



Royal College  
of Physicians

# Complex regional pain syndrome in adults

UK guidelines for diagnosis,  
referral and management  
in primary and secondary care

May 2012

Endorsed by



The  
British  
Psychological  
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THE BRITISH PAIN SOCIETY



The Physiotherapy  
Pain Association



Pain Relief  
Foundation

Also endorsed by the British Society of Rheumatologists and British Health Professionals in Rheumatology

# Complex regional pain syndrome in adults

## UK guidelines for diagnosis, referral and management in primary and secondary care

Developed by a panel of experts with support from, and representation by the Royal College of General Practitioners, the Royal College of Physicians, the College of Occupational Therapists, the British Association of Dermatologists, the British Health Professionals in Rheumatology, the British Orthopaedic Association, the British Pain Society, the British Psychological Society, the British Society for Rehabilitation Medicine, the British Society for Rheumatology, the Chartered Society of Physiotherapy, the Physiotherapy Pain Association, the Society of British Neurological Surgeons, and the Pain Relief Foundation.

With additional support from the Royal College of Radiologists, the Society of Chiropractors and Podiatrists, The Association of Orthopaedic Practitioners, the British Association of Hand Therapists, the British Association of Plastic, Reconstructive and Aesthetic Surgeons and the Association of British Neurologists.

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# Abbreviations

Colleges and professional institutions:

**ABN** Association of British Neurologists

**AOP** Association of Orthopaedic Practitioners

**BAD** British Association of Dermatologists

**BAHT** British Association of Hand Therapists

**BAPRAS** British Association of Plastic, Reconstructive and Aesthetic Surgeons

**BHPR** British Health Professionals in Rheumatology

**BOA** British Orthopaedic Association

**BPS** British Pain Society

**BPS** (Psychology) – British Psychological Society

**BSR** British Society of Rheumatologists

**BSRM** British Society of Rehabilitation Medicine

**COT** College of Occupational Therapists

**CSP** Chartered Society of Physiotherapy

**PPA** Physiotherapy Pain Association

**PRF** Pain Relief Foundation

**RCGP** Royal College of General Practitioners

**RCP** Royal College of Physicians

**RCR** Royal College of Radiologists

**SBNS** Society of British Neurological Surgeons

**SCP** Society of Chiropodists and Podiatrists

## Summary

This guideline concerns the diagnosis and management of patients with complex regional pain syndrome (CRPS). It provides recommendations for diagnosis, treatment and referral in a variety of clinical settings (GP practice, orthopaedic practice, rheumatology, neurology and neurosurgery, dermatology, pain medicine, rehabilitation medicine and long-term care). Its purpose is to provide coherent guidance for professionals working in the different health specialties who care for these patients (Fig 1, page 2). The document starts with an introduction for all interested parties, followed by specialty-specific sections. Supporting documents are appended.

Clinicians will find relevant information in reading both the introduction and respective specialty-specific sections. Recommendations are in framed boxes and are generally based on panel consensus and expert opinion; grading is not provided. A concise summary of the guideline is available as a separate document.<sup>1</sup> Grading of recommendations using the typology developed for the National Service Framework for long-term conditions<sup>2</sup> is given there.



# Methodology of guideline development

These guidelines were initiated by Dr Andreas Goebel and Dr Chris Barker in association with the Pain Relief Foundation and were developed by a UK panel of experts representing a variety of healthcare specialties and professions. In addition, various professional associations and colleges were represented on the panel. Expenses for attending meetings were funded by commercial sponsors (see Appendix 1). The guidelines are intended for use throughout the UK.

## Identification of evidence

### 1 Diagnosis

A review of the literature from July 2000 to April 2010 was carried out. Medline (PubMed) and SCOPUS databases were searched using a combination of the following search terms: complex regional pain syndromes; reflex sympathetic dystrophy and causalgia; with diagnosis; diagnostic; early diagnosis and clinical trial; metaanalysis; practice guideline; comparative study; and consensus study.

Studies were included in the assessment if they were prospective and/or likely to contribute to diagnostic techniques for CRPS. Two reviewers assessed the chosen studies according to whether they evaluated any new diagnostic technique or algorithm, re-evaluated the Budapest diagnostic criteria, or documented any potentially important differential diagnoses.

### 2 Treatment and prevention

A systematic review published in 2002<sup>3</sup> of RCTs on the treatment and prevention of reflex sympathetic dystrophy (RSD) and CRPS from 1966 to June 2000 provided a foundation of data. This was used together with a second systematic review of the literature carried out using similar methodology by a subgroup of the Guideline Development Panel, who reviewed RCTs on the treatment and prevention of CRPS from July 2000 to April 2010.<sup>4</sup> (See Appendix 2 for further details of the systematic review methodology, RCTs published between April 2010 and December 2011 are added in Appendix 16.)

The results of the RCTs from both reviews were combined. Four levels of evidence of effectiveness were defined using the Van Tulder method<sup>5</sup> based on the methodological quality and outcome of the studies.

### 3 Earlier guidelines

A further literature search was carried out in Medline and Scopus from July 2000 to April 2010 using combinations of the terms: complex regional pain syndromes; causalgia; reflex sympathetic dystrophy with guidelines; clinical guidelines; treatment guidelines; practice guidelines and consensus, to identify guidelines for the treatment of CRPS. Guidelines were selected from the resulting articles according to whether they were devised by an interdisciplinary team and the guideline development process was described. Two guidelines were found.<sup>6,7</sup> These were reviewed by two panel members and the content was taken into consideration in devising the current guidelines.

## 4 Long-term management

Following consultation we found that there is no formally published literature on the long-term care for CRPS. We therefore developed this section using a ‘bottom-up’ approach. We first asked four patients for their views on requirements for long-term care. We then invited additional patients with CRPS and one patient founder of a pain support group to attend a breakout group meeting, at which these earlier formulated patient views were presented, along with a summary, provided by a health psychologist, of the views expressed by 17 patients who had attended a CRPS focus group.<sup>8</sup> Guidance was then formulated on the basis of patient-formulated themes, and enhanced by input from participating clinicians.

### Recommendations

The recommendations were developed on the basis of panel consensus and expert opinion, with reference to the existing literature. Where possible, recommendations were informed by evidence from the reviews of RCTs. (See summary of results of two reviews of RCTs in Appendix 14). No formal grading of recommendations was undertaken. At each stage of development the draft guidelines were circulated to members of the group for peer review prior to production of the final draft. Drafts were also sent for comments to additional expert patients, who had not otherwise participated in the guideline group.

### Consultation process

The final draft of the guidelines was circulated to each of the bodies represented on the panel (BPS, RCGP, SBNS, BSRM, RCP, BOA, BHPR, BSR, PPA, BAD, COT, CSP) and to the supporting organisations (SCP, RCR, BPS – Psychology, AOP, BAHT, BAPRAS, ABN). Comments were invited to be sent directly to the chairman and were implemented in consultation with the Committee members.

### Review

This guidance will be reviewed five years from the publication date.

## Introduction

Complex regional pain syndrome (CRPS) is a debilitating, painful condition in a limb, associated with sensory, motor, autonomic, skin and bone abnormalities.<sup>9–11</sup> 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 (9%).<sup>10</sup> CRPS usually affects one limb, but in 7% of cases later spreads to involve additional limbs.<sup>11–13</sup> The European incidence rate of CRPS is 26/100,000 person-years.<sup>14</sup> The cause of CRPS is unknown.<sup>15</sup> Characteristically, there is interplay between peripheral and central pathophysiologies. The earlier concepts that the predominant problem is sympathetic dysfunction and that CRPS occurs in (stereotyped) stages are now obsolete. It is also now clear that CRPS is *not* associated with a history of pain-preceding psychological problems, or with somatisation or malingering.<sup>16–18</sup> If a patient presents with such problems, these should be addressed where appropriate, as would be good practice in other medical situations. Patients still report feeling stigmatised by health professionals who do not believe that their condition is 'real'. Independently, it is recognised that some people self-induce signs with the aim of making their limb appear as though they have CRPS.<sup>19</sup>

A definition of recovery from CRPS has not yet been agreed. Limb signs (such as swelling, sweating and colour changes) usually reduce with time, even where pain persists.<sup>20</sup> However, such reduction of limb signs is in itself not 'recovery'. Where pain persists, the condition is best considered to be active. It is noted that, without limb signs, a diagnosis of CRPS according to the Budapest criteria can often not be made (see Table 1). These

**Table 1 Diagnostic criteria for CRPS ('Budapest criteria')<sup>21</sup> (A–D must apply)\***

A) The patient has continuing pain which is disproportionate to any inciting event		<input type="checkbox"/>	
B) The patient has at least one sign in two or more of the categories		<input type="checkbox"/>	
C) The patient reports at least one symptom in three or more of the categories		<input type="checkbox"/>	
D) No other diagnosis can better explain the signs and symptoms		<input type="checkbox"/>	
Category		Sign (you can see or feel a problem)	Symptom (the patient reports a problem)
1 'Sensory'	<i>Allodynia</i> (to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement) and/or <i>hyperalgesia</i> (to pinprick)	<input type="checkbox"/>	<i>Hyperesthesia</i> does also qualify as a symptom <input type="checkbox"/>
2 'Vasomotor'	Temperature asymmetry and/or skin colour changes and/or skin colour asymmetry	If you notice temperature asymmetry: must be >1°C <input type="checkbox"/>	<input type="checkbox"/>
3 'Sudomotor/oedema'	Oedema and/or sweating changes and/or sweating asymmetry	<input type="checkbox"/>	<input type="checkbox"/>
4 'Motor/trophic'	Decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair/nail/skin)	<input type="checkbox"/>	<input type="checkbox"/>

\* Terms used are explained in the glossary.

patients (who fulfilled the criteria in the past, but now have lost some or all limb signs, yet have ongoing pain) should be diagnosed with 'CRPS-NOS' (not otherwise specified).<sup>21</sup>

The onset of symptoms for the majority occurs within one month of the trauma or immobilisation of the limb.<sup>22</sup> There is no proven cure for CRPS. Approximately 15% of sufferers will have unrelenting pain and physical impairment two years after CRPS onset and are considered to have a long-term condition, although more patients will have a lesser degree of ongoing pain and dysfunction.<sup>23,24</sup>

Prompt diagnosis and early treatment are considered best practice in order to avoid secondary physical problems associated with disuse of the affected limb and the psychological consequences of living with undiagnosed chronic pain.<sup>25</sup> However, this standard of care has yet to achieve widespread practice in the UK. Since the condition is uncommon, and the range of symptoms can mimic a large number of other possible conditions seen by practitioners from various professional backgrounds (Fig 1), patients commonly experience a delay in diagnosis and the start of appropriate therapies.<sup>14,26</sup> The aim of these guidelines is to aid diagnosis in a range of primary and secondary care settings. This document will also provide guidance on how to manage CRPS with appropriate treatment or referral to other practitioners.

