Peripheral Nervous System

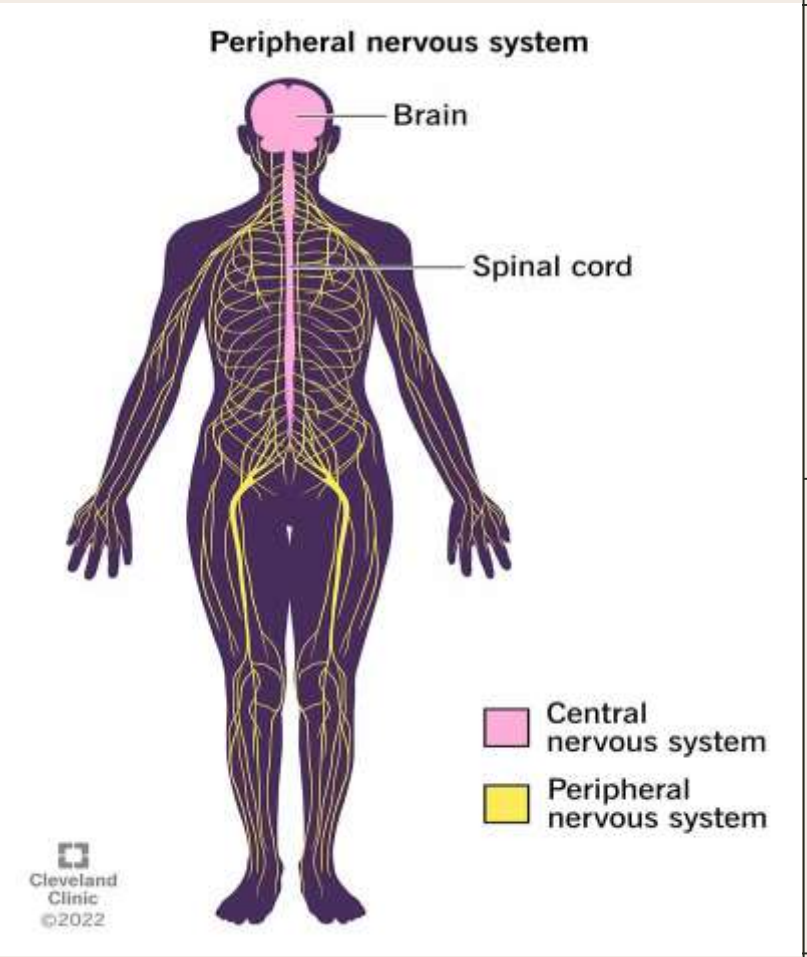
Understanding the PNS and its Importance

Definition

The Peripheral Nervous System (PNS) is the part of the nervous system outside the brain and spinal cord. It connects the central nervous system to limbs and organs, facilitating communication throughout the body.

Peripheral vs Central Nervous Systems

The Central Nervous System (CNS) consists of the brain and spinal cord, acting as the control center. In contrast, the Peripheral Nervous System (PNS) extends beyond the CNS, connecting it to the limbs and organs, facilitating communication between the brain and body. This distinction is crucial in understanding how bodily functions are regulated and coordinated.



Components

Cranial nerves: There are 12 pairs of nerves that connect directly to your brain, and 11 of them are part of your peripheral nervous system

Spinal nerves: These are 31 pairs of nerves that attach to your spine at about the same level as each segment bone (vertebra) in your spine.

