Pediatrics Audiology Developmental Overview

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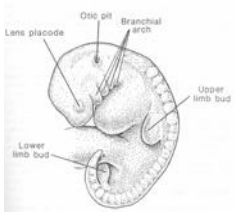
Embryology of Peripheral Auditory System

- First sign of auditory structures at week 4 gestational age.
- An embryo at the end of the fourth week of development.



The otic pit which will develop into the inner ear, and the branchial arches which will contribute to development of the ear and face, are all visible.

W4 GA

- Branchial arches appear as surface elevations on the head/neck area of the embryo. They are composed of mesenchyme derived from mesoderm, covered externally with ectoderm and lined internally with endoderm. They give rise to structures associated with head, neck, face and ear.

Branchial Structures

1. Four branchial arches

• 1st BA: tensor tympani, malleus, incus

• 2nd BA: stapedius, stapes

2. Pharngeal pouches (separate BA internally)

• 1st PP: evolves into tubotympanic recess and to middle ear cavity, the mastoid antrum, the eustachian tube

3. Branchial grooves (clefts) (separate BA externally)

• 1st BG: evolves into external auditory meatus

• swelling of mesenchymal tissue (auricular hillocks) on the 1st and 2nd BA around the 1st BG evolves into the external ear

4. Branchial membranes

• Middle layer of ear drum

Outer and Middle Ear

1. Ossification of ossicles around W16 GA
2. Ossicles are first bones to attain adult size in human body
3. External auditory meatus plugged until W20 GA
4. External ear continues to grow postnatally.

Inner Ear

- Inner ear develops simultaneously but from different tissue (ectodermal tissue)
- W6 GA- cochlea begins to coil
- W8 GA- 1.5 turns of cochlea
- W9 GA- 2.5 turns of cochlea
- W16 GA- membranous labyrinth adult like (33-37 mm)
- After W16 GA- cells surrounding membranous, cochlea ossify and form bony labyrinth.

Inner Ear

- W7-12 GA: development of ampulla, maculae & organ of Corti
- W11-12 GA: undifferentiated IHCs (OHCs at basal end of cochlea)
- W16-17 GA: differentiated HCs (same number as adult)
- W20 GA: mature synapses to IHC
- W20 GA: RUDIMENTARY HEARING
- W34GA: cochlea final size (structure, growth and development complete).

Maturation of Central Nervous System and Auditory Pathways

- At birth
- The newborn begins a rapid period of brain growth that continues well into adolescence
- Few nerve centers are myelinated at birth. In the beginning, only reflexes needed for survival are completely myelinated

In Childhood

- Myelination continues. During the first year-and-a-half of life, the corticospinal motor tract receives its myelination enabling gross control over arms, and legs.
- Neurons continue migrating into positions.

During Adolescence

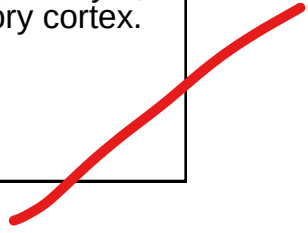
- The brain continues to change and mature during adolescence.
- Final myelination of the frontal lobes occurs in early adolescence.
- An adolescent's brain reaches the weight of an adult brain by about age fourteen due to myelin accumulation and dendritic branching. At this time the potential for contribution to insight, judgment, inhibition, reasoning, and social conscience are possible. The adolescent's frontal lobes are increasingly active, and this ability enables the adolescent to consider several things in the mind while comparing or interrelating them.

In Adulthood

- The brain continuously remodels itself-even into adulthood.
- Synapses continue to be formed in select areas of the brain but growth of new neurons is limited. Prevailing knowledge that the adult brain does not produce new neurons is currently being challenged.
- Lifelong enrichment experiences are important. These experiences continue to cause dendrites to branch, grow, and form new synaptic connections. Brain development continues in adults who regularly exercise their brains with new and varied experiences. Even in adulthood, the brain is continuously remodeling itself.

Auditory maturation: 2nd trimester (weeks 13 to 28)

- Though by W20 GA cochlea approaches its adult state, the central auditory system remains in many ways immature. Brainstem auditory neurons are enlarging rapidly but have only embryonic dendritic processes. The axons of these neurons, which form the brainstem auditory pathways, show signs of neurofilament expression but are not yet myelinated. There is, precocious neurotransmitter expression in efferent (olivocochlear) brainstem neurons. In the forebrain during the second trimester, there is formation of a cortical plate and a marginal layer, which together will constitute the auditory cortex.



