Section of Otology

President Alan G Gibb FRCS

Meeting 5 March 1976

Sudden Sensorineural Deafness

Dr Victor Goodhill
(Department of Surgery, School of Medicine, University of California, Los Angeles, California 90024, USA; and Division of Otolaryngology, Cedars-Sinai Hospital, Los Angeles, California)

Labyrinthine Membrane Ruptures in Sudden Sensorineural Hearing Loss

Sudden hearing loss is an otologic emergency, and may occur as the result of lesions of the external, middle, or inner ear, internal auditory meatus, cerebellopontine angle or central nervous system. The hearing losses may be mild, moderate, severe, or total. The term 'sudden hearing loss', as used in quotations, has acquired a special clinical usage, namely, to describe a major sudden hearing loss (in a presumably previously normal ear) due to a cochlear lesion. Until recently, such cases have been considered to be of idiopathic etiology, or presumed to be caused by viral, vascular, endocrine, allergic, or other lesions.

Recent observations of round window membrane and/or oval window ligament ruptures with perilymphatic fistulæ (Freeman & Edmonds 1972, Goodhill 1971, Goodhill *et al.* 1973, Pullen 1972, Tonkin & Fagan 1975) in such cases make it necessary to broaden the etiologic classification.

Labyrinthine membrane rupture (LMR) sudden sensorineural hearing loss (SNHL) is caused either by sudden barotrauma or physical exertion.

Etiology

I have divided my series of sudden SNHL cases into two broad groups: idiopathic etiology, and LMR etiology (Fig 1).

Idiopathic etiology: (1) Viræmia: one viral etiologic cause has been clearly identified for decades, i.e. the mumps virus. Rubella, rubeola and other viral diseases can also be causes of sudden SNHL. The basic histopathologic findings in specific and presumptive viral lesions consist of atrophy of stria vascularis, cochlear duct collapse, and displacement-distortion of the tectorial membrane. The mechanism is probably that of viræmia with the cochlear stria vascularis as the portal of entry. (2) Vascular lesions considered etiologic include capillary sludging, vascular spasm, small vessel occlusions, massive inner ear hæmorrhage related to leukæmia, thromboangiitis obliterans (Buerger's disease), serum hyperviscosity syndrome, hypercoagulation, microglobulinæmia, inflammatory vasculitis, and microembolism. Gussen (1976) recently reported histopathologic temporal bone findings in a human case of sudden vascular SNHL. Prior histologic evidence for vascular etiology has been based on animal studies. (3) Etiologic factors other than the viral and vascular include many constitutional conditions, all of which are based on presumptive clinical impressions without definitive pathologic documentation.

Labyrinth membrane rupture (LMR) etiology: Simmons (1968) postulated a theory of intracochlear membrane breaks in sudden hearing loss. The possibilities of producing intralabyrinthine membrane lesions by head blows, increases in

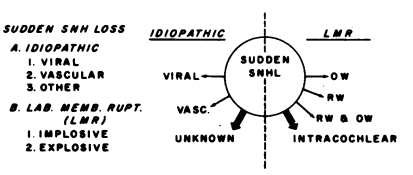

Fig 1 *Sudden sensorineural hearing loss* (SNHL): *idiopathic (viral, vascular, other) and labyrinth membrane rupture (LMR) (implosive, explosive) etiologies.* OW, *oval window.* RW, *round window*

