

## Letters to the Editor

### A Scheibe cochlea deformity with macrocephaly: a case for single channel implantation

Dear Sir,

There are two points that need raising in connection with this paper (Roy *et al.*, *JLO* 112: 1065–1068).

The first point is simply one of nomenclature: the authors describe 'bilateral primitive common cavity... dysplasia' as Scheibe deformity, however Scheibe described dysplasia of the membranous contents of the cochlea, rather than bony dysplasia of the cochlea duct. This is, perhaps, a minor objection, especially since the word Scheibe was not included in the list of key words for the paper. Historical eponyms can be misleading and the authors are in distinguished company, since Schuknecht (1980) included this type of common cavity deformity (first described by Cock in 1838) within the blanket term 'Mondini deformity'.

The second point is a more practical one and relates to the authors' decision not to use an intracochlear, multichannel electrode 'because of the risks of a CSF leak'. They suggest that this means the risk of a spontaneous CSF fistula, from the IAM, via the cochlea and into the middle ear.

There are three reasons why this may not have been a logical clinical decision:

a) If there were to have been a spontaneous, congenital CSF fistula, a second potential leak from cochleostomy is unlikely to have increased the existing risk of meningitis; such spontaneous fistulae should, in any case, be sealed and cochlear implant surgery provides a good opportunity for this. Since the middle ears had, in the past, been infected, meningitis would probably already have occurred in this child were a fistula present. The principal risk in these cases is of a surgical 'gusher' of CSF on opening the cochlea to insert the implant, but techniques exist to seal gushers and only one case of meningitis has been described following cochlear implantation in such a case (Page and Eby, 1997). The presence of middle ear infection is, of course, a contraindication to any kind of cochlear implant until the infection has been cleared up.

b) There have already been a number of reports of successful intracochlear, multichannel implantation in common cavities (Molter *et al.*, 1993; Firszt *et al.*, 1995; Tucci *et al.*, 1995; Hoffman *et al.*, 1997; Luntz *et al.*, 1997; McElveen *et al.*, 1997; French and Munro: personal communication) and the benefits and performance seem to be within the same range found in children with normally formed cochlear ducts, though probably below the mean level.

c) Curiously there seems not to be a linear relationship between the presence of the raised intracochlear CSF/perilymph pressure that leads to a gusher and the degree of maturity of these dysplastic

cochlear ducts, since some relatively well formed Mondini cochleas, with incomplete partitions, have quite forceful gushers, while some more primitive common cavities contain perilymph at normal pressure (Firszt *et al.*, 1995; Tucci *et al.*, 1995; Hoffman *et al.*, 1997; Luntz *et al.*, 1997; McElveen *et al.*, 1997; present writer's own experience).

It is true that the child's macrocephaly, possibly linked to raised intracranial pressure, might have led to a brisker gusher than usual, though this is not discussed in the article. It is also true that at the age of 11 years such a congenitally deaf child might well be turned down as an implant candidate on grounds of age. It could be misleading, however, to suggest that a common cavity cochlea is itself a contraindication to a multichannel implant, which could provide more benefit than a single channel device.

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### Author's reply

Mr Graham's point about historical eponyms is accepted and the authors will avoid eponyms for cochlear deformities in future papers.

Mr Graham's second point is practical and of great importance. There is nothing to suggest the patient which is the subject of this paper had ever had a spontaneous CSF leak or meningitis. If it was to occur on the other side we agree it would present an excellent opportunity to put in a multichannel device while sealing the leak.

As a result of reports of successful intracochlear multichannel implantation in common cavities, we considered this procedure for our patient but the limited benefit at the age of ten, the risk of macrocephaly being an indication of raised intracranial pressure, decide us in favour of a single channel extracochlear device.

The purpose of this paper was to recommend this approach to other surgeons, who for whatever reason might favour a conservative implant procedure in such cases.

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#### **Bilateral sudden sensorineural hearing loss following non-otologic surgery**

Dear Sir,

Post-operative otovestibular disturbances.

