

# PAEDIATRIC USNHL



CLINICAL PROFESSOR HARVEY COATES AO, DM, MS, FRACS

UNIVERSITY OF WESTERN AUSTRALIA



# Wadjuk Noongar Boodja



- We recognise and acknowledge the traditional custodians – the Wadjuk people of the Noongar nation
- This is – and always will be – Aboriginal land and the spiritual connection of people to country is undiminished
- We thank the Elders, past and present, for trusting us and allowing us to work with the children we see
- We work with Aboriginal communities to the very best of our energy and ability to help children be well and families to flourish

# WHAT IS UNILATERAL SNHL?

- Normal hearing in one ear and hearing loss in the other
- Not always detected early ( 5 years often pre NHS)
- Affects neural plasticity with auditory deprivation
- Can be congenital, progressive or acquired



# GENETICS OF USNHL

- NZ Study ( Colin Brown, 2016)
- Retrospective study of 60 children with USNHL
- 17 mild, 17 moderate, 7 severe and 19 profound SNHL
- DNA testing for 11 genes for non-syndromic SNHL
- 31% of children tested—positive to Connexin 26
- CT/MRI positive in 37% of children tested



# CAUSES OF USNHL

- No cause found in 50% of cases
- Hereditary in 33%
  - LVA 20%
  - Cochlear malformation 10%
  - Ossicular anomaly 5%
  - Atresia 3% ---also agenesis of cochlear N
- Acquired---infections/temporal bone fracture 10%



# PREVALENCE

- 1 in 1000 in UNHS Programs
- =one third of all children with SNHL
- Increased USNHL as children age
  - Delayed onset congenital HL
  - Trauma, infections, ototoxicity, NIHL
- in USA 2.5% of adolescents have HL >25DB in at least one ear



# A SIGNIFICANT HEARING LOSS?



- US----USNHL not eligible for services in some states
- UK----Permanent HL is bilateral SNHL >40dB
- Australia---Our advantage—Australian Hearing



# PROGRESSION OF HEARING LOSS



- Child with mild-severe USNHL at risk of progression as high as 40%
- Risk of progression to bilateral SNHL is <20%

(Lieu J et al 2018)



# USNHL—HEARING AIDS



- Hearing Aid provision improves directional and selective HL in quiet and noise
- Improved hearing related quality of life
- Even mild HL (30-40dB) should trial Hearing Aids
- Provide Hearing Aid at or before end of first year of life with USNHL

( Rohlfs, 2017)

# EARLY DIAGNOSIS AND TREATMENT

## - Results

- Improved verbal-cognitive scores
- Improved linguistic and communication scores
- Improved socio-emotional development

(Rohlf, 2017)



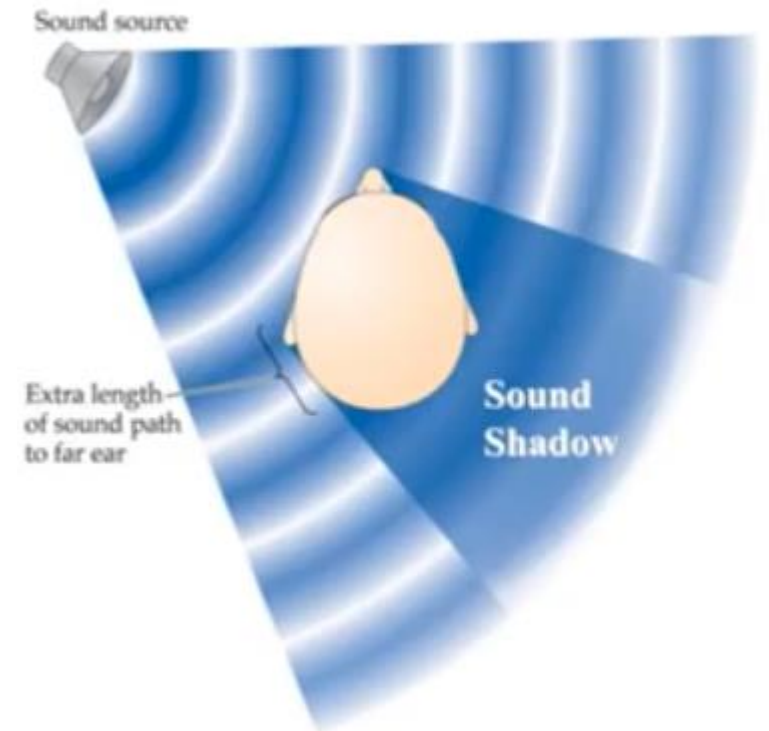
# EDUCATIONAL EFFECTS OF USNHL

- 30% repeat a year of school (3% baseline)
- 50% behavioural difficulties (Brookhouser P)
- Educational difficulties in background noise
- Problems learning to read, spelling and writing stories, and doing maths



