Management of trigeminal neuralgia and persistent idiopathic facial pain

Management of these uncommon but troublesome disorders is better left to specialists.

- Recognition in primary care is crucial to ensure prompt referral.

Trigeminal neuralgia (TN)

This disorder presents as recurrent, unilateral, brief but severe, electric-shock-like pains in the distribution of the trigeminal nerve, abrupt in onset and termination and often triggered by innocuous stimuli.

It is not common, affecting 1-2 in every 1,000 people. Women are twice as likely to be affected as men.

Principles of management

- TN is extremely painful, and untreated is physically, psychologically and socially debilitating:
  - patients may avoid the triggers of eating and drinking, seriously impairing food and fluid intake.
  - TN therefore demands accelerated specialist referral for investigation and treatment.
- Good treatment begins with education of patients, explaining their disorder and the purpose and means of management.
- The objective in management, by medical or surgical means, is abatement of attacks and pain freedom. This is not always achievable.
- MRI is mandatory since classical TN and secondary TN (due usually to cerebellopontine angle tumour, AV-malformation or multiple sclerosis) may be indistinguishable by symptom presentation.
- First-line treatment is prophylactic (antiepileptic) medication.
- Acute therapies (opioids or other analgesics) have no place in management since attacks are very short-lasting.
- Severe exacerbations with anorexia and dehydration, due to pain triggered by eating or drinking, may require hospital admission for intravenous hydration and medication.

**Education of patients**

A patient information leaflet on trigeminal neuralgia is available as Supplementary materials #26.

**Key points** of information are:

- TN produces very characteristic, very severe, electric-shock-like pains:
  - along a nerve on one side of the face, usually in the cheek or jaw;
  - repetitively, in short-lasting bouts (up to 2 minutes), which:
    - occur daily for weeks or months but sometimes remit spontaneously;
    - usually start without warning, but can be provoked by light touch, wind, cold air, eating, drinking, brushing the teeth or speaking.
- The cause of TN is often not known:
  - some people have a blood vessel in close contact with and compressing the affected nerve: an MRI brain scan is required to show this;
  - however, there are other unknown causes.
- Specialist referral is therefore necessary.
- There are a number of treatments for TN, which often work well:
  - these are preventative medications, to be taken daily;
  - painkillers do not help;
  - occasionally, surgery is required, but as a last resort;
  - TN does not require dental treatment.

**Table 1. Drugs used by specialists in trigeminal neuralgia prophylaxis**

<table>
<thead>
<tr>
<th>First line:</th>
<th>these drugs:</th>
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<tbody>
<tr>
<td><em>carbamazepine 200-2400 mg daily</em></td>
<td>• reduce efficacy of oral contraceptives;</td>
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<tr>
<td><em>oxcarbazepine 600-2400 mg daily</em></td>
<td>• may induce hyponatraemia (especially oxcarbazepine): regular monitoring is advised;</td>
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<td></td>
<td>• may induce osteoporosis in long-term treatment: prophylaxis against this is advised</td>
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<th>Second-line (either as monotherapy or as add-on medication):</th>
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<tr>
<td><em>gabapentin 600-3600 mg daily</em></td>
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<tr>
<td><em>pregabalin 150-600 mg daily</em></td>
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<tr>
<td><em>lamotrigine 200-1000 mg daily (very slow up-titration necessary)</em></td>
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</table>
Preventative medications

A narrow range of antiepileptic drugs are effective, and used by specialists (Table 1). **Maximum dosages** may be necessary to achieve pain relief, and balancing efficacy against toxicity is difficult.

**Principles of drug prophylaxis**

- Dosages should be **up-titrated slowly** until pain relief is achieved or side effects become unacceptable.
- Patients established on medication may be taught to **titrate up and down**, according to symptom severity.
- **Combinations** may cause fewer side-effects because lower doses may be required of each drug.
- Treatment may be **slowly tapered** after complete freedom from pain, and discontinued in the absence of relapse.

**Other treatment options in medically refractory patients**

- **Neurosurgical treatments** are relevant when medical treatment with maximum tolerated doses achieve insufficient efficacy, but:
  - microvascular decompression (appropriate when neurovascular compression, not merely contact, has been demonstrated) carries a small risk of severe complications such as cranial nerve palsy or stroke;
  - gamma-knife and/or percutaneous procedures (balloon compression, glycerol injection, thermocoagulation or pulsed radiofrequency treatment) targeting the trigeminal ganglion are less invasive but probably less efficacious.

**Follow-up**

While every patient with TN requires specialist initial management, long-term follow-up once stable is appropriate in primary care.

- Patients should be educated on:
  - how to **taper medication** cautiously once pain freedom is achieved;
  - how to **reintroduce medication** by careful up-titration if/when pain returns.

**Persistent idiopathic facial pain (PIFP)**

Previously termed “atypical facial pain”, this disorder presents as dull, aching or nagging, **poorly localized facial and/or oral pain**, which recurs daily for >2 hours over >3 months. Only rarely are there electric-shock-like pain attacks as in trigeminal neuralgia.

PIFP is rare, mostly affecting younger women, but it can start at any age.
Principles of management

- PIFP is painful, and can be physically, psychologically and socially debilitating.
- It is often difficult to manage, often has comorbidities, and usually requires specialist referral in the first instance.
- Good treatment begins with education of patients, explaining their disorder and the purpose and means of management.
- Freedom from pain is difficult to achieve: the objectives in management, by medical, physical and/or psychological therapies, are reduction of pain intensity and developing patients’ coping mechanisms.
- Treatment is prophylactic: acute therapies (opioids or other analgesics) have no place in management of PIFP.

Education of patients

A patient information leaflet on persistent idiopathic facial pain is available as Supplementary materials #27.

Key points of information are:

- PIFP is most often a constant, dull, nagging or aching pain in the cheek and lower jaw. Rarely there are electric-shock-like pains also.
- There are no specific triggers.
- The causes are unknown.
- There are no tests to confirm the diagnosis.
- Preventative medications, taken every day, are the best treatments for most people with PIFP:
  - these medications are more commonly used as antidepressants, but are very useful against chronic pain disorders even in people who are not depressed;
  - painkillers are unhelpful and, if taken too often, are likely to make things worse.

Table 2. Drugs used in prophylaxis of persistent idiopathic facial pain

| First line: | • intolerance is reduced by starting at a low dose (10 mg) and incrementing by 10-25 mg every 1-2 weeks; |
| • amitriptyline or nortriptyline, 10-100 mg at night | • nortriptyline has fewer anticholinergic side-effects but less good evidence of efficacy |
| Second line (either as monotherapy or as add-on medication): | • gabapentin 600-3600 mg daily |
| • pregabalin 150-600 mg daily |
Preventative medications

Drugs with some efficacy are shown in Table 2. **Maximum dosages** may be necessary.

- Use of drugs off-licence rests on individual clinical responsibility.

Principles of prophylaxis

- Patients receiving medication more often used as an antidepressant should be **advised of this, and why**; otherwise, they may default on finding out.
- Dosages should be **up-titrated slowly** until pain relief is achieved or side effects become unacceptable.
- **Combinations** may cause fewer side-effects because lower doses may be required of each drug.

Follow-up

While every patient with PIFP requires specialist initial management, long-term follow-up once stable is appropriate in primary care.