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Autism spectrum disorders in 24 children who are deaf or hard of hearing

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ABSTRACT

Objectives: Approximately 4% of children who are deaf or hard of hearing have co-occurring autism spectrum disorder (ASD). Making an additional diagnosis of ASD in this population can be challenging, given the complexities of determining whether speech/language and social delays can be accounted for by their hearing loss, or whether these delays might be indicative of a comorbid ASD diagnosis. This exploratory study described a population of 24 children with the dual diagnosis of ASD and hearing loss.
Methods: Children completed a comprehensive ASD evaluation using standardized autism diagnostic instruments (Autism Diagnostic Observation Schedule, language and psychological testing). Children with permanent hearing loss who had a developmental evaluation between 2001 and 2011 and were diagnosed with an ASD based on the results of that evaluation were included. Information on communication modality, language and cognitive abilities was collected.
Results: The median age of diagnosis was 14 months (range 1–71) for hearing loss and 66.5 months (range 33–106) for ASD. Only 25% ($n = 6$) children were diagnosed with ASD $\leq$48 months of age and 46% by $\leq$6 years. Twelve (50%) children were diagnosed with ASD, 11 were diagnosed with pervasive developmental disorder not otherwise specified and 1 child had Asperger's. Most (67%) had profound degree of hearing loss. Fourteen (58%) children had received a cochlear implant, while 3 children had no amplification for hearing loss. Nine (38%) of the 24 children used speech as their mode of communication (oral communicators).
Conclusions: Communication delays in children who are deaf or hard of hearing are a serious matter and should not be assumed to be a direct consequence of the hearing loss. Children who received cochlear implants completed a multidisciplinary evaluation including a developmental pediatrician, which may have provided closer monitoring of speech and language progression and subsequently an earlier ASD diagnosis. Because children who are deaf or hard of hearing with ASD are challenging to evaluate, they may receive a diagnosis of ASD at older ages.

1. Introduction

Autism spectrum disorders (ASD) constitute a group of developmental disorders characterized by impairments in social interaction and communication, and by restricted, repetitive, and stereotyped patterns of behavior [1]. Early identification of children with ASD is critical in their medical management and treatment [2–8], with screening endorsed as early as 18 months of age [9–11]. Although symptoms are usually apparent by 3 years of age, most children are not diagnosed with ASD until after age 4 [12]. One of the challenges in identifying children at very young ages relates to an absence of skills (such as eye contact, gestures, pointing, and pretend play) rather than the presence of noticeable features (such as spinning parts of toys, lining up toys, or repeating phrases) [13]. However, parents often do notice developmental problems as early 12 months of age, with concerns of hearing and vision most reported in that first year [14,15].

With the implementation of universal newborn hearing screening in the United States, the age of identification for hearing loss has significantly decreased [16–18]. This earlier diagnosis has made the early language acquisition trajectory for young children who are deaf or hard of hearing approach that of

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hearing children [19]. The dual diagnosis of hearing loss and ASD is important as nearly 40% of children with an identified permanent hearing loss have an additional disability [20]. Among this group of children with different learning patterns, the prevalence estimates of ASD have been reported to be as high as 1.7% [21] to 4% [22]. Since the comorbidity of ASD and hearing loss may significantly complicate language development, early identification of both conditions is of utmost importance to mitigate the effects of the dual diagnosis. Unfortunately, because both hearing loss alone and ASD alone could contribute to communication and social delays, identification of the true dual diagnosis is challenging [23–26].

The literature on the dual diagnosis of ASD and hearing loss is severely lacking. Thus characterizing this group of children is an appropriate first step to enhance our understanding and build the foundation for improved identification and intervention for their communication needs. The objective of this descriptive or exploratory study was to describe a population of children with the dual diagnosis of permanent hearing loss and ASD in the context of the degree of hearing loss.

2. Methods

2.1. Participants

This study was approved by the Institutional Review Board at Cincinnati Children's Hospital Medical Center (CCHMC). Children with permanent hearing loss who had a developmental evaluation between 2001 and 2011 and were diagnosed with an ASD based on the results of that evaluation were included. This developmental evaluation was part of a comprehensive approach to the medical evaluation of children with hearing loss at CCHMC, Ear and Hearing Center [27]. Children with an identified permanent hearing loss (managed by a pediatric audiologist) are referred for an interdisciplinary clinical protocol by their managing pediatric otologist. Potential referrals for this process include referrals to ophthalmology, developmental pediatrics, genetics, speech pathology, and aural rehabilitation. Children with any degree (mild to profound) of permanent hearing loss were included. The severity of hearing loss was clinically classified between mild and profound using a combination of frequency specific decibel loss and the slope of the audiogram. Type of hearing loss was also collected, and included both unilateral or bilateral hearing loss, sensorineural, conductive, mixed, or auditory neuropathy.

