CONGENITAL MALFORMATIONS OF EXTERNAL EAR

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- Average adult female ear is 59 mm tall and the average male is 63mm tall.
- .85 % of height of ear is achieved by 3 years of age and almost 100% by 10 years of age.
- The size of ear remains the same size till the age of 60 when it gradually enlarges particularly the lobe.
- The average adult ear protrudes 19 mm from the mastoid skin

- During the sixty week of intrauterine life the external ear begins to develop around the dorsal end of the first branchial cleft.
- On either side of this cleft lie the first (mandibular) and second (hyoid) arches. •
- The auricle develops from these arches as 6 small buds of mesenchyme known as the six hillocks of His. •
- The first arch gives rise to the hillocks 1 to 3 and the second arch gives rise to hillocks 4 to 6.

- There has been always a controversy regarding formation of which part of ear from which hillock.

- The auricle begins to develop in the anterior neck region, then it is postulated to migrate dorsally and cephalad as the mandible begins to develop during the second and third months of gestation.

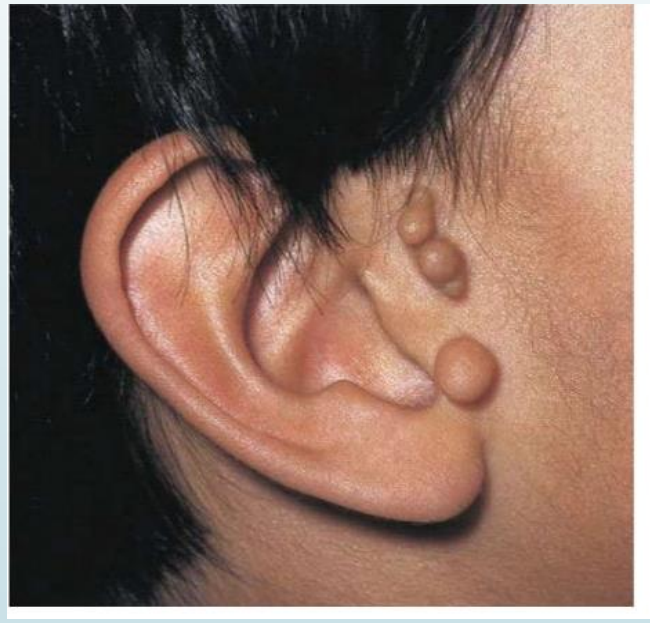
- By the 5th month of gestation the pinna lies in its adult location.

- HILLOCKS PART OF PINNA 1 ANTERIOR PORTION OF EAR LOBE 2 TRAGUS 3 ASCENDING HELIX 4 ANTI-HELIX 5 HELIX 6 POSTERIOR PORTION OF LOBE.

HILLOCKS	PART OF PINNA
1	ANTERIOR PORTION OF EAR LOBE
2	TRAGUS
3	ASCENDING HELIX
4	ANTI-HELIX
5	HELIX
6	POSTERIOR PORTION OF LOBE.

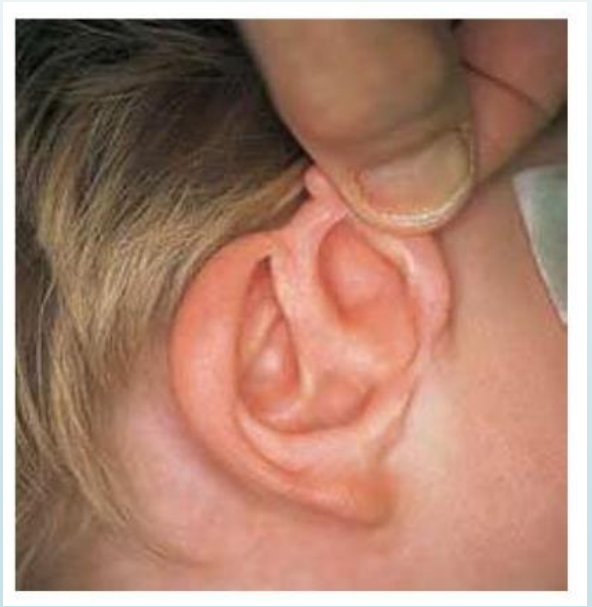
CONGENITAL DEFORMITIES OF EXTERNAL EAR

- PRE-AURICULAR TAGS
- Very common
- Involves only skin but usually contains a long tail of cartilage extending into the cheek.
- Treatment
- Excision



