CHAPTER NINETEEN

Supporting Families of Children with Auditory Neuropathy Spectrum Disorder

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Abstract

Universal agreement on best treatment options for children with auditory neuropathy spectrum disorder (ANSD) continues to be challenging for professionals and parents. Families may face contradictory information regarding the diagnosis, choices in communication, amplification, and intervention services for their child. This presentation will share “what works for families”, helps them begin to understand the complexity of ANSD and gain the confidence to become full partners in their child’s care as well as pitfalls to avoid while counseling families. This paper will discuss perspectives and stories from parents, looking back as their children transitioned from the early years to preschool, and then to elementary school and beyond.

Parents of children who are deaf or hard-of-hearing face many difficult and confusing issues and decisions. These include the anxiety of referral from newborn hearing screening, the unanticipated diagnosis of permanent hearing loss, the variety of options for amplification, communication methodology, and intervention services in their community. These issues are confounded when the child has a diagnosis of auditory neuropathy spectrum disorder (ANSD). These families may receive contradictory information about diagnosis, treatment, and outcome expectations. They may hear that “recovery” from this disorder is possible and they may observe fluctuations in their child’s hearing ability.

Supporting families of children with ANSD with a team of professionals is critically important for successful outcomes for the child and family. Professionals with a variety of perspectives and expertise can help the family understand the range of options available and support the decisions the family must make. At Bill Daniels Center for Children’s Hearing, located in the Children’s Hospital Colorado, the comprehensive support team for families includes: pediatric otolaryngologist, genetics counselor, audiologist, speech language pathologist with expertise in hearing loss, social worker, deaf educator and the family consultant.

The Family Consultant typically meets with families after the confirmation of hearing loss during scheduled appointments with the audiologist. This allows the family to meet and access support in the moment without having to schedule additional appointments. She provides families with resources and information about hearing loss, communication options, facilitates connections in the hospital and the family’s home community and lends a personal perspective into the day to day issues of life with hearing loss.

To understand the perspective and experiences of parents, the Family Consultant conducted interviews with twelve parents whose children with ANSD received ongoing care at Bill Daniels Center for Children’s Hearing. Families of children aged 2 years through 12 years were interviewed. The families were asked about their experience with the diagnosis and recommendations for intervention. They were also encouraged to consider the challenges they faced as their child transitioned through important milestones such as moving from early intervention to preschool, preschool to elementary school, and middle school and beyond when giving feedback.

Characteristics of the children whose parents were interviewed are summarized below.

- 13 children; one set of twins
- Seven boys/six girls
- Ages: 2.8 years through 12 years old
- Three children were adopted
• Four children with global developmental delays including one with autism
• One multi-generational family with deafness; communication mode is both ASL and spoken English. The four children in the family have hearing loss but only one has AN
• Four children reside in rural Colorado.
• Six children have cochlear implants
• Two children are fulltime consistent hearing aid users
• One child has used hearing aids intermittently
• One child uses ASL and does not access technology

Parents were given the opportunity to visit by phone, email or meet in person. All 12 families contacted agreed to participate and share their experiences parenting a child with ANSD. The following questions were presented to the parents:

• What support or information was helpful to your family after you learned about your child’s diagnosis of ANSD?
• What do you wish the professionals would have done differently?
• Looking back, is there anything that you would have done differently for your child?
• What advice would you give to families with children who have auditory neuropathy?
• Are there any memories or experiences where you could see how AN impacted your child at home or school?

The following experiences were common to most families:

• Parents desired to know what their child was hearing
• Parents stated that professionals need to be confident in their information or comfortable in referring families. One parent shared, “I felt that I have always had to seek out information myself and then find the professionals who were willing to help me.”
• Parent to parent support is critical and alleviates the desire to search the internet for that connection and for information which may not always be accurate or current. “I was sad when I read on the internet about some cases of ANSD resolving and I had second thoughts about a cochlear implant.” Sharing that information with the audiologist is important but there is comfort in realizing that other parents also have had the hope that their child’s ANSD would resolve.
• Parents felt that it was their job to constantly educate others; “I’m always explaining what auditory neuropathy is to family members, friends and those that work with my child.”
• Professionals may not be knowledgeable about all of the tools for communication.
• Consistent and effective communication was an ongoing challenge.
• “People tend to underestimate my child.”

