2016

Why a diagnosis of neurofibromatosis calls for the attention of a deaf educator

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WHY A DIGANOSIS OF NEUROFIBROMATOSIS CALLS FOR THE ATTENTION OF A DEAF EDUCATOR

by

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An Independent Study
Submitted in partial fulfillment of the
Requirements for the degree of

Masters of Science in Deaf Education

Washington University School of Medicine
Program in Audiology and Communication Sciences

May 20, 2016

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Abstract: This paper will seek to describe neurofibromatosis (NF), the scope of its impact, how NF relates to hearing loss, and why someone with a teacher of the deaf's expertise may have information to offer the intervention team for a child diagnosed with NF.

Acknowledgements

Thank you to my Independent Study advisor Greta Bohnenkamp, SLP-CCC, and my secondary advisor, David Gutmann, M.D., Ph.D., for collaborating with me on this paper. This has proven to be a somewhat unorthodox topic, and I am grateful for the encouragement and enthusiasm you both brought to the project. It has truly been a privilege to be under your direction and mentorship throughout this process.

I would also like to thank The Washington University NF Center and all of the resources that have been shared with me as I have pursued this project. I am humbled by your diligence and generosity, and look forward to future collaboration with you.

To my amazing parents, thank you for your support through all my endeavors—academic and otherwise. I would not be half the woman I am today without your love and guidance urging me onward.

David, my dear friend, I am beyond grateful for you. You have been such an encouragement as I have navigated the joys and difficulties of my graduate school career. You lead me in all that is lovely and true, and I cannot wait to be your wife.

Finally, I would like to dedicate this project to my Lord and Savior Jesus Christ. I am eternally beholden to His steadfast grace, for in its absence I would be nothing.

"Such confidence we have through Christ toward God. Not that we are adequate in ourselves to consider anything as coming from ourselves, but our adequacy is from God who also made us adequate as servants of a new covenant, not of the letter but of the Spirit; for the letter kills, but the Spirit gives life."
2 Corinthians 3:4-6

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Introduction

Regardless of etiology, children with hearing loss will experience some degree of delayed and disordered language abilities in the areas of pragmatics, semantics, syntax, and morphology. The impact of these language deficits can affect the spelling, writing, math, reading, auditory development and processing, working memory, and executive functioning of students who are deaf or hard-of-hearing (Baddeley, 2003; Blamey et al., 2001; Cunningham, Nicol, Zecker, Bradlow, & Kraus, 2001; Easterbrooks & Estes, 2007; Geers & Hayes, 2010; H. Hayes, Treiman, & Geers, 2014; Joshi, Treiman, Carreker, & Moats, 2008-2009; Kelly, Lang, & Pagliaro, 2003; Nittrouer, Sansom, Low, Rice, & Caldwell-Tarr, 2014; Smedt et al., 2009; Traxler, 2000). Teachers of the deaf are specifically trained to target these areas of delay in a manner unique to children who are deaf and hard-of-hearing.

Throughout their careers, teachers of the deaf will encounter students who present with various etiologies of hearing loss. Many deaf educators are familiar with how diagnoses of congenital hearing loss, hearing loss due to meningitis, and hearing loss related to neuropathy or brain tumor can affect a child’s performance in the classroom. For each of these conditions, teachers of the deaf are able to understand the effect these additional disabilities have on learning and development. This knowledge equips them to apply appropriate remediation strategies and team with other professionals to establish effective interventions.

There are other causes of hearing loss that remain largely unrecognized by teachers of the deaf and have as significant an impact on a child’s academic performance. Neurofibromatosis (NF) is one such condition. An article by Briggs, Brackmann, Baser, & Hitselberger (1994) outlines the treatment plans for NF; the role of a deaf educator is not mentioned. There are three

types of NF—NF1, NF2, and Schwannomatosis. Whereas individuals with Schwanntomatosis do not typically have hearing loss, young adults with NF2 can present with reduced hearing due to a growing vestibular nerve tumor. Vestibular schwannoma-induced hearing loss leads to similar academic struggles observed in individuals with other etiologies for hearing loss. However, even without documented hearing loss, children with NF1 can manifest with lingual, social, learning, and cognitive deficits. Although not a result of the hearing loss, these deficits create similar challenges faced by children who are deaf or hard-of-hearing.