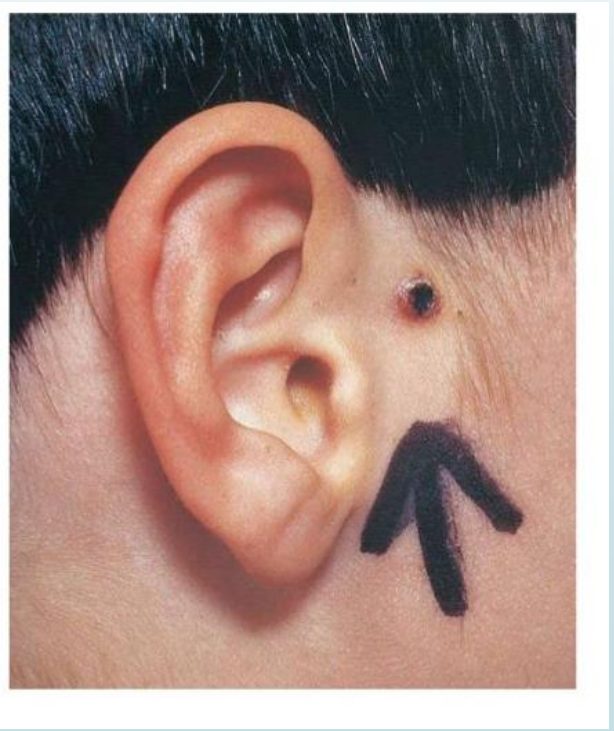
MIRROR EAR OR POLYOTIA

- Persistent pre-auricular tissues lying on posterior cheek resembling extra ear.
- Skin is peeled off the extra-auricular tissue, cartilage remnants trimmed .
- Skin excision to give cheek flatter shap



