patient had presented with erythema migrans, fever and a painful hip. Lyme disease was confirmed serologically, with a positive serum IgM immunoblot to *Borrelia* outer surface protein C and 41-kDa protein; IgG immunoblot was negative. Treatment with doxycycline for 21 days was associated with full recovery. Since then, he has been in good health.

General examination showed no superficial lymphadenopathy, arthropathy or rash. No abnormal signs were detected in the chest, cardiovascular system and abdomen (including digital rectal examination and assessment of anal tone). Neurological examination showed normal gait and heel-toe walking. The cranial nerves were intact. In the limbs, power and sensation were normal. Reflexes were reduced in the left knee and ankle: plantar reflexes were bilaterally flexor. An attempt at sitting up from the supine position resulted in upward movement of the umbilicus (Beevor's sign).

T2-weighted MRI of the thoracic spine showed focal non-specific intramedullary abnormal signal at T9–T11, with no associated expansion of the cord. MRI of the brain and craniocervical region was normal. The CSF protein was 1.25 g/l; IgG oligoclonal bands were detected (but were not present in serum). In the CSF, 250 cells/ml were detected, which were predominantly reactive lymphocytes. No malignant cells were identified. Herpes simplex-1 and Herpes simplex-2, varicella zoster, cytomegalovirus and Epstein-Barr virus were not detected in the CSF by polymerase chain reaction (PCR). No bacteria, mycobacteria and fungi were grown in CSF. Other blood investigations showed normal results, including prostate-specific antigen and immunoglobulin electrophoresis and quantification. From serological tests on blood, negative results were obtained for syphilis (*Treponema pallidum* haemagglutination assay and rapid plasma reagin assay), rheumatoid arthritis factor, antinuclear antibodies and extractable nuclear antigen antibodies; Bence Jones protein was not detected in urine. Electrocardiograph, chest radiograph, abdominal ultrasound scan and isotope bone scan were normal.

Serum was strongly positive (optical density 1.39) in the *Borrelia* C6 peptide enzyme-linked immunoassay (Immunetics, Boston, Massachusetts, USA). This synthetic peptide antigen-based test has greater specificity than some screening tests based on whole-cell lysate antigens. Serum IgM immunoblot was positive (bands at *Borrelia* outer surface protein C and at 39 and 41 kDa proteins), IgG immunoblot was negative. In CSF (tested at 1:10 dilution), IgG immunoblot was positive (bands at *Borrelia* outer surface protein C and at 20, 39, 41 and 58 kDa proteins). Insufficient CSF was available for borrelial DNA detection tests by PCR or for IgM immunoblot and more detailed assessment of intrathecal antibody synthesis.

The patient was treated with ceftriaxone 2 g a day intravenously for 10 days. Seven days after admission to hospital, he developed a left VII cranial nerve palsy: this was treated with a 10-day course of oral prednisolone and the patient fully recovered from the palsy. In May 2005 (after 8 months of follow up), serum *Borrelia* C6 peptide enzyme-linked immunoassay was positive (optical density 1.17) and IgG immunoblot was positive (bands at *Borrelia* outer surface protein C and at 20, 39, 41 and 58 kDa proteins). In July 2005, after 15 months of

follow up, the patient had no residual neurological deficit. His serum showed a C6 peptide enzyme-linked immunoassay reading of optical density 0.87. IgG immunoblot was positive, but with weaker reactions than in May 2005 (tested in parallel). IgM immunoblot was negative.

This patient presented with a painful inflammatory thoracic motor and sensory polyradiculopathy (Bannwarth's syndrome).¹ The positive Beevor's sign (weakness of the lower abdominal muscles, due to a cord or root lesion at or below T10)² was confirmed by MRI. Other causes of a positive Beevor's sign include facioscapulo-humeral dystrophy and myopathy, which were absent in this patient. The initial differential diagnosis based on the magnetic resonance appearances was malignant infiltration or varicella zoster infection, sine herpette. These possibilities were excluded by the negative findings from CSF cytology and PCR studies, by isotope bone scanning and by other investigations. Instead, Lyme borreliosis was diagnosed on the basis of the patient's history, residence in an endemic area and serological results indicating reinfection.³ Development of facial weakness supported a diagnosis of neuroborreliosis. The CSF lymphocytosis and raised levels of CSF protein were also typical findings for acute neuroborreliosis. Neuroborreliosis with malignancy simulating meningopolyneuritis has been described previously,⁴ but has not been reported as a presentation of reinfection with *B. burgdorferi*.

