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Mechanisms of Auditory Neuropathy

By Dennis Colucci, AuD, MA

The annual Arnold Starr Lecture and Exhibition, now in its ninth year, was established to honor Arnold Starr, MD, for his national and international contributions as the founding chair and architect of the department of neurology at the University of California, Irvine (UCI). He is best known for his pioneering work in auditory electrophysiology used to identify lesions in the auditory nervous system, which subsequently resulted in his discovery of auditory neuropathy. He uncovered this rare disorder while investigating hearing loss in patients with motor and sensory neuropathies. Using objective test methods, Starr was able to identify the connection of the auditory nerve to cochlear hair cells or the nerve itself as the site of lesion and consequently, the term auditory neuropathy was coined. This discovery was important because auditory neuropathy involves not only hearing loss but also a loss of discrimination ability, especially in settings with background noise, such as when patients complain, "I can hear but can't understand you." Known as the Father of Auditory Neuropathy, Starr has provided numerous research articles in collaboration with investigators from around the world, including Fan-Gang Zeng, PhD, at UCI's Hearing and Speech laboratory. Over the past 40 years, Starr has developed breakthrough clinical protocols to streamline the diagnosis of auditory neuropathy and establish treatments for this previously illusive and life-altering condition.

BLENDING NEUROPATHY & ART

The reception and art exhibition featured an extraordinary collection of watercolors by Starr, including his *Neuralscapes*. These art forms go back to the 3rd century BC, chronicling man's discoveries and knowledge about the workings of the central nervous system and the human body. Starr studied brain drawings by famous neurologists such as Ramon, Cajal, and Golgi, who all contributed to his understanding of the brain and his interest in medical art. Even as a child, Starr always had a paper and a pencil or crayon with him, and drew pictures of "hands and feet and pianos and umbrellas." This eventually included an awareness of colors by watching his mother make spring hats using artificial flowers, thereby creating a colorful object in a few minutes. This passion turned into sketching muscles and nerves when he was in medical school. Starr has honored this tradition with a series of paintings depicting the auditory nervous system, inner ear, hair cells, spiral ganglion, and the auditory nerve and its structures—surely to be preserved as part of the history of auditory



Figure 1. The author and Arnold Starr, MD, at the 9th Annual Arnold Starr Lecture and Exhibition held in UC Irvine.

neuropathy. He recounted, "I always carry my watercolors and paper when I travel to capture both the landscapes and the neuralscapes." One of his most famous watercolors depicting the inner hair cell, synaptic connections, spiral ganglion, and the acoustic nerve is seen in his 2006 artwork titled, "Auditory Nerve with Inner Hair Cells" (Fig.2). His full collection can be viewed online (<https://www.arnoldstarrart.com>).

Following the reception, Fan-Gang Zeng, PhD, introduced Maoli Duan, MD, PhD, a global researcher from the Karolinska Institute of Sweden, who gave a lecture on the mechanisms of auditory neuropathy. His research is focused on stem cell and gene therapy, drug- and noise-induced hearing loss, and pro-



Figure 2. "Auditory Nerve with Inner Hair Cells" in watercolor is part of the *Neuralscapes* collection by Arnold Starr, MD, recently exhibited at UC Irvine.



Dr. Colucci is a clinical and forensic audiologist in private practice in Laguna Hills, CA.



Figure 3. From left: Fan-Gang Zeng, PhD, Arnold Starr, MD, and guest speaker Maoli Duan, MD, PhD, at the 9th Annual Arnold Starr Lecture and Exhibition.

tective mechanisms and treatments. Duan has published over 70 journal articles on a variety of topics in neurology and otolaryngology, and is on the editorial board of *Scientific Report* and a reviewer for more than 20 international journals. He presented a historical review of the auditory neuropathy literature, including the development of technology and the research leading up to its discovery, and highlighted Starr's remarkable research and how it changed our understanding of auditory nerve disorders, hearing loss, and communication disorders.

IMPACT ON HEARING HEALTH

Starr's discovery of eighth cranial nerve neuropathy has broadened our understanding of the auditory nerve's synaptic and neural mechanisms and how desynchrony relates to temporal processing dysfunction and perceptual deficits. Because of these impairments, patients primarily complain of



Figure 4. "Immature Cochlea" in watercolor and ink by Arnold Starr, MD.

discrimination problems, which are inconsistent with their degree of hearing. The diagnostic gold standards include otoacoustic emissions, auditory brainstem response (ABR), imaging of the brainstem, and auditory nerve and behavioral testing. Although, several other tests should be used to further delineate the synaptic and neural sites of a lesion when considering treatment options such as hearing aids, cochlear or brainstem implants, or when prostheses are contraindicated. Most remarkably, auditory neuropathy is a common cause of hearing disorders, especially among certain higher-risk groups with other neuropathies. However, research has shown that the causes are not limited to genetic phenotypes but also include a diverse group of etiologies, including noise trauma, eighth cranial nerve damage, medication, low birth weight, malnutrition, aging, and more (*Brain*. 2015 Nov;138[Pt 11]:3141-58).

The 9th Annual Arnold Starr Lecture and Exhibition was an opportunity to honor and learn more about Starr, and get an update on auditory neuropathy and in a broader sense, auditory spectrum disorders. It was also a reminder of the importance of medical art and how this has enhanced our understanding of the auditory system. Stay tuned for my next article on interesting pearls of wisdom on auditory neuropathy. [📖](#)

Q&A WITH ARNOLD STARR, MD

I had an opportunity to interview Arnold Starr, MD, and get his insight into two important questions about the significance and future of auditory nerve disorders.

What is the importance of auditory neuropathy to hearing loss patients?

Starr: Hearing loss is a general category. Using objective measures, we can define the neural and synaptic mechanisms independently. This allows us to localize the involved mechanism for a concise diagnosis, understand the patient's condition and provide better treatment.

How do you see future research and treatments for auditory neuropathy and synaptopathy?

Starr: Research is going to refine our understanding of the various mechanisms responsible for hair cell and neural damage and move toward genetic and stem cell solutions. This will change how we see treatment, not only in cases of auditory neuropathy but in all of neurology. The objective measures used in the clinic require well-trained clinicians to perform the tests and because of this, patients with auditory neuropathy go undetected, incorrectly diagnosed and treated. The future of objective testing needs to be more automated, easily used remotely and with just a computer or cell phone. Patients should be able to test themselves with objective measures using handheld devices that can transmit the recordings for analysis and diagnosis. This will allow specialists to determine the best medical interventions and rehabilitation.