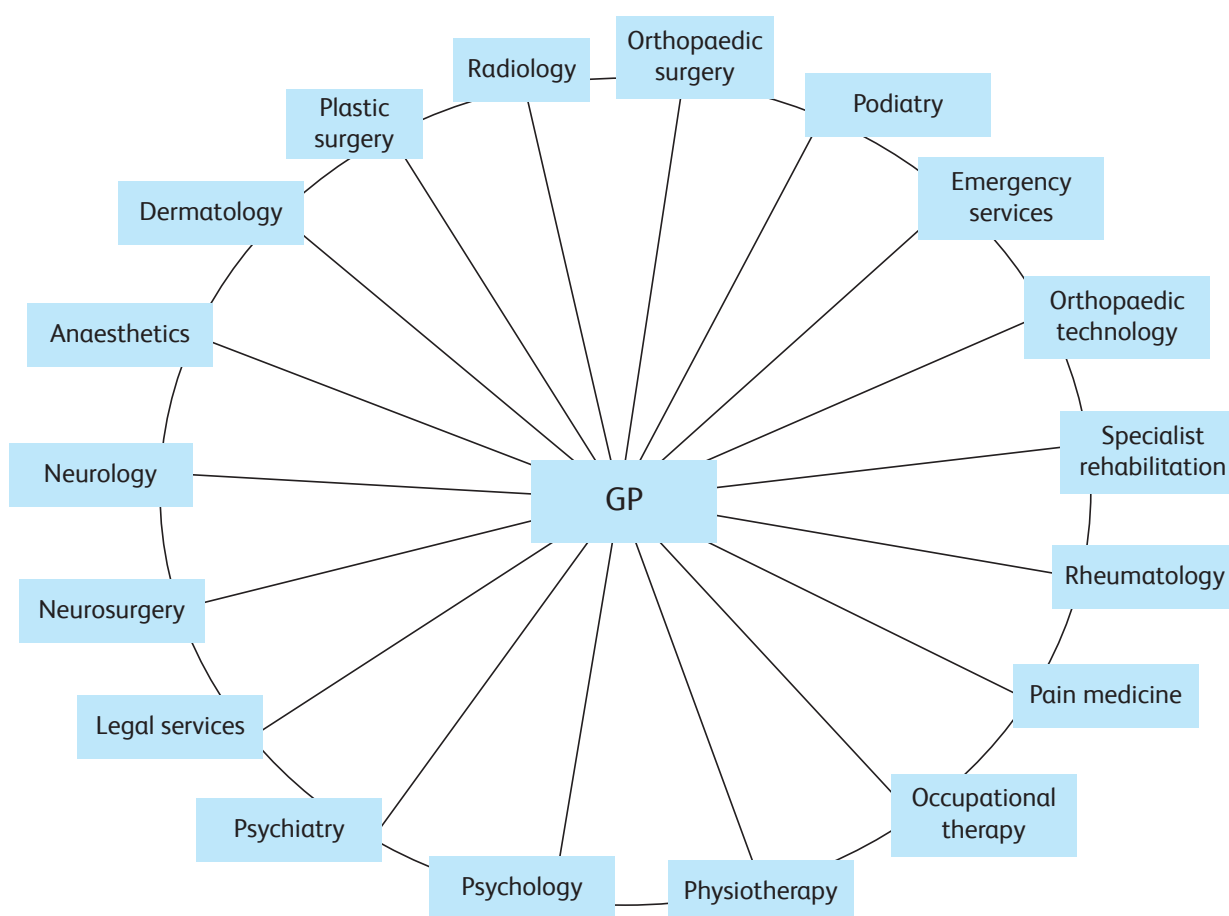


Fig 1 Range of services used by patients with CRPS

### Diagnostic criteria

CRPS is the term given to a group of painful conditions formerly termed as listed in Table 2. The diagnosis of CRPS is based on clinical examination and is given when patients meet the 'Budapest' diagnostic criteria described in Table 1.

**Table 2** Earlier names for CRPS

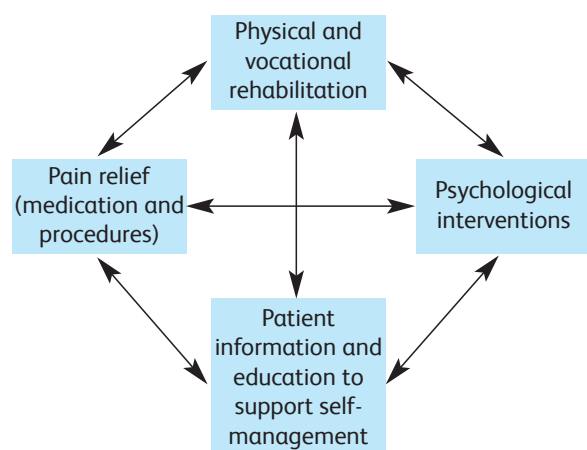
Algodystrophy	Causalgia
Algoneurodystrophy	Reflex sympathetic dystrophy
Sudeck's atrophy	Shoulder–hand syndrome
Reflex neurovascular dystrophy	Fracture disease

CRPS can be divided into two types based on the absence (type 1, much more common) or presence (type 2) of a lesion to a major nerve.\*† Currently this distinction has no relevance for management,\*\* but it can have importance in some medico-legal cases. Recent evidence suggests that even CRPS type 1 may be associated with subclinical neurological change.<sup>27</sup>

### Treatment approach

Pain is typically the leading symptom of CRPS and is often associated with limb dysfunction and psychological distress. For those in whom pain persists, psychological symptoms (anxiety, depression), and loss of sleep are likely to develop, even if they are not prominent at the outset. Therefore an integrated interdisciplinary treatment approach is recommended, tailored to the individual patient.<sup>28</sup> The primary aims are to reduce pain, preserve or restore function, and enable patients to manage their condition and improve their quality of life.<sup>29</sup>

The four 'pillars' of care (education, pain relief, physical rehabilitation and psychological intervention (see Fig 2)), which address these aims have equal importance. However, full recovery can be difficult to achieve in some, even with early appropriate treatment. Practitioners can support patients by providing a clear diagnosis, information and education about the disease, helping to set realistic goals and, where possible, involving the patient's partner and/or other family members. This document will provide guidance on how best to meet the treatment aims in a variety of clinical settings, for both acute and chronic CRPS.



**Fig 2** Four pillars of treatment for CRPS – an integrated interdisciplinary approach

\* The term 'major' within orthopaedic context is not fully defined.

† A third diagnostic subtype called CRPS-NOS (not otherwise specified) is recommended for patients who have abnormalities in fewer than three Budapest symptom categories, or two sign categories, including those who had more documented signs and symptoms in the past, if current 'signs and symptoms' are still felt to be best explained by CRPS.

\*\* As an exception, in orthopaedic practice, in CRPS 2, a nerve lesion can sometimes be directly treated (see 'Surgical management' in section on orthopaedic practice).

# Specialty guidelines

## Primary care

### Suspected or confirmed CRPS – diagnosis, management and referral in primary care in the UK: guidance for general practitioners and other primary care clinicians

Unilateral limb pain in patients presenting to the general practice surgery has many potential causes. This section of the guideline aims to add clarity to the diagnosis and management (including immediate, short-term and long-term management) of CRPS in primary care. The starting point of the guideline assumes a degree of suspicion of the presence of CRPS; an exhaustive differential diagnostic list is beyond the scope of this document (see the list of differential diagnoses in the ‘Rheumatology, neurology and neurosurgery’ section).

To improve the chance of a favourable outcome, three principal areas require attention:

- pain intensity
- limb dysfunction
- distress.

## Diagnosis of CRPS

### Recommendations

For best practice, GPs would:

- ▶ be aware of CRPS and be able to recognise the clinical signs (see Appendix 3)
- ▶ have access to a CRPS diagnostic checklist (see Appendix 4)
- ▶ apply knowledge of the ‘bio-psychosocial’ assessment of pain.<sup>30</sup>

## Referral

The main reasons to refer patients with CRPS are:

- to confirm diagnosis (pain services, neurology or rheumatology)
- to exclude ongoing pathology (eg orthopaedic, rheumatology or neurology services\*)
- when symptoms are difficult to control (pain services)
- to enable functional rehabilitation (pain and/or rehabilitation services).

It may be appropriate to manage *confirmed* CRPS in the primary care setting alone if the symptoms of CRPS are mild.

Pragmatically, to categorise CRPS as ‘mild’, a patient would have few signs of significant pain-related disability or distress and either conventional or neuropathic drugs would manage pain intensity adequately. Patients who exhibit high levels of pain, disability or distress should be referred for specialist advice; in the meantime, active rehabilitation should be initiated as early as possible.

### Recommendations

Referral of *suspected* CRPS is indicated in the following instances:

- ▶ For confirmation of the CRPS diagnosis.

---

\* In CRPS type 2 (defined as CRPS with associated damage to a major nerve), the cause for nerve damage, if unclear, should be assessed by a neurologist. CRPS can be triggered by nerve damage but does not cause nerve damage by itself.

- ▶ When pain treatment (see ‘Management of suspected or confirmed CRPS’ later in this section) is unsuccessful. In such cases, the patient should be referred to a pain specialist (in community or secondary care). This is essential even if other management is ongoing (eg by physiotherapy, orthopaedics or rheumatology). The GP should not rely on non-pain clinicians to manage the persistent pain.
- ▶ Even when the patient’s pain is mild and controlled if there are concomitant signs of pain-related distress or disability. In such cases, the patient should be referred to a multidisciplinary pain clinic (in community or secondary care). A list of pain clinics can be found at the National Pain Audit’s website ([www.nationalpainaudit.org/search.aspx](http://www.nationalpainaudit.org/search.aspx)).

#### After trauma or surgery

- ▶ When a patient is already discharged from the trauma or surgical team, the GP should consider re-referral – for example, to the attending orthopaedic specialist/surgeon or trauma service – to allow for definite exclusion of ongoing pathology.

#### Without trauma (or after minor trauma)

- ▶ Patients with suspected CRPS without preceding trauma should be referred to secondary care (eg rheumatology) to exclude or address specific pathology.
- ▶ Isolated referral to physiotherapy should be arranged with caution and only if the referrer is certain that there is no identifiable underlying cause (eg infection or other causes) (see ‘Rheumatology, neurology and neurosurgery’ for other causes).

#### Referral of confirmed CRPS

Other than in mild cases of CRPS (see ‘Referral’ earlier in this section), patients should be referred to a pain specialist for further management.