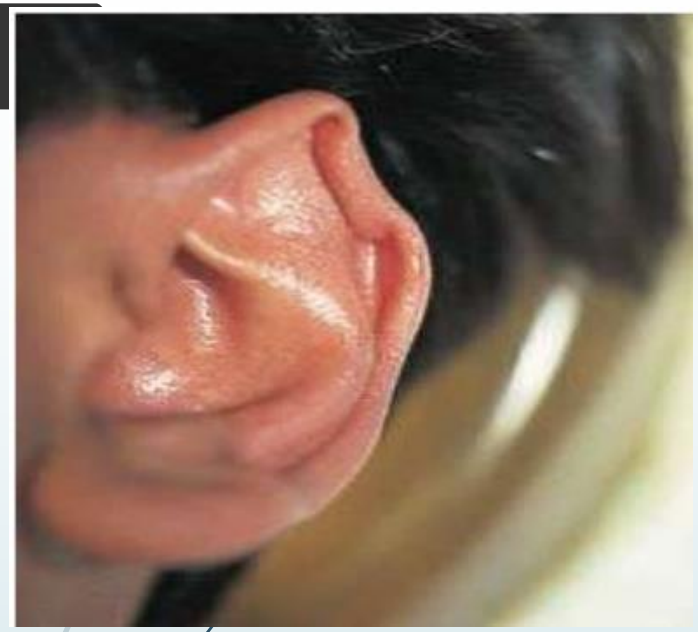
PRE-AURICULAR PITS AND SINUSES

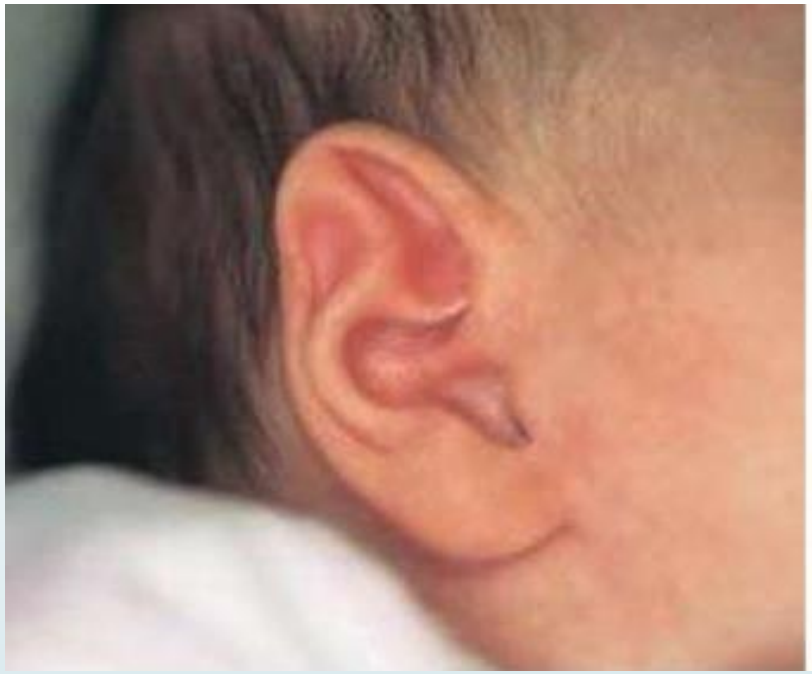
- Often bilateral & frequently asymptomatic.
- Preauricular sinuses are malformations that result from incomplete fusion of 2 of the 6 hillocks that arise from the first and second branchial arches.
- Track deeply to facial nerve.
- Excision with facial nerve monitoring.



ABNORMAL FOLDS OF PINNA

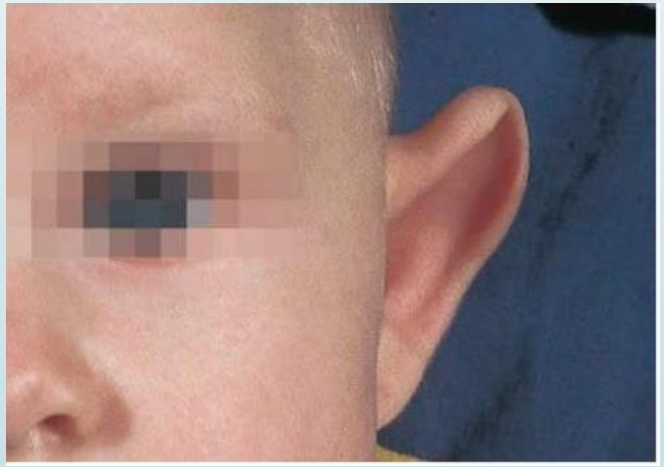
- Common
- Lop ear - Upper pole of the ear flops over •
- 'Mustarde' type suture used to create U-shaped cartilage prop at missing upper anti-helical fold. Ear hitched to the mastoid fascia •
- Other varieties are –
- Kink of helical rim
- Abnormal fusion of helical rim to the anti-helical fold
- Whole ear appears collapsed vertically
- • Surgical correction by scoring, tie-bar type tethering sutures or direct wedge excision