Function

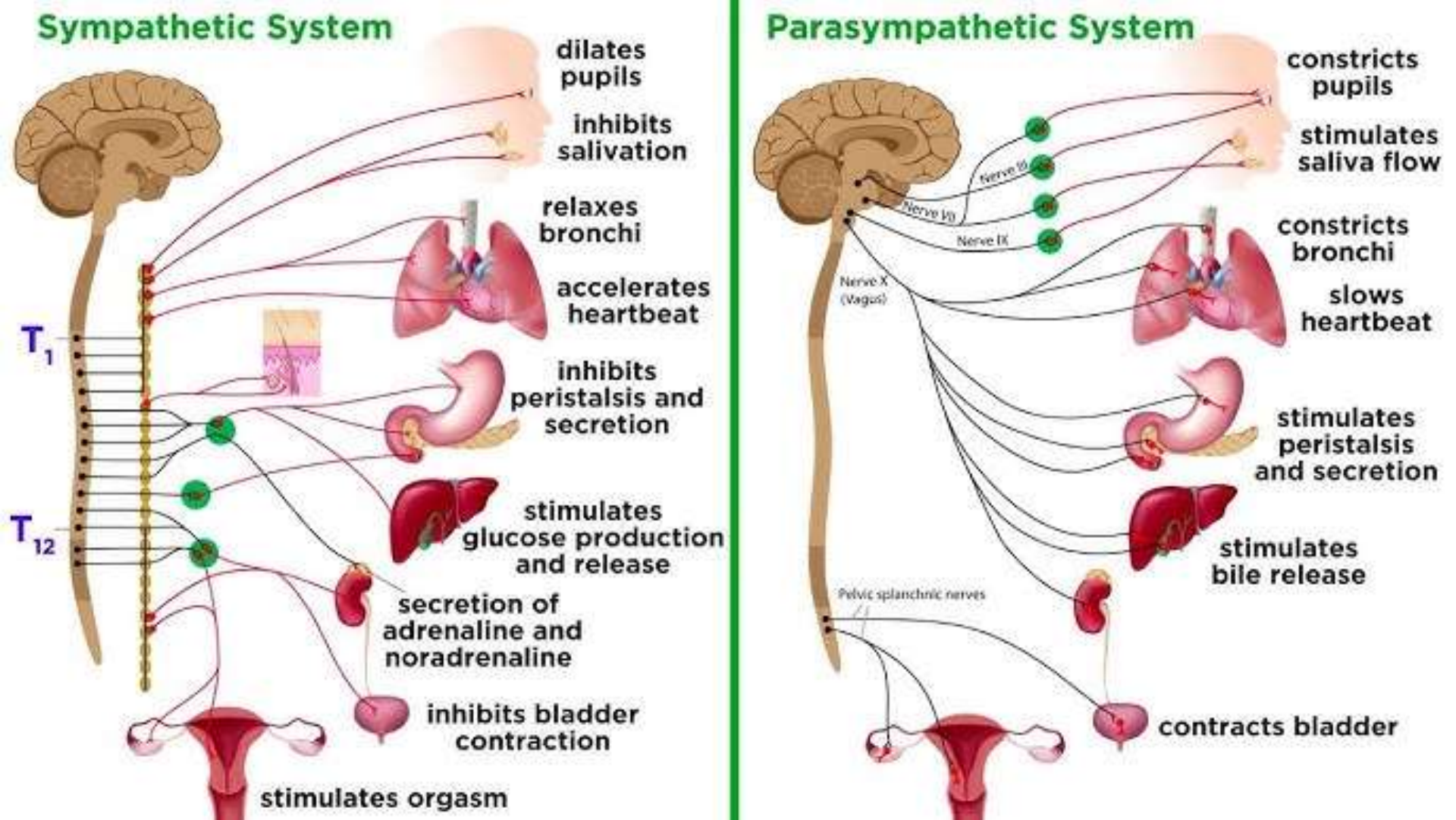
Subsystems: Autonomic and Somatic

The PNS has two main subsystems: the autonomic and somatic nervous systems. The autonomic nervous system regulates involuntary functions such as heartbeat and digestion. It is further divided into the sympathetic (fight or flight response) and parasympathetic (rest and digest) systems. The somatic nervous system, however, governs voluntary movements and transmits sensory information to the CNS.

Table 1. **Functions and dysfunctions of the cranial nerves**

Cranial nerve name (number)	Type	Function	Associated dysfunction(s)
Olfactory (I)	Sensory	Sense of smell	• Unilateral or bilateral loss of sense of smell • Loss of taste
Optic (II)	Sensory	Vision	• Loss of vision
Oculomotor (III)	Motor	Movement of the eyeball and upper eyelid	• Eye-movement problems
	Parasympathetic	Pupil constriction	
Trochlear (IV)	Motor	Movement of the eyeball	• Eye-movement problems
Trigeminal (V)	Sensory	General sensation in face, scalp, corneas, and nasal and oral cavities	• Loss of facial sensation
	Motor	Chewing	
Abducens (VI)	Motor	Movement of the eyeball	• Eye-movement problems
Facial (VII)	Sensory	Taste	• Loss of taste • Inability to close eye
	Motor	Facial expression	
	Parasympathetic	Secretion of tears and saliva	
Vestibulocochlear (VIII)	Sensory	Hearing and balance	• Loss of hearing and balance
Glossopharyngeal (IX)	Sensory	Taste and sensation from back of tongue	• Inability to swallow • Hoarse voice
	Motor	Swallowing and speech	
	Parasympathetic	Secretion of saliva	
Vagus (X)	Sensory	Taste and sensation from epiglottis and pharynx	• Inability to swallow • Hoarse voice • Delayed gastric emptying
	Motor	Swallowing and speech	
	Parasympathetic	Muscle contraction of thoracic and abdominal organs and secretion of digestive fluids	
Accessory (XI)	Motor	Head and shoulder movement	• Inability to move head and raise shoulders
Hypoglossal (XII)	Motor	Movement of the tongue muscles	• Inability to move tongue

