Brainstem Electric Response Audiometry (BERA)in the Evaluation of Hearing Loss in Infants with Cleft Palate

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It has been estimated that one child in 500 to 750 live births is born with a cleft palate (Rood & Stool,1981). A high incidence of middle ear pathology is known to be associated with this problem (Bluestone, 1971). It is generally accepted that this population suffers from Eustachian tube dysfunction which leads in turn to the development of middle ear effusions and conductive hearing loss. The various underlying causes that have been postulated are related either to an anatomic malformation of the palate and tube or a functional deficiency of their musculature (Bluestone, 1971; Rood & Stool, 1981).

Bluestone (1971) reviewed the literature and found estimates ranging from 40% to 80% of cleft palate adults and children with abnormal middle ears. Paradise, Bluestone, and Felder (1969) and Stool and Randall (1968) used otoscopy and tympanometry, and both groups found evidence of middle ear effusions in virtually all of the cleft palate infants they examined. Gerber and Mencher (1978) recommended that infants with cleft palate be placed at risk for middle ear effusions with an estimated incidence of 70-100% of cases.

Bluestone's review of the literature also examined the prevalence of hearing loss associated with this problem in older children or adult patients. The estimates ranged from 0 to 90 %, and Bluestone concluded that one should expect some degree of hearing loss in at least 50% of cases.

The use of Brainstem Electric Response Audiometry (BERA) now allows us to investigate the auditory function of infants with cleft palate . Gould (1980) found that 8 of 10 infants with cleft palate showed evidence of peripheral hearing loss through the use of evoked potentials. More recently Helias, Chobaut, Mourot, and Lafon (1988) published results showing that 19 of 23 patients, or 83%, had electrophysiologic responses consistent with conductive hearing loss when tested before palate repair and before 12 months of age.

The implications of such early auditory deprivation for language learning have been debated at length in the literature. Reviews of the various aspects of this complicated question have been done by Ventry (1980), Menyuk (1986), Brooks (1980), and Feagans, Blood, and Tubman (1988). These reviews have shown that, although there is a great deal of data which suggests the detrimental effects of early hearing loss or otitis media, many of the studies have been flawed by poor validity. Nevertheless, it is difficult for the professional to ignore the possibility of a link between the deficits in speech and language development which the cleft palate child is know to exhibit (Bzoch, 1965; Mustain, 1979; Pannbacker, 1971) and the auditory status in infancy.

The purpose of this study was to review the results of testing of a group of infants who presented in our clinic with cleft palate and to document the auditory brainstem response (ABR) thresholds of these patients. This would allow us to evaluate the need for routine audiological follow-up.

Method

Children who are born with a cleft palate in the Ottawa area are systematically referred to the Children's Hospital of Eastern Ontario and followed by a multidisciplinary team that includes ENT, plastic surgery, dentistry, nursing, social work, speech-language pathology and audiology. They undergo BERA initially at three months of age as part of a larger screening program at this centre, which has been described by Durieux-Smith, Picton, Edwards, MacMurray, and Goodman (1987). For this study, we reviewed the charts and results of BERA for all children with cleft palate referred between January, 1984 and December, 1987.

Each baby was tested in a sound attenuated chamber while sleeping naturally or very quietly awake. Disc electrodes were attached to the forehead at the midline and to the mastoids. Figure 1 lists some aspects of the test protocol. Interelectrode impedance was maintained at 5K ohms or less. Activity between the forehead and mastoid ipsilateral to stimulation was amplified, filtered between 25 and 3000 Hz, and averaged over a time base of 15 msec.

One hundred µsec rarefaction clicks were presented monaurally through a TDH-49 earphone mounted in a MX41/AR

Figure 1. Details of the test protocol (Edwards & Durieux-Smith, 1985).

