

TRIGEMINAL NEURALGIA AND CHRONIC FACIAL PAIN

Information for Patients at The Spine Surgery London

Introduction

There are many causes of chronic pain affecting the face. These may range from anything as common as simple tooth decay to some of the rarer conditions covered in this booklet.

The aim of this booklet is to provide information for those suffering from some of the more severe and persistent forms of chronic facial pain. The conditions covered are Trigeminal Neuralgia, Glossopharyngeal Neuralgia, Post-Herpetic Neuralgia, Atypical Facial Pain, Temporo Mandibular Joint Disease, Cluster Headache, Atypical Odontalgia and Oral Dysaesthesia.

TRIGEMINAL NEURALGIA

What Is Trigeminal Neuralgia?

Neuralgia is a word meaning nerve ("neur...") pain ("...algia"). There are two "trigeminal nerves", one each supplying sensation to the right and left side of the face. They both have three branches (hence "tri...") which supply the forehead, cheek and jaw.

Doctors may sometimes refer to these branches as "divisions".

What Are The Symptoms?

The pain typically only affects one side of the face. It can be severe and is of an unpredictable, paroxysmal nature. It is often described as; "burning, stabbing, lancinating, electric, shooting". Whilst it is often mild, for many patients it can be experienced as a violent and excruciating pain. These severe pains typically only last for a few seconds. It more commonly affects the jaw and cheek area, though may cover the whole of one side of the face. There are frequently "trigger points" on the skin which if touched will provoke a violent spasm of pain. The pain can often be felt in the teeth. It may be provoked by cleaning the teeth, hot or cold fluids, wind, shaving, eating and speaking

Attacks may at first be intermittent with long periods of relief occurring between them. For many these attacks become more frequent and of longer duration. For some the pain is constant i.e., it repeatedly shoots all day.

It may begin in a rather atypical pattern and many patients find that they have had extensive treatment from their dentist or others before the diagnosis eventually becomes apparent.

Who Does It Affect?

It may affect people of all ages though very rarely occurs in children. Whilst it is relatively more common in the older age groups, it not infrequently affects young adults. It is twice as common in females as males and similarly it afflicts the right side of the face twice as frequently as it does the left. Very rarely does it affect both sides. There have been very few recorded cases of it affecting more than one member of a family, i.e. it is not hereditary. As many as 1 in 200 people may be suffering from, or have suffered, trigeminal neuralgia in the U.K.

What Causes It?

A mass of theories abound, though in less than 5% of cases is a clear cause to be found. This small group of patients are sometimes found to have tumours (these are nearly always benign), infections or other abnormalities pressing upon the nerve. Trigeminal neuralgia occasionally affects patients with established multiple sclerosis though it is virtually never their first symptom, i.e. if you have trigeminal neuralgia there is *absolutely no reason* to suspect that you may be developing multiple sclerosis.

For the vast majority of patients with so-called "primary trigeminal neuralgia" no clear cause has been established. It has variously been proposed that the underlying abnormality lies within the brain, the nerves inside the head, or, the nerve as it passes through the face. Currently one of the most popular theories is that a blood vessel, normal in all respects other than its position, presses upon the nerve as it emerges from the base of the brain inside the skull. Rather like a "slipped disc" pressing upon the nerves in the back will cause sciatica, this blood vessel (about the size of a biro refill) is thought to squeeze the trigeminal nerve (about twice the size of a biro refill) to cause facial pain.

However, it would be wiser for you to select treatments on the basis of their safety rather than on these theoretical considerations of origin.

How Is It Diagnosed?

There are no signs for the doctor to detect on examination as the nerves function normally. Similarly there is no test which can conclusively prove that the condition is trigeminal neuralgia. The diagnosis is entirely based on the recognition of the clinical symptoms – your description.

Care must be taken to rule out other causes of facial and oral pain. Problems with the teeth, in the mouth, in the ear, bones of the skull and in the glands around the mouth and face need to be excluded. Therefore careful examination of these areas is required. A brain scan may be performed in order to exclude those rare lesions inside the head which can cause pressure on the nerves. The blood vessels possibly involved in causing trigeminal neuralgia cannot accurately be seen on these scans and in true primary trigeminal neuralgia the tests will usually be negative.

What Medicines Are Available?

