Institution: University of Southampton

Unit of Assessment: 01 Clinical Medicine

Title of case study: 01-31 Universal newborn screening for permanent hearing impairment

1. Summary of the impact

Permanent childhood hearing impairment (PCHI) is common and adversely affects language acquisition. Early identification enables effective early interventions including hearing aids and cochlear implants. Research at Southampton was central to the case accepted by policymakers in the UK, USA and across several continents to recommend universal newborn screening (UNS) for PCHI. From 2008-13 more than three million babies in the UK were screened and over 5,000 cases of PCHI were identified with benefit to family functioning, literacy, academic achievement, social-emotional well-being, employment, wider society and the UK economy.

2. Underpinning research

PCHI can have adverse effects on a child's neuronal development, language acquisition and educational outcomes. If PCHI is detected at an early age, children can be provided with educational support, hearing aids and cochlear implants. Prior to 2001, the standard test for PCHI in the UK was the health visitor distraction test (HVDT). Health workers attempted to distract babies with a noise and then assess their reactions. This method posed two significant problems: the subjectivity of the test and the relatively late developmental age – seven months – at which it can be carried out.

A programme of work headed by Colin Kennedy (1988-date, Professor of Neurology and Paediatrics since 2006) has led research demonstrating that tests based on transient evoked otoacoustic emissions (TEOAEs) - low level sounds detectable by a microphone in the ear canal of a normally functioning cochlea - combined with automated auditory brain stem response testing in cases whose TEOAEs are undetectable, are effective as a UNS for bilateral (PCHI >40 decibels Hearing Level (dB HL) over four sound frequencies) **[3.1, 3.2]**. This degree of PCHI occurs in more than 1 in 1000 babies.

A population-based trial across Wessex in over 50,000 newborns showed that UNS increased the odds of referral of cases of bilateral PCHI >40 dB HL prior to age six months 19 fold (95% CI 3.2 to 111.0) **[3.1]**. Over an eight-year period to 2003, a follow-up study of the birth cohort enrolled in the Wessex Trial allowed the identification of all true cases of PCHI, including UNS false negatives and cases of progressive PCHI **[3.3]**. UNS more than doubled the proportion of all true cases of PCHI that were referred before age six months from 31% to 74% **[3.3]**.

The children with PCHI were further studied at age 7-9 years with an additional cohort aged 6-10 years from four districts in Greater London **[3.4-3.6]**, including the only two districts in the UK providing UNS. In this sample (and also in subgroup analysis of the Wessex cohort), UNS was associated with higher adjusted group mean z scores for receptive language as compared with nonverbal ability (difference, 0.60; 95% CI 0.07 to 1.13) and for reading (difference 0.39; 95% CI 0.02 to 0.76) and with £2213 lower educational costs in a 12-month period **[3.4-3.6]**.

The steering committees for these and collaborative studies and its 2009-13 continuation comprised predominantly of members of the University of Southampton's Faculty of Medicine (M Campbell; C Kennedy; L Kim; D McCann; J Peacock; H Pimperton; S Worsfold) but also included members from other Faculties in the University of Southampton (Institute of Sound and Vibration Research UoA15; School of Psychology UoA4; Wessex Institute UoA1,2); the MRC Institute of Hearing Research; Whipps Cross Hospital; NPEU, Oxford; School of Education, Birmingham; UCL Developmental Sciences.

The Wessex Trial's experimental design and health economic analysis of benefit **[3.6]** enabled a demonstration of benefit of UNS to language at primary school age in a population-based study

that remains unique in the field. It made the argument for UNS rather than targeted screening and was crucial to decisions by the UK national screening committee and the US Preventative Service Task Force to recommend implementation of UNS nationally and federally respectively.

In a further follow-up study of the same cohort, now of secondary school age (13-18 years), data collection was completed in April 2013. This will provide unique information on the longer-term impact of UNS on reading, language, socialisation and health economic benefit in teenage years.

3. References to the research

Authors from the Faculty of Medicine of the University of Southampton are in italics.

3.1 *Wessex Universal Neonatal Hearing Screening Trial Group.* Controlled trial of universal neonatal screening for early identification of permanent childhood hearing impairment. *The Lancet 1998,* 352:1957-1964. The writing committee was comprised of *Kennedy CR, Kimm L, Campbell M*, Cafarelli-Dees D (ISVR of The UoS), Thornton ARD (MRC Southampton Hearing Research Unit).

