

Methods to Evaluate Congenital Ear Deformity

Jacob Antony Chakiath¹, Ravi Kumar Chittoria²

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ABSTRACT

External ear defects are common and occur in roughly 5% of the total population. The common congenital ear deformities were anotia, microtia, cryptotia, hypoplasia, constricted ear/cup ear/lop ear, prominent ear, stahl ear deformity. This manuscript shows methods to evaluate a case of congenital ear deformity.

Keywords: Congenital ear deformity; Microtia.

INTRODUCTION

The ear divides into three areas: the external, middle, and inner. Based on location, different malformations can present. A malformation is not only a change in appearance but also an alteration in function. External ear defects are common and occur in roughly 5% of the total population.¹ The most common malformations consist of combined external and middle ear deficits, called congenital aural atresia.² Microtia is a term used to describe the underdevelopment of the pinna, whereas anotia is a term used for an absence of pinna.

Author Affiliation: ¹Senior Resident, ²Professor & Registrar (Academic) Head of IT Wing and Telemedicine, Department of Plastic Surgery & Telemedicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry 605006, India.

Corresponding Author: Ravi Kumar Chittoria, Professor & Registrar (Academic) Head of IT Wing and Telemedicine, Department of Plastic Surgery & Telemedicine, Jawaharlal Institute of Postgraduate Medical Education and Research, Pondicherry 605006, India.

E-mail: drchittoria@yahoo.com

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MATERIALS AND METHODS

This template of evaluation was made after going through standard text books in plastic surgery and extensive Google search. The common congenital ear deformities were anotia, microtia, cryptotia, hypoplasia, constricted ear/cup ear/lop ear, prominent ear, stahlear deformity. The demonstrated template is a case of microtia. It's an example of template to show how to evaluate a case of congenital ear deformity.





Introduction

Before starting history or examination first introduce the patient under following headings:

Name:

Age:

Gender:

Occupation:

Address:

Education status:

Known Comorbidities:

CHIEF COMPLAINTS

Deformity of ear Since Birth

History of Presenting Illness

Inform whether history was taken from patient, parents or relatives.

Before taking details related to chief complaints ask antenatal, perinatal, post-natal, immunization & development history of the patient.

In the antenatal history ask about following details

- What is the birth order?
- Any infertility treatment taken?
- What was the age of mother and father at the time of conception?
- Is there any history of consanguinity?
- Is there any history of infection, fever, rashes, radiation exposure drug intake (phenytoin, steroids, retinoids, native drug or other teratogenic drugs), and comorbidities of the mother (anaemia, folic acid deficiency, diabetes, hypertension, hypothyroidism, seizure disorders) during pregnancy?

- Was mother hospitalised for any illness during pregnancy?

In the perinatal history ask about following details

- Was the child born full term/pre-term?
- Was it a home or hospital delivery?
- Whether the delivery was normal vaginal delivery or by caesarean section?
- If born at home, who assisted in delivery?
- Was there any use of forceps/vacuum equipment?
- Did the mother hear the child cry immediately after birth or not?
- History of any cyanosis/breathing difficulties?

In the post-natal history ask following details

- Any history of pathological jaundice?
- Any ICU admission (reason & duration for admission)?

Any history of any emergency/elective surgeries done (for Pierre Robin Syndrome, ocular deformities, craniofacial microsomia, Treacher Collins syndrome, Goldenhar syndrome, Tracheoesophageal fistula, Imperforate anus, congenital heart anomalies, renal anomalies etc.).

Any feeding difficulties postnatal (suckling difficulties).

IMMUNIZATION HISTORY

Whether baby has received immunisation according to the National Immunization Schedule.

Developmental history

Was there any delay in attaining developmental milestones for that age including IQ and performance in school?

