

Grand Round

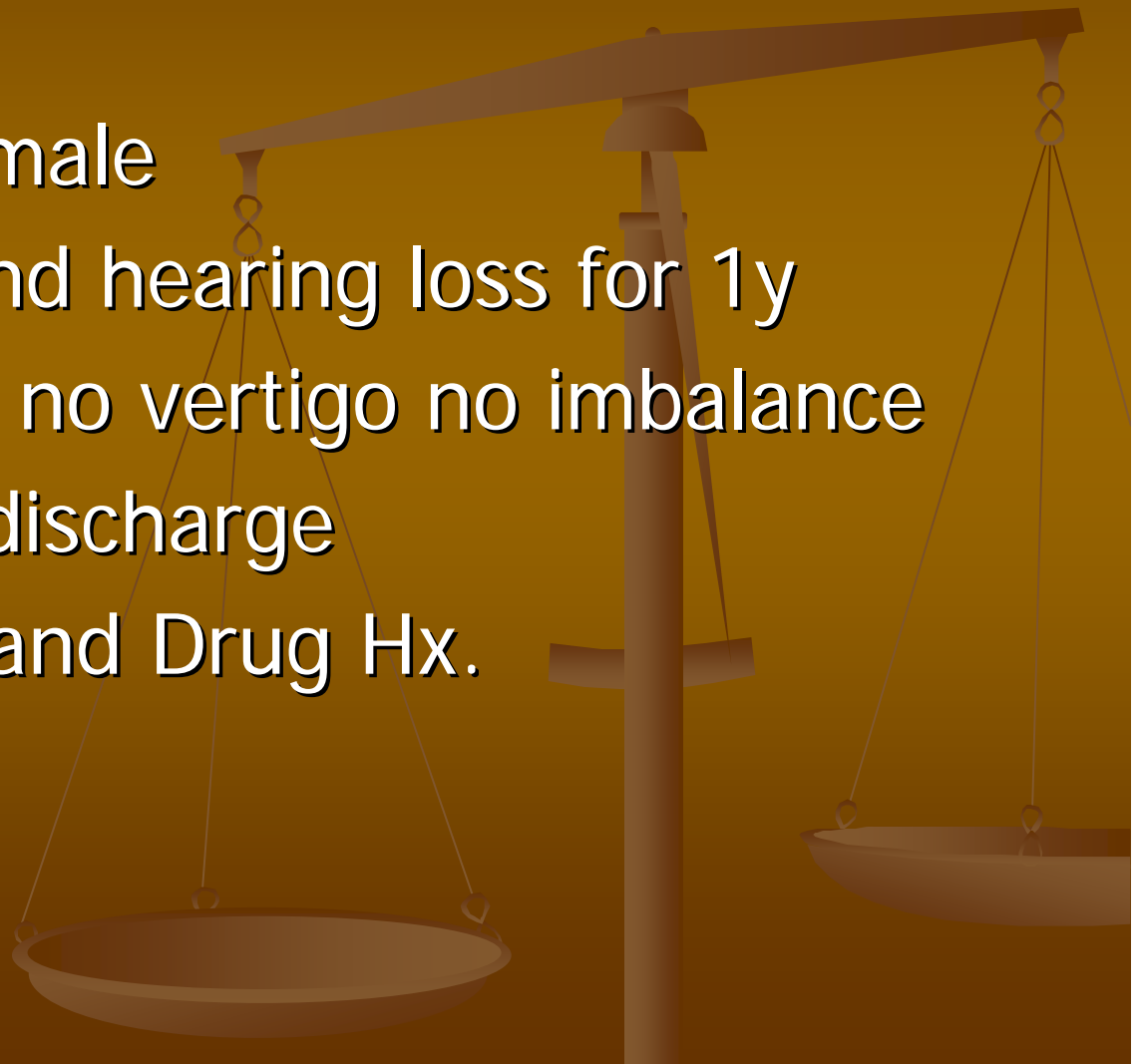
Abdulrahman Hagr
MBBS FRCS(c)



Case

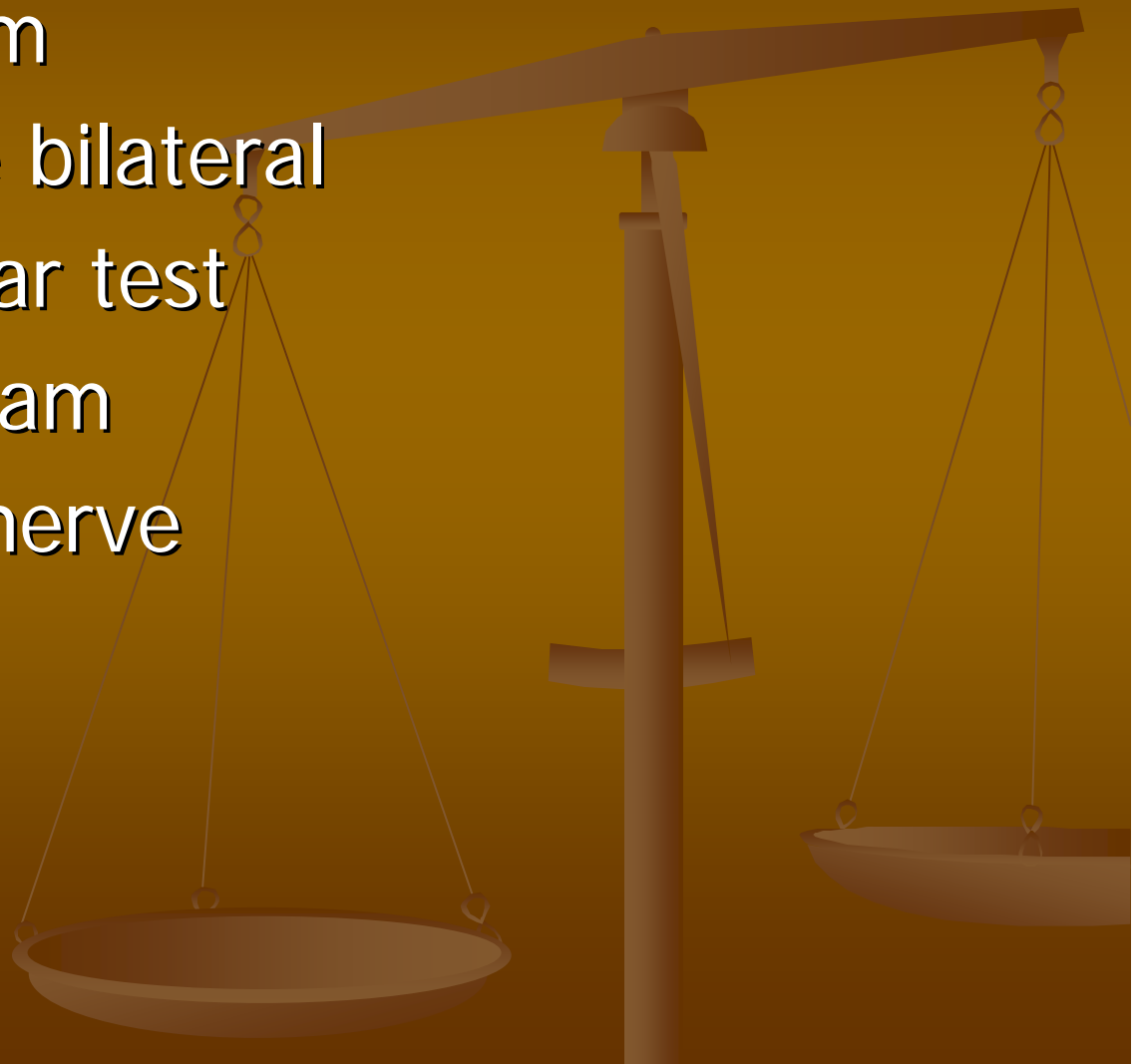
2001 Oct 22

- 59 y old single male
- Lt ear ringing and hearing loss for 1y
- No ear-fullness, no vertigo no imbalance
- No earache no discharge
- -ve FHx, PMHx and Drug Hx.



Examination

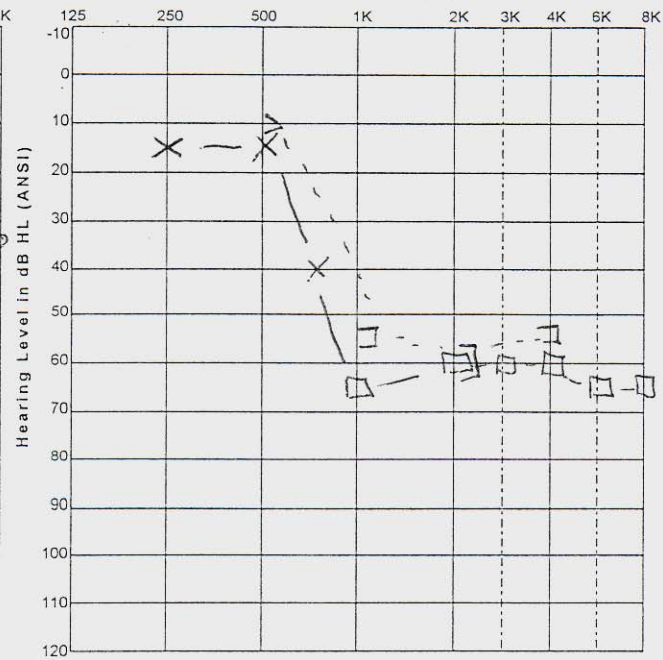
- Normal ear exam
w → Rt R +ve bilateral
- Normal vestibular test
- Normal Neck exam
- Normal cranial nerve



Next?



LEFT EAR - Freq. in Hz

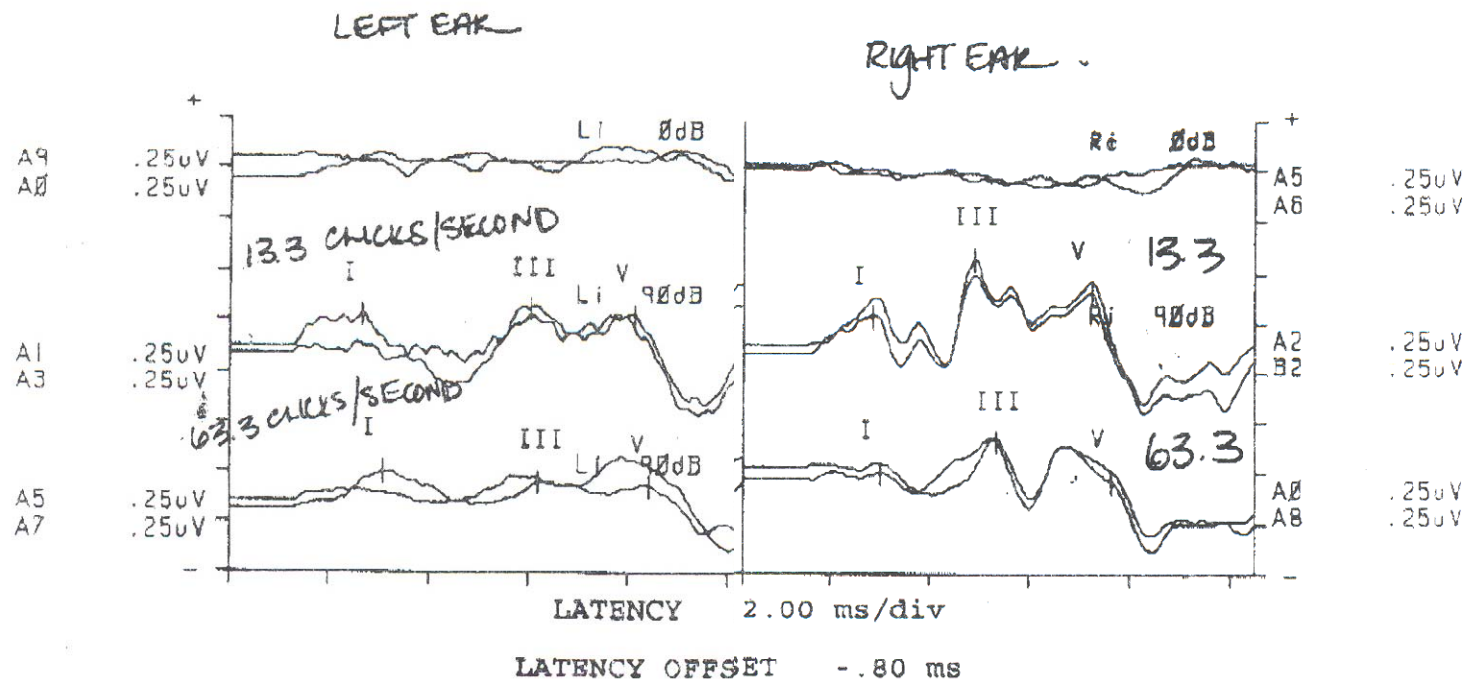


500	1K	2K	4K
NR	→		
	NR	→	

[illegible]

Next?





Left Ear

AL I= 2.08 III= 5.3 V= 7.4
 IPL 1-III=3.24 III-V=2.16 I-V=5.4

Right Ear

AL I= 1.84 III= 4.02 V= 6.36
 IPL 1-III=2.19 III-V=2.34 I-V=4.52

Left Ear

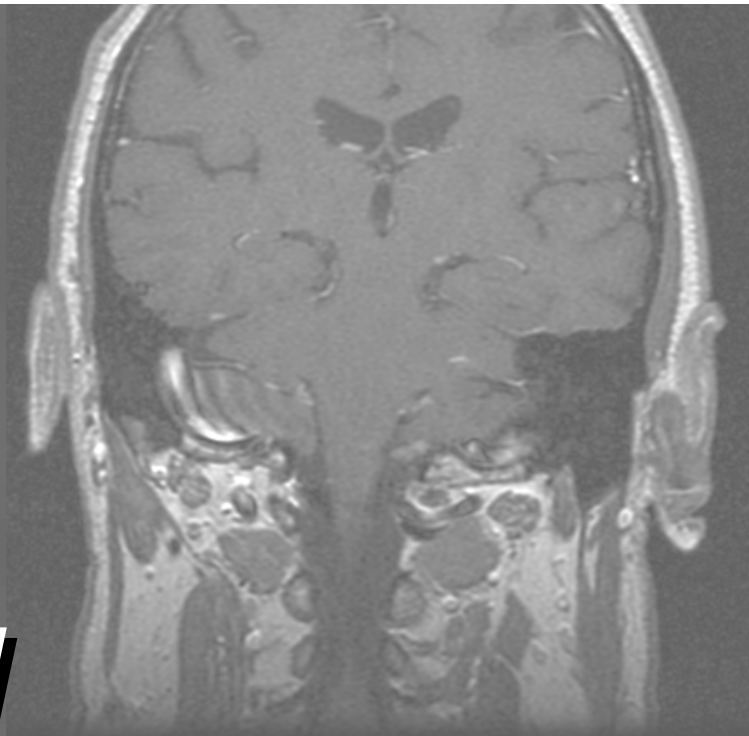
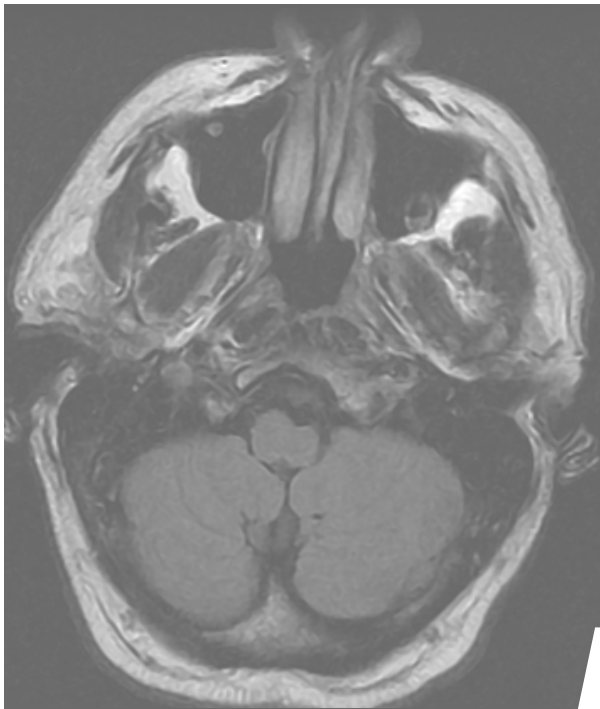
Extended I , III & V absolute latencies.
 Extended I-III, I-V inter-peak latencies

Right Ear

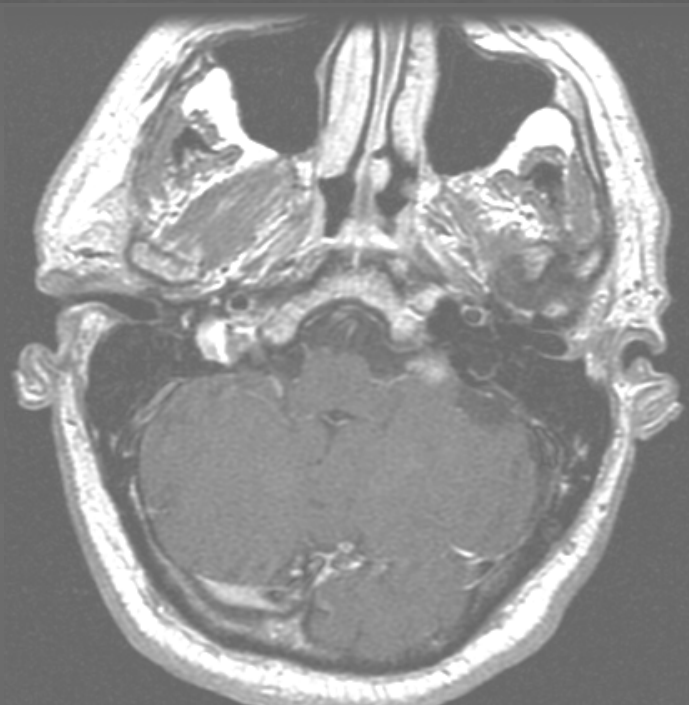
Extended I & V absolute latencies.
 Normal inter-peak latencies

Next?