It may also be appropriate instead to refer cases of confirmed CRPS to specialist rehabilitation or vocational rehabilitation services if:

- ▶ CRPS presents in the context of another existing disabling condition (eg stroke or severe multiple trauma)
- ▶ specialist facilities, equipment or adaptations are required or need review
- ▶ the patient needs specialist vocational rehabilitation or support to return to work (this service is sometimes also provided by pain management services)
- ▶ litigation is ongoing, requiring support to facilitate an early conclusion.

## Management of suspected or confirmed CRPS

### Recommendations

For best practice, GPs:

- ▶ would aim to confirm the diagnosis – this may require further diagnostic opinion (see ‘Referral’ earlier in this section)
- ▶ would have access to evidence-based information to share with patients (see ‘Sources of information for patients on the web’ in Appendix 10)

Management of pain is important to minimise suffering. This should be done in parallel with any ongoing investigation and management of potentially relevant pathology that may be contributing to the pain.

- ▶ The aim of medication is to minimise pain and support physical rehabilitation. If the patient is waiting for an appointment with a pain specialist, they should be seen regularly and be advised about the use and titration of simple analgesics.



- ▶ If simple medication does not reduce the patient's pain to a mild level after 3–4 weeks, consider using medication for neuropathic pain according to neuropathic pain guidelines.<sup>\*31,32</sup> Earlier use may be appropriate.
- ▶ Other specialists may also initiate neuropathic pain drugs, but the GP is usually best placed to arrange the follow-up required for drug titration (see section on 'Orthopaedic practice').

There is currently 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.<sup>†</sup> This should include:

- ▶ gentle limb movement (unless contraindicated for orthopaedic reasons) (see section on 'Orthopaedic practice')
- ▶ frequent attention to the affected limb
- ▶ 'desensitisation' (gentle stroking of the affected limb with different textured fabrics while viewing the limb) (Appendix 5)
- ▶ progressing to more active use (eg weight bearing and stretching) when tolerated.

If there is any doubt about the safety of movement, the advice of an orthopaedic surgeon or rheumatologist should be sought.

Mild cases of CRPS may be managed with simple and/or neuropathic pain medications and general advice regarding exercise.

Good communication between primary and secondary care is essential:

- GPs and other primary care clinicians should be involved in the care initiated by the pain specialist. The treatment plan should be clear in all cases.
- The long-term management of treatment-resistant CRPS should be shared between the primary care clinician and chronic pain service and, where appropriate, specialist rehabilitation services (see recommendations on 'Referral' earlier in this section).
- Where multiple clinicians are involved, there is an increased risk of fragmentation of care or conflicting advice; this is well understood by the GP. Often part of the GP's role will be to ensure that the patient has a clear and consistent view of the problem. The GP can minimise any inconsistencies through objective interpretation of opinions and clear communication with the patient. The GP will often decide whether further clarification is required from the specialists involved.

\* For further information regarding neuropathic pain, see the national guidance documents for the UK published by the National Institute for Health and Clinical Excellence (NICE)<sup>31</sup> and Clinical Resource Efficiency Support Team (CREST)<sup>32</sup> (these documents were not developed for CRPS; however, it is considered appropriate to use this medication in the treatment of CRPS).

<sup>†</sup> Some GPs choose to refer to physiotherapy to enhance the encouragement for patients to stay active and move the limb, etc.

# Occupational therapy and physiotherapy

Rehabilitation of those with CRPS by occupational therapists and physiotherapists

## Settings

Occupational therapists and physiotherapists provide rehabilitation to those with CRPS in diverse settings, as categorised below:

- outpatient rehabilitation: either in the community or in hospital units
- inpatient rehabilitation: multidisciplinary inpatient rehabilitation is described in the section on 'Rehabilitation medicine'
- pain management programmes (PMPs): these are multidisciplinary programmes, normally (although not always) carried out in an outpatient group setting and often attached to pain medicine departments; a more detailed description is available in the British Pain Society's leaflet *Recommended guidelines for pain management programmes for adults* ([www.britishpainsociety.org/book\\_pmp\\_main.pdf](http://www.britishpainsociety.org/book_pmp_main.pdf)).

## CRPS rehabilitation algorithm for occupational therapists and physiotherapists

Figure 3 illustrates the recommended algorithm for rehabilitation of those with CRPS, and is based on expert opinion. The algorithm is suitable for both acute and more established CRPS. Decisions are based on the degree of symptom severity and treatment response. Patients referred to physiotherapy/occupational therapy with an already confirmed diagnosis of CRPS enter phase 3 of the algorithm.

### Phase 1: undiagnosed CRPS

A therapist\* in any rehabilitation setting may identify a patient presenting with signs and symptoms of CRPS without prior diagnosis by a medical doctor.

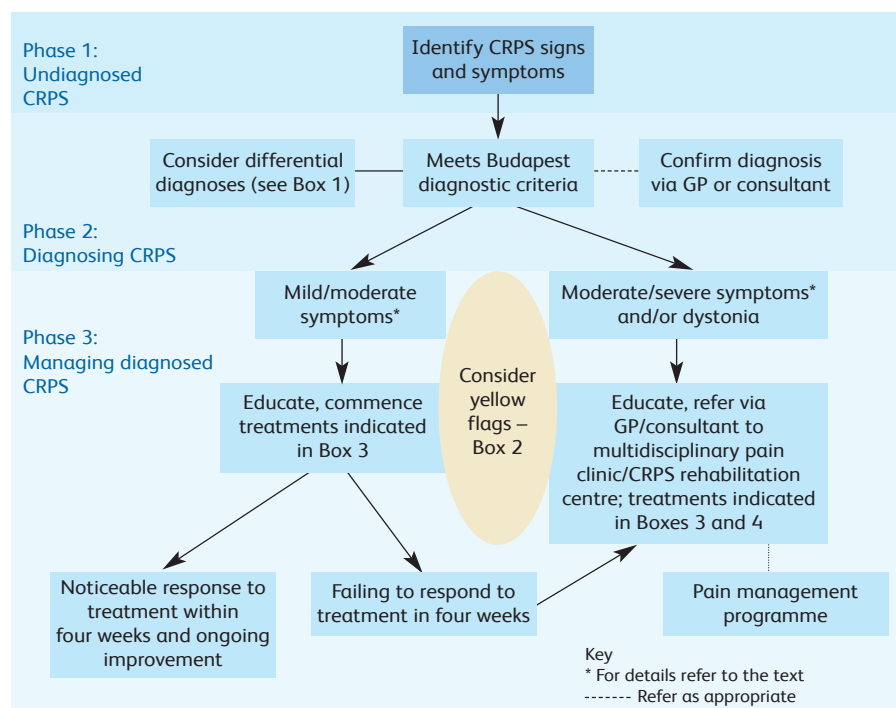


Fig 3 CRPS rehabilitation algorithm for occupational therapists and physiotherapists

\* Occupational therapist and physiotherapist are referred to as therapist.

## Phase 2: diagnosing CRPS

Confirmation of diagnosis is based on presenting signs and symptoms in accordance with the Budapest diagnostic criteria<sup>33</sup> (see Table 1 in 'Introduction' section). Although experienced therapists may make a diagnosis of CRPS (see Appendix 4), this generally should be confirmed by a doctor\* (eg the patient's consultant or GP).

It is important to consider other possible pathologies (differential diagnoses) during diagnostic assessment (Box 1).

### Box 1 Differential diagnoses<sup>†</sup>

- infection (bone, soft tissue, joint or skin)
- orthopaedic malfixation
- joint instability
- arthritis or arthrosis
- bone or soft tissue injury (including stress fracture, instability or ligament damage)
- compartment syndrome
- neural injury (peripheral nerve damage, including compression or entrapment neuropathy, or central nervous system or spinal lesions)
- thoracic outlet syndrome (due to nerve or vascular compression)
- arterial insufficiency (usually after preceding trauma, atherosclerosis in the elderly or thrombangiitis obliterans (Burger's disease))
- Raynaud's disease
- lymphatic or venous obstruction
- Gardner–Diamond syndrome (see the list of differential diagnoses in the 'Rheumatology, neurology and neurosurgery' section)
- brachial neuritis or plexitis (Parsonage–Turner syndrome or neuralgic amyotrophy)
- erythromelalgia (may include all limbs)
- self-harm

## Phase 3: managing diagnosed CRPS

The assessment of patients with CRPS by a physiotherapist or occupational therapist is described in Appendix 11.

### Assessment of CRPS severity

The degree of symptom severity is assessed to determine which of the two arms within phase 3 of the algorithm to follow.

### Mild CRPS signs and symptoms

To categorise CRPS as 'mild', a patient would have few signs of significant pain-related disability or distress, and either conventional or neuropathic drugs would manage pain intensity adequately (see refs 33 and 34 for

\* It is recognised that patients with early CRPS are often diagnosed, treated with physiotherapy/occupational therapy and discharged without formal confirmation of the diagnosis by a doctor; however, patients should in general be seen by a doctor for medication management.

† This list is not exhaustive.

further guidance on symptom severity). Patients who exhibit high levels of pain, disability or distress should be referred to a multidisciplinary pain clinic (ie two or more disciplines) or a rehabilitation CRPS unit.

Treatment should be initiated as early as possible. 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 approaches such as those outlined in Box 3.

If at any stage during treatment the therapist is unsure whether to continue with single-handed therapy or to refer for multidisciplinary rehabilitation, further advice should be sought from an experienced colleague.

### Moderate to severe presentation/poor treatment response

If one or more of the following features that indicate moderate or severe disease and/or poor recovery are present, an early referral to a multidisciplinary pain clinic or specialist unit is recommended:

- presentation with moderate to severe signs and symptoms (except if of very recent onset after trauma and quickly resolving)
- presence of dystonia
- no positive treatment response within four weeks
- condition deteriorates or improvements are not sustained despite ongoing treatment.

In addition to providing CRPS-specific rehabilitation techniques, specialist units may treat patients with advanced drug and interventional techniques, including spinal cord stimulation. A GP or consultant referral to a multidisciplinary pain clinic or CRPS specialist unit can be initiated by the therapist (see Appendix 8 for a list of centres specialising in CRPS). Some regions have direct therapy referral agreements with the local multidisciplinary pain clinic.

Descriptions of the rehabilitation provided by pain clinics and rehabilitation medicine are presented in the sections on 'Pain medicine' and 'Management of patients with CRPS and complex disability in rehabilitation services', respectively. Examples of CRPS-specific rehabilitation techniques are outlined in Box 4.