History of presenting illness

In general, irrespective of type of congenital ear deformity, ask:

- Whether it is unilateral or bilateral
- Which part of the ear is affected
- Any change in position of the ear
- Any change in shape of the ear
- Any change in size of the ear
- Any change in direction of the ear
- Any presence of accessory tissue (ear tags or

extra ear)

- Any presence of pits
- Whether external auditory canal absent or present

Ask history related to difficulty in swallowing, hearing, speech, closing of eyes, drooling of saliva, loss of taste, difficulty in frowning of eyebrows, deviation in mouth, difficulty in blowing.

Ask history of any other congenital deformities present in the body.

Ask any history of psychological, social, school/studies performance disturbance.

Ask history related to syndromes associated with congenital ear deformity (including Treacher Collins Syndrome, Goldenhar Syndrome, Craniofacial Microsomia, Hemifacial microsomia, etc).

TREATMENT HISTORY

Ask history of treatment (medical & surgical) received for the ear deformity. In case patient underwent some reconstructive surgeries then ask how many stages/numbers of surgeries have been performed. Ask what all donor sites have been used for cartilage, fascia & other tissues in previous reconstructive surgeries. Ask if any other surgeries have been performed for other problems like hearing, facial palsy related functional problems or any other congenital anomalies.

Past History

Ask any past history of comorbidities like diabetes mellitus, asthma, tuberculosis, hypertension, malignancies, blood disorders, drug allergy and hospitalization, surgeries, etc.

Family History

Ask history of any similar ear deformity or any other congenital anomalies in the family.

Personal History

Ask about patient's study performance, sleep, appetite, diet, bowel & bladder habits and any addictions, socio-economic status.

GENERAL EXAMINATION

Mention about patient's level of consciousness, coherence, orientation, built, nourishment, position & co-operation.

Mention about vitals of the patient including temperature, pulse, blood pressure & respiratory

rate.

Head To Toe Examination

Mention in head to toe examination findings including pallor, icterus, lymphadenopathy, cyanosis, edema, clubbing, hydration, presence of any congenital anomalies (musculoskeletal or visceral) or associated syndromes including Treacher Collins syndrome (down slanting palpebral fissures, Malar hypoplasia, hypoplasia of zygomatic complex, conductive hearing loss, mandibular hypoplasia, micrognathia), Craniofacial microsomia (facial clefts, macrostomia, retrognathism, craniosynostosis, external auditory canal atresia, preauricular tags), Goldenhar syndrome, etc.

Systemic Examination

In cardiovascular system mention about presence or absence of external chest wall deformities, normal heart sounds (S1, S2), added sounds (murmurs) and findings related to congenital heart anomalies (TOF, ASD, VSD, PDA etc).

In respiratory system mention about presence or absence of external chest wall deformities, chest wall expansion, chest wall circumference, previous surgery scars, normal vesicular breath sounds, any added sounds (wheeze/crepitations) or absence of breath sounds.

In the gastrointestinal system, mention about any palpable mass & bowel sounds.

In the central nervous system mention about any neurological deficits or palsies including facial nerve palsy and intelligence quotient (IQ).

In the Musculoskeletal system mention about any No spinal anomalies/any weakness of muscles

Regional Examination (Face) is done with adequate lighting, with patient in the sitting, lying down and standing position:

Regional Examination

In the Examination of face mention shape, if face is in the midline.

On inspection of the face in frontal profile:

- Scalp appears: Normal, hairs appear normal in colour, texture and distribution, hair line appears receded.
- In upper third of the face (hair line to nasion): Frontal prominence is there, eye brows appear normal, temporal region appears normal.
- Middle third (nasion to subnasale): Nose

appears normal, upper and lower lids appear normal, inter canthal distance appears normal, eyeball appears normal, zygomatic region appear normal.

- Lower third (subnasale to menton): Upper and lower lips normal, chin central position, cheeks appear normal, nasolabial folds: normal.
- Functional examination of face: Normal frowning, eye closure, cheeks puffing, smile normal.

On inspection of the face in right lateral profile

Scalp: appears normal

Temporo parietal region: appears normal

Forehead: appears prominent

Eyebrow: Normal

Zygomatic region: Appears normal

Cheek region: Appears normal

Ear: ear is absent on the right side with only vertical ridge with depression anteriorly, prominence of lobule, concha seen, auriculo-scalp angle not well defined, external auditory meatus not visible.

