



# **Malformation I**

External Ear,  
Cysts and Fistulas of the Neck

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Brockmeier

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# Malformation Outer Ear

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- fistula
- preauricular tag
- dysplasia of the outer ear
- dysplasia/aplasia of the outer ear canal

# External Fistula

## Location

- 1 preauricular
- 2 behind the ear
- 3 proximal group of lateral neck fistulas

## Symptoms:

- secretion
- inflammation

## Diagnostics

- MRI
- Contrast filling

## Therapy:

complete excision





# Fistula in Outer Ear Canal

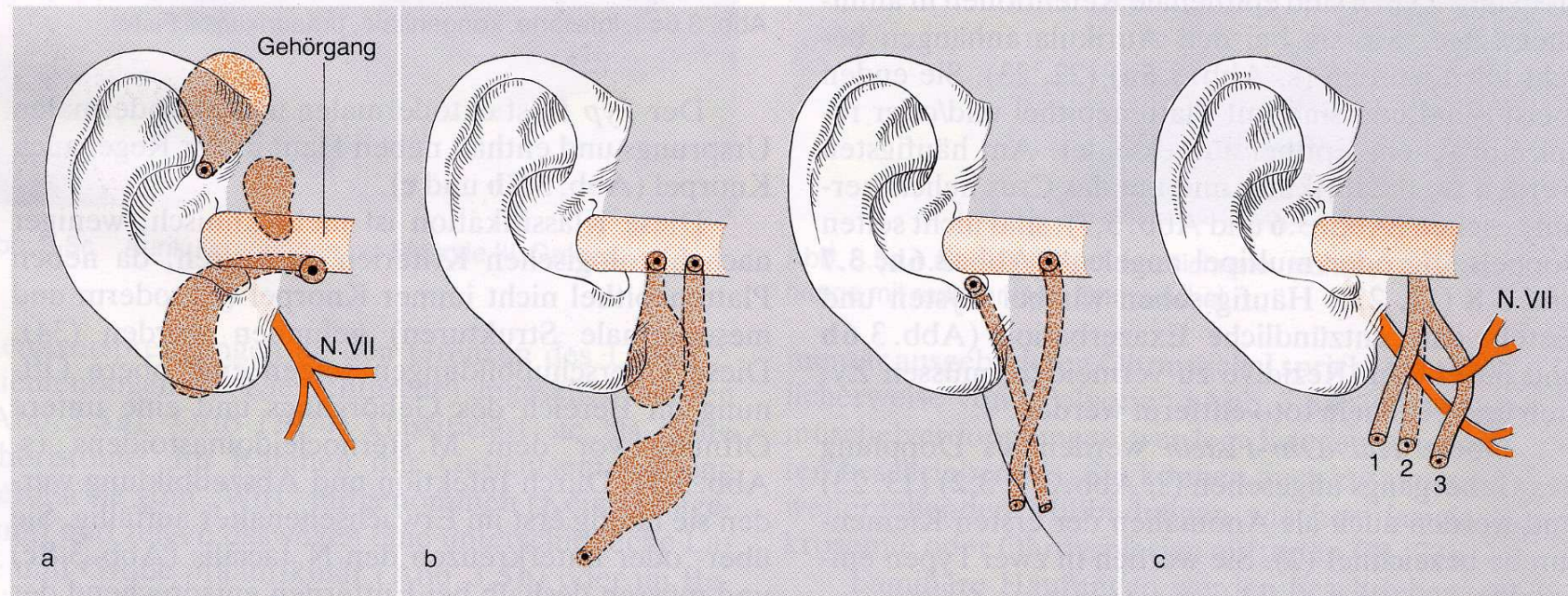


Abb. 3.8a Lage der Typ-I-Fisteln und der Typ-I-Zysten der sog. „Ohr-Hals-Fisteln“ (nach Belenky u. Medina [2]). b Lage der Typ-II-Fisteln, den sog. „gedoppelten Gehörgängen“ (nach Belenky u. Medina [2]). c Lage des N. facialis zu den Typ-II-Hals-Ohr-Fisteln (nach Belenky u. Medina [2]; Otto [23]) 1 überkreuzende, 2 und 3 unterkreuzende Fisteln.

