

standard thrombolytic treatment is contraindicated or associated with high risk.

STUART D PRINGLE
Senior registrar
NICHOLAS A BOON
Consultant

Cardiology Department,
Royal Infirmary of Edinburgh,
Edinburgh EH3 9YW

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Tinnitus: its causes, diagnosis, and treatment

Consider psychological, pharmacological, and prosthetic interventions

Tinnitus—the perception of sound originating from within the head rather than the external world—is a common, if not universal, experience,¹ but a distinction must be drawn between the experience and the complaint of tinnitus. The prevalence of complaints of tinnitus is increasing: the British Tinnitus Association estimates that one in 10 people suffer with this symptom: the national study of hearing estimates that one in 12 experience moderate to severe tinnitus.² In less developed countries, however, it is rare.³

The conditions in which tinnitus occurs are legion, but the pathophysiology remains obscure. Hearing impairment is commonly associated, and the proposed pathophysiological mechanisms span cochlear hypotheses, such as decoupling of the stereocilia of the hair cells and spontaneous otoacoustic emission, to neural hypotheses, such as derangement of the spontaneous resting activity of primary auditory nerve fibres and Möller's proposal of ephaptic transmission (“cross talk”) between adjacent nerve fibres.⁴

None the less, many people who perceive tinnitus do not complain about it. In this context, it is relevant that the complaint of tinnitus does not correlate with the acoustic characteristics of the perceived sound,⁵ but there is a significant correlation with psychological symptoms.^{6,7} Moreover, the onset of the complaint of tinnitus may be associated with negative life events such as retirement, redundancy, and divorce.

Diagnosing the cause of tinnitus requires a detailed history and examination. Objective tinnitus, in which there is an externally detectable component, is most commonly reported in palatal myoclonus, temporomandibular joint abnormalities, and vascular abnormalities such as arteriovenous fistula and vascular bruits.

Subjective tinnitus, which is perceptible only to the patient, requires a detailed assessment of hearing but may occur without auditory impairment. Unilateral tinnitus requires full investigation to exclude underlying disease of the cerebellopontine angle, in particular an acoustic neurinoma. Bilateral tinnitus with evidence of a sensorineural hearing loss is associated with presbycusis, endolymphatic hydrops, vascular labyrinthine lesions, and hearing loss induced by noise—including gunfire, leisure activities, industrial exposure, and blast injury.

In Barr's classic work of 1886 characterising hearing loss induced by occupational noise, tinnitus was rare, in contrast with today.⁸ Tinnitus resulting from head injury and direct mechanical aural injury is well recognised. The onset of tinnitus may be related to aural syringing, even without any obvious trauma. Tinnitus has also been reported after whiplash injury, electric shock, otic barotrauma, and surgical intervention, particularly stapedectomy.⁹ It has been reported as a side effect of many drugs but is important, particularly medicolegally, when associated with drugs with a recognised ototoxic effect—for example, aminoglycoside antibiotics, salicylates, and loop diuretics. Middle ear pathology is rarely associated with tinnitus but may enhance an underlying tinnitus caused by cochlear dysfunction.⁹

Tinnitus cannot be substantiated or measured objectively. Matching and masking of the perceived sound, by standard audiometric techniques, can be carried out, but this is subjective. A hearing loss may be defined, but the complaint of tinnitus cannot be directly correlated with the type or severity of any associated hearing impairment. Abnormalities of the acoustically evoked magnetic fields of patients suffering from tinnitus have recently been reported as abnormal, but this is not a consistent finding.¹⁰

The main management of tinnitus is medical and comprises psychological, pharmacological, and prosthetic considerations. The treatment of the psychological aspects of tinnitus are of paramount importance in reducing the distress caused by the symptom.³ Previous negative counselling, such as the suggestion that a patient must learn to live with the symptom and that there is no treatment or cure, must be countered by an explanation of tinnitus together with reassurance that most cases improve with time.² Sinister cerebral lesions—a commonly expressed fear in this condition—must be excluded. Cognitive therapy seems to offer the best psychological approach. Several studies have documented psychiatric morbidity³ and suicide¹¹ in patients with tinnitus, yet many doctors may fail to recognise the association. Formal psychiatric referral must therefore be considered. In addition, much support may be obtained through lay counselling—for example, that provided by the British Tinnitus Association.