Eyeball: Appears normal

Eyelids: along with lashes: Normal

Nasolabial angle: Appears normal

Lips: Normal

Chin: Normal projection

Cervico-mental angle: Normal

On inspection of the face in left lateral profile

Scalp: Appears normal

Temporo-parietal region: Appears normal

Forehead: Appears prominent

Eyebrow: Normal

Zygomatic region: Appears normal

Cheek region: Appears normal

Left ear: Appears normal in size and projection

Eyeball: Appears normal shape

Eyelids: Along with lashes: Normal

Nasolabial angle: Appears normal

Lips: Normal

Chin: Normal projection

Cervico-mental angle: Normal

On Inspection of the Face-In Posterior Profile

Bilateral ear projection and cephalo-conchal angle: Normal:

Occipital region appears normal

Vertex/Parietal region: Normal

Hairline and hair distribution: Normal

Neck symmetry: Normal

No ulceration, no swelling

Inspection in Bird's Eye View

Scalp and hair distribution: Normal

Anterior hairline appears low set

Forehead appears prominent

Eyebrows appear normal

Eyeballs appear normal

The nose appears in the midline, Nasal tip (position, angle, projection) and the lateral segment appear normal

Cheek regions appears normal

Zygoma and temporal region appear normal

Inspection in Worm's Eye View

Mandibular region: Mandible appears hypoplastic, chin and right mandible more hypoplastic

Lips appear normal

Cheek region appears normal

Nose: Tip, nostril floor, Membranous septum and lateral segment appear normal

Zygomatic region appears normal

Ears ear is absent on the right side with only vertical ridge with depression anteriorly, no reminisce of lobule, concha seen, auriculo scalp angle not well defined, external auditory meatus not visible

Left ear appears normal in size and projection

Orbital region appears normal

Frontal region appears normal with receded hair line

Palpation

- Scalp region: Findings of inspection confirmed
 - Head circumference
 - Hair quality: Normal, hair plucking test-negative

- Skin: no scar, lesion
- temporalis present, contraction present
- STA pulsation present
- Sensation normal
- Fontanelle fused
- Suture lines: Normal
- No bony irregularity/deformity/swelling
- No tenderness or any other abnormality
- **Frontal region:** Findings of inspection confirmed
 - Skin: Normal
 - No bony deformity/swelling palpable
 - Frontalis muscle present, contraction present
 - Frontal sinus: Normal (torch test)
 - Sensation normal
- **Eyebrow region:** Findings of inspection confirmed
 - Hair: Normal in texture and distribution, Plucking test: Normal
 - Supraorbital margin: Normal
 - Sensations: Normal
 - No tenderness
 - No bony deformity/swelling palpable
- **Orbital region:** Findings of inspection confirmed
 - **Upper eyelid:** All Soft tissue structures from the skin to conjunctiva are normal including lid margin and eyelashes. Upturning test: Normal, Punctum is visible and normal. Upper eyelid movements are normal. Sensations normal. Position of upper eyelid normal in front gaze.
 - **Lower eyelid:** All Soft tissue structures from skin to conjunctiva are normal including lid margin, eyelashes and punctum. Pull down test: Normal. Sensations normal.

Eyeball/Globe Examination

Pupil: Normal size, shape, symmetry, direct and indirect light reflex: present

Iris: Normal size and shape, colour brownish with no hyper or hypopigmentation, no nodules

Sclera: Colour normal, surface: smooth, no abnormal pigmentation or nodularity

Sclera: corneal junction normal

cornea: sensation present, cornea clear

Palpebral fissures: size in cm, shape in cm, medial and lateral canthal: normal, equal bilateral, punctum: normal

fornices: both superior and inferior fornices normal, no foreign body

Orbital Rims: No tenderness, no step deformity, no mass palpable in the orbital margins.

