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Drewe Lecture Theatre
Reynolds Building
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ABSTRACTS

ORAL PRESENTATIONS

INVITED SPEAKER

Peter Rudge, The National Hospital for Neurology and Neurosurgery
The Neuro-Otological Abnormalities in Multiple Sclerosis

Multiple sclerosis causes a wide variety of abnormalities of Neuro-Otological interest, since the disease commonly involves the brainstem. The most frequent signs and symptoms are due to involvement of structures concerned with eye movements, resulting in a wide variety of nystagmoi and other extraneous eye movements, and abnormalities of pursuit and saccadic movements. Some of these abnormalities are due to the involvement of the vestibular system and its connection centrally.

In contrast, clinically significant disturbance of hearing is rare in multiple sclerosis, but investigation of those patients in whom there is an auditory disturbance, has given insight into the movements of retro-cochlear hearing loss and the function of the efferent-cochlear system.

A selection of these various abnormalities will be discussed.

Classification of responses to Bithermal Caloric stimulation in disease and in health
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Episodic experiences of dizziness arising from vestibular imbalance may be associated with changes in cardiorespiratory regulation and the subjective sensations of anxiety and/or nausea.

In order to investigate this unilateral bithermal (30oC & 44oC) caloric irrigations stimulation were performed upon 103 patients attending Neuro-Otology clinics and 30 gender and age matched controls without known cardiovascular disease that had given informed consent. Continuous recording of respiratory frequency (FRmin-1; Respiband) and discrete heart rate (HR; bpm) and discrete blood pressure measurements (diastolic and systolic; mmHg) were obtained prior, during and post irrigation. Prior to the experimental protocol participants were given questionnaires rating motion sickness susceptibility (MSS) and tendency to faint. Upon completion of the protocol, questionnaires were administered where participants' sickness, faintness, and dizziness in response to the irrigations were rated, in addition to the short-form of trait anxiety.

Table 1. Classification based upon autonomic reactivity:

Type	Response	Patient %	Control %
1	None	57%	83%
2	Severe nausea or vomit only	35%	3%
3a	Severe nausea + tachypneoa	2%	7%
3b	Severe nausea + tachycardia + hypertension + tachypneoa	1%	0%
4a	No nausea but tachycardia + hypertension + tachypnoea	2%	0%
4b	No nausea but tachypnoea only	0%	7%
5a	Post-caloric migraine + anxiety + nausea	2%	0%
5b	Post-caloric migraine + anxiety + nausea + cardiorespiratory response	1%	0%

In conclusion, unsurprisingly healthy subjects were less physiologically responsive to caloric stimulation. From such responses one may infer the operation of at least 3 distinct but not exclusive neural pathways.

Firstly, vestibular activation appears particularly in patients to be potent in activating brainstem nausea centres. Secondly, activation of a vestibulo-limbic system pathway that evokes sensations and tachypnoea associated with anxiety. Thirdly, there appears to be a weak vestibulo-brainstem cardiovascular regulatory centre pathway. Pathology may result in a relative hypersensitivity and/or an attenuation of compensatory mechanisms, thus permitting increased response expression.

The Tullio phenomenon - Audio-vestibular findings and mechanisms

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SUMMARY
There is a well recognised association between sound or pressure induced vertigo (known as the Tullio phenomenon) and anterior canal dehiscence (ACD) on CT scanning. Using tympanic membrane displacement measurements (TMD), bone conduction thresholds and vestibular evoked myogenic potentials (VEMPs) in addition to 3D computerised video-oculography (VOG) during 100-110dB monaural sound stimulation, and CT scanning on 20 twenty patients with the Tullio phenomenon, new insights into the mechanism have been obtained.
Findings in 14 patients with uncomplicated Tullio (Group I) were as follows:
* Normal air conduction thresholds from 500 - 4000 Hz (14/14)
* An ipsilateral air-bone gap at 1 kHz of > 10 dBHL, indicating a conductive gain (10/10)
* A patent cochlear aqueduct / third window on TMD (9/10)
* Lowered VEMP thresholds ipsilaterally (12/12)
* Anterior canal - appropriate torsional nystagmus with a 1 kHz sound stimulus onset, as recorded by 3D VOG (10/10)
* CT scanning of the petrous temporal bone showing an ipsilateral ACD (8/8)
Where the ACD was found to be bilateral, the symptomatic ear was always associated with the larger dehiscence. Three of these 14 patients were wind or brass players, two more suffered a head injury at the onset of symptoms and two more were sound engineers, one of whom became symptomatic following acoustic trauma.
Four of the remaining six patients (Group II), had other underlying otological pathology, which is likely to have contributed to the mechanism of sound-induced vertigo, but apart from altered audiological investigations, otherwise conformed to the findings seen in the 14 uncomplicated Tullio patients.
However, the remaining two patients (Group III) had bilateral vestibular failure but no conductive gain on audiometry, a horizontal nystagmus to sound stimulus onset on VOG, and normal ipsilateral VEMP thresholds. No ACD was found on CT scanning. An alternative mechanism for the Tullio symptoms in these patients is suggested.

Vestibular perception and vestibulo-spatial navigation in the congenitally blind

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Summary
The ability of congenitally blind subjects to utilise non-visual mechanisms in navigating assumes a crucial importance in everyday life. Whilst sighted humans can perceive and accurately utilise vestibular signals in navigating in the dark (called “vestibular navigation”), the presence of such a function in the congenitally blind is unknown. Vestibular navigation may also be influenced by a brainstem mechanism called the ‘velocity storage’ integrator which prolonges the duration of the peripheral vestibular signal. We studied vestibular