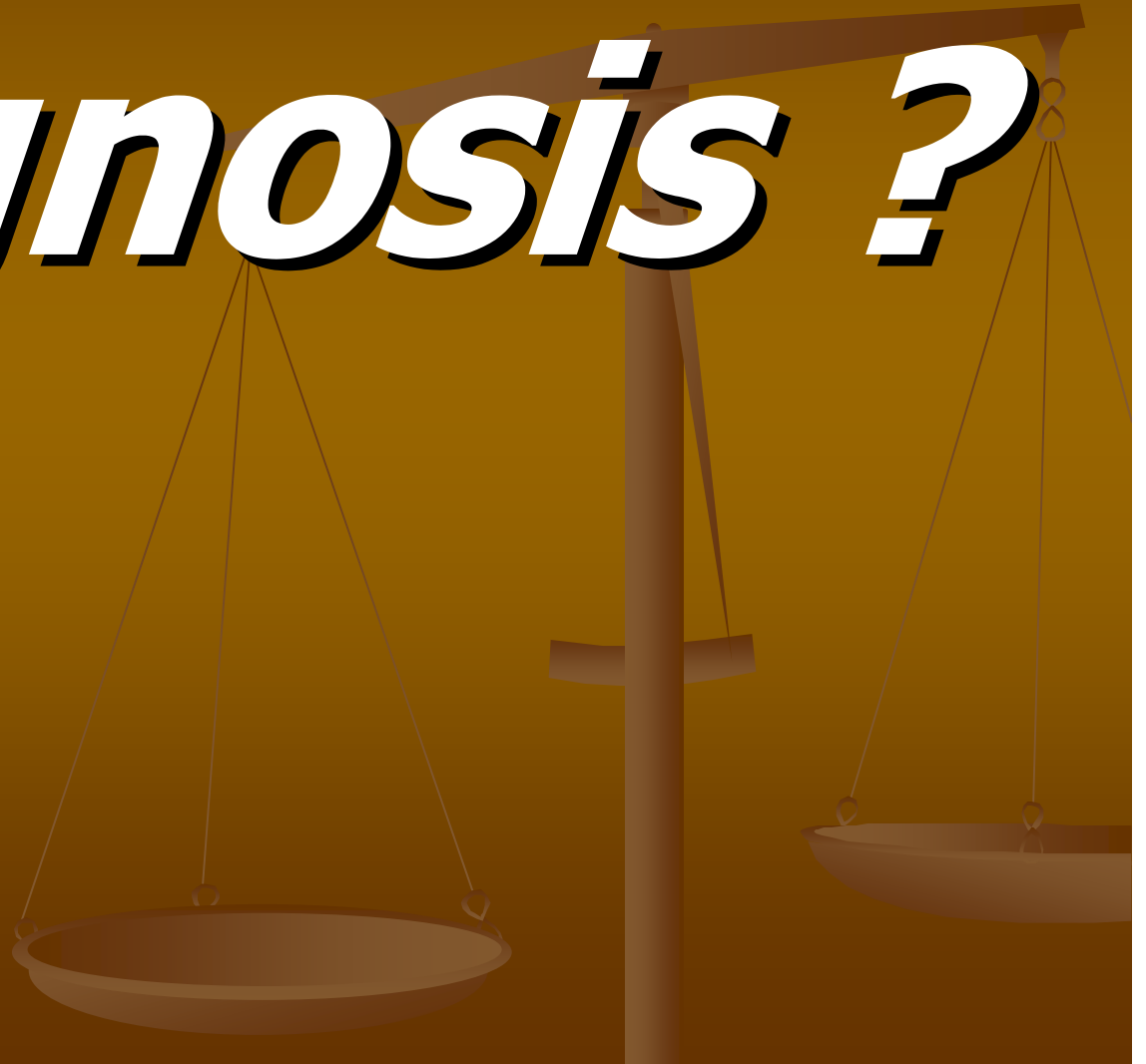




M.R.I



Diagnosis ?



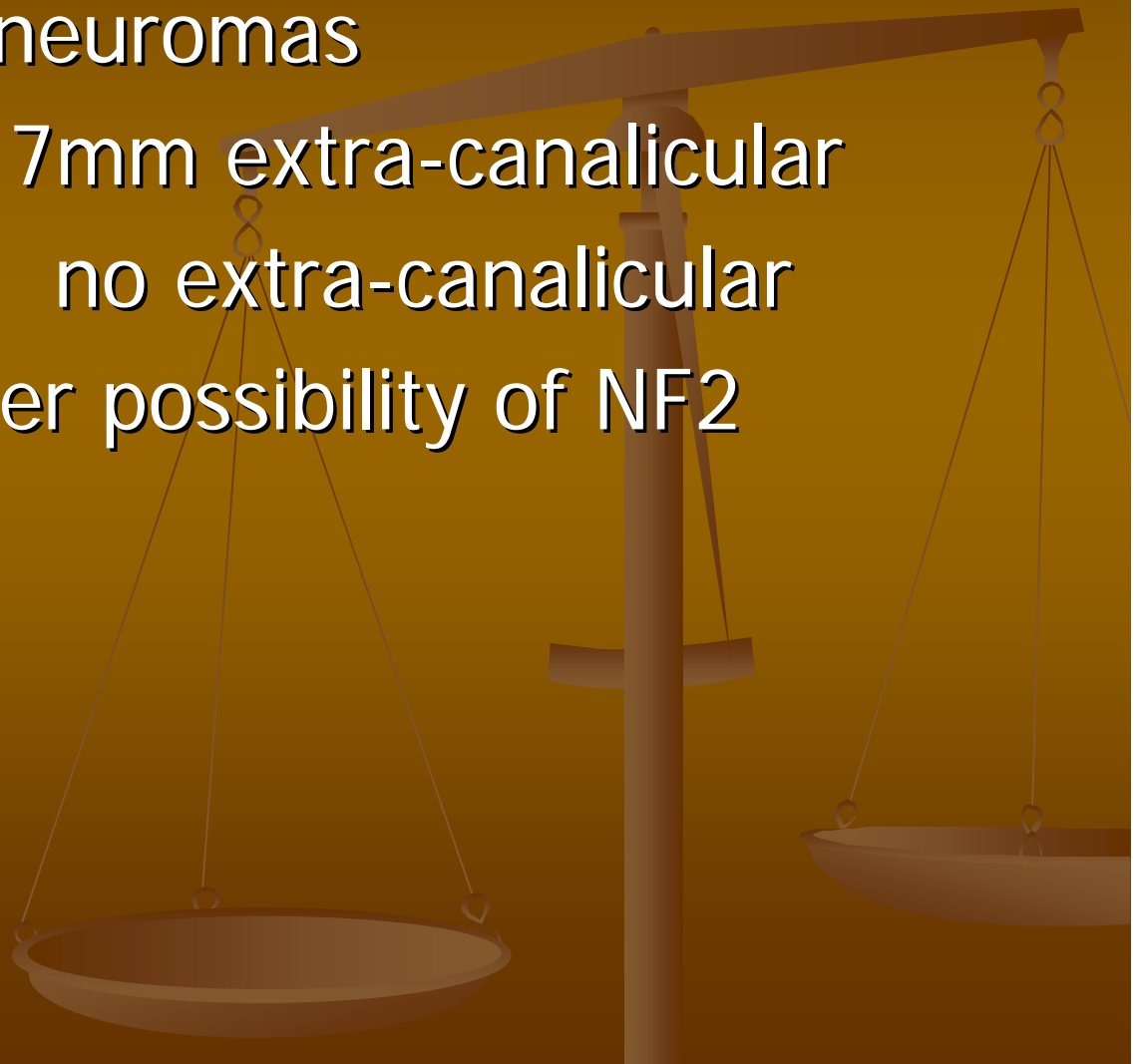
MRI Report

Bilateral acoustic neuromas

Left 12X8X9 mm 7mm extra-canalicular

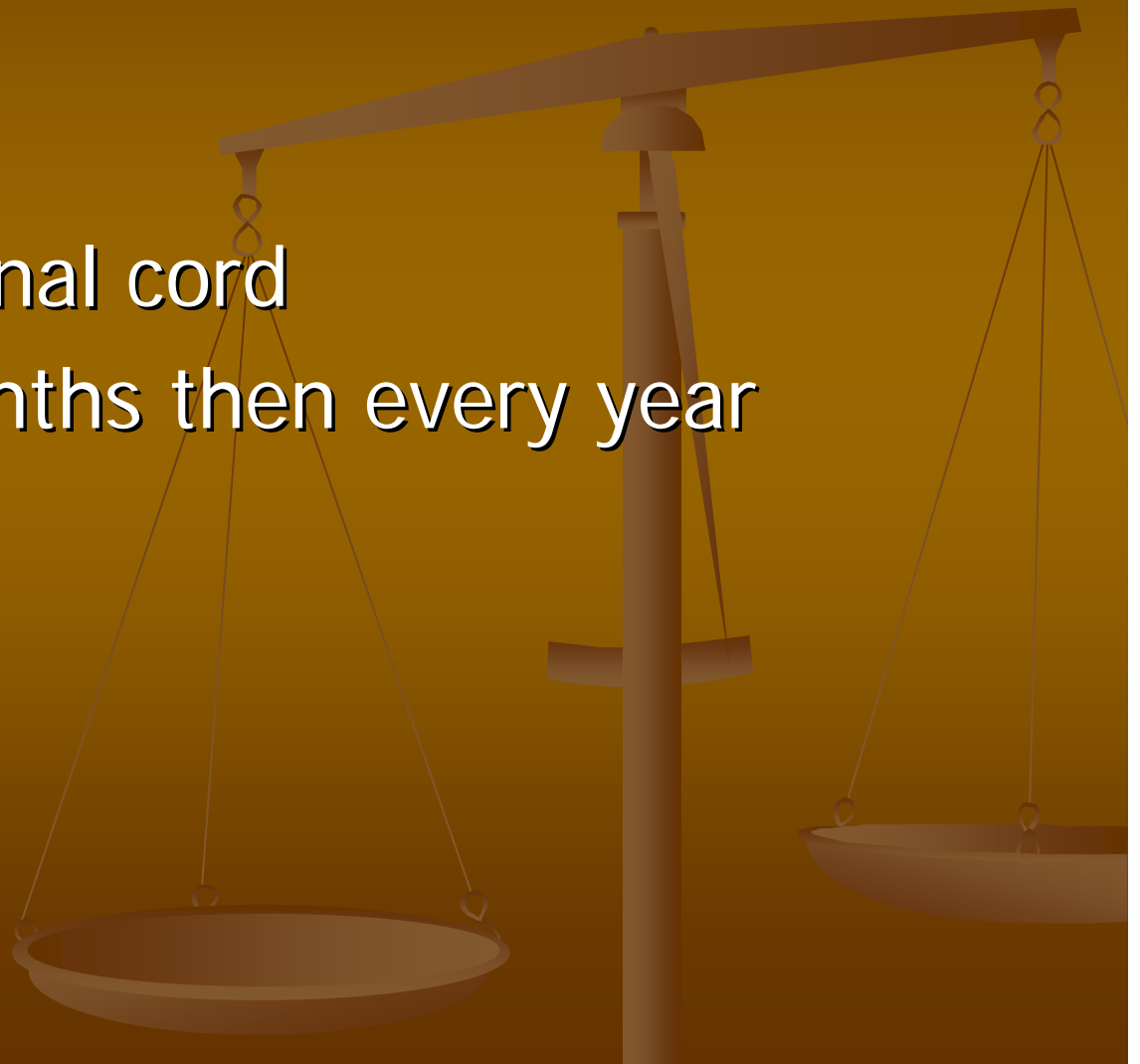
Right 7x6x6 mm no extra-canalicular

One would consider possibility of NF2



Follow up

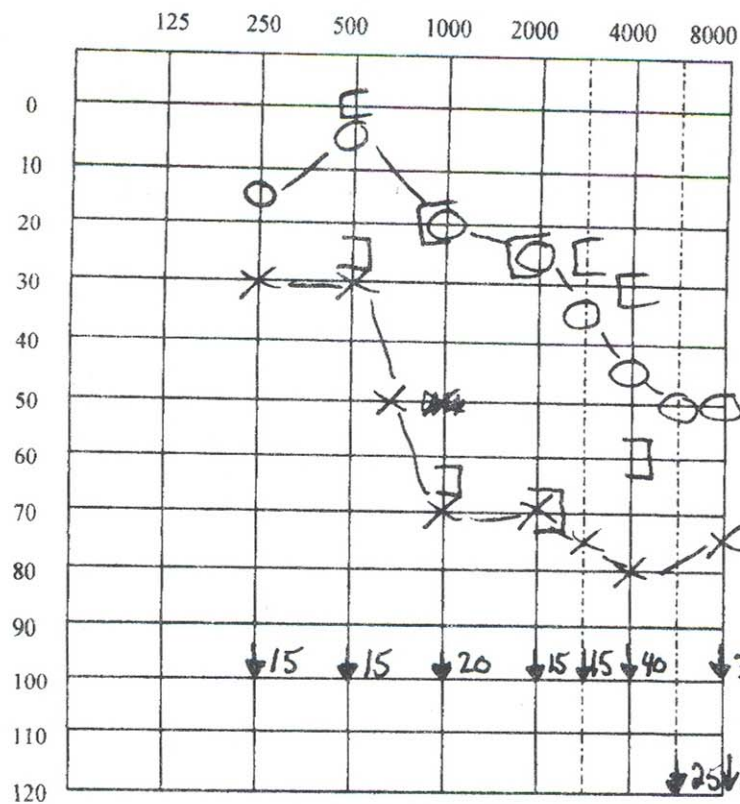
- Ophthalmology
- Genetic
- MRI Brain & spinal cord
- Follow up 6 months then every year
 - MRI IAC
 - Audiogram



Ophthalmology

- Early lenticular opacities bilaterally
- No other abnormality





AudioGram Key		
	RT	LT
AC Unmasked	O	X
AC Masked	△	▽
BC UnMasked	∠	∠
MC Masked	□	□
No Response AC	↘	↘
No Response BC	↘	↘
A - Soundfield Aided		
W - Soundfield Wabble		
B - Soundfield Unaided		
NBN - Narrow Band Noise S.F.		

SRT Speech Discrimination Tests UCL MCL

Right: 10 dBHL 76 % AT 50 dBHL _____ dBHL _____ dBHL

Left: 50 dBHL 44 % AT 90 dBHL _____ dBHL _____ dBHL

EarCanal Physical

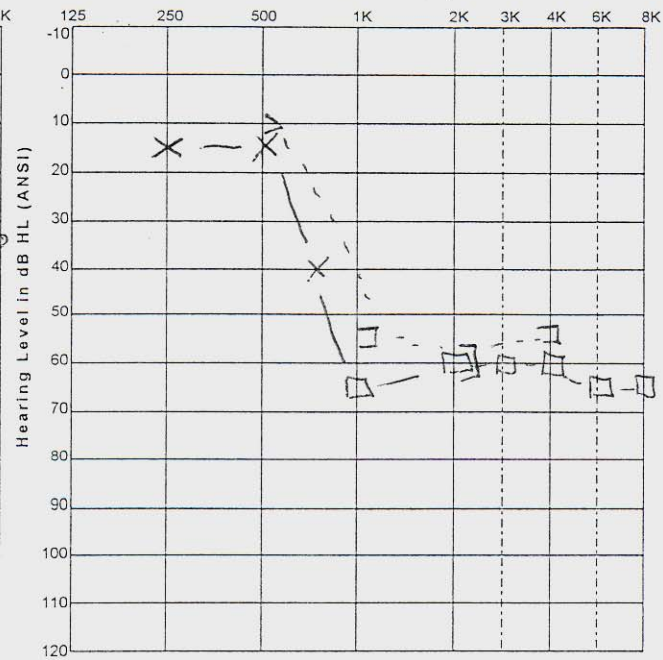
Volume Peak Press.

LT 1.0 ML 1.0 -20

RT 2.0 ML 0.9 -5

Y_{tm} = 226 HZ

LEFT EAR - Freq. in Hz



500	1K	2K	4K
NR	→		
	NR	→	

[illegible]

MRI IAC, Brain & Spine

■ **Spinal cord**

- small T12 hemangioma

■ **Brain**

- Enlarge Lt AN 12 mm extra-canalicular
- No displacement of brain stem.
- Rt side no change
- Normal Brain