The statement that “People tend to underestimate my child” was shared by several parents regardless if the child had delays or if the child is currently reading above grade level or had a sophisticated understanding of language. What might professionals be implying or doing that could make a parent feel as if their child is being underestimated? Do professionals value input and observations from parents? Are the decisions and choices of parents being honored? What strategies can professionals employ to increase parent understanding of ANSD while maximizing the child’s strengths and skills?

“Children with ANSD can develop into healthy and dynamic citizens with happy personal lives, successful academic experiences and satisfying careers. Clinicians should help families realize this goal by identifying and supporting the unique strengths and abilities of the child and family.” (ANSD Guidelines, 2008).

Experiences During Transitions:Birth to Three Years

As families looked back at the birth to three year time period, several parents were able to see the positive support that they received from their providers, especially their audiologist. Time restraint is an issue that audiologists are faced with while counseling families with ANSD. However families reported that the time spent counseling during those early days turned out to be invaluable. “The time spent in counseling was time well spent.” Another parent shared, “Audiologists should never underestimate the time spent visiting outside of the booth such as emails, and phone calls.” Other helpful comments and strategies that were expressed and considered crucial for families during the birth to three year period included:

• The need to understand the diagnosis of their child’s hearing loss
• To reiterate the diagnosis of ANSD and what their child’s audiogram means. This should take place during multiple appointments. Families reported that it took a year or two for the information about ANSD to make sense to them.
• “Maybe this will get better.” The desire for the child’s hearing loss to get better was expressed repeatedly by the families interviewed and even families with older children. One child was almost 12 years old and the family still held onto the idea that the hearing loss might improve. Another family shared that “This (hearing tests and audiograms that varied) could be part of God’s plan to heal our child.” How do audiologists encourage parents to make empowering decisions with their faith and their treatment team? If professionals attempt to draw a line between the two then treatment may become adversarial for the family. Acknowledging the values of a family may need to occur during an audiology appointment. Audiologists will learn more about families by not passing judgment, especially in situations where recommendations are not readily followed based on family values. The family may benefit from a conversation with a social worker, counselor or chaplain. Children’s Hospital Colorado has a chaplain with a personal perspective on hearing loss and is available to lend insight to the family’s desire to make faith based decisions.

• Health issues and graduating from the NICU was the most important for several families. Families with babies who had difficult neonatal complications wanted to be able to go home and wait with follow up and frequent hearing tests. Hearing is not always a priority for families with babies that struggle with health issues. Global delays also often made it difficult for some families to be consistent with hearing aid trials or get a sense of the benefits of hearing aids in the home environment.

• “It was hard to be told that my observations were wishful thinking.” Early interventionists and audiologists desire and ask for parental feedback and observations. Information from parents needs to be valued and incorporated into the visit even if it doesn’t correlate with the results of the audiogram.

• Communication options – professionals should have knowledge and understand the value of all communication options. Families appreciated honest conversations about what the best options for the best outcomes are rather than a “wait and see” approach and with ongoing testing. One family in this interview did not have a desire to incorporate sign language and felt confident that their son would do well with spoken language. The young boy has done well but must have full access to lip-reading which in retrospect serves as a visual mode for his communication needs. Another child with spoken language used cued speech while growing up and has recently added sign language for social connections with his peers. The parents of these two boys shared that as they looked back; they felt that they had to justify their communication choice to the early interventionists and providers during their child’s early years.

• Parents shared that conflicting information made it difficult to have confidence in their providers unless there was a strong team in place. Recommendations were welcomed when the child was being monitored by a team that included a variety of professionals.

• “I wish the referral for a vision screening was strongly emphasized over several of our appointments.” Parents with little one ones that needed glasses stated they didn’t realize how important vision was for children using a visual mode of communication.

• “In the early years I didn’t understand the value of collaboration until my child started school and then I was glad to have had that experience to meet my child’s school needs.” Audiologists that strive for successful teaming in the early years help parents become skilled in advocating when the child enters school.