This paper aims to describe what NF is, the scope of its impact, how NF relates to hearing loss (particularly in school performance), and why someone with a teacher of the deaf's expertise may be included in the ideal intervention team for this child.

Defining Characteristics of and Challenges for Students who are Deaf or Hard-of-Hearing

A deaf or hard-of-hearing diagnosis will have varying degrees of impact on a child's language, social, and academic development (ASHA; Sininger, Grimes, & Christensen, 2010). Progress in these areas of development is dependent on a multitude of factors. Some of these factors to include the age of onset of the hearing loss, type of loss, the site of lesion, the cause of the hearing loss, degree of the hearing loss, age at identification, intervention for the loss, and family support and resources (Cole & Flexer, 2007; Sininger et al., 2010). The earlier the onset of the hearing loss, the more significant the loss, and the fewer support resources increase the risk for the child's success. A hearing loss may cause a student to struggle to organize, analyze, and retain information presented in the classroom. All of these factors will have an effect on the child's development of language, concept formation, executive functioning, working memory, and auditory processing skills.

Language. Language achievement is the primary area of development that is affected in children who are deaf or hard-of-hearing (Koehlinger, Van Horne, & Moeller, 2013; Tomblin, Oleson, Ambrose, Walker, & Moeller, 2015). Many of the struggles that students who are deaf or hard-of-hearing will face in school are attributable to their foundation of language functioning (Easterbrooks & Estes, 2007; Geers & Hayes, 2010; H. Hayes et al., 2014; Kelly et al., 2003).

In a study that examined spoken language scores of children using cochlear implants, Geers, Moog, Biedenstein, Brenner, & Hayes (2009) found that language development is the foundation for academic success. These researchers went on to note that language development is correlated with earlier academic mainstreaming. With age-appropriate language comes more age-appropriate academic performance. With early cochlear implant activation, early intervention, and high family involvement, many of these children are able to learn age-appropriate spoken language by 5 years of age (Geers et al., 2009; Kennedy et al., 2006; Moeller, 2001; Moog & Geers, 2010). This would allow them to enter into a mainstream classroom in pre-school and the early primary grades (Francis, Koch, Wyatt, & Niparko, 1999; Geers & Brenner, 2003). It is important to remember that though the cochlear implant has allowed many children to successfully access listening and spoken language, not all children who receive cochlear implants achieve age-appropriate spoken language by school age (Geers et al., 2009). Geers and colleagues also found a correlation between a delay in specific language skills and experiencing a disadvantage in the mainstream classroom (2009).

Among children who wear hearing aids, Koehlinger and colleagues (2013) found that any degree of hearing loss placed the child at risk for language delays. The children studied produced a shorter Mean Length of Utterance and had less competency with verb-related morphology (Koehlinger et al., 2013). These researchers found that age of amplification was a predictor of

Mean Length of Utterance, implying that early identification programs have a positive influence on language outcomes (Koehlinger et al., 2013).

Tomblin and colleagues (2015) examined language and speech outcomes for children who have mild to severe hearing loss and use hearing aids. Their findings indicated that early provision of hearing aids to children combined with good audibility from the hearing aid and a longer opportunity to wear the hearing aid were associated with better speech and language development.

Studies by Moeller (2001) and Kennedy and colleagues (2006) corroborated this. These researchers evaluated language outcomes in school-aged children with a wide range of degree of hearing loss. They both found that early access to intervention enhanced the language skills of these children (Kennedy et al., 2006; Moeller, 2001; Sininger et al., 2010).

Children with hearing loss can expect to present with delays in speech and language. Regardless of type of device used, the best outcomes in speech and language production were seen in children who had early access to amplification and early intervention services.

Neurocognitive factors. Children with hearing loss experience difficulties in the area of neurocognitive factors. These factors include executive function, processing, working memory, sustained attention, and cognitive flexibility skills. One determiner in the development of these executive functions is concept formation. Castellanos and colleagues describe concept formation as “a core foundational component of human information processing that underlies many higher-order cognitive and linguistic functions such as controlled attention, reasoning, abstraction, and the ability to compare” (2014). This has implications in the areas of reading, written expression, and mathematical applications (Castellanos et al., 2014). These academic skills require competence with organizing ideas conceptually so that the passage or problem may be completed

in a logical, coherent way (Castellanos et al., 2014). The ability to organize materials, ideas, and plans is imperative to executive functioning that is used in academic and general life settings (Gioia, Isquith, Guy & Kenworthy, 2000).