- After referral, it is important to continue treatment until the patient has been assessed by the pain clinic or CRPS specialist unit.
- In circumstances where the therapist works within a multidisciplinary team, referral back to the team member who originally referred the patient for additional or alternative input may be appropriate.
- Parallel referral to a multidisciplinary, psychology-led pain management programme should be discussed in accordance with the therapist unit's referral criteria.

### Yellow flags

Recognition of psychosocial risk factors, referred to as yellow flags (Box 2),<sup>35</sup> can support therapists in understanding contributing causes for suboptimal treatment response and should be considered within the context of phase 3 of the algorithm. The presence of these factors has been used in other pain conditions to predict chronicity.<sup>36</sup>

Yellow flags may be present at initial assessment or may develop and become apparent during treatment. Recognition of these flags may guide referral to multidisciplinary pain clinics and psychology-led PMPs.

**Box 2 Yellow flags.** Adapted from Main and Williams, 2002<sup>37</sup>

- iatrogenic factors, ie previous negative experiences with health professionals
- poor coping strategies, eg ongoing 'guarding' of the limb despite education
- involved in litigation, which is affecting willingness to progress with treatment (note that this is not the case for all patients involved in litigation)
- overuse of appliances
- distress
- anxiety/depression
- lack of willingness to set goals
- passive in treatment sessions
- inappropriate beliefs despite education
- negative family influences

**Treatment approaches****Box 3 Therapeutic approaches**<sup>38†</sup>

- patient education and support<sup>39</sup>
- desensitisation<sup>40</sup>
- general exercises and strengthening
- functional activities
- mirror visual feedback<sup>41–43</sup>
- gait re-education<sup>44</sup>
- transcutaneous electrical nerve stimulation (TENS)<sup>45</sup>
- postural control
- pacing, prioritising and planning activities<sup>46</sup>
- goal setting<sup>47–49</sup>
- relaxation techniques<sup>50</sup>
- coping skills<sup>51</sup>
- hydrotherapy<sup>52</sup>
- sleep hygiene<sup>53</sup>
- oedema control strategies<sup>54</sup>
- vocational support<sup>55</sup>
- facilitating self-management of condition<sup>56</sup>
- splinting<sup>\*\*57,58</sup>

\* This list is not exhaustive.

† None of these techniques have been assessed in randomised controlled trials; however, a combination of some techniques was more successful than social work in one trial.

\*\* Limited use where clinically appropriate; avoid prolonged periods of immobilisation or covering up of the limb.

**Box 4 CRPS rehabilitation undertaken in specialist units or by therapists with CRPS expertise**

- graded motor imagery<sup>59,60</sup>
- self-administered tactile and thermal desensitisation with the aim of normalising touch perception (see Appendix 5)<sup>61,62</sup>
- mirror visual feedback<sup>43</sup>
- strategies to correct body perception disturbance, involving looking, touching and thinking about the affected body part<sup>61,63</sup>
- mental visualisation to normalise altered size and form perception of affected body part<sup>61</sup>
- functional movement techniques to improve motor control and awareness of affected limb position<sup>64</sup>
- principles of stress loading<sup>65,66</sup>
- conflict allodynia re-education to reduce fear of physical contact with others in community settings<sup>67</sup>
- management of CRPS-related dystonia

**Recommendations**

For best practice, therapists would:

- ▶ be aware of CRPS and be able to recognise the clinical signs
- ▶ be aware of the Budapest criteria for diagnosing CRPS (see Appendix 4)
- ▶ initiate treatment as early as possible
- ▶ provide patient education about the condition
- ▶ know of the nearest multidisciplinary pain clinic or CRPS specialist rehabilitation centre
- ▶ recognise non-resolving moderate or severe symptoms and, where appropriate, initiate referral to a multidisciplinary pain clinic or CRPS specialist centre for rehabilitation.

# Orthopaedic practice

## Diagnosis, prevention, management and referral of patients with CRPS in orthopaedic practice in the UK

The available evidence suggests that transient CRPS is common after limb fractures and orthopaedic operations (up to 25 % of cases).<sup>68–70</sup> The pain improves in most cases and CRPS lasting longer than a few months is an uncommon condition (overall prevalence is less than one in 1,500), although even a transient episode of CRPS may give rise to long-term disability due to structural\* and/or functional changes.<sup>24</sup>

### Diagnosis

- Both the ‘Budapest criteria’ (see Table 1) and ‘Atkins criteria’ (see Appendix 6), the latter of which were developed in an orthopaedic context,<sup>†</sup> provide similar results in the diagnosis of CRPS in orthopaedic practice.
- CRPS is a diagnosis of exclusion. The Budapest criteria were developed to differentiate CRPS from the typical causes of pain seen in a pain clinic, but the causes of pain are different in an orthopaedic setting.<sup>21</sup> Both the Atkins and Veldman criteria were developed in an orthopaedic setting.<sup>71</sup> Some validation of the Budapest criteria within an orthopaedic context has been achieved.<sup>71</sup> Concerns remain about the applicability of these criteria in a judicial context, because the Budapest criteria rely heavily on patient-reported features, and in clinical orthopaedic practice, because of their complexity.<sup>72</sup> Summaries of both the Atkins and Veldman criteria are given in Appendix 6.
- In an orthopaedic context, alternative causes of persistent limb pain include infection, orthopaedic malfixation, instability, arthritis or arthrosis, and neuropathic pain from nerve entrapment or nerve damage.<sup>72</sup> ‘Scalding’ pain in the distribution of a peripheral nerve in an orthopaedic setting should be urgently reviewed by the surgeon because of the possibility of nerve damage related to surgery or injury.
- Plaster tightness and disproportionate pain while in plaster may be early warning signs for CRPS.<sup>73</sup>
- Sensory and motor ‘neglect-like symptoms’ (sensory: ‘my hand does not feel as if it belongs to me’; motor: ‘I cannot move my limb the way I want to, and this is not due to my pain’) are features of CRPS that may be common after trauma, even in the absence of CRPS.<sup>74,75</sup> These signs are unlikely to indicate a primary psychological dysfunction.

### Recommendations

- ▶ Orthopaedic surgeons should be aware that there are diagnostic criteria for CRPS, including the Budapest, Atkins and Veldman criteria. They should be able to use one of these sets of criteria in their clinical practice.
- ▶ Orthopaedic surgeons should be aware that CRPS may never fully resolve and that it often severely reduces patients’ quality of life and may be associated with increased psychological distress.<sup>76,77</sup>
- ▶ Orthopaedic surgeons should be aware that the diagnosis of CRPS can be made in patients who have only had minor soft tissue injury. It may even occur without a traumatic event.
- ▶ A diagnostic checklist for use in an orthopaedic setting should be available in orthopaedic departments, including outpatient departments and plaster rooms.

\* Such as contracture and ankylosis.

† In this guidance, use of the Atkins and Veldman criteria is considered or recommended only in an orthopaedic context and not in other contexts, such as pain medicine or general practice.

- ▶ Classic descriptions emphasise that bone involvement is universal in CRPS after trauma. At an early stage there will be increased uptake on the delayed part of the technetium-99m-methylene diphosphonate (Tc-99m MDP) bone scan and at a later stage there will be osteoporosis. However, CRPS is a clinical diagnosis that does not depend on the results of a bone scan. The routine use of three-phase bone scans is not necessary and may delay the start of treatment,<sup>78</sup> so it is not recommended.
- ▶ General post-fracture/operation patient information leaflets should include advice to observe and report warning signs for CRPS. A sample leaflet is available (see Appendix 7).

## Management

- The development of CRPS should not be considered evidence of suboptimal surgical management.<sup>72</sup>

## Recommendations

- ▶ Healthcare professionals involved in the care of patients with CRPS within orthopaedic practice should be aware of the basic principles of CRPS therapy (refer to the four pillars of care described in the 'Introduction' section).
- ▶ Management should include reassurance that the pain will either completely or partially resolve in at least 85 % of cases, although ongoing motor dysfunction with limb disability may be common.<sup>76</sup>
- ▶ The surgeon should reassure the patient that CRPS is a recognised condition, although its causes are poorly understood.
- ▶ Physiotherapy and/or occupational therapy, unless contraindicated, should be initiated immediately when CRPS is suspected. General advice should include focusing on the affected limb, gentle movement and desensitisation. Temporary splinting in a position of safety may relieve pain and be an adjunct to mobilisation, but the treatment of early CRPS is generally by gentle mobilisation; immobilisation of a joint in a patient with CRPS carries a risk of long-term stiffness. Early functional weight-bearing is to be encouraged to accelerate rehabilitation. Orthotic devices such as insoles can support weight-bearing but require a physiotherapist's supervision. Excessive mobilisation that exacerbates pain is contraindicated.
- ▶ The orthopaedic team should initiate early treatment with simple analgesic drugs. These may include codeine, dihydrocodeine, tramadol, non-steroidal anti-inflammatory drugs (NSAIDs) and paracetamol, as appropriate.\* These drugs do not necessarily affect the specific pain of CRPS but may reduce ongoing trauma-related pains and assist in the process of mobilisation.
- ▶ Orthopaedic surgeons may initiate treatment with other drugs useful for neuropathic pain, such as tricyclic antidepressants (amitriptyline, nortriptyline or imipramine)<sup>7</sup> and anticonvulsants (gabapentin<sup>79</sup> or pregabalin), but the GP or pain specialist is usually best placed to arrange the follow up required for drug titration (see section on 'Primary care'). If a patient requires anticonvulsive or antidepressant drugs or strong opioids for the control of neuropathic pain or the treatment of CRPS, serious consideration should be given to urgent referral to a pain therapist.
- ▶ Guanethidine blocks should not be used in orthopaedic practice (see 'Pain medicine' section for clarification).<sup>4,80</sup>
- ▶ Orthopaedic surgeons should be aware of specific treatments for chronic CRPS, such as specialist physiotherapy and occupational therapy, multidisciplinary pain management programmes, spinal cord stimulation and specialist rehabilitation programmes.

\* The Guideline Development Group acknowledges that there is currently no evidence for the efficacy of these drugs in CRPS. Amitriptyline, nortriptyline and imipramine can be effective in neuropathic pain.



## Surgical management of patients with CRPS

- Amputation may worsen CRPS, with CRPS recurring in the stump.<sup>81</sup>
- If elective surgery on a limb previously affected by CRPS is delayed until signs of CRPS have resolved, the rate of operation-triggered recurrence of CRPS is <15%, with most recurrent cases being mild.<sup>82,83</sup>
- Expert opinion suggests that complications following surgery in patients with CRPS may be common. Reasons may include the adverse reaction of patients with CRPS to surgical pain and the adverse impact of body perception disturbances<sup>26</sup> and poor motor control<sup>84</sup> on rehabilitation; however, this field needs further study.

