



The Patient's Journey Through Trigeminal Neuralgia

The IASP definition of trigeminal neuralgia (TN) is “sudden, usually unilateral, severe, brief, stabbing, recurrent episodes of pain in the distribution of one or more branches of the trigeminal nerve.”¹

Here's now one sufferer describes the pain: “Supper with friends. Candles and wonderful food. Suddenly my face is split apart—the bones feel as though they are shattering and the flesh raked aside by red-hot claws. I lean forward, the food falls from my mouth. The guests stare, concerned and appalled. I cannot speak to explain why the tears stream down my face. I cannot even swallow, my own saliva dribbling onto my plate. All I can do is try not to scream. If I look into a mirror I cannot believe that there is no sign of injury, no blood pouring out of my eye.”

Both descriptions highlight the key features of trigeminal neuralgia, but the second is far more graphic than the first. Only the patient's arresting report gives us insight into

the personal experience of TN pain, illustrating the suffering and fear accompanying the first attack, which many patients remember because of its dramatic onset.

TN has an enormous psychological impact, but few scholarly papers highlight the ways it can affect the quality of life.² TN is a neuropathic condition with a unique clinical manifestation;

it is also one of the few chronic pain conditions in which sufferers can be rendered 100% pain-free either with medications or surgery.³ For this

reason, correct diagnosis is crucial so that patients can then follow a generally acknowledged care pathway as soon as possible.^{4,5} Patients' verbal and visual descriptions provide a vividness and a level of detail missing from the generic medical criteria for classification. Paying attention to these accounts can help improve the speed and accuracy of diagnosis and appropriate referral.

This edition of *Pain: Clinical Updates* explores TN through the eyes, ears, and voices of patients alongside our clinical evidence-based guidelines. We collected these stories, descriptions, and images by means of focus groups, targeted emails, patient support-group meetings, and a photographic project.

To gather material for her book, *Insights—Facts and Stories Behind*

Trigeminal Neuralgia,⁶ orofacial pain specialist Joanna Zakrzewska invited patients with TN to attend focus groups in the United Kingdom and United States. Sessions were recorded and transcribed. Patients in the U.S. support group also emailed their stories to relate their route of diagnosis, their symptoms, and the impact of the condition on their lives and those of the people

closest to them.

Another important source of information was the collaborative art and medicine project *face2face*, based in London.

Photographer Deborah Padfield worked individually with a group of facial pain patients, including three with TN, to create images and audio recordings of their pain and its impact at three points of treatment: before, during, and after pain management. This arc of time allowed the images to represent changes the patients had experienced in their perception of pain. The images also elicited significant narrative and emotion for the discussions.

The Long Journey to Diagnosis: A Common Story

One of the greatest problems clinicians and sufferers face is the difficulty in obtaining a definitive diagnosis. TN is rare, and clinicians may only see three or four cases in their entire



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Because of the rapid advances in the medical sciences, the publisher recommends independent verification of diagnoses and drug dosages.

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careers. Although the main diagnostic symptoms of TN, as defined by IASP¹ and the International Classification of Headache Disorders (ICHD),⁷ appear to be clear, there is a considerable overlap between these symptoms and those of other trigeminal autonomic cephalalgias, as well as unilateral persistent idiopathic facial pain, temporomandibular disorders, and dental pain.⁸ Drangsholt and Truelove highlight the diagnostic problems, suggesting that the majority of patients attending their tertiary facial pain clinic had previously been misdiagnosed and had undergone irreversible dental treatment, even when they had presented with classical features of TN.⁸

Dental disorders, such as periapical abscesses, can produce pain identical to that of TN. Dentists and patients are both correct to consider these extremely common disorders initially. However, dentists should not carry out irreversible procedures if a patient's clinical history, examination, and investigations are equivocal or inconsistent. On the other hand, primary medical practitioners have learned very little about the causes of facial pain and may tend to overdiagnose TN because it may be the only diagnosis with which they are familiar.

