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Case Study

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Birth history; Case #1

- 4 year old
- Unremarkable birth history
- Newborn hearing screening:
  - Refer x4 in the right ear
  - Passed each time in the left ear

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BER results; Case #1

- 1st diagnostic BER test results at 2 months

  Click: 50dB

  500 Hz: 50 dB

- Abnormal right ear immittance

- Slit like, possibly collapsing canal noted on right

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ENT and Audio follow up

- First ENT consultation:
  - ❖Right ear serous effusion

- Repeat BER testing indicated:
  - ❖30 dB click
  - ❖30 dB ASSR 500-4000 Hz
- Abnormal right immittance

- Scheduled for tympanostomy tubes but the parents canceled as they did not want for him to go under anesthesia

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ENT findings

- Granulation tissue obstructing the right ear canal (unable to assess middle ear) and prescribed Ciprodex drops
- Continued to have copious inflammatory tissue obstructing the right TM
- Concern for cholesteatoma

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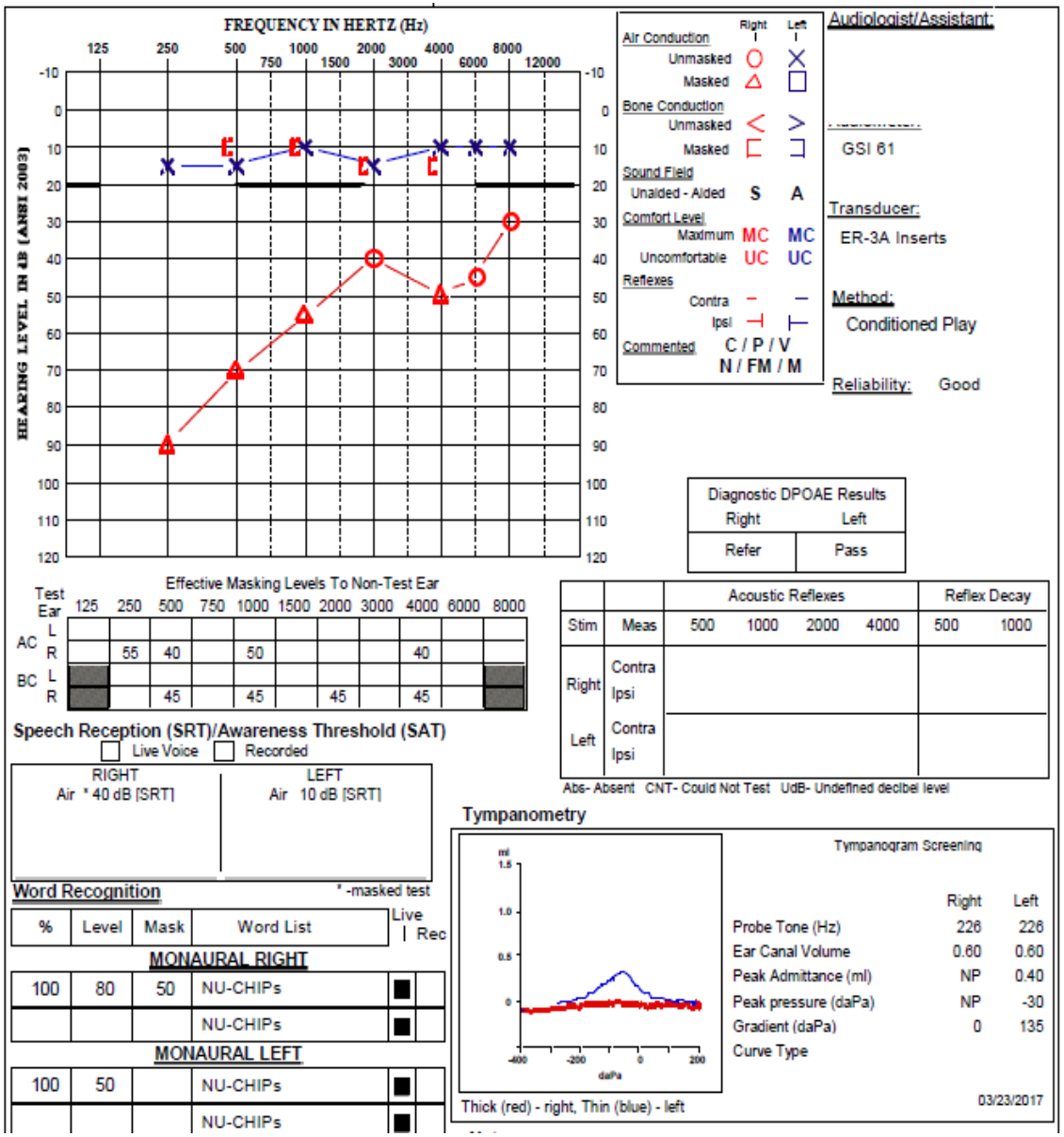
Audiometric findings



