Cochlear implants – where are we now?

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Some future day when what is now is not,
When all old faults and follies are forgot.

(Songs in Absence, Some future day; Arthur Hugh Clough1819-1861)

Outline

- Evidence based
- Basic data about cochlear implants and deafness
- Developments in technology
- In surgery
- In (re)habilitation
- In candidacy
- In medicine
- In numbers
- In outcomes

Basic data: permanent severe to profound deafness

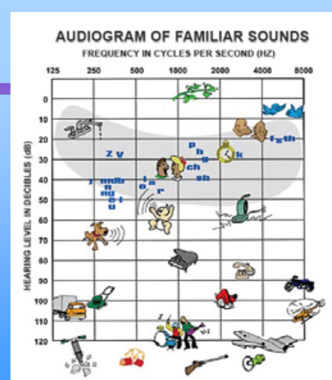
- Approx 400 children born UK each year
- 1:1000 severely/profoundly deaf at 3 years
- 2:1000 severely/ profoundly deaf aged 9-16 years
- 40% have additional difficulties
- Nearly 90% born to hearing parents

What is severe to profound deafness?

- Hearing sounds only at greater than 90dBHL at 2kHz and 4kHz

NICE 2009

- >71dB HL = severe deafness
- >91dB HL = profound



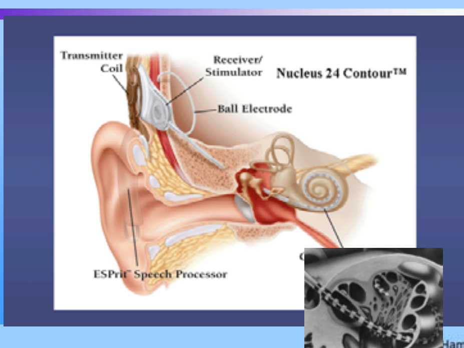
What is a Cochlear Implant?

- An electronic device
- Designed to help severely to profoundly deaf individuals who gain little or no benefit from hearing aids
- Two main parts:
  - an internal implanted part: the implant
  - an external part: the speech processor

How does it work?

- **Microphone:** detects sound
- **Speech processor:** The external speech processor captures sound and converts it into a pattern of digital signals.
- **Digital signals:** The processor sends the digital signals to the internal receiver via the transmitting coil.
- **Electrode array:** The receiver/stimulator converts the signals into electrical energy, then sends the energy to an electrode array (16-22) in the cochlea.
- **VIII nerve:** Electrodes stimulate the nerve in the spiral ganglion.

Internal parts



Normal postop Stenvers

External parts

Components of the Cochlear Implant System

Children fitted with cochlear implant

- **2006-07:**
  221 had unilateral CI (England)
  22 (Wales)
  446 under assessment
- **2007-08:**
  704 unilateral CI
  77 bilateral – 39 simultaneous
  38 sequential
- **Since 1980** > 20,000 children

(Balkany, 2005)

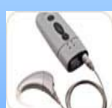
Who has a cochlear implant?

- Permanent sensorineural hearing loss
- Severe to profound
- Limited/no benefit from acoustic hearing aids
- Congenital, acquired, late onset or progressive

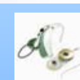
Now in technology

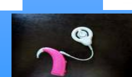
- Most implants now fitted in UK have ear level processor, not bodyworn
- Processing strategies are faster, trying to develop a relationship to human ear which allows appreciation speech and music
- Coils are curved to allow positioning against spiral ganglion, softer, without positioner
- More programmes available for different circumstances
- Radio aids
- Neural telemetry to determine if device functioning effectively

Changing size of bodyworn device

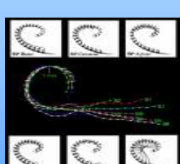
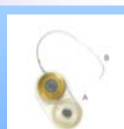



Ear level devices

Improvements in electrodes


Now in surgery

- Softer techniques (minimally invasive surgery) to conserve residual hearing and vestibular function
- Smaller incision: less shaving of hair
- Routine and improved facial nerve monitoring

(Migirov et al ,2006; Stratigouleas et al, 2006)

Now in surgery, ct

- Daycase, even for bilateral CI
- Very low complication rate <3.6% major (Lin et al, 2006; Postelmans et al, 2006)

