Management of complex regional pain syndrome of the hand

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Abstract

Downloadable open access UK guidelines for managing complex regional pain syndrome (CRPS) have been available since 2012. Care is interdisciplinary and readers involved in the care of patients with CRPS are strongly recommended to read these guidelines. Since 2012 there have been several larger randomized controlled trials (RCTs) which have helped to clarify the role of some treatments in CRPS management, although the results of some subsequent meta-analyses have been conflicting. Early data from an RCT with 152 patients suggest superiority of dorsal root ganglion stimulation over conventional spinal cord stimulation in patients with lower extremity CRPS. An RCT involving 111 patients investigating the role of low-dose intravenous immunoglobulin treatment in long-standing CRPS failed to show benefit from this treatment. Despite the lack of direct evidence that nociceptive and neuropathic pain drugs can relieve pain from CRPS, it is reasonable for orthopaedic surgeons to follow relevant guidelines and initiate treatment with these drugs and to request GP colleagues to monitor and titrate these drugs as appropriate. As per the UK CRPS guidelines, the non-surgical approach to the management of CRPS centres on the following four pillars of care: education, physical rehabilitation, pain relief and psychological intervention where appropriate.

Keywords CRPS; dorsal root ganglion stimulation; immunoglobulin treatment; pain relief; rehabilitation; spinal cord stimulation

Introduction

Complex regional pain syndrome (CRPS) is a debilitating, painful condition in a limb, associated with sensory, motor, autonomic, skin and bone abnormalities. CRPS commonly arises after injury to that limb. However, there is no relationship to the severity of trauma, and in some cases there is no precipitating trauma at all. The cause of CRPS is unknown and current hypotheses suggest that there is interplay between peripheral and central pathophysiologies.[1]

The diagnosis is a clinical one and it is beyond the scope of this paper to discuss the various hypotheses underlying the pathophysiological mechanisms involved or as to how the diagnosis is arrived at and readers are directed to other literature on this matter.[2,3]

In the production of the 2012 UK guidelines for diagnosis, referral and management of CRPS in primary and secondary care published by the Royal College of Physicians in London,[1] the evidence base for management strategies in patients with CRPS was rigorously examined and much of this article is based on the contents of that work. [Note: these guidelines were updated in July 2018 sometime after this paper was submitted for publication.[1]] Readers who are involved in the clinical care of patients who are afflicted with CRPS are strongly recommended to read these guidelines. In summary, the non-surgical approach to the management of CRPS centres on the following four pillars of care: education, physical rehabilitation, pain relief and psychological intervention where appropriate. This should be delivered early and patients should be reassured that the pain will either completely or partially resolve in at least 85%[1] of cases by 18 months to 2 years after symptom onset, although ongoing motor dysfunction with limb disability may be common. Of course, if personal experience is anything to go by, clinicians only tend to remember the 15% of patients who do not improve. Readers who wish to examine this evidence more closely are directed to the summary of results from systematic reviews of randomized controlled trials (RCTs) for treatment of pain in CRPS with a summary of NICE guidelines and International Association for the Study of Pain (IASP) recommendations for neuropathic pain which is provided in Appendix 14 of the 2012 UK guidelines[1] and a more up-to-date review of RCTs is provided in the Federation of European Societies for Surgery of the Hand Instructional Courses 2017 handbook.[4] The evidence can be conflicting — a good example of this is work done on examining whether administering 500 mg vitamin C will prevent CRPS in patients who have sustained a distal radius fracture. Two studies (carried out by the same first author) suggested benefit, whereas another study suggested no benefit. Confusingly, two recent meta-analyses based on these three studies (thus looking at the same number of patients) came to differing conclusions — one suggesting benefit and the other not[5,6]!

Education

Management of patients with CRPS should include reassuring patients that CRPS is a recognized condition[1] — although its causes are poorly understood — and that the pain will either completely or partially resolve in at least 85% of cases, although ongoing motor dysfunction with limb disability may be occur in some patients, despite the fact that early and appropriate treatment may have been given. Practitioners can support patients by providing a clear diagnosis, information and education about the disease, help to set realistic goals and, where possible, involve the patient's partner and/or other family members. Whilst there may be a lack of evidence to inform the best functional advice to offer patients with suspected CRPS, or CRPS for which concomitant pathology has not yet been ruled out, pragmatically, encouragement of gentle limb use and active lifestyle is recommended, with gentle limb movement (unless contraindicated for orthopaedic reasons), paying frequent attention to the affected limb, trying to 'desensitize' the affected limb (gentle stroking of the affected limb with different textured fabrics while viewing the limb and

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then progressing to more active use (e.g. weight-bearing and stretching) when tolerated.[1]