? Next



Follow up 2002

- ? Prophylactic radiation
- SSNHL → Steroid



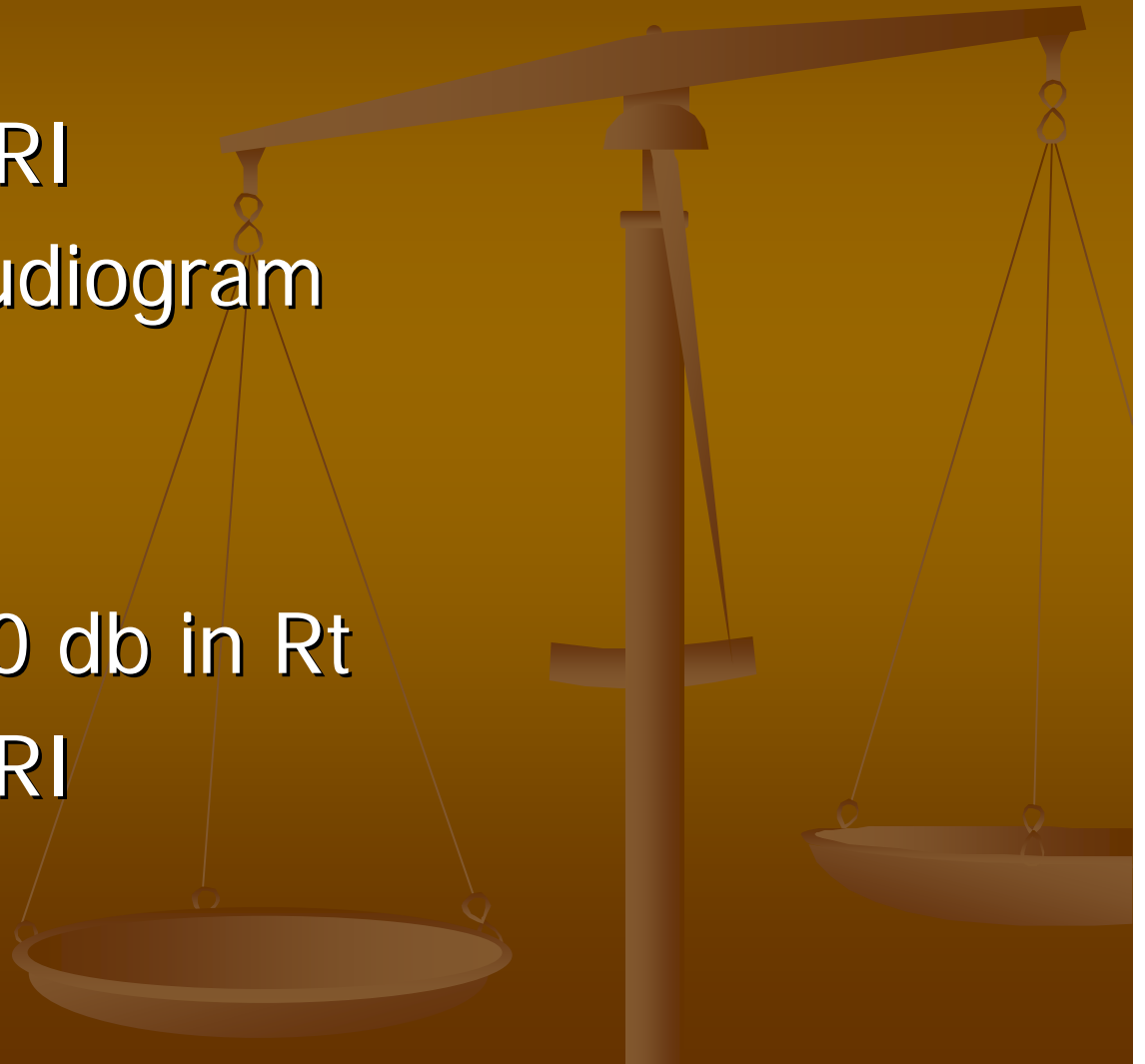
Follow up 2003 & 2004

2003

- No change in MRI
- No change in audiogram

2004

- Improvement 20 db in Rt
- No change in MRI



Neurofibromatosis Type 2

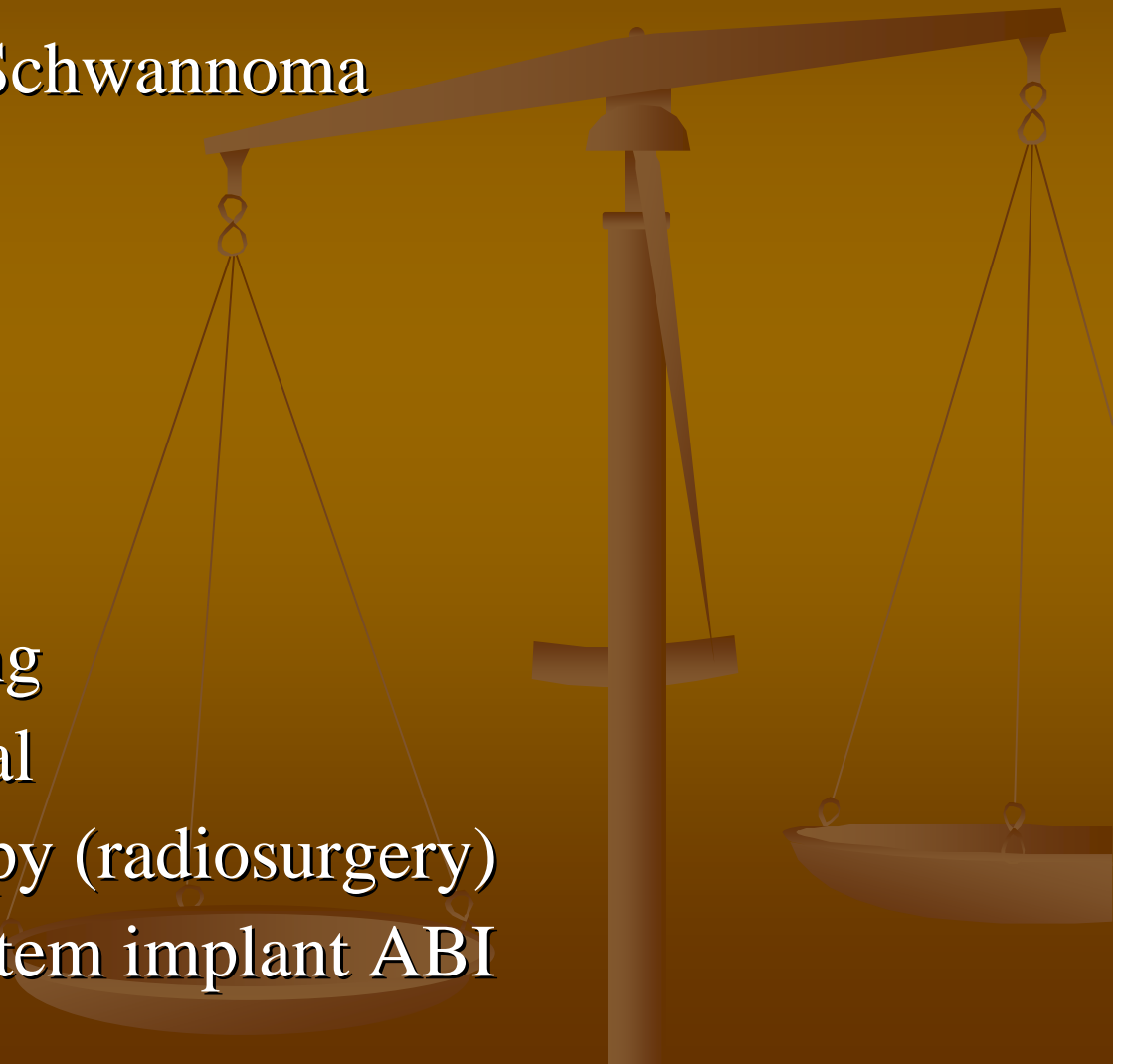


Dr. Abdulrahman Hagr
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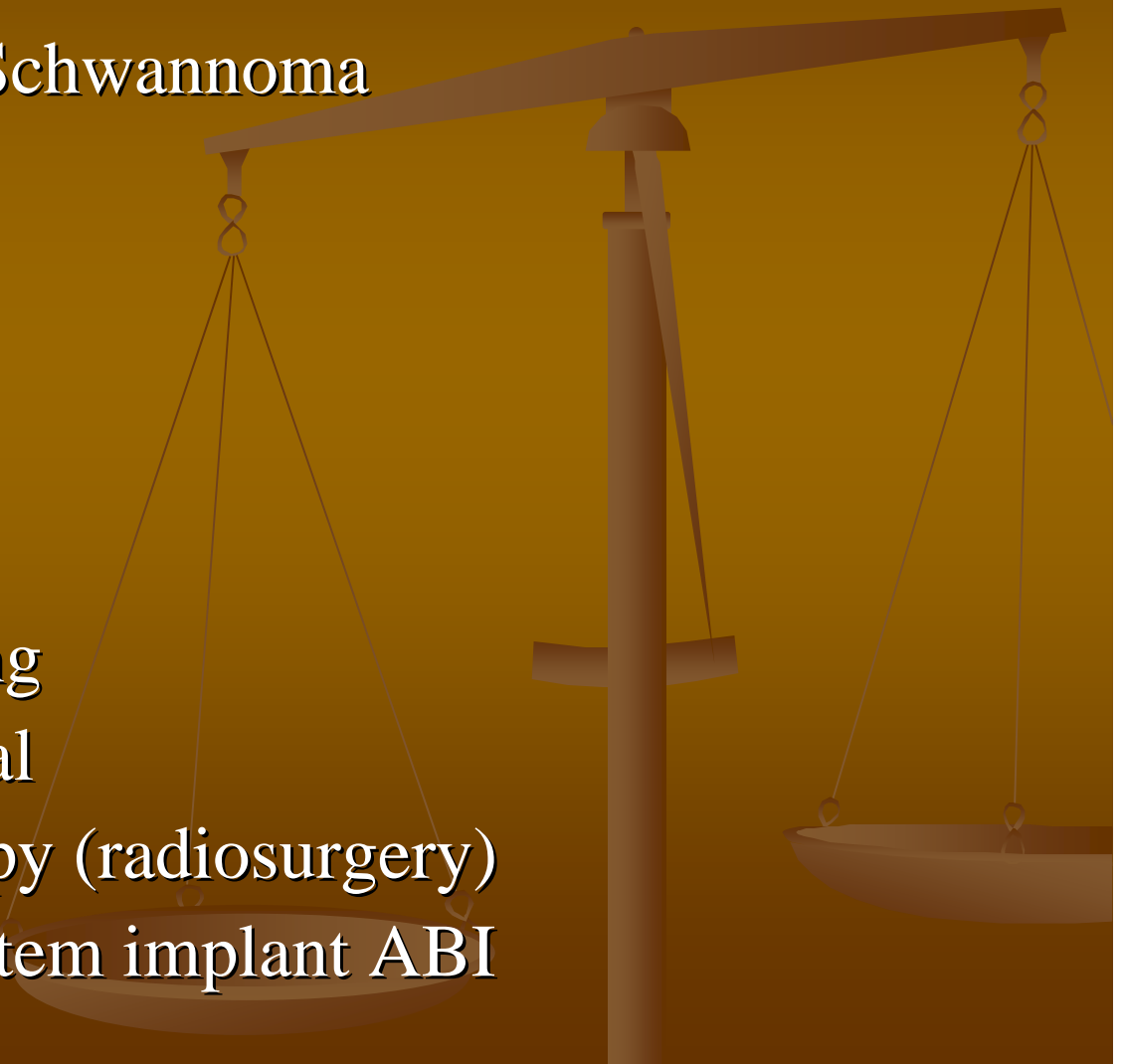
Neurofibroma II

- Introduction
- Neurofibroma vs Schwannoma
- N.I.H Dx
- NF2 Gene
- NF 2 Presentation
- Diagnosis
- Treatment
 1. Watchful waiting
 2. Surgical removal
 3. Radiation therapy (radiosurgery)
 4. Auditory brainstem implant ABI



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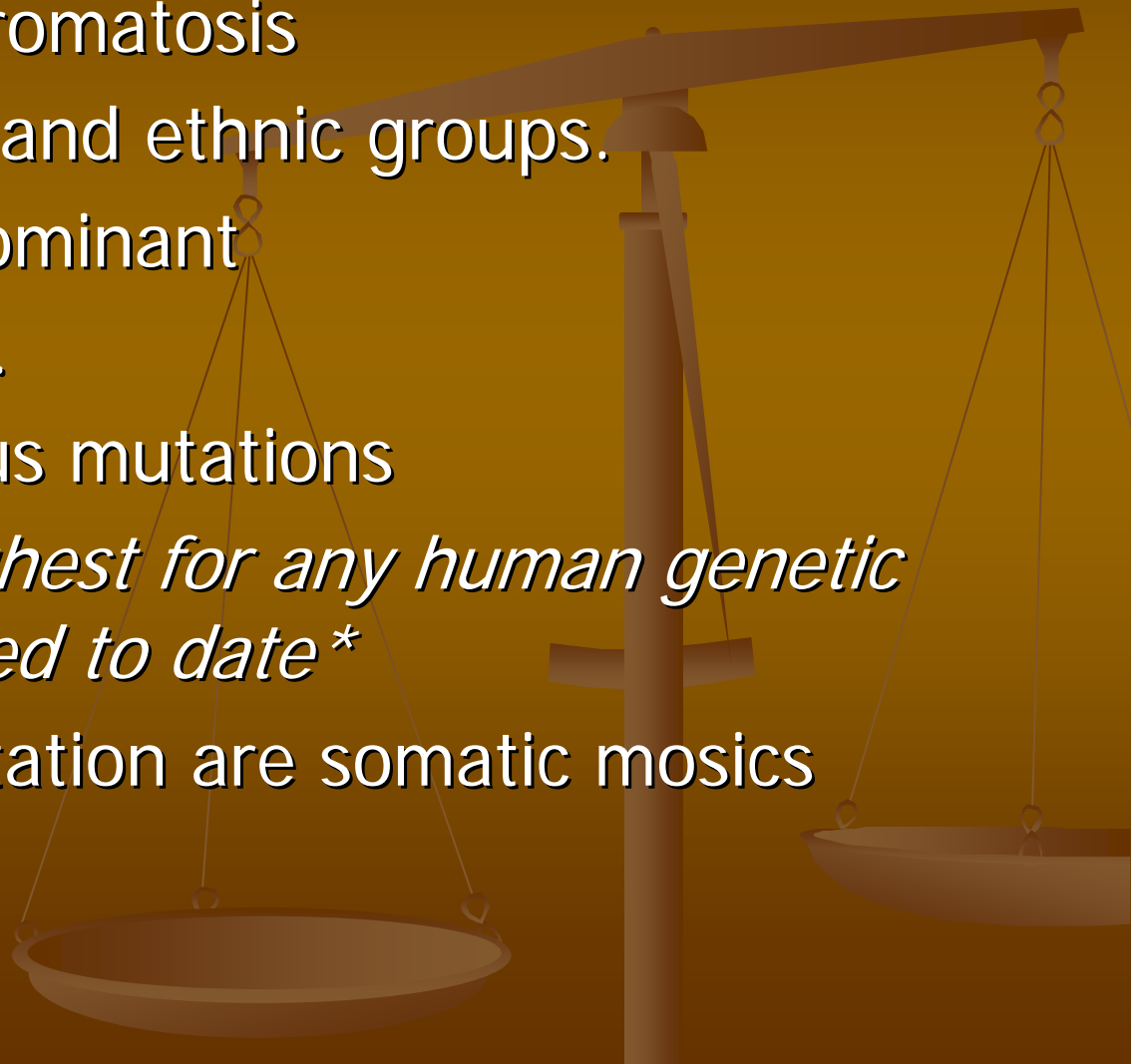
Neurofibroma II

- Central neurofibromatosis
- All races, sexes, and ethnic groups.
- AD autosomal dominant
- Chromosome 22.
- 50% spontaneous mutations

*Mutation is the highest for any human genetic disorder described to date**

- 20% of new mutation are somatic mosaics

*MacCollin 1998



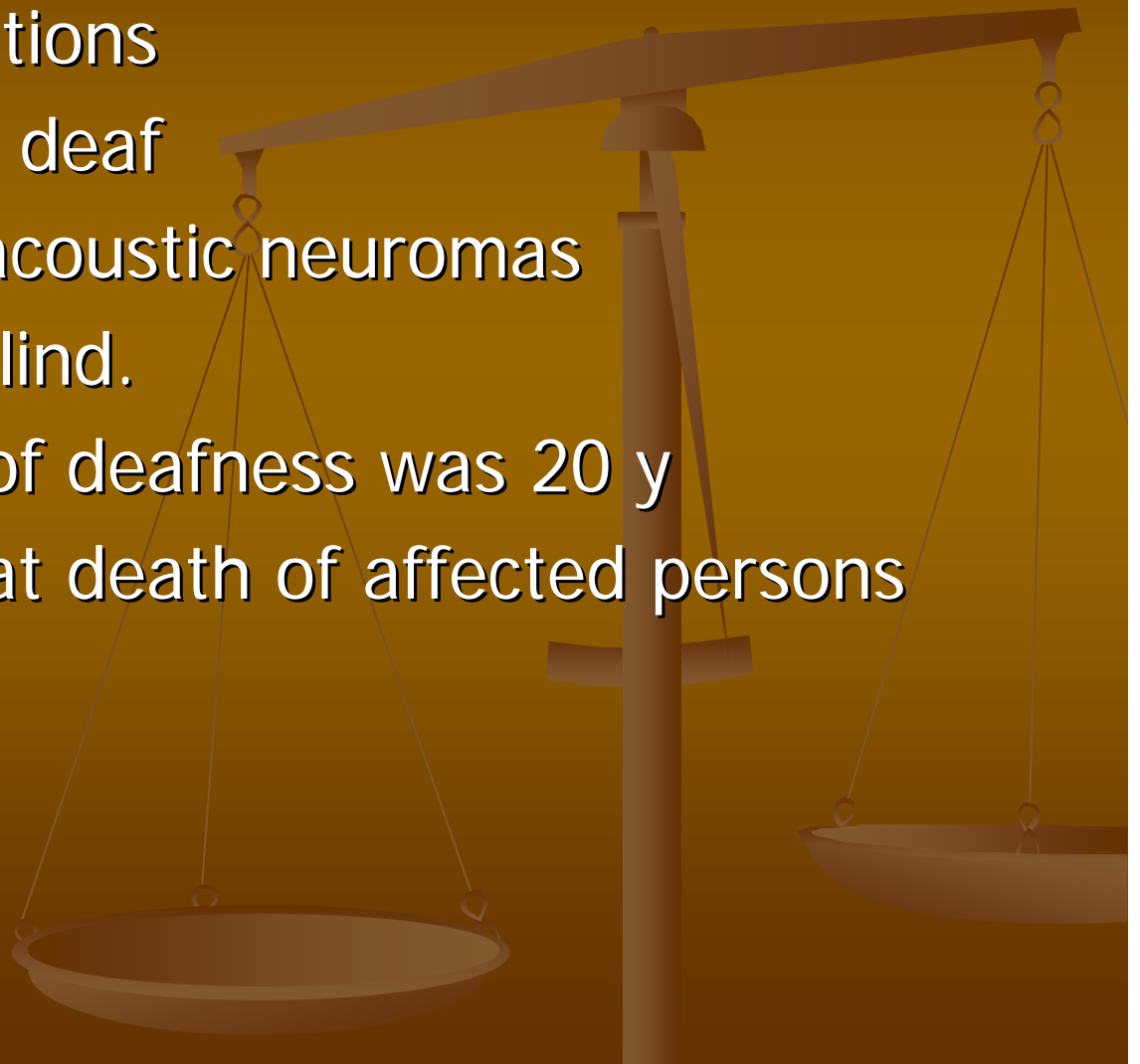
Wishart (1822)

- Wishart's (president of the Royal College of Surgeons of Edinburgh)
- First report of bil AN
- Michael Blair: 21 y old with bilateral deafness.
- Rt eye blindness 4 months after birth.
- Completely blind and deaf at the end of his life.
- Autopsy revealed tumors of the
 - Dura mater
 - Brain
 - Attached to auditory nerves just where they enter the meatus auditorius internus

History

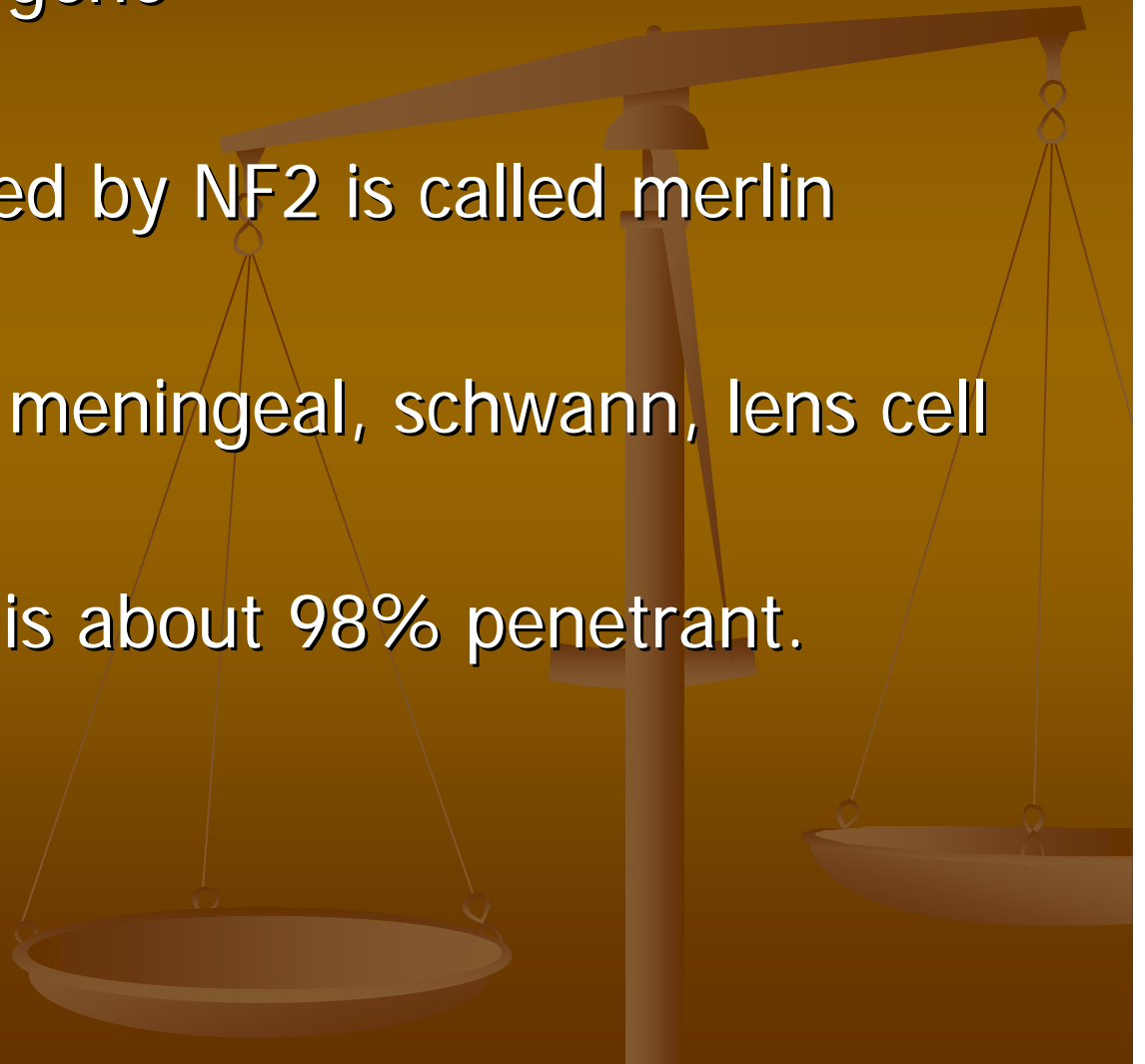
Gardner (1933)

