

From Infancy to Maturity: Successes and Challenges in Paediatric Cochlear Implantation 20th Graham Fraser Memorial Lecture By Philip J. Robinson

A summary

Mr Robinson began by saying he would address the successes and challenges that still remained in paediatric implantation. The first congenitally deaf child was implanted in Sydney in 1987. In the UK the first implant of a child was in an eight year old post-lingually deafened boy. Despite initial protests from the Deaf community five hundred children now receive cochlear implants each year in the UK. (Worldwide about 20,000 children are implanted each year.) Almost all of them receive two cochlear implants thanks to the excellent work done by Chris Raine and Louise Craddock who, on behalf of the British Cochlear Implant Group, persuaded NICE of the need for bilateral implants for children.

Increasingly the trend is to implant at a younger age because newborn screening detection of profound deafness in babies makes this possible. Hearing aids are fitted at six weeks or earlier. Children who go on to have an implant generally receive one between the age of one and two but now the aim is to do it before they are one.

Even at birth the congenitally deaf child will have missed out on many weeks of auditory development in the womb. There is a period of neural plasticity during which the brain can adapt to new stimuli, but after this it becomes less possible. Significant differences are noted in children implanted before the age of three, especially before the age of two – there is better auditory performance and speech development. Of the children implanted when they are under two 90% are able to attend mainstream school while only 20% of those implanted over the age of four can. Other studies have shown that children implanted



Dr Pat Fraser, Mr Shakeel Saeed and Mr Philip Robinson with his certificate

between 12 to 16 months are more likely to reach age appropriate language performance at 4.5 years than those who receive an implant after their second birthday.

Early implantation gives rise to a number of concerns. Obtaining an accurate assessment of hearing levels is difficult, the anaesthetic and surgical risks increase and there are problems with programming and habilitation afterwards. There may also be other undetected disabilities such as autism. Imaging requires a general anaesthetic at this age and there is concern about the side-effects of computed tomography – MRI is better. Anaesthetic risks are greater with babies under six months and from the surgical point of view there are quite a number of problems such as the very thin skin and skull of young infants and the risk of infection. For the most part the infant undergoing cochlear implantation is just a small adult.

How safe is the surgery?

Between January 2010 and December 2011

DIARY DATES

14 June 2014 Summer Meeting and AGM will be held at the Clore Management Centre, 25-27 Torrington Street, London WC1E 7JL
1 November 2014 Technology Day at University Hospital of Wales, Heath Park, Cardiff CF14 4XW. The workshop will be led by The Ear Foundation.

there was a UK nationwide audit of paediatric CI surgery with 14 centres involved. There were 961 recipients or 1397 implants as most underwent bilateral simultaneous implantation. The majority of the centres recorded 3-4 hours for the whole surgical procedure in bilateral cases and the length of hospital stay was commonly 24 hours. There were 15 major complications and no permanent facial palsies or deaths. There was only one case of meningitis and in two cases wound infection necessitated the removal of the device. Device failure accounted for six cases.

Results

Of 85 children assessed at four years post implant 72 were able to understand common phrases and 50% could use the telephone. Speech intelligibility rating scores were also impressive with 50% of the 90 assessed having speech that was easily intelligible. Those whose speech was not easily intelligible often had additional disabilities.

In education cochlear implanted children do well but there are significant variations – they do not develop subtle communication skills as well as hearing children and are slower developing pragmatic skills which are important for literacy and educational development. In respect of psycho-social adjustment they are no different from their hearing peers. In secondary school there are difficulties. There is a more challenging curriculum, language is more demanding at school and socially and listening conditions are often far from ideal.

With 30% to 50% of hearing impaired children there is another problem such as learning difficulties, auditory neuropathy spectrum disorder (ANSD), cerebral palsy and autism. 10% of hearing impaired children have a degree of ANSD and 5% have autism. Severely disabled children show benefits from cochlear implantation but not in the traditional tests so it is difficult to demonstrate cost effectiveness.

For children with cerebral palsy (20% have hearing or language problems) outcomes are good except for those with severe cognitive disabilities. Of children with autism 13% have a hearing impairment and 3.5% have a profound hearing loss (5% of hearing impaired children are autistic). Autism in some children is not diagnosed before they are implanted. Compared to non-autistic children their performance is poor but there is improvement in behaviour compared to their behaviour before receiving an implant. The majority of parents of cochlear implanted autistic children would recommend an implant.

