The suppression of palatal (or intra-tympanic) myoclonus by tinnitus masking devices
A preliminary report
by

Summary
Twelve cases of well established palatal myoclonus with objective clicking tinnitus were treated with tinnitus masking techniques. After a few months, three patients became completely symptom free, one for five years now. A further four patients continued to get periods of relief from their clicking sounds with continuous masking, and four patients found the distracting effects of the white noise helped them. The mechanism of palatal myoclonus is discussed with reference to the part played by masking therapy in this condition.

Introduction
Myoclonus causing an irregular clicking sound in the ear has been regarded as a rare event. Since its first description in the 19th century, it has remained a clinical curiosity. The aetiology remains obscure and its management has proved difficult. The possibilities of a hypersensitive reflex arc involving the mandibular nerve have been postulated. Many patients have coexisting psychological symptoms, and parallels have been drawn with tic and other neurological disorders (De Jong, 1950). Histological brain stem changes have been described in association with myoclonus (Nathanson, 1956) but most theories are unsupported by pathological data.

Whatever the aetiology, the presence of an audible clicking noise, sometimes together with discomfort in the ear or throat, is distressing and distracting to the sufferer. The accidental discovery that these sounds could be controlled by white noise masking occurred when a patient with coexisting high frequency cochlear tinnitus and palatal myoclonus was fitted with a tinnitus masker. She reported a dramatic improvement, with the clicking noise being completely abolished after two weeks' masking (Patient 1, Table 1).

This pilot study discusses 12 cases of palatal myoclonus in which there were audible irregular clicking tinnitus sounds in the ear. They presented at the tinnitus clinic at University College Hospital, London, which has been seeing over 300 new patients per year since 1977. The use of wide band masking noise in this condition and its possible mode of action is discussed.

Patients and methods
In each case the patient has a full ENT history and examination. The ear is examined under the microscope with times 40 magnification. This is important because, where the intra-tympanic muscles are involved, very small movements of the ear drum may be seen.

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<table>
<thead>
<tr>
<th></th>
<th>Age</th>
<th>Symptoms</th>
<th>Duration</th>
<th>Masker fitting</th>
<th>Effect of masking/counselling</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>NK</td>
<td>Clicking (L) ear 80/min.</td>
<td>9 months</td>
<td>Monaural</td>
<td>Rate slowed after two hours. Clicks abolished after 14 days</td>
<td>Return of symptoms after three years' freedom without masking. Now controlled with masker. Symptom-free at two years without masking.</td>
</tr>
<tr>
<td>2</td>
<td>SN</td>
<td>Rapid clicks (R) ear. Transient vertigo.</td>
<td>3½ years</td>
<td>Monaural</td>
<td>Control at two months. Stopped masking after three months.</td>
<td>Free for six months without masking.</td>
</tr>
<tr>
<td>3</td>
<td>ZB</td>
<td>Clicking (L) ear with fluttering sensation 150/min.</td>
<td>3 months</td>
<td>Binaural</td>
<td>Immediate slowing of clicks. Symptom-free at two months.</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>PM</td>
<td>Bilateral clicking in ears &gt;150/min.</td>
<td>4 years</td>
<td>Binaural</td>
<td>Immediate response. Symptom-free at four weeks.</td>
<td>Fixe for five years without masking. continued use of bilateral maskers for three years.</td>
</tr>
<tr>
<td>5</td>
<td>PB</td>
<td>Bilateral clicks. Laryngeal myoclonus. Vertigo.</td>
<td>5 years</td>
<td>Binaural</td>
<td>Rate slowed after four weeks. Symptom-free period 2-3 weeks.</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>MF</td>
<td>Rapid (L) ear clicks.</td>
<td>10 years</td>
<td>Monaural</td>
<td>Non-compliant with masker. Control of clicking after four weeks masking.</td>
<td>Lost to follow-up. Occasional clicking returned after four months. Not using masks.</td>
</tr>
<tr>
<td>7</td>
<td>EX</td>
<td>Bilateral clicking 70/min. 100/200.</td>
<td>4 years</td>
<td>Binaural</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>RC</td>
<td>Rapid clicking in the throat.</td>
<td>9 months</td>
<td>Binaural</td>
<td>Clicking frequency reduced after eight weeks masking.</td>
<td>Intermittent use of masker.</td>
</tr>
<tr>
<td>9</td>
<td>JT</td>
<td>Bilateral ear clicks Clonus of floor of mouth. Vertigo.</td>
<td>2 months</td>
<td>Binaural</td>
<td>(L) ear symptoms and vertigo abolished at two months.</td>
<td>No effect on suprhyoid myoclonus. Continued use of masker. Partial voluntary control. Returned masker. Symptoms now uncontrolled after 18 months.</td>
</tr>
<tr>
<td>10</td>
<td>KL</td>
<td>Clicks in (L) ear high pitched tinnitus.</td>
<td>2½ years</td>
<td>Monaural</td>
<td>No effect at four months.</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>KL</td>
<td>Bilateral ear clicks &gt;100/min.</td>
<td>5 years</td>
<td>Binaural</td>
<td>Initially experienced reduced frequency of clicks after three months masking.</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>KH</td>
<td>Irregular clicks (R) ear 80/min.</td>
<td>6 months</td>
<td>Monaural</td>
<td>Rate slowed after six weeks.</td>
<td>Has not completed three months trial.</td>
</tr>
</tbody>
</table>
synchronously with the clicking sounds. Auscultation of the ear is performed with the stethoscope and also with an intrametral microphone and amplifier (Hazell, 1984). In those cases in which involuntary movement of the palate and/or suprahypoid muscles could be observed, the clicking sounds were synchronous with these movements.

