Objective: To assess the effects of the side of implantation (first-side vs second-side vestibular schwannoma); the presence of nonauditory sensations; the general health, expectations, and motivation of the patients; and a support group on the use of a multichannel auditory brainstem implant (ABI) in 12- to 18-year-old patients with neurofibromatosis 2.

Design: Since 1992, 21 individuals (age range, 12-18 years) who were deafened by neurofibromatosis 2 have undergone implantation with a multichannel ABI at the House Ear Institute, Los Angeles, Calif. The patients were categorized regarding side of implantation, presence of remaining hearing (in first-side implant recipients), incidence of nonauditory sensations, and ABI use or non-use. They were also rated on factors of general health, personal motivation, expectations, and family support.

Results: Nineteen (95%) of 20 teenagers tested received hearing sensations from their ABIs. Eleven teenagers used their ABIs regularly, but 8 did not. Of the non-users, 2 had good remaining hearing on the side with the second vestibular schwannoma, 2 had persistent nonauditory sensations, and 4 became program dropouts. None of the dropouts had remaining hearing, significant nonauditory sensations, or poor health; however, they generally rated poorly in terms of personal motivation, expectations, and family support. One patient with good family support returned with excellent ABI results after 4 years’ absence.

Conclusions: The multichannel ABI is an effective means of providing hearing sensations to young patients deafened by neurofibromatosis 2. Preoperative counseling regarding the importance of such factors as expectations, personal motivation, and family support is invaluable and can promote successful adaptation to the device. With patience and support, even young nonusers (including program dropouts) can become successful device users.

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side tumor removal, which, in our experience, often occurs within 2 to 3 years of the first-side surgery. Not surprisingly, the level of hearing remaining on the second side influenced the frequency of use of the ABI, and patients with good remaining hearing tended to use their implants less frequently. Many patients, however, found their ABIs increasingly useful as hearing on their second side deteriorated with further VS growth.

Another factor in ABI use and benefit is the presence of nonauditory sensations (eg, tingling, dizziness, or visual “jittering”) with activation of some electrodes. By altering stimulus parameters, these “side effects” could usually be eliminated or reduced. However, about 9% of our patients experienced significant levels of nonauditory sensations that affected their ability to use their implants.

In general, optimal ABI candidates (both young and old) have relatively good health, acceptable vision, an interest in spoken communication, acceptable anatomical status, reasonable anatomical status, reasonable expectations, high motivation, and a good support group. It has taken time for us to fully comprehend the potential impact of these factors in teenagers, and we continue to learn. Deficiency in any of these areas can impact ABI use and benefit, or even lead to nonuse of the device or to the patient dropping out of the program. The existence of chronic disease can have a serious impact on the self-esteem, behavior, and coping abilities of affected teenagers and their families. Lack of good family support may affect compliance with required follow-up visits to accommodate changes in patient responses and to optimize performance. Initially, low motivation to persist in using the ABI can prevent recipients from adjusting to their implants and from experiencing performance improvements that can continue for up to 10 years.

The focus of this retrospective review is on the more clinical aspects of ABI use in young persons with NF2. While attempting to cope with their disabilities, these individuals are also struggling to establish an identity and at the same time to “fit in.” The ABI can facilitate this process, but it requires significant cooperation and team effort. The cornerstone of a successful ABI program is careful patient preparation, and we believe that this may be especially important for young implant candidates and their families in promoting ABI benefit.

**RESULTS**

In all, 19 patients received hearing sensations from their ABIs. One individual experienced only nonauditory sensations, and one was awaiting connection to his device. Therefore, 95% (19/20) of patients tested received auditory sensations from their implants. The speech perception and psychophysical performance of these patients was similar to those that have been observed in adult recipients. Five of the teenaged ABI recipients were among the top performers with their implants.

Figure 1 shows the number of patients tested and the number of device users by age. In all, 58% (11/19) of the teenagers used their ABIs regularly. This percentage compared with a use rate of 79% in our adults with ABIs. One of the best young performers did not fit the general description of the optimum ABI candidate since he was not only deaf but also completely blind. This patient was exceptional, however, in that he was extremely motivated to make the best use of his ABI, and he had excellent family support.

**Figure 2** shows the age distribution of the 8 ABI nonusers along with data on side of implantation and presence of nonauditory sensations. Five of these individuals underwent implantation on their first VS side, with hearing remaining on their second side, and 2 of them did not use their ABIs regularly because of good remaining hearing. Figure 2 also shows that 4 nonusers reported significant levels of nonauditory sensations. As is typical, steps were taken to eliminate or reduce these sensations; however, 2 of the 4 patients continued to experience nonauditory sensations that prevented regular ABI use.

Figure 2 also includes 4 ABI recipients (aged 12, 15, 16, and 18 years) who unfortunately became nonusers by virtue of dropping out of the program, a decision that we accepted and respected. None of the dropouts had useful hearing remaining on the second side, nor did they have problems with significant nonauditory sensations.

**Figure 3** shows the ratings for the dropouts on possible contributing factors: family support, general health,
factors in multichannel ABI use and benefit in patients ranging in age from 12 to 18 years. As in adults, the multichannel ABI was found to be effective in providing hearing sensations to the majority (95%) of our young patients, although 8 teenagers (42%), including 4 program dropouts, did not use their devices.

Some factors that contributed to nonuse, such as remaining hearing in first-side implant recipients or persistent nonauditory sensations, were beyond our control. While neither of these 2 factors was a major issue in the program dropouts, the dropouts rated poorly on family support, expectations, and motivation to make the best use of ABI sound. These factors are amenable to clinical intervention, however.

In the case of the 18-year-old patient who returned to the program, of major significance was his father’s steadfast encouragement to become reinvolved. After a few weeks of ABI use, this young patient subsequently wrote that he could not believe how far he had come with his ABI, a common observation among ABI recipients. He also noted that “cold feet” had led him to live life deaf for 4 years, but that even though ABI sound initially was confusing, through daily use he quickly adjusted and found it beneficial. This comment is also a common observation by ABI users.

In retrospect, we noted that this young man was angry and argumentative during preoperative counseling, which was understandable in view of the difficulties he was facing. However, we may have underestimated the eventual significance of these potential “red flags.” Despite our efforts to prepare him, he was clearly very disappointed with his first experience with ABI sound, which prompted his sudden (and lengthy) departure. This experience has served as a valuable lesson to us in the preparation and management of subsequent young ABI candidates and their families.

In general, ABI recipients (young and old) must accept that sounds initially will not be at all “normal,” that significant levels of sound-only speech recognition are uncommon, and that improving with an ABI takes time and experience. In counseling our teenaged candidates, we now include a frank discussion of our experiences with younger device recipients, emphasizing the importance of such factors as good family support and high motivation for regular use of the device. We also emphasize the value of adopting a positive, mature, and responsible attitude in achieving long-term benefit and coping with NF2. We also have advised families regarding the potential contribution of psychological counseling. These have been very positive steps in the treatment of our young patients with NF2.

In conclusion, appropriate preoperative counseling regarding the impact of such factors as expectations, personal motivation, and family support is invaluable in helping to prepare teenaged ABI candidates and their families. The goal is to raise awareness of potential obstacles to successful adaptation to (and use of) the implant. We also conclude that with patience and support, some young nonusers of the ABI can become successful device users in the long term.

This retrospective report reviewed the effects of several factors in multichannel ABI use and benefit in patients.
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REFERENCES


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