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CT Scan temporal bones

- Complete opacification of the right mastoid air cells and middle ear cavity
- Soft tissue opacification of the osseous portion of the right external auditory canal
- Bony destruction involving the mastoid air cells and anterior wall of the right external auditory canal, with likely additional destructive changes about the right incus and malleus
- Differential diagnosis:
  - Congenital cholesteatoma versus Langerhans cell histiocytosis

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Biopsy findings

- **Rhabdomyosarcoma**
  (*embryonal*, stage IV)

- Approximate 4.2 x 2.4 cm homogeneous enhancing erosive soft tissue mass centered within the right temporal bone and extending throughout the right external auditory canal and middle ear
- Associated early erosion through the right tegmen and floor of the right middle cranial fossa, and involvement of the overlying dura

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Rhabdomyosarcoma

Rhabdomyosarcoma (RMS) is a **malignant** tumor that arises from a normal skeletal muscle cell.

Because skeletal muscle cells are found in virtually every site of the body, RMS can develop in almost any part of the body.

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Rhabdomyosarcoma

- Sarcomas are cancers that develop from connective tissue in the body
- Rhabdomyosarcoma is the most common soft tissue sarcoma of childhood and adolescence
- More commonly found in
  - head/neck area (near eyes or nasal sinuses)
  - reproductive organs
  - trunk (arms and legs)
- **Uncommon in the middle ear**