The most powerful medication is Carbamazepine (Tegretol). This is an old and well established drug which was developed primarily for use in epilepsy. It dampens the activity in excitable nerves and in this instance its effect is manifest on the trigeminal system. Carbamazepine may cause a rash and a number of other rarer side-effects which you may wish to discuss with your doctor. You will need to have a regular blood test. You may need to take large quantities of the drug. Its principal side-effect is drowsiness and in some people this results in insufficient being taken and pain breaking through. However, for the majority of patients Carbamazepine is safe and all that is required.

Other anticonvulsants have been tried and these include Lamotrigine. This drug has to be started at a low dose and gradually increased if the principal side effect of a rash is to be avoided. Gabapentin is another though this may be built up quite quickly. Sodium Valproate (Epilim) is another and may be used instead of or in combination with Carbamazepine. They may well be less effective though may be better tolerated, i.e. cause less drowsiness.

Baclofen is a muscle relaxant. Seldom will it work on its own though it may be effective in combination with Carbamazepine or its alternatives

None of these drugs are available over the counter and you will need a prescription from your doctor.

None of the standard pain killers, however strong, seem to work on trigeminal neuralgia and you should stop them as they will add to the cocktail effect of the medicines without contributing to the pain relief.

What Form Does Surgery Take?

There are many types.

Interrupting the nerve:

A number of different methods are used to interrupt the nerve impulses. Whilst this may relieve the pain it will nearly always leave an area of lost feeling or numbness on the face. The interruption can be made at one of three sites.

A. In the Periphery of the Face

The small branches of the nerve which pass through the cheek and forehead may be cut or frozen. This can usually be carried out with local anaesthetic. A cut is made on the inside of the mouth or a needle passed to the nerve from the inside of the mouth.

The procedure will always result in an area of numbness which upon recovery is usually accompanied by the return of pain. It is a safe treatment in that very few patients come to harm during the procedure. There are three principal problems.

Firstly the effect may be short-lived. Secondly the numbness may lead to a different and sometimes worse form of pain. Called "anaesthesia dolorosa" this numb pain may be even worse than the original problem. Treatment is very difficult. It is a rare though very troublesome complication. Finally, it can only be used if the pain is confined to a small area of the face.

B. At the "Ganglion"

This lies just inside the bottom of the skull. It is a swelling on the nerve, the size and shape of a split broad bean. It contains the cells that supply the nerve fibres of the trigeminal nerve.

In this technique a needle is passed via the cheek up through a small hole in the base of the skull to reach the ganglion. An X-ray is used to guide it. The ganglion may then be injected with alcohol or glycerol, interrupted by heat ("thermocoagulation") or cold ("cryotherapy"), or compressed with a small inflatable balloon positioned at the tip of the needle. These procedures are carried out with the patient partially awake. The heavy sedation used means that the procedure is usually well tolerated though the surgeon is able to confirm by stimulation that the needle has reached the correct place. It can be performed as a Day Case or with a simple overnight stay in hospital.

A broader area of the face may be treated with this technique than can commonly be achieved by interrupting the nerve in the periphery of the face. Again there is usually a significant numbness affecting the face though this is often not as complete as when the nerve is cut. Anaesthesia dolorosa may also complicate this procedure.

There are a number of additional risks. As the needle penetrates the skull complications affecting the brain may arise. This occurs in much fewer than 1% though meningitis, brain abscess and brain haemorrhage have all been recorded. The result of these unusual events may be death or permanent disability, (mental or physical handicap). Further, some patients have found the procedure extremely unpleasant though with good sedation it is fairly well tolerated. Indeed, in those patients in whom the pain returns many are content to come back for a second procedure. Finally, in those patients where primarily the forehead is affected it can be difficult to position the needle into that part of the nerve and thus a large area of numbness is created before the pain is relieved.

C. The Nerve Trunk

This is the portion of the nerve which emerges from the brain and extends to the ganglion. It is approximately 1 cm in length and about twice the width of a biro refill.

In order to undertake this procedure a formal operation is required under general anaesthesia. An opening at the base of the skull is made just behind the ear. Using a microscope the nerve is identified as it passes between the brain and the inside of the skull. A cut is then made in the nerve in order to divide the fibres that supply feeling to the part of the face which is affected by pain. This inevitably results in some numbness though often less than might be anticipated. The operation involves a hospital stay of between five to ten days and you will usually be off work for a period of some six weeks.

The result is usually lasting relief as the numbness tends not to recover, i.e. it frequently provides for a permanent cure.

The principal disadvantage of this procedure is the need for an operation in such a strategic area of the brain. Even in the most expert hands and with the greatest possible care the risk to life and limb exceeds that of procedures performed more peripherally.