Mr Robinson lastly discussed one of the great remaining challenges, that of a congenitally deaf child presenting too late to get significant benefit from cochlear implantation: the auditory cortex will develop pathways if stimulated early and often but there is only a limited period for the auditory cortex to develop. This critical or sensitive period when the plasticity is greatest is before the age of three and a half years of age in a child. He concluded his presentation by saying that paediatric cochlear implantation is one of the greatest technological achievements in medicine.

This was the last of the Graham Fraser lectures at the Royal College of Medicine. The lecture and the reception afterwards have always been well attended especially by old colleagues and friends of Graham Fraser who set up the first British cochlear implant programme at the Royal Ear Hospital, the ENT department of University College Hospital, in the early 1980s. Being the 20th lecture and the last one in this beautiful setting the occasion was very nostalgic. We are very grateful to Dr Pat Fraser for organising these wonderful gatherings. Fortunately a new series of Graham Fraser lectures will start under the auspices of the British Cochlear Implant Group which he helped to establish and of which he was the first president. It will form a part of their annual conference – Editor

British Cochlear Implant Group Meeting, March 2014

The British Cochlear Implant Group (BCIG) is the main forum in which professionals working in CI teams and universities in the UK (and Ireland) can come together to share their experience and drive up professional standards. Each year BCIG have a two day Academic Meeting organised by one of the CI Centres, this year's meeting was held at Leeds in late March,

hosted by the Yorkshire CI team. Where possible the NCIUA tries to send an observer to these meetings, which role I undertook this year. Tricia Kemp was also in attendance, wearing her CICS hat as well as an NCIUA one.

Inevitably much of the discussion is fairly technical, but there were several items of

reasonably direct interest to CI users. Members who were able to attend our Conference a couple of years ago will remember that a team from Southampton University gave a presentation on their work developing an interactive music awareness programme (IMAP) to help CI users get the best from their music listening experience.

Mary Grasmeder gave a talk at Leeds announcing that IMAP had now been launched as an online resource for anyone to use, and has agreed to provide an article for our Newsletter giving details of how members can use IMAP (see below).

Another common problem of CI users, especially ones who have only recently been switched on, is the difficulty of getting enough structured listening practice to properly train the brain to make the best use of the sound it is receiving from the implant. Hence it was good to see a presentation by Sandra Driver on behalf of several of the UK CI teams describing the launch of an online system called "Sound Success" which is designed to allow users to practice listening at a time of their choice. Again Sandra has agreed to provide an article for our Newsletter, and in this case she will also be demonstrating the system at our Conference in June.

A consensus is slowly emerging in the CI community that the assessment criteria

currently used to decide who can receive a CI are overdue for review, and that this review also needs to tackle the question of bilateral implants for adults – which is excluded from the guidelines currently approved by NICE. There is a long way to go before the CI community can pull together a solid enough body of evidence to justify asking NICE to conduct a formal review of its guidance, but one or two presentations at Leeds did offer straws in the wind as to where some of the evidence might come from. For example a paper from the UCL Ear Institute suggests that in children the pure tone audibility threshold used as one of the criteria in assessing candidacy could usefully be relaxed by 5dBHL relative to its current level, which would increase the proportion of children qualifying for bilateral implants. Similarly a study by the RNTNE and St Thomas's teams looked at the benefits seen by some teenagers who received implants, but wouldn't have been implanted if they had been assessed as adults. This sheds an interesting light on the current requirement that adults must have a BKB speech perception sentence score (in quiet) of less than 50% to qualify for an implant. Before we can sensibly ask NICE to review their guidance we have to fit together quite a complicated jigsaw of evidence, but I came away from the BCIg meeting feeling that one or two of the pieces are now at least falling onto the table.

Paul Tomlinson

An Interactive Music Awareness Programme for Cochlear Implant Recipients (IMAP)

Created by Rachel M van Besouw, Benjamin R Oliver, Mary L Grasmeder, Sarah M Hodgkinson

Many cochlear implant (CI) users wish to enjoy music but are dissatisfied by its quality as perceived through their implant. However, the provision of interactive music rehabilitation materials that have been developed with and evaluated by CI users remains limited.

In response to this need, an 'interactive music awareness programme' (IMAP) was developed through a series of consultations and music workshops with CI users.