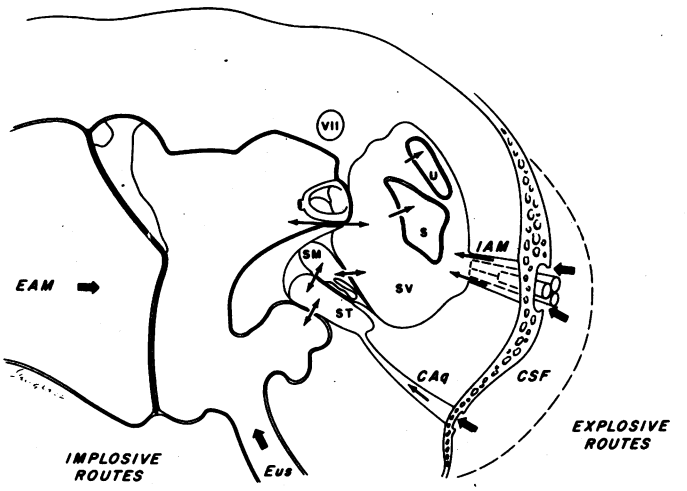

Fig 2 *Explosive and implosive routes for labyrinth membrane ruptures.* CAq, *cochlear aqueduct.* CSF, *cerebrospinal fluid.* EAM, *external auditory meatus.* Eus, *eustachian tube.* IAM, *internal auditory meatus.* S, *saccule.* SM, *scala media.* ST, *scala tympani.* Sv, *scala vestibuli.* U, *utricle.* VII, *facial nerve*

CSF pressure, and other trauma have been reported (Fee 1968, Moscovitch *et al.* 1973, Stroud & Calcaterra 1970).

In 1971, I reported three cases of surgically confirmed labyrinthine fistulæ in stress related or physical exertion related cases of sudden hearing loss (Goodhill 1971). In an additional paper (Goodhill *et al.* 1973) my colleagues and I confirmed the phenomenon of perilymphatic fistulæ which can occur in either round or oval or both windows in cases associated with prior physical or barotrauma stresses.

The intimate hydrodynamic relationships between the subarachnoid space CSF pool, the cochlear aqueduct, the medial vestibular lamina cribrosa and the labyrinthine perilymph are illustrated in Fig 2.

Exertion may involve an increase in CSF pressure. The cochlear aqueduct and the internal auditory meatus cribriform foramina are both potential pathways for transmission of CSF pressure changes to the perilymph system. Of the two, the cochlear aqueduct seems the more likely.

For a number of years the cochlear aqueduct was considered to be closed by a barrier membrane. Recent studies by Palva (Palva & Dammert 1969, Palva 1970) indicated that such a membrane was not always present, and also indicated that there was a difference between the size and shape of the cochlear aqueduct in infants and adults. On the basis of these findings, I postulated the possibility that an infantile type of cochlear aqueduct might persist in some adults (Goodhill 1971). Such relatively large cross-sectional area aqueducts might account for variable pressure transmissions from CSF to scala tympani perilymph.

Stress episodes of the Valsalva type, which result in an abrupt transient increase in CSF pressure may then be transmitted to scala tympani and from that perilymph area to vulnerable intracochlear labyrinthine membranes and eventually to round window membrane and/or oval window annular ligament regions.

It is possible for such labyrinth membrane lesions to follow both explosive and implosive pressure transmission phenomena (Fig 3).

During stapedectomy, surgeons have frequently observed the phenomenon of perilymph refilling the vestibule within seconds or minutes following inadvertent suction at the oval window. In addition, the so-called 'perilymph gusher' has been encountered occasionally, with profuse perilymph-CSF drainage from the oval window. There is, thus, clinical as well as anatomical evidence of the intimate relationships between CSF pressures and the vestibular perilymph pool.

The conductive loss phenomenon in post-stapedectomy perilymph fistulæ (Goodhill 1967) exemplified one aspect of the broad spectrum of conductive, mixed and cochlear hearing losses which can occur in post surgical fistulæ. It is quite likely that similar physiologic explanations can be considered in the more complicated phenomena of spontaneous labyrinth membrane ruptures.

The *explosive route* makes it possible for a sudden increase in CSF pressure (due to physical stress or barotrauma) to be transmitted through