Auditory maturation: 3rd trimester to 1 year of life

- Structures in the human brainstem auditory pathway, from the proximal end of the cochlear nerve to the inferior colliculus, undergo myelination in W26-29 GA
- By W26 GA, axons in the cochlear nerve and brainstem pathways have faint myelin sheaths.
- By W29 GA, definitive myelination is present in all auditory pathways, including the proximal end of the cochlear nerve, trapezoid body, lateral lemniscus, and inferior colliculus

Auditory maturation: 3rd trimester to 1 year of life

- Subsequent to the W29 GA, density of myelination increases in all pathways until at least 1 year postnatal age. By the end of the first postnatal year, dendrites on brainstem auditory neurons have grown into their mature branching pattern, and myelin density and axonal conduction velocity approach adult levels.

Auditory maturation: 3rd trimester to 1 year of life

- Auditory cortex still has an immature laminar organization and contains mature axons only in the marginal layer. Although the chief landmarks (sulci and gyri) of the cerebral cortex are present at birth, the cortex remains relatively immature in terms of its intra- and interregional connectivity

Auditory maturation: childhood

- By age 2, the cortex has expanded to an adult-like depth and pattern of lamination, but maturation continues in the axons that carry auditory information from the brainstem to the deep cortical layers until age 5
- From 6 months to 5 years, maturing thalamocortical afferents to the deeper cortical layers are the first source of input to the auditory cortex from lower levels of the auditory system

Auditory maturation: childhood

- From 6 to 12 years, axons mature in the superficial cortical layers that form transcortical connections, both within and between the cerebral hemispheres. This progressive cortical maturation presumably underlies the changes in evoked cortical activity that occur during childhood, as well as behavioral advances in auditory perception and integration.
- From 5 to 12 years, maturation of commissural and association axons in the superficial cortical layers allows communication between different subdivisions of the auditory cortex, thus forming a basis for more complex cortical processing of auditory stimuli. Maturational changes in auditory cortex extend to late teenage years.

Every child is unique!

- **biopsychosocial development** occurs across three separate, overlapping domains

1- biological domain: physical bodily changes, maturation, and growth

2- cognitive domain: mental processes of knowing, which include imagining, perceiving, reasoning, and problem solving

3- psychosocial domain: emotions, personality, and social interactions and expectations

1-2 months

- Motor skills: movements are dominated by primary reflexes

– pulled to sit (considerable head lag)

– automatic walking

1-2 months

- Visual skills (has not seen before birth but rapidly adapts)

– pupils react to light– lids close against intense light– turns, stares to diffuse light

– eyes follow moving object (that is near)

– defensive blink (from around 4-6 w)

1-2 months

- Social skills:

– eye-to-eye contact (2-3 w)

– social smile (4-6 w) and responsive vocalisations (6 w)

– cries, coos (4-6 w)

– startles to loud noises and calms to familiar voices

– eyes often corner reflexly in direction of the sound source

3 months

- Motor skills

– when pulled to sit no head lag

– held sitting

– lifts head in prone position

3 months

- Visual skills:

– visually alert, especially to faces

– converges eyes to finger play

– recognises feeding bottle

3 months

- Social skills:

– smiles, coos, chuckles, laughs

– recognises familiar situations with pleasure

– focus on vowel-like sound and true babbling begins (2-4 mo)

– responds happily to all friendly visitors (up to 6 mo)

6 months

- Motor skills
  – sits with some support
  – turns head with control
  – rolls over front to back (6 mo)
  – rolls over back to front (6.5 mo)

6 months

- Visual skills:
  – visually alert to near and far
  – follow rolling ball at 5 to 10 feet

6 months

- Social skills:

– still friendly with strangers

– takes objects to mouth

– noisy protesting cries when distressed and from 5 mo shouts to attract attention

Note

- **5-6 mo: vocalisations are identical in deaf and hearing infants.** Both increase their vocalisations when the parents speak to them. This is a preadaptive, reflexive response stimulated by the presence of the parent's face, much as the smile response that appears at the same age.

Note

- Babbling seems to represent reflexive preadaptive behaviour unique to human infant. It is the earliest function in the preprogrammed schedule of linguistic activities that can be described as being innate process.
- Babies learn the specific sounds of the language to which they are exposed by 6 months of age

9 months

- Motor skills:

– pulls self to sit (7 mo)
– sits unsupported (8 mo)
– picks up toys
– attempts to crawl (7-8 mo)
– stands holding on (9 mo)

9 months

- Visual skills:

– visually very attentive

– looks for toys

– visual competence rapidly improving

9 months

- Social skills:

– double babble, responsive

– babbles for self-amusement when alone (from 7 mo) as well as with other people dad-dad, abababa, mamama

– understands e.g. bye-bye

– plays peek-a-boo and imitates hand clapping

– turn takes

Note

- 8 mo: spontaneous vocalisations become different in deaf and hearing infants (both in quality and quantity)

12 months

- Motor skills:

– crawls

– pulls to standing, stepping sideways and lets self down safely (from 11 mo)

– may stand alone

– walks with hands held (11 mo)

