

Letter from the ENT Director



The condition of auditory neuropathy is poorly understood and represents multiple challenges to our understanding of the physiology of the auditory system. Initially, most people have doubted that CI can work in these settings, but there is now a clear and compelling evidence in the literature as to the efficacy of cochlear

implants in auditory Neuropathy spectrum disorders. We will use this issue of our bulletin to discuss our experience with ANSD here at the CI center at the American Hospital-Dubai.

I would like to welcome and introduce to all Mr. Lee Bolton who has joined our ENT department. Lee is focused on the diagnostic and management challenges of swallowing and voice disorders. The Field of diagnosing and managing the different types of Dysphasia have advanced significantly over the last 10 years. Lee brings to our department tremendous expertise in this field including Fiberoptic Endoscopic Evaluation of Swallowing and Videofluoroscopic Evaluation of Swallowing (VFES). Please join me in welcoming Lee to our hospital and our city.

Dr. Muaz Tarabichi

ENT Director at the American Hospital Dubai

AUDITORY NEUROPATHY SPECTRUM DISORDER

Lubaina Sharafally, M.A. , CCC-A Clinical Audiologist

Hearing loss is a common problem in newborns. Some cases are due to auditory neuropathy spectrum disorder (ANSD), a problem in the transmission of sound from the inner ear that makes sound disorganized when it reaches the brain.

Someone with ANSD has difficulty distinguishing one sound from another and trouble understanding speech clearly. In some cases, ANSD causes only mild hearing difficulties or is only a problem in noisy situations. In others, it leads to significant hearing loss. The causes of ANSD are unknown, but children who are born prematurely or have a family history of the condition are at higher risk for it. Symptoms can develop at any age, but most kids with ANSD are diagnosed in the first months of life. As ANSD becomes better understood, it is diagnosed more frequently and now accounts for about 10% to 15% of cases of hearing loss.

What is ANSD? When someone has ANSD, sound enters the ear normally, but because of damage to the inner row of hair cells or synapses between the inner hair cells and the auditory nerve, or damage to the auditory nerve itself, sound isn't properly transmitted from the inner ear to the brain. As a result, the sound that arrives at the brain isn't organized in a way that the brain can understand. It is disorganized and in some cases the sound never even makes it to the brain. In other cases ANSD is due to a problem with the auditory nerve.

ANSD has only been understood and diagnosed in recent years. As a result, many questions remain about the condition. The symptoms of ANSD can range from mild to severe. Some kids with ANSD hear sounds but have trouble determining what those sounds are. For others, all sounds seem the same, like

static or white noise. For example, a voice might sound the same as water running, a dog barking might sound the same as a car horn, or a bird chirping might sound the same as a pan banging. For some, ANSD improves over time. For others, it remains the same or gets worse.

Even if a child passes newborn hearing screenings, symptoms of hearing problems might only become apparent over time. Symptoms such as :

- infant doesn't startle when there are loud or sudden noises, or turn toward sound
- baby isn't cooing, babbling, or laughing by 8 months
- child is not trying to imitate sounds and actions by 12 months or isn't responding to simple commands

A series of behavioral and neurological tests will help diagnose ANSD and rule out other hearing problems. Since ANSD displays auditory characteristics consistent with normal outer hair cell function and abnormal neural function at the level of the VIIIth (vestibulo-cochlear) nerve.

These characteristics are observed on clinical audiologic tests as normal otoacoustic emissions (OAEs) in the presence of an absent or severely abnormal auditory brainstem response (ABR).

If these tests show that the child has ANSD, over time other tests can determine how severe the condition is. Currently, no test

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can determine the severity of the condition in infants. While there is no known cure for ANSD, assistive listening devices (ALDs) can help kids with ANSD make sense of sounds and develop language skills. You'll work with a medical team to determine which devices are right for your child. Treatment for ANSD depends on how severe it is and the age of the child at diagnosis. A crucial part of making any device effective is ongoing therapy with a speech-language pathologist, who helps children with hearing loss develop speaking and hearing skills.