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Medical Treatment

- Antineoplastic chemotherapy (Vincristine)
- Proton beam therapy

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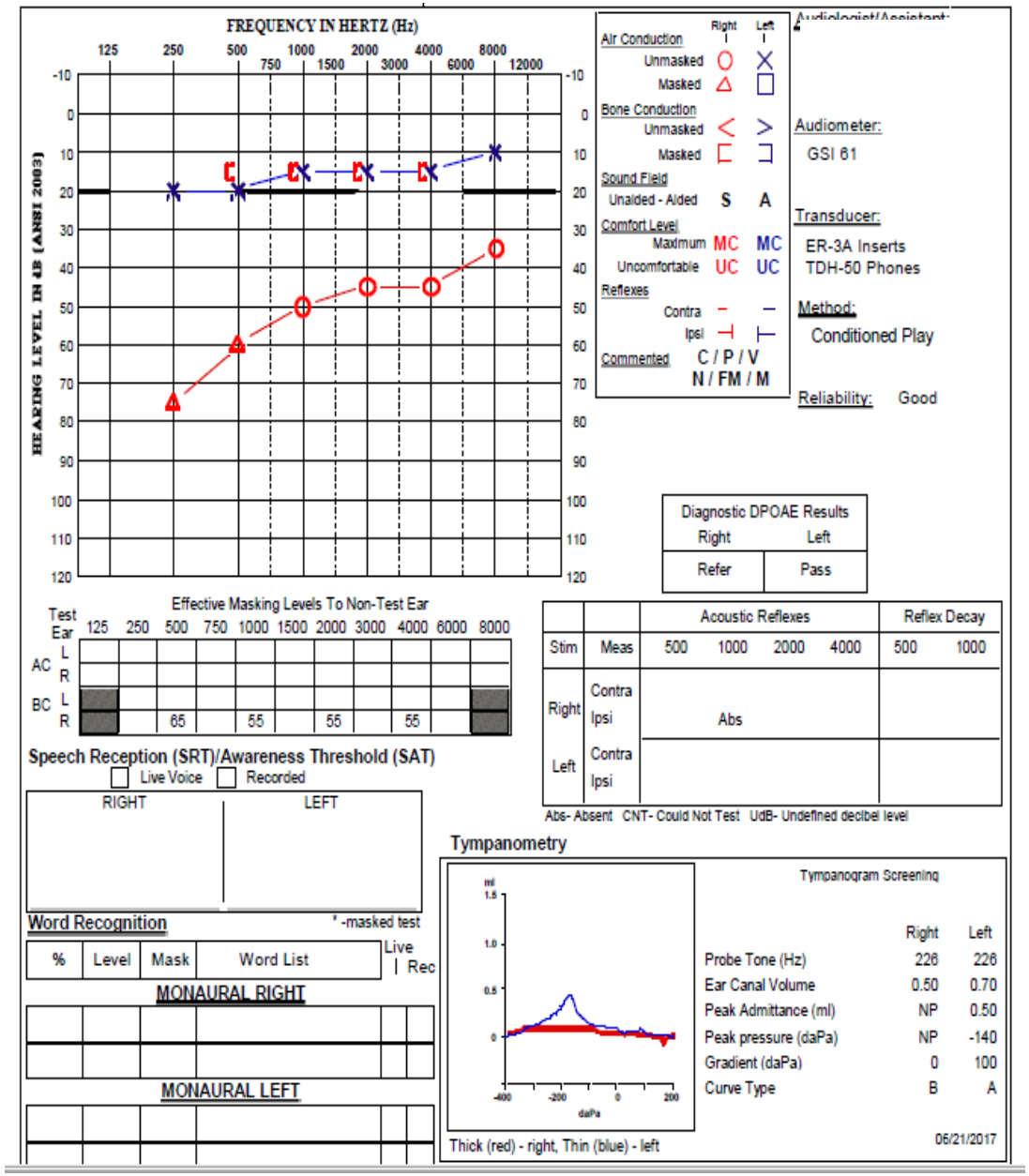
3 months later