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Now in (re)habilitation

- Moving back from using local resources to direct therapy in CI programmes: **listening**
- Auditory verbal therapy
- Implanted children did better in school than HA users but gap with hearing children widened as they become older: need flexible approach (Thoutenhooft, 2006)
- >56% needed additional education support in mainstream (Mirkau et al, 2006)

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Now: Changing Criteria

- Babies and infants-NHSP
- Severe to profound hearing losses - emphasis on hearing at 2 & 4 KHz
- Bilateral implants-simultaneous & sequential (NICE guidance, Jan 2009)

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Critical factors associated with outcomes

General

- Age at implant
- Language
- Length of deafness before implant
- VIII integrity & brain plasticity
- Residual hearing
- Length of hearing aid and implant use
- Post-operative rehabilitation and follow-up

Specific

- Developmental level/learning ability.
- Family commitment/dynamics
- ?underlying diagnosis

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Age of implantation: less than 3 years?

- More likely to be using oral communication than children implanted later (Gibson et al, 1998)
- Early implantation, before 3 years, beneficial as language gap would be smaller (McConkey Robbins et al 2004)
- Hardly ever had normal aud perception if implanted after 6 years (Govaerts et al 2002)
- Improved listening and spoken language acquisition (McConkey Robbins et al 2004, Lin et al, 2006, lesinsky et al, 2006)).
- Maximum plasticity central auditory system up to 3 ½ yrs, processing worse esp if >7yrs (Sharma et al, 2006).

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Age of implantation: less than 2 years?

- Rate of language development faster (Gibson et al, 1998)
- 90% integration into mainstream kindergarten (Govaerts et al, 2002)
- Consistently significantly better scores (IT_MAIS) compared with implanted age 2-3 years (Muller et al)
- Central plasticity suggests <2yrs is better (Kral, 2006)

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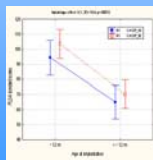
Age at Implantation: under 12 months?

- No problems with surgery
- Very high perception scores within 3 months of CI
- Babbling started within 1-3 months of switch on. (Colletti et al, 2005)



Does diagnosis make a difference? No??

- Waardenburg (2%) (Cullen et al, 2006)
- Connexin 26 (20-50%)
  -less language impaired (Kawasaki et al, 2006)
  -same additional problems as other children (Wiley et al, 2006)
- Usher 1 achieve more independence (Damen et al, 2006)
- Meningitis – same outcome as congenitally deaf
  - same range of additional needs (Nikolopoulos et al, 2006)
- Progressive hearing loss without other problems
- Vestibular function – helps predict which ear
  - post op morbidity
  - diagnosis (Moeller, 2004)

Comparison of Speech Perception Performance According to Genetic Diagnosis

Table 4. Comparison of Speech Perception Performance According to Genetic Diagnosis

Variable	Mean (SD) *SLC26A4* Mutation (n=18)	*GJB2* Mutation (n=4)	No Mutation (n=45)	*P* Value
Male, No. (%)	7 (39)	3 (75)	21 (47)	.46[a]
Age, y	9.7 (3.6)	8.3 (2.3)	9.5 (3.0)	.74[b]
Age at implantation, y	5.7 (3.2)	3.2 (1.0)	4.4 (2.8)	.18[b]
Duration of implant use, y	3.7 (1.4)	4.7 (2.0)	4.6 (1.7)	.12[b]
Pure-tone average before implantation, dBHL	96.7 (10.6)	103.5 (4.4)	103.0 (9.5)	.27[b]
Achievement of open-set speech recognition, No. (%)	18 (100)	4 (100)	34 (76)	.04[a]
Speech recognition scores, %				
Consonant	88.0 (7.2)	90.5 (1.9)	66.2 (38.6)	.04[c]
Vowel	86.2 (6.1)	81.5 (8.4)	64.6 (38.0)	.047[c]
Tone	91.7 (6.3)	95.5 (1.9)	71.0 (41.3)	.06[c]
Phonetically balanced word	79.2 (10.5)	73.5 (6.8)	56.3 (34.4)	.02[c]
Sentence	89.9 (10.8)	93.3 (5.4)	58.3 (37.7)	.001[c]

Abbreviation: dBHL, decibel hearing level.
[a]Fisher exact test.
[b]Analysis of variance.
[c]Analysis of variance, post hoc test for significance with the Tukey multiple comparison procedure: consonant, *SLC26A4* mutation vs no mutation, $P = .047$; vowel, *SLC26A4* mutation vs no mutation, $P = .04$; phonetically balanced word, *SLC26A4* mutation vs no mutation, $P = .02$; and sentence, *SLC26A4* mutation vs no mutation, $P = .002$.