Recording electrodes	: F_z-M_i
Interelectrode Impedance	: <5 k Ω
Average sweep	: 15 ms
Filters	: 25-3000 Hz
Click nHL	: 40 dB peak SPL
Click Polarity	: rarefaction
Replications	: 1
Intensity and Presentation Rate	: 30 dB nHL at 61/sec (increase in 10 dB steps if no response) 70 dB nHL at 11/sec
Sum	: 4000 at 61/sec 2000 at 11/sec

cushion. The clicks were presented initially at 30 dBnHL at a presentation rate of 61/second, and two replicate averages of 4000 responses each were obtained. A response at 30 dBnHL was considered to demonstrate normal peripheral auditory sensitivity. If no response was recorded at 30 dBnHL, the intensity was increased in 10 dB steps up to a maximum of 95 dBnHL until threshold was reached. Clicks also were presented at 70 dBnHL (or higher in cases of hearing loss) at a rate of 11/second, and replicate averages of 2000 responses were recorded in order to assess Wave I and the Wave I-V interval. A complete set of results could not be obtained in all cases because some of the babies awoke during testing. Figure 2 shows typical ABR recordings obtained with this population.

Those children with elevated thresholds on the initial BERA returned for a retest and repeat otoscopic examination three months later. If an elevated threshold persisted at this first and subsequent visits, myringotomy and tube insertion were recommended by the otolaryngologist.

For some of the children, results of behavioural audiometry were available. Attempts at visual reinforcement or conditioned orienting response audiometry were usually started when the child was at least twelve months of age. Generally, results of conditioned play audiometry under earphones were not reported for children under thirty months. Tympanometry was carried out only after six months of age. Paradise, Smith, and Bluestone (1976) and Schwartz and Schwartz (1978) reported poor reliability for this measure with younger infants. Otomicroscopy was used by the otolaryngologist on the team for the first evaluation at three months of age and at all later test sessions. It should be noted that due to the retrospective nature of this work, a strict protocol was not followed in all cases.

Results

Fifty children, including 25 male and 25 female patients, with cleft palate or cleft lip and palate were tested with BERA. For

Figure 2. Examples of a normal ABR tracing, a conductive hearing loss ABR, and a sensorineural loss ABR in infant patients. (Picton & Durieux-Smith, 1988). Positivity at the vertex is indicated by a downward deflection.