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# Preauricular Tags

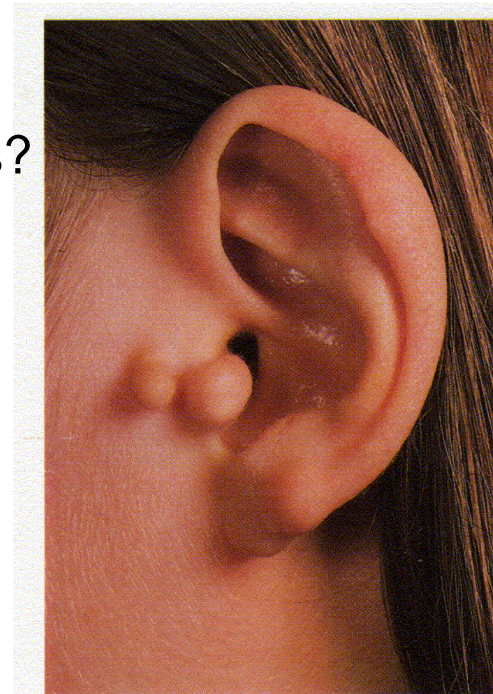
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## **Diagnostics:**

Accompanied by (conductive) hearing loss?

## **Therapy:**

Wait and see  
only remove if material is not needed



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# Dysplasia of the Outer Ear

## Classification

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**Grade I:** malformation of single structure of the outer

eg. prominent ear, macrotia, cup ear, cats ear, lobular anomaly ( eg adherent, split, hypertrophic), ....

**Grade II:** complete microtia , more pronounced malformation, structures may be missing

**Grade III:** many structures missing, often accompanied by malformation of the outer ear canal

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# Malformation of the Outer Ear Therapy

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**Grade I:** eg. correction of ear-head-angle (ideally 30°)  
when: before school

**Grade II:** reconstruction of single structures  
when: variable

**Grade III:** epithesis  
plastic construction of missing structures  
period: variable



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# Atresia of the Outer Ear Canal Therapy

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## **Surgical:**

Typ A: canal plasty with epithelial coverage

Typ B: additional tympanoplasty

Typ C: see B, often recurrent relapses, longstanding

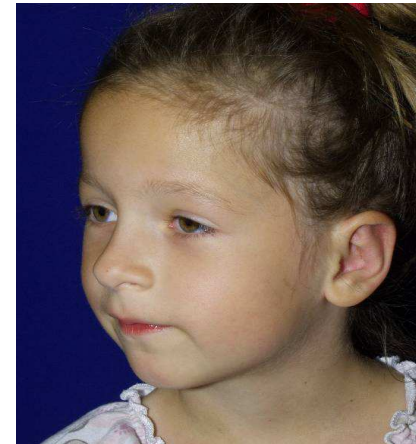
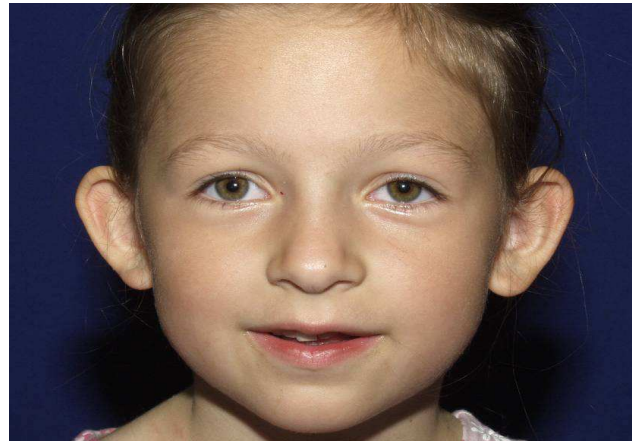
## **Rehabilitation of Hearing Prosthetical:**