– may walk alone

12 months

- Visual skills:

– picks up fine objects
– throws toys
– points
– interested in pictures

12 months

- Social skills:

– responds to her own name (10 mo)
– comprehends simple instructions (11 mo)
– initiates vocalisations
– drinks from cup
– may be shy

15 months

- Motor skills:

– walks alone from 13 -14 mo broad base, uneven steps and arms held up to balance

– creeps upstairs safely, but usually not downstairs

– pushes large wheeled toy (16 mo)

15 months

- Visual skills

– builds tower of two

– manipulates small objects

– watches outside events

15 months

- Social skills:

– spontaneous use of a single word in a right context (12-14 mo)

– helps with dressing

– physically restless

– emotionally labile

And onwards...

- 18 mo: 6 word vocabulary
- 24 mo: speaks in understandable 2-word phrases and has a vocabulary of 20 words

Warning Signs!

1. Mother's concern (usually correct!)
2. Not smiling by 8-10 weeks
3. Visually unaware
4. Abnormal gait
5. Not talking by 18 months
6. Abnormal social behaviour

Permanent Childhood Hearing Loss (PCHL)

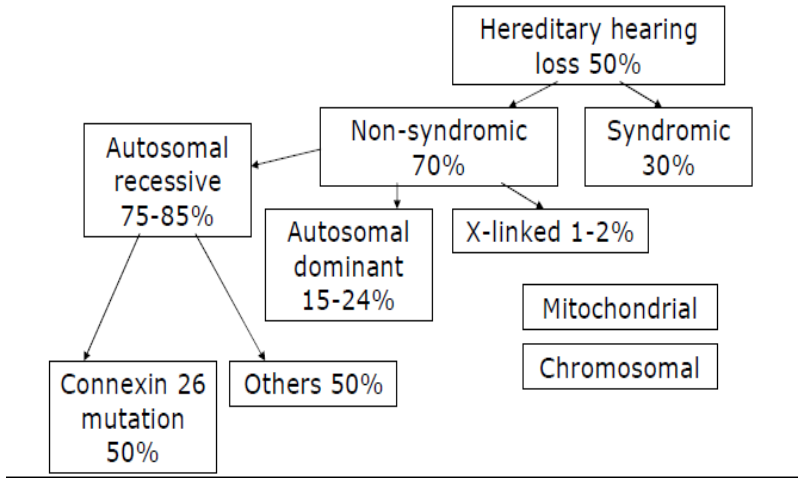
- Defenetion:
- Permanent bilateral moderate or greater hearing loss, the level of the hearing thresholds measured in the better hearing ear at 0.5, 1, 2 & 4 kHz equal to 40 dB HL or worse and the condition is permanent
- Pay attention to

– degree of hearing loss
– unilateral vs bilateral
– age at onset

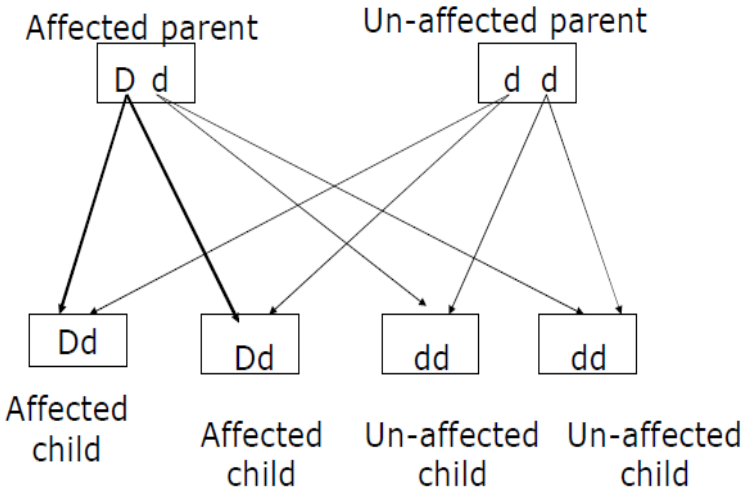
Aetiology of Permanent Childhood Hearing Loss

