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Abstract Selection

Facial neuropathy due to axonal degeneration and microvasculitis following gamma knife surgery for vestibular schwannoma: a histological analysis. Case report Watanabe, T., Saito, N., Hirato, J., Shimaguchi, H., Fujimaki, H., Sasaki, T. Department of Neurosurgery, Gunma University School of Medicine, Maebashi, Gunma, Japan. nswata@med.gunma-u.ac.jp. *Journal of Neurosurgery* (2003), November, Vol. 99 (5), pp. 916–20, ISSN: 0022-3085
Complete facial palsy (House-Brackmann Grade VI) developed in a 63-year-old man with a vestibular schwannoma 25 months after he had undergone two gamma knife surgeries performed 33 months apart and involving a cumulative dose of 24 Gy directed to the tumour margin at the 50 per cent isodose line. Magnetic resonance imaging demonstrated tumour enlargement with central nonenhancement, which initially had been recognized 21 months after the second radiosurgery. Microsurgery was performed to achieve total removal of the tumour. Histological and immunohistochemical examinations of the facial nerve specimen removed from the edge of the tumour revealed a loss of axons, proliferation of Schwann cells, and microvasculitis. In this case, microvasculitis and axonal degeneration were probably the major causes of the radiation-induced facial neuropathy.

Evolution of a cochlear schwannoma on clinical and neuroimaging studies. Case report. Khurana, V. G., Link, M. J., Driscoll, C. L. W., Beatty, C. W. Department of Neurologic Surgery, Mayo Clinic, Rochester, Minnesota 55905, USA. *Journal of Neurosurgery* (2003) October, Vol. 99 (4) pp. 779–82, ISSN: 0022-3085.
The authors report on a patient with a rare schwannoma that arose from the cochlear division of the vestibulocochlear nerve. Distinctively, the lesion appeared to arise from the cochlea itself and was monitored with clinical and neuroimaging studies for 12 years before it was diagnosed and treated. The atypical occurrence of schwannomas of the vestibulocochlear nerve originating in the inner ear structures underscores the high level of clinical suspicion required for the diagnosis of these lesions in patients presenting with persistent auditory and vestibular symptoms.

VoiSS: a patient-derived Voice Symptom Scale. Deary, I. J., Wilson, J. A., Carding, P. N., MacKenzie, K. Department of Psychology, University of Edinburgh, 7 George Square, Edinburgh EH8 9JZ, UK. i.deary@ed.ac.uk. *Journal of Psychosomatic Research* (2003) May, Vol. 54 (5), pp. 483–9, ISSN: 0022-3999.
OBJECTIVE; Many voice-rating tools are either physician-derived, disease-specific measures or they merely combine general quality-of-life domains with vocal symptoms. The aim of this series of studies was to devise and validate a patient-derived inventory of voice symptoms for use as a sensitive assessment tool of (i) baseline pathology and (ii) response to change in adult dysphonia clinics. METHOD: Three stages in the development of the instrument are described. First, an initial exploratory, open-ended questionnaire study was used to compile a prototype list of voice complaints (*Clin Otolaryngol* 1997;**22**:37). Second, the prototype list was administered to 168 subjects with dysphonia and underwent principal components analysis. Qualitatively, it was also assessed at this stage for its ability to capture voice-related impairment, disability and handicap. Third, a modified 44-item scale was administered to 180 new subjects. RESULTS: The symptoms were highly endorsed. Principal components analysis with oblique rotation yielded a Voice Symptom Scale (VoiSS); 43 of the items comprise a 'general voice pathology' scale. More specifically, five oblique components provided assessments of: 'communication problems,' 'throat infections,' 'psychosocial distress,' 'voice sound and variability' and 'phlegm.' CONCLUSION: The VoiSS is simple for patients to complete and easy to score. It is sensitive enough to reflect the wide range of communication, physical symptoms and emotional responses implicit in adult dysphonia.