Baha, Bonebridge, implantable hearing aid



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# Grade 1



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Courtesy of R. Staudenmaier

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## Grade 2



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Courtesy of R. Staudenmaier



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## Grade 2

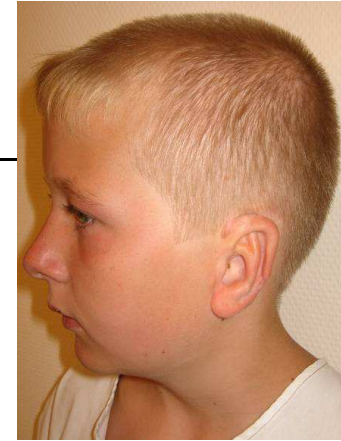
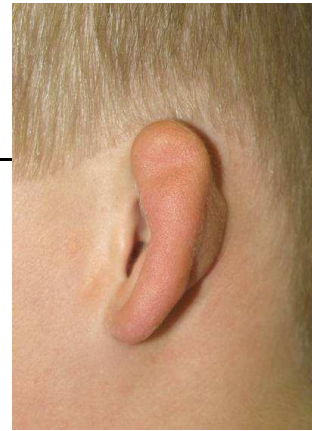


Courtesy of R. Staudenmaier

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## Grad 2- 3

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More pronounced malformation need:

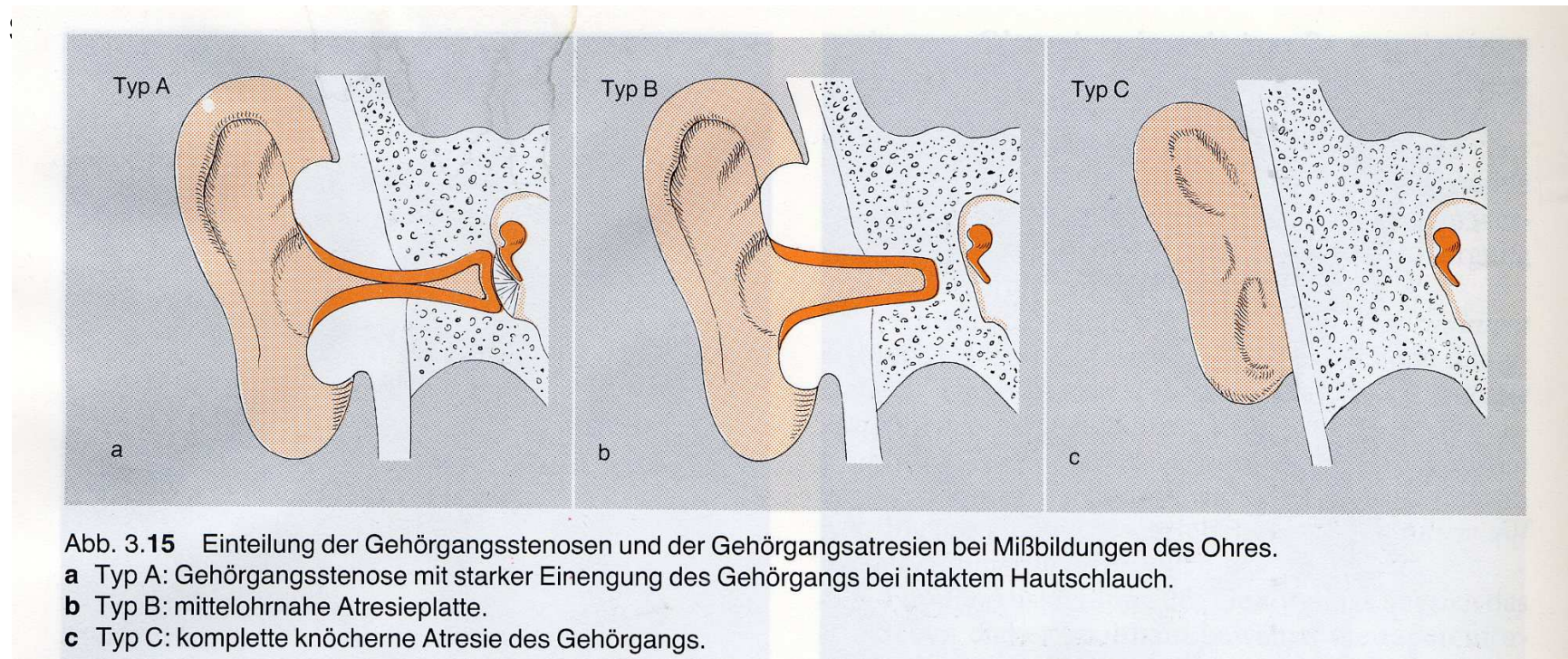
- forming of a three dimensional structure
- adequate skin overlying