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Audiologic Management

- Hearing aid consultation
- Right hearing aid fitting

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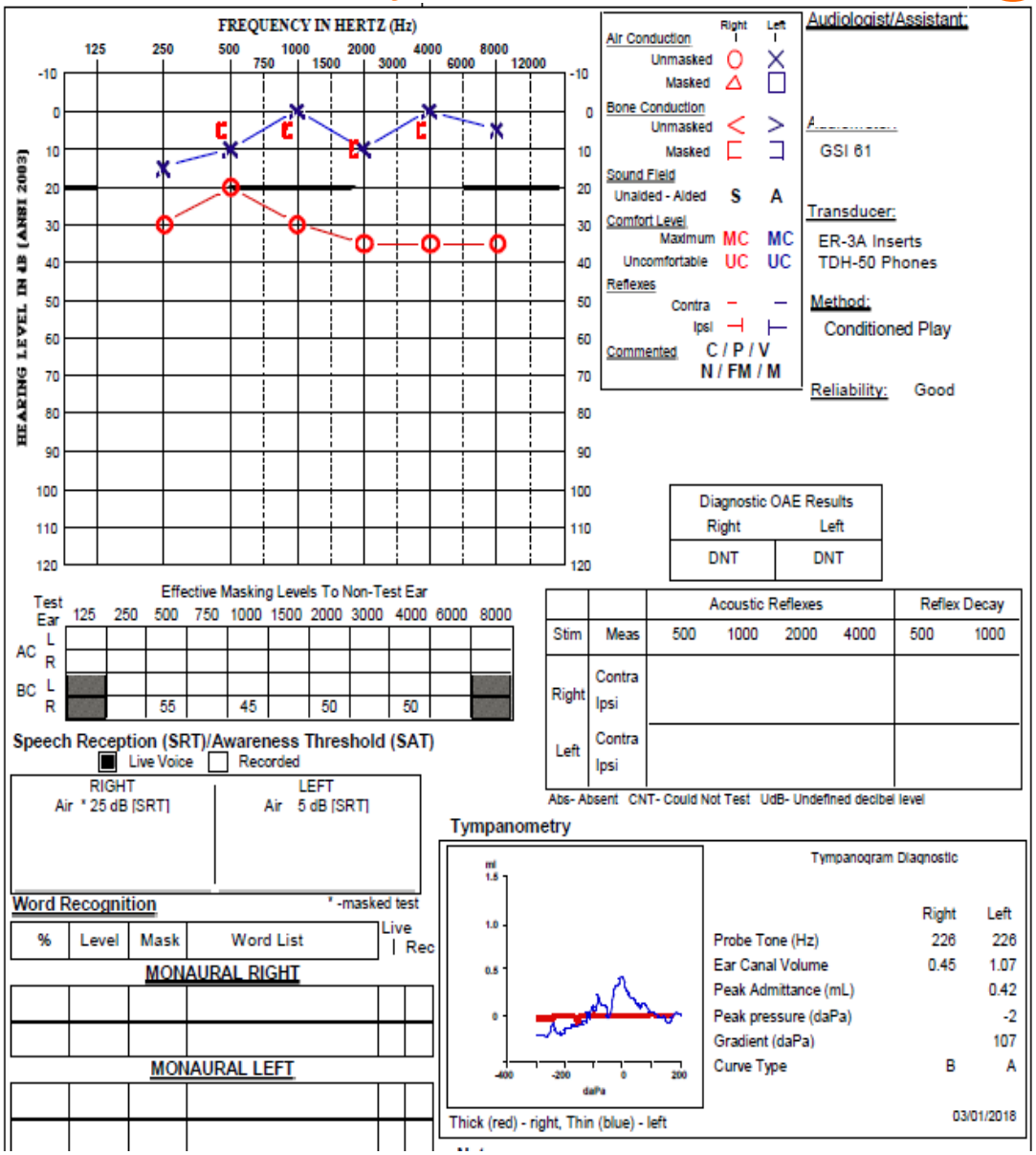
5 months post HA fitting