Physical rehabilitation

In the specialty-specific guidance for occupational therapists and physiotherapists, the UK CRPS guidelines state that patients presenting with mild to moderate disease and some patients with recent onset severe disease that is quickly resolving (eg shortly after trauma) can be treated using a variety of different approaches, although many of these approaches have not been subject to the crucible of an RCT. The thrust of these approaches — amongst other things — is for patients to self-manage their situation, by exercising and paying attention to their painful limb, as well as pacing themselves and setting realistic goals. Therapists can teach coping and relaxation skills and educate patients that they must avoid prolonged periods of immobilization or covering up of a painful limb.[1]

The guidelines also advise that where if there are features that indicate moderate or severe disease and/or poor recovery that an early referral to a multidisciplinary pain clinic or specialist unit is made. Features that might prompt this are presentation with moderate to severe signs and symptoms (except if of very recent onset after trauma and quickly resolving), the presence of dystonia, the lack of a positive treatment response within 4 weeks of initiating treatment, or if the patient's signs or symptoms deteriorate or improvements are not sustained despite ongoing treatment.

There is strong high-quality evidence that graded motor imagery is of benefit in this condition so where rehabilitation is undertaken in specialist units or by therapists with CRPS expertise, this may be offered. There is also some moderate quality evidence that mirror visual feedback therapy is of benefit — although this was in seen in patients with CRPS after stroke. In addition to providing CRPS-specific rehabilitation techniques, specialist units may treat patients with embrace strategies that include self-administered tactile and thermal desensitization with the aim of normalizing touch perception, strategies to correct body perception disturbance which involve looking, touching and thinking about the affected body part, mental visualization to normalize altered size and form perception of affected body part, functional movement techniques to improve motor control and awareness of affected limb position, and attempt to address conflict allodynia, where patients may need re-education to reduce fear of physical contact with others in community settings.

Yellow flag psychosocial risk factors can help therapists to understand contributing causes to a suboptimal treatment response.[1]

Pain relief

Large numbers of RCTs with large numbers of patients that can help support the evidence base underpinning medical interventions for this condition simply do not exist. There are many case series which suggest good outcome for various interventions, but in the context of a condition where the pain will either completely or partially resolve in at least 85% of cases by 18 months to 2 years after diagnosis[1] it is difficult to be certain it was the intervention that resulted in the good outcome, or whether it was natural resolution of the condition.

Where there is evidence arising from randomized studies supporting an intervention, the numbers of patients participating in these studies is small. Thus for example, the 2012 UK guidelines support the use of IV pamidronate 60 mg if administered within 6 months of onset of symptoms. This was based on a small though high-quality study and the data from all the studies using biphosphonates early in the course of the condition favoured the early use of biphosphonates. The 2012 UK guidelines also support the use of the spinal cord stimulation — based on an RCT involving 54 patients. The guideline authors also recognized that there is some evidence that the efficacy of this treatment generally declines over time. It is of interest that a recent large RCT involving 152 patients comparing spinal cord stimulation with dorsal root ganglion stimulation gave a higher treatment success rate in patients with CRPS at 3 and 12 months but this study only involved patients with CRPS affecting their lower limbs.[7] Promising results from early studies suggested that there might be benefit to be had from low-dose intravenous immunoglobulin treatment for long-standing CRPS. This resulted in a large RCT involving 111 patients being carried out.[8] Unfortunately it failed to show any benefit from this treatment.

A meta-analysis of several small studies suggests that there may be moderate evidence for the efficacy of low-dose IV ketamine infusion in long-standing CRPS but the duration of effect does not persist beyond several weeks. Concerns have also been expressed that repeated or prolonged administration of ketamine might cause bladder and liver damage.

Whilst early treatment with simple analgesic drugs like codeine, dihydrocodeine, tramadol, non-steroidal anti-inflammatory drugs (NSAIDs) and paracetamol where appropriate may not necessarily affect the specific pain of CRPS, they but may reduce ongoing trauma-related pains and assist in the process of mobilization. The 2012 UK guidelines suggest that existing guidelines for the drug-based management of neuropathic pain could be used as a starting point in patients with CRPS, but apart from one study involving gabapentin — which did not show any benefit — none of the studies that form the basis for those neuropathic pain guidelines actually involved patients with CRPS.

As regards interventions like stellate ganglion block and intravenous regional blockade, evidence from RCTs suggest that these are of no benefit. In the case of intravenous blockade, the possibility has been raised that it is the actual effect of the prolonged inflation of the tourniquet that may be giving some patients benefit.