Tympanometry and acoustic reflex measurements were made. The absolute compliance of the ear was measured with the impedance meter at its most sensitive setting. In several patients a 'flittering' change of this impedance value was noted (Fig. 1) and this is thought likely to be due to changes in intra-tympanic muscle tone (Klochoff, 1981). Vestibular tests were performed in two patients with symptoms of vertigo.

After confirmation of the diagnosis, patients were interviewed by an audiological scientist experienced in the fitting of tinnitus masks. At the first session, information and counselling were given and an impression was taken for an open earmould to be fitted to each affected ear. At a second interview, Viennatone AMTI masks were fitted to the patients. Where the symptoms were monaural one instrument was fitted, but two instruments were fitted for binaural symptoms. The instruments were set to the patient's comfortable listening level, or at a level sufficient to inhibit the myoclonus, whichever was the less. The output characteristics of the Viennatone AMTI and its use in masking sensori-neural tinnitus is described elsewhere (Hazell et al., 1985). Patients were instructed to wear the masks for gradually increasing periods during the day, but to remove them at night. At three weeks another interview was conducted to ensure correct use and fitting and to record changes in symptoms. A further counselling session was conducted four weeks later and the patient reviewed in the clinic at three months. Patients were encouraged to continue masking for a further three-month period where this was necessary.

Results

Table 1 documents the 12 patients in this study. There were an equal number of males and females, with an age range of 11—80. All the patients had clicking tinnitus which could be easily heard by the examiner, and was often heard by the spouse or other members of the family without amplification. The rate varied between 70 and 180 clicks per minute. The duration of symptoms was between three months and 10 years. Thirteen patients had extensive myoclonus involving the floor of the mouth, pharynx and larynx caused by contraction of the suprahypoid muscles. Two patients experienced transient vertigo, and in these ENG and vestibular function tests were performed which revealed a left canal paresis on caloric testing in one case but no abnormalities in the other. Auditory Brainstem Responses were also performed in these two patients but no abnormality was found. The compliance of use of the masking devices over the first three months was 90 per cent.

In four patients with only otological symptoms the clicking ceased after three months of masking. Three have remained completely symptom-free, in one case for five years. The fourth had a return of the clicks after three years, which were again abolished by using a masker. A further four patients continue to use their masks, as they persist with their condition.
TINNITUS MASKING DEVICES

their maskers, although their symptoms persist, as they preferred the distracting or masking effect of white noise to the clicking sounds. Two patients who initially obtained relief, subsequently found that their symptoms returned, after four months and two years respectively, but masking was subsequently ineffective in controlling the clicks. One patient had no response at all, although he did develop some voluntary control, unaided. One patient was lost to follow-up.