# Atresia of the Outer Ear Canal

## Classification





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# Malformation of the Middle Ear Ossicles

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**joints and/or ossicle themselves malformed/missing**

**Stapes:** most common malformation  
alteration of Stapes und adjacent joints

**Malleus:** mostly fixation  
fusion with epitympanic wall

**Incus:** most commonly fused with Malleus

**Therapy:** surgical  
prothetical hearing aid conservative, implantable

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## Malformation of the Outer Ear Canal and Middle Ear Symptoms

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Conductive hearing loss : 20 – 60 dB

Stapedial reflex      can be missing

Tympanometry      variable

normal/type B/

hypermobile when gap in ossicular  
chain

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# Bone Conduction Implantable Hearing Aid Baha/Bonebridge

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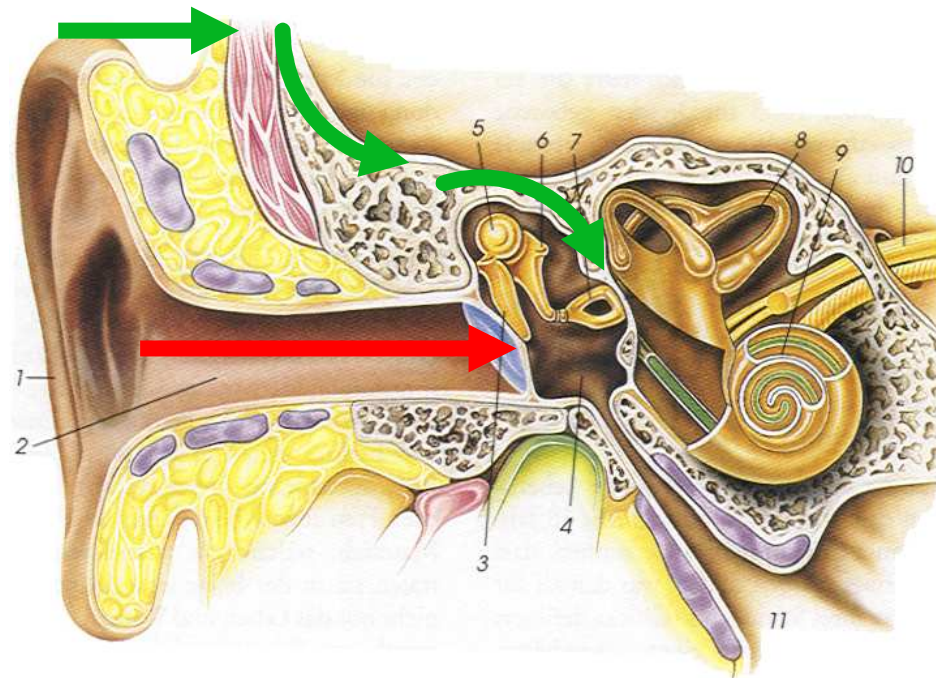
## Indication:

Bilateral aural atresia

Unilateral atresia

Unilateral problem:

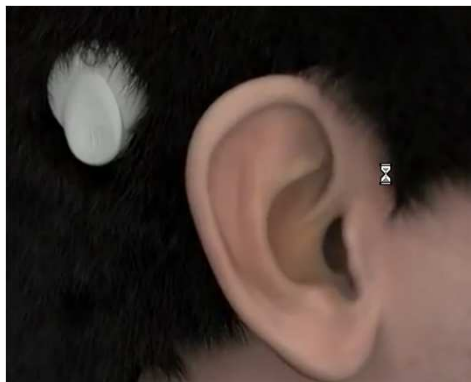
hearing in noise compromised  
spacial hearing reduced



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# Bone Conduction Implantable Hearing Aid Baha/Bonebridge

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Bonebridge



Baha attract



Die Schnappkupplung...



... und das Baha Divino™  
in situ.