- Congenital vs Aquired

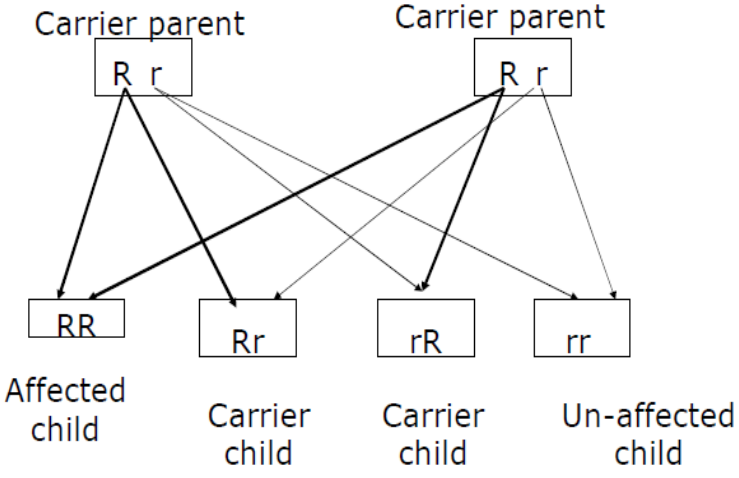
Part 1: Genetic hearing loss



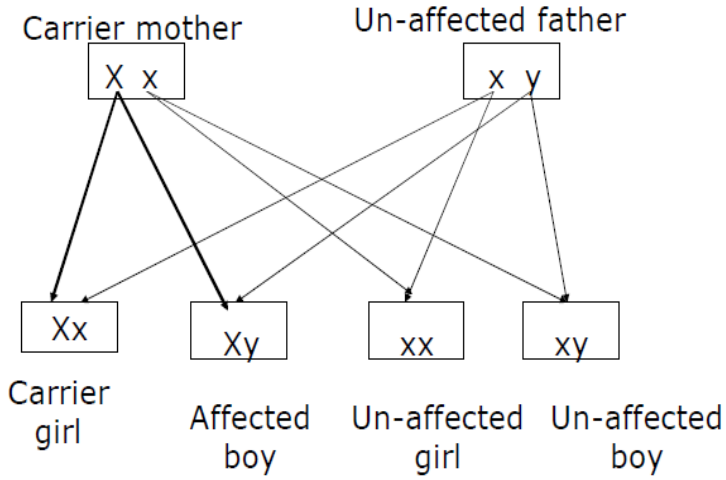
AD Inheritance



AR Inheritance



XL Inheritance



Syndrome

- Syndrome- pattern of abnormalities with a specifically defined cause
- Usher Syndrome
- Waardenburg Syndrome
- Treacher Collins Syndrome
- Branchiootorenal Syndrome
- Pendred Syndrome/DFNB4
- *Alport Syndrome*

Usher Syndrome

Type	Hearing impairment	Vestibular responses	Onset of retinitis pigmentosa
Type I	Profound hearing loss Congenital	Absent	Onset in first decade
Type II	Sloping audiogram Congenital	Normal	Onset in first or second decade
Type III	Progressive hearing loss	Variable	Variable

- Usher Syndrome is inherited in an *autosomal recessive* manner.
- Usher syndrome has been estimated to be responsible for 3%- 6% of all childhood deafness and approximately 50% of all deaf-blindness.

Usher Syndrome type I

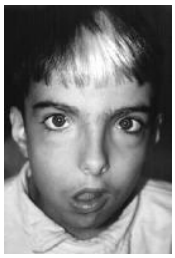
- *Congenital* (i.e., prelingual) profound bilateral sensorineural hearing loss
- No significant vestibular responses
- Retinitis pigmentosa, is a progressive, bilateral, symmetrical degeneration of the retina that initiates at the periphery; rods (photoreceptors active in the dark-adapted state) are mainly affected first, causing night blindness and constricted visual fields (tunnel vision). Onset in first decade
- Normal general health and intellect; otherwise normal physical examination

Usher Syndrome type II

- *Congenital* (i.e., prelingual) sensorineural hearing loss that is mild to moderate in the low frequencies and severe to profound in the higher frequencies
- Intact vestibular responses
- Retinitis pigmentosa, is a progressive, bilateral, symmetrical degeneration of the retina that initiates at the periphery; rods (photoreceptors active in the dark-adapted state) are mainly affected first, causing night blindness and constricted visual fields (tunnel vision). Onset in first or second decade
- Normal general health and intellect; otherwise normal physical examination

Waardenburg Syndrome type I

- WS1 is inherited in an *autosomal dominant* manner.



Waardenburg Syndrome type I

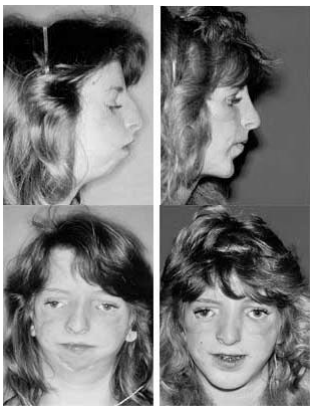
- Major Criteria
  - *Congenital* sensorineural hearing loss
  - White forelock, hair hypopigmentation
  - Pigmentation abnormality of the iris
  - Dystopia canthorum
  - *Affected first-degree relative*

Waardenburg Syndrome type I

- Hearing loss. The hearing loss in WS1 is *congenital*, typically non-progressive, either unilateral or bilateral, and of the sensorineural type. The most common type in WS1 is profound bilateral hearing loss (>100 dB). The dominance of the hearing loss is variable among and within families.
- Various temporal bone abnormalities have been identified in persons with WS1 and hearing loss [*Madden et al 2003*]. The temporal bone abnormalities include enlargement of the vestibular aqueduct and upper vestibule..