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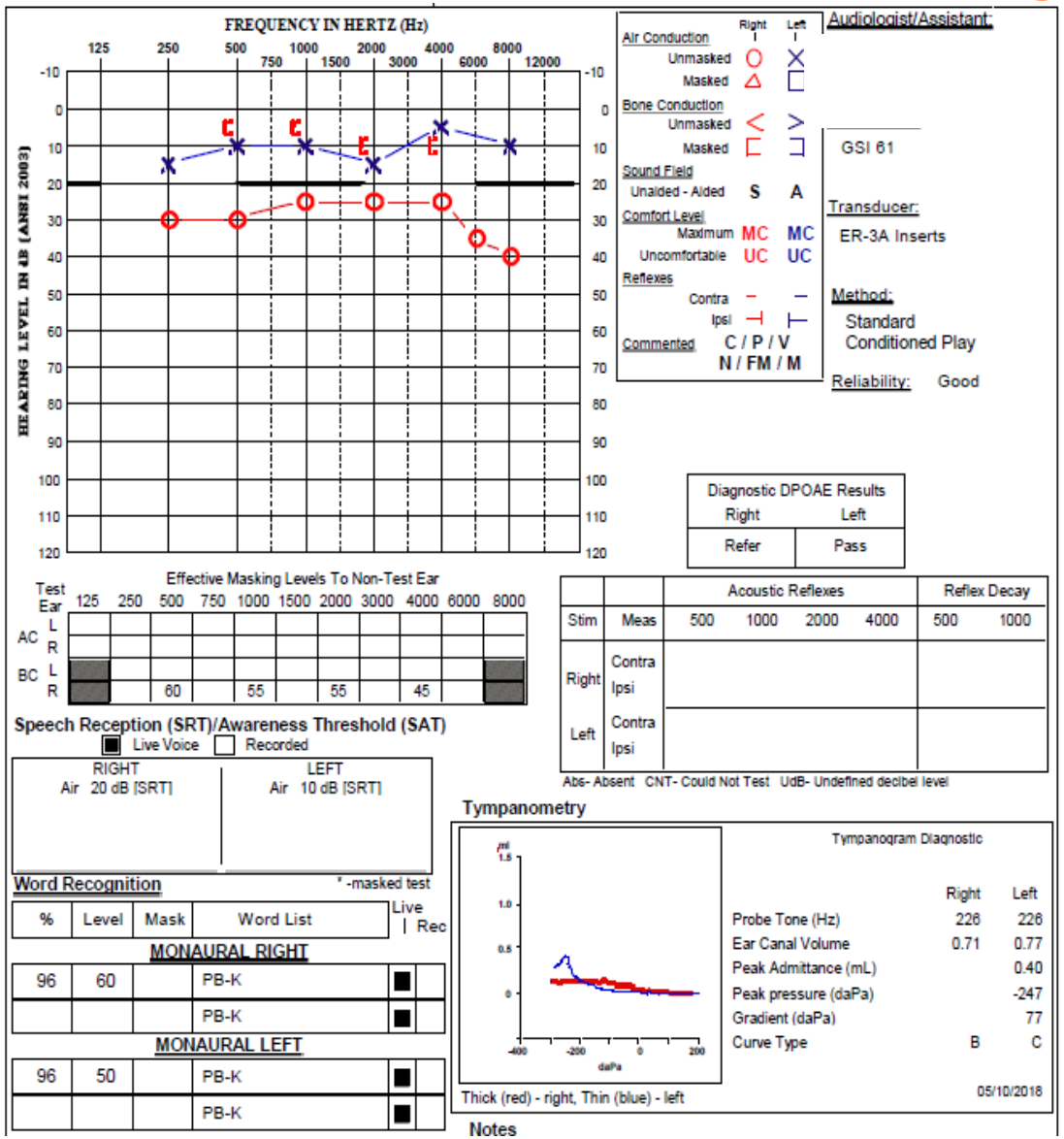
7 months post HA fitting



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Case #2

- 4 year old
- Imaging ordered after about a month of foul discharge from right ear that did not respond to irrigation or antibiotics
- Temporal bone CT and MRI obtained in China

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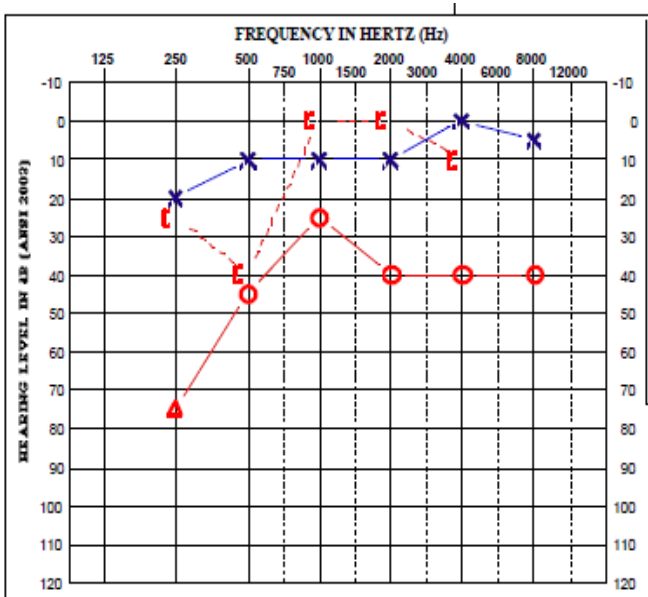


	Right	Left
Air Conduction		
Unmasked	O	X
Masked	△	□
Bone Conduction		
Unmasked	<	>
Masked	[	]
Sound Field		
Unaided - Aided	S	A
Comfort Level		
Maximum	MC	MC
Uncomfortable	UC	UC
Reflexes		
Contra	–	–
Ipsi	⊣	⊢
Commented	C / P / V	
	N / FM / M	