Do the genes that cause otosclerosis reduce susceptibility to otitis media? Manolidis, S., Alford, R. L., Smith, R. J. H., Ball, C., Manolidis, L. The Bobby R. Alford Department of Otorhinolaryngology and Communicative Sciences, Baylor College of Medicine, Houston, Texas 77030, USA. spirosm@bmc.tmc.edu. *Otology & Neurotology* 2003; November, Vol. 24 (6), pp. 868–71, ISSN: 1531-7129
HYPOTHESIS: This genetic factors that cause otosclerosis reduce susceptibility to otitis media. BACKGROUND: Susceptibility to some infectious diseases is modulated by host genetic factors. Genes that reduce the morbidity and mortality of infectious diseases may confer a selective advantage and achieve high-frequency in at-risk populations. Acute otitis media in the pre-antibiotic era frequently led to complications with high morbidity and mortality. The long-term sequela of acute otitis media is chronic otitis media. Chronic otitis media has a prevalence of 0.5 to two per cent in the Caucasian population. Clinical otosclerosis occurs in one per cent of Caucasians. Histologic otosclerosis occurs in 10 per cent of Caucasians. MATERIALS AND METHODS: Retrospective analysis of 2,362 subjects with surgically confirmed otosclerosis. Subjects' medical records were reviewed for evidence of chronic otitis media. The incidence of chronic otitis media in the general population was compared with the subject population. RESULTS: Of the 2,362 subjects with surgically confirmed otosclerosis, one subject with evidence of chronic otitis media was identified. The incidence of chronic otitis media in the subject population is 0.04 per cent ($p<0.0001$). CONCLUSIONS: The incidence of chronic otitis media in subjects with a surgically confirmed diagnosis of otosclerosis is significantly lower than expected. The genes that cause otosclerosis may confer resistance to the pathogens that cause chronic otitis media and/or acute otitis media. The mechanism of resistance is unknown.

Fluoroscopically assisted cochlear implantation. Fishman, A. J., Roland, J. T. Jr., Alexiades, G., Mierzwinski, J., Cohen, N. L. Department of Otolaryngology, New York University Medical Center, New York 10016, USA. andrew.fishman@med.nyu.edu. *Otology & Neurotology* (2003), November, Vol. 24 (6), pp. 882–6, ISSN: 1531-7129.
HYPOTHESIS: Real-time intraoperative fluoroscopy is a useful adjunct to cochlear implantation in selected cases. The advantages include the avoidance of complications such as extracochlear array placement, intrameatal array insertion, and avoidance of significant bending or kinking. This is particularly useful when implanting a severely abnormal cochlea. BACKGROUND: The technique was initially developed for laboratory study of electrode prototypes in cadaver temporal bones to evaluate insertion dynamics and mechanisms of intracochlear trauma. The technique was subsequently adapted for use in live surgeries. METHODS: Live surgeries were performed using fluoroscopic guidance on nine patients. RESULTS: Five patients were implanted with the Nucleus 24 RCS during preclinical trials. Two patients with severe cochlear malformations were implanted with a Nucleus C124M straight array. Two patients with severe cochlear ossification were implanted with the Nucleus C124 double array. Appropriate insertions were achieved without electrode damage in all cases. CONCLUSIONS: Intraoperative fluoroscopy is a useful adjunct to cochlear implantation, which can be performed with minimum risk to the patient and operating room staff if the outlined precautions are taken. Intraoperative fluoroscopy is indicated in cases where the intracochlear behaviour of the electrode array cannot be predicted, a condition encountered when implanting new elec-

trode designs, cases with severely malformed inner ears, or cases of severe intraluminal obstruction requiring a double-array insertion.