navigation and quantified the perceptual vestibular velocity storage time constant in congenitally blind and sighted subjects in the dark. **Vestibular navigation** was assessed by having subjects (6 blind vs. 12 sighted) steer a motorised Barany chair, in response to imposed angular displacements of different amplitude and dynamic profiles. Two tasks were studied. 1) 'Go Back to Start' (GBS), a path reproduction task (egocentric navigation) and 2) 'Complete the Circle' (CTC) a path completion task (allocentric navigation). All subjects sighted and blind) accurately performed GBS as measured by linear regressions between stimulus and response displacement (r^2 range 0.61 - 0.94). There was no significant difference (P>0.5) between sighted and blind group GBS performance as assessed by their respective linear regression r^2 and slopes for required angle vs. response angle. **Vestibular velocity storage function** was measured by having subjects (5 blind vs. 31 sighted) turn a tachometer wheel to indicate instantaneous angular velocity perception, in response to velocity steps of 90deg/s in the dark. The cumulative velocity storage time constant of the congenitally blind (7.2s) was shortened by about 50% with respect to normal controls (16s). These values, comparable to estimates of the cupular time constant, indicate an essentially absent vestibular storage integrator in the patients. Two congenitally blind sbjects had ultra-short time constants (2.39s and 3.30s respectively) which were associated with superior angular path completion performance. These two subjects continuously participated throughout childhood and adulthood in spatial activities involving head (or body) movements, suggestive that the degree and duration of vestibulo-spatial activity may influence the development of ultra-short time constants and enhanced navigation in blind subjects. Our findings thus agree with animal experiments suggesting that vision is necessary for the development of a normal velocity storage mechanism. The navigational experiments show that humans maintain representations of derivatives of the raw velocity vestibular input which can be accessed for spatial navigation. Whilst congenital blindness does not interfere with simple path reproduction navigation, it is associated with less efficient path completion task performance, analogous to true spatial navigation, than in sighted subjects. Early and prolonged physical spatial activities in childhood and adulthood however, appear to be of value in improving vestibular navigation in the congenitally blind.

Are the secondary/tertiary referrals of patients to an otology led vertigo clinic different to those referred to a multidisciplinary vertigo clinic?

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Introduction and Aims:
Dizziness is a common complaint affecting up to 23% of the UK population at any time1. General practitioners can refer these patients to a number of specialists including neurologist, cardiologist and otolaryngologist. This study sought to determine if secondary/tertiary care referrals of patients to an otology led vertigo clinic differed from those referred to a multidisciplinary vertigo clinic.

Methods:
Anonymised data on patients referred to the North-Bristol NHS trust otology led vertigo clinic from May 2002- April 2003 were studied. The source of referral, patient's sex and age were tabulated. The number of patients with a single diagnosis of a vestibular, central, idiopathic or other cause for vertigo including multiple pathology was recorded. Chi-squared statistical tests were used to compare the categorised patient data with published data for a tertiary centre multidisciplinary vertigo clinic2 to determine if the patient characteristics differed.

Findings:
90 new patient referrals were seen at the North Bristol Trust otology led vertigo clinic for the period studied. The mean age of patients in the Bristol sample was 52.6 years and 50.7 years in the literature controls. 59 patients (66%, 95% CI 59.8-72.2%) were women, a similar proportion to the multidisciplinary clinic population. The proportions of patients in the major diagnostic groups were different in the Bristol sample compared to the multidisciplinary literature control clinic (Chi-squared, p=0.004). Interestingly a peripheral/labyrinthine disorder was the most common major single diagnosis made in both clinic settings accounting for 43% and 57.5% of patients seen in the Bristol and control clinic respectively. Patients without

an attributable diagnosis accounted for 15.5% of the Bristol series, central disorders 2.2% and other disorders 38.9%.

Conclusions:
The patients referred to the otology led vertigo clinic in Bristol do not differ in age or sex from the patients referred to the literature control multidisciplinary vertigo clinic. The proportion of patients with labyrinthine/vestibular disorders in the multidisciplinary clinic however exceeded that in the otology led clinic.

References:
1. Yardley L et al. Prevalence and presentation of dizziness in a general practice community sample of working age people. Br J Gen Pract. 1998;48(429):1131-5.
2. Heaton JM et al. Evaluation of the dizzy patient experience from a multidisciplinary neuro-otology clinic. J Laryngol Otol.1999;113:19-23.

Long term results of Round window catheterization and Gentamicin infusion in Meniere's disease
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Over the past 4 years, 30 patients have undergone Round window catheterization with Gentamicin infusion as treatment for Meniere's disease at the Leicester Royal Infirmary. We present the results at long-term follow up in these patients.

Patients and methods:
All patients had their catheter placed through endaural tympanotomy and underwent 10 days of continuous Gentamicin infusion (10 mg/ml) at a rate of 5 microlitre/hour. The mean age at treatment was 53.4 years and 14 were males. 25 patients had minimum of 2 years follow up. The results were analyzed using AAOHNS (1995) guidelines and the Dizziness Handicap Inventory.

Results:
11 (37%) patients had complete control of vertigo, while 17 (57%) had substantial improvement. One patient had persistent imbalance and in the other, vertigo recurred after 12 months. Hearing was unaffected in 18 patients, improved in 4 and reduced in 8, the mean reduction in pure tone average being 17 db. The tinnitus improved or remained unchanged in 22 (73%) patients but worsened in one.

Conclusion:
Round window catheterization and gentamicin perfusion is an effective treatment for Meniere's disease and should be considered in patients who failed medical treatment.

Interobserver error and time taken in the measurement of acoustic neuromas
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Acoustic neuromas are benign, slow-growing tumours, which account for about 6% of all intracranial tumours. In certain cases, particularly when dealing with small asymptomatic acoustic neuromas or in the presence of good hearing, it is accepted clinical practise to manage patients conservatively by interval scanning. The rationale for this is that many tumours grow very slowly or not at all. Clearly, it is important to be able to accurately monitor tumour growth in these patients. A number of papers have been published describing and comparing different methods of tumour measurement, but no commonly accepted method exists at this time. In this paper the authors attempt to evaluate the inter-observer error and the time taken using some of the commonly accepted measurement techniques.
The MRI scans of 10 patients, with previously diagnosed acoustic neuromas, were reviewed by 10 observers (who were either ENT surgeons or Radiologists). Each scan was assessed in 5 ways (1. maximum extracanalicular diameter, 2. maximum antero-posterior diameter, 3. the square root of the product of the antero-posterior and the medio-lateral diameters, 4.

manually segmented area, 5. perimeter) by each observer. We also examine a potential model taking account of measurements in the coronal scans, in addition to the axial scans. The results for each different measurement technique were analysed for inter-observer error and the average time taken to assess each scan was calculated for each technique.
The results show a wide variation in the time taken for evaluation. Each technique exhibits inter-observer error. The greater the number of measurements, the longer it takes to complete the analysis and the greater the chance of inter-observer error. However, this must be balanced against the fact that the simplest measurement techniques do not reflect the true size of the tumour most accurately. We propose a protocol for the sizing of acoustic neuromas in our department.