Wu, C.-C. et al. Arch Pediatr Adolesc Med 2008;162:269-276.

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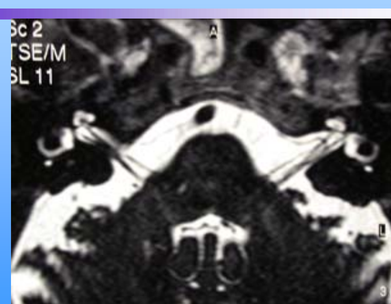
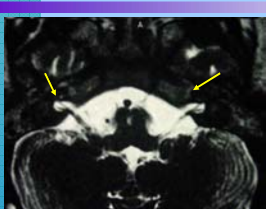
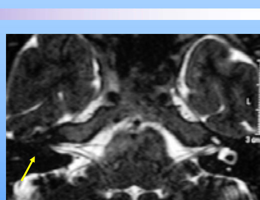
Anatomy axial T2 MRI IAM's



Labyrinthitis Obliterans

Axial T2W images of IAMs:

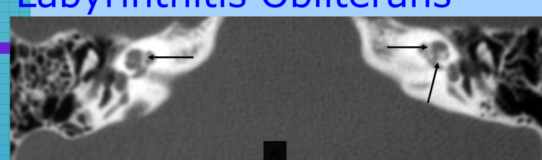




A) Complete obliteration of the membranous labyrinth
B) Obliteration of right membranous labyrinth

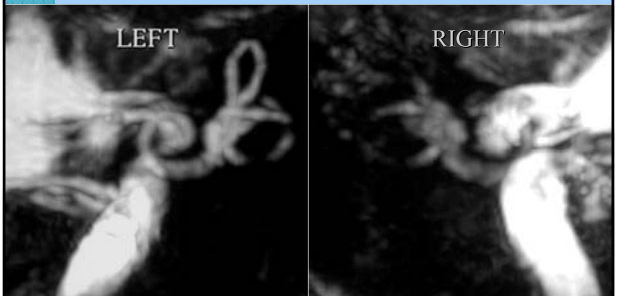
C

Labyrinthitis Obliterans



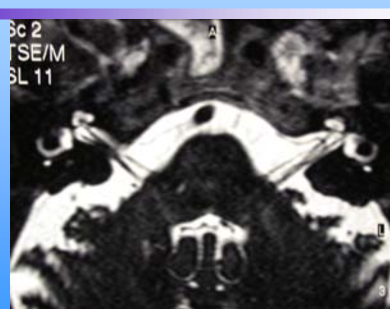
Axial CT (A & B)

Labyrinthitis Obliterans



MIP MRI

Anatomy axial T2 MRI IAM's



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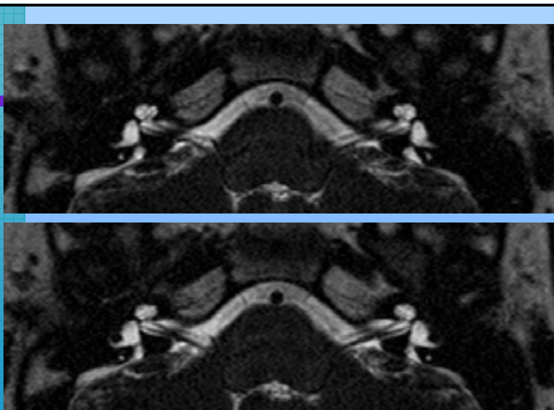

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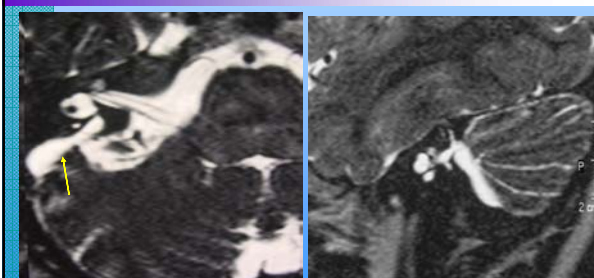
Enlarged Vestibular Aqueduct



Axial parasagittal

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Diagnosis make a difference? maybe…