Otoplasty for prominent ears in children. The technique adopted in the Portmann Institute. Mayaleh, H. A., Khalil, H. S., Portmann, D., Negrevergne, M. Institut Georges Portmann, 114 avenue d'Ares, 33074 Bordeaux, France. *Revue de Laryngologie – Otologie – Rhinologie* (2003), Vol. 124 (2), pp. 135–6, ISSN: 0035-1334.
The Otoplasty technique adopted in the Portmann Institute is a simple rapid technique that maintains the natural contours of the auricle with minimal risk of infection. The technique involves excision of a large ellipse of skin from the mastoid surface of the auricle and reflection of remaining skin to the edge of the helix. The subcutaneous and muscular tissues on the mastoid bone are excised and the mastoid surface of the auricular cartilage scored with monopolar diathermy. After haemostasis, the wound is closed using continuous long-term absorbable sutures. A dressing and bandage are applied and the child is monitored for 10 days. A head bandage is applied at night for one month with use of a sunscreen cream on the scar at daytime.

Hypopharyngeal pharyngoplasty in the management of pharyngeal paralysis: a new procedure. Mok, P., Woo, P., Schaefer, M. J. Grabscheid Voice Center, Department of Otolaryngology – Head and Neck Surgery, Mount Sinai Medical Center, New York, New York 10129, USA. *The Annals of Otology, Rhinology and Laryngology* (2003), October, Vol. 112 (10), pp. 844–52, ISSN: 0003-4894.
Dysphagia after a high vagal nerve injury may be associated with a patulous hypopharynx that serves as a reservoir for pharyngeal secretions, contributing to primary or secondary aspiration. We describe a new hypopharyngeal pharyngoplasty procedure for the paralyzed pharynx to improve swallow. The paralyzed pyriform sinus is resected to remove insensate and redundant mucosa. The inferior constrictor muscle is then advanced anterior to the oblique line of the thyroid cartilage to improve pharyngeal tone and prevent pharyngeal dilatation. The surgery is performed in conjunction with medialization laryngoplasty and arytenoid adduction. The utility of this procedure is reviewed retrospectively in eight patients. They were evaluated by clinical evaluation, fiberoptic endoscopic evaluation of swallow, and modified barium swallow study. All had significant preoperative dysphagia. Three patients were gastrostomy tube-dependent. After operation, all patients had subjective and objective improvements in swallow and progressed to peroral feeding. There were no operative complications. We conclude that hypopharyngeal pharyngoplasty diminishes pyriform sinus pooling and improves pharyngeal transit. Dysphagia patients with unilateral pharyngeal paralysis secondary to cranial nerve palsies may benefit from this new procedure.

Benign paroxysmal positional vertigo in older women may be related to osteoporosis and osteopenia. Vibert, D., Kompis, M., Haeusler, R. Department of Otorhinolaryngology – Head and Neck Surgery, Inselspital, University of Berne, Berne, Switzerland. *The Annals of Otology, Rhinology and Laryngology* (2003), October, Vol. 112 (10), pp. 885–9, ISSN: 0003-4894.
Benign paroxysmal positional vertigo (BPPV), so-called canalolithiasis and cupulolithiasis, usually occurs after head trauma or viral vestibular neuritis. In many cases, the cause remains obscure, and it often affects women more than 50 years old. The goal of this work was to study a possible relationship between BPPV and osteopenia or osteoporosis. Thirty-two women, whose ages ranged from 50 to 85 years (median age, 69 years), who had BPPV and were free of any other otoneurologic history, were selected. The diagnosis of osteopenia or osteoporosis was confirmed by a bone mineral density measurement made with dual X-ray absorptiometry of spine and hip (T-score). The BPPV was unilateral in 26 patients and bilateral in six patients. Our results showed osteopenia or osteoporosis in 24 of the 32 patients (75 per cent) with BPPV. The T-scores were compared in three age groups to those of 83 healthy women. The patients were BPPV had a significantly lower ($p<.026$) T-score in all groups. Possible pathophysiological mechanisms are discussed to explain the apparent correlation between BPPV and osteopenia or osteoporosis.