- family of 5 generations
- 38 members were deaf
- All have bilateral acoustic neuromas
- 15 later became blind.
- The average age of deafness was 20 y
- The average age at death of affected persons
 - 2nd → 72 y
 - 3rd → 63 y
 - 4th → 42 y
 - 5th → 28 y



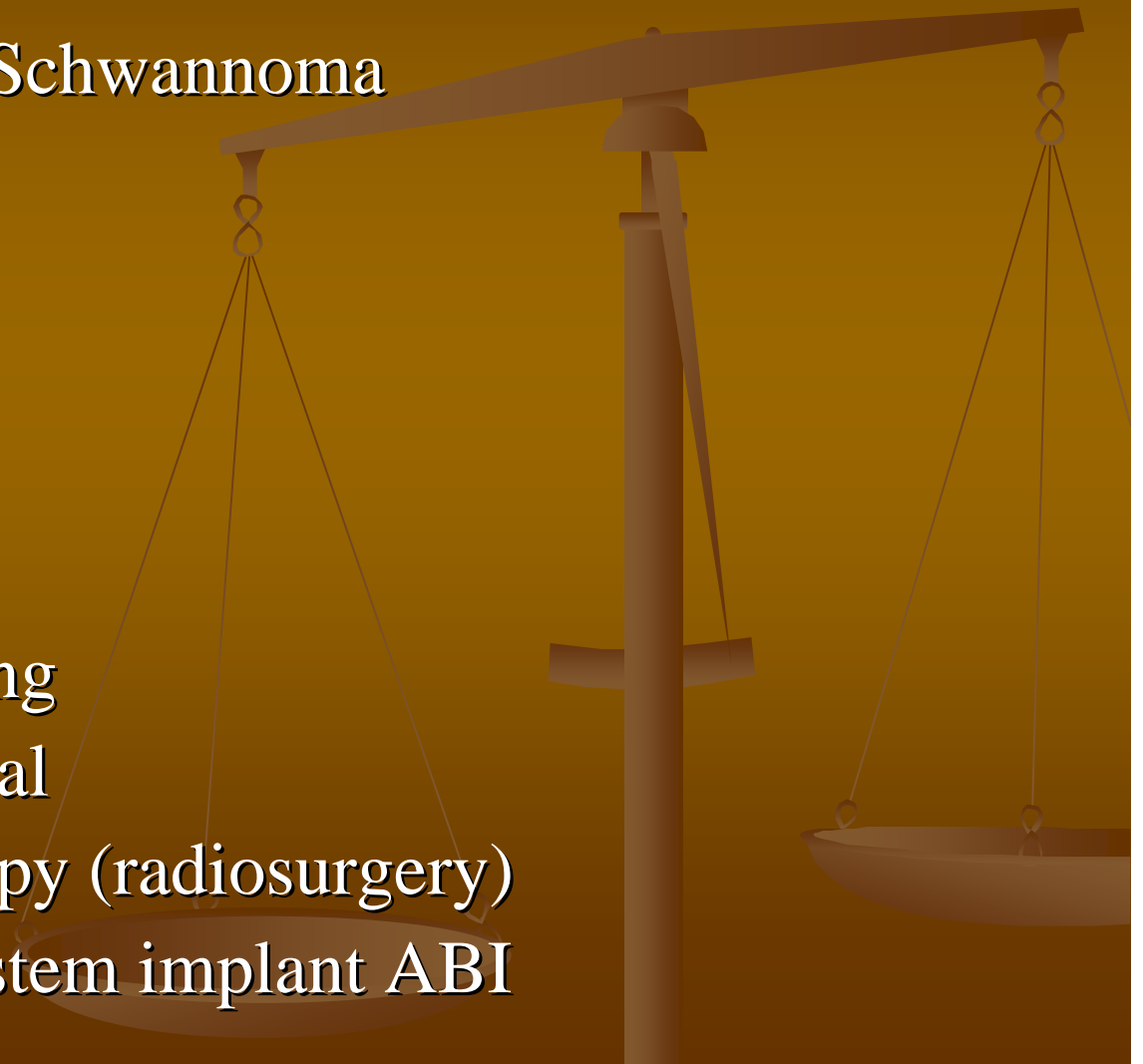
The NF2 Gene:

- Tumor suppressor gene
- The protein encoded by NF2 is called merlin
- Merlin expression: meningeal, schwann, lens cell
- The NF2 mutation is about 98% penetrant.



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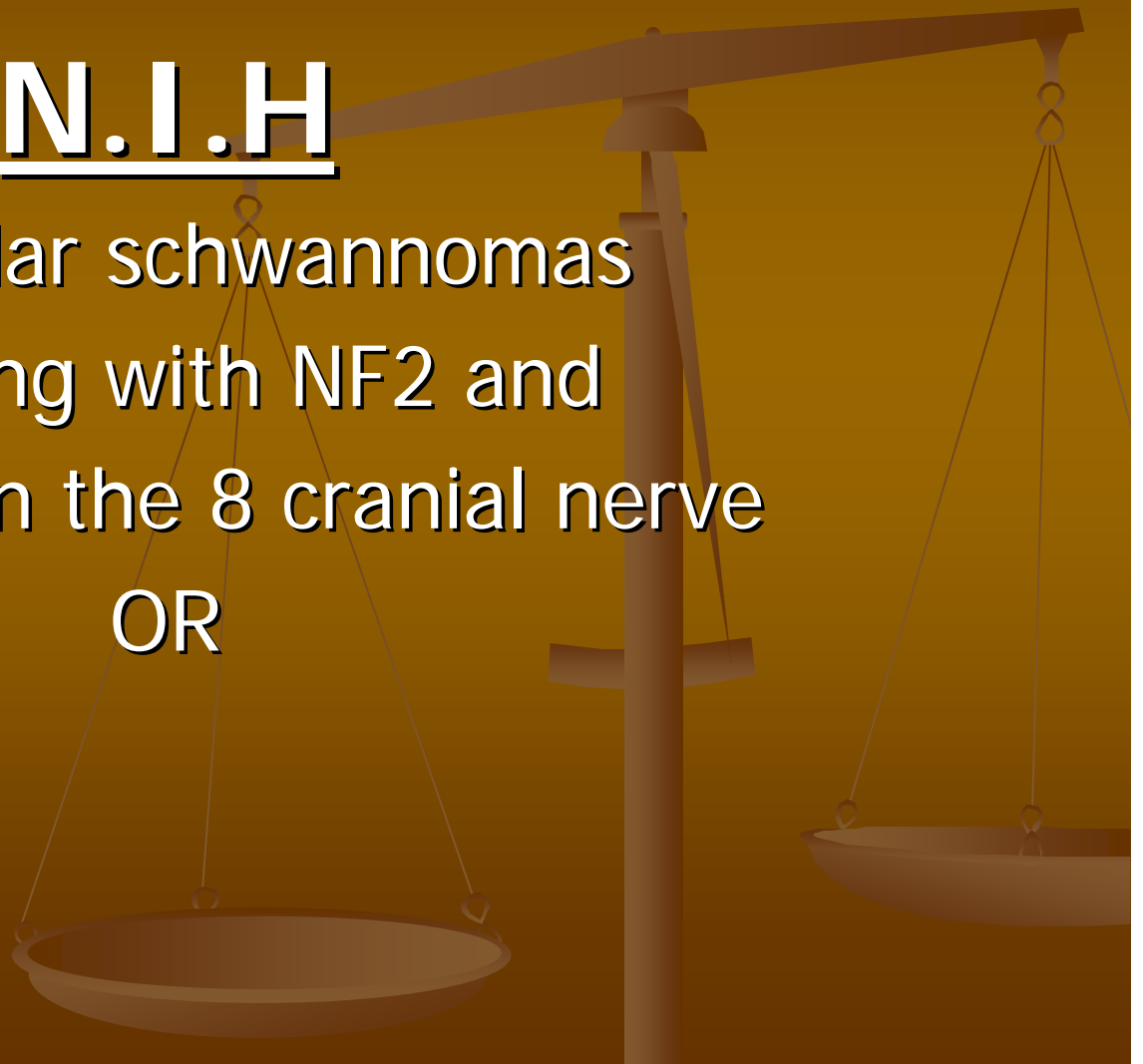


Neurofibroma II

N.I.H

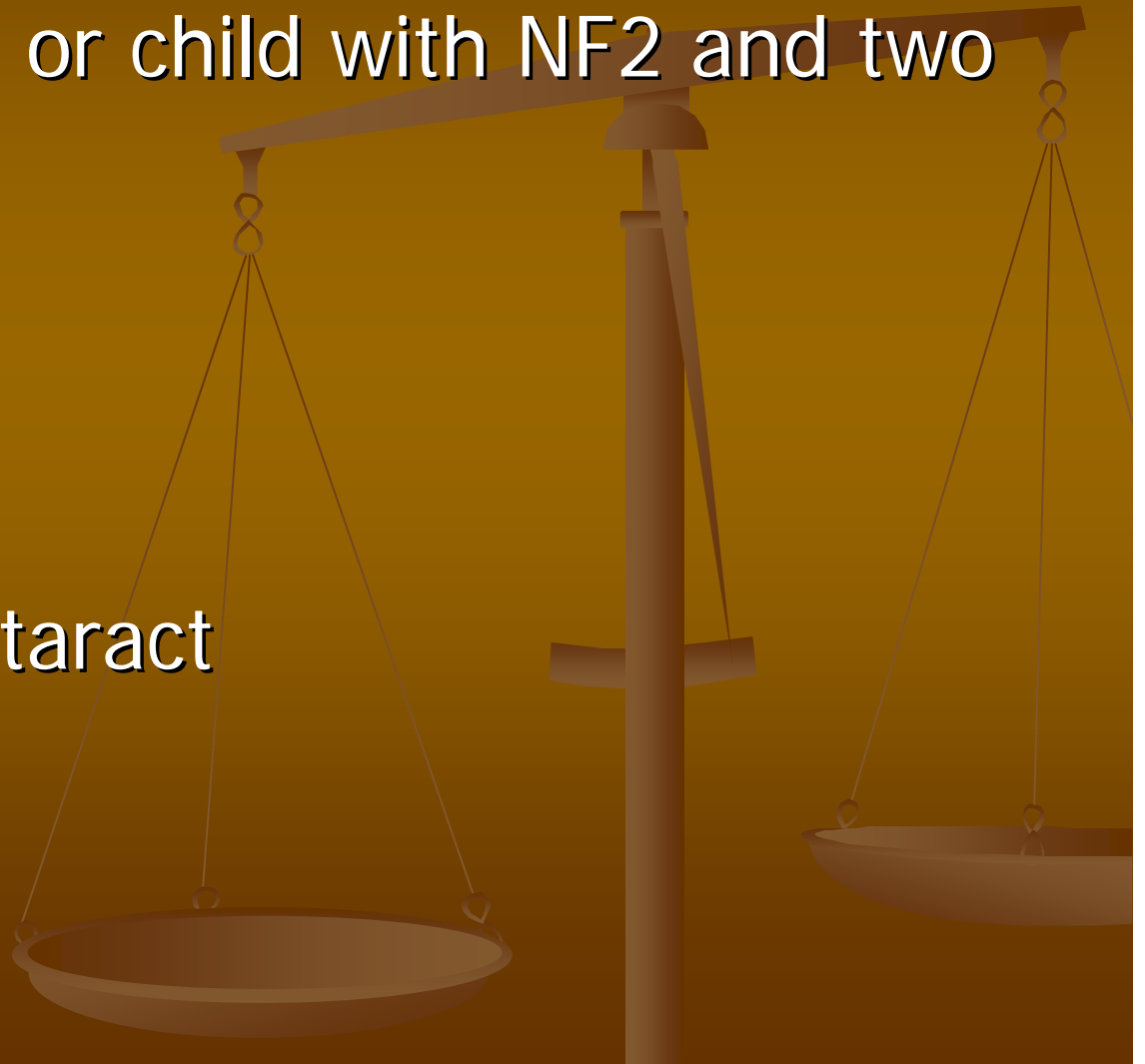
- Bilateral vestibular schwannomas
- A parent or sibling with NF2 and unilateral tumor on the 8 cranial nerve

OR



Neurofibroma II

- A parent, sibling or child with NF2 and two of the following:
 - Glioma
 - Meningioma
 - Schwannoma
 - Early onset cataract



Neurofibroma vs Schwannoma

- Benign tumors
- Arise from the Schwann cell



Neurofibroma vs Schwannoma

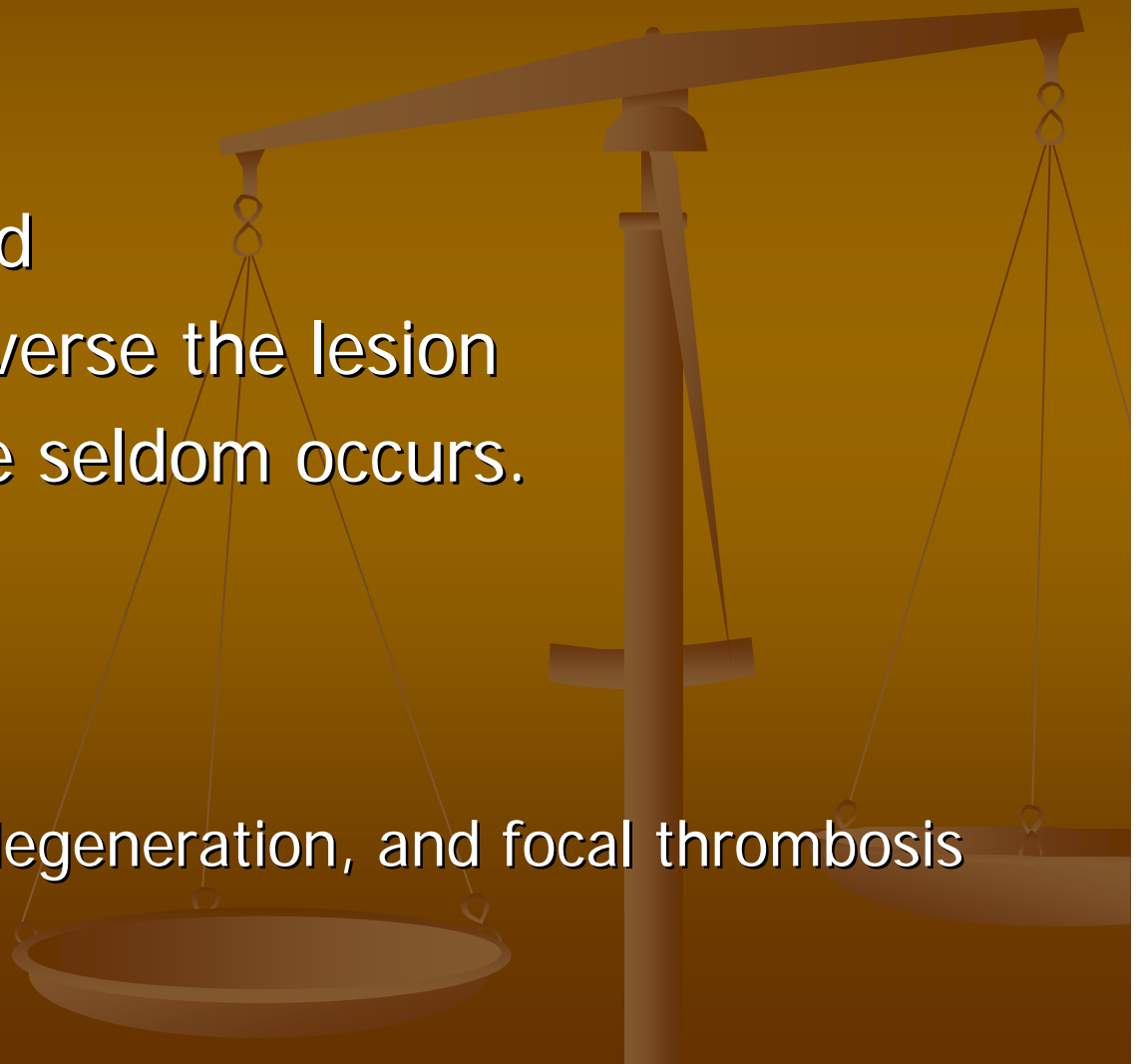
Neurofibroma

- Aggressive → Invade the cochlear nerve
- Ultimately → profound SNHL
 - Not encapsulated
 - Tumor between nerve fibers
 - Sarcomatous transformation
 - Histology
 - Spindle cell pattern with elongated and wavy nuclei
 - Uncommon Cystic and degenerative changes

Neurofibroma vs Schwannoma

Schwannoma

- Slow-growing
- Well-encapsulated
- Axons do not traverse the lesion
- Malignant change seldom occurs.
- Histology
 - Antoni type A
 - Antoni type B
 - Necrosis, cystic degeneration, and focal thrombosis are prominent