This case serves to underscore several clinical points. Firstly, Lyme borreliosis may present by mimicking a malignancy. Secondly, a previous episode of borrelial infection may not confer immunity. Reinfection is uncommon, but is more likely to occur in patients whose previous episode was promptly treated rather than in those with longstanding infection, who have a well-developed antibody response before treatment.⁵ Thirdly, patients may not specifically recall a tick bite. Thus, it is important that a history of tick exposure risk, which may be residential, occupational or recreational, is sought from patients. Finally, Beevor's sign has a useful localisation value.

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Informed consent was obtained for publication of the patient's details in this report.

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What's in a name—familial rectal pain syndrome becomes paroxysmal extreme pain disorder

The condition generally known as familial rectal pain syndrome is a rare autosomal dominant disorder that was first described by Hayden and Grossman.¹ The term familial rectal pain, was coined by Dugan.² The OMIM database (<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=167400>) uses the term "pain, submandibular, ocular and rectal, with flushing".

The disorder starts in the neonatal period (possibly in utero) and is lifelong. Its most characteristic clinical features are attacks of excruciating pain that affect various parts of the body, including the rectum, genitalia, face and limbs. In addition, other features reflecting autonomic dysfunction occur, including harlequin colour changes and pupillary abnormalities. Some patients with the disorder experience non-epileptic tonic seizures during severe episodes of pain. These may be associated with cardiac asystole.

Over the past decade, a worldwide consortium of clinicians, geneticists and scientists have been attempting to find the cause of familial rectal pain and recently discovered that it is caused by mutations in the sodium channel *SCN9A*.³ The consortium has worked closely with patients affected by the disorder and their families. During this collaboration there was considerable dissatisfaction with the name "familial rectal pain syndrome".

Consequently, a conference was arranged at the offices of the charity called Contact a Family (<http://www.cafamily.org.uk>) in London on 6 September 2005. Members of the consortium, along with affected people and their families, debated whether the name should be changed and, if so, to what. Others participated by email.

Whether the condition should be named according to its clinical features or on a more scientific basis reflecting the new knowledge that it is a channelopathy or that it appears to be a disorder principally affecting the autonomic nervous system was discussed initially. All preferred the first approach, partly because it was felt that a name should have meaning for the general public and also for the medical and scientific communities and partly because mutations in the sodium channel have not been found in all those affected with the disorder.

There was near unanimity that the term rectal should be dropped. This was partly because its inclusion seems to imply that the pain is confined to the rectum, when in fact this is only one affected site. In addition,

those with the disorder voiced the view strongly that they found the term both stigmatising and embarrassing. Clinicians and those affected generally considered that the term incorrectly implies the condition is gastrointestinal rather than neurological and, as a consequence, those professionals who should be interested in it are often not.

All those involved considered that the term pain should remain. Most clinicians were initially sceptical about qualifying this with words such as severe, extreme or excruciating, often feeling that the use of these would be frightening, especially at diagnosis. Those affected and their families disagreed, arguing that as the condition is associated with episodes of pain that seem to be as severe as anything experienced by man, this must be reflected in the name. Indicating the severity of the pain will emphasise to clinicians the importance of recognising the disorder, particularly as partially effective treatments are available. Finally, after much debate, most considered that the best term to describe the pain was "extreme".

Clinicians and scientists were of the opinion that as we now understand what causes the condition and know its clinical features and prognosis, the term syndrome is no longer appropriate and that either disease or disorder should be substituted. Those affected and their families preferred the term disorder as being less stigmatising.

Most initially wanted to retain the term familial. There was, however, eventually an agreement to drop it, partly because a five-word name was considered excessively cumbersome and partly in acknowledgment that sporadic cases also occur.

Whether other features of the disorder, such as the harlequin colour changes and the non-epileptic tonic seizures, should be reflected in the name was discussed. It was, however, felt that some of these features, such as colour changes, although interesting, have a minor effect on those affected, whereas others, such as seizures, are not universal. Clinicians strongly thought that the episodic nature of the attacks should be reflected in the name. The term paroxysmal was preferred to seizure as this was less likely to lead to confusion with epilepsy.