De la Cruz and Bance (*JLO* **112**: 769–771) reported a case of post-surgical hearing loss, only finding 25 similar case reports. They described seven mechanisms by which sudden sensorineural deafness might occur, concluding that all theories were speculative at best. There is, however, a well-documented and perfectly satisfactory explanation, endolymphatic hydrops, which curiously they ignored completely.

Abundant audiological evidence for cochlear hydrops appears in the three cited cases of Cox and Sargent: *Case 1*. Aural fullness, lightheadedness, flattish bilateral hearing loss, *Case 2*. Fullness, profound flat loss on right maximal at 500 and 1000 Hz, typical low tone and high tone loss on left with normal hearing at 2000 Hz, *Case 3*. Muffled hearing and tinnitus, fullness in right ear, severe flat loss on right maximal at 500 Hz, *Their case*. Nausea/vomiting/vertigo, severe fluctuant right deafness maximal at 250 Hz with characteristic 4000 Hz peak, profound left fluctuant loss maximal below 2000 Hz.

Even separately these features strongly indicate hydrops. *Cases 2 and 3* followed spinal surgery. Panning *et al.* (1983) noted typical fluctuant peaked losses after urological operations under spinal anaesthesia; which they could not explain; a similar case appeared in Cox and Sargent's review. Cochlear hydrops due to low CSF pressure was the only mechanism proposed (Gordon, 1983). This idea is not new.

De la Cruz and Bance located one previous report of sporadic sudden deafness in otosclerosis (DuVall *et al.*, 1981). In the discussion to this report, the only

proposed mechanism in the non-stapedectomized ear was hydrops, for which Paparella stated there was ample clinical and pathological evidence. A previous review in this journal (Hochermann and Reimer, 1987), the first to claim a bilateral loss, labelled it "Meniere-like' low frequency hearing impairment". Audiosensitivity, an often ignored symptom of hydrops (Gordon, 1983; 1997), was also noted.

Walsted (1998) prospectively performed serial audiometry on 34 patients undergoing spinal anaesthesia, 60 having neuromas removed from their opposite ears and 32 having neurosurgery. Reversible low-tone or peaked losses typical of hydrops were common, especially after large losses of CSF. No other type of deafness was seen.

There is an extensive literature on post-operative nausea and vomiting (Watcha and White, 1992). It would be surprising if its cause was unrelated to the occasional case of deafness, especially if associated with vertigo, as above. Although there is no consensus as to its origin, there are many facts implicating dehydration or labyrinthine fluid pressure changes. Risk factors are previous motion sickness, opioids that sensitize the labyrinth, motion or change in position, otological surgery, middle ear pressure change (with nitrous oxide), spinal anaesthesia, hypotension and dizziness. Sickness is reduced by drugs effective in motion sickness and otovestibular disorders. Extra fluids dramatically reduced post-operative thirst, dizziness and nausea (Yogendran *et al.*, 1995).

De la Cruz and Bance's case developed musical hallucinations shortly after vertigo and tinnitus. In a recent case (Marneros *et al.*, 1997), also otosclerotic, hallucinations were clearly related to alcohol intake and withdrawal, implicating labyrinthine pressure changes, and were abolished by stapedectomy. A review of musical hallucinations (Gordon, 1997) shows that irrespective of any concurrent psychiatric, psychological, otological, pharmacological, religious or mystical states, there is always an associated hyperirritable inner ear or incipient hydrops. The pathological basis is perilymphatic hypotension from dehydration, weight loss, hypotension or loss of CSF. Thus a lumbar puncture led to a mystical state (Judson and Wiltshaw, 1983), including a powerful rhythm or vibration: 'It could be likened to a pulse or even a tune sounding through the universe'.

Despite many appeals in neurological journals (Gordon, 1998). I have not found neurological examples of musical hallucinations. If any otologist knows of a case of musical hallucinations where thorough examination excludes fluctuant low tone deafness and all symptoms of hydrops or ear disease, could they please report them, as otherwise psychiatrists, reliant on a neurological model of auditory hallucinations, are in deep trouble.