NF-2 Types

1. **Gardner**

2. **Wishart**

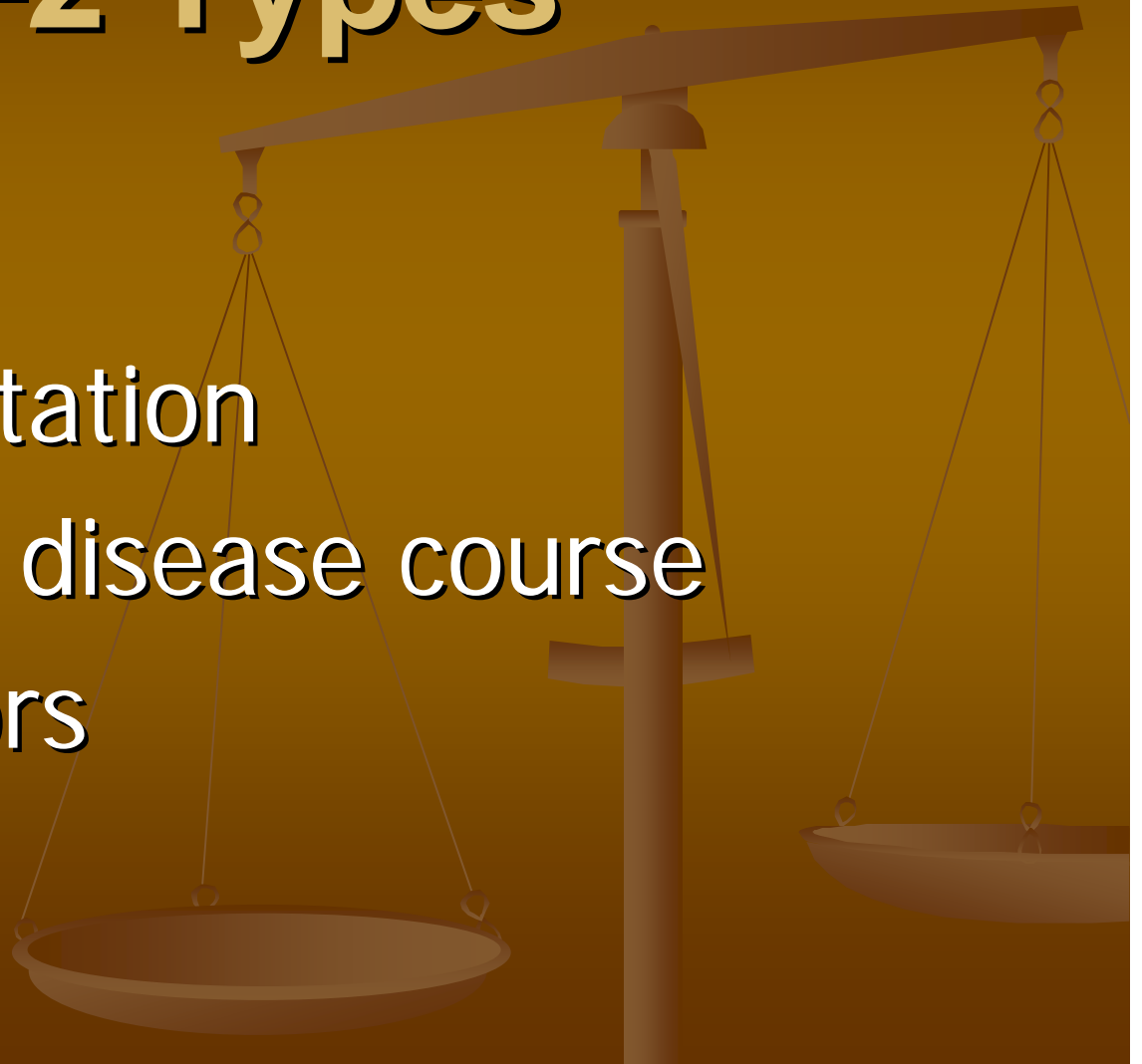
3. **Lee**



NF-2 Types

1. Gardner

- Late presentation
- Less severe disease course
- Fewer tumors



NF-2 Types



2. Wishart

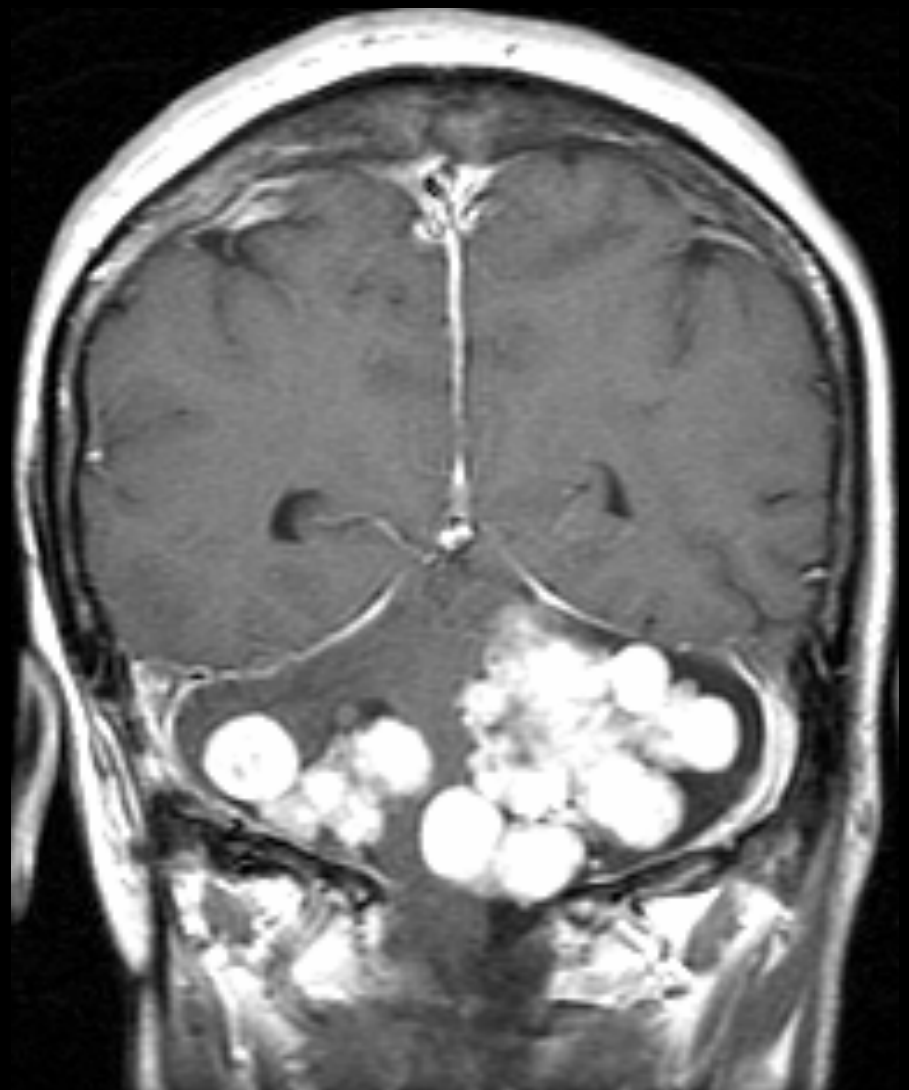
- Onset 20's
- Rapid and aggressive tumors
- Greater number of tumors present
- Impaired reproduction (more sporadic)

NF-2 Types



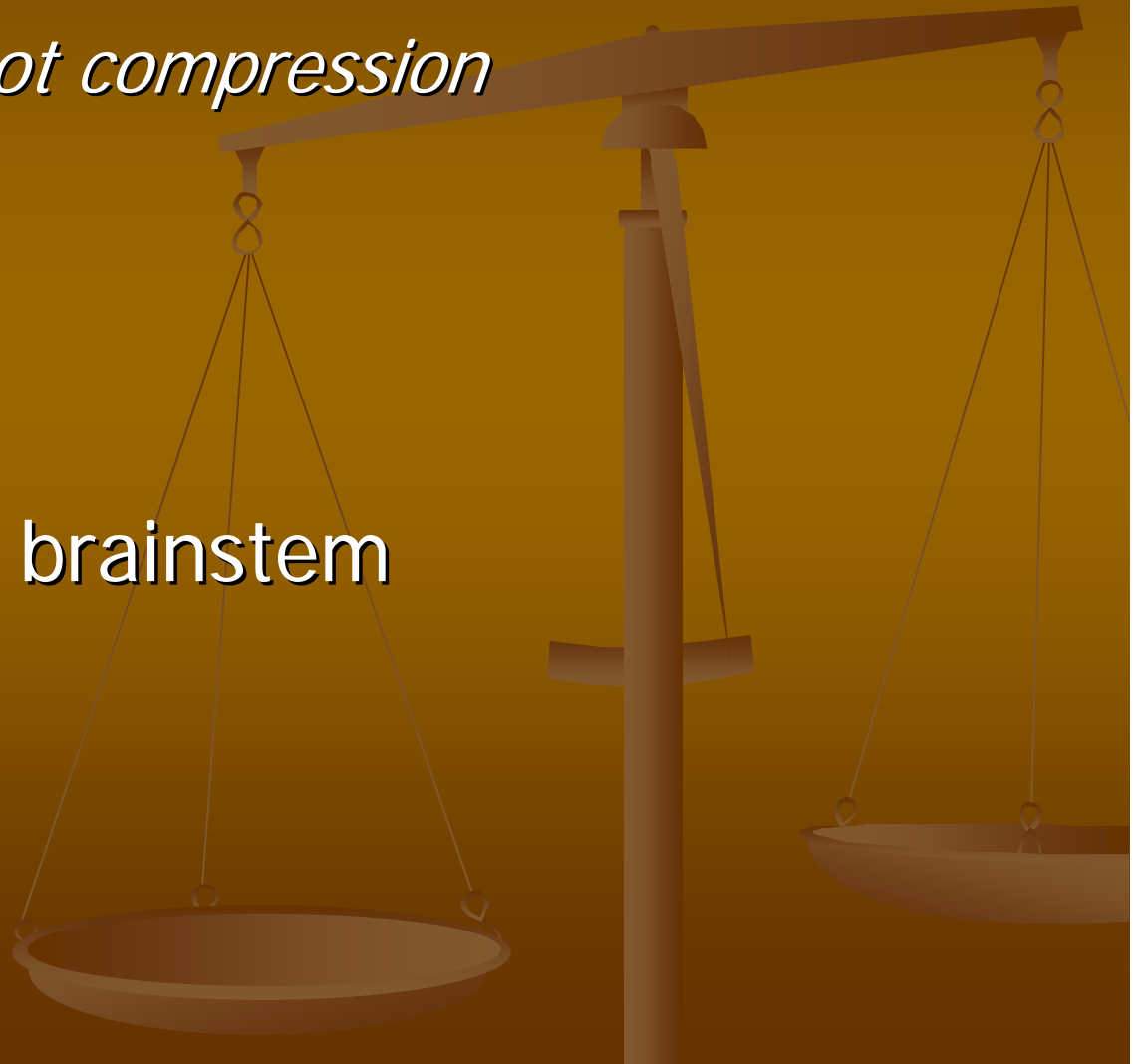
3. Lee & Abbott

- Variable age of hearing loss
- Early morbidity of numerous tumor
- 50 % has pigmented hairy skin lesion



NF2 Signs and Symptoms

- Late *infiltration not compression*
- Hearing loss
- Tinnitus
- Vertigo
- Pressure on the brainstem
- Headache
- Cataract



Diagnosis

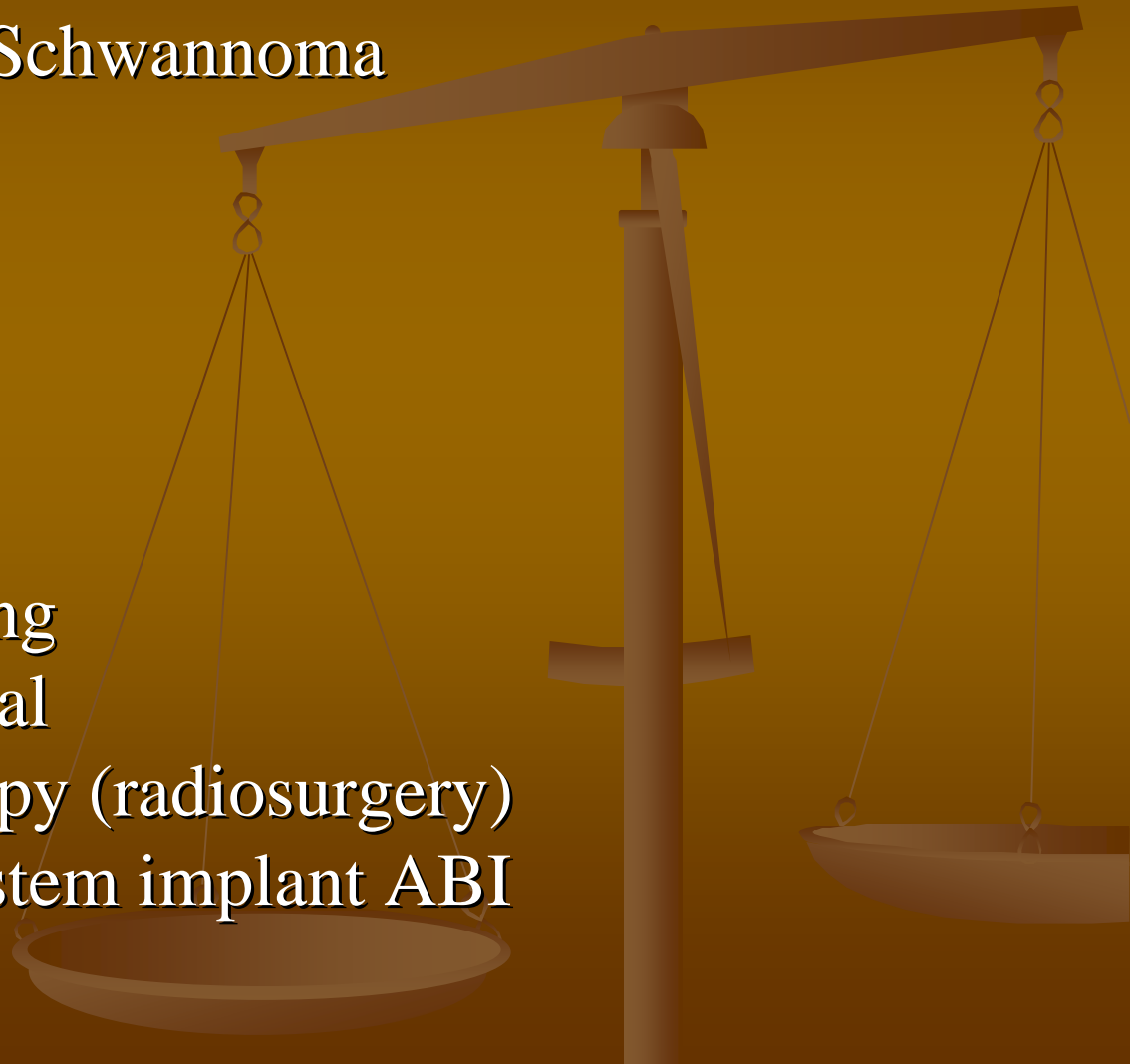


- NF2 gene product (MERLIN protein)
- Prenatal testing
 - Amniocentesis
 - Chorionic villi sampling
- MRI +Gado* (Ix of choice)
- ABR is poor screening*

Lalwani the Ame Jour of otol 1998 19:352-357

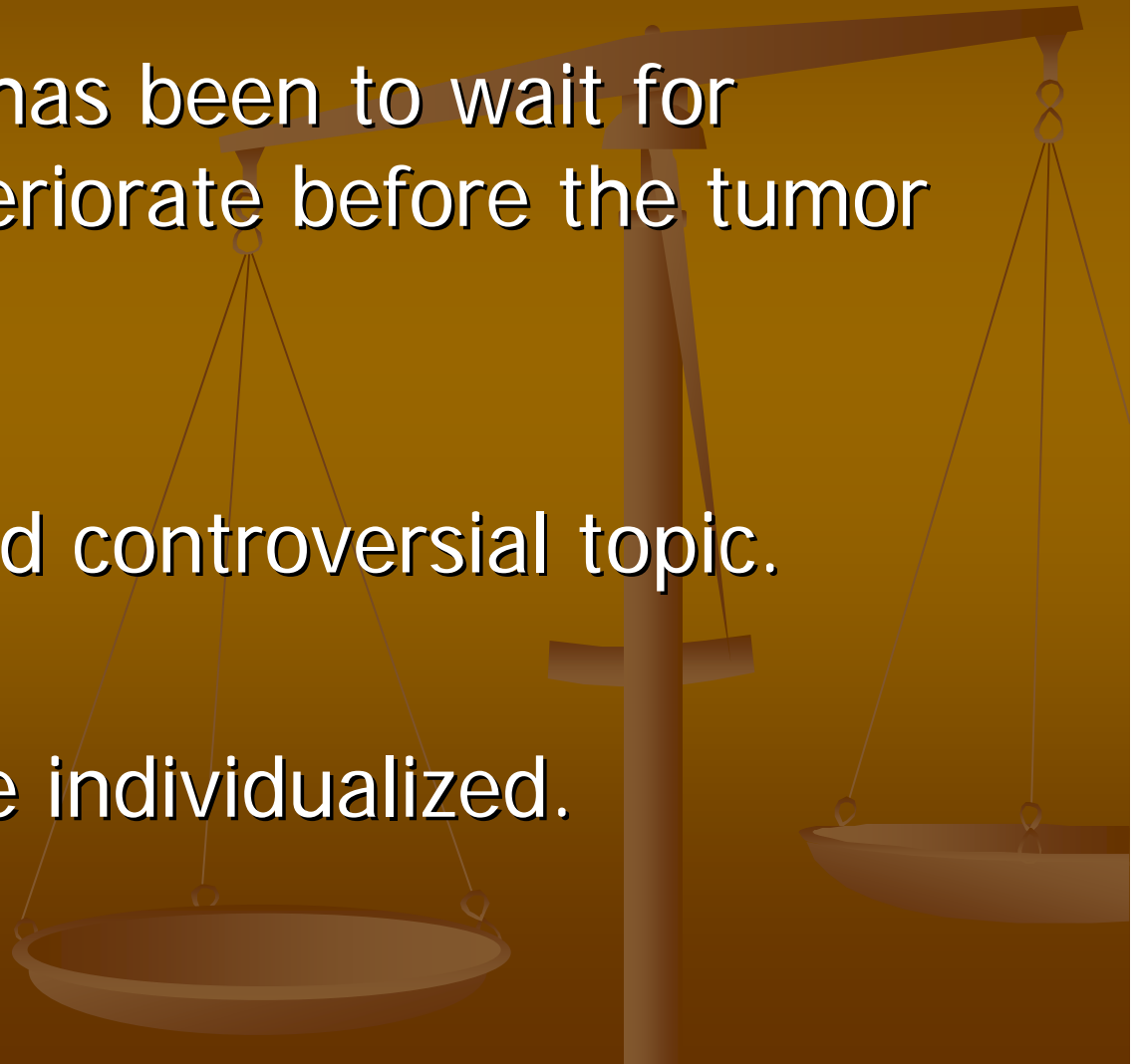
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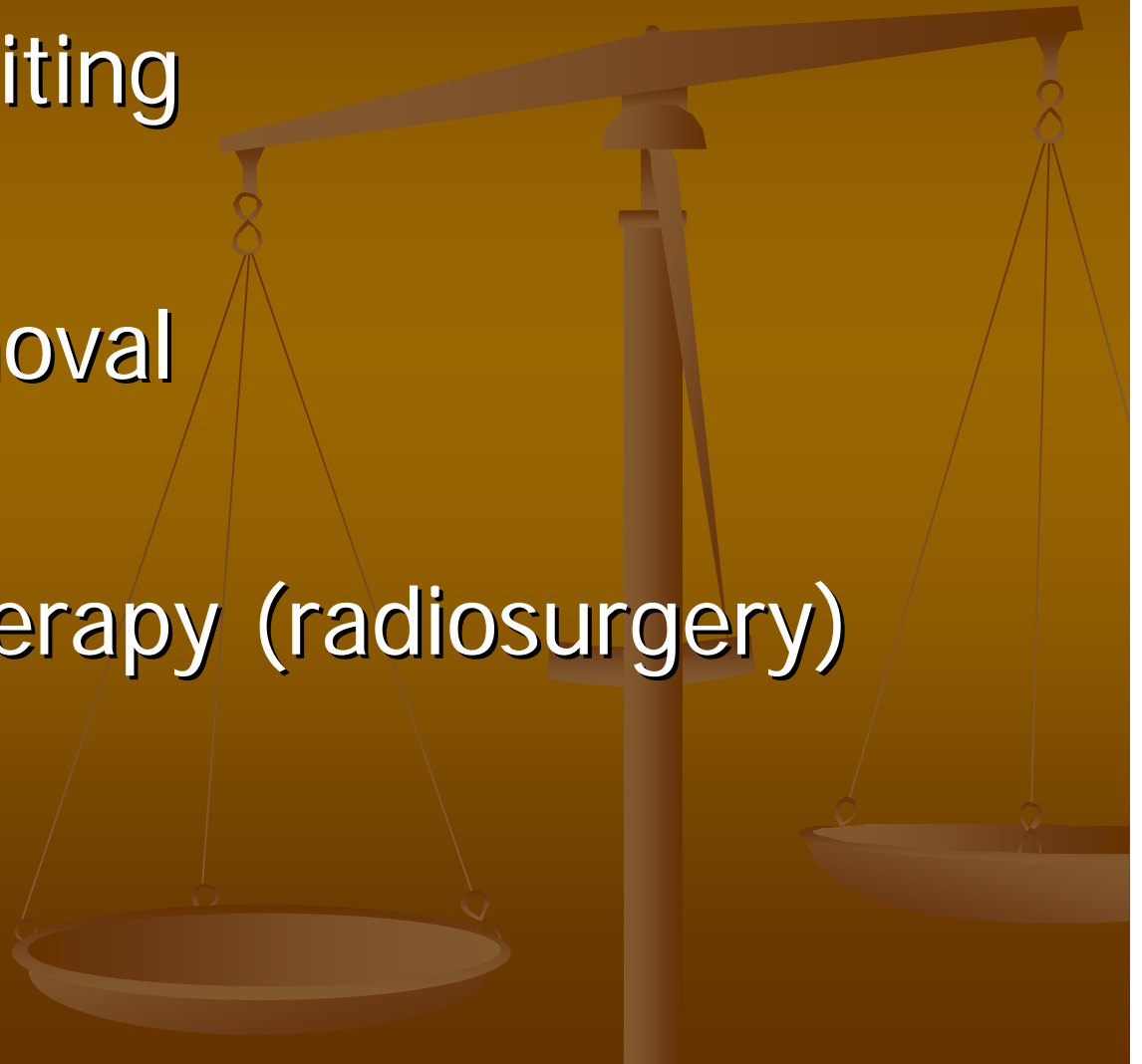
Treatments

1. The tendency has been to wait for hearing to deteriorate before the tumor is removed.
2. Challenging and controversial topic.
3. Treatments are individualized.



