormal ECG findings. While an ECG is an excellent, cost-effective screening tool, its worth is greatly diminished if no system exists for systematic follow-up of patients with positive screens. We have implemented a protocol for follow-up based on our institutional cardiology recommendations (Figure 2). Additionally, communication between otolaryngologists and other health care providers is integral, so that screening ECGs are reviewed in a timely manner and adequate follow-up is obtained. The false-positive rate of ECG screens has been reported to be as high as 99%, but the consequence of missed cardiac diagnoses can be devastating. Thus, we implemented a protocol where patients with concerning abnormalities on ECG screen are referred for cardiology evaluation.

This study had several limitations. First, it was a retrospective review of ECGs that were ordered prior to the proposed protocol and hence were practitioner dependent. The study was performed at a large pediatric hearing loss and cochlear implant center over a long period with multiple practitioners. As such, patients frequently receive additional care at home institutions and with home health care providers who are outside the medical records system. Therefore, follow-up ECGs or medical workup may not have been captured in our medical records, hence underestimating the percentage of patients who actually received appropriate follow-up. This study reports on the prevalence of ECG findings and initial medical workup only. Future studies will analyze the clinical significance of these ECG findings prospectively. Additionally, we relied on documentation of further patient evaluation by otolaryngologists and thus may have missed additional follow-up data not documented in the medical record.

Conclusion
We present the largest study of ECG abnormalities among children with SNHL. Our data suggest that ECG abnormalities may be more common than previously thought among children with unilateral or bilateral SNHL, are present in a range of hearing loss severities, and are not limited to JLNS. While prospective data and further analysis are needed, pediatric otolaryngologists who treat children with congenital SNHL may want to consider ECG screening protocols to identify other ECG abnormalities aside from QT prolongation in children with SNHL. For reference, the algorithm implemented at the University of North Carolina is included (see Figure 2). If our findings are confirmed with future prospective studies, we propose that routine ECG screening be considered for children with unilateral or bilateral SNHL regardless of severity. Otolaryngologists should be alert for ECG changes other than LQTS. Standardized clinical pathways are suggested to ensure appropriate follow-up of children with abnormal ECGs discovered on screening.

Author Contributions
Zainab Farzal, conception and design, acquisition of data, analysis and interpretation of data, drafting and revising manuscript; Jonathan Walsh, conception and design, acquisition of data, analysis and interpretation of data, drafting and revising manuscript; Faisal I. Ahmad, acquisition of data, analysis and interpretation of data, drafting and revising manuscript; Jason Roberts, acquisition of data, analysis and interpretation of data, drafting and revising manuscript; Sunita J. Ferns, conception and design, analysis and interpretation of data, drafting and revising manuscript; Carlton J. Zdanski, conception and design, acquisition of data, analysis and interpretation of data, drafting and revising manuscript.

Disclosures
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