Pulsed low pressure to control the symptoms in Meniere's disease: Using the Meniett® *in the treatment of episodic vertigo*

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Objective: This is the first UK study evaluating the use of the Meniett® in the treatment of Meniere's disease. The device generates low transtympanic pressure pulses via a ventilation tube.
Methods: 16 patients fulfilling the AAO criteria for definite Meniere's disease who have failed conservative treatment have been referred for treatment of their disabling vertigo. Symptoms and their severity were recorded throughout the study. Audiometry, Posturography, disability score and the Glasgow Benefit Inventory were used as outcome measures.
In the first stage baseline audiometry, posturography and level of disability according to the AAO disability score were assessed. In the second stage ventilation tubes were inserted and the response was assessed over a minimum of three months. The Meniett® device was then used for a minimum of three months recording severity and frequency of attacks. A successful outcome was defined as a return to normal ability.
Results: All patients had unilateral disease and disability scores of >4. Only two patients noted an improvement of their symptoms with grommets in place. The majority found little change or even a deterioration with grommets in situ.
All patients using the Meniett® noted an improvement of their vertigo, however a change of their disability was noted by 72% with return to normal daily activity and employment. There was only little improvement of hearing, tinnitus or aural fullness.
Conclusion: This new mode of treatment may present an alternative in the treatment of Meniere's disease. However long-term results (> 2 years) will need to be reviewed to confirm these promising results.

Chemical labyrinthectomy in Meniere's disease by intratympanic gentamicin infusion.

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Objective: Intratympanic gentamicin therapy is becoming the treatment of choice for uncontrolled Meniere's disease. Successful control of incapacitating vertigo without affecting residual hearing is the challenge. This study aims to evaluate the outcomes of our experience of this treatment modality with respect to vertigo control and hearing preservation. The results should serve as a guide to appropriate case selection in the future.

Methods: 6 consecutive patients with unilateral intractable Meniere's disease were treated with intratympanic gentamicin between September 1999 and October 2001. All patients had magnetic resonance imaging with contrast prior to treatment to exclude retrocochlear or central pathology. Buffered gentamicin solution (20 mg/ml) was administered three times daily for three consecutive days through an intratympanic catheter. The hearing was monitored strictly during and at intervals following treatment. The patients' subjective response to therapy was followed.

Results: Complete control of vertigo was achieved in all patients (follow-up range 18-30 months) based on the AAO-HNS criteria. At the outset 1 patient had unserviceable hearing and 5 had a moderate to severe unilateral hearing loss. All patients had normal hearing in

the contralateral ear. Residual hearing on the treated side was reduced (n=5) by a mean pure tone average of 40dB. Hearing in the contralateral ear was unaffected in all patients.

Conclusion: Hearing loss is a recognized complication of intratympanic gentamicin therapy for vestibular ablation. In intractable Meniere's disease it offers better risk/benefit outcome than other invasive therapies. This particular treatment regime appears to result in chemical labyrinthectomy rather than vestibular ablation. It may be particularly suited for the management of vertigo in patients in whom hearing preservation is not required.

Results of steroid perfusion of cochlea using Silverstein micro wick for Sensory Neural hearing loss

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Background: Sudden Sensorineural hearing loss is traditionally treated with oral steroids. Cochlear perfusion with steroids through Silverstein wick has been shown to stabilize or improve hearing. We present our experience with this treatment.
Patients and methods: Over the past 3 years, 36 patients with sudden or fluctuating sensorineural hearing loss underwent Silverstein wick insertion and cochlear perfusion with steroids at the Leicester Royal Infirmary. The round window niche was identified either through a myringotomy or tympanotomy. After removal of any adhesions, the Silverstein wick was placed directly in contact with the round window niche. Dexamethasone (4mg/ml) was used topically over the wick 2 drops thrice daily for the next 10 days to 2 weeks, after which the wick was removed. 7 patients also had oral steroids for a week concurrently or prior to wick treatment.
Results: Data were available for 26 patients. The mean age at treatment was 52.9 years with a median of 56 years and ranging from 22 to 77 years. 16 patients were males.
Hearing improved in 3 patients (11.5%); unchanged in 23 patients, of whom 4 had preoperative unrecordable hearing. These 4 patients were not included in the statistical analysis. Paired t-test was used to analyze the hearing results and statistically significant difference was noted for change in hearing thresholds at 1000 Hz ($P < 0.05$). Tinnitus worsened in 3 patients and was unchanged in 10 patients. 8 patients did not have tinnitus preoperatively, of whom one developed it post operatively.

Table shows the results of paired t-test to compare the average of the differences between the hearing thresholds before and after the treatment.

Frequency	Mean Difference	Standard Deviation	95% C.I.	P- value
500 Hz	5.00	14.3	-1.2 to 11.2	0.12
1000 Hz	6.82	13.5	0.8 to 12.8	0.03
2000 Hz	3.18	11.6	-1.96 to 8.3	0.21
4000 Hz	1.36	9.28	-2.8 to 5.5	0.49
PT average of 0.5,1,2 & 4 KHz	3.86	9.89	-0.5 to 8.2	0.08

Conclusion: Steroid perfusion of the inner ear is variably effective for the treatment of sudden sensory neural hearing loss and is particularly useful when oral steroids are contraindicated.