Concept formation is based largely on children's experience, reasoning skills, and language development (Nelson, 1996). Because of this, "children with sensory and/or language delays and disorders may be at high risk for disturbances in the development of concept formation" (Castellanos et al., 2014). In the subject area of reading, for instance, children are expected to learn the fundamentals of reading and then comprehend what has been read. This is based on background experiences, language proficiency, the ability to draw parallels, draw associative relationships, make inferences, remember what has already been read, and employ repair strategies (Easterbrooks & Estes, 2007; Geers & Hayes, 2010). Easterbrooks & Estes claim, "One's ability to think is integrally associated with one's language" (2007). Based on this statement, the comprehension of text proves to be more difficult for students who are deaf or hard-of-hearing than for a child with well-established language skills.

Language and concept formation is, furthermore, integrated into a subject like mathematics. Kelly and colleagues (2003) noted that students who are deaf or hard-of-hearing experience much difficulty with mathematical problem solving, especially when attempting word problems (Kidd & Lamb, 1993; Kidd, Madsen, & Lamb, 1993; Rudner, 1978). The mental rigidity and language delays children who are deaf or hard-of-hearing experience affect their metacognition and ability to integrate higher-level concepts. This makes equation and multi-dimension problem solving less fluid and facile (Kelly et al., 2003).

Children who are deaf or hard-of-hearing often experience a lack of auditory language exposure and a degraded auditory signal, which impacts the neurocognition, language, and

executive functioning of these children (Geers & Sedey, 2011; Hauser & Marshark, 2008; Peterson, Pisoni, & Miyamoto, 2010; Pisoni, Kronenberger, Roman, & Geers, 2011). This early sensory deprivation may cause disturbances in higher processes, "such as higher-order relational concepts" (Castellanos et al., 2014; Johnston, 2004; Luria, 1973). Castellanos et al. found that deficits in executive functioning and language correlated with delays in the concept formation of children who are deaf or hard-of-hearing and use cochlear implants.

Intervention Strategies and Techniques for Children who are Deaf or Hard-of-Hearing

Teachers of the deaf are trained to intervene and mitigate the consequences of the deficits caused by hearing loss outlined above. Teachers of the deaf do this through a variety of skill building techniques, explicit academic remediation, and the implementation of classroom accommodations. Incorporating all of these interventions into practice allows deaf educators to target the various factors that hearing loss has on classroom success.

First and foremost, it is important that children who are deaf or hard-of-hearing on the listening and spoken language path be given good access to sound; this will be the starting point for all other skill building. Audiologists will equip students with an assistive listening device, such as a cochlear implant, hearing aid, or frequency-modulation (FM) system. These assistive listening devices provide students with the sound needed to listen to and learn oral language. Once these children are given access to a usable stimulus, the audiologist and teacher of the deaf must directly train the user in how to manage his or her device(s) and interpret the signal he or she is receiving. For new listeners, this is done primarily through auditory training.

Auditory training. Auditory training is described by Ferguson & Henshaw as "teaching the brain to listen through active engagement with sounds, whereby listeners typically learn to

make perceptual distinctions between sounds presented systematically" (2015). These researchers claim that auditory training can help compensate for a degraded auditory input and results in improvements in hearing, competing speech, and complex cognitive tasks and executive functions that can influence attention and working memory (2015).

A teacher of the deaf may serve as the primary auditory trainer. Auditory training seeks to move children through the stages of auditory skill development. These developmental stages are based on Erber's levels of listening: detection, discrimination, pattern perception, identification/recognition, and comprehension (Erber, 1982). The skills become more complex as children move through the hierarchy. Other auditory skills that may be targeted during auditory training may include auditory memory practice, comprehension in background noise, localization of sound, media skills, gender and emotion identification/comprehension, and music appreciation.

