"COCHLEAR IMPLANT IN SYNDROMIC DEAFNESS: REPORT OF THREE CASES"

Francesco Galletti - R.Bruno, C.De Palma, G.Gangemi, M.Vannella

Introduction: Genetic deafness affects 40% of people with hearing loss and has many causes, some of which (30% approximately) associated with additional clinical features of other organ system. Syndromes that are associated with deafness are about 400 and in some case the genes have been identified.
Objective: The objective of this retrospective study is to report the author's experience with cochlear implant in subjects with three different syndromic deafness.
Methods: Three patients with a clinical diagnosis of syndromic deafness were included. Among these patients, the first one was affected by CHARGE syndrome, the second one by Usher's syndrome and the third one by Di George's syndrome.
All the patients were implanted with multichannel cochlear implants between 2002 and 2006. Two of them were evaluated with PCVRAR questionnaire, one with TAUV test. These logopaedic tests measure pre-implant and post-implant detection, discrimination, identification, recognition and intelligible speech.
Results: Logopaedic outcomes with cochlear implant showed good perception skills in all but one case, although the results were poor compared to non-syndromic implantees.
Conclusion: In view of our results, even if the number of patients reported in this study is too small to provide definitive results, in our opinion, profound syndromic deafness correction should be carefully studied and the patients and/or the parents should receive before and post surgery appropriates information not to create false expectations.

Key words: Cochlear implant, Usher's syndrome, CHARGE syndrome, Di George's syndrome.