3.2 *Kennedy CR, Kimm L*, Thornton R, Davis A. False positives in universal neonatal screening for permanent childhood hearing impairment. *The Lancet* 2000; 356:1903-04.

3.3 *Kennedy C, McCann D, Campbell MJ, Kimm L*, Thornton R. Universal newborn screening for permanent childhood hearing impairment: an 8-year follow-up of a controlled trial *The Lancet* 2005; 366:660-662.

3.4 *Kennedy CR, McCann D, Campbell MJ*, Law C, *Mullee M*, Petrou S, Watkin P, *Worsfold S, Yuen HM*, Stevenson J. Language ability after early detection of permanent childhood hearing impairment. *New England Journal of Medicine* 2006; 354:2131-41.

3.5 *McCann DC, Worsfold S, Campbell MJ*, Law CM, *Mullee M*, Petrou S, Stevenson J, Watkin P, *Yuen HM, Kennedy CR*. Reading and communication skills after early life detection of permanent hearing impairment. *Arch Dis Child* 2009; 94:293-297.

3.6 Schroeder L, *Petrou S, McCann D,* Law C, Watkin PM, *Worsfold S, Yuen HM, Kennedy CR.* The economic costs of congenital bilateral permanent childhood hearing impairment. *Pediatrics 2006; 117:1101-12.*

Grants

Dates	Award Holder	Funding Body	Title	Value
1992-1996	Kennedy	Wellcome Trust	Controlled trial of UNS for PCHI;	£407,472
2001-2005	Kennedy	Wellcome Trust	Effect of early treatment of PCHI on outcome at 8 to 9 years	£371,000
2010-2013	Kennedy	Wellcome Trust	Effect of early intervention for PCHI on outcome at secondary school age	£362,323

4. Details of the impact

The results of the Wessex Trial were used extensively in the key 1997 document commissioned by the Department of Health to review the relevant data and make recommendations on a screening programme. This review, which led directly to the implementation of UNS now in place throughout the UK, singled out the Wessex trial for its size, quality, and design; the review also specifically

recommends that providers build on the experience of this trial. In the history of the NHS Neonatal Hearing Screening Project, written by the Medical Research Council and posted on the current "Achievements & Impact" page of its website, it is one of only two clinical trials cited **[5.1]**.

The growth in the use of evidence-based medicine has led to systematic reviews of trials becoming increasing influential in formulating health policy and the Wessex trial is one of only two trials worldwide rated as "good" following a major systematic review of the evidence in 2001. The follow-up study of the effect of UNS on language is also one of only two trials rated as "good" in the 2008 update of that systematic review **[5.2]** and, of these two, only the Wessex Trial evaluated the benefit of universal, as opposed to targeted, newborn screening.

Following the Hearing Outcomes Project that documented outcomes following the Wessex Trial, Kennedy gave evidence to the US Preventative Services Task Force (USPSTF) in October 2006 both on data already published and also on data, subsequently published in 2009, documenting benefits to reading ability in the same sample **[3.6]**. This played a significant role in the USPSTF systematic review **[5.2]** that focused heavily on the benefits to language reported by Kennedy et al in The New England Journal of Medicine **[3.4]** and in the change in the linked USPSTF recommendation on UNS from 'Insufficient Evidence' in 2001 to 'Recommend' in 2008 **[5.3]**. The fact that the report by Schroeder et al **[3.6]** of a reduction in the cost of educational special support associated with early intervention and other economic impact on families and society increased the influence of Southampton's work on health policy, both in the UK and overseas, is also clear from subsequent reviews published in 2012 **[5.4, 5.5]**.