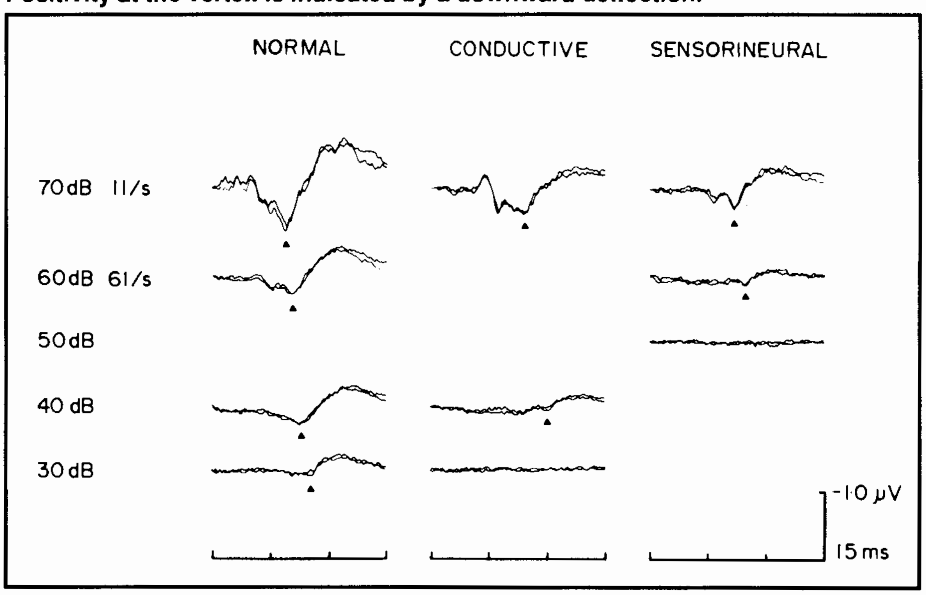
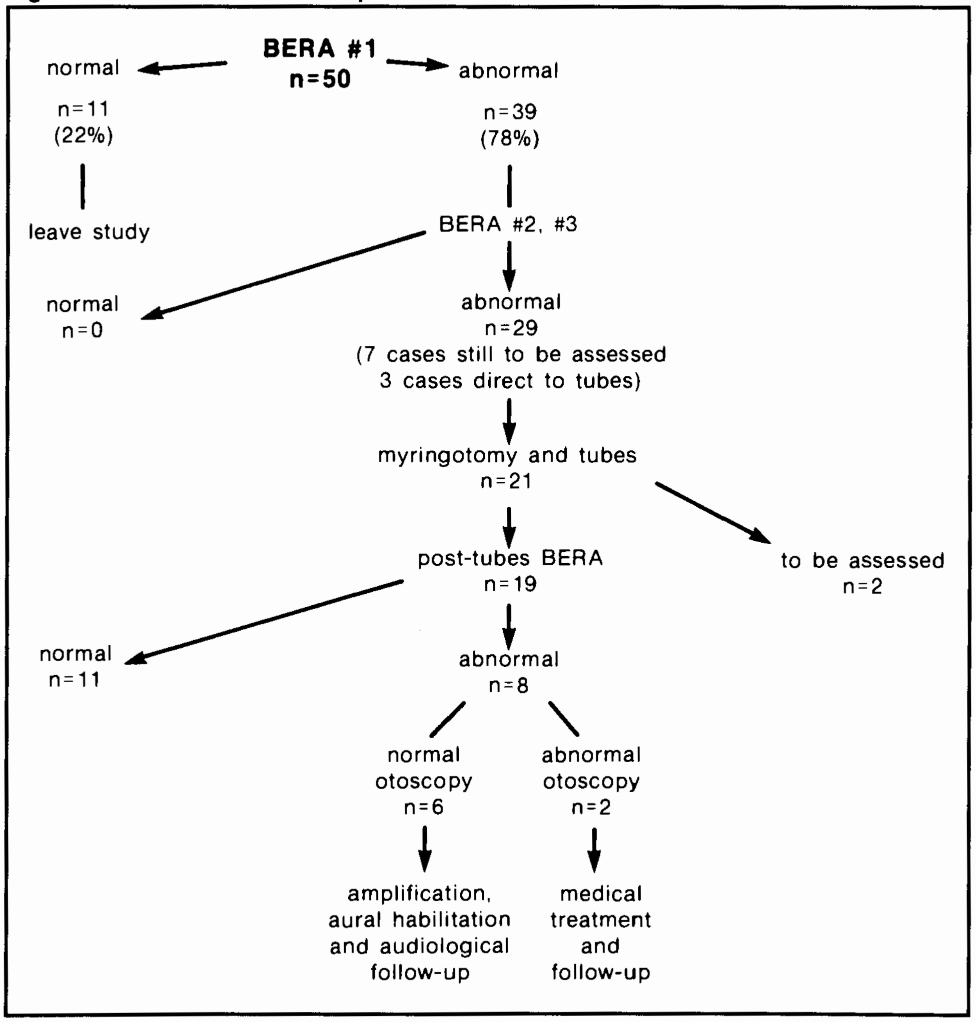


Figure 3. Flow chart of the test protocol and results.



some of the children, information was available on two, three, and as many as four tests. A total of 109 tests were examined.

Figure 3 shows that of the 50 babies tested, 39 (78%) were found initially to have ABR thresholds of greater than 30 dBnHL in at least one ear. Of these, 35 showed a bilateral elevation in threshold. Of the 39 babies with abnormal results, 29 went on to be retested, 3 underwent myringotomy and tubes without further verification, and 7 have yet be re-assessed.

Twenty-one children underwent tube insertion after a second or third BERA evaluation. Of these, only 11 were found to have a normal threshold within two months post-operatively. Two cases were found to have otorrhea, and further medical management was required. Two cases have yet to be assessed. No child who began with an elevated threshold resolved to normal levels spontaneously or before medical intervention at least during the first year of life. One child eventually showed normal ABR thresholds, but only at fifteen months of age.

Six cases without otorrhea failed to resolve for the first BERA following tube insertion. Three of these six cases have since been found to be sensorineural in nature on the basis of air and bone conduction audiometry and impedance audiometry. One of these is a unilateral hearing loss. The other two are bilateral and have been fitted with hearing aids and have entered into a program of aural habilitation.

Three other children who were found to have a persistent hearing loss following tube insertion were fitted with aids and began therapy. Behavioural evaluations showed that these children eventually resolved to normal hearing some months later. In two of the cases, improvement was noted only after the palate repair. The time taken before normal hearing was documented in these children was seven to eleven months.

Previous research in our department had indicated that BERA testing was best carried out at three months corrected age (Durieux-Smith et al., 1987). We questioned whether this recommended age also was the ideal for the cleft palate chil-

Figure 4. Difference in percentages of normal ears when testing took place at less than three months of age or later.