Mismatch negativity in cochlear implant patients

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Introduction: Cochlear Implantation is an established approach for the management of profoundly deaf children and adults who obtain no benefit from conventional hearing aids. A paediatric cochlear implant programme requires special test techniques and professional skills when compared to the adult population. Evoked potentials are sensitive indicators of

the physiological integrity of the afferent auditory system and provide the opportunity to evaluate infants and young children objectively. One of the promising components of the auditory event related potentials is the Mismatch negativity (MMN). The MMN is a non task related neurophysiological index of auditory discrimination and has the potential to diagnose central auditory processing dysfunction in children with cochlear implants in whom peripheral deafness is overcome with the use of the implanted device and speech processor.

Methods: The MMN was elicited in 37 cochlear implant patients by the synthesized speech stimulus pair /ba/ and /da/ delivered in an oddball paradigm fashion, through speakers at an intensity of 75dB SPL. The evoked potentials were acquired using a digital EEG amplifier, and off line analysis was done using Scan 4.2 software. The final clinical outcome of these patients was noted from the regular assessments by the Teachers of the Deaf, Audiologists and Speech and Language therapists and was based primarily on their Category of Auditory Performance score (CAP: 0 to 7).

Results: The MMN was present in all patients (5) who demonstrated a CAP score of 7. In addition, it was also present in 5 patients with an average CAP score of 6. It was absent in the remaining 21 patients with a CAP score of less than 6. In 6 patients, off line analysis could not be conducted satisfactorily to demonstrate the presence or absence of MMN due to artefacts generated by the cochlear implant package and speech processor.

The MMN is a sensitive, objective indicator of normal auditory sensory memory and discrimination. It can be used in paediatric cochlear implant patients to predict behavioural outcome in the very young population who may not give much feedback by way of behavioural tests since it does not require any patient participation. We need to further study replicability and reliability of MMN in the paediatric age group before using it as a clinical test.

Cochlear Implantations - Various Topics

1. Short-Term versus long-term antibiotic prophylaxis in Cochlear Implant Surgery.
2. Cochlear Implantation in Psoriasis patients.
3. Flap complication in cochlear implant surgery secondary to pre-operative radiotherapy.
4. Cochlear Implantation and Management of chronic suppurative otitis media: Single stage procedure?

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Abstract 1:
Objective: To evaluate antibiotic prophylaxis options for cochlear implant surgery. Does long-term antibiotic prophylaxis have any advantage over single peri-operative dose in preventing post-operative infection?
Study Design: Retrospective case review
Patients: A Total of 292 adult and paediatric patients who went cochlear implantation during a 15-year period (1988-2003) were reviewed.
Main outcome measure: Minor and major post-operative wound infections in first 4 weeks.
Results: There were 4 major and 8 minor complication among 292 patients (complication rate: 4.1). The infection rate was high and severe in patients who had either C incision (11.1%) or extended endaural incision (7.5%) and in patients with pre-existing medical condition. The infection rate was high in patients who had long term antibiotics (5.6% and 13% in 5 days and 7 day regime) compared to short term (single dose) group.
Conclusion: Long term antibiotic prophylaxis did not prove any advantage over single peri-operative dose. Predisposing medical conditions and extensive surgical incisions was associated with a greater severity of infections and higher risk of wound complications.

Abstract 2:
Cochlear implantation has become a safe and effective method for auditory rehabilitation of severe to profound sensorineural hearing loss. The flap complications being the commonest1 and the risk increase further when it's associated with medical conditions

predisposing to infection. We present two patients with psoriasis who underwent cochlear implant surgery, discussing the risk of surgical site infection and treatment options to minimise infection.

Abstract 3:
Radiotherapy can alter normal wound healing following surgery. Surgical and radiation factors are important consideration in predicting wound complication. We present a case of flap necrosis following cochlear implant in a patient who had radiotherapy for benign brain tumour.

Abstract 4:
In a series of 360 patients who had cochlear implantation in our center, three patients (4 procedures) had cochlear implantation with obliteration of mastoid cavity and management of cholesteatoma as a single staged procedure. Two patients were bilaterally deaf secondary to CSOM and had bilateral mastoid cavities and in one patient congenital cholesteatoma was identified during cochlear implantation. A mastoidectomy or revision mastoidectomy with obliteration of mastoid cavity (using modified Rambo technique) and cochlear implantation was performed as a single stage procedure. Surgical procedure, complications, follow-up and outcomes are discussed.

Proteins responsible for autoantibody production in immune-mediated inner ear disease.
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Background: Autoimmune inner ear disease and audiovestibular dysfunction in connection with systemic autoimmune disease both belong to the large group of inner ear disorders of unknown aetiology. On the other hand, immune-mediated inner ear dysfunction, if diagnosed, is one of the few forms of treatable inner ear disorder with a good response to immunosuppressive therapy and early treatment may reverse or ameliorate the progression of disease. The inner ear antigens targeted by the immune system in autoimmune inner ear disorders have not yet been identified and this has significantly impaired the development of diagnostic tools and more specific therapeutic strategies.
Aim: To study the presence of autoantibodies in sera from patients with immune-mediated inner ear disease and systemic autoimmune disease accompanied by SNHL and/or peripheral vestibular dysfunction.
Material and Methods: Autoantibodies in sera from patients with immune-mediated inner ear disease were studied by using the Western blot technique. Patients with inner ear disorders with no clinical presentation of autoimmune disease (eg noise, genetic, traumatic or drug induced) were included as controls. Inner ear tissue from guinea pigs were used as antigen. The different parts of the inner ears were dissected and separated into 6 different tissue preparations (organ of Corti, stria vascularis, the three semicircular canals with their ampullary tissue, the saccule, the utricle and the endolymphatic sac). To identify the patients with antibodies against inner ear tissue all serum were screened against whole inner ear tissue preparations. Subsequently, the samples showing ear reactive antibodies were tested against the separate dissected inner ear tissues.
Results: To date serum from 25 patients with immune-mediated inner ear disease have been screened against whole inner ear tissue preparations. 17 of these patients were shown to have antibodies specific against inner ear proteins. The most frequently detected antibodies were against proteins migrating around 188 kDa, 98 kDa, 68 kDa and 17 kDa. In addition, some patients were shown to have a single strong reactivity against inner ear specific proteins of either 28 kDa, 49 kDa or 62 kDa. The experiments using separate inner ear tissues incubated with sera indicated that the patients do have antibodies binding to specific proteins in specific parts of the inner ear. In sera from 4 control patients screened against whole inner ear tissue preparations no inner ear specific antibodies could be detected.
Conclusion: Serum from 68% of patients with immune-mediated inner ear dysfunction and audio-vestibular complications of systemic autoimmune disease have been found to exhibit antibodies to inner ear specific proteins. This novel application of a standard immunochemical technique may provide a definitive clinical diagnostic tool for autoimmune inner ear disease.