Baha divino

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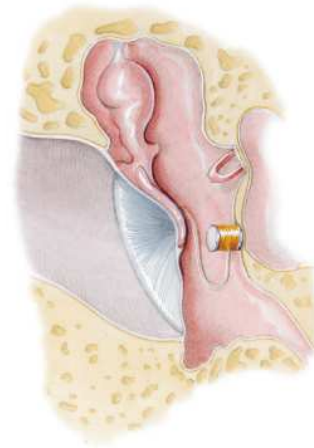
# Vibrant Soundbridge in Middle Ear Malformations

Lit z.B. Wollenberg (2007) HNO 55:349-356

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Fixation at any ossicular structure  
Or  
Round window nice



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Courtesy of Firma Medel



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# Literature Therapy Options

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Hearing outcomes of atresia surgery versus osseointegrated bone conduction device in patients with congenital aural atresia: a systematic review. Nadaraja GS, Gurgel RK, Kim J, Chang KW., Otol Neurotol. 2013 Oct;34(8):1394-9.

Congenital aural atresia . Abdel-Aziz M. J Craniofac Surg. 2013 Jul;24(4):e418-22.

Atresiaplasty versus BAHA for **congenital aural** atresia. Yellon RF.; Laryngoscope. 2011 Jan;121(1):2-3.

Does the evidence support use of the Baha implant system (Baha) in patients with **congenital** unilateral **aural** atresia? Danhauer JL, Johnson CE, Mixon M. J Am Acad Audiol. 2010 Apr;21(4):274-86.

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## Case

bilateral complete atresia of the outer ear canal, Grade 2 - 3 dysplasia of outer ear

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### postpartal

- CT: Inner ear normal  
Bilateral extensive malformation of middle ear, bony atresia
- conductive hearing aid in cap week 2, 7 month bilateral in headband