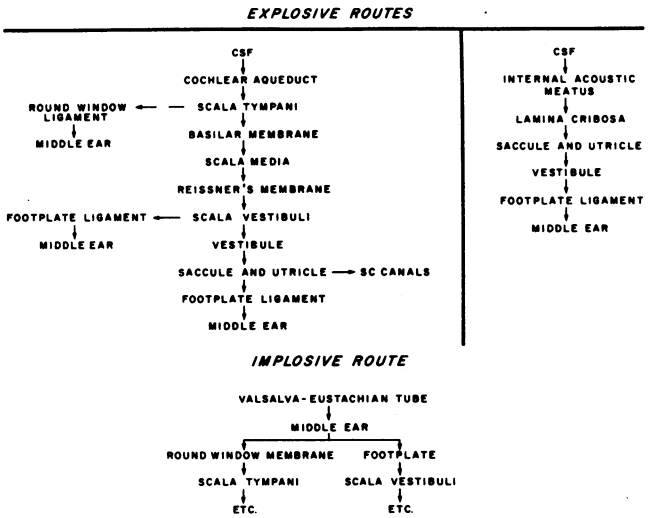

Fig 3 *Potential explosive and implosive routes for labyrinth membrane ruptures*

cochlear aqueduct to scala tympani, with ensuing rupture of round window membrane and/or basilar membrane, and other labyrinthine membranes. In an experimental cat study, Harker *et al.* (1974) showed round window membrane bulging as a result of increased CSF pressure in 83% of experimental animals. A similar mechanism through the internal auditory meatus may involve the medial vestibule wall cribriform communication to scala vestibuli. The ruptured intralabyrinthine membranes may produce a chain reaction of sequelæ in the perilymphatic and endolymphatic systems, with hearing loss, vertigo, and tinnitus (Fig 4).

The *implosive route* from sudden Valsalva forces involves a sudden increase in tubotympanic pressure with round window membrane and/or oval window ligament ruptures. There may be a similar reverse chain reaction with disruption of internal labyrinthine membranes (Reissner's and basilar) with resulting hearing loss, vertigo, and tinnitus (Fig 5).

In our perilymphatic fistulæ cases, physical exertion or barotrauma history was clear cut in some, but vague in others. Labyrinth window ruptures have occurred with or without vertigo in deep sea divers, unrelated to nitrogen em-

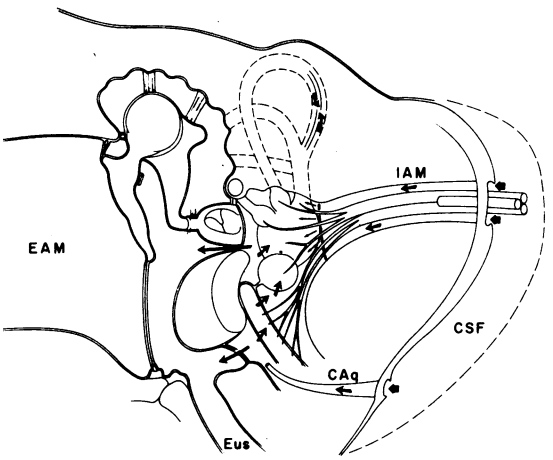

Fig 4 *Explosive route for labyrinth membrane ruptures (in detail).* CAq, *cochlear aqueduct.* CSF, *cerebrospinal fluid.* EAM, *external auditory meatus.* Eus, *eustachian tube.* IAM, *internal auditory meatus*

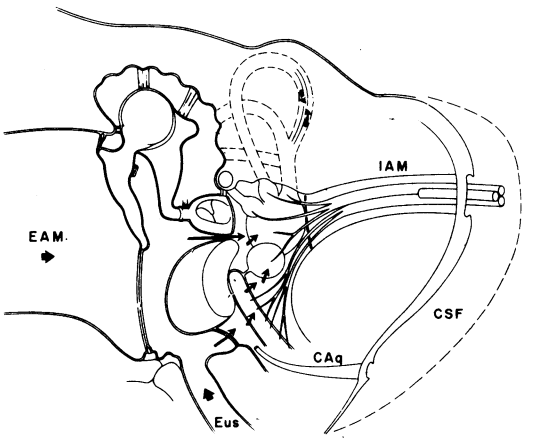

Fig 5 *Implosive route for labyrinth membrane ruptures (in detail).* CAq, *cochlear aqueduct.* CSF, *cerebrospinal fluid.* EAM, *external auditory meatus.* Eus, *eustachian tube.* IAM, *internal auditory meatus*

bolization, and some cases have followed aircraft travel. Patients have become suddenly deaf following physical exertion such as heavy lifting or unusual exercise.

Clinical Aspects of Idiopathic and LMR Etiology Cases

The problem of etiologic diagnosis is very difficult, since a basic history pattern may be common to both syndrome groups. Hearing loss is the first symptom in some, tinnitus or dizziness in others. Both idiopathic and LMR cases may have similar histories of onset. However, there are some differences in pre-onset histories which are important.