Fortunately, kids with ANSD can develop strong language and communication skills with the help of medical devices, therapy, and visual communication techniques. Proper diagnosis and early intervention are essential!

Pure tone thresholds:	Normal to severe/profound hearing loss (Any configuration; can be asymmetric)
Speech recognition in quiet:	Variable; slightly reduced to greatly reduced
Otoacoustic emissions:	Normal
Middle ear muscle reflexes:	Ipsilateral Absent Contralateral Absent Non-Acoustic Present
Cochlear microphonic:	Present (Inverts with stimulus polarity reversal)
ABR:	Absent (or severely abnormal)
Masking Level Difference (MLD):	No MLD (i.e., 0 dB)
Efferent Suppression of TEOAEs:	No suppression
Speech recognition in noise:	Generally poor

Table 1. Expected test results in auditory neuropathy patients.

Case Study:
Auditory Neuropathy Spectrum Disorder (ANSO)



Dr. Rana Batterjee, Au.D., CCC-A, ASHA-F, F-AAA,
Clinical Audiologist

In March of 2008, H.A., a 6 year old female was seen at this facility for a hearing evaluation. She was referred by her speech therapist because she was not doing well with hearing aids and sometimes

behaved like a deaf child. In addition, conflicting test results from different facilities had the family and therapist requesting a second opinion. Case history reported by her mother was significant for speech/ language delay and hearing loss initially diagnosed at the age of 1.8 years. H.A. did not speak and used gestures to communicate. Birth and development history were not significant.

H.A.'s mother indicated that she was not fitted with hearing aids until she took her to Thailand at the age of 4 years. The puzzling thing was that the family insisted that she could hear and understand without them. At the time of the appointment she was wearing only one aid which was functioning. The ear mold was too small and the battery was dead. Testing in Thailand indicated a severe loss in the right ear and a profound loss in the left with poor morphology bilaterally. She was also tested at different facility in the UAE one week earlier using Pure Tone Audiometry. Results indicated normal hearing sloping to moderately severe in the right ear and a profound loss in the left ear.

The family and therapist were puzzled by the diagnosis and the pattern of behaviors H.A. displayed such as:

- Responding to her parent's commands without visual cues
- Responding to the telephone/mobile phone ringing
- Dancing to music
- Identifying animal pictures but not the sounds they made

TESTING AT THE AMERICAN HOSPITAL DUBAI:

Conditioned Play Audiometry (CPA):

CPA was attempted on 18/03/2008. H.A. was difficult to test. Her responses were not consistent or reliable. She displayed hyper behaviour with a short attention span. Results for the right ear revealed a moderately severe loss rising to moderate with a Speech Awareness Threshold (SAT) of 40 dBHL. The left ear revealed a moderately severe loss with a SAT of 55 dBHL. An ABR was scheduled to verify results and rule out ANSD.

AUDITORY BRAINSTEM RESPONSE (ABR):

Immittance testing revealed type A tympanograms bilaterally. Acoustic reflexes were absent bilaterally. OAEs were present for the right ear and absent for the left. Click stimulus ABR waveforms were absent at an intensity level of 95 dBnHL bilaterally with the presence of a Cochlear Microphonic at 1 msec. The results were consistent with ANSD.

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INTERVENTION:

After extensive counseling on the issue of ANSD and setting realistic expectations for H.A., her family decided to go ahead with the recommendation of cochlear implantation. The process took five months.

HOOK UP:

- H.A. responded to live voice
- Sound field responses to warble tones were within normal limits
- H.A. was able to condition to respond to tones with good reliability which was very different from testing pre operatively when she was extremely unreliable.