The exact risk will depend upon the age and general health of the patient but the incidence of death or serious disability (mental or physical handicap) has been 1% or less in most reports.

In addition to the risk to life and limb the adjacent nerves coming from the brain stem may be damaged. These include those supplying hearing and balance, i.e. the patient may suffer unsteadiness or deafness in one ear. More rarely weakness affecting the face on the side of the operation or difficulty in swallowing may be encountered. This damage to adjacent nerves has arisen in something less than 5% of cases. A not infrequent complaint has been pain around the wound behind the ear. This usually settles though can be a trouble for some months. Again anaesthesia dolorosa may follow this procedure.

A technique of "Stereotactic Radiosurgery" or "Gamma Knife" treatment has been developed. This uses a highly focused beam of radiation to interrupt the nerve just behind the ganglion. It has shown promising early results though remains very much under evaluation. It has the disadvantages that the benefits are delayed and that numbness may result bringing with it concerns about future anaesthesia dolorosa. Additionally there are the long term concerns about irradiation.

Decompression of the Nerve

The operation of "neurovascular (nerve vessel) decompression" was developed in a bid to provide pain relief without nerve damage and consequent numbness or risk of anaesthesia dolorosa.

In this procedure the blood vessel found compressing the nerve as it emerges from the brain stem is mobilised free from the nerve and a soft pad placed between them (see earlier section entitled "What Causes It?"). In 90-95% of patients immediate pain relief is provided. Most published accounts have shown that about 75% of patients will still be pain free some three to five years later. Whilst recurrence may still occur after 5 years of relief rarely does the procedure produce loss of sensation and no incidence of anaesthesia dolorosa has been reported.

Of course the disadvantage of this procedure is that it again requires a formal operation and the risks attendant on this are the same as those described earlier in Section C. The same opening is made and again you are in hospital for five to ten days.

In perhaps 10% to 15% no blood vessel is found or one that is very small and thought not to be significant. In these circumstances I would routinely partially section the nerve so that some relief is gained by the operation. (see Section C) In some special circumstances patients elect not to have this done in the event of a negative exploration and can then expect to still have pain thereafter. Some surgeons only offer a decompression if the MRI shows a blood vessel. However sometimes these show a vessel which in reality is not there and on others no vessel when one in fact is present i.e., MRI scans of the small, deep vessels may yield false positive and false negative results. Whilst the accuracy of MRI is increasing I prefer that you give me consent to section the nerve if I feel it necessary during the operation.

What Should I Have Done?

If your troubles are controlled by medication then this is certainly the best mode of treatment. Broadly speaking, two groups of patients go forward for other treatments; those in whom the tablets never gain control of the pain and those in which the medications cause intolerable side effects. In these eventualities you will need to give careful consideration to which treatment you would wish to have. This is something you will need to discuss with a specialist.

A sensible guiding principle is to put aside the theories relating to the cause of trigeminal neuralgia and concentrate on weighing the potential benefits and risks of the various treatments. How likely is it to work, for how long will the benefit last and to what risk does it expose me as an individual?

A general recommendation has been that for young, healthy patients the neurovascular decompression operation is the most appropriate. It provides pain relief without numbness and usually does so in a lasting manner. For those who are unwell, are elderly or frail, cannot afford the time or are unwilling to take the risk the usual suggestion is to treat the condition with simpler techniques (see sections A&B). The benefits will usually be of lesser duration and will usually then come with an associated numbness. However the procedure can be repeated and subsequently one can go on for more definitive surgery.

You will need to consider these factors carefully and should feel under no pressure to accept one or the other as you will receive good support whatever your decision. All the treatments are available.

There are two other guiding principles when considering intervention beyond medication that I feel are relevant.

Medications can cause serious side effects especially when taken in high doses and for long periods. People may even suffer disastrous complications to blood and liver function and some die each year from the anticonvulsant drugs listed above. In many studies patients who have had interventions reflecting on their time on medication wish they had had the surgery sooner. "Coming off the drugs gave me my life back"

By contrast there are those who rue the day they had surgery. Complications may occur and the operation may fail. Both misadventures may arise in the same sole and their lot is a very unhappy one. In addition, however big the operation and however good its statistics are, no procedure can be said to cure the condition. There is always a chance it may fail even after years of relief. The treatments, medical or surgical, should be considered as containment not cure.