Idiopathic sudden SNHL histories: In idiopathic sudden SNHL syndrome (presumably viral type) the patient may give a history of a cold, influenza, sore throat, sinusitis or exposure to upper respiratory infections.

In idiopathic syndrome (presumably vascular type) there may be a history of previous cardiac or hypertensive disease with or without the use of heparin or coumadin, history of diabetes, arteriosclerosis, hypercholesterolæmia, or hyperlipid states.

However, in some patients in the idiopathic group, there is no history of viral or vascular disease.

LMR sudden SNHL histories: In most LMR syndrome cases there is a clear-cut history of a specific physical event followed immediately or later by a sudden cochlear hearing loss. The event may be stress or exertion related to barotrauma (swimming, diving, snorkel, or scuba diving, or unusual aircraft travel incidents). The hearing loss may have followed an event of lifting, coughing, sneezing, difficult micturition or defæcation, sexual intercourse or any other physical exertion. A severe acoustic trauma or blast episode may have preceded the hearing loss. In diving cases, differential diagnosis must include the possibility of nitrogen embolization. The barotrauma, exertion, or physical stress history may not have seemed unusual for the patient, and thus be completely ignored. Probing questions are necessary to elicit such possible causes.

Otologic and General Medical Findings

The workup of both syndrome history groups is comparable. It includes otologic and general medical history, otologic and complete medical examination, which includes a search for bradycardia, arrythmia, bruits, urine and blood studies, sedimentation rates, fluorescent treponemal antibody absorption tests and special tests where indicated for hypercoagulation, lipidæmia, hyperviscosity and other circulatory syndromes.

Otologic findings in both syndromes show normal tympanic membranes. Audiologic studies show moderate, or severe sensorineural hearing losses, usually greater in the middle and higher frequencies. Bekesy tracings are usually Type II, occasionally Type III, and very rarely Type IV. Impedance studies and tympanometry curves are normal. There is usually a positive stapedial reflex on the involved side, and frequently an absent stapedial reflex on the opposite (normal) side.

Vestibular studies are varied. Positional nystagmus occurs in some patients, with or without labyrinthine preponderance. There are no consistencies in vestibular findings. Radiographic findings are usually normal in both syndromes.

Management Approaches

Variable management approaches based on idiopathic etiology: The management of sudden cochlear hearing loss, unrelated to recognizable specific ear lesions, has been based in the past entirely upon idiopathic, presumptive etiologic concepts and on empirical therapeutic theories. Presumptive diagnoses of vascular cochlear accident, viral disease, endocrine, allergic, or other causes have been made, and followed by empirical treatment with single or multiple modalities including vasodilators, steroids, hyperbaric oxygen, vitamins, intravenous procaine, intravenous histamine, adenosine triphosphate, other drugs, and surgical procedures such as stellate ganglion blocks. All have been used on empirical grounds. No single definitive therapeutic approach has been in general use.

Spontaneous recovery and the treatment evaluation dilemma: Spontaneous recovery is frequently noted. Such recovery may be partial or total, and may occur within hours, days, weeks, or months. Controlled statistics are difficult to evaluate because of the vagueness of an idiopathic etiology concept. In 'idiopathic syndrome' cases the etiology must be considered presumptive. Objective etiology can be confirmed surgically in some LMR cases. A definitive oval window or round window fistula observed in surgery introduces some objectivity into etiology. However, even in such cases, as in idiopathic syndrome cases, the final definitive answer must await post-mortem temporal bone histopathologic studies.

Suggested new management approach: Our present management approach is based upon: (*a*) those aspects of idiopathic syndrome history and general medical findings which indicate a likely presumptive etiology and (*b*) those aspects of LMR syndrome history which strongly suggest the possibility of labyrinth membrane rupture.