Treatments

1. Watchful waiting
2. Surgical removal
3. Radiation therapy (radiosurgery)



Watchful waiting

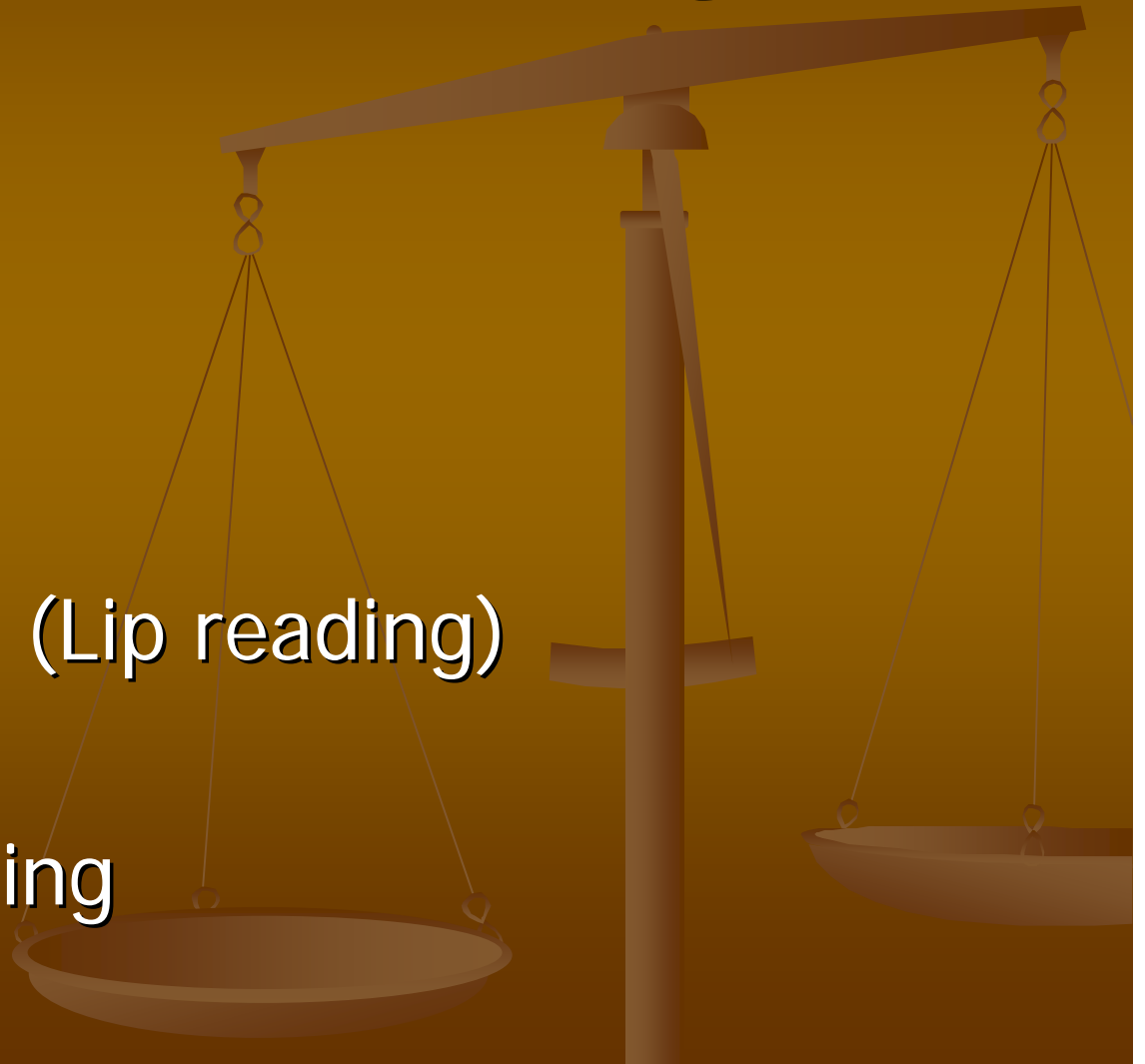
- Risk of bilateral hearing loss with Sx
- Growth decreases with with increase age
- Small non-progressing
- Serious medical issues
- Only hearing ear

Mautner et al 2002 96:223-228

Watchful waiting

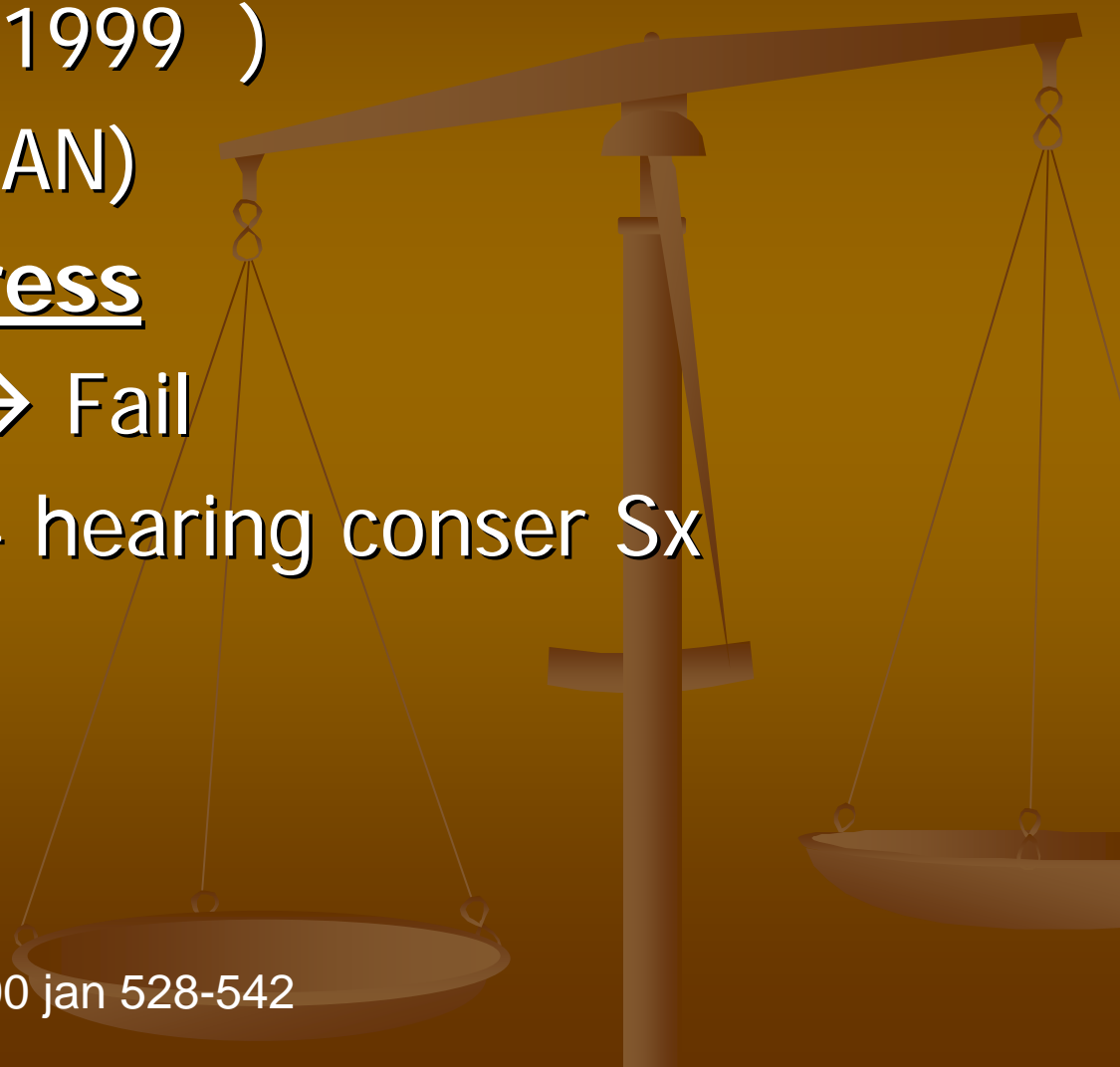
Follow up

- MRI
- Audiogram
- Education
- Rehabilitation (Lip reading)
- Counseling
- Family screening



Watchful waiting

- 15 years (1984-1999)
- 29 patients (48 AN)
- 68% no progress
- 5 patients XRT → Fail
- 10 Translab + 4 hearing conser Sx
- Early detection



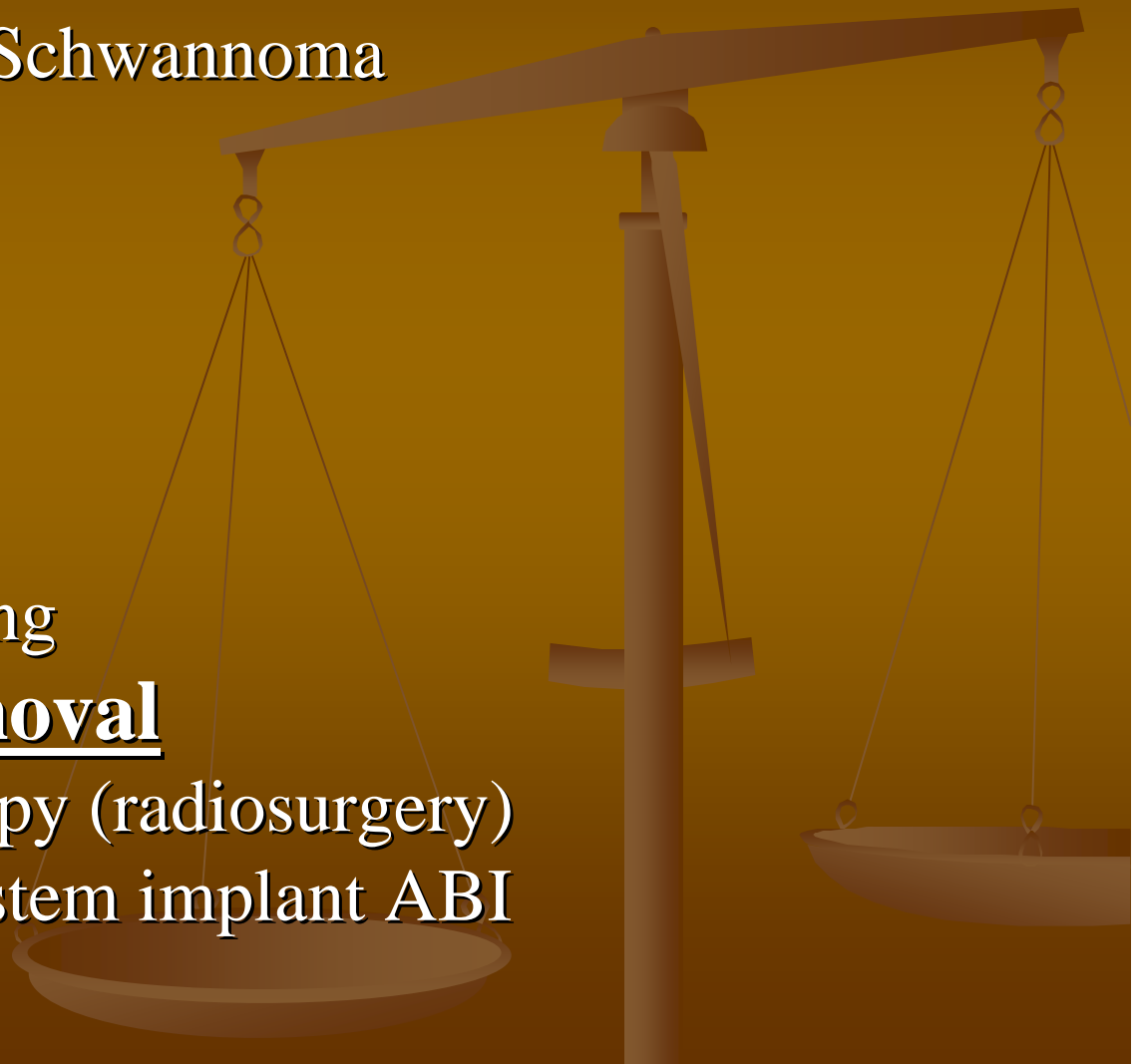
Watchful waiting

Masuda et al

- Retrospective of 63 NF2
- 2y F/U
- 73% no significant changing in hearing

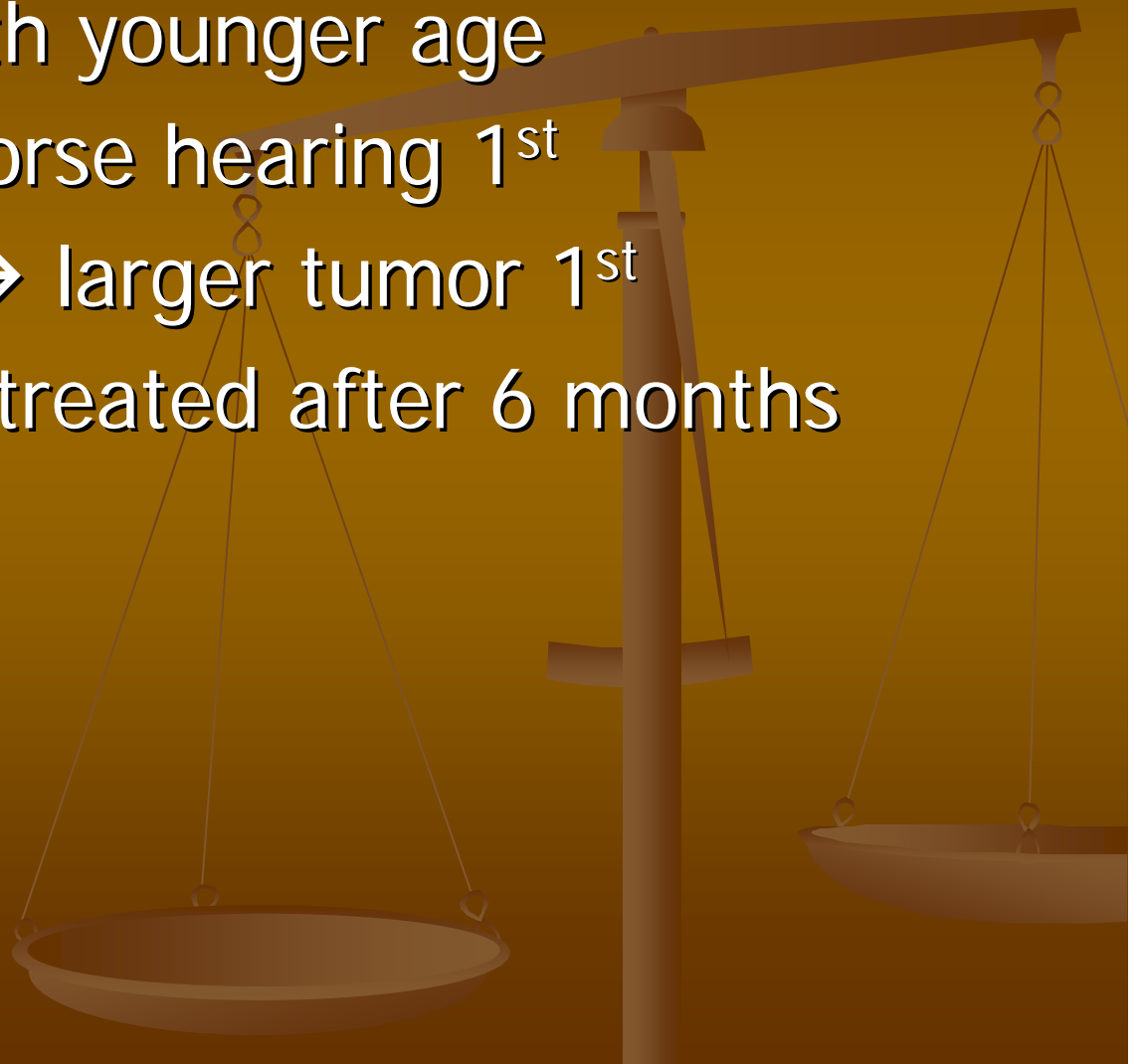
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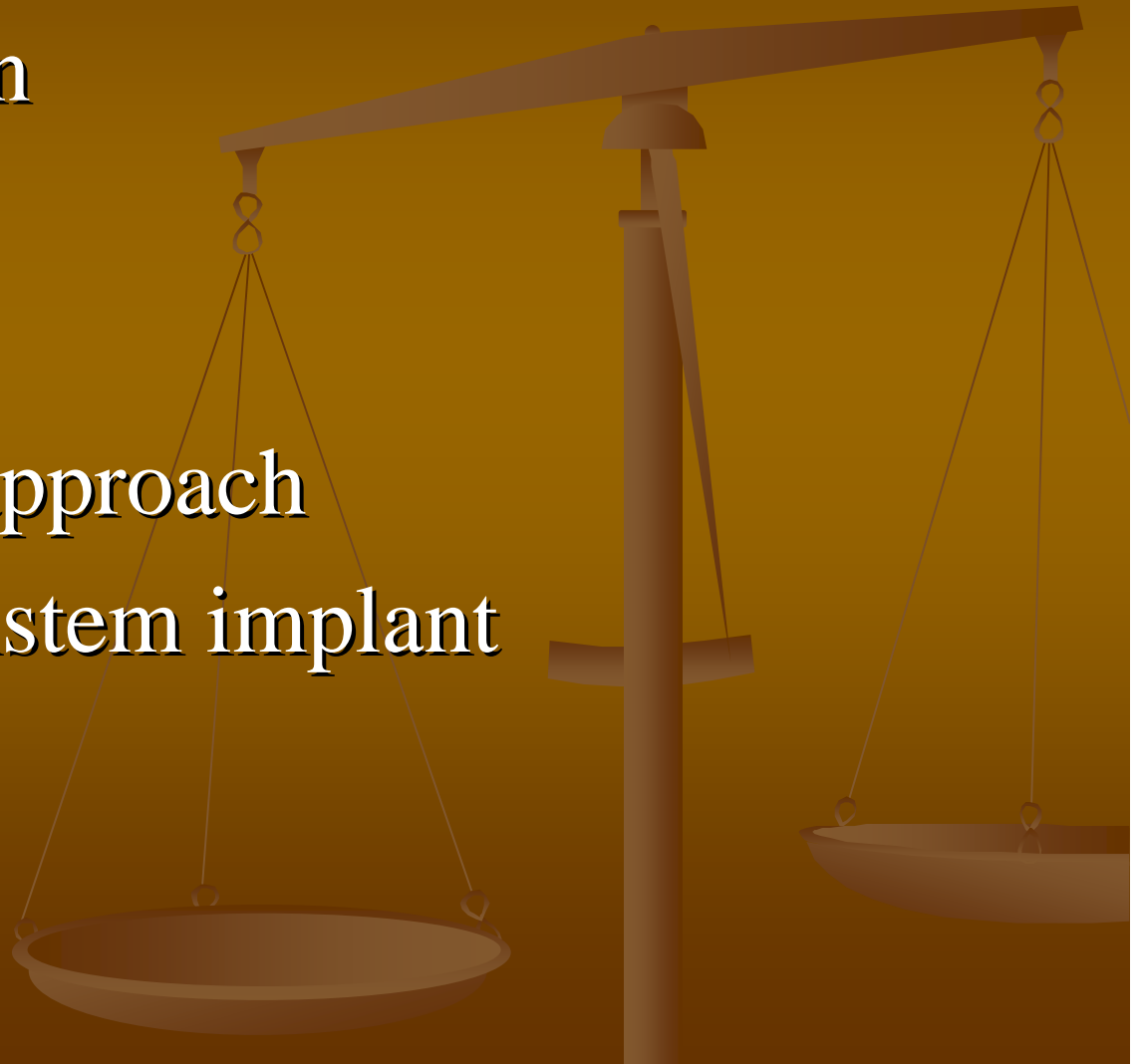
Surgery