A recent study on the prevalence of facial pain in Dutch general practices found that after experts had reviewed the notes and asked the physician supplemental questions, 48% of the cases had been wrongly diagnosed as TN. The positive predictive value was 57.3% for trigeminal neuralgia compared with 83.7% for postherpetic neuralgia.⁹

To take into account the need for more than one episode of pain, Koopman et al⁹ included only those patients who had at least a one-year history, suggesting that time helps improve the likelihood of a correct diagnosis.

Drangsholt and Truelove⁸ showed that symptoms caused by non-nociceptive triggers sharp, shooting pain lasting only a few seconds, and the inability of opioids to adequately reduce pain provides an 81% probability that a case is TN rather than pulpal or temporomandibular pain.

The following testimonies recount typical stories:

"I went for my appointment to a dental consultant. He examined me and did a full mouth x-ray. He seemed really confident that I definitely did NOT have TN, and that it is all dental."

When patients come to realize they have received an incorrect diagnosis, they begin the long process of consulting ear-nose-throat and maxillofacial specialists—each of whom may also be unfamiliar with this rare condition—before finally reaching a neurologist or neurosurgeon experienced in managing TN. Patients with private insurance will often quickly visit numerous health-care professionals.

One patient encountered a neurologist who recognized the condition but not its severity or the urgent need for treatment: *"My neurologist gave me a thorough examination and advised that he definitely thought it was TN, and the next step would be an MRI scan.... I have to wait another six months for a scan. The thought of being in pain every day before I even get to the next step of treatment is really getting me down.... I got my scan results and, no surprise, they haven't found anything. The neurologist wrote to say he didn't need to see me again."*

During this process, patients may have run through the entire range of painkillers—nonsteroidal anti-inflammatory drugs, opioids, and tranquilizers—none of which will have any effect on the pain. It is not surprising that some feel they are going mad and become increasingly lonely. *"Every doc-*

tor I saw made me feel as if I were crazy. There were some who would just sit and look at me. Many more had me feeling I was alone in this—that there was nothing out there that would ever help me. I was going to spend every day of my life in this tortured state.”

Diagnosing TN Correctly

The principal way a health-care professional can make a proper diagnosis is to receive (rather than take) a careful history. This requires allowing patients adequate time to complete their opening statement.^{10,11} It is important to allow patients to tell their story in their own words, and as David Loxterkamp says, “This takes time—face time, time looking into their faces instead of a clock or computer.”¹² As William Osler said to his students a century ago: “Listen to the patient: he is telling you the diagnosis.”

Because no objective diagnostic tests exist for idiopathic TN, listening remains the only reliable tool. In contrast, examinations and x-rays improve the diagnosis of dental pain. As part of the diagnostic work-up, patients should have a magnetic resonance

imaging (MRI) scan or, at the least, a computed tomography scan to rule out TN secondary to tumors or other compressive causes and to reveal plaques indicative of multiple sclerosis. In classical TN, high-resolution MRIs will indicate the presence of neurovascular compression of the trigeminal nerve in the posterior fossa.

How the Diagnosis Affects Patients

Diagnosis will have a dramatic effect on patients. Patients are often relieved to learn that their pain has a “label,” but they may find it hard to accept that in some instances there is no cure—only good pain control and learning to live with the disorder. Some patients, but not all, will gain complete pain relief through surgery and will return to a high quality of life.

General practitioners and consultants who have not previously encountered TN often have difficulty in comprehending the physical suffering and emotional frailty of patients who present with this devastating disorder. Patients searching for information on the Internet sometimes add to their own distress; for example the

following patient, after reading two Internet definitions, said this:

“But the PROGNOSIS for TN—it was alarming! There was no good news anywhere in the descriptions of it, none at all: TN just got worse...the remissions got shorter, then ultimately disappeared entirely. The worst thing had to do with the rarefied nature of TN: Who else would have it? With whom could I share the horror or my fears?”

With the onset of the first attack, a patient suddenly must face agonizing pain for which there seems to be no relief, often leading to social isolation and loss of employment: “Isolation, depression, and fear of increasing pain are just some of the emotions I try to work through.”