Management of Mild-moderate Sudden SNHL
All patients with mild to moderate losses (speech reception threshold (SRT) 30–50 db, speech discrimination score (SDS) 40–80%) with either syndrome possibility are placed on bedrest at home with head elevated 30 degress and allowed out of bed only for necessary attendance at the clinic for otologic and/or general medical diagnostic studies. No physical stress (exercise, sexual activity, walking, sports) is allowed. Only bathroom privileges are allowed. No initial medications other than necessary tranquillizers and sedatives are advised during the workup period. In every case of sudden SNHL, complete otologic, radiologic, vestibular and general medical examinations are necessary. Every attempt is made to rule out a specific otologic cause.

If no such otologic cause can be elicited, specific medical studies are carried out to elicit such possible etiologic factors as viral, vascular, drug, and other 'idiopathic' medical etiologies. Occasionally, unsuspected problems such as clotting defects, bradycardia, auricular fibrillation, vascular occlusions, and other definitive general medical conditions are discovered. Where there is no suggestive LMR history, an idiopathic syndrome etiology is assumed. The presumption is made that there is a relationship between the recognized general medical condition and the hearing loss, and appropriate medical therapies are instituted. In those patients in whom examination suggests a bacterial upper respiratory infection, empirical broad spectrum antimicrobial therapy is started and changed when possible to specific antibiotic therapy, if pathogenic bacterial organisms can be cultured from the nose or nasopharynx. If there is no medical contraindication, a course of empirical steroid (ACTH or prednisone) therapy may be started.

The patient is advised to avoid any physical exercise in spite of an apparent systemic etiology. Such systemic lesions may or may not be etiologically related to the sudden SNHL lesion. Conversely, a specific history of barotrauma or physical stress does not necessarily prove a LMR etiologic relationship. Where very definitive circulatory, upper respiratory disease, or other general medical conditions are discovered and appropriate treatments instituted, spontaneous prompt beginning improvement in hearing may be noted in a few days. General empirical supportive therapy also includes high vitamin B and C intake, avoidance of tobacco, caffeine, alcohol, salicylates, and of the common allergenic foods (milk, chocolate, nuts, shellfish) which might contribute to membrane œdema.

Audiometric evaluations are done every 3–4 days to monitor hearing levels. If significant serial audiometric gain is demonstrated, this conservative therapy is continued until hearing returns to within 'normal' limits, i.e., a level of 20–25 dB SRT with 70–80% SDS score. However, if tinnitus or vertigo continue, further observation is necessary.

If the hearing does not improve or drops in SRT and SDS measurements, and/or if there is increased tinnitus intensity, and/or if increased vestibular symptoms occur, medical management must be considered inadequate. Management should then be changed to that of a severe-total sudden SNHL case.

Management of Severe-total Sudden SNHL
Idiopathic etiology cases: Patients with severe or total losses (80–100 SRT, 0%–30% SDS) who show no improvement on empirical medical therapy, including corticosteroid therapy and bedrest at home for 7–8 days, are hospitalized and placed at absolute bedrest, no lavatory privileges, with head elevated 30 degrees. The course of corticosteroid treatment is continued to completion. If there is no hearing gain, as measured by bedside audiometry, after 3–4 days of such hospitalization, surgical exploration should be considered in spite of absence of history suggestive of LMR etiology.

LMR etiology cases: Patients with severe or total loss (80–100 dB SRT, 0–30% SDS) are immediately hospitalized and kept at absolute bedrest with head elevated at 30 degrees. Hospital medical consultation is secured for search for possible unrecognized vascular, viral, or other systemic diseases. Daily bedside audiograms are done.

If the LMR syndrome diagnosis appears tenable on the basis of a significant physical or barotrauma stress history, and if there is a completely negative general medical history and examination, the likelihood of membrane rupture is considered to be significant and early surgical exploration is indicated. No absolute guidelines can be laid down for timing of surgical explora-

tion, but in our experience, it appears that it should be considered within the first 10–12 days following the physical or barotrauma stress episode.

Surgical Exploration Technique

Labyrinth membrane surgical exploration is a microsurgical procedure of rather brief duration and is performed under local anæsthesia in our practice.

Local anæsthesia using xylocaine 1.5%-epinephrine 1/200 000 block of the external auditory meatus and canal is carried out utilizing the anterior position of the surgeon (Goodhill 1973). A tilting operating table facilitates adequate exploration. Endomeatal tympanotomy, as in stapes surgery, will usually require removal of a posterior annular ledge of bone for complete posterior tympanic visualization.