Audiologist/Assistant:

Transducer:
ER-3A Inserts

Method:
Conditioned Play

Reliability: Good

Diagnostic DPOAE Results	
Right	Left
Refer	Pass

Effective Masking Levels To Non-Test Ear

Test Ear		125	250	500	750	1000	1500	2000	3000	4000	6000	8000
AC	L											
	R		40									
BC	L											
	R		55	45		45		45		45		

		Acoustic Reflexes				Reflex Decay	
Stim	Meas	500	1000	2000	4000	500	1000
Right	Contra						
	Ipsi						
Left	Contra						
	Ipsi						

Abs- Absent CNT- Could Not Test UdB- Undefined decibel level

Speech Reception (SRT)/Awareness Threshold (SAT)

☐ Live Voice ☐ Recorded

RIGHT	LEFT
Air 20 dB [SAT]	Air 0 dB [SAT]

Word Recognition * -masked test

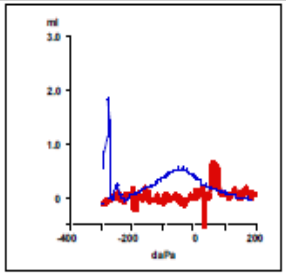
%	Level	Mask	Word List	Live	Rec

MONAURAL RIGHT

MONAURAL LEFT

SOUNDFIELD AIDED RIGHT

Tympanometry



Thick (red) - right, Thin (blue) - left

Tympanogram Diagnostic

	Right	Left
Probe Tone (Hz)	226	226
Ear Canal Volume	1.07	1.02
Peak Admittance (mL)	0.64	1.82
Peak pressure (daPa)	NP	-25
Gradient (daPa)		14
Curve Type	B	A

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Notes

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Case #2

Diagnosis:

Embryonal rhabdomyosarcoma of the right temporal bone/middle ear/mastoid.

Treatment:

- Chemotherapy (Vincristine)
- Proton beam therapy
- Repeat imaging with PET/CT scan after induction chemotherapy that showed no PET avid lesions and near resolution of soft tissue mass