Extra: ocular muscle function: Normal

NOSE

External Examination

- Finding of inspection confirmed, nose in midline
- Skin: Appears normal in all region including colour, texture, mobility with no swelling, tenderness and hypo or hyperpigmentation
- Bone (including nasal, vomer, frontal and maxillary spine): are normal with no elevation/depression, no tenderness or crepitus, no step deformity
- Cartilage (Upper and lower lateral): Normal
- Cottle test: Negative
- Nasal valve angle normal 10-15 degree
- Nasolabial angle
- Naso-columellar angle
- Nasal lobule to nostril ratio= Normal (1:2)
- Tip projection = Alar base width

Internal Examination

- Normal nostrils, normal hair
- Membranous and cartilagenous septum normal
- On examination with Thudicum speculum-Septum/turbinate: Normal

Zygomatic Region

Inspection findings confirmed

No bony abnormalities/step deformity/Mass

No evidence of buccinator hypertrophy

No maxillary sinus tenderness

Lip and Chin

Function of orbicularis oris

Inspection findings confirmed

Sensations

- Vision
 - Visual acuity
 - Schirmer test
- Hearing
 - Rinne's test
 - Webber's test
 - External auditory canal Examination
- Lymphnode: preauricular/postauricular/
neck nodes

Intraoral Examination

Cleft in the palate is extending from ___ to ___

Width of the cleft, the gap in the palate is approximately ___ cms

Palatal shelves are horizontal and of adequate/
inadequate width

Palatal length is adequate / inadequate

Shape of the cleft

Position of the lower ridge of the vomer

Inclination of the palatal shelves

Length/symmetry/mobility of the soft palate

Degree of motion of the lateral pharyngeal walls

Distance between the posterior edge of the soft
palate and posterior edge of the pharyngeal wall

Presence of passavant's pad

Amount of adenoid tissue (if visible)

Size and state of the tonsils

**Ear examination is done in zones (upper,
middle, lower third)**

Upper third, mention about helix, antehelix, crus
of helix

In middle third mention about tragus, antitragus,
scaphoid fossa

Left ear: Normal. No anomalies detected

Right ear: Helix, antihelix, concha, scaphoid,
triangular fossa, tragus, antitragus absent.

**Ear Remnant With Hypo Plastic Lobule Positioned
at the Same Level as the Opposite Normal Ear.**

External auditory meatus not visualized

Hairline normal

Preauricular side burn +

No sinus, pits, scars noted in the preauricular/
postauricular region.

Hearing Test

Rinne's test

Weber's test

DIAGNOSIS

Feature	Anotia	Microtia	Constricted ear	Prominent ear	Cryptotia
Helix	Absent	Absent	Hooded	normal	Normal
Ante helix	Absent	Absent	Flattened	Norma	Normal
Concha	Absent	Remnant found in concha type	Normal	Normal	Normal
Tragus	Absent	Remanent found in concha type	Normal	Normal	Normal
Projection	Absent	Absent	Normal	Prominent	Absent
Auriculo cephalic angle	Absent	Absent	Normal	>150'	Absent

Congenital right sided lobule type microtia, with
no other associated congenital anomalies, with
conductive hearing loss

Investigations

To add on to my diagnosis

Hearing assessment

HRCT temporal bone

USG abdomen

Cardiac echo

Chest X ray

For anaesthetic fitness

CBC, BUSE, LFT, viral markers, blood grouping
typing

DISCUSSION**Cryptotia**

Cryptotia is a congenital ear deformity in which upper pole of ear cartilage is buried underneath the scalp. The superior auriculocephalic sulcus is absent but can be demonstrated when you pull up the helical pole. Various surgical corrections are reported from Japan, due to the high prevalence of cryptotia, as frequently as 1:400. Non-surgical ear moulding treatment may be applied if the child is in early neonate stage. The goal of surgical treatment is to create the retro auricular sulcus by skin grafts, Z-plasty, V-Y advancement, or rotation flap.³ Common cartilage deformity associated with cryptotia is helix scapha adhesion, which may be addressed by cartilage remodelling techniques.