### Recommendations

- ▶ Amputation should not be used to provide pain relief in CRPS.<sup>81</sup> Amputation may be considered in rare cases of intractable infection of the affected limb.<sup>85</sup>
- ▶ Surgery should be avoided on a CRPS-affected limb where possible and be deferred where it cannot be avoided until one year after the active process has resolved.<sup>82</sup>
- ▶ Surgery may be indicated in CRPS type 2 when there is an identifiable remediable nerve lesion (eg certain cases of neuropathic pain due to either nerve compression by scar tissue, neuroma formation or perioperative nerve injury, such as through a needle stitch) but should be undertaken only when, on balance, the expected benefit from pain reduction outweighs the risk of exacerbation.
- ▶ Where surgery on an affected limb is necessary, this ideally should be performed by a surgeon with experience in operating on patients with CRPS and an anaesthetist who is also a pain specialist (see the section on 'Pain medicine').

## Prevention

There is insufficient evidence for any prophylaxis for CRPS.\*

### Recommendations

- ▶ Although there is no evidence that early physiotherapy can prevent CRPS, early diagnosis and treatment with physiotherapy and/or occupational therapy, delivered by therapists competent in treating patients with chronic pain and/or CRPS, is recommended to reduce suffering, improve function, prevent complications such as contractures and speed recovery from CRPS.

## Referral

### Recommendations

- ▶ If treatment with simple analgesic drugs does not satisfactorily reduce pain, the patient should be referred to the pain clinic for multidisciplinary treatment. Ideally multidisciplinary pain therapy treatment should begin within three months of the onset of the condition.
- ▶ If psychological factors (eg significant distress)\*\* are thought to be important in a patient, early referral to a psychologist specialising in pain may be advisable.<sup>†</sup>

\* Although some evidence suggests that vitamin C can reduce the incidence of CRPS after distal radius fracture, the panel finds that this evidence is insufficient and does not warrant recommendation of this intervention.

\*\* Such as fear and anxiety, depression, rigid beliefs and significant avoidance or protective behaviour, which present significant barriers to recovery, rehabilitation and daily functioning.

† Such a referral is often arranged through a pain specialist.

- ▶ Non-resolving CRPS currently under the care of other departments or professionals (eg pain clinics and/or physiotherapists) but that originally developed in an orthopaedic context should be considered for review by an orthopaedic specialist for an opinion regarding alternative or contributing causes (see also the section on 'Pain medicine').

# Rheumatology, neurology and neurosurgery\*

Management of suspected and confirmed CRPS in rheumatology, neurology and neurosurgery

## Diagnosis

Earlier, now superseded, terms for CRPS are given in Table 2 in the 'Introduction' section. In patients with an established diagnosis of CRPS, neurologists are sometimes asked to determine whether there is accompanying nerve damage and, if there is nerve damage, to determine whether the same injury that caused the nerve damage had caused CRPS (ie CRPS type 2) or whether the nerve damage is concomitant or preceded the CRPS.

### Box 5 Selected differential diagnoses for CRPS in rheumatology, neurology and neurosurgery, although not necessarily in all three disciplines

- bone or soft tissue injury (including stress fracture, ligament damage and instability)
- compartment syndrome
- neuropathic pain (eg due to peripheral nerve damage including compression or entrapment neuropathy or due to central nervous system or spinal lesions)
- arthritis or arthrosis
- thoracic outlet syndrome (due to nerve or vascular compression)
- infection (bone, soft tissue, joint or skin)
- arterial insufficiency (usually due to atherosclerosis in the elderly, trauma or thrombangiitis obliterans (Burger's disease))
- lymphatic or venous obstruction
- Raynaud's disease
- Gardner–Diamond syndrome<sup>†</sup>
- brachial neuritis or plexitis (Parsonage–Turner syndrome or neuralgic amyotrophy)
- erythromelalgia (may include all limbs)
- self-harm<sup>19</sup>

## Recommendations

- ▶ Rheumatologists, neurologists and neurosurgeons should be familiar with the Budapest criteria<sup>21</sup> for the diagnosis of CRPS (see Table 1 of the 'Introduction' section; see Appendix 4 for more details).
- ▶ In persistent limb pain, in the absence of a neurological or neurosurgical explanation, a diagnosis of CRPS should be considered (see Appendix 4). This also applies if a lesion is identified but the pain is disproportionate.

\* Neurology and neurosurgery here relates to general neurology and neurosurgery. Pain-related neurosurgery and guidance for neurologists working in pain clinics can be found in the section on 'Pain medicine'.

<sup>†</sup> Psychogenic purpura (Gardner–Diamond syndrome, autoerythrocyte sensitisation or painful bruising syndrome) is a rare and poorly understood clinical presentation of unexplained painful ecchymotic lesions, mostly on the extremities and/or face.

## Management and referral

For surgical management, refer to the 'Orthopaedic practice' section.

### Recommendations

- ▶ Rheumatologists, neurologists and neurosurgeons should be aware of advanced treatments for CRPS, including specialist physiotherapy and occupational therapy, multidisciplinary pain management programmes, spinal cord stimulation and specialist rehabilitation programmes.
- ▶ In neurological and neurosurgical practice, patients with CRPS should generally be referred to a pain unit for comprehensive assessment and/or specialist treatments (see list of available clinics available at: [www.nationalpainaudit.org/search.aspx](http://www.nationalpainaudit.org/search.aspx)).
- ▶ However, if individual specialists with a special interest in CRPS wish to manage the condition, the four 'pillars' of treatment (pain relief, physical and vocational rehabilitation, psychological intervention, and patient education and self-management), as described in the 'Introduction' section, have equal importance and should be delivered with an integrated interdisciplinary approach.
- ▶ To facilitate this, the following aspects of care should be available to patients:
  - expertise in the physical rehabilitation of patients with chronic pain conditions
  - management of pain (see section on 'Pain medicine')
  - psychological intervention specific to pain in the form of a multidisciplinary, usually cognitive behavioural therapy (CBT)-oriented, PMP<sup>86</sup> (see the British Pain Society (BPS)'s guidance on PMPs available at: [www.britishpainsociety.org/pub\\_professional.htm#pmp](http://www.britishpainsociety.org/pub_professional.htm#pmp)); most PMPs integrate the physical rehabilitation aspect with CBT\*
  - patient education being an important part of treatment; patient information resources are available (see Appendix 11)
  - specialists being aware that there are also centres with a special interest in CRPS for referral (see Appendix 8).

\* Cognitive behavioural therapy (CBT) is not a single therapy or even a single set of standardised 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.<sup>87</sup>

# Dermatology

## Diagnosis

In dermatology, CRPS has previously been known by a number of other, now superseded, terms, which are listed in Box 6. Higher awareness of CRPS is required by dermatologists, as early diagnosis helps management and recovery.

In addition to the typical trauma, antecedent factors that may lead to presentation to dermatologists include stroke, myocardial infarction, tuberculosis and *herpes zoster*, as well as more distinct dermatological events such as vasculitis, Weber–Christian syndrome, nail biopsy, excisional skin biopsy and epitheloid haemangioma. Although it is not clear how some conditions lead to CRPS, the common unifying factor in most instances is trauma. In leg ulcers, the injury from the ulcers themselves is probably involved.

The skin presentation of CRPS seems to take distinct forms that are sometimes sequential. There can be vasodilation and sudomotor dysfunction (hot, red and dry skin) or signs of vasoconstriction and hyperhidrosis (cold, blue and sweaty skin). These signs and symptoms can interchange.

Recognition of changes in skin may help in the diagnosis. The skin in CRPS may become either thin and glossy or thickened. Nails may show decreased or increased growth or thickening, become brittle or develop striations. In addition, hair growth can be increased or decreased. These changes may be due to vasoconstriction (resulting in skin hypoxia) or decreased range in motion of the skin from inactivity of underlying joints, tendons or ligaments. In chronic cases, skin sometimes becomes thin, atrophic and dry. Fingertips may diminish in volume, and deeper structures, including fascia, may become thickened, resulting in contractures. However, many patients have no trophic skin changes.

### Box 6 Dermatological manifestations of CRPS. Source: Pheps RG, Wientz S. Reflex sympathetic dystrophy. *Int J Dermatol* 2000;39:481–6

- erythema
- skin atrophy
- oedema
- hypohydrosis
- warmth
- hyperhydrosis
- pallor
- Beau's lines in nails
- cyanosis
- factious ulcers
- hypertrichosis
- bullae
- hypotrichosis
- leukonychia
- nail ridging
- onychodystrophy

## Recommendations

- ▶ The diagnosis should be based on the Budapest criteria.<sup>21</sup>

The Budapest criteria achieve 80–90% accuracy. Given that no objective tests have been validated, the diagnosis is clinical.

## Referral

### Recommendations

- ▶ Patients who are first diagnosed in dermatology should be referred to a pain management team.

Dermatologists may also receive referrals for dermatological input in the management of severe skin manifestations in patients with CRPS.

## Management

Treatment principles should be those outlined in the 'Introduction' section.

### Recommendations

- ▶ A multiprofessional approach is essential.
- ▶ Dermatologists should be aware that patients with problematic oedema may require spinal cord stimulation by a pain specialist or neurosurgeon.
- ▶ Recommendations for the treatment of skin ulcers, skin infection and problematic oedema are described in Appendix 9.

# Pain medicine

Diagnosis and management of CRPS in pain clinics in the UK. Guidance for pain physicians\* and neurosurgeons with a special interest in the management of pain

## Diagnosis

### Recommendations

- ▶ Pain specialists and neurosurgeons with a special interest in the management of pain should be aware of the Budapest criteria for the diagnosis of CRPS<sup>21</sup> (see 'Introduction' section and Appendix 4).
- ▶ These criteria rely on a clinical examination to diagnose CRPS. The use of additional tests is not recommended. However, in some instances, certain tests such as magnetic resonance imaging (MRI) and nerve conduction measurements may be appropriate to exclude other diagnoses.

## Management

### Recommendations

- ▶ Patient information and education is an important part of the management of CRPS. Template information leaflets are included in Appendix 10, with additional links to other sources of patient information on the web.
- ▶ Patient and doctor should agree on a treatment plan.
- ▶ If there is a lack of progress, the treatment plan should be re-evaluated.

### Drug treatment\*\*

- ▶ No drugs are licensed to treat CRPS in the UK.
- ▶ Drugs with efficacy in neuropathic pain should be used according to the National Institute for Health and Clinical Excellence (NICE) guidelines for neuropathic pain<sup>31</sup> and the recommendations of the Neuropathic Pain Special Interest Group of the International Association for the Study of Pain (IASP).<sup>†88,89</sup>
- ▶ Pamidronate (single 60 mg intravenous dose) should be considered for suitable patients with CRPS less than 6 months in duration as a one-off treatment.<sup>††</sup>
- ▶ Intravenous regional sympathetic blocks (IVRSB) with guanethidine should not be used routinely in the treatment of CRPS, as four randomised controlled trials have not demonstrated any benefit.