Source: Bayram-Weston (2020)


Sympathetic System
dilates pupils
inhibits salivation
relaxes bronchi
accelerates heartbeat
T1
inhibits peristalsis and secretion
T12
stimulates glucose production and release
secretion of adrenaline and noradrenaline
inhibits bladder contraction
stimulates orgasm
Parasympathetic System
constricts pupils
stimulates saliva flow
Nerve VII
Nerve IX
Nerve X (Vagus)
constricts bronchi
slows heartbeat
stimulates peristalsis and secretion
stimulates bile release
Pelvic splanchnic nerves
contracts bladder

Conditions and Disorders

- **Type 2 diabetes**
- **Autoimmune and inflammatory conditions**.: lupus, Guillain-Barré syndrome, rheumatoid arthritis
- **genetic conditions:** NF1, NF2
- **Infections:** viruses such as HIV or bacteria such as *Borrelia burgdorferi*, which causes Lyme disease
- **Trauma**
- **Tumors**

Common signs or symptoms

motor nerves: Weakness.
Cramps, spasms, tremors or twitches.
Wasting (shrinking of muscles).
Loss of control.

Sensory nerves: Tingling or numbness (paresthesia)
Neuropathic pain
Loss of touch.

autonomic nerves: Circulatory system (BP control)
Digestive system
Skin and temperature control (hyperhidrosis / anhidrosis)

Common tests

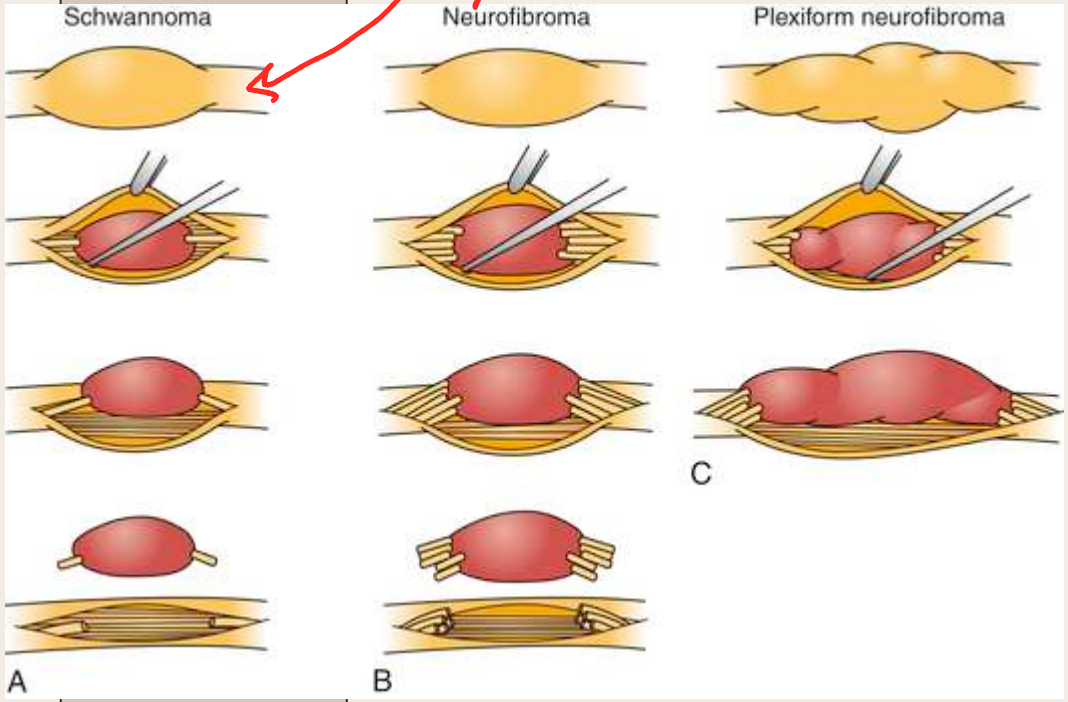
- Nerve conduction tests.
- Electromyogram
- Nerve ultrasound.
- Nerve biopsy.
- Genetic testing
- Magnetic resonance imaging (MRI)