### 6 months

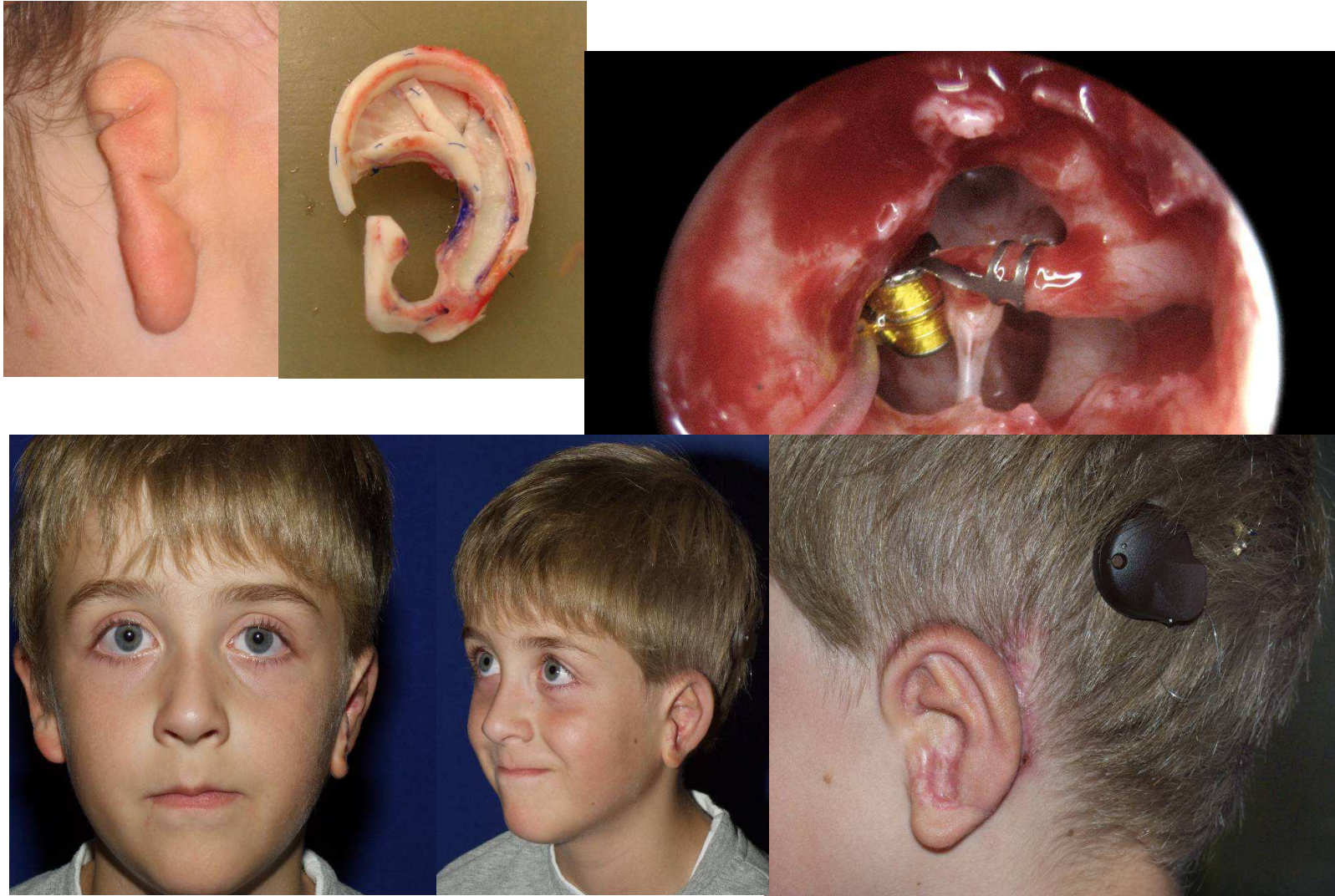
ABR bone/air

### approx age 6

implantable hearing device  
otoplasty



# Case

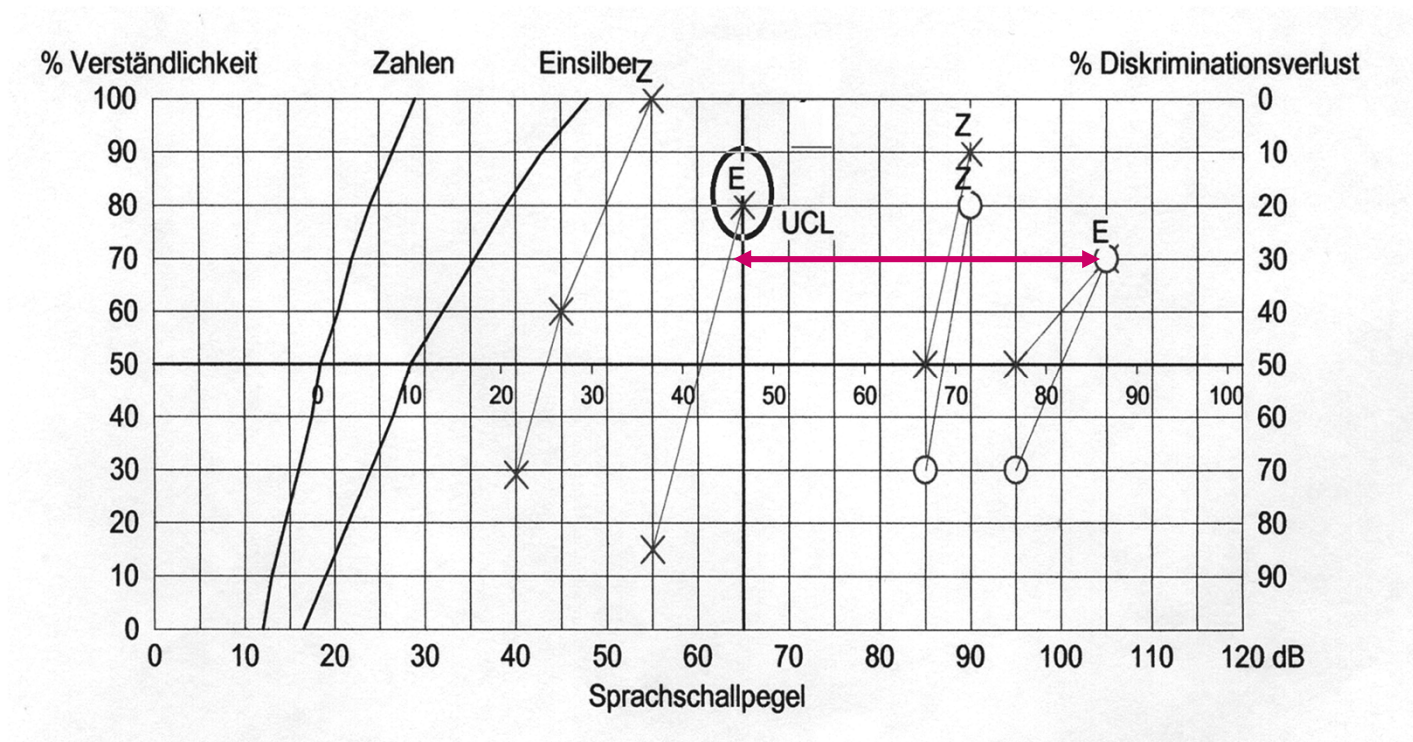


Courtesy of R. Staudenmaier



# Grade 3 with Atresia of Outer Ear Canal

Audiologisches Ergebnis bei bds. Fehlbildung



Courtesy of R. Staudenmaier

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# Unilateral Aplasia of Outer Ear Canal

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## **postpartal**

- OAES contralateral normal
  - => counselling
  - adressing child from good side

## **monitoring speech development**

**Further steps depending on**  
**symptoms**  
**needs of parents/child**

## **discussion**



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# Summary

## Therapy of Malformation of Outer Ear, Middle Ear

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### **Cosmetics:**

- minor malformation good results with local plastic surgery
- major malformation reconstruction with (rip) cartilage

reduction of felt stigmatisation  
increases self esteem

### **Hearing Loss:**

treating functional deficit

- conventional hearing aid (bone/air conduction)
- implantable conductive hearing aid (Baha/Bonebridge)