Some additional drugs demonstrate efficacy to treat pain in CRPS. For information only, a summary of the results from two systematic reviews is given in Appendix 14. However, no treatment recommendations are given due to combinations of the following factors:

- ▶ The evidence is only preliminary.
- ▶ Some drug treatments are not feasible for use in a clinical setting.
- ▶ Other drugs (eg intravenous immunoglobulin) are unavailable or restricted in the UK.

\* In pain centres, pain physicians may have various backgrounds, including anaesthetics, rheumatology and neurology.

\*\* The diagnosis of CRPS type 1 or 2 has no consequence for drug management. As an exception, in orthopaedic practice, in CRPS 2, a nerve lesion can sometimes be directly treated (see 'Surgical management' in 'Orthopaedic practice' section).

† These documents were not developed for CRPS. However, it is considered appropriate to use this medication in the treatment of CRPS.

†† The panel recognises that there may be other, newer types of bisphosphonates that may be appropriate/available in equivalent doses.

- ▶ Physical and vocational rehabilitation:
  - Both early and late CRPS should be treated with physiotherapy and/or occupational therapy<sup>90</sup> delivered by therapists competent in treating patients with chronic pain and/or CRPS. Physiotherapists and/or occupational therapists should be involved in the management of patients as early as is possible in the treatment pathway.
  - The occupational therapist associated with the pain service should:
    - address functional needs (self care, leisure and work)
    - teach pacing and relaxation strategies to support self-management of the condition
    - be aware of regional support services for return to work or to maintain existing employment.
  - Where such physiotherapy and/or occupational therapy services are not available within the pain management setting, a member of the pain team should be aware of regional support services such as specialist rehabilitation/vocational rehabilitation programmes (refer to 'Rehabilitation medicine' section).
  - Qigong (Tai chi) exercises should be considered.<sup>91</sup>
- ▶ Interventions:
  - Spinal cord stimulation should be considered in patients with CRPS who have not responded to appropriate integrated management, including specialised pain physiotherapy. This treatment can be carried out only in specialised centres (see BPS website\* and NICE guidance\*\* for further information). Pain specialists should be aware that there is some evidence that the efficacy of this treatment generally declines over time.<sup>92,93</sup>
- ▶ Psychological interventions:
  - The treatment of chronic pain should include provision of psychological interventions specific to pain in the form of a multidisciplinary pain management programme, for those who require it, based on an appropriate assessment method.<sup>†92</sup> Psychological interventions usually follow principles of CBT (see footnote to p18).

In some centres, interventions (including injection of local anaesthetic solution to the sympathetic chain, epidural catheters delivering a local anaesthetic and clonidine, or interscalene indwelling catheters) are used with an aim of 'breaking the cycle' of pain or aiding physiotherapy. Although there is currently no conclusive evidence for this practice from randomised controlled trials, considerable anecdotal evidence suggests that pain levels can remain low after such intervention. More research is needed before these methods can be formally recommended. We also note that some pain centres continue to provide IVRSB with guanethidine in subsets of patients with CRPS who show positive responses to such treatment. There is emerging evidence that guanethidine itself is not active, but that the other components of the treatment, including the tourniquet, are active (Mbizvo FKM *et al*, in preparation).

## Surgery

If surgery is considered necessary on limbs currently or previously affected with CRPS, or in patients in whom other limbs are currently or have previously been affected, clinicians should note that there is no evidence that any particular anaesthetic technique is superior in preventing recurrence of CRPS, postoperative pain exacerbation or new CRPS in a previously unaffected limb. (Please refer to the section on 'Orthopaedic practice' for orthopaedic surgeons for a discussion of the risks of operations in suspected or diagnosed CRPS.)

\* [www.britishpainsociety.org/books\\_scs\\_patient.pdf](http://www.britishpainsociety.org/books_scs_patient.pdf)

\*\* [www.nice.org.uk/nicemedia/pdf/TA159Guidance.pdf](http://www.nice.org.uk/nicemedia/pdf/TA159Guidance.pdf)

† As in other chronic pain situations, some patients with significant distress may need individual therapy as a precursor to group-based treatment.



## Treatment for symptoms and/or signs other than pain

For CRPS-related limb dystonia, intrathecal baclofen treatment can be considered only if all other options, including oral medications, have failed.<sup>95</sup> Intrathecal treatment should be delivered only in specialised centres; its adverse event profile is poor. In some cases, no effective tolerable treatment can be established. The overall efficacy of regional botulinum toxin for CRPS-related dystonia is poor; some patients might experience improvement.<sup>96</sup> Serial splinting by experienced physiotherapists may symptomatically improve some cases of dystonia, but care should be taken to give time to exposing the limb for the conduct of desensitisation therapies (see Appendix 5).

In cases of refractory, disabling limb swelling, advice from a lymphoedema nurse should be sought (see Appendix 9). 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 (see Appendix 9). Where ulcers occur in parallel with limb oedema, reduction of the oedema with spinal cord stimulation can promote ulcer healing.

## Prevention

There is no recommendation for any prophylaxis for CRPS because of insufficient evidence.

## Experimental treatments

Although the systematic review, which underpins the guidelines, included only RCTs, many other treatments for CRPS have been studied. A list of these treatments, with references, is provided in Appendix 15. However, these treatments should generally not be used outside a research setting.

## Referral

### Recommendations

- ▶ Patients should be referred in a timely manner based on individual assessment.
- ▶ Complex regional pain syndrome that developed in the context of orthopaedic practice, eg fracture, should be considered for referral for a further orthopaedic opinion to exclude the contribution of mechanical causes such as instability, malfixation, arthrosis, infection and neuropathic pain from nerve entrapment.
- ▶ Practitioners should consider referral of patients to tertiary or specialised secondary pain management care for the following reasons:
  - specialised treatment eg spinal cord stimulation or a pain management programme
  - subgroup management for non-resolving, worsening or highly distressing CRPS; CRPS with dystonia; blistering skin changes; ulceration; lymphoedema or myoclonus; and children/adolescents.
  - further consultation and assessment or if the patient requests a second opinion.
- ▶ Patients with blistering skin changes, ulceration, skin infection or disabling limb swelling should be referred to a dermatologist and those with isolated disabling limb swelling to a lymphoedema nurse. Pain specialists should be aware that spinal cord stimulation can reduce limb swelling.<sup>97</sup>

- ▶ Pain specialists should be aware of centres with a special interest in CRPS treatment (see Appendix 8) and of the nearest centre with a neuromodulation service. Pain specialists should also be aware of the role of specialist rehabilitation services in supporting patients with severe complex disability and those requiring vocational support (see section on 'Rehabilitation medicine').
- ▶ At discharge, information about CRPS (see Appendix 3) and its management should be sent to the patient's GP with the clinic letter.

## Competence

### Recommendations

- ▶ Pain specialists, including GPs and neurosurgeons with an interest in neuromodulation, and all allied health professionals should be aware of available treatments for CRPS and their effectiveness.

# Rehabilitation medicine

## Management of patients with CRPS and complex disability in rehabilitation services

### Specialist rehabilitation

A proportion of patients with CRPS will have complex combined physical, emotional, psychological and behavioural disability, and may require the support of a specialist rehabilitation service. The provision of specialist rehabilitation services for musculoskeletal conditions is currently patchy across the UK, and even where these services exist, they are often poorly integrated into the CRPS care pathway. The aims of this section are to highlight the availability of this resource to pain management specialists, to detail its strengths, and to confirm for the benefit of the rehabilitation team those requirements specific to CRPS.

Definitions of ‘specialist rehabilitation’ are given in Box 7.

#### Box 7 Definitions of rehabilitation

- Rehabilitation medicine is the medical specialty concerned with the prevention, diagnosis, treatment and rehabilitation management of people with disabling medical conditions.
- Rehabilitation is a process of assessment, treatment and management by which the individual (and their family/carers) are supported to achieve their maximum potential for physical, cognitive, social and psychological function; participation in society; and quality of life. It is divided into two main approaches:
  - restorative: a goal-orientated process by which the individual is supported to achieve optimal function and independence
  - disability management: a collaborative approach in which the team works with the patient and their family to support adjustment to change, prevent avoidable complications and minimise the effects of a disabling condition.
- Specialist rehabilitation teams are interdisciplinary and led or supported by a consultant trained and accredited in rehabilitation medicine. They work closely with other specialties (eg neurology, rheumatology, orthopaedics, pain specialists, neuropsychiatry, etc) to support patients and their families with complex medical, physical, emotional, behavioural and psychological needs arising from long-term disabling conditions.

Some patients with CRPS and complex disability can develop ‘learned disuse’ and marked disability behaviour in their attempts to avoid pain, which may be compounded by well-meaning but inappropriate support from local disability services and, in some cases, family.<sup>98</sup> For these patients, early engagement of specialist rehabilitation services with the patient, their family and their local services may break this cycle. Close liaison between pain management and specialist rehabilitation teams can enhance independence and participation,<sup>25,98</sup> mitigate effects from perverse incentives,\* and ensure long-term care and support.

### Diagnosis

The diagnosis of CRPS is made on the basis of the Budapest criteria (see Table 1 in ‘Introduction’ section and Appendix 4 for more details). Some patients will progress to develop long-standing CRPS. Over time, the typical vasomotor changes may become less prominent. Patients can therefore no longer fulfil the Budapest

\* A perverse incentive is an incentive that is likely to be withdrawn if the patient makes functional improvement.

criteria but nevertheless have ongoing significant pain and/or motor and trophic dysfunction. The Budapest criteria suggest the use of the diagnostic subtype 'CRPS not otherwise specified' (CRPS-NOS) for patients who do not fully meet the criteria but whose signs and symptoms could not be explained better by another diagnosis. For patients who fulfilled the Budapest criteria in the past but no longer do so, the term CRPS-NOS may also be used. Where patients present later in the course of their condition (as is often the case in rehabilitation medicine), careful history-taking is required to establish whether autonomic changes were present earlier on. Because painful symptoms will have persisted for a while, it is expected that there will often be some features of psychological distress. In the remainder of this section, the term 'CRPS' is used as shorthand for both CRPS and CRPS-NOS.

## Patient selection and referral

Examples of patients who may benefit from input from specialist rehabilitation include those:

- with CRPS-related severe complex disability
- with CRPS presenting in the context of another existing disabling condition (eg stroke or severe multiple trauma)
- with complex psychological or psychiatric comorbidities – either predating or postdating the onset of CRPS
- who require specialist facilities, equipment or adaptations or review
- who are unable to work and require specialist vocational rehabilitation or support
- who have ongoing litigation and require support to facilitate an early conclusion.