SINGLE CASE SESSION

MELAS Syndrome.Review of the literature. The role of the otologist.

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MELAS syndrome (mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes) is a rare disorder of mitochondrial DNA (mtDNA). It was first described in 1984. Although it does not feature in the acronym, hearing loss is a common finding in MELAS. Reports have shown that at least 50% of MELAS patients have sensorineural hearing loss(SNHL).The histopathologic finding is atrophy of the stria vascularis. Normal hearing is dependent upon the stria vascularis which provides the ionic environment necessary for sound signal transduction. The spectrum of hearing loss due to MELAS has been recently studied with otoacoustic emissions, auditory brainstem responces (ABR) and MRI. The findings were consistent with a predominantly cochlear origin for the hearing deficit. We outline the recent progress in understanding the pathology of this syndrome.
Patients with this syndrome may present to the otolaryngologist with SNHL which is genetic in origin. Cochlear implant may be required and can give very good results. In recent years there have been a few cases reported in the literature of succesful cochlear implantation, one at this centre, in patients with MELAS syndrome. Outcome will be reviewed in this atypical group of patients.
High index of suspicion is required for early diagnosis and intervention. An understanding of the pathology and cochlear implantation of mitochondrial deafness is essential. We believe it is important for the otolaryngologist to maintain an awareness of MELAS syndrome in order to facilitate rehabilitation of these patients.

An unusual case of X-linked adrenoleukodystrophy with auditory processing difficulties as the first and sole clinical manifestation.

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X-linked adrenoleukodystrophy (X-ALD) is characterised by demyelination that is associated with a deficient beta-oxidation of very long chain fatty acids. We report the unusual case of a male adult with X-ALD who was diagnosed at the age of 26 by brain MRI performed because his brother had been diagnosed with a rapidly deteriorating form of X-ALD. His sole symptom was hearing difficulties in the presence of a normal audiogram since childhood. He has remained stable for 7 years now. Central auditory testing in our patient revealed severe deficits in several auditory processes. These findings correlated with involvement of the auditory pathway at the level of the trapezoid body and posterior corpus callosum in particular, on his brain MRI. This case highlights not only the need for thorough audiological investigation of the patient who complains of hearing difficulties in the presence of a normal audiogram, but also that audiological investigations could be of value in the phenotypic evaluation of cases with adrenoleukodystrophy.

The effect of reproductive hormones on auditory system: Modulation of cochlear function during the hormone replacement therapy

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A case of a patient with a premature menopause is presented, as a "pilot" study of the project.

Anecdotal evidence and some studies suggest that reproductive hormones, oestrogen and progesterone, may modulate auditory function during the ovarian cycle. The levels of oestrogen and progesterone fluctuate during the normal cycle, with oestrogen rising during the first phase, while progesterone production starts to rise in the second phase of the cycle, following ovulation. During the menopause, the levels of oestrogen and progesterone are negligibly low.
The presence of oestrogen receptors in the cochlea may allow a direct effect on cochlear function, while their widespread presence within the central nervous system provides a physiological basis for influence upon the central auditory pathways. Oestrogen plays an important role in modulating different neurotransmitters systems, and it is considered to facilitate neural transmission. In contrast, progesterone and its metabolites are known as potent inhibitory modulators, through the interaction with steroid binding sites on the GABA receptors, thus enhancing GABA-ergic "tone".
A 36 year old patient with a premature menopause, who was referred because of difficulty in hearing and intermittent tinnitus, underwent auditory investigations. Pure tone audiometry, stapedial reflexes and brainstem evoked auditory responses showed normal results. Her cochlear function was monitored, using otoacoustic emissions, during two cycles of the hormone replacement therapy, consisting of 16 days of oestrogen and 12 subsequent days of oestrogen and progesterone. Transient evoked otoacoustic emissions (TEOAEs) were recorded on the 7th, 17th and 24th day of the first cycle and 7th and 27th day of the second cycle.
The total TEOAE responses indicated a marked reduction during the progesterone administration, in two consecutive cycles: in the fist cycle by 5 dB and in the second by 3 dB. This is in contrast to minimal TEOAE changes in a control subject and normative data in the literature. These findings may reflect an inhibitory effect of progesterone on cochlear function. Such alterations in TEOAEs have not been previously reported.

Audible eye movements: 'Doctor, I can hear my eyes'

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An unusual but fascinating symptom is one described by patients complaining they can hear their own eye movements. Two cases, with different postulated mechanisms, are presented.

Case 1: A 53 year old female presented with imbalance following left vestibular schwannoma resection complicated post-operatively by a left cerebellar infarct and hydrocephalus. Direct questioning about tinnitus led to the patient describing left tonal tinnitus in which upgaze increased the pitch, while down-gaze lowered the pitch. Notably she said 'I feel I could play a tune with my eyes'.

Case 2: A 32 year old male presented with visual instability and a tendency to fall forward and to the left provoked by loud sounds such as telephone ringing in his left ear. These symptoms could be reduced if the patient tensed his abdominal muscles. Left beating torsional nystagmus was provoked by presentation of loud sounds to the left ear and CT scanning of the petrous temporal bones revealed bilateral dehiscence of the superior semicircular canal. The patient also described hearing a soft 'squeak' in the left ear every time he moved his eyes. This is more noticeable if he made a long sweeping eye movement, low in pitch, 'rather like moving a hard-pressed finger across a clean, wet china dinner plate'.