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**Author's reply**

I thank Mr Gordon for his illuminating and interesting commentary on our case. There are a few points in his letter I would like to address. Firstly, the ear has only a limited number of possible responses to insult of any kind, 'muffled hearing loss', 'tinnitus', 'fullness' are common accompaniments of any sudden hearing loss, and cannot be taken as definitive evidence of hydrops, although I do agree they are suggestive. Much of the literature Mr Gordon cites describe hearing changes after loss of CSF, which was not the situation in our reported case with no spinal anaesthesia and no opening of the dura. Thirdly, I have a very difficult time believing that musical hallucinations are an end organ phenomenon, a result of 'hyper irritable inner ear or incipient hydrops'. In my experience with these hallucinations, they are often complex musical arrangements, and many times patients will describe them as a tape recording. In fact, often they are songs from childhood, and at least two musician patients have been able to listen to them accurately enough to transcribe them in musical notation. It is difficult to comprehend how an irritable inner ear could produce music of this rhythmic arrangement. The most important point against a peripheral mechanism is that most of the cases I have encountered of musical hallucinations (four I can remember) have followed total ablation of the inner ear by a translabyrinthine removal of the inner ear, by definition excluding a peripheral mechanism, at least on the operated side.

Nevertheless, I do appreciate the insights Dr Gordon brings to this discussion and thank him for sharing his observations.

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**Eye protection for ENT junior doctors**

Dear Sir,

The importance of eye protection is well recognized in surgical fields (Bell and Clement; 1991, Berridge *et al*, 1993). Otolaryngology risks contamination in the operating theatre (Hinton *et al.*, 1991) and during ward procedures. Epistaxis is a common problem managed on the ward by junior doctors. Close proximity to a patient with epistaxis who may be sneezing or coughing inevitably results in a wide dispersion of blood. It would be reasonable to expect doctors packing noses to be provided with eye protection in expectation of this hazard. A recent audit of 20 SHO's working in different departments countrywide, including inner-city areas, was performed by telephone questionnaire.

Fourteen of those questioned said no protection was provided. Goggles or visors were provided for only six. However these were irregularly utilized and only one doctor regularly wore eye protection. Sixteen had been recently splashed in the face and eyes. Conjunctival transmission of hepatitis B can occur and a case of HIV contracted in this way has been documented (Gioannini *et al.*, 1988).

It is argued that no distinction should be made between high risk and ordinary patients and universal precautions should be adopted in operating theatres (Wastell, 1992). These practices should be maintained on the wards. Glasses are known to provide insufficient protection against blood splashes (Brearley and Buist, 1989) and a full face visor should be available for doctors managing epistaxis.

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**Erratum**

The influence of pregnancy on sensation of ear problems – ear problems associated with healthy pregnancy: K Tsunoda *et al.*, *JLO* 113: 318-320.

The author has informed us post publication that there are a number of errors in the data originally provided. The author wishes to apologize for this mistake and in order to correct what was otherwise a publishable paper we print below the Materials and methods and Results sections with new data. EDS.

