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Updated guidelines on complex regional pain syndrome in adults☆

James Henderson

Department of Plastic and Reconstructive Surgery Southmead Hospital, Bristol BS10 5NB, United Kingdom

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Abstract A multidisciplinary team representing 28 professional bodies has updated the guidelines for the management of complex regional pain syndrome (CRPS), published by the Royal College of Physicians (RCP) of England, 2018. The author represented the British Association of Plastic, Reconstructive and Aesthetic Surgeons in this process and is an author of the guidelines. This article summarises the updated guidelines and highlights aspects relevant to plastic and reconstructive surgery.

A 3-year process involving 28 professional bodies has updated the 2018 guidelines for professionals working with patients having complex regional pain syndrome (CRPS). These guidelines are published by the Royal College of Physicians and are available for free download at https://www.rcplondon.ac.uk/guidelines-policy/complex-regional-pain-syndrome-adults.[1]

The key update to the guidelines is that they now contain sections for clinicians working in every conceivable setting in which CRPS could be encountered, including emergency medicine, general practice, psychiatry, dermatology, rheumatology, sports medicine, neurosurgery, pain medicine, rehabilitation medicine, occupational and physiotherapy, vascular surgery, orthopaedic surgery, hand surgery and plastic surgery. Each of the specialities provided representatives to the process, who attended focus group meetings as well as ongoing communication by email.

The author represented the British Association of Plastic, Reconstructive and Aesthetic Surgeons (BAPRAS) in this process. It was interesting to see how our experience of patients with CPRS differed from that of other specialities, and we worked hard to frame guidelines that were appropriate for all clinicians and encouraged multidisciplinary care of the patients when appropriate. As expected, with 28 different contributing organisations, there was a certain amount of compromise needed to produce a cohesive and consistent document, and inevitably, the guidelines are a large document (97 pages). Within this document, however, are specific sections for different clinicians, which summarise the information needed; therefore, it is not necessary to read the entire document.

The guidelines were based on two systematic reviews, conducted in 2012 and 2017, and treatment recommendations were developed by panel discussion. No formal grading of recommendations was undertaken.

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E-mail address: james.henderson@nbt.nhs.uk

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CRPS is a debilitating painful condition in a limb, associated with autonomic, sensory and motor abnormalities, as well as physical alterations to the skin and bone. CRPS usually arises after trauma (including surgery) to the limb, but the incidence of CRPS does not relate to the severity of the trauma. The incidence in Europe is 20–26/100,000 person-years. Although usually only one limb is affected, in 7% of patients, the condition spreads to other limbs. The cause of CRPS is unknown, but the idea that it is solely a sympathetic dysfunction and occurs in stages is currently obsolete. Treatment for CRPS can be very effective if initiated at the early stage, but some patients continue to suffer indefinitely.

Type 2 CRPS occurs after a recognised peripheral nerve injury, and in the absence of nerve injury, type 1 CRPS is diagnosed.

Patients still report that some health professionals do not believe that their condition is 'real'. Conversely, it is recognised that some patients factitiously present with CRPS for reasons of secondary gain, but CRPS is not associated with a history of previous psychological problems, somatisation or malingering.

Anecdotally, plastic and hand surgeons, particularly those who are involved in medicolegal work, report large numbers of patients who have self-diagnosed or been misdiagnosed with CRPS; in the latter case, clinicians were possibly misled by patients seeking secondary gain.

Diagnosis

The guideline team reached a consensus on the diagnostic criteria for CRPS. The decision was made to adopt the 'New IASP' (*Budapest*) criteria,[2] which were generated by a panel discussion of experts in 2007 and have been widely adopted. Patients who do not meet the diagnostic criteria may still be treated for their symptoms but should not be diagnosed as having CRPS. Sensory and motor abnormalities after surgery or trauma are common, and although these abnormalities can be features of CRPS, they are not diagnostic.

Patients might present to surgeons with symptoms following trauma or surgery, and it is incumbent on us to diagnose and treat these patients quickly. The guidelines include a simple chart that is designed for display in busy clinics (Figure 1) so that clinicians can easily check the diagnostic criteria, initiate treatment and refer patients promptly. It is recommended that post-operative patients be instructed to seek help if they develop symptoms of worsening pain or sensory, motor or vasomotor changes.

The development of CRPS should not be considered as an evidence of suboptimal surgical management.

Patients with a prior diagnosis of CRPS might be referred to surgeons whether this is a correct diagnosis or not. These patients often have expectations that they can be cured with surgery, typically expecting that this can be achieved by amputation. This can lead to difficult consultations, and the guidelines aim to clarify the roles and risks of surgery for such patients.

Treatment by surgeons

Surgeons should initiate treatment with simple analgesia (non-steroidal anti-inflammatory drugs and paracetamol) and possibly tricyclic antidepressants (e.g. amitriptyline) or anticonvulsants (e.g. gabapentin), although the patient's general practitioner might be better placed to arrange follow-up and titration of doses. If a patient requires anticonvulsants or strong opiates, then urgent referral to a pain consultant should be considered. You should be aware that no drugs are licensed for the treatment of CRPS in the UK.

Surgeons should organise urgent physical therapy for patients, and this is the mainstay of treatment for CRPS.

Guanethidine blocks should not be used by plastic surgeons, although pain clinicians may consider them; benefit has not been shown in randomised controlled trials.