Treacher Collins Syndrome

- TCS is inherited in an *autosomal dominant* manner.



Treacher Collins Syndrome

- The prevalence of TCS is estimated to be between 1:10,000 and 1:50,000
- Hypoplasia of the zygomatic bones and mandible [*Posnick 1997*] resulting in the following:
- Midface hypoplasia (89%) with a bilaterally symmetrical convex facial profile, prominent nose, and characteristic downward slant of the eyes secondary to hypoplasia of the lateral aspects of the orbits
- Micrognathia (78%) with variable effects on the temporomandibular joints and jaw muscles
- External ear abnormalities (77%) including absent, small, and malformed ears (microtia) or rotated ears
- Lower eyelid abnormalities including the following:

– Coloboma (notching) (69%)

– Sparse, partially absent, or totally absent cilia (lashes) (53%)

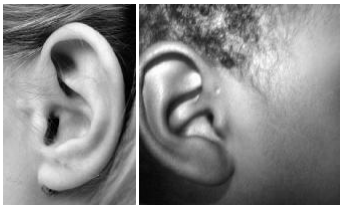
- *Family history* consistent with *autosomal dominant* inheritance (40%)

Treacher Collins Syndrome

- Conductive hearing loss in individuals with TCS is usually attributed to middle ear anomalies including hypoplasia or absence of the ossicles or middle ear cavities. The inner ear structures are typically normal.
- External ear anomalies including absent, small or rotated ears are typical of individuals with TCS, and some may also present with atresia or stenosis of the external auditory canals.

Branchiootorenal Syndrome

- BOR spectrum disorders are inherited in an *autosomal dominant* manner.



Branchiootorenal Syndrome

- **Otologic findings, found in more than 90% of individuals with BOR syndrome** [*Chang et al 2004*], include:

– **Hearing loss** (>90%)

– **Type: mixed** (52%), **conductive** (33%), **sensorineural** (29%)

– **Severity: mild** (27%), **moderate** (22%), **severe** (33%), **profound** (16%)

– **Non-progressive** (~70%), **progressive** (~30%, correlates with presence of a dilated vestibular aqueduct on computed tomography) [*Stinckens et al 2001*, *Kemperman et al 2004*]

Branchiootorenal Syndrome

- Otologic findings, continued:
- Abnormalities of the pinna

– Preauricular pits (82%)

– Lop-ear deformity (36%)

– Preauricular tags (13%)

– Abnormalities of the external auditory canal. Atresia or stenosis (29%)

- Middle ear abnormalities. Malformation, malposition, dislocation, or fixation of the ossicles; reduction in size or malformation of the middle ear space
- Inner ear abnormalities. Variably present:

– Cochlear hypoplasia

– Enlargement of the cochlear and vestibular aqueducts

– Hypoplasia of the lateral semicircular canal [*Ceruti et al 2002*, *Kemperman et al 2002*]

Branchiootorenal Syndrome

- **Second branchial arch anomalies:**
- Branchial cleft sinus tract appearing as a pin-point opening anterior to the sternocleidomastoid muscle, usually in the lower third of the neck
- Branchial cleft cyst appearing as a palpable mass under the sternocleidomastoid muscle, usually above the level of the hyoid bone

Branchiootorenal Syndrome

- **Renal anomalies.** Renal malformations can be unilateral or bilateral and can occur in any combination. The most severe malformations result in miscarriage or neonatal death
- In a study in which 21 affected individuals had renal anomalies were noted in 67%
- **Other findings** [*Chang et al 2004*]:

– Lacrimal duct aplasia
– Short or cleft palate
– Retrognathia
– Facial nerve paralysis
– Gustatory lacrimation

Pendred Syndrome

- Pendred syndrome is inherited in an *autosomal recessive* manner.
- 7.5% of all *congenital* deafness (*Fraser, 1965]*) . If these data are representative, Pendred syndrome is a common cause of *congenital* hearing impairment.
- **Sensorineural hearing impairment that is usually *congenital*, non-progressive, and severe to profound.**
- **Bilateral dilation of the vestibular aqueduct** (DVA) with or without cochlear hypoplasia, in which the labyrinth has one and one-half cochlear turns as opposed to the normal two and three-quarters turns. The presence of both DVA and cochlear hypoplasia is known as Mondini malformation or dysplasia.
- Development of **euthyroid goiter** in late childhood to early adulthood

Alport Syndrome

- The **prevalence** of **Alport syndrome** has been estimated at approximately 1:50,000 live births [*Levy & Feingold 2000*].
- **Renal:**
- Hematuria.100% of *affected* males and more than 90% of *affected* females have hematuria.
- Proteinuria, hypertension, and renal insufficiency develop with advancing age in all males.