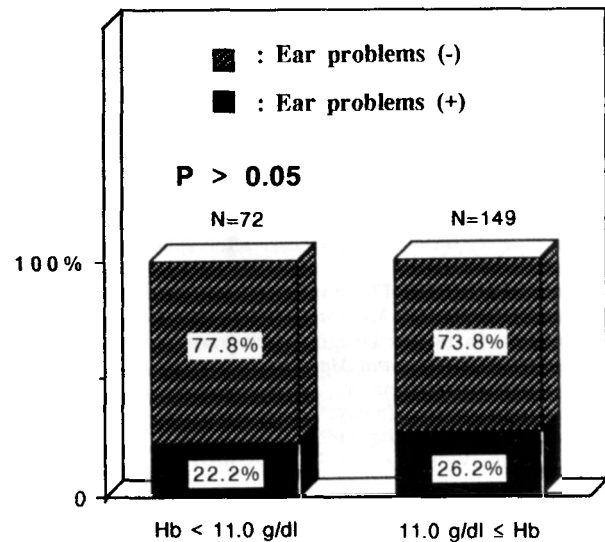
**Materials and methods**

To address this question, data were obtained from a group of healthy women who attended the gynaecology clinic in our hospital as pregnancy cases between February 1995 and January 1998 and who volunteered to participate in our study. A control group was drawn from healthy female co-medical staff members of our hospital who had never been pregnant. Before the study, all volunteers were screened for good ear/nose health using ear and nose physical examination, pure-tone audiometry, and impedance audiometry. The final subject population comprised 228 healthy pregnant women and 29 healthy women who had never been pregnant. The data used for comparing the two groups were taken from a questionnaire about ear problems that was presented to all subjects. In addition, for 221 cases in the pregnant group, we obtained measures of blood haemoglobin level (Hb) and measures of blood pressure when they were eight months pregnant.

**Results**

Results from the questionnaire showed that 25.0 per cent of women in the pregnant group reported ear problems: fullness in the ear, tinnitus, and/or autophonia. Interestingly, all those reporting ear problems during pregnancy also reported that they resolved completely on delivery of their babies. Even amongst those complaining of ear problems, we could not detect any ear hearing loss or other problems in the pure-tone audiometry and impedance audiometry. Among the non-pregnant females, the incidence of ear problems was 3.4 per cent (one case, who reported experiencing ear problems during each

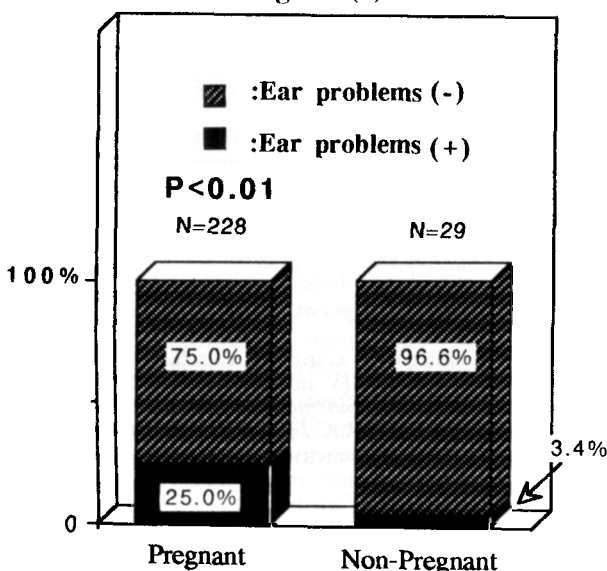
**Figure (b)**



premenstrual period). Chi-square analysis showed a significant group difference ( $p < 0.01$ ) in the incidence of ear problems (Figure 1-a).

To examine possible causes of ear problems in the pregnant group, the 221 cases for whom haemoglobin levels and blood pressure had been measured were divided into subgroups according to these results. In particular, they were divided into anaemic ( $Hb \geq 11.0$  g/dl) and non-anaemic ( $Hb < 11.0$  g/dl) groups, and also into hypotensive (systolic pressure  $< 100$  mmHg) and non-hypotensive (systolic pressure  $> 100$  mmHg) groups. Of the anaemic women from pregnant group, 22.2 per cent reported ear problems, while the incidence of ear problems in the non-anaemic pregnant woman was 26.2 per cent. Chi-square analysis showed that this difference is not significant ( $p > 0.05$ ) (Figure 1-b). However, in the hypotensive subgroup, there was a 47.1 per cent incidence of ear problems, while there was a 15.0 per cent incidence of ear problems in the non-hypotensive group. This difference is highly significant ( $p < 0.001$ ) (Figure 1-c).

**Figure (a)**



**Figure (c)**