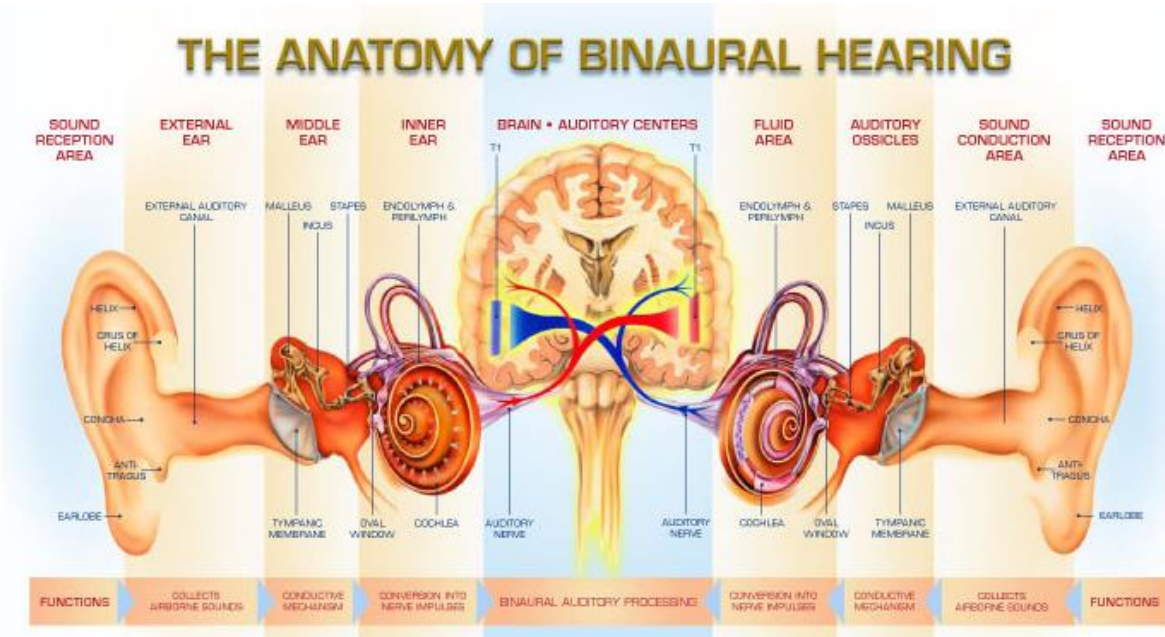
# EFFECT OF USNHL ON A CHILD

- Difficulty telling where sound coming from
- HL in background noise
- Difficulty understanding speech from a distance
- Difficulty hearing someone speaking on their 'bad' side
- 'Spatial hearing and shadow effect'



# WHY USNHL CAUSES LANGUAGE DELAY

- Thought to be impaired binaural ability to understand speech in noise and localise sound
- This may diminish the ability to learn overheard conversation (incidental learning)
- Variables:
  - Innate cognitive ability
  - Severity of SNHL
  - Socio-economic factors



# WHY BEHAVIOURAL DISORDERS?

- Brain network interconnection different in children with SNHL in areas of brain associated with auditory processing, executive function and memory function





# CONUNDRUM

- But children with USNHL who use hearing aids report lower quality of life scores than those who don't wear a device
- Despite studies showing large subjective improvement in hearing function

# INTERVENTION STRATEGIES FOR HEARING REHABILITATION

- Younger aided children better with sound localisation than older children
- FM devices at school show increased word recognition.
- BAHA for severe-profound SNHL
- Should trial softband BAHA for a week before considering BAHA surgery



The small titanium implant



The abutment without sound processor



The sound processor on the abutment



# COCHLEAR IMPLANT—NEXT FRONTIER



- In 3 children—binaural hearing benefit for speech in noise
- Better localisation
- Subjective hearing ability better
- Invasiveness of the cochlear implant procedure weighed against risks of speech delay and educational problems

# SINGLE SIDED DEAFNESS

- When there is non-functioning hearing in one ear and no clinical benefit from amplification with the other ear having normal audiometric function

- Solution:

- CROS aids
- BAHA
- Cochlear implant
- Other devices e.g SoundBite



# WHAT MIGHT THE PARENT NOTICE AT HOME?



- HL in background noise/ TV
- Preference of child to sit on one side for story telling
- Hearing problems in the car when they can't see your face
- Direction sounding difficulty
- Increased hearing loss when they have a cold/ blocked ears

# WHAT MIGHT THE TEACHER NOTICE?

- Ignoring teacher in background noise
- Unable to concentrate / copies others' work
- Misses important instructions
- Problems with soft sounds eg 's', 'f', 'th'
- Maths issues if misses a crucial step
- Problems with talking/ understanding other children in background noise



# WHAT CAN PARENTS DO?

- Frequent visits to school re progress
- Keep reminding teachers each year re HL/preferential seating in class
- Check homework in a quiet area at home
- Ensure, if needed, extra school attention and speech therapy
- Discuss H Aids, FM and CI with audiologists



# WHAT CAN PARENTS DO?

- Regular hearing tests
- Ensure vaccinated against measles, mumps, meningitis
- Treat ear infections urgently. ( only hearing ear)
- Ensure paediatric ENT care if surgery required
- (risks of surgery in only hearing ear)





THANK YOU



Questions ???