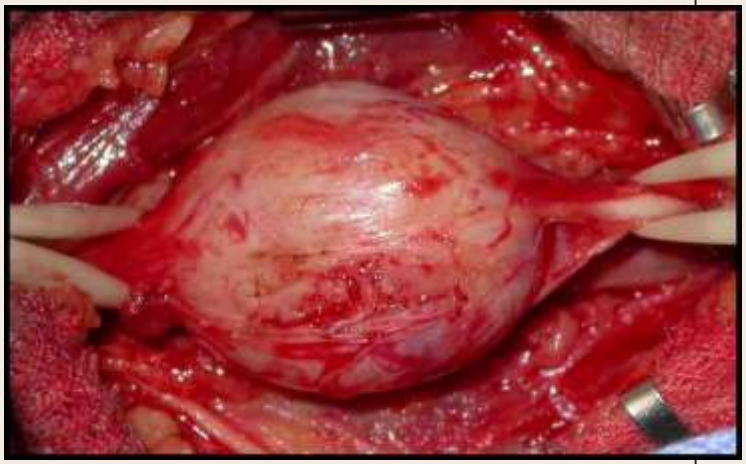
Table 1 - Diagnostic criteria for NF1 and NF2

Disorder	Criteria
NF1	1. 6 or more cafe´-au-lait spots measuring at least 5 mm before puberty or 15 mm after puberty 2. Axillary or inguinal freckling 3. 2 or more neurofibromas or 1 plexiform neurofibroma 4. 2 or more iris Lisch nodules 5. Optic pathway glioma 6. Characteristic skeletal dysplasia (long bone or sphenoid wing) 7. Affected first-degree relative *At least 2 must be present
NF2	Confirmed diagnosis: Bilateral vestibular schwannomas Probable NF2: Family history of NF2 and unilateral vestibular schwannoma, plus one of following: • Meningioma • Ependymoma • Glioma Shwanoma. • Posterior subcapsular cataract/cortical opacity