- implantable hearing aid e.g. Vibrant Soundbridge

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## Other Middle Ear Malformations

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<b>Muscles:</b>	rare aplasia M stapedius , M tensor tympani aplasia stapedial tendon
<b>Vascular anomalies:</b>	persistant A stapedia aberrant A carotis interna high jugular bulb
<b>N. facialis:</b>	<b>20% when ME Malformation</b> most common dehiscence of Fallopiian Canal course very variable
<b>Chorda tympani:</b>	course very variable

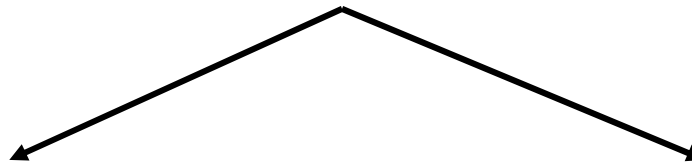
significance mostly in combination with middle ear surgery

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# Fistulas and Cysts of the Neck

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lateral

medial

branchiogenic malformation

- Branchiogenic cysts
- Branchiogenic fistulae

thyreoglossal malformation

- Thyreoglossal duct cysts
- Thyreoglossal duct fistulas
- (Zungengrundstruma)

# Branchiogenic Malformation Cysts/Fistula

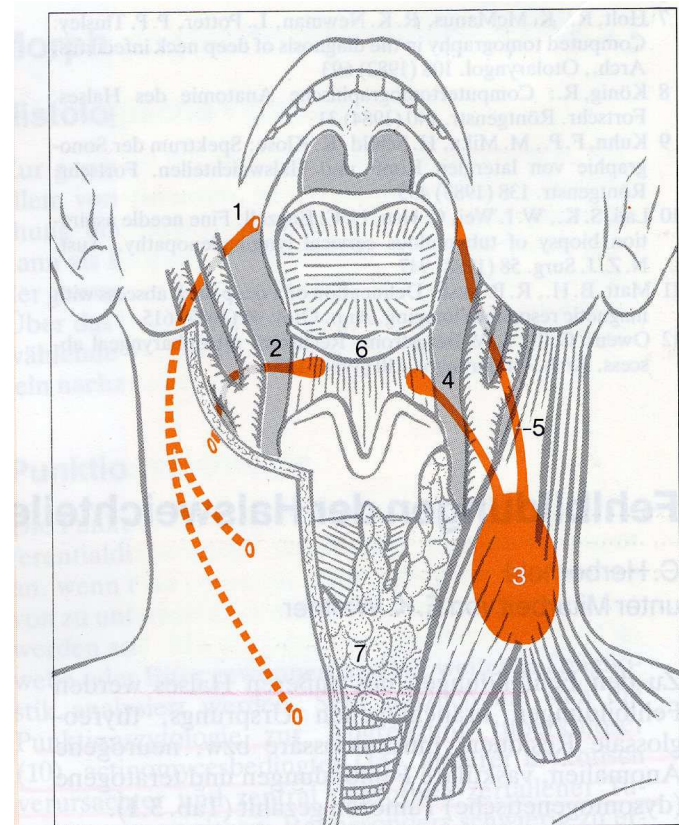


Abb. 3.1 Laterale Halszyste und -fistel (aus Naumann, H. H.: Differentialdiagnostik in der Hals-Nasen-Ohren-Heilkunde. Thieme, Stuttgart 1990).