• Clinicians with an expertise in working with children who have hearing loss were an invaluable resource for parents and children that attended toddler or preschool groups.

• “I was so scared to lose what hearing my child had.” Confidence in technology was slow to emerge during the birth to preschool years. Multiple trials with hearing aids were needed. Parents that later opted for cochlear implants stated that it was still difficult to give up what hearing their child had even when the language wasn’t progressing with therapy.

Experiences During Transitions: Preschool Years – Three to Five Years

• Three of the mothers shared that their preschoolers were more dependent on them for clarification and comfort in social situations than typical preschoolers. While the sensitivity and attentiveness of these mothers gave their child increased access to communication it became more difficult as the child entered the school years. A mother of a seven year old child expressed her concern and desire to stay with her child during activities at a weekend camp for children with hearing loss. The staff shared that access to communication would be provided in every way at every camp activity, day and night; from fishing to horseback riding and the evening campfires. When
her child saw the other campers and ran off to join them, she shared that “In hindsight, finding different ways to help my child understand what was going on would have been better than being her interpreter during those early years and in tough situations.”

- Developmental delays become more obvious as the child entered preschool. “I began to notice that the language and speech delays were more apparent as he entered preschool and when he tried to play with the other preschoolers.”

- Skills in advocacy and parental involvement became critical during the preschool years. A parent said that his advice to families would include: “People will help your child if they know you. Get to know people at your child’s school!” Confidence and knowledge about ANSD began to emerge for parents during the preschool years. This became crucial as parents began to advocate for their child’s school needs and get those needs written into the Individual Education Plan for school.

- Decisions for technology were typically made during the preschool years. “I wish we had done the cochlear implants sooner. She loves her cochlear implants (CIs) but I was scared to lose what hearing my child had.” Two of the girls from this group were implanted at the age of four and enjoyed hearing with cochlear implants. Both girls had several hearing aid trials. One did not consistently wear her hearing aids and the other child refused to wear her hearing aids. Upon activation of her CI’s one child went through a period of time when she would say good night to her cochlear implants. The other girl was so excited about hearing that when she was in her room after surgery she kept saying, “I hear birds!” “I hear my heart.” Of course those sounds were not yet present but her parents had done a nice job of talking about the possibility of hearing new sounds and she was eager to hear those sounds.

**Experiences During Transitions: Elementary School - Six Year to 11 Years**

- During this stage, advocacy needs to move from the parents advocating for their child to self-advocacy skills for the child and be incorporated into the child’s Individual Education Plan.

- The elementary school age child needs to become familiar and aware of when they are not hearing well and when they are hearing well. They need to learn what optimal hearing situations look like for them.

This is not only challenging for this age group with ANSD but also their parents who continue to desire and know what their child is hearing. Parents should share what they themselves are hearing which helps for comparison and discussion about different sounds. One father said that he had been so focused on what his son couldn’t hear that he never shared what he himself was hearing when they were hiking or playing sports together.

- Strategies for tough days and plans need to be in place especially as the challenges of hearing in background noise take place on a daily basis for elementary age children. For the older child, there is comfort in knowing what to do when listening environments are tougher than usual.

- Creative outlets, sports or quiet time (listening breaks) may help the older child on days when extra energy is required for hearing and communicating well. One child is very artistic and will draw his way through his stress. Another journals and writes imaginary stories and yet another child takes comfort in reading quietly in his room.

- Social skills require guidance and support from family members and people who are connected with the older child. Communication breakdowns need to be recognized by the older child (not just the parents) so that the child can attempt to repair social mishaps.

- Parents shared that as their child began to transition to the middle school years they revisited questions such as; “What are the goals for my child now? What can we expect and does our child have the right tools to get information taking place academically and socially? Are the current communication methods working for our child?”

**Experiences During Transitions: Middle School (6th grade) – 11 Years to 13 Years**

- School placement and support services may change from elementary school to the middle school years. The parent of a 12 year old said, “Schools are focused on what services they have to meet your child rather than what are the best services for your child.” I chose to move my child from the public school setting to a private school since smaller class sizes were more important than the services that the public school had to offer my child.”