- well capsulated
- dorsal horn

diffused



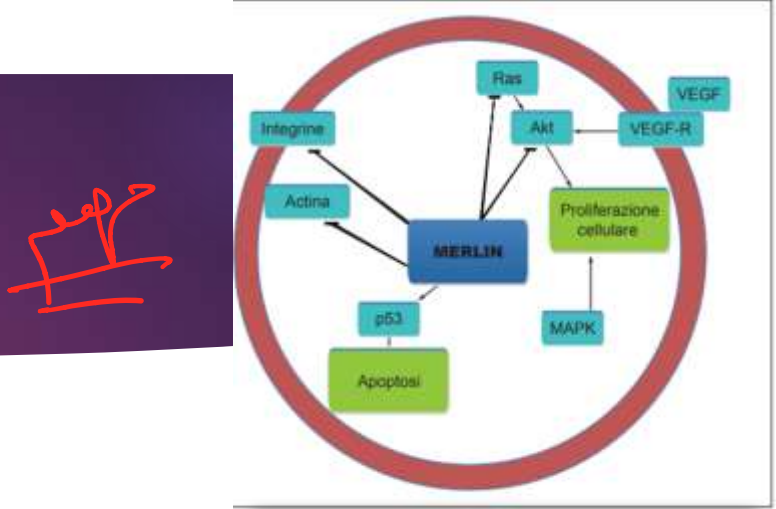


Vestibular Schwannoma

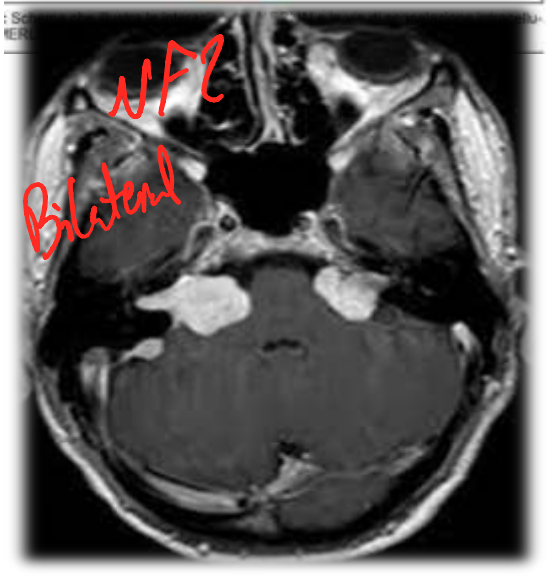
Most common site N VIII

- More improperly, "**acoustic neuroma**";
- Benign tumor originating from the Schwann cells of one of the two vestibular branches VIII n.c.;
- 70% originates from the inferior vestibular nerve in its intra-canal portion at the transition point between the central myelin and the peripheral myelin «Transition zone» with growth in the C.P.A. cistern. ;
- **Incidence 1 in 100,000 inhabitants/year;**
- 8% of adult intracranial tumors;
- 80-90% of all A.P.C. tumors;
- IV-VI decade (average age 50 years);
- **Unilateral in 95% of cases; bilateral in 5% (NF2)**

Vestibular Schwannoma



- Increased incidence in patients with Neurofibromatosis type 2 (high penetrance AD)
- NF-2: inactivating mutation in the tumor suppressor gene located on chromosome 22q12 which encodes the merlin or schwannomin protein [high RAS protein expression and cell proliferation]
- age <40 years - suspicion of NF
- 5% of patients diagnosed with schwannoma are affected by NF-2
- NF-2-associated and sporadic VS are cytologically identical



Vestibular Schwannoma

- Increased expression of some pro-inflammatory cytokines (TGF-β1, IL-1β and IL-6). The neoplastic cell produces pro-inflammatory cytokines that act in an autocrine manner by stimulating cell proliferation

- Increased expression of VEGF which induces neoplastic growth by promoting angiogenesis

Mol Med Rep. 2015 Mar 4. doi: 10.3892/mmr.2015.3415. [Epub ahead of print]

Immunohistochemical profile of cytokines and growth factors expressed in vestibular schwannoma and in normal vestibular nerve tissue.

Taurone S[1], Bianchi E[1], Attanasio G[1], Gioia CD[2], Ierinó R[2], Carubbi C[3], Galli D[3], Pastore FS[4], Giangaspero F[2], Filipo R[1], Zanza C[1], Artico M[1].

- Benign tumor
- Slow growth (1-3 mm/year)
- 2% histological aspects of malignancy: cellular atypia, high number of mitoses

MACRO

- Brown, round/oval, capsulated extra-axial mass;
- may have bright yellow areas and hemorrhages;
- 15-20% is associated with cystic degeneration (d.d.x.: arachnoid cyst)

MICRO

- Elongated neoplastic Schwann cells
- There are two morphological tissue patterns:
- Antoni A (scarce stroma, areas of high cellularity, elongated cells, palisa
- Antoni B (less cellular, loose connective tissue, lipid-rich stroma); often areas of cystic degeneration