	<3 months at BERA #1	≥months at BERA #1
normal ears	30.3%	26.7%
abnormal ears	69.7%	73.3%

x^2 is not significant

Figure 5. Comparison of mean thresholds for children tested at various ages.

Age at test	N=ears	Mean Threshold (dB nHL)
≈3 months	48	49.79
≈6 months		46.67
≈6 months	7	50.00
≈9 months		42.87

No significant difference when test results for each child compared at different ages

dren. An earlier review of a subset of this group done by Tropper, Moran, Odell, and Durieux-Smith (1988) indicated that the percentage of normal ABR thresholds obtained when the children were less than four months of age was higher than for the group who were tested at a later age. Our concern was that children who passed the electrophysiologic screening at three months of age might later develop a significant hearing loss which would go undetected.

It was found through examination of the results of the larger group of children that this concern was not warranted. Figure 4 shows the percentage of ears of children who passed or failed at the criterion level of 30 dBnHL. The proportion of ears with normal thresholds when tested at less than three months was 30.3%. The proportion of ears with normal thresholds tested at a later age was 26.7%. The difference in these two groups did not reach statistical significance at the 0.05 level ($\chi^2 = 0.017$).

Figure 5 shows the mean thresholds for two separate groups, those whose members had been tested twice, once at three months and again at six, and those whose members had had tests at six and nine months of age. With this second method of analysis, no significant difference was found between the results for the younger and older age group in either case (3,6 months: F=1.31, df=47,1; 6,9 months: F=0.91, df=6,1). We were not able to compare results for children across several ages as so few of the patients had been tested more than twice.

Discussion

The review of these cases has highlighted several points. First, 78% of the 50 children with cleft palate who were tested before medical intervention showed an abnormal threshold. This figure is consistent with estimates of middle ear pathology and hearing loss in older cleft palate patients (Bluestone,1971). The rate of detection of elevated response remained high whether the children were tested at three months of age or at a later time during the first year of life.

We might have expected an even higher failure rate given the reports of almost universal occurrence of middle ear effusions in infants with cleft palate (Paradise et al., 1969; Stool & Randall, 1968). It should be noted that the click stimulus that was used in this study best correlates with audiometric thresholds at 2000 Hz and 4000 Hz (Coats & Martin, 1977; Gorga, Worthington, Reiland, Beauchaine, & Goldgar, 1985). The children in whom the middle ear pathology had caused only a low frequency hearing loss would not have been detected. The results, therefore, may represent an underestimate of auditory dysfunction for this population.

The incidence of bilateral sensorineural hearing loss was 2 out of 50 or 4%. This yield is slightly higher than that found through the screening of all NICU graduates at our centre, where 1.5% had a significant loss (Durieux-Smith et al., 1987). It has been suggested that the use of a risk factor is justifiable when it increases the detectability of a condition by at least ten times that found in the general population. (Joint Committee on Infant Hearing, 1982; Richards & Roberts, 1967). Our results indicate that the presence of cleft palate meets this requirement given an incidence of congenital hearing loss in the general population of one in 750 (Feinmesser & Tell, 1982).

In addition, a much greater proportion of the children were affected bilaterally than unilaterally. The resulting auditory deprivation raises concerns about the potential for delay in speech and language development and underlines the need for early assessment and vigilant follow-up.

Tube insertion did result in improvement in threshold in the majority of cases. However, we noted three children in whom there was an unexpected lag between treatment and the time when hearing was documented to be within normal limits. The reason for this delay is not evident. The tympanostomy tubes seemed insufficient to clear the middle ear effusions in these cases. The restoration of normal middle ear function may be seen only after more normal Eustachian tube function is achieved through cleft palate repair. The clinical audiologist must be aware of the possibility of delayed improvement in threshold in this population and plan counselling and management strategies accordingly.

Summary

The results of this study should alert the audiologist to the high incidence of elevated ABR thresholds in infants with cleft palate. This high incidence of auditory dysfunction was noted when the children were tested at three months of age or at some time later, but still before their palate repair. Given the fluctuating or recurrent nature of middle ear effusions we feel that structured audiological follow-up and careful parent education are recommended in order to ensure prompt management of hearing loss throughout the first years of life in this population.

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