Critical Review:
Can individuals with hearing impairment associated with Usher Syndrome benefit from a cochlear implant?

Inconsistencies in the documented age range of implantation were also present.

Although pre-implantation data was collected, the authors did not provide this information.

No statistical analyses were used.

With respect to determining the Usher etiology, the authors also did not include a screening for all known Usher syndrome type I genes in the mutation analysis. This may have helped confirm the diagnosis in more individuals. These are important factors that significantly limit the credibility and results of this study.

Based on an adaptation by the Oxford Centre for Evidence-based Medicine Levels of Evidence (March 2009) and NHMRC additional levels of evidence and grades for recommendations for developers of guidelines (June 2009), the case series with pre- and post-test study design has a level of evidence of 3. However, the level of evidence provided by this study is not very compelling due to 6-* 567):=5 /. 0 .6&6.215 &1) +/&95, as described above.

Study #2

Damen, Pennings, Snik & Mylanus (2006) conducted a retrospective non-randomized, between groups case-control to examine

Benefit Inventory (GCBI). Statistical analyses included linear regression and non-linear regression (to acquire satisfactory curve fits).

Mutation analysis for type I genes was performed on blood samples from each individual. This analysis did not include a screening for all exons of USH1 genes which can introduce a limitation to this study (as mentioned in study #1). The authors identified gene mutations in two type I genes: *MYO7A* and *CDH23*. Six individuals had pathogenic mutations in *MYO7A* and one individual demonstrated a mutation associated with the *CDH23* gene. The remaining 7 individuals did not demonstrate mutations in the genes screened in this study.

Significantly lower EHL scores were found in group 1 (in 5 of 7 individuals). The group had a mean EHL score of 84 dB HL. In group 2, the youngest individual (of 3 individuals) also had a significantly lower EHL score. The mean score for this group was 97 dB HL. The 4 individuals in group 3 did not show any significant improvement in hearing post-implantation. They had a mean score of 115 dB HL.

Linear regression analyses were used to assess relationships between age and different performance criteria post-implantation. A linear regression analysis (GCBI and age of implantation) showed that performance of a cochlear implant could have significantly increased benefit when implanted at a younger age. The authors suggested the best performance occurred when individuals were implanted within the f

simple sentences. In group 2, 5 individuals evolved from no production to at least spared words. Four of

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