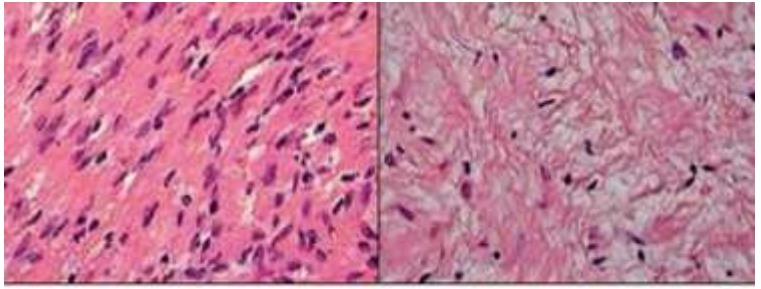


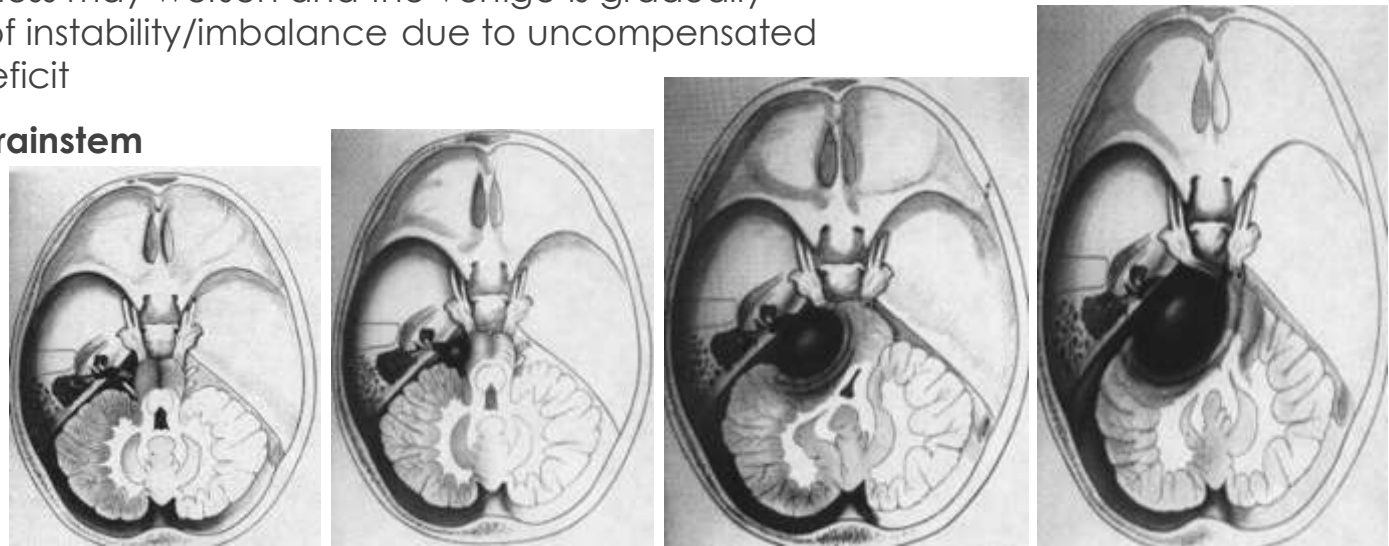
Fig. 11: Tipo A di Antoni.

Fig. 12: Tipo B di Antoni.

Growth pattern with clinical correlation

The growth of acoustic neuroma can be divided into four anatomical stages:

1. **Intracanalicular**: hearing loss, tinnitus, rare dizziness
2. **Cisternal**: the hearing loss may worsen and the vertigo is gradually replaced by a sense of instability/imbalance due to uncompensated unilateral vestibular deficit
3. **Compressive on the brainstem**
4. **Hydrocephalic stage**



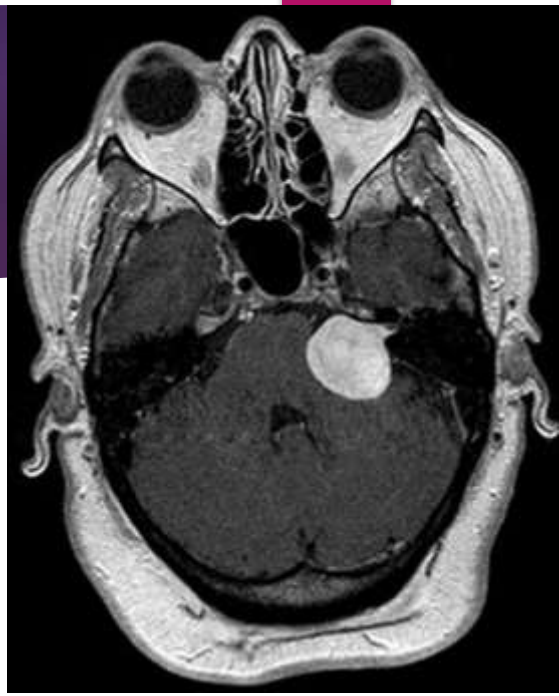
Hearing reduction/loss Difficulty understanding words	98%
Tinnitus	70%
Vertigo	67%
Headache	32%
Facial paralysis	10%
Diplopea	10%
Nausea & vomiting	9%
Otalgia	9%
Altered taste sensation	6%