Progression through the auditory skills hierarchy can be assessed several ways. Audiologists should perform aided detection and speech perception testing. There are also several standardized measures that can assess auditory skill development, such as the *Early Speech Perception Test* (Central Institute for the Deaf, 2012) and the *Infant-Toddler Meaningful Auditory Integration Scale* (Zimmerman-Phillips, Osberger, & Robbins, 2000). While these standardized measures are helpful in determining a child's present levels of functioning, teachers of the deaf should focus most of their attention to diagnostic therapy and the implementation of auditory skills programs. Curricula include the *Speech Perception Instructional Curriculum and Evaluation* and *Speech Perception Instructional Curriculum and Evaluation for Life*; the *Cottage Acquisition Scales for Listening, Language, and Speech*; the *Auditory, Speech, and Language*; and the *Miami Cochlear Implant, Auditory, and Tactile Skills Curriculum*. Teachers of the deaf

may use these programs in conjunction with other therapeutic techniques to help children manage real-life listening situations. The goal is for children to carry over these skills and learn through listening. Niparko reinforces the importance of making listening meaningful by integrating listening skills with other communication skills: "For this reason it is helpful to utilize material from school curricula as stimuli during auditory sessions" (2000). He lists examples of using vocabulary or spelling words as closed-set stimuli or social studies articles to prompt discussion (Niparko, 2000). Though direct auditory training is important, students must understand that listening is meaningful.

Classroom accommodations. In addition to direct intervention, a teacher of the deaf needs to ensure that the classroom listening environment is optimal for the student's listening and learning.

Signal-to-noise ratio (SNR). In order for students to best receive, interpret, and learn from the information being presented in the classroom, they must be in an environment that provides a favorable signal-to-noise ratio. The American Academy of Audiology Clinical Practice Guidelines defines SNR as, "the amount in decibels by which the amplitude of the desired signal (usually speech) exceeds that of an interfering signal" (American Academy of Audiology, 2008). Classrooms are not naturally conducive to a good SNR. Interfering signals might include the air conditioner, papers rustling, whispers, students shuffling in the hallways, chairs and desks being pushed, etc. "While the normally developing child may be able to cope with a sub-optimal acoustic environment, others may not. Children/youth with deficits of hearing, language, auditory processing, attention, or learning, and for children/youth listening in a non-native language, a poor acoustic environment adds an unacceptable burden, with significant negative effects on learning and development" (American Academy of Audiology,

2008). Teachers of the deaf can team with the audiologist and general educator in order to execute environmental modifications that enhance the SNR in the classroom.

Acoustically treating classrooms. The teacher of the deaf can acoustically treat general education classrooms. Extra noise, distance between the listener and speaker, and reverberation all make listening in a typical classroom difficult. Acoustically treating the classroom mitigates these barriers to listening and learning. DeConde Johnson (2003) lists some of these modifications to include the following: installing acoustic tile ceiling, adding carpet or rugs to the floor, gluing rubber tips or tennis balls to the legs of chairs, hanging curtains on the windows, putting corkboard on the walls, and keeping windows and doors to hallways closed. Acoustically treating the classroom can have a significant impact on SNR. Perhaps the best accommodation, however, is the provision of a hearing assistive technology (HAT) system.

HAT system management. HAT is recommended for children who have difficulty listening in noise. There are currently four types of HAT systems: personal, soundfield, induction loop, and infrared (American Academy of Audiology, 2008). The use of HAT systems in the classroom allows the speaker's voice to be sent from the microphone (i.e., transmitter) to the listener via a soundfield or personal receiver. This direct transmission of the signal in this manner overcomes distance, noise, and reverberation in the classroom.

The provision of these systems is written into a child's Individualized Education Program (IEP). While only audiologists are qualified to fit and program a child with a HAT system (American Academy of Audiology, 2008), teachers of the deaf are able to use and provide basic HAT system troubleshooting.

The Neurofibromatoses

The interventions and services provided by deaf educators have a positive influence on their students. But could they work for other populations of students who face similar challenges in the classroom? Individuals with NF represent an under-recognized group of students that could benefit from the specialized services of teachers of the deaf. The similarities between children are deaf or hard-of-hearing and children with NF could enable deaf educators to serve as the bridge between the clinical and education services these students need regardless of a concurrent diagnosis of a hearing loss.

As previously noted, the three types of NF (NF1, NF2, and Schwannomatosis) each have different diagnostic criteria and clinical manifestations (Gutmann et al., 1997; MacCollin et al., 2005). Individuals with Schwannomatosis do not present with learning disabilities, and will therefore not be discussed in this paper.