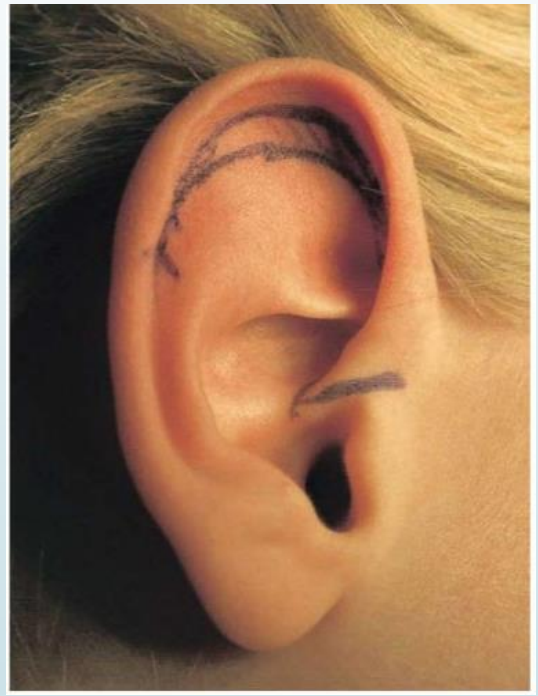
PROMINENT ('BAT') EARS

- • Prominence is due to an absent anti-helical fold but in some cases, the conchal bowl is excessively deep (hilloks 4)
- Digital pressure on relevant part of ear determines cause and gauge strength of cartilage. •
- Recommended to do surgery only after 5 years of age. •



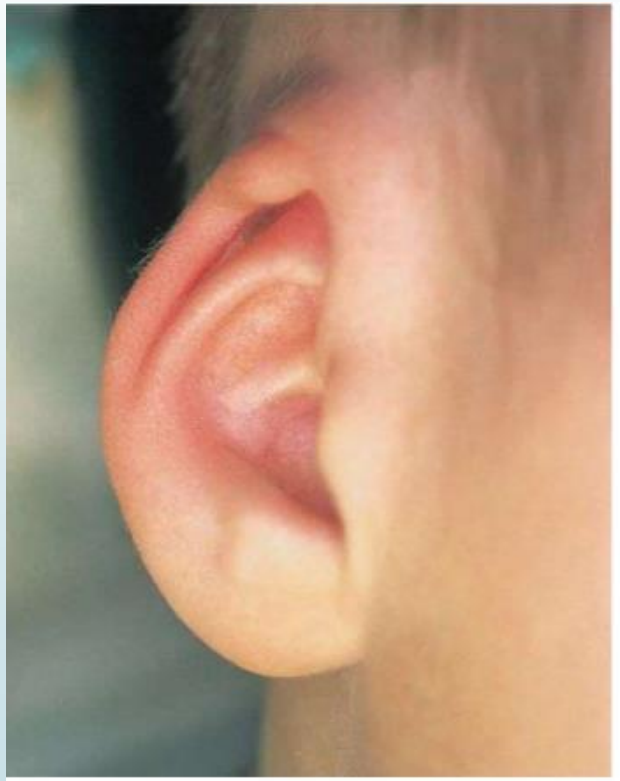
MACROTIA

- Excessively large ears.
- In normal ear, upper pole, conchal hollow and lobe take up an equal amount of the height of ear, splitting in to thirds.
- Big scaphal hollow or big lobe are the reasons of macrotia.
- Anterior crescent of skin and cartilage be removed from scaphal hollow.Wedge of tissue is removed from oversized lobe



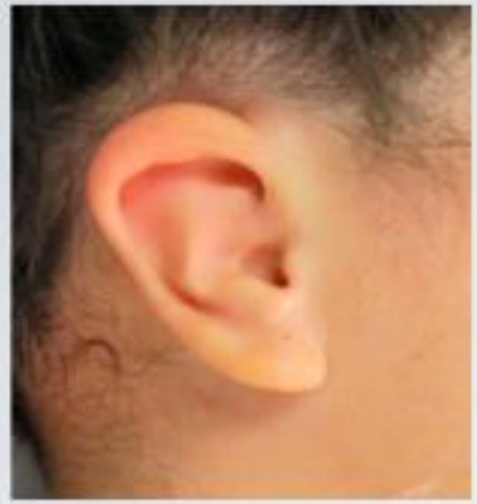
CRYPTOTIA (‘THE HIDDEN EAR’)

- Lower two thirds of ear visible, upper auricular sulcus seems lost.
- When ear is pulled away from the side of the head, the upper pole cartilage becomes evident, having hidden beneath scalp skin.
- Upper pole is tethered and lower lobe is prominent.
- Small ear buddies splint applied at birth for non-surgical correction.
- Later surgical correction by releasing the tethered portion of ear.