During severe episodes, the sufferer is often unable to work, either because of the pain or because of the side effects of prescription drugs. Claiming insurance benefits can be difficult because of the rarity of TN and the uncertain course the disorder follows. In some countries, the benefits system is not structured to allow for variable periods of remission in which the sufferer may choose to work: “I was broke. I had been ‘let go’ from my job. I was so afraid I’d be fired that I returned to work too soon and kept having relapses... The \$237 a month I received from welfare wasn’t enough to live on, anywhere. I existed from day to day, never knowing where I would be sleeping that night.”

The severity and unremitting nature of the pain leads to depression: “It destroys and kills the life you once knew; the alternative name of ‘suicide illness’ is truly apt.” “[I] wanted to take my own life, but I looked at my husband who was desperately trying to help me fight this, yet feeling so useless, and I had to hold on.”

Even if they manage to control the pain, patients live in fear of its return: “Even now that I’m in remission, at the

Key features of trigeminal neuralgia as described by patients

- **Character and location of the pain:** “The pain was sharp, stabbing, electrical-type shocks adjacent to my nose in the left cheek.”
- **Duration of the pain:** “I was having more than 100 attacks a day. Most of them lasted 20 seconds to two minutes each. Sometimes a dull ache followed the spikes for 20 minutes or so. Other times the pain would disappear just as suddenly as it came on.”
- **Periodicity of the pain:** “I have had several periods of remission for which I am very grateful. My last round of pain lasted only six months and was fairly minimal compared to what I had been through before.”
- **Severity of the pain:** “The intensity is unlike anything I could ever imagine; the pain is so strong it steals your mind, it takes over all your thoughts. I prayed for death.”
- **Provoking factors:** “Forget eating or brushing my teeth. Speaking, at this point, would have been a cruel torment, electric sparks flying with each connection the tongue makes with any part of the mouth. After a while the pain didn’t even seem to be triggered; it just came on by itself as if it had a mind of its own.”
- **Impact of pain:** “I ended up getting dehydrated and losing quite a bit of weight.”

back of my mind, while crunching toast or cleaning my teeth is the thought 'better go slowly just in case.'" "The pain causes great fear of being out of control, and of course fear of the terrible pain."

Sufferers lose their confidence and control of life: "It is hell. It is the worst God-awful thing that has ever happened in my life. This pain had taken over my life."

Relatives find it hard to help: "My husband hates the disorder, he feels helpless and angry at the disorder."

Sufferers need empathy and support: "I really need some help, somebody who can understand, somebody's that's been through this problem before. People in my situation need a lot of understanding, a lot of attention, a lot of comfort as well."

Managing the Pain

Once a correct diagnosis has been reached, it becomes possible to start to control the pain. Again, careful listening on the part of the clinician is essential because a "preference

misdiagnosis" can occur.¹³ Patients may value the risks, benefits, and side effects of the various options very differently from the physician. The first approach is with drugs. Carbamazepine has long been the gold standard, but a wide variety of drugs are available; details of their use are available in publications by the Cochrane Collaboration,^{14,15} Clinical Evidence,¹⁶ and international guidelines.^{4,5} "While my Trileptal [oxcarbazepine] medication still effectively keeps the sleeping demons at bay inside my trigeminal nerves, life is good. Until the medication loses its effectiveness and we need to ratchet up the [dose] or start combining a number of them."

All these drugs have side effects, which in some cases preclude their use, and they tend to become less effective for pain control as the disorder progresses. "I began to have serious trouble playing my violin. I found I couldn't remember whole tunes; my mind would wander off in the middle of a tune, and I was getting feedback from my band-mates that my tempos were

much too slow." "I couldn't take the phenytoin or carbamazepine as I had a severe allergic reaction to both drugs, including a rash all over me, which looked like bad sunburn, swelling of my face and limbs, and peeling of the skin from the soles of my feet and palms of my hands."

It is important for patients not only to learn how to use their medications to maximum effect but to be allowed to control the dosage themselves. Providing patients with greater control reduces their fear, as "Anne's description of her trigeminal journey" shows.