Neurofibromatosis type 2 (NF2). NF2 affects approximately 1 in 38,000 persons (Lloyd & Evans, 2013). NF2 is an autosomal dominant disorder, caused by a genetic mutation in the *NF2* gene on chromosome 22 (Lloyd & Evans, 2013). The defining hallmark of NF2 is the presence of bilateral eighth nerve schwannomas, occurring in more than 95% of patients (NIH, 1988; Plotkin et al., 2013). In addition to vestibular schwannomas, individuals with NF2 are prone to schwannomas affecting other nerves, meningiomas, ependymomas, and cataracts.

Vestibular schwannomas are one of the primary clinical features of the disease. Surgical resection of the tumor frequently results in hearing loss (Briggs et al., 1994; Shepard, Tucci, Grant, & Kaylie, 2012). Even in situations where preservation of hearing is not possible, newer technologies exist to restore hearing capacity in these individuals. These include the use of hearing aids, cochlear implants, auditory brainstem implants. The treatment of individuals with

NF2 requires a multi-disciplinary medical team (Gutmann et al., 1997; Lloyd & Evans, 2013; Shepard et al., 2002). Team members typically include neuro-oncologists, ophthalmologists, otolaryngologists, neurosurgeons, neurologists, pathologists, radiologists, and audiologists (Gutmann et al., 1997; Lloyd & Evans, 2013). A teacher of the deaf may also be on the team for a child with NF2 if a hearing loss presents. The role of the deaf educator would be similar to the role played for children with other etiologies of hearing loss.

Neurofibromatosis type 1. NF1 affects approximately 1 in 3,000 individuals worldwide (NIH, 1988; Hirbe & Gutmann, 2014). The diagnosis of NF1 was established in 1987, and requires two or more of the following features:

1. Six or more café au lait macules (i.e., pigmented birthmarks) at least 5 mm in greatest diameter in prepubertal individuals and over 15 mm in greatest diameter in postpubertal individuals
2. Two or more neurofibromas of any type *or* one plexiform neurofibroma
3. Freckling in the axillary (i.e., underarms) or inguinal (i.e., groin) regions
4. Optic pathway glioma (i.e., a tumor that starts in the brain or spine, affecting the optic nerve)
5. Two or more Lisch nodules (iris hamartomas) (i.e., mass of disorganized tissue projecting off of the iris)
6. A distinctive osseous lesion such as sphenoid dysplasia (i.e., widening of the sphenoid wing in the orbit) or thinning of long bone cortex, with or without pseudarthrosis (i.e., bone fracture incapable of healing itself)
7. A first-degree relative (i.e., parent, sibling, or offspring) with NF1 by the above criteria

Children and adults with NF1 are at risk for developing a variety of benign and malignant tumors, including brain and nerve tumors. In addition, there are a number of cognitive and behavioral problems in children with NF1. As such, visual-spatial perception deficits are frequently observed (Billingsley, Slopis, Swank, Jackson, & Moore, 2003; Gilboa, Rosenblum,

Fattal-Valevski, & Josman, 2010; Hofman, Harris, Bryan, & Denckla, 1994; Hyman, Shores, & North, 2005; North, Hyman, & Barton, 2002; Ozonoff, 1999). In addition, academic difficulties are seen in the areas of language, spelling, reading, writing, math, attention, auditory temporal processing, working memory, and executive functioning (Billingsley et al., 2003; Gilboa et al., 2010; Hofman et al., 1994; Hyman et al., 2005; North et al., 2002; Ozonoff, 1999). These learning disabilities occur despite the absence of mental retardation (Gutmann et al, 1997, Hofman et al., 1994, North et al., 2002).

Neurocognitive deficits. Children with NF1 have been observed to present with poor planning and organizational skills (North et al., 2002). On standardized tests measuring recognition and recall, visual attention, task switching, and mental flexibility, children with NF1 performed lower than the general population (Chapman, Waber, Bassett, Urion, & Korf, 1996; Ferner, Chaudhuri, Bingham, Cox, & Hughes, 1993; Hofman et al. 1994; Joy, North, & deSilva, 1995; Ozonoff, 1999; Zöller, Rembeck, & Backman, 1997). Children with NF1 have trouble with executive functions, including cognitive flexibility, inhibition, working memory, shifting, abstract concept formation, and sustained, switching, and selective attention (Gilboa et al., 2010). Other studies, however, have noted that these executive deficits, with the exception of sustained attention, do not impact the student beyond the correlation with the child's IQ score (Hyman et al., 2005; North et al., 2002). Even when IQ is controlled for, however, children with NF1 still experience attention problems that may compromise performance in many areas (North et al., 2002). Though children with NF1 experience verbal and non-verbal impairments that have negative impacts in classroom performance, the treatment plan for these individuals is mainly clinical (Ozonoff, 1999).