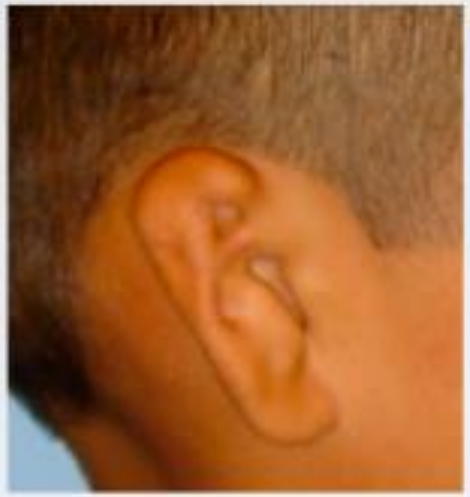
MICROTIA & CONGENITAL AURAL ATRESIA

- DEFINITION
- Microtia: Defined as the abnormal development of the Pinna resulting in a malformed auricle.
- Congenital aural atresia: Defined as a failure of development of external auditory canal.
- Congenital aural atresia is always associated with a certain degree of microtia



Grade 1

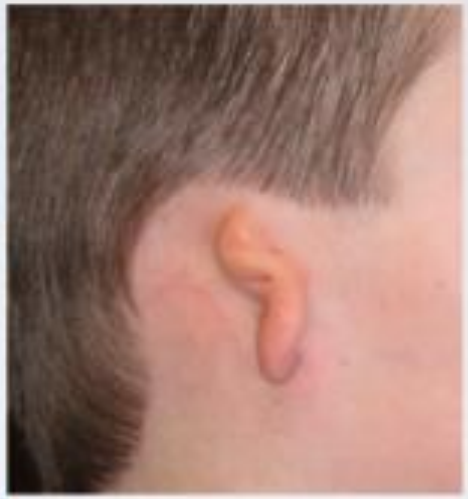
Smaller than normal, but the ear has mostly normal anatomy



Grade 2

Part of the ear looks normal, usually the lower half

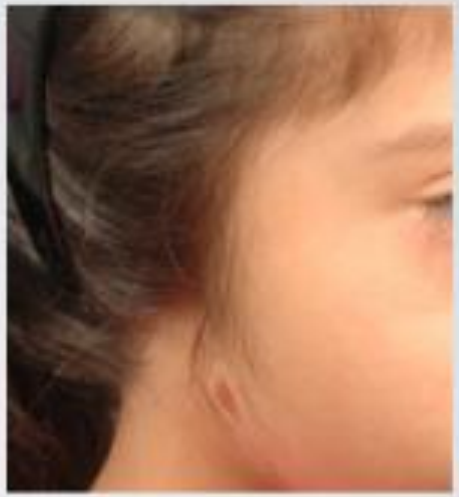
The canal may be normal, small or completely closed



Grade 3

Just a small remnant of "peanut-shaped" skin and cartilage

There is no canal, which is called aural atresia



Grade 4

Complete absence of both the external ear and the ear canal, also called "anotia"

EPIDEMIOLOGY

- • Microtia and congential auditory atresia occur in approximately 1 in every 6000 live births.
- These deformities commonly occur unilaterally, more so on the right side.
- Men are affected as common as women. The degree of auricular deformity usually correlates with the degree of middle ear deformity. The incidence of inner ear deformities are very rare in patients with congenital auditory atresia.
- Microtia is associated with other anomalies of face 50% of the time. Women with four or more pregnancies are at increased risk of bearing a child with microtia.
- The incidence of microtia is higher in Japanese population.