The Surgery Option

When drugs no longer provide relief, a variety of surgical options are available. It is important to gauge patients' reactions to each option and take their preferences into account.¹² Options include percutaneous procedures at the level of the Gasserian ganglion, which are destructive. Balloon compression, glycerol injection, or radiofrequency thermocoagulation and Gamma Knife

Anne's description of her trigeminal neuralgia journey

"One of the first mental images I'd like to express visually is of trying to shut it out, to prevent 'it' from attacking me. I see a large armoire or big heavy doors and behind it the attacker waits, like lava from a volcanic eruption. ... The aggressive rocks overwhelm me, articulating lonely despair. They represent difficulty, a cruel terrain crushing my spirit. ... The rocks depict the isolation well, the feeling that you're being overwhelmed by this force of nature. ... I like the idea of a final hopeful image, incorporating flowers I have grown, especially roses. Different cultures have various interpretations for the meaning of flowers; roses usually symbolize hope."



Photographs by Deborah Padfield with Ann Eastman from the series *face2face*. ©Deborah Padfield.

surgery (at the level of the nerve root) provide pain relief in 70% of cases for as long as five years but may cause facial numbness.¹⁸

The most successful procedure, providing relief for 70% of patients for 10 years, is microvascular decompression of the nerve, a nondestructive procedure. This procedure involves major neurosurgery and is therefore not suitable for all patients; it carries a mortality rate of 0.5% and a 2% risk of hearing loss.¹⁹ However, satisfaction is high after this operation.²⁰ Patients' emails also reflect positive outcomes:

"I was a bit resistant about going for the surgery because I've never had major surgery, but now after the surgery, thinking back about the pain, I'm totally relieved."

"I was very pleased to be able to eat a sandwich, and some ice cream, within 1½ hours.... The TN pain had gone completely."

"I think people fear this surgery more than necessary. It's certainly not a minor procedure, but when you're hurting this badly, it's almost something one eagerly anticipates. I felt as if I got my life back. Once I got off the TN medication, my memory and energy levels gradually came back. It was like I was a 70-year-old who became a 36-year-old."

The series of images created during the *face2face* project reflect the successful journeys through surgery of three patients: Anne, Alison, and Chandra. The texts are taken from audio recordings during one-on-one workshops.

Finding Help

Joining a support organization can provide information that will allow the patient to work with clinicians to develop a treatment plan. Such organizations provide contact with other sufferers through online forums, books,^{6,21}

meetings, and conferences.²² Patients can provide one another with empathy, understanding, information, and help in interpreting some of the medical jargon they hear at their consultations. Participation in these groups can inspire greater confidence, which is so important in the road to recovery.¹²

"The local support group I found online is fabulous. What a treat to be able to sit in a group of total strangers and know I'm not alone. We all have some type of debilitating facial pain, and we all know what each is experiencing. That's been the biggest gift to me in this whole mess. Patient, kind, and lovely people who feel my pain. Literally."

Contributions

Joanna Gardner, a patient with TN, was for many years the email helpline supporter for TNA UK, and she drafted an initial article on this topic a few years ago. The patients' stories were

Alison's description of her trigeminal neuralgia journey

"I think for me it really captures the pain I used to feel if you were biting into an apple or something. It was just like needles were going into the roof or the side of your mouth ... sometimes it was fine and sometimes something bigger, like a blade stabbing."

"It was trying to get as much in as possible from the experience, the medication, the pins, the strawberry, it was just anything that I had that related to the thing. ... It's just a door, closure to it all. It was an important part of it. ... I love the strawberry being there in a different form."

"I couldn't see anything other than being stuck in that pain cycle. ... I was locked in a place with this pain and couldn't move forward. I think through coming here and having a look at a beginning, middle, and an end, it somehow moved me onto the next phase and psychologically I could look at it differently. I don't know, without this, how I would have moved."



Photographs by Deborah Padfield with Alison Glenn from the series *face2face*. ©Deborah Padfield.

Chandra's description of his trigeminal neuralgia journey

"Very often I've connected the pain to wires. I've blown a fuse as well and realized that this is the same problem that I have with my face. When I'm having a conversation this is what happens, when the wires touch each other like this, is when the pain is most severe. When they are apart, this is when I'm in bed and I don't feel any pain at all. This one where the wires are closer is when the problem starts, when I get up in the morning and when I try to brush my teeth."