In most LMR cases there is mucoperiosteal vascular congestion throughout the middle ear. Fibrous adhesive bands may reflect evidence of spontaneous repair activity.

The oval window exploration is directed to the stapes footplate and to the stapedial annular ligament. A patient examination and observation over a period of many minutes is found to be essential. Fibrous webs are carefully sectioned in the obturator foramen area to provide an unobstructed view of the footplate and especially the anterior footplate marginal region. The fistula ante fenestrum may play a role in this phenomenon. The demonstration of a perilymphatic leak is comparable to that seen in post-stapedectomy fistulæ. Visible clear fluid is gently aspirated with a fine suction tip and the site is observed for refilling with fluid to confirm an active perilymph fistula. Actual footplate 'holes' have not been seen. In small leaks the 'blotter soaking' effect on dry gelfoam pledgets is useful. If a leak is found, the mucoperiosteum over the annular ligament is elevated to expose the ligamentous area for placement of tragal perichondrium autografts (which measure approximately 1×3 mm) around the primary fistulous area.

Attention is then directed to the round window niche. Gentle section of fibrous webs is necessary in some cases to demonstrate a definite fistula. A round window membrane reflex is present in almost every case. Only rarely is even a small 'hole' seen in the round window membrane. Marginal round window membrane defects are the rule. Tragal perichondrial autograft closure of the fistulous area is performed.

The patient is kept at absolute bedrest with head elevated 30 degrees for an average period of 48 hours postoperatively. Upon returning home the patient is restricted to sedentary activity for five to ten days. A prophylactic antibiotic, usually ampicillin, is prescribed for ten days post-operatively. The patient is restricted from flying, going to the mountains, and strenuous physical activity for one month postoperatively. Swimming is prohibited for three months and diving is completely prohibited.

Results of Management Approaches

In our 1973 report (Goodhill *et al.* 1973) a total of 21 sudden hearing loss cases were explored surgically, and 15 fistulæ were found. Our present total number of 76 cases includes 59 surgical explorations; fistulæ were found in 47 cases. No fistulæ were found in 12 cases. The remaining 17 cases were not explored surgically but were managed medically.

Among the 47 patients with fistulæ, all had hearing loss; tinnitus was present in 37 and vertigo in 24.

Of the 47 patients, 29 were males and 18 were females. The age distribution was: 3 patients aged 11–20; 13 aged 21–40; 23 aged 41–60; and 8 aged 61–72.

Of great interest is the continued evidence of left ear preponderance, described by Goodhill *et al.* (1973). Of the 47 fistulæ, there were 14 right ears and 33 left ears, a left ear preponderance of more than 2:1. In the 12 patients without fistulæ, 5 were right ear and 7 were left ear, showing no significant right : left difference.

As regards site of fistula, only 4 of the 47 cases were in the round window alone, 19 were in round and oval windows combined and 24 were in the oval window alone. This is in contrast to some misconceptions which limit fistula consideration to the round window; in our series, the round window alone was the least significant fistula site.

Hearing result studies in the 47 surgically explored cases with fistulæ showed 12 cases (25%) with SRT gains of more than 30 dB and SDS score gains of more than 50%. In 9 cases (18.75%) there were only slight gains, and the majority of patients, numbering 26 (62%), showed no gains. Of the 12 cases (25%) with significant gains, almost all were explored in the first 13 days. Of the larger group with no gains, the intervals between exploration and onset ranged from 15 days to several years.

Of the 12 cases explored with no fistula, only 1 showed a significant gain spontaneously.

Of the 47 fistula cases, a history of stress was obtained in 29 out of 47 cases.

Of the 76 in our series, 17 cases served as medical management controls. There were stress histories in 6 of the 17. Because spontaneous hearing improvements in this group were relatively rapid, no surgery was performed. Of these 17 cases, 13 had significant gains and 4 had no gains. These patients were managed primarily with bed-rest; prednisone was used in 4.