Regeneration of the vocal fold using autologous mesenchymal stem cells. Kanemaru, S. I., Nakamura, T., Omori, K., Kojima, H., Magrufov, A., Hiratsuka, Y., Hirano, S., Ito, J., Shimizu, Y. Department of Otolaryngology – Head and Neck Surgery, Kyoto University Faculty of Medicine, Kyoto, Japan. *The Annals of Otology, Rhinology and Laryngology* (2003), November, Vol. 112 (11), pp. 915–20, ISSN: 0003-4894.
The aim of this study was to regenerate the injured vocal fold by means of selective cultured autologous mesenchymal stem cells (MSCs). Eight adult beagle dogs were used for this experiment. Selective incubation of MSCs from bone marrow was done. These MSCs were submitted to three-dimensional incubation in one per cent hydrochloric acid atelocollagen. Three-dimensional incubated MSCs were injected into the left vocal fold, and atelocollagen only was injected into the right vocal fold of the same dog as a control. Four days after injection, the posterior parts of the vocal folds were incised. The regeneration of the vocal fold was estimated by morphological and histologic evaluations. Our results showed at three-dimensional incubated MSCs were useful in the regeneration of the injured vocal fold. This study shows that damaged tissues such as an injured vocal fold would be able to be regenerated by tissue engineering.

Efferent system degeneration in the human temporal bone. Gacek, R. R. Division of Otolaryngology – Head and Neck Surgery, Department of Surgery, University of South Alabama College of Medicine, Mobile, Alabama 36688-0002, USA. *The Annals of Otology, Rhinology and Laryngology* (2003), November, Vol. 112 (11), pp. 947–54, ISSN: 0003-4894.
Sense organ deposits have been described in temporal bones from patients with vestibular neuronitis, Ménière's disease, and benign paroxysmal positional vertigo that are not found in a comparable series of temporal bones without vestibulopathy. Because the recurrent vestibulopathies are caused by vestibular ganglionitis and the vestibulocochlear anastomosis was degenerated in these temporal bones, the deposits may represent the end buds of regenerating efferent axons injured in passage through the vestibular ganglion. Such neural buds have been described with transmission electron microscopy in animals after vestibular nerve transection and in a human temporal bone with endolymphatic hydrops. The buds may be visible by light microscopy, because their size is comparable to that of hair cell nuclei and they stain blue with haematoxylin because of their nucleic acid content. The variable location and size of these deposits (buds) in the labyrinthine sense organs is described to aid in the recognition of efferent system injury in human temporal bones.

Stroboscopic observation of vocal fold vibration with the videoendoscope. Sato, K., Umeno, H., Nakashima, T. Department of Otolaryngology – Head and Neck Surgery, Kurume University School of Medicine, Kurume, Japan. *The Annals of Otology, Rhinology and Laryngology* (2003), November, Vol. 112 (11), pp. 965–70, ISSN: 0003-4894.
Asahi Optical Co, Ltd (Toyko, Japan), has manufactured a trial thin videoendoscope with an instantaneous RGB (red-green-blue) system. We have developed a new laryngostroboscopic system using this videoendoscope. In this article, we report the performance of the clinical trial of this new and useful videoendoscopic system for stroboscopy, and compare it to conventional stroboscopy using a flexible fibrescope or rigid endoscope. This new thin videoendoscope has the following characteristics. A small charge-coupled device (CCD) chip is built into the lip of this endoscope, and an instantaneous RGB system is used. The outer diameters of its tip (4.1 mm) and the insertion tube (3.7 mm) of the videoendoscope are relatively small. The videoendoscope system is compact. Stroboscopic observation was conducted with this videoendoscope in conjunction with a laryngostroboscope. Compared to stroboscopy with a conventional flexible fiberscope or rigid endoscope, this stroboscopic system has several clear advantages. It presents clear dynamic colour images on a colour video monitor and provides excellent resolution and recording, thus yielding high diagnostic accuracy. The diameter of the videoendoscope is relatively small and results in less discomfort for patients, even for children. The videoendoscope allowed the doctors to perform pernasal endoscopy. Therefore, there is less limitation in the range of subjects for examination. Patients are able to phonate holding a normal head position, and are thereby able to produce varying vocal sounds in

the habitual or normal manner during examination. The new stroboscopy system using a videnendoscope is a very useful examination tool that has the advantages of both a conventional flexible fibrescope and a rigid endoscope.

