The term neurofibromatosis *(NF)* is used for a group of genetic disorders that affect the cell growth of neural tissues. These hypertrophied nerve trunks are tortous, form a plexus and often are embedded in a fibrous matrix. There may be an associated over growth of the surrounding soft tissue.

Type I neurofibromatosis is a genetic, multisystemic disorder that has major cutaneous manifestation, such as cafe´-au-lait spots and neurofibroma. Its hallmark symptom is multiple benign dermal neurofibromas.

Neurofibromatosis type 2 is much less frequent than neurofibromatosis type 1, Intracranial schwannomas of the VIII cranial nerve complex are the hallmark of *NF2*.

The most frequent tumors of the cranial nerves are referred to as schwannomas They may develop in most cranial nerves, except I and II, which do not have Schwann cells, except for very rare cases of ectopic pediatric olfactory schwannomas.

Schwannomas are benign tumours of the nerve sheath that arise on cranial and spinal nerve roots as well as along the course of peripheral nerves. First recognized as a separate entity by ***Verocay*** in 1910, they have been described in almost every part of the body. Intracranial schwannomas are relatively common.

The most frequently affected are vestibular, trigeminal, and facial nerves, followed by the caudal cranial nerve group.Schwannomas originating from the oculomotor, trochlear, and abducent nerves in the absence of neurofibromatosis are extremely rare.

The trigeminal nerve is next most frequently affected cranial nerve. Presence of occulomotor, trochlear or abducent nerve tumors should raise the possibility of *NF2.* Similarly, involvement of more than one cranial nerve should warrant a work up for *NF2.*

The pattern of presentation in children is variable, and quite different from adult onset type. While hearing loss or tinnitus is the most common presenting symptom in adult patients.

Neurologic complications include tumors of the cranian nerves, peripheral nerves, nerve roots, and plexi; spinal cord compression;

Diagnosis of neurofibromatosis is baised mainly on clinical, radiological diangosis. Yet genetical diagnosis may plays and important role in diangosis of some cases for example prental diagnosis.

Care of persons with *NF1*, or those strongly suspected of being affected, should be provided by a clinician who is familiar with *NF1.* This entails reevaluation annually or more frequently if symptoms dictate. The mainstay of care is anticipatory guidance and surveillance for treatable complications.

The surgical treatment of individuals with *NF2* is complex and should probably be limited to specialty tertiary care centers with experienced otolaryngologists and neurosurgeons.

Stereotactic radiosurgery (*gamma knife radiosurgery*) has been offered as an alternative to surgery in selected patients with vestibular schwannoma. Radiation therapy of other *NF2*-associated tumours should be considered carefully, since radiation exposure may induce, accelerate, or transform tumours in a patient with an inactivated tumour suppressor gene.

Excellent results have been reported for the treatment of acoustic neuromas using modern microsurgical techniques. The treatment strategy for intracanalicular acoustic neuromas is controversial because of the natural course of the condition and associated treatment morbidity. Expert acoustic neuroma surgeons recommend early surgical resection to preserve hearing in patients with these neuromas.