In reference to children who are deaf or hard-of-hearing, others have noted that difficulty with abstract concept formation might partly explain the challenges with cognitive flexibility, working memory, and the other executive functions, such as attention and inhibition (Castellanos et al., 2014). These investigators also found that prelingually deaf CI users were delayed in concept formation compared to normal hearing controls, despite no discrepancy in IQ (Castellanos et al., 2014). Similarly, Gilboa and colleagues (2010) demonstrated that abstract concept formation and lack of cognitive flexibility is present in children with NF1.

Language. Children with NF1 frequently present with problems in expressive and receptive language skills (Billingsley et al., 2003; Gilboa et al., 2010; Hyman et al., 2005; North et al., 2002; Ozonoff, 1999). In this regard, there are sensory and/or language delays that result in disturbances in the development of concept formation (Castellanos et al., 2014). This also has a direct impact on vocabulary, written language, reading fluency and comprehension, phoneme segmentation, phonologic memory, and naming (Gilboa et al., 2010; Hyman et al., 2005; North et al., 2002). Mazzocco and colleagues (1995) found that children with NF1 display deficits in word retrieval, knowledge of grammaticality, rapid automatized naming, receptive language, phonemic segmentation, phonologic memory, reading achievement, written language, and mathematical achievement. Similarly, Dilts and colleagues (1996) reported struggles with overall language performance and language structure, reading achievement, written language, and mathematical achievement. Finally, North and colleagues (1994) noted areas of difficulty included overall language performance and language structure, reading comprehension and fluency, spelling, and mathematical achievement. Unlike executive function, when IQ is controlled for, children with NF1 still present with deficits in reading, spelling, planning, visual perception, and sustained attention (Hyman et al., 2005).

Management of NF1. It is recognized that children diagnosed with NF1 experience academic learning challenges. However, the greatest advances in clinical care have been in the area of tumor management, with less focus on learning disabilities in this population. This can be hindered by the fact that some of the diagnostic criteria may not be develop until later in a child's development. Since a diagnosis of NF1 cannot occur until two or more of the identifying criteria are met, some children may not receive a diagnosis until later in adolescence. Children with NF1 have an increased risk of using special education services and receiving remedial teaching for learning, behavior, speech, or motor problems (Gilboa et al., 2010). One group of researchers found that 50% of children with NF1 had to repeat at least one grade in school (Coudé, Mignot, Lyonnet, & Munnich, 2007). Because of the heightened incidence of learning disabilities in this population, children with NF1 should be evaluated and early intervention provided as promptly as possible. Furthermore, someone skilled in these domains should evaluate and monitor progress throughout the academic careers of these students. Many children with NF1, however, are not acknowledged to have learning disabilities (Ozonoff, 1999). Much of children with NF1's school failure is related to executive function difficulties in the areas of attention, cognitive flexibility, and emotional regulation (Kusnyer & Standberry, 2013; Ozonoff, 1999). If an educator is not trained in how to identify these difficulties, they may go unrecognized and untreated. Furthermore, because these problems are misunderstood, students can be viewed as being "willfully disobedient" and will rarely receive the special education services necessary to remediate these problems (Kusnyer & Standberry, 2013; Ozonoff, 1999).

Children who are deaf or hard-of-hearing are more easily able to qualify for services than children who have less identifiable learning disabilities, such as an executive functioning deficit. Not only do children with hearing loss have outward physical proof of their disability (e.g., the

wearing of listening devices), but the literature has demonstrated that children with hearing loss will often experience some a language delay that has a negative impact on the child's academic performance (Connor, Craig, Raudenbush, Heavner, & Zwolan, 2006; Easterbrooks & Estes, 2007; Francis et al., 1999; Geers, 2003; Geers & Brenner, 2003; H. Hayes et al., 2014; Geers & Hayes, 2010; Johnson & Goswami, 2010; Joshi et al., 2008-2009; Kelly et al., 2003; Rudner, 1978; Kidd & Lamb, 1993; Kidd et al., 1993; Spencer, 2004; Spencer & Oleson, 2008; Traxler, 2000). Although there must be proof that the hearing loss has an adverse affect on educational progress, children who are deaf or hard-of-hearing usually qualify for services from a teacher of the deaf due to the toll that language impairments can have on academic performance. Because children who are deaf or hard-of-hearing have an explicit disability that is easily recognizable, it ensures that nearly all children who are deaf or hard-of-hearing receive services as outlined on their Individualized Education Programs (IEPs) in accordance with the Individuals with Disabilities Education Act (IDEA).