Psychological intervention

Psychotherapeutic and sociotherapeutic methods represent an important part of multimodal pain therapy when targeting pain-related fears in all phases of the disease process, especially if accompanying psychosocial factors or co-morbidities exist (eg, depressive mood, pain-related avoidance, posttraumatic stress disorders, perceived injustice, and financial worries). Patients with many psychosocial problems may be harder to treat). Graded exposure (GEXP) treatment has shown good evidence for efficacy in CRPS. For this approach, a psychologist identifies and classifies fear-triggering situations (e.g. pain induction through certain movements and situations). Patients are then gradually

exposed to these situations by a physiotherapist. The efficacy of GEXP in comparison to conventional rehabilitative therapy was confirmed in one large case series for chronic CRPS (n = 106) and a recently published small and single centre RCT. GEXP reduced pain and improved function.[9]

Psychological interventions specific to pain would normally be provided in the form of a multidisciplinary pain management programme, for those who require it, and would normally be based on an appropriate assessment method. The psychological interventions would usually follow principles of cognitive behavioural therapy (CBT) which is not a single therapy or even a single set of standardized interventions. Rather, CBT is a broad category of different treatment regimens. However, CBT regimens almost always include cognitive therapy (the 'C' of CBT) as a core component. Usually CBT also includes interventions designed to alter behaviours (the 'B' of CBT) and some combination of operant treatment, coping skills training, relaxation strategies, pacing or activity—rest cycling, exercise and activity management, and pleasant activity scheduling.[1]

Management of symptoms and/or signs other than pain

The UK 2012 guidelines suggest that for CRPS-related limb dystonia, intrathecal baclofen treatment can be considered only if all other options, including oral medications, have failed and that this treatment should be delivered only in specialised centres. The guidelines also suggest that the overall efficacy of regional botulinum toxin for CRPS-related dystonia is poor. The guidelines go on to state that whilst serial splinting by experienced physiotherapists may symptomatically improve some cases of dystonia, care should be taken to give time to exposing the limb for the conduct of desensitization therapies.

In cases of refractory, disabling limb swelling, advice from a lymphoedema nurse should be sought and there are reports that spinal cord stimulation can reduce limb swelling. In patients with CRPS and skin ulcers with or without infection, tissue viability and/or dermatological opinion should be sought as early as possible. Where ulcers occur in parallel with limb oedema, reduction of the oedema with spinal cord stimulation can promote ulcer healing.

Long-term support in CRPS

A proportion of patients with CRPS will have ongoing symptoms requiring long-term support, usually low-level, and the UK 2012 guidelines provide further advice and guidance regarding this.

Role of surgery in the management of CRPS

One of the common conditions that can cause CRPS is fracture of the distal radius. It can occur, with equal severity, in those fractures that were either treated operatively or non-operatively. It the former, the patient is likely to have been warned about this as part of the consent process, although it can be argued that they may not have fully understood the nature of the condition. In the latter (non-operatively treated group) it usually comes as a shock to the patient and they, understandably, seek to find out what went wrong and whom to blame. At this point, counselling and treating the patient in a multidisciplinary team setting, including the treating surgeon, will be of considerable benefit to the patient.

There is a reluctance to carry out any additional surgery when CRPS is active. The relapse rate of CRPS is well evidenced if elective surgery is carried out when the condition is still active.[1,2] The exception to this would be the occurrence of peripheral nerve entrapment such as carpal tunnel syndrome. If the symptoms are distressing and conservative treatment including steroid injection fails, then it would be reasonable to carry out carpal tunnel release.

The benefit of manipulation under anaesthesia (MUA) of the joints of the hand and wrist in the early phase of CRPS to prevent long-term stiffness is not well evidenced in the literature; however the surgical author does believe that MUA around the 6-month mark can be of help in those with moderate CRPS.

The long-term sequela of CRPS of the hand is joint stiffness, typically hyperextension of the metacarpophalangeal jopint (MCPJ) and flexion of the proximal interphalangeal joints (PIPJs). Therapy, including the judicious use of splints, can play an important role in reducing this. MCPJ stiffness can cause significant functional restriction and correction of this by arthrolysis after the disappearance of CRPS (typically around 2 years after the occurrence). The PIPJs are best left alone unless the flexion deformity is particularly very severe.

Conclusion

There is no cure for CRPS and as long as this remains the case, the care of patients with CRPS will have to be interdisciplinary and will involve many different specialties working together to try and achieve the best possible outcome for the patient. Fortunately patients can be reassured that the pain will either completely or partially resolve in at least 85% of cases by 18 months to 2 years after symptom onset, although they may still have ongoing motor dysfunction with limb disability. There remains a paucity of RCTs with large numbers of patients. ◆

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