- Strongest predictors of speech perception and intelligibility were general development and cognition – 40% variability (Edwards et al, 2006)
- 8/40 children had brain abnormalities (Lapointe et al, 2006)
- Autistic spectrum disorder - poor speech perception(Danestri et al, 2006)
- Some cochlear anomalies

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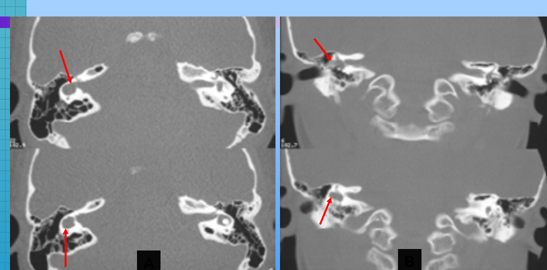
Common Cavity Deformity



Axial (A) and Coronal (B) CT

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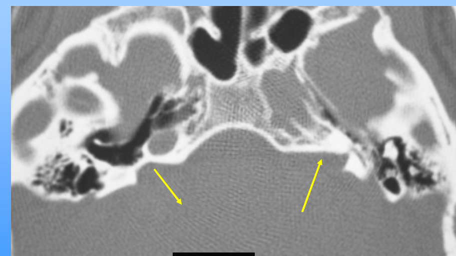
Diagnosis make difference? Absolutely…

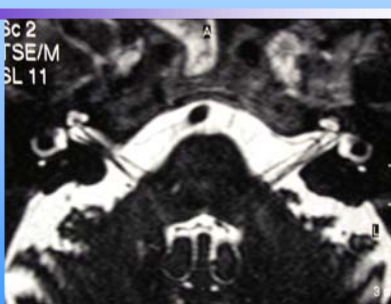
- Severe cochlear anomalies
- Hypoplasia or aplasia of cochlear nerve (Bamiou et al, 2001)
- Severe developmental delay so that unable to benefit from habilitation
- Progressive neurodevelopmental/health conditions

Michel Dysplasia

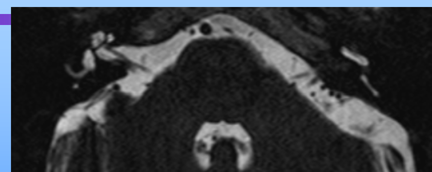


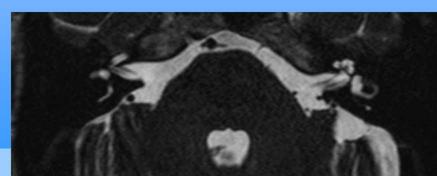
Axial CT

Anatomy axial T2 MRI IAM's

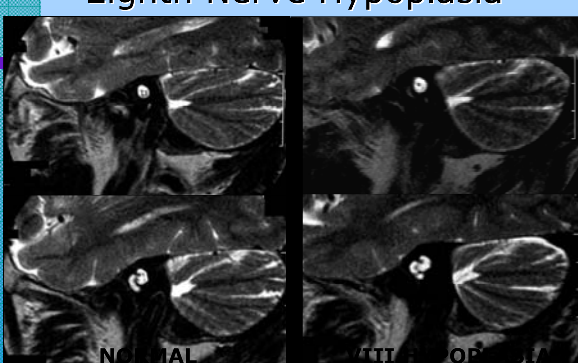


Hypoplastic VIII Nerve





Eighth Nerve Hypoplasia



Comparison of Speech Perception Performance According to Imaging Results

Table 5. Comparison of Speech Perception Performance According to Imaging Results

Variable	Mean (SD) Narrow IAC (n=9)	Other IEMs (n=24)	No IEMs (n=34)	P Value
Male, No. (%)	3 (33)	10 (42)	18 (53)	.52[a]
Age, y	7.3 (2.0)	9.0 (3.3)	9.9 (3.4)	.34[b]
Age at implantation, y	3.4 (1.3)	4.9 (3.1)	4.9 (3.1)	.40[b]
Duration of implant use, y	4.6 (1.6)	3.8 (1.4)	4.8 (1.8)	.08[b]
Pure-tone average before implantation, dBHL	106.3 (9.7)	98.4 (11.5)	103.1 (7.5)	.06[b]
Achievement of open-set speech recognition, No. (%)	2 (22)	23 (96)	31 (91)	<.001[a]
Speech recognition scores, %				
Consonant	17.8 (35.3)	82.4 (19.1)	81.1 (26.4)	<.001[c]
Vowel	18.0 (35.7)	81.7 (18.3)	78.0 (26.3)	<.001[c]
Tone	21.3 (42.3)	88.2 (19.8)	85.7 (27.9)	<.001[c]
Phonetically balanced word	14.7 (29.1)	73.2 (19.0)	68.8 (25.2)	<.001[c]
Sentence	15.7 (31.1)	80.3 (23.8)	73.4 (29.4)	<.001[c]

