

## Newborn hearing screening: decision time for Australia

*Nationally coordinated, universal screening will allow prospective assessment of outcomes for hearing-impaired infants*

AUSTRALIA DOES NOT DO WELL in the early detection of congenital hearing impairment. Only about 25% of infants born with hearing impairment are diagnosed by the age of 12 months, and for many children deafness remains a disability leading to severe and lasting language impairment.<sup>1</sup> The technology for newborn hearing screening has now been in regular use in many parts of the world for much of the past decade, and there is at least some persuasive evidence that very early detection helps these children achieve normal language skills.<sup>2,3</sup> This evidence is far from perfect.<sup>4</sup> Only one randomised controlled trial of detection rates with and without newborn screening has been reported, and no randomised controlled trial has yet examined outcomes of hearing screening. Nonetheless, universal newborn hearing screening has become not only possible but expected in the United States and Canada, the United Kingdom and many European countries. However, it has not yet been widely implemented in Australia. As a result, the excellent diagnostic and rehabilitative services available to all Australian children once the diagnosis of hearing impairment has been made contrasts strongly with our patchy and very incomplete ascertainment of hearing impairment in the first year of life.

The Western Australian Newborn Hearing Screening Programme is therefore an important step, as is the recent announcement that a program will commence throughout New South Wales by the end of 2002. In a report of the WA program in this issue of the Journal, Bailey et al (*page 180*)<sup>5</sup> demonstrate that a high-quality, sustainable universal newborn hearing screening program can operate in Australian birthing hospitals. It appears to be a model program, with exceptionally high coverage, high acceptability, low referral rates, and low rates of babies lost to follow-up. It exceeds most benchmarks set in 2000 by the US Joint Committee on Infant Hearing,<sup>6</sup> and is in line with the Australian National Consensus Statement on Newborn Hearing Screening.<sup>7</sup> Nonetheless, it raises a number of difficult issues.

Bailey and colleagues report that the hearing screening program has achieved more than 96% coverage in the five participating metropolitan hospitals. However, together these screened infants represent only about half Western Australia's annual births — unfortunately, the “easy” half. The program operating throughout the US State of Colorado,<sup>8</sup> one of the few approaching a true population coverage, has demonstrated that hospitals with fewer than 400 births annually do worse on average than large hospitals in terms of coverage (substantially lower) and referral rates (substantially higher). Australia is characterised by vast distances and a very large number of small hospitals. For instance, the State of Victoria has a similar birthrate to Colorado, but nearly twice as many birthing hospitals (about 110, compared with 60), most of which are small. Statewide or national newborn hearing screening therefore

poses considerable logistic and economic obstacles in Australia, and these are not necessarily surmountable.

But what are the consequences of limiting ourselves to larger hospitals? Let us assume that 50% of a State's population receives a very high quality hearing screening program which achieves 95% coverage, 90% sensitivity, 95% follow-up, and 80% compliance with early fitting of hearing aids and intervention (as some parents choose neither). This equates to less than a third of that State's hearing-impaired children benefiting from the program. If any one of these parameters is lower, then the number of children potentially benefiting falls even further. Despite individual gain, median age at diagnosis and overall outcomes for the State would improve little. If universal screening is to lead to population benefits, then universal it needs to be — despite the challenges.

A poor alternative is to screen only babies with a risk factor for deafness. Asking about the presence of a risk factor becomes the universal “screen”, followed by a targeted screening test of hearing itself. At face value, this may seem cheaper, but it is not easier and certainly detects fewer children. While almost all children with hearing loss detected in the Western Australian series so far have had a risk factor, in larger series this applies to only about 50% of babies.<sup>8,9</sup> Neonatal intensive care and special care nurseries typically contain about 30%–40% of all infants found to have moderate or greater hearing loss in newborn screening programs<sup>9,10</sup> and are relatively easily targeted, as are babies with obvious head and neck abnormalities. Other risk factors, such as family history of early hearing impairment, pose greater problems. Accurate elicitation requires skilled enquiry and mothers may not recall, or even know, that a risk factor is present until after the diagnosis is made, thus reducing sensitivity. The Victorian Infant Hearing Screening Program has recently highlighted the very low positive predictive value of family history and some other risk factors, with close to 200 babies needing to be referred to diagnose one child with hearing loss.<sup>11</sup> Low sensitivity combined with poor positive predictive values equates to spending a lot of money to miss many children. Finally, risk-factor screening raises issues of equity, as the many children with hearing impairment, but without a discernible risk factor, would be denied access to hearing screens in such a program.

A final issue is that of program sensitivity. At 0.7 per 1000, Western Australia's detection rate for children with bilateral congenital hearing impairment of more than 35 dB HL (hearing level) is lower than the usual rate of 0.9–1.0 per 1000 detected (with hearing impairment of more than 40 dB HL in the better ear).<sup>8,9</sup> Most likely, this is a chance finding reflecting the small size of the series — it “just happened” that relatively few babies with hearing impairment were born during this time period. Alternative expla-

nations include equipment problems or program insensitivity. The Western Australian program is unusual in that a baby is not referred until screening has shown three “fail” responses. This lowers the false positive rate and increases program specificity — both of which are desirable. But in screening programs rises in specificity are typically accompanied by falls in sensitivity. It may be that, in this case, the pendulum has swung too far towards specificity at the expense of sensitivity, and that, after years of worrying about excessive referral rates, they are — at last — too low.

Currently, we are not serving well the hundreds of children born each year with moderate or greater hearing impairment in Australia. Universal newborn hearing screening seems one way of improving this situation. It is not reasonable to defer implementing a program until the evidence is stronger, since stronger evidence is unlikely to be available soon. Rather than stopping us from implementing programs, this should spur us to acquire such evidence. Because Australia has not yet widely introduced newborn hearing screening, we have an unusual capacity to study these questions prospectively. National benchmarks and a minimum dataset should be established, and we should start carefully and systematically examining outcomes for hearing-impaired babies born now against which to compare gains over the coming years.

We should also keep an open mind. In 10 years’ time, Australia should be able either to guarantee continuation of

an effective program, or to move resources rapidly from a program that, despite best efforts, has proved ineffective.

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