The programme consists of 24 half-hour sessions which enable users to create and manipulate music. Applications include graphical mixers that allow users to control the instrumental/vocal mix, pitch and speed of songs; a drum machine with samples of environmental sounds which

can be looped and layered; and an audiovisual player in which users can combine audio and video clips to explore how music contributes to the mood, meaning and aesthetics of a film.

Twenty-one CI users were recruited to take part in a trial of the IMAP. After 12 weeks use, users showed improved instrument recognition abilities and feedback suggests further positive impact on participants' lives.

A multi-centre trial of the IMAP followed its launch online at www.morefrommusic.org in January 2014.

This research is supported by AHRC grants AH/H039392/1 and AH/K002880/1.

Manufacturer's News

Advanced Bionics



AB Family Fun Day

Center Parcs
in Nottinghamshire

Saturday 14th June 2014

A Day of Celebration for All AB Users and Anyone Considering Cochlear Implants

Advanced Bionics will be hosting a day of fun and celebration for all AB users and their families at Center Parcs in Nottinghamshire on Saturday 14th June 2014.

The free, all-weather event has been organised as a chance to meet professionals from AB and mingle with other AB users and their families. It will provide a great opportunity to hear presentations and view exhibition stands on AB technology, including the new waterproof AquaCase™ container for the Naída CI Q70 processor.

There will be plenty of activities for all the family, including a soft play area for children, and Bionic Buddy will be on hand to meet the kids and pose for photographs! Lunch and refreshments will be served, and everyone attending will have access to Center Parcs facilities after the event, including the Sub Tropical Swimming paradise.

The celebration will run from 10:30 am to 3:00 pm and is open to any AB users, or anyone interested in learning more about receiving an AB cochlear implant, and their families.

If you would like to join in the fun, please register online at www.advancedbionics.com/FamilyFunDay

ADVANCED BIONICS RELEASES DYNAMIC APP FOR NEW NAÍDA CI SOUND PROCESSOR

-- myNaída CI app provides an interactive user guide for AB's groundbreaking Naída CI Q70 sound processor – Advanced Bionics (AB), a global leader in cochlear implant technology and a company of the Sonova Group, announced the launch of a new app that delivers a fun, dynamic and user-friendly guide for its latest innovation, the Naída CI Q70 (Naída CI) sound processor.

The myNaída CI app is an innovative mobile solution for providing product and instructional information to help recipients get the maximum benefit from the exciting

features and accessories available for AB's groundbreaking new processor. Combining AB and Phonak technologies, the innovation leaders in cochlear implants and hearing instruments, Naída CI brings a host of new features to cochlear implant recipients for the first time, including bimodal and bilateral wireless streaming.

Available free for both iPad® and Android™ devices, the app puts comprehensive guides, engaging videos and simple-to-follow instructions right at the fingertips of users. It can be downloaded through iTunes® and

Google Play™ digital distribution platforms. The current English-language release will be followed by additional versions in Spanish, French, Italian, German, Dutch and Portuguese.

"The Naída CI is a quantum leap forward in cochlear implant technology, with never before-seen features for wireless connectivity and performance designed to improve the way our recipients hear their world," said Tracey Kruger, Vice President of Global Marketing. "The myNaída CI app is a fun, engaging way to help recipients get familiar with all that their new processor has to offer."

AB has a track record of creating successful apps, including the Build myNeptune shopping App which won two prestigious Stevie® Awards in the 10th Annual International Business

AwardsSM earlier this year.

"We deliver the most innovative, forward-looking products in our industry," said Hansjuerg Emch, Group Vice President of the Sonova Medical Division within which AB resides. "It is only fitting we use the most modern tools to support them." Launched worldwide in 2013, Naída CI is the most advanced processor in the world with a chic, modern instyle™ design.

To learn more about AB apps, visit AdvancedBionics.com/ABapps.

For information on all Advanced Bionics products or services, contact a local AB representative, email info.uk@advancedbionics.com or visit AdvancedBionics.com.