Abbreviations: dBHL, decibel hearing level; IAC, internal auditory canal; IEM, inner ear malformation.
[a] Fisher exact test.
[b] Analysis of variance.
[c] Analysis of variance, post hoc test for significance with the Tukey multiple comparison procedure: consonant, vowel, tone, phonetically balanced word, and sentence: narrow IAC vs other IEMs, all $P < .001$ and narrow IAC vs no IEMs, all $P < .001$.

Wu, C.-C. et al. Arch Pediatr Adolesc Med 2008;162:269-276.

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If it be now, 'tis not to come; if it be not to come, it will be now; if it be not now, yet it will come: the readiness is all.

(Hamlet v. i)

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Selection Criteria for Children

- Bilateral severe-profound sensorineural hearing loss
- Little or no benefit for speech understanding from conventional hearing aids following hearing aid trial
- No minimum age for referral
- No medical or radiological contraindications
- Local professional & educational support
- Appropriate expectations and commitment from the family

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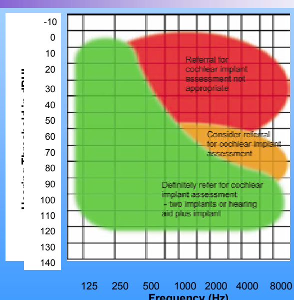
Referral Guidelines for Paediatric Cochlear Implants

NHS
Royal National Throat Nose & Ear Hospital



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Now: bilateral implants?

- Localisation of sound
- Speech perception in background noise (Peters et al, 2007)
- Stimulation both auditory pathways
- Improved enjoyment of music
- Insurance against device failure
- Better ear will always be stimulated
- ?do they make faster progress?

(BCIG 2006)

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Bimodal=HA + CI

- ?better music appreciation (CI users reognise the original music but not melody alone, (Vongpaisal et al, 2006)
- ?better pitch perception
- ?better resolution of concurrent voices
- Better than unilateral CI alone (Ching et al, 2006)

But

- Bilateral CI better for speech perception (Litovsky et al, 2006)

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If bilateral -?simultaneous ?sequential

- Sequential better than bimodal at localisation <20 deg (Litovsky et al 2006)
- Simultaneous cheaper and less stressful
- Optimally within 12 months
- Should be < 8yrs old with 1st implant 4yrs
- May not be helpful if >13yrs (Graham et al, 2008)

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So, for children with limited benefit from acoustic HA

- Speech, language and listening skills appropriate to age, development and cognitive ability
- Multidisciplinary team assessment
- HA for at least 3 months
- Equality of access (E2L, physical, cognitive, communication)

NICE 2009

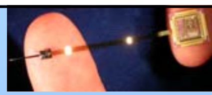
NICE Guidance (Jan 2009) Final Appraisal Determination (FAD)

Unilateral:
- Severe-profound HL (all children)
- No adequate benefit from acoustic HA

Simultaneous:
- All children with severe - profound HL (12 months-17 years)

Sequential:
- Have a unilateral CI (categories above)
- Contra-lateral ear within criteria
- Considered to provide benefit by clinicians & after informed discussion with the individual & carers.

The future?



- Improved diagnostic ability to contribute to outcomes
- Bilateral cochlear implant,
- Concealed (internal) cochlear implant
- Neurotrophic factors through CI (Paasche et al, 2006)
- Younger children? (Singh et al, 2006)
- Hybrid implants with short electrodes for HF hearing loss- children's hearing too unpredictable (Yao et al, 2006)
- Event-related potentials as predictors?
- More cohesion between CI centres (eg pan London)
- More knowledge…

Now, *here*, you see, it takes all the running you can do, to keep in the same place. If you want to get somewhere else, you must run at least twice as fast as that.

(Through the Looking-Glass, ch 1, Lewis Carroll 1832-1898)