The impact of UNS for PCHI dominates all newborn screening for any disorder: the number of cases of PCHI detected by UNS in the USA in 2009 was estimated to be 5,073 accounting for 43.3% of all detected cases of the 29 conditions for which newborn screening is recommended **[5.6]**. The impact on patients in the UK began in 2001 as the Neonatal Hearing Screening Programme (NHSP) was rolled out, and continues to the present day. PCHI affects over one in 1400 babies born in England each year. In a typical week in 2012, 13,290 babies were screened in England, with 336 of these being referred for further audiological testing, and 27 identified as having PCHI **[5.7]**. Thus from 2008 to 2013, it is estimated that over five million babies in England will have been screened **[5.8]**, and over 8,000 identified with PCHI. The consequences of early treatment of PCHI on the development of neural pathways and language, family functioning, literacy, academic achievement, social–emotional well-being and employment **[5.4]** impact not only on the child and their family, but also financially on the wider society and the UK economy.

In the annual report on the NHS NHSP for 2008-09, Susan Daniels, Chief Executive of the National Deaf Children's Society **[5.9]** said this about the universal screening program: "Detecting deafness as soon in life as possible gives children the best chance of fulfilling their potential. It enables families to access early intervention and support services earlier, offers them greater choices and has a proven impact on the development of language and communication skills, as well as contributing to the closure of the attainment gap between deaf children and their peers."

Then Secretary of State for Health, Andrew Lansley, backed the program by saying in 2012: "The NHS Newborn Hearing Screening Programme … allows babies with hearing problems to receive the support they need earlier to give them the best possible start in life." **[5.10]**

The introduction of the NHS NHSP has led to a dramatic reduction in the median age of identification of PCHI. In a respected 2005 review **[5.4]**, the median age at identification was shown to fall from 60 to 10 weeks on introduction of the NHSP. The reduction for the upper quartile age was even more dramatic: from 210 to 25 weeks. These represent substantial improvements in the prospects for these babies. These changes have led to similarly dramatic reductions in the age of clinical interventions such as the fitting of hearing aids or cochlear implantation.

Similar benefits are also occurring in wider Europe, North America and other continents with associated improvements in quality of life and financial savings for society **[5.3, 5.5, 5.11]**. For

example, by 2010 UNHS had been implemented in 20 of the 32 provinces in China, where 115,000 children under the age of seven suffer from severe-to-profound deafness and where 30,000 babies are born each year with a hearing impairment. In 2009, the Chinese government set up a project to offer cochlear implants to 1,500 children aged 1-5 years over the next three years. By 2011, the government had agreed to fund implants for an additional 17,000 children over four years **[5.11]**.

In summary, the widespread introduction of UNS programmes, based on pivotal research led by Kennedy in Southampton has had an impact on improving the quality of life of many thousands of babies and their families in the UK and Worldwide.

5. Sources to corroborate the impact

5.1 http://www.mrc.ac.uk/Achievementsimpact/Storiesofimpact/Hearingscreen/index.htm (accessed 25/04/2013)

5.2 Nelson HD, Bougatsos C, Nygren P. Universal newborn hearing screening: systematic review to update the 2001 US Preventive Services Task Force Recommendation. *Pediatrics* 2008; 122:e266–76.

5.3 US Preventative Services Task Force. Universal screening for hearing loss in newborns: US Preventive Services Task Force recommendation statement. *Pediatrics* 2008; 122:143–148 [RECOMMENDATION. Screen for hearing loss in all newborn infants (B recommendation).]

5.4 Pimperton H, Kennedy C. The impact of early identification of permanent childhood hearing impairment on speech and language outcomes. *Archives of Disease in Childhood* 2012; 97:648–653.

5.5 Olusanya BO. Neonatal hearing screening and intervention in resource-limited settings: an overview. *Archives of Disease in Childhood* 2012; 97: 654-59.

5.6 Howell RR, Terry S, Tait VF et al. CDC grand rounds: newborn screening and improved outcomes. *MMWR* 2012; 61 (21) 390-93.

5.7 NHS NHSP: http://hearing.screening.nhs.uk/statistics (accessed 23/05/2013). The NHS NHSP is also a beneficiary of this work.

5.8 National Deaf Children's Society website http://www.ndcs.org.uk/family_support/audiology/newborn_hearing_screening/

5.9 2008-09 annual report: http://goo.gl/EMiW9c

5.10 Comment by former Secretary of State for Health Andrew Lansley http://hearing.screening.nhs.uk/5million

5.11 Liang Q, Mason B. Enter the dragon – China's journey to the hearing world. *Cochlear Implant International* 2013; Suppl 1:526-31.