- High growth with younger age
- Equal size → worse hearing 1st
- Equal hearing → larger tumor 1st
- Opposite ear is treated after 6 months



Surgery

- Decompression
- Tran-Lab
- Retro-sigmoid
- Middle fossa approach
- Auditory brainstem implant



Decompression

Brackmann 1990

- 1976-1988
- 5 Patients
- Growing and small <2 cm
- May improve and prolong useful hearing

Brackmann, Hitselberger **Laryngoscope**100:948–952, 1990.



Middle cranial fossa

- < 2.5 cm
- 1987: Hitselberger* 1st hearing preservation
- 1995: Hitselberger** 1st bilateral hearing preservation

**Hitselberger Am J Oto 1987 8:35-38*

***Hitselberger Am J Oto 1995 16 431-443*

Middle fossa craniotomy

- Hitselberger (1992-1996)* 18 Pat → 23 Sx
- Hitselberger (1988-1999)** 28 Pat → 40 Sx
- Bilateral acoustic tumors
- not the only hearing
- Small tumor < 2.5 cm
- Hearing preservation
 - PTA < 50 dB
 - 50% speech discrimination

*Brackmann Hitselberger *Am J Oto* 1998 638-643

**Brackmann Hitselberger *Neurosurgery*, Vol. 49, No. 2, August 2001

Middle fossa craniotomy

Brackmann Hitselberger

- 28 patients
- 11 patients → bilateral procedures.
- Mean age of 22.6 years.
- Mean size 1.1 cm
- All Gross total removal
- VII 80% Normal → 89.7% long term
- CSF leak, meningitis, redo..... 1 patient
- Audiometric data was 12.8 months

Brackmann Hitselberger Neurosurgery, Vol. 49, No. 2, August 2001

Tumor Size (cm)	No. of Cases
≤ 0.5	5 (12.5%)
0.6–1	24 (60.0%)
1.1–2.0	8 (20.0%)
2.1–3.0	2 (5.0%)
> 3	1 (2.5%)

TABLE 3. Preoperative versus Postoperative American Academy of Otolaryngology-Head and Neck Surgery Hearing Classification

Preoperative Class	No. of Cases				Total
	Postoperative Class A	Class B	Class C	Class D	
A	20 (58.8%)	3 (8.8%)		11 (32.4%)	34 (85%)
B		1 (33.3%)	1 (33.3%)	1 (33.3%)	3 (7.5%)
C			1 (33.3%)	2 (66.7%)	3 (7.5%)
Total	20 (50.0%)	4 (10%)	2 (5.0%)	14 (35.0%)	40

Middle fossa craniotomy

Brackmann Hitselberger Conclusion

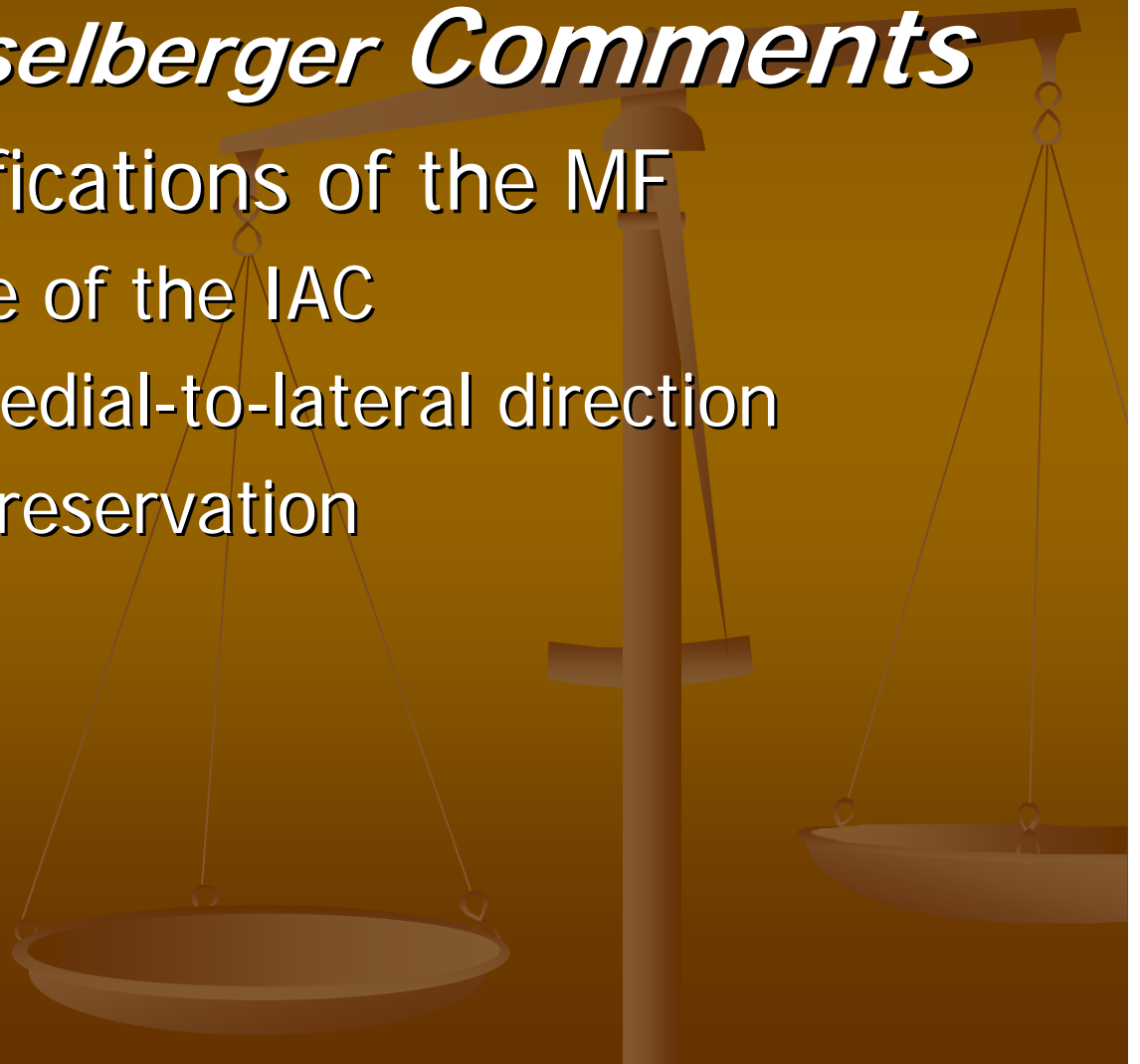
- Comparable to unilateral AN via the MF
- Better preoperative hearing →
better hearing preservation
- No relationship between the size of the tumor and hearing preservation.
- No factors predictive of hearing outcomes.

Middle fossa craniotomy

Brackmann Hitselberger Comments

Since 1992, modifications of the MF

- Larger exposure of the IAC
- Removal in a medial-to-lateral direction
- More Hearing preservation



Middle fossa craniotomy-2

Doyle

- 10 patients → 13 Sx
- Average age 26 y
- < 2 cm
- Hearing preservation 69%

Table 2. Preoperative and Postoperative Hearing Classification in all Middle Fossa Tumor Removal Operations

Hearing Result	Definition	Number of Cases		
		Preoperative (n = 13)	Postoperative	
			All Operations (n = 13)	Total Removal (n = 12)
Good	SRT ≤ 30 dB HL; SDS ≥ 70	9	2 (22%)	2 (22%)
Serviceable	SRT ≤ 50 dB HL; SDS > 50	13	9 (69%)	4 (44%)
Measurable	Any measurable hearing	13	9 (69%)	8 (61%)

Doyle Am J Oto 1993

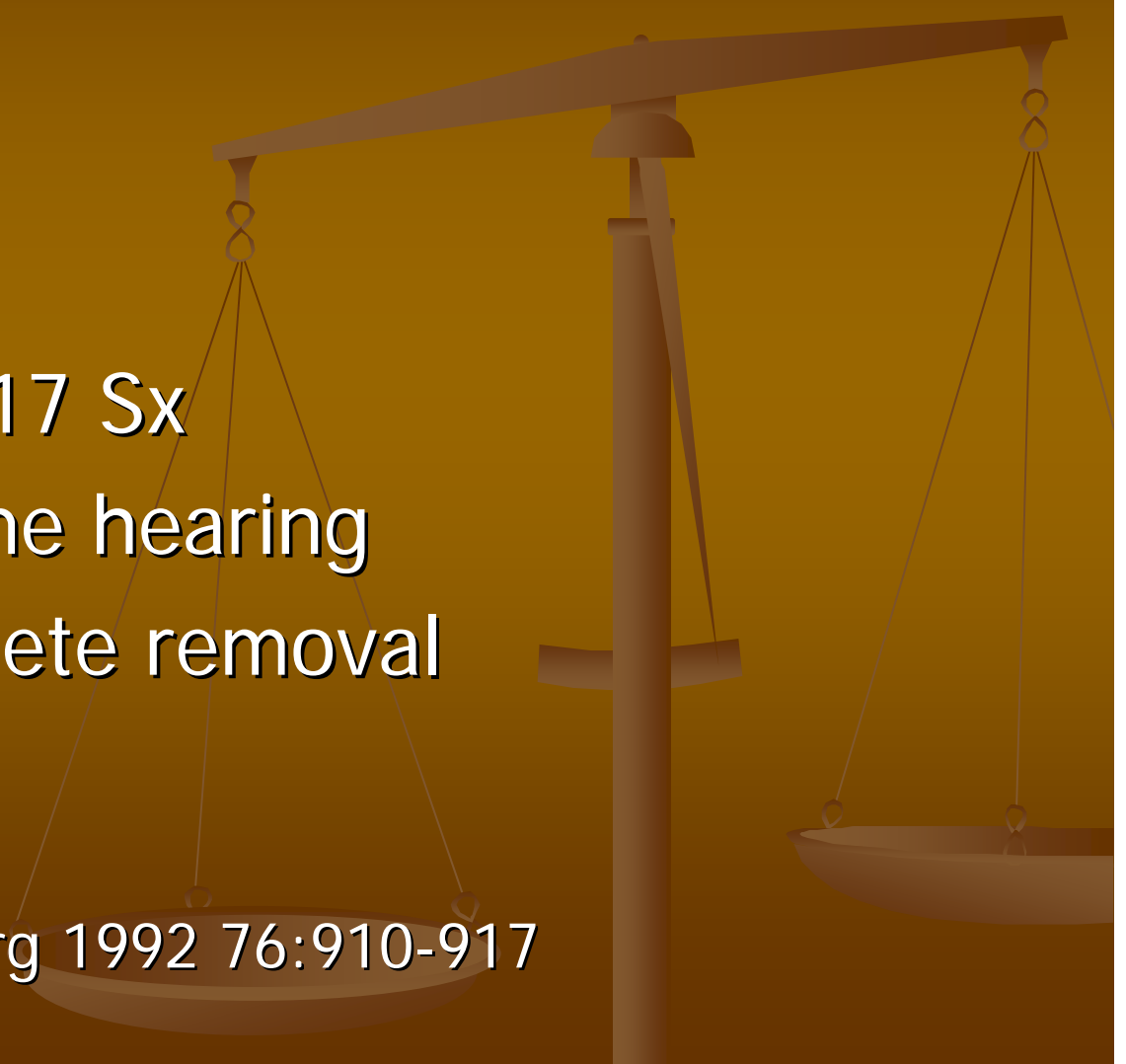
Retro-sigmoid

- Small series

Fischer et al

- 14 patients → 17 Sx
 - 3 (17.6%) some hearing
- Incomplete removal

Fischer et al J Neurosurg 1992 76:910-917

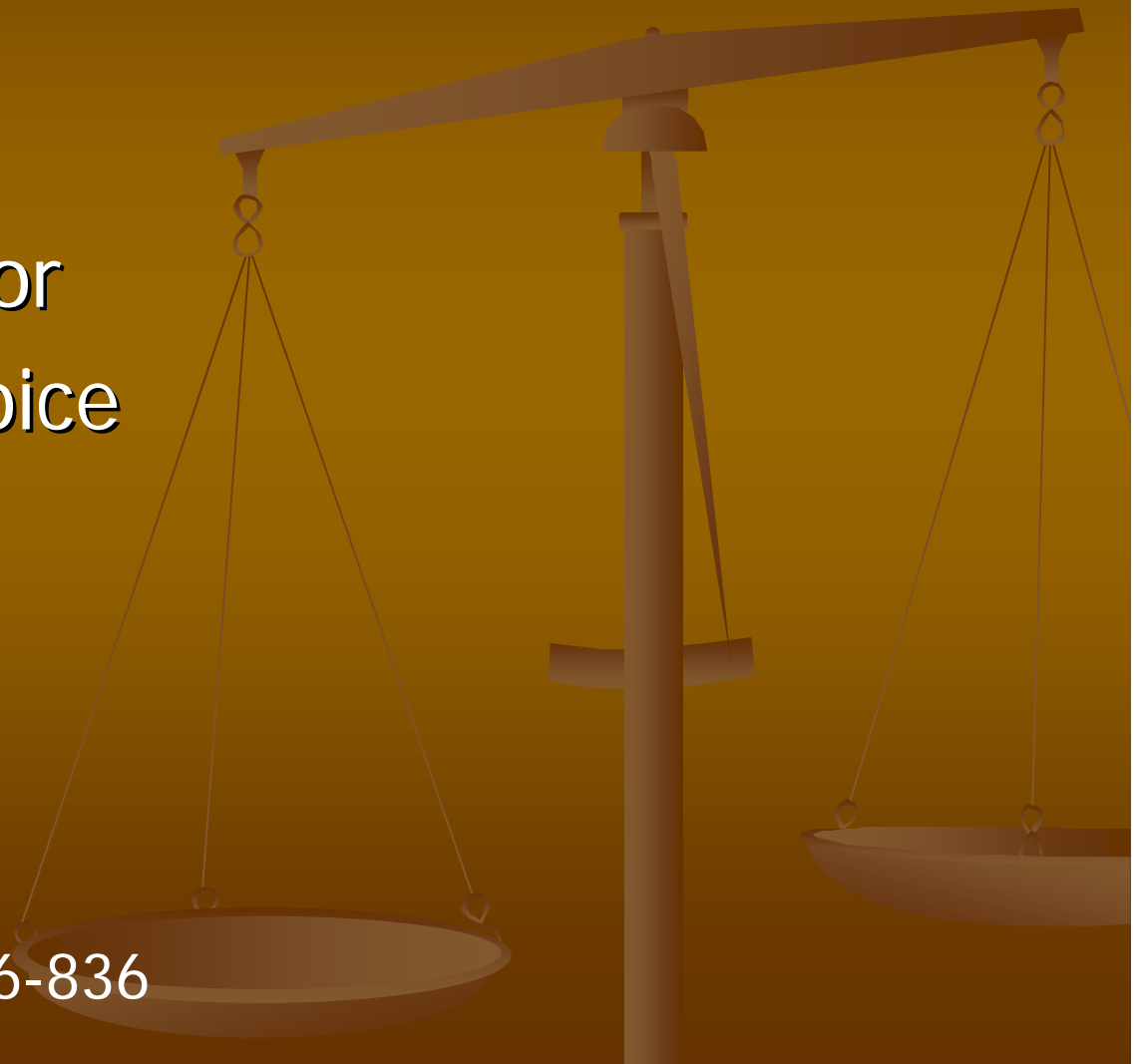


Retrosigmoid ABI

Colletti

- 5 patients NF2
- 12-30 mm Tumor
- Approach of choice

Colletti et al 2000 21:26-836



Cochlear Implant

- Electrocochleography
- AN initially cause a cochlear deafness
 - Vascular compression
 - Thrombosis of the labyrinthine arteries
- Preserved auditory nerve → C.I.