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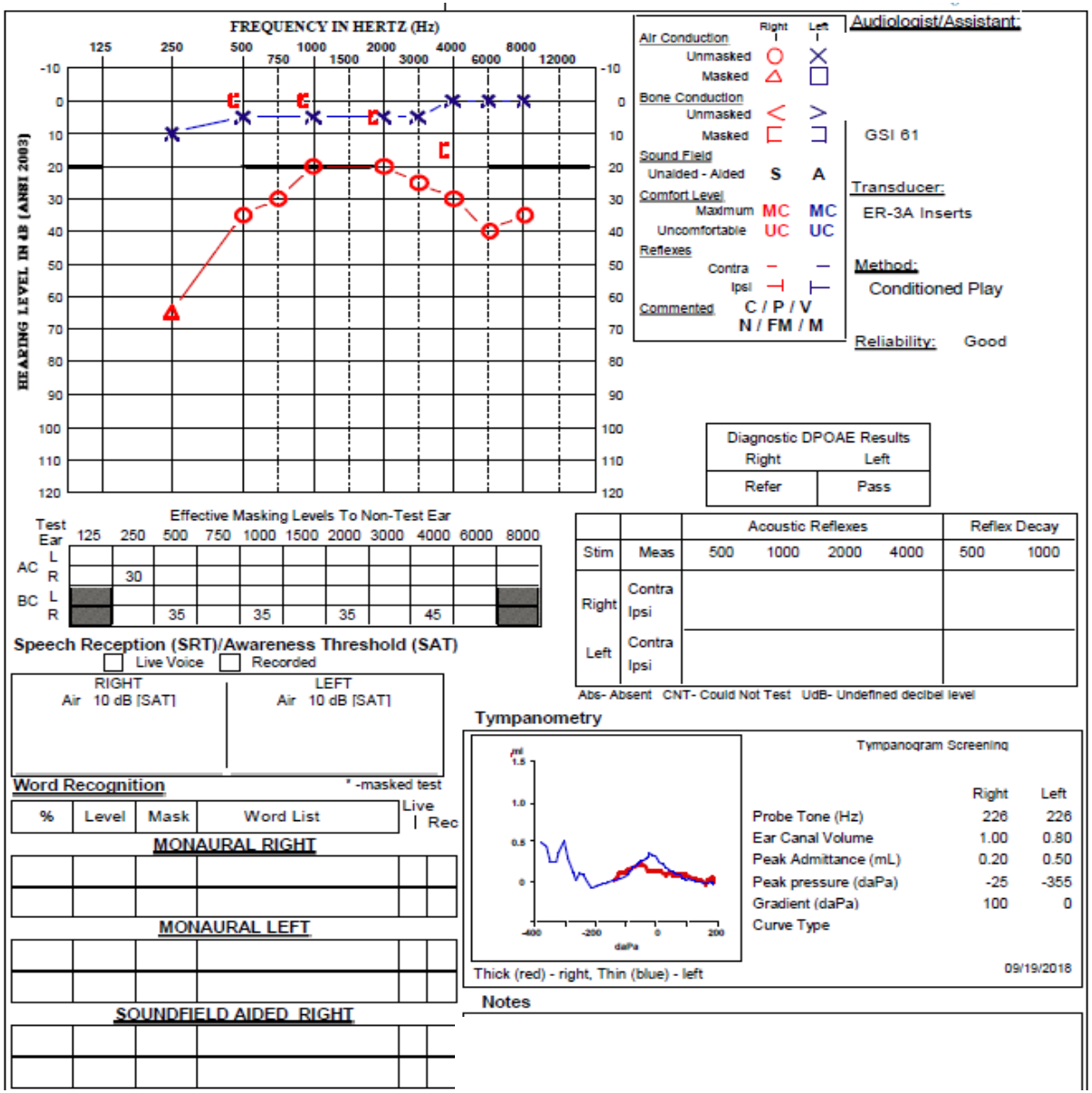
Latest audiogram



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Recommendations

- Hearing aid consultation completed
- Earmold impression, hearing aid fitting and follow up appointments scheduled
- Stringent audiologic monitoring
  - Status of mass after treatment
  - Chemotherapy changes
  - History of radiation

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Rhabdomyosarcoma

- Rhabdomyosarcomas comprise three distinct histologic subtypes: embryonal, alveolar, and pleomorphic
- Embryonal type is the most common one found in the head and neck
- Of all childhood rhabdomyosarcomas, approximately 30%-50% occur in the head and neck
- The most common locations in the head and neck include the nasopharynx and orbits

J Surg Case Rep. 2012 Oct; 2012(10): 9.
Published online 2012 Oct 1. doi: 10.1093/jscr/2012.10.9
PMCID: PMC3649646
PMID: 24960749
Rhabdomyosarcoma in middle ear of an adult: a rare presentation
S Bhargava, M Grover, J Mehta, and V Maheshwari

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Incidence of RMS

- Approximately 4-7 cases per million
- Most RMS are diagnosed in children and teens
- More than half are in children younger than 10
- Peak incidence is the 0-4 year old age range
- About 3% of all childhood cancers are RMS
- Slightly more common in boys than girls
- Only 7% of RMS arise in the middle ear

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Considerations

- Rhabdomyosarcoma is the most common soft tissue sarcoma in the pediatric age group
- Rhabdomyosarcoma of the temporal bone is an aggressive tumor that clinically simulates chronic otitis media
- **A high index of suspicion should be raised in the context of otitis media that is unresponsive to ordinary medical treatment**

Int J Pediatr Otorhinolaryngol. 2007 May;71(5):807-14. Epub 2007 Mar 8.
Temporal bone rhabdomyosarcoma in children.
Sbeity S1, Abella A, Arcand P, Quintal MC, Saliba I.

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The End