Discussion

The aetiology of myoclonus remains unclear. There are many theories. Brain stem disorders involving the inferior olive and cerebellum have been shown in post mortem studies of patients with palatal myoclonus (Nathanson, 1956) and the concept of removal of central inhibition allowing uncontrolled reflex contractions to occur was proposed by Ramsey Hunt (1933). Swanson et al. (1962) considered the most common cause was vascular, leading to infarction of areas in the brain stem, but in our patients no other neurological sequelae were noted. Hypersensitive reflex arcs have been described in myoclonus affecting other parts of the body (Swanson et al., 1962). A pathway transmitted via the mandibular nerve to the tensor veli palatini is described by Querry (1972). Klochoff (1981) describes a 'tensor tympani syndrome' due to increased tone in the muscles with impedance fluctuation and a number of variable aural and vestibular symptoms.

The condition of palatal myoclonus has been described after whiplash injuries to the neck, with posterior fossa lesions, multiple sclerosis, intracranial aneurysms and following ENT operations (Swanson et al., 1962). One case in this series (No. 5) had a facial tic as a child and another patient's mother suffered with palatal myoclonus which apparently ceased following eustachian tube catheterization.

The source of the clicking is likely to be due to breaking of surface tension as the walls of the eustachian tube open under the action of the peritubal muscles, as suggested by Palec and Simonton (1961). Recent work supporting this theory using sonotubometry demonstrated that clicks occurred simultaneously with eustachian tube opening (Slack et al., 1986). Lyons et al. (1976) proposed another source of clicking to be the rapid clonus of the tensor tympani muscle and demonstrated changes on impedance audiometry resembling tensor tympani reflexes. It must be remembered, however, that any phenomenon creating a sound in the ear will produce an apparent change of compliance on an impedance meter, provided it contains low frequencies, and provided the impedance meter depends on measuring alterations in the intensity of a probe tone applied to the ear. It is possible that the middle-ear muscles fibrillate with the larger peritubal muscles in most cases it is unlikely that the tensor tympani and or stapedius muscles are producing the clicking effect on their own. The presence of vertigo in certain patients may be due to stimulation of the peripheral vestibular mechanism by movements of the stylus secondary to tympanic muscle contractions, or may be generated centrally.

Many treatments have been tried, including anti-epileptic drugs, sedatives, psychotherapy and anesthetizing the nasopharynx (Rahko and Hakkinen, 1979; Palec and Simonton, 1961). Surgical intervention has involved a grommet to alter the dynamics of the middle ear (Kwee and Struben, 1972) (which often makes things worse) and division of the intratympanic muscles (Williams, 1980) or of the tensor palati (Ward et al., 1975). Alteration of muscle tone in the pharynx (e.g. by swallowing, yawning and valsalva manoeuvre) can inhibit the symptoms in some patients. White noise will increase the tone of the muscles involved in the acoustic reflex, possibly resulting in a less excitable state, and this might reduce spontaneous contractions. Impedance changes occur at white noise levels at 60-80 dB. above threshold, to the contralateral ear.

The tensor tympani and tensor palati share a common nerve supply, from the mandibular division of V, and alterations of tone in one muscle may indirectly affect tone in the other. Muscle groups not involved in the acoustic reflex or sharing the same nuclear innervation...
are unlikely to be affected by white noise masking. The effects of masking combined with regular counselling have been described in patients with subjective tinnitus (Hazzell et al., 1985) and the influence of different therapists in helping patients achieve habitation to cochlear tinnitus has been highlighted by Stephens and Corcoran (1985). White noise has been used as an adjunct to dental anaesthesia and has a mild tranquillising effect.

The two patients with uncontrollable symptoms are both young males (age 19 and 24) and now pose a difficult management problem. There were no clear predictors to identify responders, and in particular the duration of the symptoms had no bearing on the outcome. The patient with vertigo, however, found that the symptoms disappeared altogether. The one patient who had no benefit at any time learnt to partially control his clicking by relaxation and returned his mask.

Two patients in the group had had grommets inserted previously which had made the clicking sounds louder. To date none have been offered a tympanotomy for muscle tenotomy although this remains an option when all else fails. Spontaneous remission from palatal myoclonus was described by Litman and Hausman (1982) in a six-year-old and this must be borne in mind in assessing the results of any treatment of this condition.

Tinnitus masking techniques are well established in the management of sensori-neural tinnitus and are free from side-effects. Their use in the treatment of palatal myoclonus seems to be helpful in the majority of cases and deserves further investigation.

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References


Address for correspondence:

C. A. East,
Royal Ear Hospital,
Hunley Street,
London W1E 6AU.