"Now you can see the smile on my face with the way I've been speaking, I'm totally relieved believe it or not. ... I wish I had this surgery a long, long time ago. ... I'm very, very pleased with the surgery. I think I'm, well, I'm looking forward now to getting back to my normal life."



Photographs by Deborah Padfield with Chandra Khoda from the series *face2face*. ©Deborah Padfield.

obtained from their contributions to Prof. Joanna Zakrzewska's book *Insights*, from emails, and from forums of TNA UK. Prof. Zakrzewska undertook this work at UCL/UCLHT and received a portion of funding from the Department of Health's NIHR Biomedical Research Centre. The *face2face* project was a collaboration between Dr. Deborah Padfield and

Prof. Zakrzewska, along with pain-management patients and clinicians. The project received funding from AHRC, Arts Council England, UCH small grants awards, Derek Hill Foundation, and support from the National Portrait Gallery London, Paintings in Hospitals and the Menier Gallery, LAHF, UCLH, UCL, and the Slade School of Fine Art.

For more information on the project as a whole, visit www.ucl.ac.uk/slade/research/student/archived-research/mphil-phd-research/project-9. For information on a project developing the work further, "Pain: Speaking the Threshold," visit www.ucl.ac.uk/slade/research/staff/current-research/project-14.

Support for patients and additional resources for patients

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2. TNA US (Formerly the Trigeminal Neuralgia Association), Suite C, 925 NW 56th Terrace, Gainesville, FL 32605-6402, USA. Website: www.endthepain.org
3. Trigeminal Neuralgia Association Australia, PO Box 1611, Castle Hill, NSW, 1765, Australia. Website: www.tnaaustralia.org.au
4. Books published by TNA US: *Striking Back: The Trigeminal Neuralgia Handbook and Insights, Facts and Stories Behind Trigeminal Neuralgia*.

Upcoming Issues:

Marijuana for Pain • Integrative Pain Medicine

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The Challenge of a TN Diagnosis

By Mark T. Drangsholt, DDS, MPH, PhD

Trigeminal neuralgia (TN) is a unique disorder among pain conditions, with characteristic qualities of electric-shock-like pain lasting only a few seconds to two minutes, usually triggered by light touch, movement, or air currents. At the same time, it is puzzling why this condition is so commonly misdiagnosed; indeed, the diagnostic delay for TN is measured in *years*, not months.²² This delay has been apparent in the literature for decades, and despite education, articles, and pleas from authors, there has been no decrease.

How can this be? Are clinicians really so ignorant as to miss this condition? The consequences of misdiagnosis, as Joanna M. Zakrzewska's article shows, can truly be disastrous and inflict untold suffering.

Much of the problem lies in the numerical challenge of spotting a rare disorder among far more common disorders. The incidence of TN in the general population is 3.4 to 5.9 per 100,000 people, or about 0.005%.²³ In people seeking care, the number can be higher, but not significantly so. A primary care physician, general dentist,

or nurse practitioner may see one case every five to 10 years—that is, if correctly diagnosed.

Because of this low prevalence in the primary care setting, even if we had a biomarker that was 99% sensitive and 99% specific, the positive predictive value (PPV) of the test (the probability of having the disease if the test is positive), with a prevalence of 0.01% (a wishfully high value), would be about 1% (see the figure on page 8). The PPV is so low because it is highly dependent on prevalence, while sensitivity and specificity are not.

Put into everyday parlance, this amazing test would have only a 1 in 100 chance of showing that someone has TN, if he or she truly has it. This problem of spotting rare disorders in the general population is one of the main reasons why screening tests for rare disorders are not generally advocated.

So, what do we do to reduce the risk for misdiagnosis? One of the strategies is the rationale for referring more-complex patients into specialty or tertiary-care clinics. Even if the clinicians there are no more skilled

in diagnosing these special conditions, from a numerical standpoint, with the higher prevalence (say 1%) via the funneling of more TN cases to a pain clinic, the identical test done in a specialty clinic will be far more effective at picking up a case because the prevalence and the concomitant positive predictive value (about 50%) are much higher (see the accompanying figure).