Finally, I feel I have given a balanced view. However, you may well benefit from talking to others and obtaining a second opinion. The latter never bothers me. So, if you are uncertain do not be afraid to ask me to arrange extra help. As usual the Brain and Spine Foundation offer helpful advice though I should point out that I was heavily involved in the writing of their booklet on the condition. (The Brain and Spine Foundation contact details are given at the end of this guide)

GLOSSOPHARYNGEAL NEURALGIA

This is a much rarer condition and may often be confused with trigeminal neuralgia. The pain is of an identical nature though affects a different area. Glossopharyngeal neuralgia is usually felt at the base of the tongue, the back of the throat and may radiate to the ear. It affects only one side of the head. The same areas are affected by hypersensitivity and the pain may therefore be triggered by touching the ear or swallowing.

The investigations of this pain will be along the same lines as that for trigeminal neuralgia. The medicines used in its treatment are also the same as those used in trigeminal neuralgia. Because of its rarity surgical treatment for those patients in whom the pain breaks through the medications is less established. You will certainly need to discuss this carefully.

POST-HERPETIC NEURALGIA

What Is It?

This is a form of chronic facial pain which follows an attack of shingles on the face.

Shingles is caused by the chicken pox virus which most people get as a child. Also known as the herpes zoster virus, shingles usually occurs in older people when the organism has been lurking in the body for many years and suddenly attacks a single nerve. It will most often affect a nerve supplying the skin of the trunk though it affect a nerve supplying the head and face. It will never cross from one side of the face to the other.

The illness follows a common pattern. It begins with a dull pain in the skin over the face to be followed within a few days by the eruption of a weeping rash. During this acute phase the pain may be very severe though it usually then settles. It will usually be over within a few weeks. The attack may be shortened by the use of certain medications like Acyclovir. In some patients the rash may leave scarring though for the vast majority the pain settles and the rash goes away.

Rarely, the pain may continue and it is this chronic pain which is called post-herpetic neuralgia. The pain is usually of a burning, aching or throbbing nature and may often be associated with extreme tenderness. Rubbing of clothes or wind blowing against the face may be unbearable. Oddly, firm pressure may be well tolerated. Despite the extreme tenderness in the affected area ordinary sensation may in fact be blunted.

As with many chronic pains the symptoms may be worsened by physical or mental stress. In addition sleep is often interrupted as bedclothes brush against the face.

Who Gets Post-Herpetic Neuralgia?

Anyone who has had shingles may get post-herpetic neuralgia though in reality it is very rare for young patients to develop it. Probably 100,000 people per year develop post-herpetic neuralgia on some part of the body, though only a small percentage of these patients will have it affecting the face.

What Is The Cause Of Post-Herpetic Neuralgia?

The virus damages the nerve and as a result the area supplied by that nerve goes partly numb. However the damaged nerve pathways then start to generate pain. Precisely how this comes about is unknown.

What Treatments Are Available?

The effects of treatment are much better when given early. If after an attack of shingles on the face you still have pain a month after the rash has settled you should go to your doctor immediately.

Ordinary painkillers usually have little effect and so may in fact only waste valuable time.

The principal drug used for this condition is Amitriptyline which is a drug more commonly used for depression, though has very powerful effects on certain forms of nerve pain.

You may need to take the Amitriptyline for two or three weeks before it provides relief.

Other drugs such as creams and lotions, or the anticonvulsant medication Carbamazepine (Tegretol) may also be used.

Surgical procedures to cut the nerve, such as those used in trigeminal neuralgia, are always ineffective.

OTHER CAUSES OF CHRONIC FACIAL PAIN

These conditions are considered together because they have many common features. They are not necessarily rarer than the conditions considered in the previous sections.

Temporo-mandibular Joint Dysfunction

This condition is sometimes also called “facial arthromyalgia” or “Costen’s Syndrome”.

The pain is in the form of a dull ache which affects the jaw and muscles over the side of the face. It may also cause clicking of the jaw joint and difficulty in opening the mouth because of spasm in the jaw muscles. The pain may extend over the side of the head and down into the neck. Often pain may be felt in the ear where there may also be a sense of fullness or buzzing. Sometimes dizziness may occur.

The cause of this pain is unknown though for some the problem relates to disease in the jaw joint. It may also occur when the teeth do not align properly. This can happen when teeth have been lost or if dentures do not fit well. The treatment must therefore begin with a careful assessment from a dental specialist.

Atypical Odontalgia

Odontalgia is a medical word for toothache. Atypical odontalgia is a severe discomfort in the teeth or a tooth socket in the absence of any of the usual dental causes. Oddly the pain may be made worse if dental treatment is pursued though it is very common for teeth to have been removed by the time the diagnosis becomes clear.