Discussion: Case 1 describes gaze-evoked tinnitus, a phenomenon first described in 1982 (1), and subsequently found to be surprisingly common (prevalence 19-36%) in patients post vestibular-schwannoma resection (2). It often develops months post-operatively and is usually located to the operated ear. It has also been described in a patient with CPA meningioma (3) and meningeal metastases of malignant melanoma (4). Postulated mechanisms are neural sprouting, abnormal auditory cortical activity, and failure of cross modality inhibition (5).
Case 2 has Tullio phenomenon. These patients often complain of abnormal auditory sensations such as 'hearing footsteps vibrating through the body', and the noise of chewing

making understanding conversations at mealtimes difficult. Patients with Tullio phenomenon have been found to have supra-normal bone conduction thresholds at and lower than 1 kHz (6), and this may be an effect of the dehiscence of the semi-circular canal acting as a lower impedance alternative pathway for the sound energy. It is postulated that this mechanism enables these patients to hear the movements of their eyeballs within the bony sockets, these sounds being conducted through the skull.

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INTERNATIONAL GUEST LECTURE

Migrainous Vertigo

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Both migraine and vertigo are common complaints in the general population and may occur in an individual patient by chance alone. The term migrainous vertigo designates a vertigo syndrome which is causually related to migraine and accounts for at least 10% of referrals to specialized dizziness clinics. The diagnosis of migrainous vertigo is based on recurrent vestibular symptoms, a history of migraine and the temporal association of migrainous and vestibular symptoms. Migraine specific precipitants of vertigo and response to antimigraine drugs support the diagnosis. Hypothetical mechanisms for migrainous vertigo include cortical spreading depression and modulation of vestibular activity by 5-HT, dopamine, noradrenaline or CGRP and ion channel dysfunction. The latter mechanism, which has been identified in related disorders such as familial hemiplegic migraine and episodic ataxia, is supported by the frequent familial occurrence of migrainous vertigo and response to acetazolamide. Treatment relies on the arsenal of acute and prophylactic antimigraine drugs, but controlled trials on their efficacy for migrainous vertigo is still lacking.
Repositioning manoeuvre for canalolithiasis made simple using a labyrinth simulation model

POSTERS

Defective auditory interhemispheric transfer in a patient with a PAX6 mutation.

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PAX6 encodes a transcriptional regulator that is widely expressed in the human cerebrum. Heterozygous PAX6 mutation is associated with congenitally absent/hypoplastic anterior commissure and reduced size corpus callosum. We found deficient auditory interhemispheric transfer in a 53 year old woman with a PAX6 mutation who had absent anterior commissure with normal callosal area. We argue that these auditory deficits can be explained on the basis of the absence of the anterior commissure and/or a functionally deficient corpus callosum.

Does Lamotrigine protect against acute excitotoxicity in the mammalian cochlea?
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Objective: Previous work has demonstrated the synergistic effect of acute noise exposure and hypoxia in producing an accelerated acute threshold shift in the guinea pig. The aim of the current experiments was to assess whether lamotrigine (an anti-convulsant which actions include modulation of voltage-gated Na channels) was capable of protecting the cochlea from this severe excitotoxic injury.

Methods: A total of 15 albino guinea pigs were used. Auditory threshold measurements were obtained by electrocochleography via a surgically implanted round window catheter with integral electode positioned following opening of the bulla. General anaesthesia was maintained throughout the experiments. 7 animals served as controls while the other 8 animals were given 20mg/kg lamotrigine by oral gavage 120 minutes before exposure to 15 minutes of white noise at 110dB in combination with hypoxia of FiO2 12%. Additionally a group of animals received lamotrigine alone while a further group were give the general anaesthetic regimen alone. Thresholds were obtained for 8,16,24 and 30kHz prior to and immediately following termination of the noise/hypoxia and at 15 minute intervals thereafter for a minimum of 3 recordings at each frequency.

Results: Analysis was performed using the Student t-test. Protection was demonstrated at all frequencies tested with p-values ranging between p=0.005 (30kHz) and p=0.03 (16kHz).
Conclusion: Lamotrigine is capable of modulating an acute excitotoxic injury in the guinea pig cochlea. which may have implications not only for protection against other cochleotoxic injuries but also allow protection in other regions of the CNS.

A study on habituation to motion sickness
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Background: Controlled breathing, initiated when a subject begins to feel ill has been shown to have a moderate ameliorating effect on motion sickness (Yen Pik Sang et al. 2003). The purpose of this study was to investigate whether controlled breathing during motion exposure speeds habituation to motion sickness. Methods: In an initial test of baseline susceptibility, subjects (n =30) were rotated (72°/s) in a chair whose rotational axis was tilted off vertical (17° OVAR) for a max of 30 min. Chair rotation stopped when subjects rated 4 on the scale: 1 = OK, 2 = initial symptoms, but no nausea, 3 = mild nausea, 4 = moderate nausea & stop motion, or after 30 min whichever was the sooner. Subjects were matched for motion tolerance times and then randomly assigned to a breathing or a control group (n=15 per group). The same afternoon each group underwent four, consecutive 10 min motion desensitisation sessions, spaced 5 min apart, during which the breathing group followed taped instructions 'breathe regularly and gently throughout motion'. Subjects' susceptibilities were re-evaluated the next day using the baseline test during which controlled breathing continued to be used by the breathing group. Results: Time to sickness rating 4 mean ± sd (min) at baseline: control 11.0 ± 7.8, breathing 12.3 ± 9.2 and on re-testing: control 10.9 ± 9.11, breathing 15.1 ± 9.7. There was an overall effect of habituation to the motion stimulus shown by a significant reduction in symptom scores ($p<0.04$) and increase in tolerance to motion ($p<0.05$) on re-testing. The trend was for the breathing group to perform better on re-test but this did not reach statistical significance.
Conclusion: A stronger effect of controlled breathing on habituation might have been detected if more desensitisation trials had been used. The case for a beneficial effect of breathing on habituation rate is not proven.