Stahl Ear

Stahl ear, a rare congenital auricular deformity, is characterized by the third crus extending toward the helical rim. Stahl ear is classified into three types:

Type 1: Obtuse-angled bifurcation of antihelix; looks as though superior crus is missing

Type 2: Trifurcation of antihelix

Type 3: Broad superior crus and broad third crus (protruded scaphoid fossa)

Ear moulding may work well if ear moulding is started in early infancy. Surgical treatment is broadly categorized into two types: cartilage/skin excision and cartilage alteration. Type 1 Stahl ear needs special a mention, to reconstruct missing superior crus, by using excised third crus or rib cartilage graft or creating superior crus by sutures or cartilage cutting.

Constricted Ear

Constricted ear is a concept proposed by Tanzer in 1975.⁴ In constricted ear, helix and scaphafossa are hooded, and crura of antihelix is flattened in various degrees. One gains an impression that the rim of helix has been tightened. Constricted ear is often referred as cup or lop ear. Tanzer classified constricted ear into three groups based on the severity of defect/deformities.

Tanzer Classification of Constricted Ear

Group Description

I Involvement of helix only

II Involvement of helix and scapha

II-A No supplemental skin needed at margin of auricle

II-B Supplemental skin needed at margin of auricle

III Extreme cupping deformity; often associated with incomplete migration, forward title.

Stenosis of External Auditory Canal, and Deafness.

Tanzer group 1: Mild deformities of helix, often called lop ear. Defect involves helical cartilage with minimum skin defect. Musgrave technique is a useful method to expand the helix. Through either anterior or posterior skin incision, multiple cuts were made to the curled cartilage, fan upward and backward, fixed to the curved strut made of concha cartilage graft. The skin is then re-draped across the reconstructed framework. For milder constricted ear (group 1,2 A), focusing surgical correction to construct helical curve is the reasonable option while keeping the original elastic cartilage framework, avoiding hard rib cartilage framework. When superior crus is deficient, partial helix plus superior crus frame from rib cartilage⁵ can normalize the deformity.

Tanzer group 2B: Has both skin and cartilage defect in the upper onethird of the auricle. The loss of folding may involve anti-helical crura, and hooding is more pronounced. The height of the ear is sharply reduced. Park⁶ proposed versatile solution for group 2B constricted ear. For helical skin defect, Park modified the Grotting flap (postauricular flap), creating both skin flap and fascia flap with the same pedicle. For helical cartilage defect, eight rib cartilage is harvested, the helix is fabricated, and the entire length of helix is constructed.

Tanzer group 3: Most severe cupping and failure of migration. Brent recommends to treat severe constricted ear as if it is a form of microtia, when the construction is severe enough to produce a height difference of 1.5 cm. Nagata recommends treating severe constricted ear as a concha type microtia, to replace the defective framework with a full rib cartilage framework.⁷

Ear Molding in Miscellaneous Ear Anomalies

Matsuo, who first reported ear molding treatment for congenital ear deformities, states that when the ear deformities are not hypoplastic, non-surgical correction is easy and reliable.⁸ Stahl ear responds well to the nonsurgical correction only during the neonatal period, whereas protruding ears and cryptotia respond until approximately 6 months of age (Matsuo). It is widely believed that the early initiation of molding is more effective because maternal estrogen in the neonate keeps ear cartilage soft and elastic.

Most agree that if ear molding is started after 3 months of age, the response tends to be poor.

Helix-antihelix adhesion responds poorly to the ear molding treatment and may not be the indication of the ear molding. Skin irritation is probably the most frequent complication, possibly due to tape or adhesive.

MICROTIA

Epidemiology and Pathophysiology

Microtia (small ear) is a congenital condition with unknown cause. Prevalence of microtia varies significantly among ethnic groups (0.83–17.4 per 10,000 births) and is higher in Asian countries for unknown reason. 80% to 90% of microtia is unilateral, and 10% to 20% is bilateral. There are more than 18 different microtia associated syndromes with single gene.