Cochlear™ Nucleus® 6 Update

Nucleus 6 has been available to all UK implant centres since July 31st 2013. Since then we have had lots of people notice the differences. Here are some of the differences of Nucleus 6 and some comments from our recipients:

- 1. SOUND CHANGES QUICKLY, NOW YOUR SOUND PROCESSOR CAN TOO.** With SmartSound iQ your processor will automatically keep up with your hearing needs so you can enjoy life with minimal listening effort. "I am not sure how other people who have a cochlear implant cope without it". (newly implanted adult patient talking about SCAN program)
- 2. SOMETIMES THE BIGGEST MIRACLES ARE ALSO THE SMALLEST** Enjoy wearing it all day, every day. "With my auto-on function I just change the battery and its ready to use again- no fiddling with buttons again" (newly upgraded Nucleus 6 recipient)
- 3. MAKE THE MOST OF YOUR NATURAL HEARING.** Enjoy the best of both worlds with the all-in-one cochlear implant and hearing aid which works simultaneously to enrich your hearing.
- 4. FREEDOM, IN AND AROUND WATER.** Be confident to take a walk in the rain, enjoy the pool or have the freedom to swim in the sea, as your sound processor is now fully waterproof. "This is the best product! Now my son can participate in water activities and still hear the people around him" (Facebook Mum Comments)
- 5. ENHANCE YOUR HEARING EXPERIENCE, WIRELESSLY.** Enjoy clear speech in noisy situations and have effortless phone conversations all through a truly wireless connectivity that streams audio directly into your sound processor.
- 6. A PROCESSOR SO SMART, IT CAN TELL YOU WHERE IT'S BEEN**
Auditory information about your child's Nucleus cochlear implant can be downloaded by your audiologist. For example, information on how many hours per day the processor has been working. That's especially reassuring when your child is in the classroom and you're at work. Your audiologist can study listening environments and detect patterns that will make the fine tuning process much more accurate.

Messing around in the Water with Nucleus 6

A little while ago Cochlear launched the Aqua Accessory and we have had such a fantastic response to it. Sometimes these things really take us by surprise! Shortly, the disposable device will be replaced by a re-useable one and shortly after this (in time for the summer) you will see Aqua+ appear with any Nucleus 6 that is chosen at clinics. Aqua + is a silicone rubber sleeve for the processor and a waterproof cable and coil that makes the Nucleus 6 the only ear-level processor that is waterproof.

Handy Alerts and Tips on Nucleus 6

How do you know your Nucleus 6 is not working or if the battery has drained?

Well, it's very simple with Nucleus devices. If the coil is not sending a signal to the implant it will flash orange every second (it may have become disconnected from the magnet). If the orange light flashes are continuous then this means that the battery needs replacing. You can also settle your mind by checking what the processor is picking up by using the monitor earphones. These are supplied and plug straight into the accessory port.

Adult Cochlear Implantation: evidence and experience – the case for a review of provision by Brian Lamb and Sue Archbold

This review of the literature about the consequences of severe and profound hearing loss in adults and the low provision of cochlear implants makes a compelling case for change. There are an estimated 460,000 people in the UK with a severe to profound hearing loss in the UK but only about 5% of adults who could benefit from an implant actually get one. Germany, Austria and Belgium are implanting many more adults.

Hearing loss in adults is linked with unemployment, increased risk of poor health, depression and other conditions including dementia. However there is little awareness of the impact of hearing loss or of the latest hearing technologies that can relieve it.

Some of the recommendations of the authors are:

- a review by NICE on its current guidance on cochlear implantation on the criteria around both unilateral and bilateral implantation
- greater education of GPs and audiologists on the potential benefits for adult and older patients
- the need for commissioning models to look at the wider health costs in decision making. In the case of hearing loss it would mean the consequences of not addressing it earlier.

Andrew Dunlop, a GP with a cochlear implant, said, 'I am passionate about encouraging and promoting the possibilities of cochlear implantation – I think there is an "iceberg of unmet need" out there. I feel GPs need more information about the procedure, and more patients should be referred to ENT consultants and audiologists who have experience and understanding of the role that cochlear implants can have in transforming lives.'

For further information about the report please see www.earfoundation.org.uk

This review was mentioned in our last Newsletter in a report of the Ear Foundation's conference on adult deafness by Nigel Williams

FROM MED-EL

MED-EL Music Grants 2014 – Apply now!

In 2013 MED-EL UK launched its music grant programme and it obviously struck the right chord: the judging panel were overwhelmed by the level of interest and had the difficult task selecting only two winners from the many deserving applicants.

The piano proved a popular choice for last year's winners. The under 19s MED-EL Music Grant went to 8 year old Adam Kassam from London, allowing his family to purchase a full size electric piano so Adam could continue with his music lessons.

The over 19s winner, Claire Ingham from Woking who has been deaf since birth, is also discovering music since receiving her MED-EL cochlear implant system. She achieved her first musical goal of playing Christmas carols for her family on the piano during the festive season.