Alport Syndrome

- **Ocular:**
- Anterior lenticonus it occurs in 15%-20% of those with Alport syndrome and typically becomes apparent in late adolescence or early adulthood.
- **Cochlear:**
- **Bilateral high-frequency sensorineural hearing loss (SNHL) typically becomes apparent by audiometry in late childhood or early adolescence**
- SNHL eventually develops in 80%-90% of *affected* males as well as in some *affected* females.
- In some families, SNHL may not be detectable until well into adulthood.

Part 2: Congenital Infections

- **Cytomegalovirus (CMV)**
- **Rubella (German measles)**
- **Toxoplasmosis**
- **Syphilis**
- **Herpes**

Cytomegalovirus CMV

- Pathogen: **Cytomegalovirus** (*Herpesviridae*)
- Transmission:

– acquired: CMV via bodily fluids, requires close contact

– congenital: little risk of CMV-related complications for women who have been infected **at least 6 months** prior to conception; risks high when first infection

– **congenital: complications more likely when mother infected <20WG**

– also the virus can also be transmitted to the infant at delivery from contact with genital secretions or later in infancy through breast milk. However, these infections usually result in little or no clinical illness in the infant

CMV

- **Acquired:**
- Asymptomatic
- Similar to infectious mononucleosis and is characterized by swollen lymph nodes, fever, malaise, and muscle and joint pain but without sore throat
- Problematic in immunocompromised patients

CMV

- **Congenital:**
- Most common intrauterine infection: 1% of live births
- 60-70% of these women give birth to seronegative baby
- 10-20% of seropositive babies symptomatic at birth: microcephaly, seizures, "blueberry muffin" rash, and hepatosplenomegaly. Can be fatal.
- 80% to 90% of seropositive babies will have complications within the first few years of life: hearing loss, visual impairment, learning difficulties

CMV: audiology

- **Most common non-genetic aetiological factor for congenital permanent hearing impairment accounting for approx 30% of all cases** (Fowler et al 1995, 2006)
- 10-20% of asymptomatic cases of congenital CMV will have hearing impairment
- CMV-related hearing impairment may be congenital, approx 50% progressive or late onset occurring during the first 6 years of life (Fowler et al 1999)
- variability in the severity of CMV-related hearing impairment ranges from unilateral high frequency losses to profound bilateral losses, ANSD has been reported

German measles

- Pathogen: **Rubella** *(Togaviridae)*
- Transmission: acquired via airborne droplet emission from the upper respiratory tract of active cases. The virus may also be present in the urine, faeces and on the skin. The disease has an incubation period of 2 to 3 weeks
- **The greatest risk for congenital rubella syndrome is when a rubella virus infection occurs during the first three to six weeks after conception. More rarely, throughout pregnancy.**

German measles

- **Acquired:**
- Early symptoms unspecific (3-4 days): mild fever, swollen lymph glands behind the ears and/or neck, malaise, red, watery eyes
- In children, the rubella rash may be the first symptom
- At day 4 classic rash rubella symptom maculopapular RASH appears (lasts 3 days)

– lighter in colour than the measles rash It

– begins at the hairline and then spreads to the face and upper neck.

– gradually moves downward and outward, reaching the hands and feet.

German measles

- Besides congenital rubella syndrome, consequences of a rubella infection during pregnancy include miscarriages and stillbirths
- The classic symptoms for **congenital rubella** syndrome is:

– SNHL (severe and profound, may progress)

– cataract and micropthalmia

– learning difficulties

– microcephaly

– blueberry muffin rash

– hepatopathy and splenopathy

– micrognathia

Syphilis

- *Pathogen:* **Treponema pallidum**
- Transmission:

– Acquired: almost always through sexual contact

– **Congenital: Transplacental transmission of to the foetus does not appear to occur before 12 WG.**

– **Risk greatest for foetal infection if the pregnant mother is in the early infectious stages of syphilis infection (primary, secondary, or early latent syphilis stages).**

– In 40% of cases of untreated maternal syphilis can lead to spontaneous abortion, stillbirth, and premature labour.

Syphilis

- Congenital syphilis (early <2 yrs; late >2yrs):
- The latter is more common, initial symptoms of late congenital syphilis can present anytime from after 2 years of age or into the sixth decade of life.
- Hutchinson's triad:

– hearing loss,

– notched incisors

– interstitial keratitis

Syphilis: audiology

- 12% of children with congenital syphilis develop hearing loss by 10 years of age
- auditory nerve involvement?
- the hearing loss seen in late congenital syphilis presenting in childhood is described as a sudden, bilateral, symmetric and profound loss with no vestibular symptoms.
- In contrast, the hearing loss presentation in adults with late congenital syphilis is also reported as sudden, but typically asymmetric, fluctuating, variable in progression, and often accompanied by tinnitus and vertigo.