Histopathology of the inner ear in unoperated acoustic neuroma. Mahmud, M. R., Khan, A. M., Nadol, J. B. Jr. Department of Otolaryngology, National University of Malaysia, Kuala Lumpur, Malaysia. *The Annals of Otology, Rhinology and Laryngology* (2003), November, Vol. 112 (11), pp. 979–86, ISSN: 0003-4894.
Although hearing loss is the most common presenting symptom in patients with acoustic neuroma, the pathophysiology of hearing loss associated with acoustic neuroma is unknown. Although primary dysfunction of the auditory nerve is intuitively logical, available histopathologic and clinical data suggest that although neural degeneration is common, it alone does not adequately account for hearing loss in many cases. The purpose of this study was to evaluate 11 cases of unoperated unilateral acoustic neuromas. Temporal bones were identified by means of a search mechanism provided by the National Temporal Bone, Hearing and Balance Pathology Resource Registry and were prepared for light microscopy by standard techniques. Quantification of spiral ganglion cells, hair cells, stria vascularis, and spiral ligament was accomplished for each specimen. In addition, the maximum diameter and volume of each tumour were calculated from histopathologic sections. Increasing tumour size did predict a reduced spiral ganglion count. However, although there was a tendency for decreasing spiral ganglion cell count and for increasing tumour size to predict a higher pure tone average and lower speech discrimination score, these correlations did not reach statistical significance. In tumour ears in which the speech discrimination score was 50 per cent or less, there was always significant degeneration of other structures of the inner ear in addition to neurons, including hair cells, the stria vascularis, and the spiral ligament. Endolymphatic hydrops and eosinophilic precipitate in the perilymphatic spaces were found in two of three such cases. It is concluded that acoustic neuromas appear to cause hearing loss, not only by causing degeneration of the auditory nerve, but also by inducing degenerative changes in the inner ear. It is hypothesized that the proteinaceous material seen histologically may represent the products of up-regulated genes in acoustic neuroma, some of which may interfere with normal cochlear function.

Selective denervation: reinnervation for the control of adductor spasmodic dysphonia. Allegretto, M., Morrison, M., Rammage, L., Lau, D. P. Department of Surgery (Otolaryngology), University of Alberta, Edmonton, AB. *The Journal of Otolaryngology* (2003), June, Vol. 32 (3), pp. 185–9, ISSN: 0381-6605.
OBJECTIVES: The objective of this study was to evaluate the efficacy of a new surgical procedure for adductor spasmodic dysphonia (AddSD). This surgery involves the bilateral selective division of the adductor branches of the recurrent laryngeal nerves with immediate reinnervation of the distal nerve trunks with branches of the ansa cervicalis (selective denervation-reinnervation). METHODS: Our first six patients to undergo this procedure were enrolled in the study. All patients suffered from AddSD and had previously received botulinum toxin A (Botox, Allergen, Markham, ON) therapy. Patients were recorded preoperatively and all underwent the same surgical procedure performed by the same lead surgeon. All patients were surveyed postoperatively and then re-recorded. Expert and untrained judges undertook perceptual evaluation of voice quality. Voice samples were also objectively evaluated for aphonic voice breaks. RESULTS: No major surgical complications were noted. Patient satisfaction was excellent, and five of the six patients no longer require botulinum toxin therapy. In five of the six patients, the majority of untrained and expert listeners perceived the post-operative voice to be superior. Objectively, the rate of aphonic voice breaks was also reduced in five of the six patients.