ETIOLOGY

- Exposure to teratogens like vitamin A, thalidomide, isotretinoin
- Vascular insults and genetic aberrations.
- Isolated microtia can occur with branchial arch anomalies.
- •Syndromic associations of with congenital aural atresia include

- MICROTIA

- Malformation, such as anotia and microtia, are likely to be caused by the disturbance of development at seven to eight weeks gestational age, whereas deformations (lop, cup and prominent ears) are caused by a problem later in the development or by external compression.

MANAGEMENT

- Investigations :

- Age appropriate Hearing assessment
- HRCT Temporal bones
- X-ray Cervical spine
- USG for congenital renal malformations

- COMPLICATIONS OF MICROTIA SURGERY •
- MAJOR – hematoma formation, skin flap necrosis, infection, and pneumothorax. • .
- MINOR – malpositioning, scar contracture or hypertrophy, and poor contour

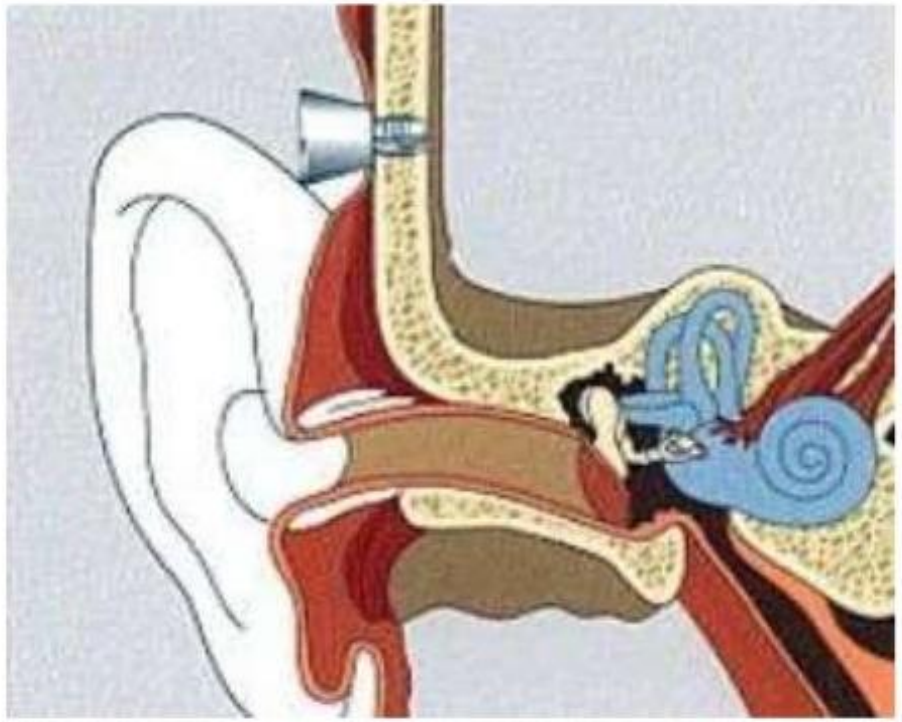
ALTERNATIVES FOR CONGENITAL AURAL ATRESIA:

IMPLANTABLE HEARING AIDS IN CHILDREN • Several types of implantable hearing aids may be used for hearing rehabilitation of CAA in children over 5 years of age, as an alternative to atresiaplasty: –

percutaneous (crosses skin) bone-conduction devices (BAHA, Ponto),

– transcutaneous (closed-skin) bone-conduction devices (Sophono),

– active middle ear implants (Vibrant Soundbridge).



The image of the BAHA shows how a button (or stud if you prefer) is inserted into the bone of the skull above the ear.

The BAHA device is attached to the button within the skull. It is unobtrusive (once the hair is put back into place). (image is from Island Hearing.)

The PONTO device is attached to an abutment (i.e. stud) behind the ear (image is from Ponto web site.)

The SOPHONO device is attached using a magnet (image is from Sophono web site.)

END