3 months post Hook up:

- Uses Sound Processor during all waking hours
- Enjoys making a lot of noise.
- Attending speech therapy two to three times a week
- Attends school daily
- More attentive and quiets down in class
- Vocabulary starting to increase. She can pronounce two syllable words
- Improved behavior

6 months post Hook Up:

- Using words instead of signs and gestures
- Learning names: "Saif" "Fatum"

12 months after Hook up:

- Starting to use short sentences (2 to 3 words)
- Asking questions
- Progressing well in first grade in school for children with special needs

18 months post Hook up:

- Passed first grade
- Continues to attend speech therapy sessions twice weekly using visual cues
- Speech discrimination scores without visual cues is 50% at normal conversational level (50 dBHL) using the Ashoor Arabic Speech Test for Children

After cochlear implantation H.A.'s life improved dramatically. She is a happy little girl who loves her implant and uses her sound processor all day long. Her therapist reported that her vocabulary has expanded and although I may not be able to comprehend what she says 100% of the time I know that cochlear implantation was the correct therapy choice for her. I often wonder what life would have been like for her if she was implanted much earlier...

PERSPECTIVES FROM THE FIELD OF EARLY CHILDHOOD SPECIAL EDUCATION



Nadine Ewanchyshyn, MS, S-LP (C), CCC-SLP
Speech Language Pathologist

Joan N. Kaderavek, from The University of Toledo, Toledo, Ohio, proposed that young children with special needs have positive academic outcomes

when a combination of professionals including speech-language pathologists (SLPs), general education teachers, occupational and physical therapists, and early childhood special educators (ECSEs) work together. She concludes, through the forum encompassing views from several professions that "SLPs can work toward strategic alliances with ECSEs when they understand the field from the perspective of related professions in early childhood special education".

The field of early childhood special education has evolved over the past 25 years. Young children from birth to age 8 encounter challenges that "set them apart from typically developing young children as well as older individuals with disabilities".

The forum addresses critical factors which affect the degree to which inclusive early childhood education is successful for all children. These include quality of the early childhood setting, roles and responsibilities of personnel who work with young children with disabilities, efficacy of child-focused interventions,

and approaches to assess children and monitor progress.

Federal law in the United States mandates that young children with disabilities are required to have access to the general education curriculum. In community-based settings, approximately one third of all young children with individualized education program (IEP) goals receive specialized instruction in general early childhood programs (Individual With Disabilities Education Act Data, 2007). Aspects of early childhood special education include the following: Universal design for learning and high-quality environments:

- Differentiated instruction;
- Embedded Learning Opportunities;
- Roles, Responsibilities, and Accountability of Personnel Supporting Inclusion;
- Together the articles allow SLPs to consider a more encompassing perspective: "SLPs are more likely to build strategic alliances that facilitate the adoption of early childhood evidence-based practices when we understand the field from the perspective of teachers and ECSEs".

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Children With Cochlear Implants and Complex Needs



Ghada Ahmad, MA, Speech-language Therapist

In recent years, the number of children receiving cochlear implants who have significant disabilities in addition to their deafness has increased substantially. This article focuses on a special group of children with cochlear implants: those who have difficulties or disabilities in addition to

their deafness which make the task of meeting their complex needs particularly challenging. A figure of 30%–40% of deaf children is consistently quoted as being the proportion with additional disabilities, although definitions of what constitutes an additional disability do vary. They define them as “any physical, mental, emotional or behavioral disorder that significantly adds to the complexity of educating a hearing-impaired child”. “Cognitive/Learning Disabilities” - They found that the former group was significantly slower in developing speech perception skills following implantation. Interestingly, there was no significant association between outcome and the etiology of deafness per se. Thus, they speculate that the wide variety of outcomes among children with the same syndrome or etiology of deafness are individual variations in the functioning of higher centers of the brain. Autistic Spectrum Disorders (ASDs) remain one category of additional difficulties where great caution is exercised by cochlear implant