A hearing aid may improve the patient's hearing, lessen the attention the patient gives to hearing problems, and mask

tinnitus by amplifying desirable environmental sounds.² Without counselling and continued support, however, hearing aids alone are not effective.³ Furthermore, despite anecdotal evidence, controlled studies of tinnitus maskers have failed to show that they are better than placebo.³

Intravenous lignocaine results in the disappearance or amelioration of tinnitus, but no oral antiarrhythmic or anticonvulsant drugs have been found to be of benefit. Formal psychiatric assessment, with appropriate drug treatment, works in some patients. Benzodiazepines have been the drugs of choice in anxious patients, but they may make a depressed patient with tinnitus worse. In view of the association between tinnitus and depressive illness, treatment with tricyclic antidepressants has been proposed: nortriptyline is effective, although trimipramine is not.³

Rarely, surgery for arterial stenoses, glomus jugulare tumours, and arteriovenous malformations is indicated, but there is no evidence that destructive labyrinthectomies or eighth nerve sections are of benefit. It is much better for patients with troublesome tinnitus to be investigated for a cause and managed by someone with a positive attitude

towards the condition, prepared to give an informed explanation and appropriate psychological or psychiatric support.

LINDA M LUXON
Professor of audiological medicine

Institute of Laryngology and Otology,
London WC1X 8EE

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Sickle cell disease: the case for coordinated information

Currently little is known of its epidemiology in Britain

Despite an estimated 5000 people with sickle cell disease in Britain, living mainly in large cities, few reliable data exist on its incidence at birth and its natural course.^{1,3} Lack of information has hindered the establishment of a comprehensive national policy to guide the development and evaluation of services for sickle cell disease.^{4,5}

The problems are exemplified by the issues surrounding neonatal screening. Estimates of the number of babies affected vary widely from 75 to 300 a year.^{1,2} Early diagnosis, combined with prophylaxis against pneumococcal infection, parental education, and adequate follow up reduce early childhood mortality in sickle cell disease.^{6,7} This has led to calls for the introduction of a comprehensive neonatal screening programme in Britain.⁸

Locally, decisions on whether and how to screen neonates for sickle cell disease have largely depended on the commitment of individual people, with little central support in terms of policy direction or resources. This has resulted in differing approaches nationwide; in contrast, the need for a national neonatal screening programme for phenylketonuria, which detects about 90 affected infants a year, is widely recognised.⁹

Infants with sickle cell disease are born mainly in inner city areas with large populations at risk. Universal screening may be appropriate here, whereas selective screening may be more appropriate in districts with a low proportion of infants at risk.^{10,11} Despite this some districts that might most cost effectively adopt universal screening operate selective screening and vice versa. To facilitate a consistent approach to screening across the country, assessment of the cost effectiveness of different approaches to screening in relation to the proportion of the population at risk is needed. Account also needs to be taken of the coverage of screening programmes: selective screening may miss up to 15% of infants at risk.¹² Both selective and universal programmes will succeed only if the organisation and resources are available to ensure adequate follow up of affected children.^{2,13}

Inconsistent policies between adjoining districts with large populations at risk result in failure to detect all affected infants. For example, infants born in a district with a policy to screen resident infants six to 10 days after birth but living in a district that practises cord blood screening will remain untested. Several health authorities have already made substantial out of court settlements for failing to diagnose sickle cell disease neonatally or antenatally.

Deficiencies in our knowledge of the number and distribution of affected births are mirrored by ignorance of the natural course of sickle cell disease in Britain. Few figures on mortality or survival are available.³ This has been highlighted by the recent debate surrounding the use of bone marrow transplantation in sickle cell disease.¹⁴ Without accurate information on the long term outlook for patients who receive comprehensive supportive care it is difficult to judge the role of bone marrow transplantation, which offers a possible cure but carries an estimated 5-10% risk of death and serious morbidity related to the procedure. This dilemma is likely to recur as other new approaches to the treatment of sickle cell disease become available.

These issues show the need for a clear policy on sickle cell disease in Britain based on reliable data. We need coordinated information to define the size of the affected population and the natural course of the disease if we hope to develop and evaluate health services for this group of patients.¹⁵ Long term follow up of affected infants identified by neonatal screening could provide the foundation for a national cohort study along the lines of that funded by the Medical Research Council and the Overseas Development Administration in Jamaica, which, in a different setting, has provided invaluable information on the epidemiology of the disease.¹⁶

There is now an opportunity to address these issues. The Standing Medical Advisory Committee is due to report soon on the care of patients with haemoglobinopathies, and this should help to develop a national policy for sickle cell disease. A government that is committed to addressing the health