The Teacher of the Deaf's Role

Teachers of the deaf are trained to teach and support children who are deaf or hard-of-hearing in a specific manner in order that these students may overcome academic challenges presented by language and concept formation delays. The teacher of the deaf often collaborates with the audiologist and general educator in order to provide the child with a strong usable signal so learning in a classroom may be facilitated. A teacher of the deaf works directly with a child who is deaf or hard-of-hearing to give explicit instruction across multiple academic areas, provide auditory training, coordinate HAT system use in conjunction with the audiologist, acoustically-treat general education classrooms, and advocate for any additional

accommodations (Cunningham et al., 2001; DeConde Johnson, 2003; E. Hayes, Warrier, Nicol, Zecker, & Kraus, 2003; Hornickel & Kraus, 2013; King, Warrier, Hayes, & Kraus, 2002). The provision of these services has proven successful for children with hearing loss (Easterbrooks & Estes, 2007; Geers & Hayes, 2010; H. Hayes et al., 2014; Joshi et al., 2008-2009; Kelly et al., 2003).

These accommodations are also successful for children with other learning disabilities and diagnoses (Hayes et al., 2003; Hornickel & Kraus, 2013; King et al., 2002). For instance, children with a diagnosis of Autism Spectrum Disorder, Dyslexia, and learning disabilities have a diminished capacity for processing speech and noise (Cunningham et al., 2001; Hornickel & Kraus, 2013; Russo, Zecker, Trommer, Chen, & Kraus 2009). The inclusion of a HAT system in classroom settings has been proven to mitigate the negative effects of impaired processes in these populations of students (Hayes et al., 2003; Hornickel & Kraus, 2013; King et al., 2002). Hayes and colleagues (2003) and King and colleagues (2002) showed that auditory training results in improved neural responses. When put in a meaningful context, these improved listening and communication skills will help serve all children in general education settings (Niparko, 2000).

Deaf educators also incorporate remediation strategies and tools into the child's academic program to help compensate for deficits in executive function that may affect classroom performance. For instance, a child who is deaf or hard-of-hearing with an executive dysfunction in the area of cognitive flexibility and shifting may struggle to balance main ideas and supporting details in written language. A teacher of the deaf may incorporate a tool such as a graphic organizer to help the child order the information (Kusnyer & Standberry, 2013). For a child with working memory deficits, a teacher of the deaf might help the child compensate by giving instructions in smaller chunks as opposed to long strings of information (Kusyner & Standberry,

2013). Teachers of the deaf use these types of strategies to help overcome the executive function challenges faced by children who are deaf or hard-of-hearing. These tools and techniques can be applied to all children with executive function deficits, however (Kusyner & Standberry, 2013).

Conclusion

The goal of this literature review was to describe what NF is, the scope of its impact, how NF relates to hearing loss, and why a teacher of the deaf might have some information to offer the intervention team for a child diagnosed with NF1. Though hearing loss is not typical in individuals with NF1, these students experience a variety of academic challenges that may be rooted in processing disorders and language deficits. If this is the case, then these children should receive language-based intervention. Additional therapies and classroom accommodations should also be considered in the treatment of students with NF1. The literature is still sparse in the areas of academic functioning and management for school-aged children with NF1. However, the implementation of strategies that have proven to help children who are deaf or hard-of-hearing overcome academic, language, executive function, concept formation, and processing difficulties may also be found to be helpful in the treatment of children with NF1. The placement of children in an acoustically treated classroom, the inclusion of a HAT system, and the incorporation of executive function remediation strategies are accommodations and interventions that should be considered in the academic management and treatment plan for students with NF1. Further research is needed to determine the role a teacher of the deaf may have in educating other professionals in specific treatment plans and interventions that may benefit different populations of students.

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