So if you would like to learn how to tinkle the ivories or have a blast on the trumpet and you are the recipient of a MED-EL hearing implant, follow the example of Adam and Claire and apply for this year's MED-EL music grant awards. Applications are now open!

There are two categories of Music Grants: one for applicants under 19 years old and one for 19s and over. To be eligible you must be the recipient of a MED-EL hearing implant. Applicants under the age of 18 years old must have the consent of their parent or legal guardian.

The winners of the grants will receive a musical instrument of their choice up to the value of £500 and MED-EL will pay for one 30 minute lesson per week for one year. There is also a £30 allowance for sheet music. Travel expenses will not be covered. All tuition fees will be paid directly to the music tutor.

Applications are welcome from all MED-EL implant users with an interest in music, whether they are complete beginners or have prior musical experience.

The deadline for applications is 29th July 2014. For more information, terms and conditions, and an online application form visit: www.medel.com/uk/musicgrant, alternatively please email conferences@medel.co.uk for a paper application form or call the marketing team on



Claire Ingham - over 19's Winner

01226 242874.

A final word of encouragement comes from Altaf Kassam who applied in 2013 on behalf of his son Adam: "In August last year we heard the wonderful news from MED-EL that Adam was the proud winner of the under 19s music grant. This award has meant that Adam can continue his musical progress. We strongly encourage all parents of eligible children to apply. What have you got to lose?"

Good luck everyone!

connecting people

Hearing Link

Ask our helpdesk

Use our helpdesk to find information and support and to connect with others who are deafened.



A group of about ten people, mostly young adults, are posing for a group photo. Some are wearing Hearing Link t-shirts. One person is holding a sign that says "A UK charity for people with hearing loss www.hearinglink.org".

We're a UK-wide charity. Contact us:

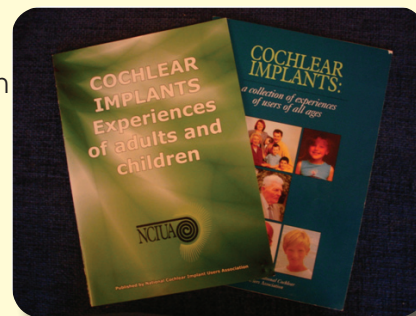
helpdesk@hearinglink.org
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SMS: 07526 123255
www.hearinglink.org

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HRH
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WE NEED YOUR HELP!

for a new cochlear implant book of users' experiences

Our most recent booklet, 'Cochlear Implants: experiences of adults and children', is rapidly going out of print despite a print run of 6,000 copies. It has been very successful like its predecessor, 'Cochlear Implants: a collection of experiences of users of all ages', which also had a large print run. There is a continued demand for it and we need your help to compile a new collection of experiences to help those considering an implant. Unlike our first two booklets, this one will be confined to adult experiences.



We need the stories of users, young and not so young, from all walks of life and with different types of deafness. Some people have suffered sudden hearing loss while others have gone progressively deaf over the years. Some users have been deaf since childhood but have been able to manage with the help of hearing aids until they lost what little hearing they had or found that a cochlear implant was a better option.

If you are interested in helping us, to begin with, please let us have brief details of your experiences of deafness. How it affected your life and career, and how your cochlear implant has helped you. We aim to compile a balanced collection of users' experiences to reflect the many varied reasons that lead them to decide on a cochlear implant as a solution to their deafness.

We look forward to hearing from you. Please contact us by email:

alisonheath71@hotmail.co.uk, or write to: Mrs Alison Heath, Hill End Farm, Hogshaw, Buckingham MK18 3JY

The Ear Foundation

Hearing & Communicating in a Technological Era



Sound Advice Adult Days 2014 Helping adults make the most of their hearing technology

These group days give adults, their partners and friends the opportunity to meet up with time to talk on a range of topics. They are run by experienced professionals who can offer friendly help and up to date advice. Adults are also able to visit our [Resource Centre](#) throughout the day for hands on advice.

BBQ

Saturday 5 July (3:00pm till 6:00pm)
Come and join us for our annual BBQ, in Normanton on Soar, where there will be lots of stalls, games and food, everyone welcome!

(The Willows, Butt Lane, Normanton on Soar, LE12 5EE)

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Disclaimer

Whilst the Association uses its best endeavours to provide accurate information on the subject of cochlear implants it does not provide medical advice or make recommendations with regard to any particular implant or equipment and no article in this newsletter should be construed as doing so.

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