Whether a patient with complex disability is primarily under the care of the interdisciplinary pain team with input from specialist rehabilitation, or vice versa, will depend on the required key elements of the treatment programme and the expertise and resources within the respective teams (Table 3). In general, most ambulant patients are best managed primarily by the pain team, with support, where needed, from specialist rehabilitation. However, some patients will have conditions that render them unsuitable for treatment in a pain management programme and are better managed primarily by the rehabilitation team. This includes, for example, patients with severe concomitant physical or psychiatric disability (including some cases in which either the team or the patient feel that group treatment is not appropriate); in addition, patients, in whom the consistency and structure of a 24-hour rehabilitative milieu is required to retain and carry over gains, may require management in an inpatient rehabilitation setting. Simultaneous support from the pain team is essential when the rehabilitation team infrequently sees patients with CRPS, or does not have the resources to provide cognitive behavioural therapy.

**Table 3 Key elements of expertise for the treatment of patients with CRPS and complex disability that may be provided by specialist rehabilitation and pain teams**

Specialist rehabilitation team	Specialist pain team
<ul style="list-style-type: none"> <li>• complex disability self-management</li> <li>• assessment and provision of special facilities and equipment</li> <li>• vocational rehabilitation</li> <li>• support for litigation (to facilitate an early conclusion)</li> </ul>	<ul style="list-style-type: none"> <li>• cognitive behavioural psychology</li> <li>• pain-relief strategies (summarised in the section on 'Pain medicine')</li> <li>• specialist physiotherapy and occupational therapy techniques including novel therapies for CRPS (see the section on 'Occupational therapy and physiotherapy'), such as mirror training<sup>43</sup> or graded motor imagery,<sup>99</sup> desensitisation (see Appendix 5), pacing and relaxation*</li> </ul>

\* These therapies may also be provided by some specialist rehabilitation teams.

## Management

A coordinated multidimensional programme is required,<sup>100</sup> which should be delivered in the context of a cognitive behavioural approach. Key elements include:

- engagement – education and information for the patient and their family
- medical management
- psychosocial and behavioural management
- physical management
- activities of daily living and societal participation.

(Further details are given in Appendix 12, which is adapted from the British Society of Rehabilitation Medicine (BSRM) report on musculoskeletal rehabilitation.)<sup>100</sup>

Vocational and litigation support may be provided by specialist rehabilitation teams and, in other cases, these services are integrated into the regional pain management programme.

### Vocational rehabilitation\*

- Inactivity can compound the pain experience and the physical consequences of disuse.
- The loss of employment and its financial consequences serve to compound the psychosocial disadvantage experienced by patients and their families.
- Referral to the disability employment adviser may be required to access the various work support programmes available from the Department of Work and Pensions (DWP). However, because staff at the DWP may lack understanding of CRPS, liaison may be required through specialist vocational support programmes.

### Litigation

- Because of the association between CRPS and injury or minor surgery, claims for compensation are not uncommon.
- Litigation tends to fuel stress, which may adversely affect outcomes and ability to engage in rehabilitation.<sup>101</sup>
- Consultants in rehabilitation medicine may have a useful role in this context, as medicolegal training is a standard part of the curriculum for rehabilitation medicine.

## Recommendations

In the management of patients with CRPS and complex needs:

- ▶ Patients with complex disabling CRPS should have access to specialist interdisciplinary rehabilitation programmes led or supported by a consultant in rehabilitation, as described in Appendix 12 (examples of referral criteria are listed under 'Selection and referral').
- ▶ Specialist rehabilitation teams and pain management services should work together in close liaison to share their expertise and resources for the management of patients with CRPS and complex needs.

\* The BSRM has recently published guidelines to vocational rehabilitation in long-term neurological conditions, which provide detailed guidance on specialist vocational rehabilitation. (British Society of Rehabilitation Medicine. *Vocational assessment and rehabilitation for people with a long-term neurological condition: interagency guidelines*. London: BSRM; 2010.)

- ▶ Whether the patient is primarily under the care of the interdisciplinary pain team with input from specialist rehabilitation, or vice versa, management should depend on the key elements of the programme that are required and on the expertise and resources within the respective teams (see Table 3 earlier in this section).
- ▶ In either situation, care should be delivered in the context of a cognitive behavioural approach involving both the patient and their family.
- ▶ The rehabilitation programme should be goal orientated, with active engagement of the patient and their family in setting goals so that the patient remains in control and responsible for the rate of progress.
- ▶ Patients should have access to vocational assessment at an early stage in the condition to support them to stay in work if possible.
- ▶ Ongoing specialist vocational support should be provided in conjunction with the disability employment adviser to access the various work support programmes available from the Department of Work and Pensions.
- ▶ If productive work is impossible, patients should have appropriate support to withdraw from work and vocational rehabilitation efforts should focus on leisure and social activities instead.
- ▶ If the patient is engaged in litigation with respect to their CRPS, the rehabilitation team should provide support to facilitate its conclusion as soon as possible.

## Long-term support in CRPS

A proportion of patients with CRPS will have ongoing symptoms requiring long-term support. Patients report the following with respect to long-term management:

- They are concerned that many doctors and therapists have little understanding of CRPS and its management (see 'Sources of information for patients on the web' in Appendix 10). They are therefore worried about being discharged from specialist pain and other services, as it is often difficult to re-access specialist expertise when required after discharge.
- They are afraid of relinquishing social benefits and financial support as their condition improves in case they are unable to manage back at work, especially if their condition fluctuates from day to day.
- They want to be in control of their own condition and to remain as active and independent as possible. The following would enable them to do this:
  - better information and advice to help them manage their own condition, particularly about the forms of support that may be available to help them return to or remain in work
  - flexible support (eg through personalised budgets) to allow them to make their own choice about the types of support that will work best for them at any given stage in their disease.

As long as the lifeline to the specialist service remains in place, patients are generally happy to accept fairly low levels of contact. Feasible models of support included:

- annual review by the pain clinic, possibly via telemedicine (eg by telephone or postal questionnaire)
- a named single point of contact within the pain team (eg nurse, doctor or physiotherapist) who they could contact if needed
- being included in a register or flagged in some way on the general practice records, so that they could get rapid access if needed in case of a flare of their symptoms
- access to self-help and peer support groups – possibly run by the voluntary sector, with occasional professional support from the specialist team
- access to facilities such as hydrotherapy and adapted gym facilities, where they could continue their own self-exercise programmes.

The aim of this guidance is to support self-management by empowering patients to manage their own condition, but with the knowledge that help and advice is available when needed. Although formal health economic evaluation is currently lacking, experience suggests that providing a relatively low-cost lifeline does not produce dependence on or excessive use of specialist services but can be effective in avoiding more expensive crisis management.

The longer-term management of people with CRPS can be conceptualised around the framework provided by the National Service Framework (NSF) for long-term conditions (Appendix 13).

Recommendations for long-term care and support may be mapped broadly onto the NSF's quality requirements, including:

- person-centred integrated information and care planning (QR1)\*
- ongoing access to specialist care (QR2)
- community rehabilitation and support (QR5 and QR8)
- vocational support (QR6)
- support for families and carers (QR10).

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\* QR numbers refer to NSF mapping (Appendix 13).

## Recommendations

### Person-centred integrated information and care planning (QR1)

- ▶ People with CRPS should have access to appropriate information about their condition.
- ▶ Those who have complex long-term needs for care and support should have:
  - a named single point of contact for advice and information
  - integrated care planning, involving all of the agencies involved in their care, at a frequency agreed between the patient and their support team (usually no less than once a year).
- ▶ The named point of contact may change over time according to the individual's needs, but professionals taking on this role must have appropriate understanding of the long-term management of CRPS and should also have access to expert support and advice from specialist pain and rehabilitation services when this is required.

### Ongoing access to specialist care (QR2)

- ▶ People with CRPS who require continued contact with specialist pain or rehabilitation services should have access to these through an appropriate route, which may include telephone or email access to a named team member and/or access by self-referral, within one year of treatment completion, subject to funding agreements.
- ▶ At the treatment centre, provisions should be made for rapid processing of repeat GP referrals, eg by triaging upon receipt to physiotherapy/occupational therapy, a pain management programme and/or outpatient doctor appointment.

### Community rehabilitation and support (QR5 and QR8)

- ▶ People with CRPS should have access to a range of facilities to maintain their levels of activity and societal participation, which may include:
  - self-help and peer support groups<sup>102</sup>
  - facilities for self-directed exercise (eg adapted gym and swimming/hydrotherapy pool)
  - support for social and leisure activities
  - psychological intervention and counselling.
- ▶ These services are often appropriately run by voluntary organisations, with input from professionals as required.
- ▶ People with CRPS should have access to flexible support systems (eg personalised health and social care budgets) to maximise autonomy and choice of the most suitable forms of activities as their needs change over time.

### Vocational support (QR6)

- ▶ People with CRPS should have ongoing access to vocational support to help them remain in or return to work. Support may include:
  - various work support schemes provided through the Department of Work and Pensions (DWP) (eg the 'Access to work' scheme, which provides support for special equipment or the cost of getting to work for those unable to use public transport; see section on 'Rehabilitation medicine' for more details)
  - education for employers, occupational health doctors and advisers from the DWP about the specific requirements of people with CRPS.

### Support for families and carers (QR10)

- ▶ Families and carers of people with CRPS should have access to advice, support and information, including:
  - support to manage their own needs
  - support to maintain relationships.



# Appendices

## Appendix 1 Commercial sponsors

### **Astellas Pharma Ltd**

Lovett House  
Lovett Road  
Staines  
Middlesex TW18 3AZ

### **Grünenthal Ltd**

Aston Court  
Kingmead Business Park  
Frederick Place  
High Wycombe  
Bucks HP11 1LA

### **Medtronic Ltd**

Suite 1, Building 5  
Croxley Green Business Park  
Watford  
Herts WD18 8WW

### **Pfizer Ltd**

Walton Oaks  
Dorking Road, Walton-on-the-Hill  
Tadworth  
Surrey KT20 7NS

## Appendix 2 Systematic review methodology 2010

### Methodology

We reviewed randomised controlled trials (RCTs) published on the treatment and prevention of CRPS from July 2000 through to April 2010. A previous systematic review published in 2002,<sup>3</sup> which reviewed RCTs on the treatment and prevention of reflex sympathetic dystrophy (RSD) and CRPS from 1966 through to June 2000, formed the basis of the methodology for this review.