teams when considering a child for an implant. Hearing children with autism generally have major difficulties communicating effectively and in developing spoken language skills and may have sensory integration difficulties. It is therefore not surprising that deaf autistic children are not usually considered good candidates for cochlear implantation. Thus, in the past, a confirmed diagnosis of autistic spectrum disorder has typically been given as a contraindication to implantation. However, with the steady decrease in age at which children are being assessed and implanted, the number of children in whom autism is only diagnosed after they have received an implant is slowly increasing. The authors stress the importance of counseling parents about the potential benefits of an implant, emphasizing that the implant is likely to have little or no impact on the diagnosis of ASD and that oral communication is unlikely to be a realistic goal. In conclusion the available published literature on outcomes following cochlear implantation in children with additional disabilities suggests that cognitive functioning is one of the strongest predictors of progress in developing speech perception and speech production skills. Unfortunately, this is also one of the most difficult areas to accurately assess before implant in those children with the most complex needs.

Lindsey C. Edwards

<http://jdsde.oxfordjournals.org/cgi/content/full/12/3/258>

Overview Of Oropharyngeal Dysphagia



Lee Bolton, BA MMedSci MRCSLT, Specialist Speech Therapist Speech, Voice and Swallowing Disorders

Oropharyngeal dysphagia refers to any impairment in moving food, liquids or saliva safely and efficiently through the mouth and pharynx into the esophagus. It is a common and disabling disorder caused by a number of neuromuscular disorders or structural diseases of the oral

cavity, pharynx, larynx or cervical esophagus.

Causes of oropharyngeal dysphagia cover a wide variety of acute and chronic conditions, including brain damage resulting from trauma, infection, tumour, cerebrovascular disorders, congenital abnormality or neurosurgery, degenerative neurological and muscular diseases, psychiatric disorders, and trauma, disease or surgery to the aerodigestive tract. Common ENT presentations arise from head and neck malignancies, Zenker's diverticulum and GERD. With increasing numbers of people living longer, this results in a rapidly growing ageing population who have an increased risk of age-related illnesses associated with dysphagia.

Dysphagia has significant implications in terms of management of patients, health outcomes and healthcare costs. The main health complications associated with dysphagia are dehydration, malnutrition, and aspiration leading to choking and respiratory infections (aspiration pneumonia). Large-volume and frequent pulmonary aspiration of food, fluids and pathogenic oropharyngeal secretions is a serious event with associated risks of pneumonia and death. Early and accurate identification and evaluation of oropharyngeal dysphagia symptoms, as well as implementation of therapeutic interventions, are therefore essential to prevent or reduce the complications of dysphagia and

improve patient health and well-being.

It is important to remember that dysphagia is not a medical diagnosis per se, but rather a symptom of an underlying disease or disorder. Once identified, the precise nature and characteristics of the dysphagia must be comprehensively evaluated. It is not sufficient to categorise patients as dysphagic or non-dysphagic based on the presence or absence of dysphagia symptoms during a screening procedure (such as a barium swallow or flexible endoscopy), as this does not give the level of detailed information required about swallowing pathophysiology to initiate an appropriate treatment or management plan. Where the medical condition responsible for the dysphagia is not yet known, further investigations are required to establish the etiology before treatment can proceed.

Swallowing is a rapid and complex behaviour that requires a dynamic assessment approach as the patient swallows different food and fluid boluses. The most widely available and commonly used investigations for oropharyngeal dysphagia are the clinical/bedside assessment, videofluoroscopic and flexible endoscopic techniques. Standardised protocols have been developed for each that can provide a comprehensive evaluation of swallowing function. Each technique has its own strengths and weaknesses, therefore it is important to select the most appropriate tool based on the patient's symptoms and needs. The ideal situation for the dysphagia clinician and patient is access to clinical, videofluoroscopic and flexible endoscopic procedures.

In the next edition, I will provide an update on the evaluation techniques for oropharyngeal dysphagia, emphasising the importance of adopting a multidisciplinary team approach.