Intratympanic steroids: do they acutely improve hearing in cases of cochlear hydrops? Hillman, T. M., Arriaga, M. A., Chen, D. A. Pittsburgh Ear Associates, Pennsylvania 15212-4746, USA. *The Laryngoscope* (2003), November, Vol. 113 (11), pp. 1903–7, ISSN: 0023-852X.
OBJECTIVE: To study the acute effects on hearing of intratympanic dexamethasone in patients with cochlea hydrops. STUDY DESIGN: Retrospective review. METHODS: Patients who met established criteria for the diagnosis of Ménière's disease or had a history of fluctuating hearing loss and met hearing loss criteria for Ménière's disease, indicating cochlear hydrops, underwent a series of one to three intratympanic injections of dexamethasone in the affected ear. Follow-up audiograms were obtained one week after each injection and, in many patients, several months after injection. RESULTS: Fifty patients met inclusion criteria and were studied. Using the American Academy of Otolaryngology – Head and Neck Surgery reporting guidelines, hearing improved acutely in 20 of the 50 patients (40 per cent), was worse in two (four per cent), and did not change in 28 (56 per cent). For those who improved, the average decrease in threshold was 14.2 dB. Whether the patient had typical Ménière's disease or cochlear hydrops did not affect the response to therapy. There were no significant complications from the injections. CONCLUSIONS: Intratympanic administration of dexamethasone may acutely affect sensorineural hearing loss associated with endolymphatic hydrops. A prospective, controlled study is required.

The H+/K+-ATPase (proton) pump is expressed in human laryngeal submucosal glands. Altman, K. W., Haines, G. K. 3rd, Hammer, N. D., Radosevich, J. A. *The Laryngoscope* (2003), November, Vol. 113 (11), pp. 1927–30, ISSN: 0023-852X
OBJECTIVES/HYPOTHESIS: Diagnosis and treatment of gastroesophageal and laryngopharyngeal reflux disease has significantly increased over recent years. The larynx is highly sensitive to the effects of LPRD and is similarly responsive to proton pump inhibitor pharmacotherapy. The hypothesis of the study was that proton pump activity exists in the human larynx and plays a functional role in normal and/or pathological laryngeal tissue. STUDY DESIGN: Pathological investigation. METHODS: Two fresh human cadaveric larynges (one male and one female larynx) were obtained as part of an exempt protocol from the Human Subjects Committee and were formalin fixed and paraffin embedded. Banked human stomach tissue was also obtained for use as comparative positive and negative control specimens. Sections were immunostained with monoclonal antibodies reactive with both alpha and beta subunits of the H+/K+-ATPase (proton) pump. Specimens were reviewed for staining pattern and intensity. RESULTS: Stomach parietal cells (known to produce gastric acid) exhibited strongly positive staining for both the alpha and beta subunits of the proton pump. There was no staining in stomach cells that were not morphologically consistent with the parietal cell. In the human larynx there were strong focal and identical staining patterns in the serous cells and ducts of the minor seromucinous glands by both alpha and beta monoclonals to the proton pump. There was variable staining in the laryngeal epithelium that was thought to be consistent with artifact staining resulting from tissue processing. CONCLUSION: The H+/K+-ATPase (proton) pump is present in serous cells and ducts of submucosal glands in the human larynx. Proton pump inhibitor pharmacotherapy may have a site of action in seromucinous glands of the human larynx, with possible relevance for patients treated for chronic laryngitis with or without laryngopharyngeal reflux disease.