Reference:
Yen Pik Sang FD, Billar JP, Golding JF, Gresty MA. Behavioural methods of alleviating motion sickness: effectiveness of controlled breathing and a music audiotape. *J Travel Med. 2003. 10(2):108-11*

Predicting Individual Differences in Motion Sickness Susceptibility by Questionnaire

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Background: Motion sickness susceptibility questionnaires (MSSQ) are useful in the prediction of individual differences in motion sickness due to a variety of provocative environments.
Aims: To develop a short version (MSSQ-Short) of the MSSQ-Long (Golding, 1998), with adult reference norms and motion validity data. Method: Development and prototyping was based on repeated item analysis and various scoring of the MSSQ-Long to produce the best short questionnaire. Norms and percentiles for the MSSQ-Short were based on a sample n=257. Subsequently predictive validity was assessed in a series of experiments in the laboratory using controlled exposure to a variety of provocative motions: cross-coupled Coriolis motion, low frequency translational oscillation, off-vertical axis rotation and visual-motion simulator (total n=178).
Results: Shortening by two-thirds was achieved by collapsing the scaling of exact exposure within each different type of motion and by eliminating questions on vomiting frequency as opposed to nausea. Motion types (cars, boats, planes, trains, funfair rides, etc), sickness severity scaling, and childhood versus adult experiences were preserved. Responses for visual/optokinetic sickness items (cinerama, video game, virtual reality) suggested low population prevalence and added little information. They were excluded but could become important in the future. The MSSQ-Long with MSSQ-Short relationship was around r=0.90, the shortfall in covariance largely being due to underestimations of susceptibility in terms of vomiting in extremely motion susceptible individuals. Reliability: Cronbach's standardised item alpha was 0.87, the correlation between Part A (child) and Part B (adult) was r = 0.68, and test-retest reliability may be assumed to be better than 0.8. Correlation between MSSQ-Short and other non-motion sources of nausea and vomiting (eg stress, viral, headaches, etc) in the last 12 months was r = 0.2 ($p<0.01$). Predictive validity for motion was around median r = - 0.45 for the MSSQ-Short, similar to the MSSQ-Long for the same experiments. Predictive validity for aggregated experiments was higher, around r=-0.55.
Conclusions: The MSSQ-Short maintains reliability, and produces an efficient compromise between length (reduced time cost) and validity (predicted motion susceptibility). French, Dutch, Flemish, German and Russian language variants of the MSSQ-Short are available.

Simultaneous occurrence of a central and a peripheral positional nystagmus during the dix hallpike manoeuvre.

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Introduction.
Peripheral positional nystagmus in the context of posterior canal benign paroxysmal positional vertigo (BPPV) is a common otological disorder. This case report describes its coincidental association with a central positional down beating nystagmus (DBN).

Case report.
A 57 year old man was referred in February 1999 because he insidiously developed over the last few years a severe gait disturbance and dysarthria. His neurological status recently aggravated with the apparition of brief positional vertigo, occurring mainly on rising from bed or rolling on the right side when lying flat. Neurological examination revealed a cerebellar ataxia with bilateral dysmetria as well as marked dysequillibrium with broad-based and ataxic gait. Speech was slurred. Oculomotor examination showed a saccadic pursuit. There was no spontaneous nystagmus in the different positions of gaze with or without fixation (Frenzel's glasses). Left Dix Hallpike and straight head hanging manœuvres triggerred an immediate and short lived DBN. Right Dix Hallpike testing triggered a similar DBN, but after 3 to 5 seconds the nystagmus changed to a strong rotatory upbeating nystagmus in which the upper pole of the eye beats toward the right ear. This latter nystagmus was associated with definite vertigo and both lasted for approximately 10 seconds. Pure tone audiometry was

normal. Brain MRI scan showed severe cerebellar and pontine atrophy; there was no Chiari malformation.
The patient was reassessed two more times in June 1999 and April 2002. He had no definite positional vertigo, particularly when rolling on the right side in a supine position, although he complained of slight and short headedness triggered by all types of head movements. On these two visits, the right, left and straight head hanging manœuvres only revealed an immediate and short lived positional DBN; the previous rotatory upbeating nystagmus could not be elicited. On the last visit, neurological examination has further deteriorated and showed a bilateral pyramidal syndrome with hyperactive deep tendon reflexes and Babinski's signs on both sides.

Discussion and conclusion
The patient initially presented with a cerebellar ataxia and further deteriorated with brainstem involvement, which is highly suggestive of an olivo-ponto-cerebellar form of multiple system atrophy. The unique feature in this patient was the positioning examination, which revealed 2 different types of nystagmus. Firstly, a central positional nystagmus as the nystagmus was immediate, purely vertical and DBN, triggered by right, left and straight head hanging manœuvres, and was not associated with definite vertigo. Such a positional DBN has been reported in central nervous system lesion, particularly cerebellar degeneration in multiple system atrophy [1]. Secondly, another nystagmus which occured after a latency of a few seconds, was rotatory upbeating, and induced definite vertigo, all these features corresponding to a typical posterior canal BPPV [2]. Although no therapeutic manœuvre was performed, the spontaneous disapperance of both positional vertigo and nystagmus is concordant with posterior canal BPPV. This case illustrate the interest of an attentive analysis of the nystagmus during positional manoeuvres which clearly distinguish a central and a peripheral positional nystagmus because of their different characteristics, particularly for latency and direction [1,2,3]. As the central positional DBN occurred with no latency, it was seen before being overshadowed by the strong rotatory up-beating nystagmus.

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Dizziness following Restricted Environmental Stimulation Technique (REST): A Case Report
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Introduction: Restricted Environmental Stimulation Technique or REST, as it is popularly known, is a well-documented tool for individuals who wish to explore the full potential of their bodies, their minds and their natural abilities. REST is based on a scientific approach to deep relaxation and involves depriving the body of the sensory inputs of gravity, temperature, touch, sight and sound. This reduces the brain's workload considerably, conserving vast amounts of energy, which is then re-directed inwards. The effect is called the parasympathetic response, or Relaxation Response.

A lot of studies have been done on Restricted Environmental Stimulation Technique in a flotation tank. However we came across a very interesting case at Mayday University Hospital, London in the special Tinnitus and Vertigo clinic. On doing a medline search, we found out that such a case has never been reported in the literature in the past and is therefore unique in itself. Since we believe that this case could add to our knowledge of the Restricted Environmental Stimulation Technique (REST) and a person's response to it, we are hereby reporting this case.