Oral Dysaesthesia

This is a group of problems which include a burning or altered sensation in the tongue and gums. It may be associated with a nasty taste. Some patients have a sense that they produce too much or too little saliva. Dentures, crowns and bridges may become uncomfortable such that it is impossible to wear them despite all attempts to modify the shape.

Some sufferers may develop a “phantom bite” whereby they feel the teeth do not meet properly.

How Are These Conditions Diagnosed?

As with trigeminal neuralgia the diagnosis is made on clinical grounds. That is to say there is no specific test that will prove the existence of any of these conditions.

Very careful examination of the mouth and the associated structures will be required by a relevant dental or medical specialist. Often investigations such as X-rays or scans may be performed, though these are to exclude other conditions and may well be normal. On other occasions abnormalities may be shown which are not relevant to your condition. The pain may be very severe and there is a temptation for patient and doctor alike to clutch at any anomaly seen on an X-ray or scan. Blind alleys of treatment may be pursued with failures then increasing the distress of all concerned. Usually this is not now a problem and once the diagnosis has been established and other problems excluded treatment may begin.

What Causes The Pain?

It is true to say that a full understanding of these conditions is not yet available. However, it is clear that these pains are not imagined and are very real. Whilst the trigger in many cases is unknown the pain may be mediated by cramped muscles and dilated blood vessels. As with many chronic pain states they tend to be exacerbated by tiredness, intercurrent illness and stress.

What Treatments Are Available?

Simply to have an accurate diagnosis and some understanding of the condition often leaves many sufferers much better able to cope with their pains. Treatment is based on medications often supported by “pain management techniques” (see below). There is no operative solution to these problems though occasionally allied pathologies may require treatment. You may need to see several specialist before the correct advise is found.

What Medications Are Used?

The most frequently used medications are anti-depressant drugs. These are NOT prescribed because it is felt that you are necessarily depressed, but because these drugs are now known to have a specific effect on certain forms of chronic pain. If the pain has made you depressed then they will of course help that problem too. The most commonly used medicine is Amitriptyline or Nortriptyline. They will often need to be taken for several months before they become fully effective, and they may need to be taken for a year if the course is to be of lasting benefit.

These are “safe” drugs which very seldom have serious side effects though frequently can cause mild drowsiness, a dry mouth and occasionally constipation. They are best avoided if you suffer from certain conditions such as glaucoma, or prostate problems.

The effects of drowsiness and constipation may be worse in the elderly. Your doctor will advise you.

Other related drugs may be used if side effects are encountered or anticipated. In addition if the pain is severe at night occasional night sedation will be required.

What Are Pain Management Techniques?

Pain can feel worse if you feel it has become out of control and if stress develops. These pain management techniques aim to teach you methods of coping better with the pain when it breaks through the treatments. They aim also to help you better control stress and thus prevent this from exacerbating the pains. These methods are often taught by Psychologists. A referral to a Clinical Psychologist should NOT be taken as meaning that the doctor believes your pains are due to some psychiatric disorder. People with chronic pain of any source can gain benefit from such treatment.

The treatment sessions will usually consist of teaching you relaxation techniques and coping strategies for when the pain is severe. You may need to attend on several occasions so the idea is for you to learn the methods for yourself.

It is true to say that the use of these techniques varies very much from one specialist unit to another. However they are becoming increasingly popular and you will be able to discuss them with your doctor.

What Other Help Is Available?

There is a self-help group for Trigeminal Neuralgia sufferers in the UK run by Mrs. A. Conn, 27 St. Kenya Avenue, Hove, East Sussex, BN3 4PN.

There is an American Trigeminal Neuralgia Self-help Group who may be contacted at the Trigeminal Neuralgia Association, P.O. Box 340, Barnegat Light, NJ 08006, USA
Phone: USA - (609) 361 1014 Fax: USA - (609) 361 0982

There is a free information pack provided by the **Brain and Spine Foundation** which covers this problem at **www.brainandspine.org.uk**

The Brain and Spine Foundation is a charitable organisation and they would receive any contribution you can make with gratitude. They fund valuable research and education programs into the neurological disorders.

I am unaware of Self-help Groups relating to the other conditions covered in this booklet and would be pleased to hear from you should you find one.

Finally, always consider talking to your General Practitioner. They help people every day in making decisions about complex medical issues. They will also be able to place your problems within the context of any other health issues you have.

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