Toxoplasmosis

- Pathogen: **Toxoplasma gondii**
- Carriers: warm-blooded animals, including humans, primary host cat
- Transmission:

– Intake of raw or partly cooked meat, especially pork, lamb, or venison containing *Toxoplasma* cysts.

– Intake of contaminated cat faeces

– drinking water contaminated with *Toxoplasma*.

– transplacental infection in utero

– receiving an infected organ transplant or blood transfusion (rare)

Toxoplasmosis

- **Acquired infection** is usually asymptomatic but may cause mild, self-resolving cervical or axillary lymphadenopathy.
- The chance of **congenital infection** increases as the pregnancy proceeds, but the consequences become less severe

Toxoplasmosis

- Manifestations of congenital toxoplasmosis :

– Overt neonatal disease

– Mild or severe disease occurring in the first months of life

– Relapse of a previously undiagnosed infection presenting as late as adolescence

- Severe **congenital toxoplasmosis** is marked by the classic triad:

– Chorioretinitis

– Intracranial calcifications

– Hydrocephalus

Toxoplasmosis: audiology

- Toxoplasmosis generally listed as a risk factor for congenital or late-onset hearing loss
- SNHL in 14 - 28%
- Delayed-onset or progressive toxoplasmosis-associated SNHL
- Effect of antiparasitic treatment starting prior to 2.5 months on hearing

Herpes

- Pathogen: **Herpes Simplex Virus 1 & 2**
- Both types of herpes simplex virus (HSV), HSV-1 and HSV-2, can cause oral or genital infection.
- Transmission of HSV occurs from close contact with an individual who is actively shedding virus. Viral shedding generally occurs from lesions but can occur even when lesions are not apparent

  and sometime occurs without lesions

Herpes

- Commonly cause recurrent infection affecting the skin, mouth, lips, eyes, and genitals
- Common severe infections include encephalitis, meningitis, and,
- In immunocompromised patients, spread infection

Herpes

- **Neonatal HSV** infection usually transmitted during parturition
- Diagnosis is by viral culture
- Neonatal HSV infection has high mortality. Incidence estimates range from 1/3,000 to 1/20,000 births. HSV2 80% of cases; 20% HSV1
- 95% of survivors have severe neurologic diseases; 30% develop neurologic impairment, which may not manifest until 2 to 3 yr of age.

Herpes: audiology

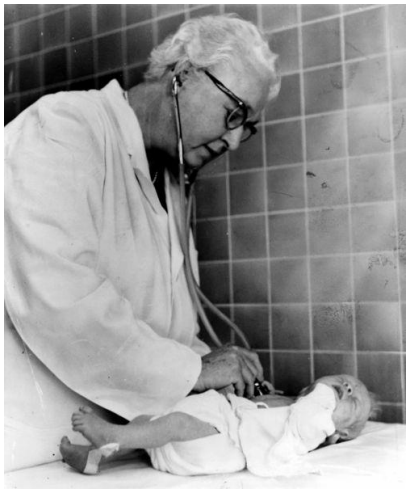
- There are no reports of delayed-onset SNHL following perinatal or asymptomatic HSV infection.
- The development of SNHL in children with exposure to HSV is rare. Routine serological screening for HSV infection in otherwise healthy neonates newly diagnosed with SNHL is unjustified. There is insufficient data to define the incidence and natural history of SNHL in children with HSV infections.
- More research needed.

Risk factors for permanent childhood hearing loss

- History of treatment in an Neonatal Intensive Care Unit (NICU) or special care baby unit (SCBU) for more than 48 hours
- Family history of early childhood deafness
- Craniofacial anomaly (e.g. cleft palate) associated with hearing impairment

Infant Hearing Screening

- **Apgar score**
- devised by Virginia Apgar in 1953



Apgar score

	Score of 0	Score of 1	Score of 2	
Skin colour	blue allover	blue at extremities	normal	Appearance
Heart rate	Absent	<100	>100	Pulse
Reflex irritability	No response to stimulation	Feeble cry when stimulated	sneeze/ cough/ pulls away when stimulated	Grimace
Muscle tone	none	Some flexion	Active movement	Activity
Respiration	absent	weak	strong	Respiration

Auditory Responses

- Typical Sound-Field Minimum Response Levels for Normal-Hearing Children from Birth to 24 Months of Age

Age (months)	Noisemakers (dB HL)	Warble Tones (dB HL)	Speech (dB HL)
0 – 4	40	70	45
4 – 6	45	50	25
6 – 8	25	45	20
8 – 10	20	35	10
10 – 14	20	30	10
14 - 20	20	25	10
20 - 24	15	25	10

Neonatal Screening with ABR

- Auditory brain-stem response (ABR) audiometry has become increasingly popular as a neonatal testing system.
- Disadvantages:

1- The lack of frequency specificity evident when click stimuli are used.