- More schools are utilizing computers as a teaching tool. Older students with hearing loss have commented that they can’t hear fast enough when taking
tests on the computer. Practice tests and conversations between the student and the teacher need to occur to examine if less than optimal grades are the results of hearing or academic performance.

- Older students may demonstrate embarrassment using an FM system. It may be time to evaluate the possibilities of note takers and wearing an FM or note takers vs. the FM system. The ease of a note taker allows the child to sit back, listen and absorb the information which may help the student focus on academic skills.

- Audiologists may be able to suggest useful strategies that assist students in obtaining missed information and seek clarification. Consistently asking the student “What did I say? Or “Did you hear me?” puts the child in a position to not tell the truth. Children this age do not want to be wrong. Asking “What did you hear” prepares the students to understand that what they hear isn’t always what was said.

- Teachers should ask the child next to the student with hearing loss for repetition to avoid consistently asking the student with hearing loss to repeat directions that were given.

- “The fluctuations in his hearing are evident in his temper when he comes home from school”. Information begins coming at a faster pace during these school years. Keeping up can be more difficult and lead to frustration especially at home when extra patience is required to get along with siblings. “I can hardly wait to be old enough to live by myself” commented a 12 year old boy when he complained that it was too hard to be with his brothers.

- “Everybody is always talking about me and my hearing.” Empower this age group to take ownership of their hearing loss by assisting them in understanding their hearing loss should be considered a goal. Creating biographical power points and getting the student involved in their own Independent Education Plan (IEP) may help students become aware of their hearing and enhance self-advocacy skills. “She (the audiologist) is always writing notes about me during the visit”. Audiologists should inform the older student what they are writing when taking notes during appointments. Students this age should be at the center of the appointment as the audiologist shares results of the tests and the child’s audiogram.

- Independence/freedom/privacy is most important for the pre-teen and teenager. Audiologists may serve as a valuable resource by sharing ideas for parents of preteens that desire privacy or independence. Parents of children with hearing loss tend to enter the child’s room because they know that their child may not hear them knocking. Create a plan, crack the door and wave a hand through the cracked door or flick the light rather than knocking and walking right into a teenager’s room. Audiologists can model this in the clinical setting. When the teen is the patient, rather than gently knocking on the door when coming into a room during the appointment, knock loudly, or wave a hand through a door that is cracked open. Parents will attempt these strategies at home. Audiologists may be a source for suggesting ways to move the family and child towards independence by encouraging the preteen to use technology or assistive devices. One of the fathers shared how the family was able to let the son bike a distance from home with a friend as long as the boys used the FM system. Parents may share the frustration of waking teens up in the morning and not realize there are alarm clocks that are available for people with hearing loss. Two of the families shared that the value of the extra time their audiologist spent counseling them during the early years was beneficial once again during their child’s adolescence. Referrals for resources, assistive devices and connections with deaf/hard of hearing mentors or role models were needed as their youngster entered adolescence.

**Summary**

The following statements are taken from a report from a panel of distinguished scientists and clinicians at the Lake Como, Italy, 2008 Guidelines Development Conference on Auditory Neuropathy Spectrum Disorder (ANSD Guidelines, 2008):

“Counseling families of infants with this ANSD is one of the greatest challenges associated with this disorder. Because the developmental effects of ANSD cannot be predicted from test results obtained in the earliest months or even years of life, families struggle with the uncertainty of what the diagnosis means relative to their infant’s growth and development. Many infants with ANSD have had difficult perinatal courses with complications including prematurity, birth asphyxia, infections or other conditions requiring neonatal intensive care. The significance of the ANSD diagnosis may be difficult for families to appreciate as they struggle to understand their infant’s complex medical and developmental needs. Strong support systems, including parents of children with similar diagnoses and professionals with expertise in clinical social work and
family counseling, should be available to meet the ongoing and challenging needs of families.”

Parents interviewed for this paper reinforced the importance of a strong support system including a multi-disciplinary team of professionals. As professionals, regardless of our discipline, we can endeavor to give parents the confidence that their child with auditory neuropathy will do well with ongoing support, family involvement, quality services, and access to language and technology. As one mother so eloquently expressed, “Hope didn’t get me through understanding hearing loss and deafness. People did - people got me through it.”

Reference