2.2. Clinical evaluation for autism spectrum disorder

As part of clinical care, children completed a comprehensive evaluation for an ASD using standardized autism assessments. The assessment battery was not consistently administered across all children due to the time-frame, during which the children were clinically seen and the variability in children's ability to complete specific testing instruments. Evaluations included (as seen clinically appropriate) the Gilliam Autism Rating Scale (GARS) [28], Autism Diagnostic Observation Schedule (ADOS) [29], cognitive testing, test of adaptive behavior, and language testing. The GARS was administered by a psychologist and the ADOS was administered by a speech-language pathologist. Children were seen by a developmental pediatrician who has extensive experience with children who are deaf or hard of hearing. Because autism diagnostic tools (e.g., GARS and ADOS) have not been validated for use in children who are deaf or hard of hearing, both qualitative and quantitative information from the assessments were used in conjunction with DSM-IV criteria. Thus, a team of professionals representing expertise in ASD and expertise in hearing loss arrived at a consensus opinion for the diagnosis of ASD. All evaluations were used to clarify developmental patterns and were interpreted with the impact of hearing loss on scoring considered. Because the scoring algorithms for the ADOS are not valid for use in children with sensory impairments such as hearing loss, the discussion regarding the scores from the ADOS or the GARS is beyond the scope of the objectives of this study and will be addressed in a future report. Severity of an autism spectrum disorder is not routinely measured; however in general, a diagnosis of autism would be viewed as more severe than a diagnosis of pervasive developmental disorder not otherwise specified.

2.3. Cognition and language assessments

When possible, standardized cognitive assessments occurred at the discretion of the clinician. For the purpose of this analysis, nonverbal intelligence quotients were used. For children who did not have a standardized cognitive assessment, a developmental assessment was available using the Revised Gesell Developmental Schedules [30,31], which provides an adaptive (performance) age equivalent. A developmental quotient was calculated based on the developmental age compared to the chronological age of the child at the time of administration (developmental age/chronologic age $\times$ 100). Language assessments were conducted for clinical purposes and tools were selected as deemed appropriate by the speech-language pathologist. The Preschool Language Scales-4th edition (PLS-4) [32] and the Clinical Evaluation of Language Fundamentals (CELF) [33] were most commonly used. The CELF was more often used in children who had higher language levels. Both assessments yield standard scores and age equivalents for receptive and expressive language. Because the PLS-4 has a floor of 50 as a standardized score, a language quotient was used for the analysis. The language quotient was derived by dividing the age equivalent from the PLS-4 by the chronological age of the child at the time of testing, multiplied by 100 (age equivalent/chronological age $\times$ 100). Quotients of 100 represent language ages equivalent to the chronological ages. The test results reported were results from the assessment closest to the ASD diagnosis. Communication strategies used were defined as oral, sign (any sign language or sign support) and behavior (acting out for the purpose of communicating).

2.4. Statistical analysis

Because this study was meant to be a descriptive study of children who had the dual diagnosis of ASD and hearing loss, the statistics reported remained descriptive in nature. Descriptive statistics included medians with ranges for continuous variables (e.g., age) and frequencies with percentages for categorical variables (e.g., etiology). We conducted simple Spearman's rank-order correlations to understand the relationship between language scores and ages at time of either hearing loss or ASD identification. Significant differences in subject characteristics between children with severe to profound hearing loss and children with lesser degrees of hearing loss were explored using Wilcoxon Rank Sum test for continuous variables, or a Chi-square or Fisher's Exact test for categorical variables. Point biserial correlations (r_{pb}) were used to explore the association between severity of hearing loss and language scores. Statistical significance was set at $p \leq 0.05$. Statistical analysis was conducted using SAS® Version 9.3 software (Cary, NC).

3. Results

3.1. Study participants

Thirty-four children with hearing loss were identified as having had an evaluation for suspected ASD. Of these, three children were