- 1 Laterale Halsfistel als Relikt des 2. Kiemenbogensystems
- 2 blind endender Gang einer kurzen lateralen Halsfistel als Relikt des 3. Kiemenbogensystems
- 3 laterale Halszyste mit blind endender Gangverbindung in Höhe der Membrana hyothyreoidea (4) bzw. bis zur Tonsillenloge reichendem Gangrudiment (5)
- 6 Zungenbeinkörper
- 7 Schilddrüse

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# Symptoms of Branchiogenic Malformations

( Hosemann et al HNO (1998) 36: 140-146)

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## branchiogenic cysts

Age: toddler – early adulthood

tumor: elastic

size slowly increasing,

lateral triangle or

infront/under M sternocleidomastoideus

swelling	89%
pain	27%
inflammation	7%
dysphagia	5%
restriction of neck movemet	1%

## Branchiogenic fistulas

Age: often babies

opening :

front edge M sternocleidomastoideus

blind end or continuation to  
oropharynx

can run through carotid bifurcation

secretion	89%
inflammation	21%
swelling	16%
pain	5%



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# Diagnostics and Therapy of Branchiogenic Malformations

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## **Branchiogenic cysts**

### **Diagnostics:**

Ultrasound

CT/MRT

### **Therapy:**

exstriation

Including excising duct

## **Branchiogenic fistula**

### **Diagnostics:**

contrast instillation

MRI

### **Therapy:**

Complete exstriation

# Thyreoglossal Residue

## Symptoms and Diagnostics:

Same as branchiogenic

## Therapy:

complete excision

including middle part of hyoid

evt up to root of tongue

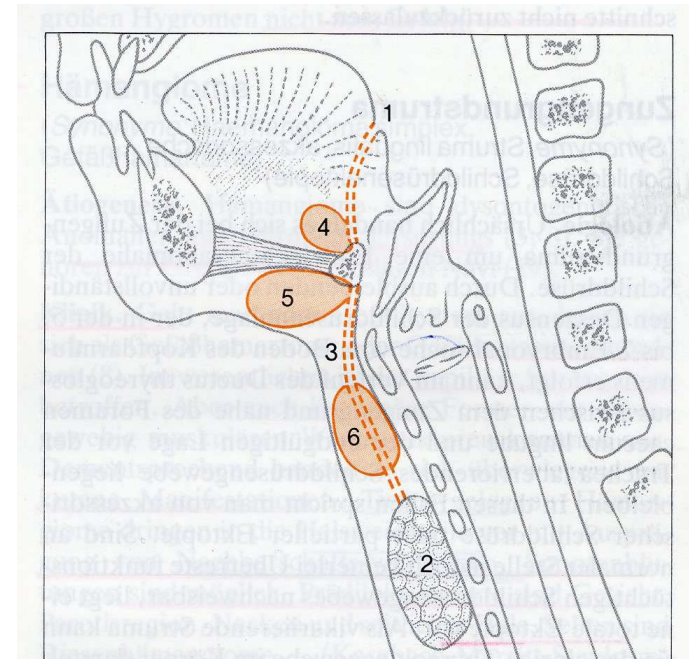


Abb. 3.2 Lagevarianten der medianen Halszyste (aus Naumann, H.H.: Differentialdiagnostik in der Hals-Nasen-Ohren-Heilkunde. Thieme, Stuttgart 1990).

- 1 Foramen caecum am Zungengrund
- 2 Schilddrüse
- 3 embryonaler Ductus thyreoglossus
- 4 suprahyoidale mediane Halszyste
- 5 subhyoidale mediane Halszyste
- 6 kollare mediane Halszyste

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# Thank you for your attention

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