The name that received most support was paroxysmal extreme pain disorder (PEPD). Given that this name emerged from a democratic process including, as far as we know, most professionals interested in the condition and affected patients and their families from around the world, we consider that henceforth the name "paroxysmal extreme pain disorder" should be used in preference to the obsolete name familial rectal pain (syndrome).

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Email triage for new neurological outpatient referrals: what the customers think

When patients with neurological symptoms are referred to a neurologist by their general practitioner (GP), the neurologist acquires two customers—the patient and the GP. If the neurologist plans to introduce a new or changed service it would be considered good practice to seek the opinion of the people using the service—the customers. Such an approach is commonplace in other walks of life, but is rarely used in medicine.

We used this approach when considering changing from a letter-based referral system for new referrals to one based on email. Email can be used to triage referred patients into those who need to be seen in a clinic and those who can be managed without entering the clinic system, either by investigations or by simply offering advice. This approach has been used successfully for general medical patients in Finland¹ and we have shown that in neurological practice this novel method is feasible, safe, sustainable and saves neurologists' time.^{2,3} A similar system is being introduced within parts of the UK National Health Service for a variety of specialties under the title of Integrated Clinical Assessment and Treatment Services.⁴ The email service provides a response to the GP within 48 h, but if the referral is dealt with by simply providing advice or by arranging investigations, the patient will not be seen in person by the neurologist. When we presented our results at neurological meetings, one objection raised against the email system was that patients and their GPs would be unhappy that a referred patient may not see a neurologist in person. To test this hypothesis we carried out a customer survey.

Methods

The study was carried out in two different parts of the UK, general neurology clinics throughout Northern Ireland and waiting list initiative clinics in Pinderfields Hospital, Wakefield, Yorkshire.

Patients were new referrals from GPs who had been referred conventionally by letter and whose degree of urgency had been graded by the neurologist as routine. They had generally been waiting between 3 and 6 months for their appointment.

Their opinion was obtained while they were waiting to be seen by the neurologist. The approval of the ethical committee was not required.

Patients were asked to choose between the two possible referral methods that were

presented to them on a single sheet of paper. The sheet was given to them by the clinic nurse or receptionist, with a short written explanation, and the completed sheet was collected from them before they saw the neurologist, who was unaware of the results when seeing the patient.

GPs were chosen in Northern Ireland by geographical spread, excluding County Fermanagh, where the email system had already been introduced. They were contacted by telephone by one of the authors (CD) and the questionnaire was filled in over the telephone.

GPs in Wakefield were chosen from hospital referrers. They were contacted first by an explanatory letter enclosing the questionnaire and if this was not returned, one of the authors (LL) followed up by telephone.

Results

Overall, 80% of patients in Northern Ireland (n = 100) and 91% of those in Yorkshire (n = 100) preferred the email system. GPs also preferred the email system, although less enthusiastically so—80% in Northern Ireland (n = 50) and 51% in Yorkshire (n = 82).

Discussion

Most new patients and GPs in two parts of the UK would prefer an email triage system compared with the current system of hospital referral. We carried out this study on patients who were within the system but had not yet completed it and who therefore had good knowledge of what the conventional system included. All the GPs had first-hand knowledge of referral of patients to their local neurologist. The wording of the questions was direct but encapsulated the implicit trade-off between an early opinion with the possibility of not being seen by a specialist against a delayed opinion with the guarantee of being seen by a specialist.

We do not find these results surprising. The existing system of referral is impersonal and inefficient. Hospital referral letters are usually unstructured and many are still hand written and often difficult to read. An email with a proforma encapsulates what the GP wants to gain from the consultation and the rapid response from the neurologist brings a degree of personal interaction that is completely lacking in the conventional system. A competent neurologist, from a description by a competent GP, can easily diagnose many neurological symptoms of patients. Patients can then either be reassured without being seen or have appropriate investigations carried out without entering the hospital system. A follow-up of more than 150 patients by this system has shown that, when carried out by an experienced neurologist, this practice is safe and saves neurologists' time.³ Benefits accrue to both patients and their GPs and also to neurologists who will see patients in the clinic more relevant to their skill and experience. A wider adoption of this system requires neurologists and GPs to change their practice and such changes are known to be difficult. Whatever the arguments proposed against these changes, a lack of customer readiness should not be one of them.

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