There is no recommended prophylaxis for CRPS, although early vigilance and rehabilitative treatment are recommended.

Surgery in patients with CRPS

Elective surgery can be performed in limbs that have been previously affected by CRPS. Recurrence of CRPS is less than 15% if surgery is delayed until the acute symptoms have settled; most recurrences are mild. If possible, it is recommended to delay surgery until at least 12 months after symptoms have settled. Anecdotally, patients with previous CRPS have worse outcomes than those without, and this might be due to poor body image, response to surgical pain or difficulty with rehabilitation. More research is needed to evaluate the outcomes of elective surgery on limbs or patients previously affected by CRPS.

Surgery might be indicated in the treatment of type 2 CRPS where a recognised nerve insult has occurred – e.g. neuroma – or if a nerve is thought to be caught in a suture or scar contracture.

Surgery for patients with CRPS should be performed by a surgeon experienced in treating patients with CRPS, with an anaesthetist who is also a pain specialist.

Amputation of CRPS – affected limbs

Patients are increasingly requesting amputation of affected limbs, either in the belief that this will cure pain, or for reasons of hygiene/to allow chronically ulcerated skin to be removed and wounds to heal. We occasionally see patients who have attempted to amputate their own limbs, which may force us into terminalising stumps.

There is insufficient evidence to predict outcomes after amputation, but the literature suggests that many patients will have pain and not be able to wear prostheses on that limb. Many patients have recurrent symptoms in that limb or in another limb, and phantom limb pain persists in the majority.

If amputation is considered, it is recommended that a multidisciplinary team must be involved, comprising, as a minimum, a pain consultant, an appropriate psychologist or psychiatrist, a physiotherapist and/or occupational therapist as well as a surgeon; a tertiary amputee rehabilita-

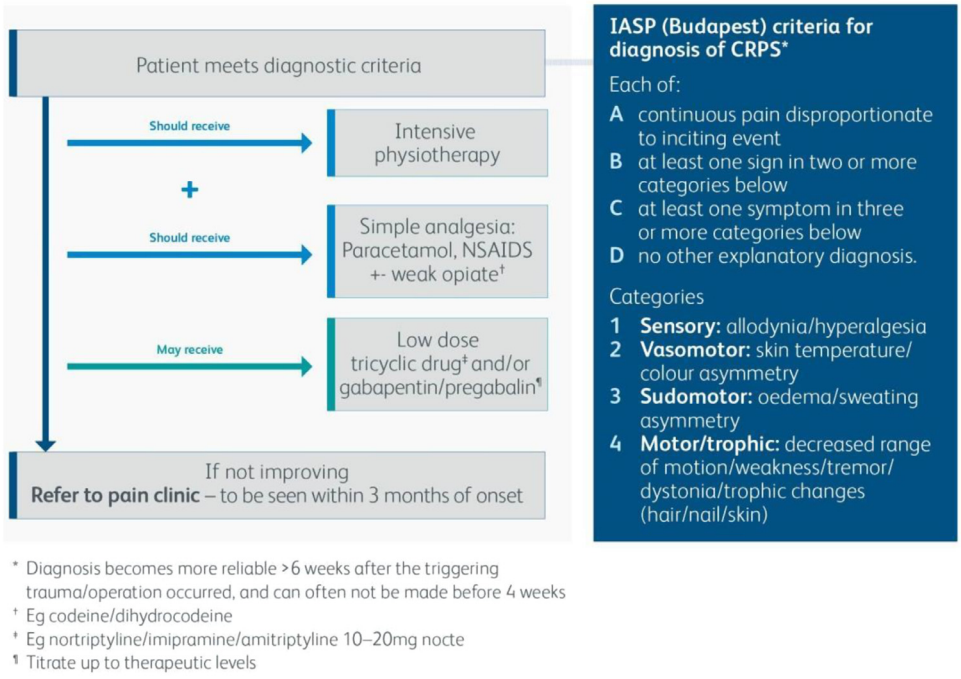

Figure 1 Chart for display in clinic showing Budapest diagnostic criteria and initial management of CRPS.

tion and rehabilitation team should be involved well before surgery and ideally should have experience of patients with CRPS.

All other treatments should be assessed before patients proceed to amputation, and the team must consider whether the patient has unrealistic expectations, psychological disorders or negative coping mechanisms, all of which are predictors of a negative outcome.

Patients and their relatives need to be aware that amputation is unlikely to cure pain, that CRPS may recur in the stump or another limb, and that prosthesis use may be impossible. Ulceration may recur in the stump.

Amputation should not generally be performed within 24 months of the diagnosis of CRPS being made. The one strong indication for amputation is intractable infection, but the multidisciplinary team should be involved as above except in life-threatening emergencies.

Summary

CRPS is a severe albeit rare complication of limb trauma or surgery. The development of CRPS does not imply that treatment has been suboptimal. Although diagnosis has historically been difficult, thus leading both to overdiagnosis and missed diagnoses, clear criteria are included in the guidelines (Figure 1). Early treatment is critical, particularly physical therapy.

Surgery on patients with CRPS should be delayed for 24 months from diagnosis or 12 months from resolution of symptoms if possible. Amputation should be regarded as a last resort, to be approached very cautiously with a multidisciplinary team that is experienced in such cases.

Conflict of interest

JH is a contributing author of the 2018 Complex regional pain syndrome in adults (2nd edition) but had received no financial or other benefits or support.

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