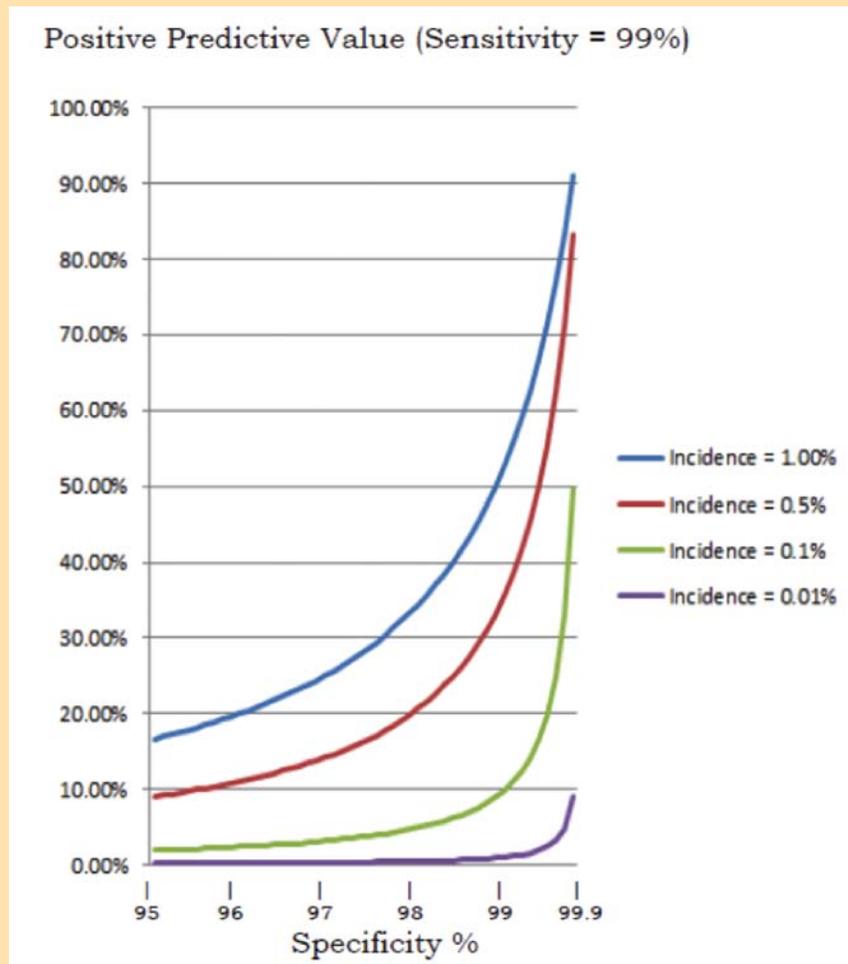
At the same time, the specialist clinician, by virtue of a higher volume of TN cases, becomes far more familiar with the pattern and nuances and can then develop skills to spot atypical variants of TN that less experienced clinicians would be unable to discern. Thus, well-defined referral networks for patients with rarer pain conditions such as TN are critical to optimal care.

Can we do more? Training and refresher courses for all levels of health-care providers are needed and

may be provided by realistic and modern case-based tutorials. One parting shot is to note that in TN, even without the wished-for serum biomarker noted above, the unique history is so powerful that it has biomarker-like levels of high sensitivity and specificity. The next time your patient complains of electric-shock-like pain that lasts for seconds without provocation in an otherwise healthy orofacial structure, think outside the box: Consider deferring invasive procedures and seek an appropriate referral.

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This graph shows the relationship between the positive predictive value (PPV) and the incidence of a disorder. At very low incidence, such as is the case with trigeminal neuralgia, around 0.01% (purple line), even with a highly accurate test with 99% sensitivity and 99% specificity, the PPV of a test will be very low, around 1% in this case. Increasing the incidence of a disorder to 1% (blue line), such as in a specialty clinic, increases the positive predictive value to 50%. Thus, if someone reports electric-shock like pain from light touching of the skin of the face or inside the mouth, the chance that he or she has TN will be much higher in a specialty clinic, and it will be easier to detect. (Graph courtesy of Katie Stoll, MS, as posted to www.thednaexchange.com)