Our work on LMR ruptures is still in an early stage. We have found no fistula when a fistula had been expected. In some, there were findings of oval window and/or round window fibrosis with no fluid. It is possible that a fistula had healed spontaneously with no improvement in hearing, and that this was because of major irreversible intralabyrinthine membrane lesions. In most cases with proven fistulæ and surgical repair there were no hearing improvements, and again this was probably because of major intralabyrinthine membrane lesions. In almost all of the cases which showed no improvement there had been delay in treatment.

The problem is complex. The major cochlear hearing losses cannot be explained adequately on the basis of window membrane ruptures alone, but must be considered to result from complex intralabyrinthine membrane lesions which accompany surgically demonstrated oval window and/or round window fistulæ. The transtympanic observation of a round window membrane or oval window membrane fistula does not answer questions relating to the fates of basilar membrane, Reissner's membrane, tectorial membrane, hair cells, supporting structures, stria vascularis, and other intracochlear structures (Fig 6).

A few fistulæ have recurred, either spontaneously, or in response to new physical exertion or barotrauma factors, and have been re-explored and regrafted.

Long-range Follow Up

In all patients in whom no improvement is noted, regardless of management or findings, long-range follow up is advisable. The possibility of an undiagnosed lesion other than the blanket label of 'sudden hearing loss' must be considered in all such cases. If there has been no improvement in hearing, tinnitus, or vestibular symptoms, such conditions as vestibular Schwannoma (VIII nerve tumour), Ménière's disease, and CNS vascular disease must be considered in the follow up.

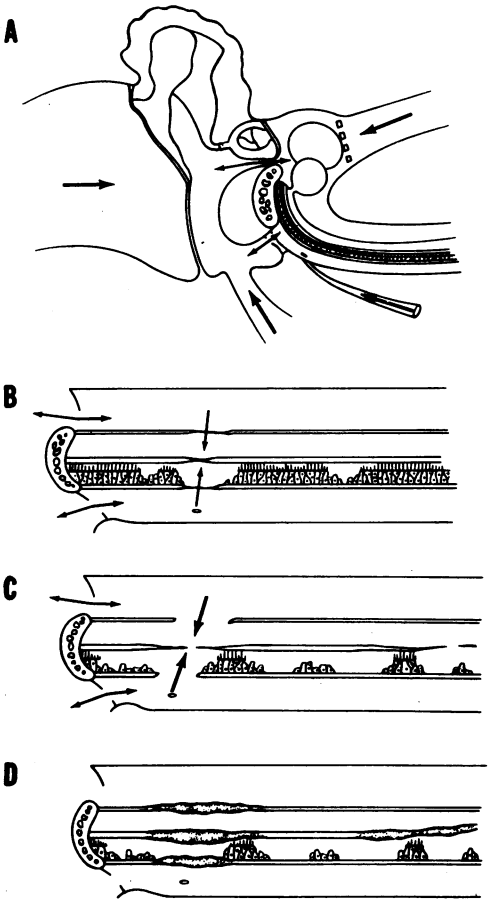

Fig 6 *Potential labyrinthine sequelæ – theoretical possibilities.* A, *diagrammatic explosive plus implosive ruptures.* B, *minor ruptures with minimal organ of Corti damage.* C, *major ruptures with persistent intracochlear fistulæ.* D, *healed intracochlear lesions*

Stenvers internal auditory meatus radiographs (as well as Schuller mastoid radiographs) are considered part of the diagnostic test battery in the evaluation of every case. In those patients in whom specific diagnosis has not been made following medical or surgical management, and in whom there was no improvement in hearing, tinnitus, or vestibular symptoms, it will be advisable for long-range follow up to include polytome radiographs, EMI scans, and cerebello-pontine angle myelograms.

A patient may actually think that a hearing loss has occurred suddenly, when in effect, a slight loss with no subjective awareness simply became

suddenly worse. This may happen in cochlear hydrops or in true Ménière's disease.

A patient with previous episodes of transient mild cerebral ischæmia may have had an VIII nerve vascular episode to explain the sudden sensorineural hearing loss. A new episode of severe vascular occlusion may occur with CNS symptoms other than hearing loss, tinnitus, or vertigo.

These and other diagnostic possibilities must be kept in mind in the follow up of those cases in whom no improvement followed either medical or surgical treatment.