Participants Methods and Results: The case we are reporting here is a 34-year-old lady who was referred to the ENT Department at Mayday University Hospital, London, by her G.P. She was seen in a specialized Tinnitus and Vertigo clinic at Mayday University Hospital. This is a multidisciplinary clinic is held once a week by the Consultant Otologist, Hearing and Balance therapist and Audiologist. Any tests like Bithermal Caloric tests etc, if required can be done during the clinic.

This 34-year-old lady presented with a history of feeling unsteady and off-balance immediately after a one-hour session of Restricted Environmental Stimulation Technique (REST) in a flotation tank. She had undergone REST because she had been under a great deal of stress and had found it to be useful for that previously, with no side effects. On this occasion, however, she started to feel dizzy while still in the tank and this feeling persisted afterwards. Labyrinthine sedatives proved to be of no help to her, but she noticed that she could reduce the dizziness by relaxing and also that it was not noticeable during driving or travelling. She was otherwise fit and well and gave no history of any medical, neck or back problems, to which these symptoms could be attributed. There were no previous episodes of dizziness.
General physical and Otolaryngological examination was normal and audiovestibular tests confirmed normal function. She was referred to the balance therapist for retraining exercises and is responding well to these.

Comments: This seemingly unusual case raises the possibility that her symptoms could be due to the brain's reluctance to leave the stress free environment of REST and return to the stressful outside world.
We recommend a further study to study the effect of REST in a flotation tank on the balance of a patient who has been through a lot of stress lately.

Olfactory function in essential tremor (ET) and idiopathic Parkinson's disease (IPD)
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Introduction: Error in diagnosis of tremor is common. We wish to determine whether olfactory testing would facilitate differentiation of tremor variety, given that most patients with IPD have impaired olfaction while another common type of tremor known as ET, where marked shaking of the limbs can be mistaken by an inexperienced doctor for IPD. Sometimes the two conditions coexist. At other times ET seems to progress from IPD. Preliminary studies on ET patients indicate that the sense of smell seems to be normal whereas 80% of those with IPD have disordered olfaction.

Methods: Olfactory testing - psychophysical and olfactory evoked potential (OEP) tests have been used to differentiate the two diseases.
1) Controls: 60 subjects aged 24-79 years. 2) E.T.: 40 patients aged 25-79 years conforming to the Brain criteria. 3) IPD: 50 patients aged 39-80 years with predominant rest tremor fulfilling the criteria of the UK PD Research Group. The psychophysical data was obtained using 40-odourant University of Pennsylvania Smell Identification Test (UPSIT) and OEP responses were obtained using Burghart Olfactometer with H2S gas at 2ppm concentration.

Results: Results obtained clearly show that IPD patients have olfaction disorder (anosmia or microsmia) whereas ET patients have normal sense of smell (normosmia). The mean UPSIT score was similar between control subjects and ET patients while it was significantly lower for PD patients. Also, there was no significant difference of latency or amplitude measurement in OEP between control and ET patients while over half of the PD patients had no recordable tracing but in those with a reading latency was significantly prolonged compared to controls and ET patients, but amplitude measurement was normal.

Conclusion: There were clear differences between control and ET patients compared to tremulous PD for UPSIT-40 score and OEP latency but no significant difference between the control and ET group for any measurement. A normosmic patient with tremor is more likely

to have ET than PD while someone with impaired olfaction is likely to have PD or related syndrome. Overlap of scores between the three groups prevents complete separation of disease on the basis of smell testing alone although impaired smell sense in ET might review of diagnosis

A Simple Programme for Nystagmus Analysis: Retrospectively Fitted to an Existing Rotating Chair System

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This poster presents some aspects of a program under development at my hospital for analyzing rotating chair plots to obtain nystagmus gain and phase for sinusoidal testing and maximum velocity and decay constants for impulse rotation testing. The chair system was purchased many years ago before nystagmus analysis programs were common with such a program now desired for the system. Plots are shown for both the eye postion and chair velocity curves for the two types of chair motion as well as the best-fit curves to obtain gain & phase or maximum velocity and decay constant values. The best-fit curves are obtained by using the curves on the computer screen with the needed calculations done by the computer. The results are sent to a printer with a layout specified by the user.

The effect on hearing of changes in ABR during microvascular decompression for trigeminal neuralgia.

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Monitoring hearing before during and after microvascular,decompression for trigeminal neuralgia was carried out in 75 consecutive patients. Hearing thresholds were established prior to surgery and 2 days after surgery. During surgery the eighth nerve status was monitored using ABR.In the majority of cases there were no changes to the hearing after surgery. In 11 patients the hearing improved on average by 5dbHL over the speech frequencies. In 9 patients the hearing became worse over the speech frequencies by an average of 10dbHL This was correlated with the latency shift seen on ABR during the operation while continuously monitoring eighth nerve function. In those subjects where there was no change in hearing threshold the average change in the 1-V Interval was 0.06ms from beginning to end of operation. In those subjects where there was a change in hearing for the worse the average increase in the 1-V interval was 0.9ms. Although well recognised that loss of the 1-V response will result in hearing loss post-operatively. More subtle changes not noticed by the patient in hearing threshold may be detected with a latency shift of 1ms.

VIDEO PRESENTATION (withdrawn)

Repositioning manoeuvre for canalolithiasis made simple using a labyrinth simulation model

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Objective: Particle repositioning manoeuvre for canalolithiasis is an efficient mode of treatment for most forms of geotropic vertigo. The orientation of the semicircular canals in relation to head position needs to be well understood to achieve the intended result.
Method: A model has been designed to simulate the anatomy of the semicircular canals and the movement of the endolymph. It has been used with great success in the Departmnent of Otolaryngology and Audiological Medicine.
Discussion: This technique allows accurate manoeuvring for canalolithiasis of the posterior and lateral semicircular canal. It is extremely useful for teaching and manipulation in patients with limited neck movement.

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