Incidence of headache

* In tumors measuring 1 to 3 cm: 20%
* > 3 cm: over 40%

It is usually localized in the sub-occipital region or widespread

Radiological characteristics



- slow growing extra-axial mass
- **acute** connection angle with the adjacent bone
- Morphology: "**ice cream cone**"
- Endocranial opening CUI

CT:

non-calcified, iso-/modestly hyperdense mass

Widening of the internal acoustic meatus

remarkable, uniform contrast grip

MRI with Gd: ***gold standard***

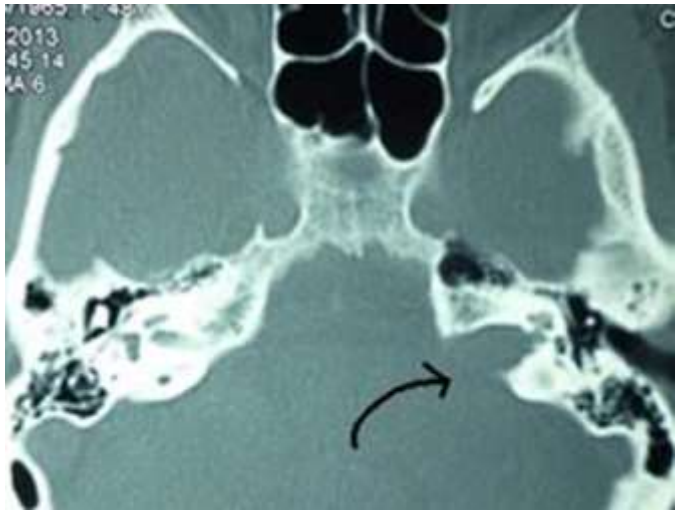
- iso-/hypointense on T1
- Hyperintense on T2

in 15% T2 hyperintense intratumoral cysts

- >95% notable contrast grip (2/3 solid; 1/3 ring-shaped or non-homogeneous)

DIMENSIONS:
Small < 1.5 cm
Moderate 1.5-3 cm
Large >3 cm

- Widening of the internal acoustic meatus "**Trumpet sign**"

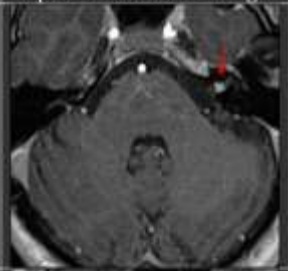
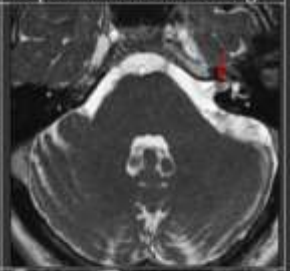
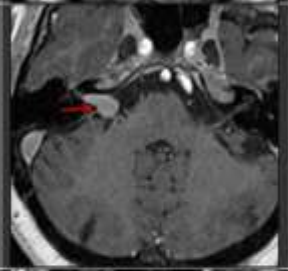
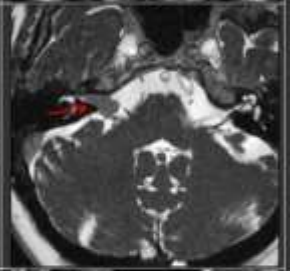
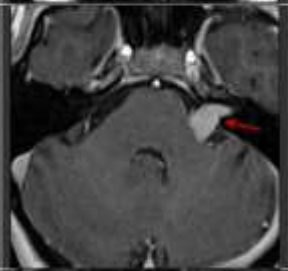
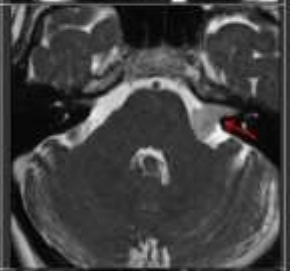


Koos Classification

- KOOS classification

- I: < 1cm
- II < 2 cm
- III < 3 cm
- IV > 3 cm

- Surgical risk assessment and best surgical strategy

Grade	Criteria	Representative ceT1 image	Representative hrT2 image
I	Tumours are completely confined to the internal auditory canal.		
II	Tumours have both intra- and extrameatal components, extending into the cerebellopontine angle (CPA) but do not contact the brainstem.		
III	Tumours contact the brainstem but do not compress it.		
IV	Tumours cause brainstem compression and/or displacement of adjacent cranial nerves.