Summary and Conclusions

The division of sudden sensorineural hearing loss cases into a new classification of idiopathic and LMR syndromes is now necessary. The demonstrations of definitive labyrinth window perilymphatic fistulæ into the middle ear in sudden hearing loss cases cannot be ignored, even if the precise intracochlear sequelæ cannot be known now. When a fistula is demonstrated, surgical closure is mandatory, regardless of unanswered intralabyrinthine membrane questions. The lessons of documented cases of meningitis following any temporal bone fistulæ cannot be ignored in this category of sudden hearing loss.

These new LMR observations are real and must now be considered in the difficult diagnostic problem of sudden sensorineural hearing loss cases. We will await some answers from experimental animal studies now under way, but ultimate answers can only come from human temporal bone histopathologic documentation.

REFERENCES
Fee G A (1968) *Archives of Otolaryngology* **88**, 477
Freeman P & Edmonds C
(1972) *Archives of Otolaryngology* **95**, 556
Goodhill V
(1967) *Laryngoscope* **77**, 1179
(1971) *Laryngoscope* **81**, 1462
(1973) *Annals of Otology, Rhinology and Laryngology* **82**, 547
Goodhill V, Harris I, Brockman S J & Hantz O
(1973) *Annals of Otology, Rhinology and Laryngology* **82**, 2
Gussen R
(1976) *Annals of Otology, Rhinology and Laryngology* **85**, 94
Harker L A, Norante J D & Ryu J H
(1974) *Transactions of the American Academy of Ophthalmology and Otolaryngology* **78**, 448
Moscovitch D H, Gannon R P & Lazlo C A
(1973) *Annals of Otology, Rhinology and Laryngology* **82**, 53
Palva T (1970) *Acta Otolaryngologica* **70**, 83
Palva T & Dammert K
(1969) *Acta Otolaryngologica*, Suppl. 246
Pullen F W (1972) *Transactions of the American Academy of Ophthalmology and Otolaryngology* **76**, 1444
Simmons F B (1968) *Archives of Otolaryngology* **88**, 41
Stroud M D & Calcaterra T C
(1970) *Laryngoscope* **80**, 479
Tonkin J P & Fagan P
1975) *Journal of Laryngology and Otology* **89**, 733

Mr Andrew W Morrison
(*The London Hospital, London E1 2AD*)

Sudden Sensorineural Deafness: Outline of Management

In 1971, when Victor Goodhill introduced us to the entity of spontaneous rupture of the round window as a treatable cause of sudden sensorineural deafness, he unleashed a great potential of surgical energy. For the otologist the concept of a simple operation to cure or partially reverse acute hearing loss is so attractive that it carries the risk of replacing adequate investigation by exploratory tympanotomy. While acknowledging the debt which otology owes to Dr Goodhill for providing this new dimension, it is important to recognize that the spectrum of otological disease is embraced by this topic. Acute auditory failure can be described as idiopathic only when investigation has been complete and thorough. With increasing knowledge of diseases of the otic capsule, of the inner ear and of its neural connexions, the idiopathic group of patients, though still large, is diminishing.

Causes of Sudden Sensorineural Deafness

To attempt to describe all the causes of sudden deafness would be a mammoth and inappropriate task. Some idea of the diverse nature of the problem can be obtained from Table 1, which is based on a careful analysis of 319 patients whose presenting symptom was sudden deafness. Conductive causes have been excluded, as have surgical iatrogenic inner ear lesions. Two important facts stand out: despite modern diagnostic effort a cause for the acute auditory failure has not been forthcoming in as many as 1 in 4 – the idiopathic group; and amongst those with an identifiable cause a relatively limited number of pathologies account for more than a third of the cases. These pathologies are acoustic neuroma, late syphilis of the temporal bone, infection with the viruses of mumps, herpes zoster or measles and, finally, Ménière's disease. Acute endolymphatic hydrops, primarily affecting the cochlear duct, is frequently overlooked as an etiological factor. The hydrops may be idiopathic, as in Ménière's disease, or secondary to involvement of the otic capsule or surrounding bone by syphilis, otosclerosis, the sequelæ of chronic eustachian insufficiency, or rarer conditions such as osteogenesis imperfecta or Cogan's disease.

General Management

This is best considered under four headings: history, localization, diagnosis and treatment.