Chromosomal aberrations; however, there is no causal genetic mutation confirmed to date. A relatively common syndrome associated with microtia is hemifacial microsomia and Treacher-Collins syndrome. Isolated microtia rarely run in families. Treacher-Collins syndrome, inherited in an autosomal dominant fashion, often presents with bilateral microtia.

CLASSIFICATION

Many attempts have been made to classify microtia based on embryonic development and severity of the deformities. Nagata's classification is based on surgical correction of the deformity⁹⁻¹¹

Anotia: Absence of auricular tissue

Lobule type: Vestige ear with lobule, without concha, acoustic meatus, and tragus

Concha type: Vestige ear with lobule, concha, acoustic meatus, and tragus

Small concha type: Vestige lobule with small indentation of concha (need lobule type construction)

Atypical microtia: Cases do not fall into previous categories

Patient Assessment and Workup

About 20% to 60% of children with microtia have associated anomalies or an identifiable syndrome; therefore, individuals with microtia should be examined for other dysmorphic features. Microtia is a common feature of craniofacial microsomia, mandibular dysostoses (eg, Treacher-Collins and Nager syndromes), and Townes-Brocks syndrome, and these conditions

should be considered among the differential diagnosis when evaluating an individual with microtia. If there is family history of syndrome, genetic counseling may be necessary.

Physical Examination

Identify the type of microtia, size, dimension, and type of normal auricle, if unilateral. Normal side may not be normal and may have subtle ear deformity such as helix-antihelix adhesion. Evaluate the symmetry of face, facial animation (partial facial paralysis is frequent finding), and dental occlusion. Hemifacial microsomia is often associated with difficult airway for intubation.

Diagnostic Studies

Audiologic testing to determine conductive versus sensorineural defect.

Temporal Bone Imaging

High resolution CT scan for evaluating middle ear ossicles to assess the possibilities of future otologic surgery.

MRI to determine the course of facial nerve, often displaced, especially in the absence of pneumatized mastoid.

Rule out the presence of cholesteatoma (squamous epithelium trapped in the middle ear), present in 4% to 7% of atresia.

Atresia and Middle Ear Anomalies

In bilateral microtia, early and conscientious use of bone conductive hearing aid is imperative for hearing and speech development. Most of hearing deficits in children with bilateral microtia are managed with hearing aids.

Treatments of microtia ideally involve reconstruction of the external ear and the restoration of normal hearing.¹² Hearing impairment in microtia is related to abnormal auditory canal, tympanic membrane and middle ear. The problem is conduction. Typically, microtia patients have a hearing threshold of 40 to 60 dB on the affected side. By comparison, normal function allows us to hear sounds between 0 and 20 dB. Regarding middle ear surgery for hearing restoration, most surgeons presently feel that potential gains from middle ear surgery in unilateral microtia are outweighed by the potential risks and complications for the surgery, and this surgery should be reserved for bilateral cases. Careful selection of the atresia surgery candidate is important to achieve optimal outcome and more importantly to avoid unnecessary surgery

