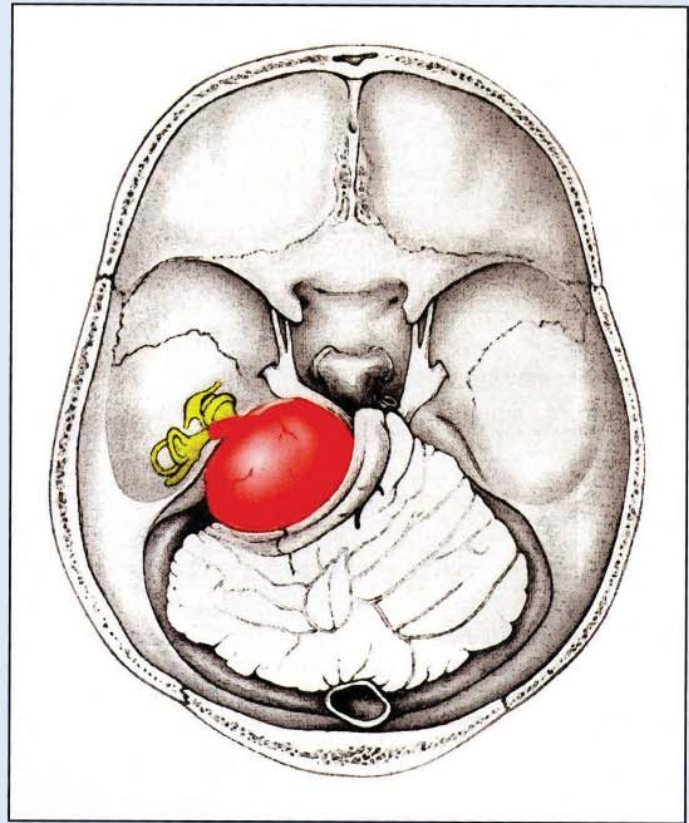


What is **NF2?**



Neurofibromatosis Type II (NF2) is a genetic disease of the central nervous system characterized by non-cancerous

fibrous tumors (also referred to as vestibular schwannomas or acoustic neuromas) that grow on both left and right vestibular nerves. Growth of these tumors results in pressure on hearing and facial nerves that causes hearing loss and facial paralysis.

Untreated, these tumors can result in loss of hearing, balance and ultimately death. Because of their location, removal of these tumors typically necessitates severing the auditory nerve, resulting in a complete loss of any remaining hearing on the side affected.

When NF2 is hereditary it results from a genetic mutation, occurring in approximately one in 25,000 live births in the U.S. Those who are born with this condition have one mutated copy of the NF2 gene in each cell. In about half of cases, the altered gene is inherited from an affected parent, who has a 50/50 possibility of passing the disease to their children. The remaining cases result from new mutations in the NF2 gene and occur in people with no history of the disorder in their family.

If the auditory nerve has been severed, a cochlear implant cannot be used to treat an individual with NF2. Instead, an NF2 patient may benefit from an auditory brainstem implant (ABI), which is designed to stimulate the auditory

portion of the cochlear nucleus in the brainstem and send sound signals directly to the brain. In the past decade, advances have been made in radiographic diagnosis, surgical approaches to acoustic neuromas and the understanding of the molecular biology of NF2.

At House Ear Institute's Center for Neural Tumor Research, researchers are seeking new drug-based therapies to combat NF2 that will lead to future human clinical trials. Utilizing a tumor cell line developed at HEI and unique animal models, Dr. Marco Giovannini and his co-workers are screening selected agents for those with the greatest potential for limiting growth or entirely eliminating the tumor cells resulting from NF2.

House Ear Institute is one of the few centers in the world with extensive experience both managing NF2 in presently affected patients as well as researching new, preventive treatments. ♦