2- The lake of norms that is correspond to children's gestational ages

3- Expensive

Neonatal Screening with Otoacoustic Emissions

- The OAEs have been shown to be specific, sensitive, and cost effective measures in infants.
- An OAE screen followed by an ABR screen has been reported to be successful, screenings of both OAE and ABR are more effective than the use of either measure alone.

Behavioural tests

- 0 – 6 mths: Behavioural observation audiometry
- 6 – 12 mths: Distraction test
- 6 – 36 mths: Visual Reinforcement Audiometry (VRA)
- 30 – 36+ mths: Play audiometry

Behavioural tests

- Why not just use objective tests like ABR or OAE?

1- It reveals the Auditory thresholds or minimum response levels

2- Behavioural tests give information on the entire auditory pathway

3- Response to sounds gives information on functional auditory behaviour

4- Speech testing gives insight into disability

5- Quicker, less technical equipment needed (e.g. compared to ABR)

A typical paediatric hearing assessment:

1. Gathering information
   - Referral letter
   - History taking
2. Testing phase
   - Behavioural testing
   - Otoscopy / Tympanometry
   - ? Other (e.g. TE OAE)
3. Patient management
   - Review
   - Discharge
   - Refer
   - Link back to patient history

BOA (0 – 6 months)

1. Baby observed in a quiet state
2. Then during presentation of sounds

- Unconditioned response
- Shows responsiveness/awareness to sounds at particular measured levels
- Often a test of last resort (may do prior to objective testing) Or, following aiding

BOA (0 – 6 months) Stimuli

- Broad band stimuli (speech sounds) most effective but not frequency specific
- Sound field presentation
- E.g. NBN/warble tones (60 – 80 dB)

BOA (0 – 6 months) Response

Easier to see:

- stilling (from mild activity)
- arousal (from quiet or light sleep)
- eye widening
- blinking/startle

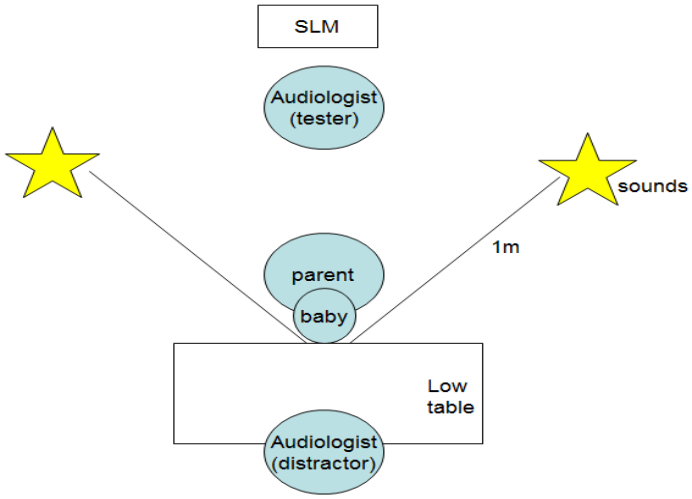
Harder to see:

- limb movement
- sucking
- grimacing
- breathing change

BOA (0 – 6 months) Response

- Highly influenced by arousal state
  - (best during light sleep to quiet awake; 'heavy eyelids')
- Supra- threshold information only
- Rapid habituation
- Highly subjective.

Distraction Test (6-12 months)



Distraction Test

- Two testers, one presents sounds (soundfield) the other controls infants attention
- Preferable that infant is alert, can sit up straight on parents knee & perform head turns (~6mths of age)
- 2nd tester presents stimulus out of sight & waits for a head turn response- response then reinforced
- No sound trials essential
- Can give good threshold information (if done well) but only gives the better ear

Distraction Test Stimuli

- HF rattle (6-8 kHz @ 35dB A)
- Live voice 'unforced hum' (~5 kHz)
- Live voice 's' (~4 kHz)
- FM warble tones or NBN

Visual Reinforcement Audiometry (6-36 months)

- Widely used in hospital based clinics: test of choice for 6-36mth age group
- Based on principle that child will turn to a sound (or tactile) stimulus **involuntarily**
- but this response is **reinforced** in order for it to be repeated, using a visual reward

VRA in principle

Stage 1 (conditioning) :
sound + visual reward = head turn [classical conditioning]

THEN:

Stage 2 (testing):
sound = head turn → *reinforced by* visual stim.

Stage 2 (testing):
sound = NO head turn → NO *reinforcement*