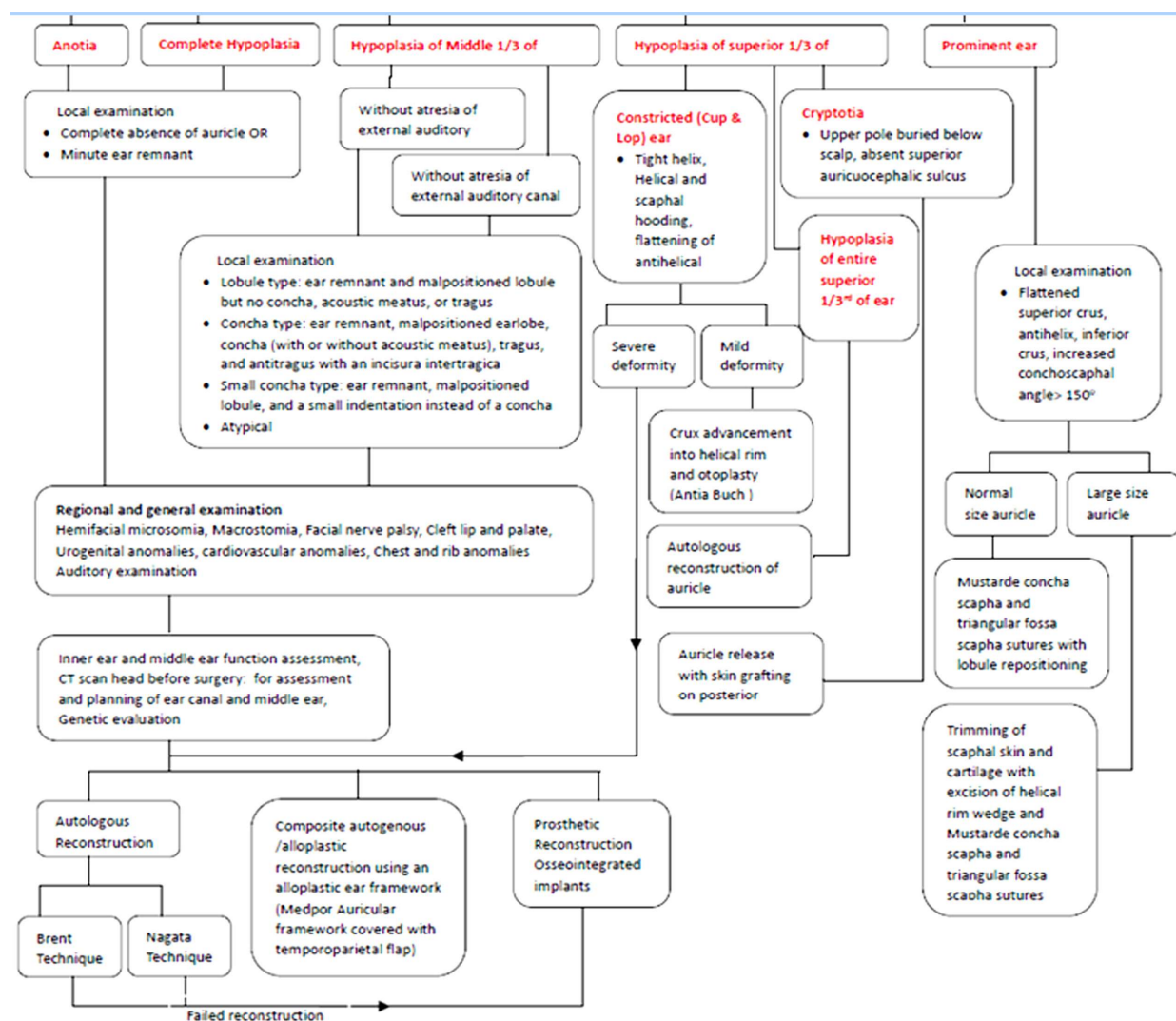
and its complications. Jahrsdoerfer criteria are a widely accepted guideline to select atresia surgery candidates.¹³ The bone anchored hearing aid (BAHA; Cochlear, Mölnlycke, Sweden; and Ponto; Oticon, Kongeballen, Denmark) has been used since 1977, which does not need functioning middle ear or patent canal. In microtia patients, BAHA was initially started to use for bilateral microtia with bilateral conducting hearing

loss: Unilateral BA HA is usually placed because a single aid will stimulate both cochlea simultaneously. The drawback of BAHA is the interface between titanium and skin: It may cause skin irritation or infection. BAHA has a retention rate of over 95% on long term follow-up, with a soft tissue reaction rate of 30%.¹⁴

Brents technique of auricular reconstruction

MANAGEMENT

Treatment Planning Algorithm



Source: @ Chapter 12: Microtia and congenital ear anomalies, Treatment Planning in Plastic Surgery by Dr Ravi Kumar Chittoria¹⁵

Stage 1: Auricular framework placement - donor site from contralateral 6,7,8,9 costal cartilage.

Stage 2: correction if malposition ear lobule.

Stage 3: Framework elevation.

Stage 4: Concha, triangular fossa, tragus reconstruction.

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