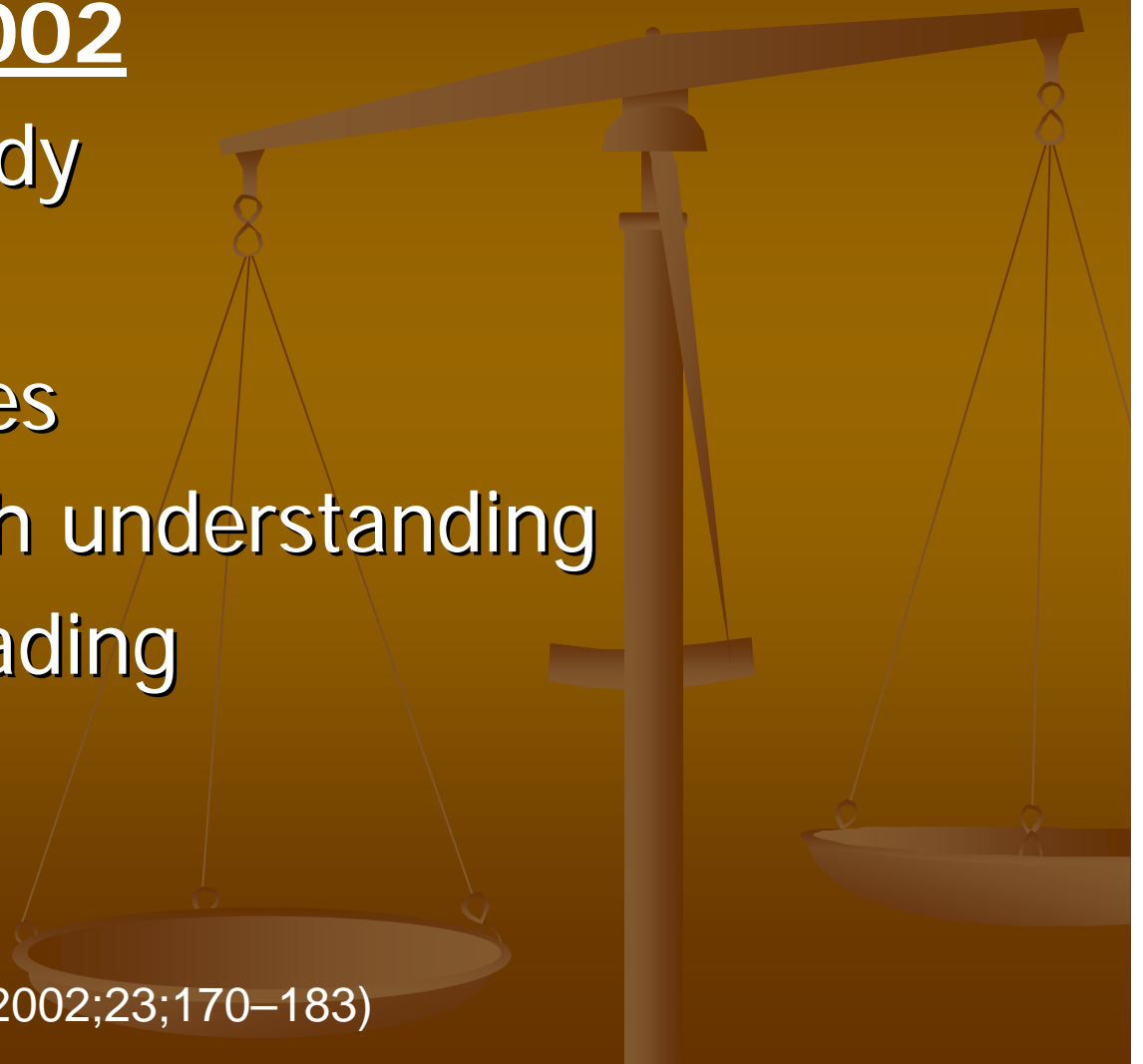
Marangos, *The Journal of Laryngology & Otology* 2000, pp. 3–7

A.B.I

Nevison et al 2002

- Multi-center study
- 27 patients NF2
- 8.6/21 electrodes
- Open-set speech understanding
- 2 without lip reading

Nevison, Ear & Hearing 2002;23;170–183)



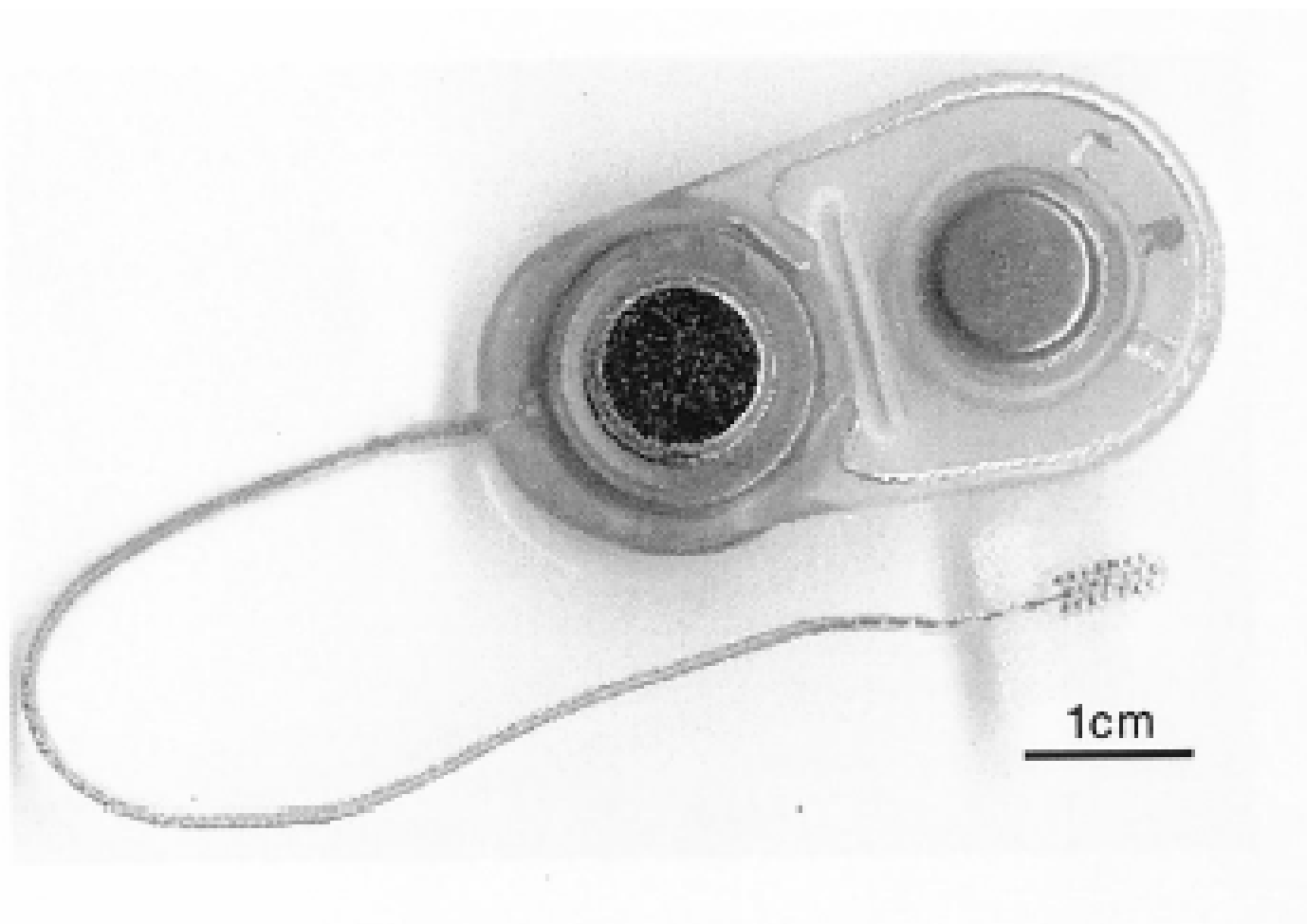


Figure 1. The Nucleus 21-channel Auditory Brainstem Implant (ABI) based on the Nucleus 22 receiver/stimulator package.

TABLE 2. Demographics of the 27 implanted patients.

	Subject (Nationality)	Sex	Age at Implantation (yr)	Etiology	Tumor Size (largest diameter)	Implant Side	1st or 2nd Side	Operation Date	Surgical Approach	Follow-up Clinic
1	HK (D)	F	52	NF2*	1.5 cm	L	1st	7 Sep 92	T.lab	Freiburg
2	GS (D)	M	58	NF2	1.2 cm	L	1st	23 Nov 93	T.lab	Freiburg
3	AL (F)	M	31	NF2	7.0 cm	L	2nd	25 Nov 93	T.lab	Freiburg
4	SG (D)	M	30	NF2	1.5 cm	R	1st	10 Feb 94	T.lab	Freiburg
5	JB1 (GB)	F	33	NF2	2.0 cm	R	1st	25 Feb 94	T.lab	Manchester
6	RF (D)	F	34	NF2	2.0 cm	R	1st	27 Jun 94	T.lab	Freiburg
7	PF (F) ‡	M	34	NF2	3.0 cm	R	2nd	28 Jun 94	T.lab	Freiburg
8	CH1 (D)	M	29	NF2	5.0 cm	L	2nd	14 Feb 95	T.lab	Freiburg
9	FU (D)	M	41	NF2	1.5 cm	L	2nd	16 Feb 95	T.lab	Freiburg
10	MP1 (Ru)	M	17	NF2**	5.0 cm	R	1st	14 Dec 95	T.lab	Toulouse
11	ID (D)	M	40	NF2	5.0 cm	R	2nd	7 Feb 96	T.lab	Freiburg
12	MK (GB)	M	32	NF2	3.0 cm	L	2nd	8 Feb 96	T.lab	Manchester (imp. Freiburg)
13	AT (GB)	M	24	NF2	2.5 cm	R	2nd	9 Feb 96	T.lab	Manchester (imp. Freiburg)
14	TL (D)	M	29	NF2	1.0 cm	L	2nd	7 May 96	R.sig	Hannover
15	BC (D)	F	35	NF2	4.5 cm	R	2nd	25 Jul 96	R.sig	Hannover
16	AA (D)	M	51	NF2	0 cm	R	2nd	25 Jul 96	R.sig	Hannover
17	IG (F)	F	30	NF2	0 cm	R	2nd	23 Sep 96	T.lab	Paris
18	MP2 (D)	F	24	NF2	3.5 cm	R	2nd	7 Nov 96	R.sig	Hannover
19	TK (D)	F	31	NF2	4.0 cm	L	2nd	12 Nov 96	R.sig	Hannover
20	NW (D)	F	13	NF2**	NR	R	1st	24 April 97	T.lab	Freiburg
21	CH2 (D)	M	39	NF2	NR	L	2nd	24 April 97	T.lab	Freiburg
22	FD (F) + blind	F	44	NF2	1 cm	R	1st	5 May 97	T.lab	Paris
23	LP (N)	F	42	NF2*	2.5 cm	L	1st	21 May 97	T.lab	Uppsala
24	JB2 (S)	M	24	NF2*	3 cm	R	1st	22 May 97	T.lab	Uppsala
25	CR (E)	F	20	NF2	2.5 cm	L	1st	23 June 97	T.lab	Pamplona
26	CM (F)	M	30	NF2	3 cm	R	2nd	30 June 97	T.lab	Paris
27	SM (I)	F	27	NF2	2.5 cm	R	2nd	23 Aug 97	R.sig	Verona
Mean			33.1 yr ($\sigma = 10.5$)		2.71 cm ($\sigma = 1.69$)					

* Irradiated

Radiation therapy

Subach 1999

- University of Pittsburgh
- Median follow-up period of 36 months
- Normal VII 81% (92% MF)
- hearing preservation 43% (65% MF)
- No further growth 98% (99.7% MF)
- Uncertain long-term efficacy
- Not for brainstem decompression

Subach J Neurosurg. 1999 May;90(5):815-22

***Brackmann Hitselberger Neurosurgery, Vol. 49, No. 2, August 2001*

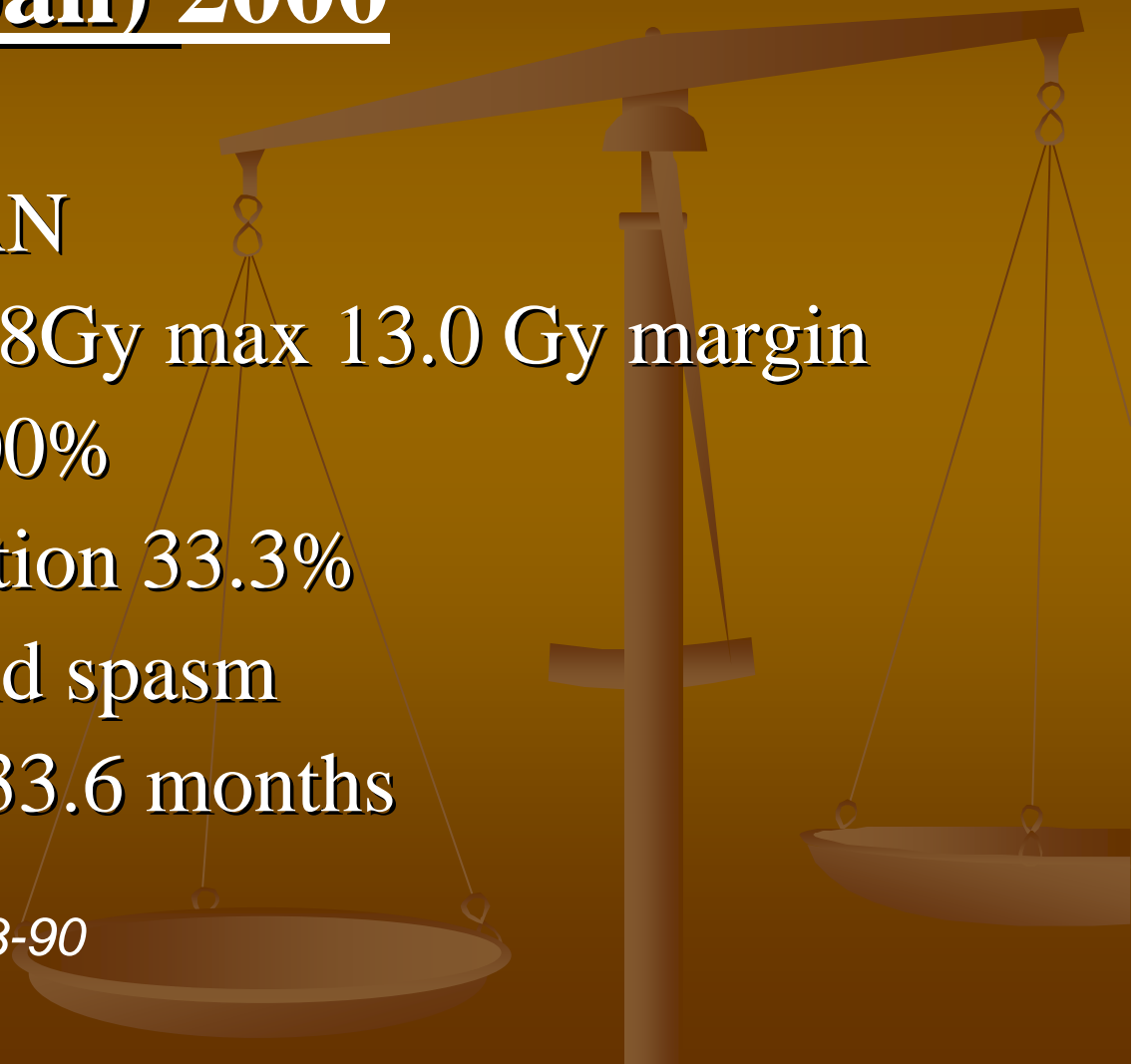
**** → Reserved for microsurgery contraindications**

Radiation therapy

Kida et al (Japan) 2000

- 20 patients
- < 3cm growing AN
- Gamma knife 26.8Gy max 13.0 Gy margin
- Tumor control 100%
- Hearing preservation 33.3%
- 10% VII palsy and spasm
- Mean follow-up 33.6 months

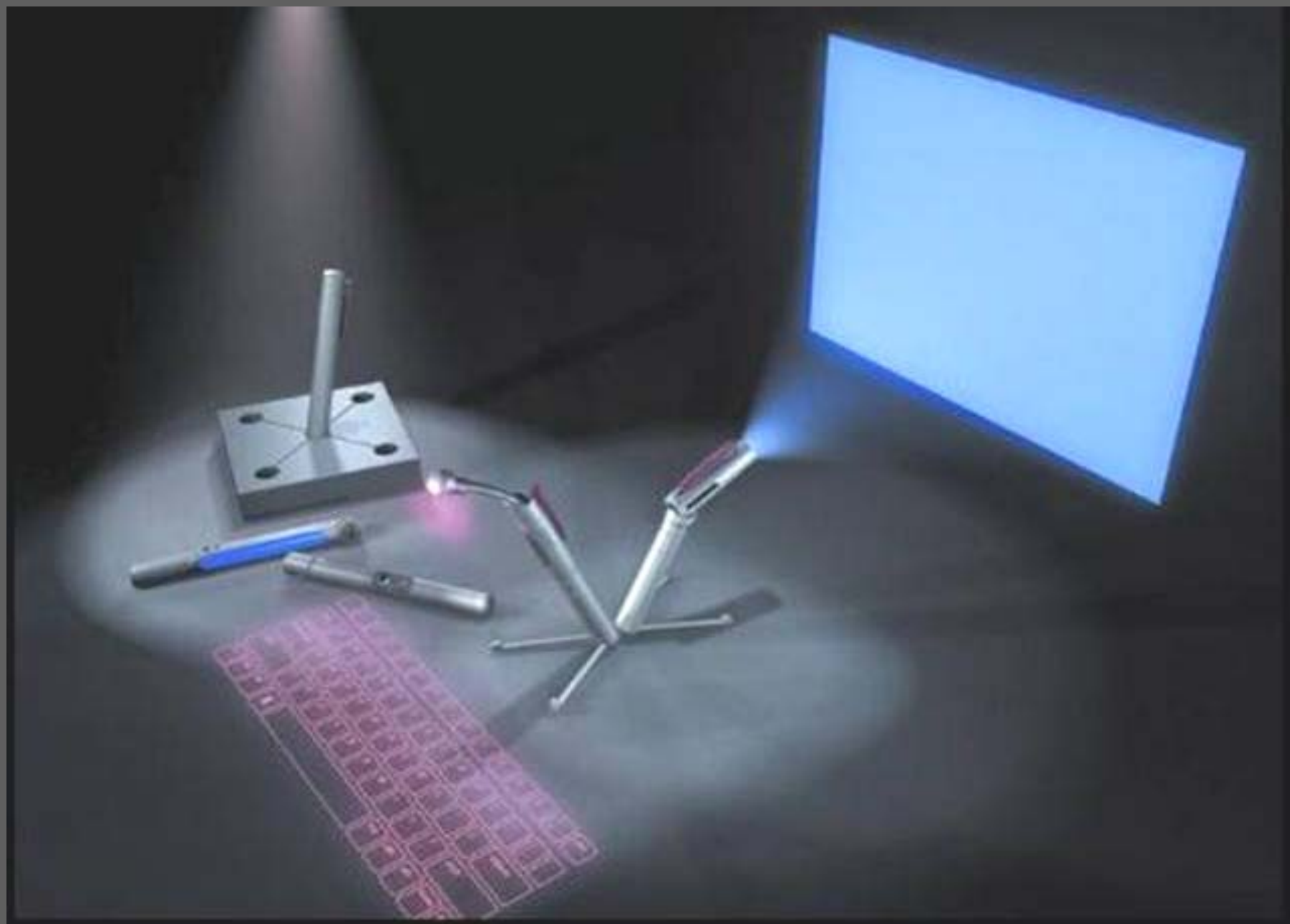
Kida et al. Surg Neurol 2000 383-90



Grand Round

Abdulrahman Hagr
MBBS FRCS(c)





The

End