Comparison of postoperative facial nerve outcomes between translabyrinthine and retrosigmoid approaches in matched-pair patients. Ho, S. Y., Hudgens, S., Wiet, R. J. Central Florida Ear Institute, Melbourne 32901, USA. *The Laryngoscope* (2003), November, Vol. 113 (11), pp. 2014–20, ISSN: 0023-852X.
OBJECTIVES/HYPOTHESIS: The objective was to assess whether the translabyrinthine approach for acoustic tumour removal offers better post-operative facial nerve function compared with the retrosigmoid approach. STUDY DESIGN: Retrospective case review from a tertiary otology referral centre. METHODS: Patients who had undergone either retrosigmoid or translabyrinthine approach for removal of acoustic neuroma from January 1, 1980, to December 31, 1999, were included in the study. Two groups of patients were created, one containing retrosigmoid cases and the other, translabyrinthine. Attempts were made to match each retrosigmoid case to a translabyrinthine case with regard to tumour size, patient age, and date of operation. This matching served to eliminate these variables from influencing post-operative facial nerve outcomes. From an initial pool of 450 patients, 35 pairs of patients were matched for the study. Facial nerve functions were reported at immediate, three-month, and

one-year post-operative periods. RESULTS: Patient demographics demonstrated that matched patients had almost identical tumour size, patient age, and date of operation. Comparisons of postoperative facial nerve functions between the matched groups revealed that retrosigmoid approach carried 2.86 times higher risk of facial nerve dysfunction during the immediate post-operative period. However, by one year, the facial nerve outcomes were similar between the two groups. CONCLUSION: Compared with the translabyrinthne approach, retrosigmoid approach carries a higher risk of postoperative facial nerve dysfunction during the immediate post-operative period. However, long-term facial nerve outcomes are identical between the two approaches.

Evidence for surviving outer hair cell function in congenitally deaf ears. Rea, P. A., Gibson, W. P. R. The Sydney Cochlear Implant Centre, Department of Otolaryngology, University of Sydney, New South Wales, Australia. *The Laryngoscope* (2003), November, Vol. 113 (11), pp. 2030–4, ISSN: 0023-852X.
OBJECTIVES/HYPOTHESIS: The hypotheses of the study were that congenital hearing impairment in infants can result from the isolated loss of inner hair cells of the cochlea and that this is shown by the presence of abnormal positive summating potentials on round window electrocochleography. The objectives were to establish the proportion of infants with hearing loss affected, the nature of the cochlear lesion, and its etiology. And to highlight the important implications for otoacoustic emissions testing and universal neonatal screening. STUDY DESIGN: A prospectively conducted consecutive cohort study with supplemental review of notes was performed. METHODS: Four hundred sixty-four children underwent round window electrocochleography and auditory brainstem response testing under general anaesthesia to assess suspected hearing loss. The presence of abnormal positive potentials was recorded. Otoacoustic emissions data were collected separately and retrospectively. RESULTS: Three hundred forty-two children had significant bilateral congenital hearing loss. All results were from hearing-impaired children. Abnormal positive potentials were recorded in 73 of 342 children (21 per cent). Eighty-three per cent of children with otoacoustic emissions also had abnormal positive potentials, but only 14 per cent of children without otoacoustic emissions had abnormal positive potentials ($p<.001$). In the neonatal intensive care unit setting, 43 per cent of infants were found to be have abnormal positive potentials, whereas only 10 per cent had abnormal positive potentials if not in the neonatal intensive care unit setting ($p<.001$). Abnormal positive potentials were present in 63 per cent of infants born before 30 weeks gestation and in 41 per cent of infants born at term ($p<.001$). Abnormal positive potentials were identified in 57 per cent of infants with documented hypoxia and 11 per cent of children with no episodes ($p<.001$). Otoacoustic emissions were present in 48 per cent of infants from the neonatal intensive care unit, despite their hearing loss. CONCLUSION: Both otoacoustic emissions and abnormal positive potentials may originate from outer hair cell activity following inner hair cell loss. This may occur in more than 40 per cent of hearing-impaired children in the neonatal intensive care unit setting. Chronic hypoxia is the most likely cause. Otoacoustic emissions testing may not be a suitable screening tool for such infants.