MEDLINE (PubMed), Scopus, Cumulative Index to Nursing and Allied Health Literature (CINAHL) and Allied and Complementary Medicine Database (AMED) bibliographic databases and the Cochrane Central Register of Controlled Trials were searched electronically using combinations of the following search terms: complex regional pain syndromes; causalgia; reflex sympathetic dystrophy with therapy; drug therapy; rehabilitation; randomised controlled trial; clinical trial; prevention and control. All foreign language papers were included.

Three reviewers filtered the resulting studies. Trials were included at this stage only if they were appropriately randomised,<sup>103</sup> ie if the randomisation was described and deemed appropriate. If the randomisation was described but deemed inappropriate, then the study was excluded. However, if the study was described as randomised but the method of randomisation was not described, these papers were allowed to remain in the review.

A study was regarded as relevant to this review if either pain intensity or prevention of CRPS was given as an outcome measure. Studies were excluded if they compared two active interventions and there was no significant difference in outcome between the two intervention groups with no control group. Paediatric studies were excluded and also studies with composite outcomes, eg 'CRPS score' compiled using various parameters, if pain intensity was not also given separately.

In a second step, the filtered studies were evaluated for their methodological quality using a 15-item checklist,<sup>104</sup> identical to that used in the 2002 Forouzanfar review. Six reviewers, in three groups of two each scored a third of the identified papers, such that each paper was scored by two people. Scores were then agreed between reviewers and any disagreement was settled by a third reviewer. A trial was considered to be of good quality if the methodological score was 50 or greater and of low quality if the score was less than 50.

Studies were considered to be positive if pain intensity was significantly reduced by the intervention described when compared with placebo or a control group. Studies were classed as negative if there was no difference in pain intensity after the intervention when compared to placebo. A similar classification was used for prevention studies.

Four levels of evidence of effectiveness were defined using the Van Tulder method,<sup>5</sup> based on the methodological quality and outcome of the studies as summarised in Table 4 (overleaf).

The results from the two systematic reviews are summarised in Appendix 14.

**Table 4 Levels of evidence of effectiveness of treatments for CRPS**

<b>Level of effectiveness</b>	<b>Evidence required</b>
Strong	Multiple good-quality RCTs
Moderate	One good-quality RCT and one or more low-quality RCTs
Limited	One good-quality RCT OR multiple low-quality RCTs
No evidence	One low-quality RCT OR no relevant RCTs OR contradictory outcomes

## Appendix 3 Information for GPs

### Complex regional pain syndrome (CRPS)

CRPS is a chronic condition characterised by limb pain, and dysfunction within the motor, sensory and autonomic nervous systems.

A CRPS limb has some of the following features:

- pain disproportionate to that expected after the relevant trauma
- abnormal swelling
- abnormal colour (may appear red, mottled or cyanosed, or all at different times)
- abnormal temperature
- abnormal sweating
- motor dysfunction
- abnormal skin or nail appearance.

Often patients describe the limb as feeling like it doesn't belong to them, and a hypothetical desire for amputation of the limb. The pain can be very severe. There is usually difficulty in moving the CRPS limb, which can be related to pain but also motor dyspraxia.

### Epidemiology and impact

CRPS is usually post-traumatic (eg following radial fracture), although 10% cases have no obvious causal event. CRPS is also usually, unilateral although in approximately 7% of cases there is later involvement of additional limbs. The incidence of CRPS is similar to that of multiple sclerosis, but 85% of CRPS cases improve or resolve within 18 months. Half of these cases continue with long-term functional problems and nearly half of patients do not return to work as a result of their chronic functional disability and residual pain.

### Aetiology

Exact mechanisms for the pathogenesis of CRPS are not understood. A combination of elements including inflammation, dysfunction within sympathetic and somatosensory nervous system, and cortical (not psychological) factors are thought to contribute to the generation and perpetuation of symptoms.

### Diagnosis

A combination of the presenting features will be seen. See Appendix 4 for a diagnostic checklist.

### Management in primary care

#### What to tell patients

It is important for the CRPS sufferer to understand the role of physiotherapy in rehabilitation. This may appear counter-intuitive, as even with gentle physiotherapy the pain may worsen, and reassurance may be necessary.

There is no cure for CRPS, but the majority of patients will get better. Reinforcing pain management principles, such as pacing, goal setting and relaxation, is helpful. A purely biomedical focus (cure seeking or solely reducing pain intensity) is unlikely to be of sufficient help and may impede progress. Improving disability and distress from CRPS is helpful in long-term recovery.

### **Crisis management**

Pain flares in CRPS are normal and should not be considered as a worsening of the condition. Usually a flare will settle over days or a few weeks. Continuing treatment, possibly with reducing the intensity of physical therapy (not the frequency) is important to maintain recovery and speed the resolution of flare. If the situation becomes difficult to control, it is important to involve a pain clinic in the first instance. This will avoid referral to other specialists for additional assessment, and prevent further escalation of suffering.

## Appendix 4 CRPS diagnostic checklist

A) The patient has continuing pain which is disproportionate to any inciting event		<input type="checkbox"/>	
B) The patient has at least one sign in two or more of the categories		<input type="checkbox"/>	
C) The patient reports at least one symptom in three or more of the categories		<input type="checkbox"/>	
D) No other diagnosis can better explain the signs and symptoms		<input type="checkbox"/>	
Category		Sign (you can see or feel a problem)	Symptom (the patient reports a problem)
1 'Sensory'	<i>Allodynia</i> (to light touch and/or temperature sensation and/or deep somatic pressure and/or joint movement) and/or <i>hyperalgesia</i> (to pinprick)	<input type="checkbox"/>	<i>Hyperesthesia</i> does also qualify as a symptom <input type="checkbox"/>
2 'Vasomotor'	Temperature asymmetry and/or skin colour changes and/or skin colour asymmetry	If you notice temperature asymmetry: must be >1°C <input type="checkbox"/>	<input type="checkbox"/>
3 'Sudomotor/oedema'	Oedema and/or sweating changes and/or sweating asymmetry	<input type="checkbox"/>	<input type="checkbox"/>
4 'Motor/trophic'	Decreased range of motion and/or motor dysfunction (weakness, tremor, dystonia) and/or trophic changes (hair/nail/skin)	<input type="checkbox"/>	<input type="checkbox"/>

Please note:

- Distinction between CRPS type 1 (no nerve injury) and CRPS type 2 (major nerve injury) is possible, but has no relevance for treatment. As an exception, in orthopaedic practice, in CRPS 2, a nerve lesion can sometimes be directly treated (see 'Surgical management' in 'Orthopaedic practice' section).
- If the patient has a lower number of signs or symptoms, or no signs, but signs and/or symptoms cannot be explained by another diagnosis, 'CRPS-NOS' (not otherwise specified), can be diagnosed. Includes patients who had documented CRPS signs/symptoms in the past.

If A, B, C and D above are all ticked, please diagnose CRPS. If in doubt, or for confirmation, please refer to your local specialist.

### Explanation of terms

'Hyperalgesia' is when a normally painful sensation (eg from a pinprick) is more painful than normal; 'allodynia' is when a normally not painful sensation (eg from touching the skin) is now painful; 'hyperesthesia' is when the skin is more sensitive to a sensation than normal.

### A special feature in CRPS

In category 4, the decreased range of motion/weakness is not due to pain. It is also not due to nerve damage or a joint or skin problem. This is a special feature in CRPS and is due to a poorly understood disturbed

communication between the brain and the limb. A helpful question to assess this feature is: 'If I had a magic wand to take your pain away, could you then move your ... (eg fingers)?'. Many patients will answer with 'no' to that question.

### Unusual CRPS

Around 10% of patients cannot recall a specific trauma or may report that their CRPS developed with an everyday activity such as walking or typewriting. In some people CRPS can have a bilateral onset. In about 7% CRPS can spread to involve other limbs. Around 15% of CRPS cases do not improve after 2 years. It is appropriate to make the diagnosis of CRPS in these unusual cases.

Note: psychological findings do not preclude the diagnosis of CRPS.



# Appendix 5 Desensitisation

## General patient information on therapy to help sensations to the skin feel more normal\*

This is a therapy known as desensitisation.

The goal of these activities is to make sensations to the skin of the body area affected by complex regional pain syndrome (CRPS) feel more normal. The aim is to re-educate the sensory system, part of which involves areas of the brain.

### General instructions

Many of these activities involve touch, and are suitable for the upper limbs (arms and hands) and lower limbs (legs and feet), although some are specific to one limb as indicated. These activities can be done on a daily basis and incorporated into your normal routine. Where possible, feel the sensation on a part of your body not affected by CRPS first, and remember how that normal sensation felt when then applying to the affected area.

### Regular practice: little and often

Regular practice of these activities will increase the benefit. A short period of desensitisation (even 1–2 minutes) as many times as possible throughout the day is recommended. It might be helpful to set aside particular times during the day to perform them. A quiet, relaxed environment with few distractions will help you to concentrate on the task.

As you progress you may find other activities within your daily routine in which to incorporate these principles.

### Discomfort

It is usual for these activities to be uncomfortable and somewhat painful whilst doing them and shortly afterwards. You may find that there are certain activities that you are unable to tolerate. Choose one that you feel comfortable with and gradually progress to others as you are able to do so. If you experience intolerable pain and discomfort, then stop that activity and find one that is more tolerable.

### Concentration is important

To help normalise the system, it is important that you concentrate on the quality of the sensation. This can be done by first undertaking the activity on a limb unaffected by CRPS. Concentrate on how this sensation feels, remember it and then undertake the activity on the affected area, whilst looking at it and thinking about it.

---

\* Adapted from a leaflet provided by experts at the Royal National Hospital for Rheumatic Diseases, with permission.

## Suggested activities

### 1) Activities of daily living

Desensitisation therapy can be incorporated into activities of daily living as part of your normal routine.

#### Whilst in bed

Feel the bed sheet against your unaffected limb. Close your eyes and concentrate on the quality of that sensation. Now feel the bed sheet against your affected area and recall how that normal sensation felt whilst thinking about the area you are touching.

#### Whilst dressing

Concentrate on your affected limb by looking at it and thinking about it as you get dressed. Feel the texture of the garment against your skin both on the unaffected and the affected areas.

#### Whilst having a bath or shower

Select a water temperature that you can tolerate. Feel the water on your unaffected body and now on your affected limb whilst looking at it and thinking about it. Recall how that normal sensation of the water felt against your skin.

Gently rub either a soft flannel, sponge or 'scrunchy' on the unaffected areas of your body. Use various movements such as circular actions, rubbing, patting and stroking. Concentrate on how these normal sensations felt whilst applying the movements to the affected areas of your body.

### Activities for the upper limb only:

#### Whilst washing up

Feel the temperature of the water on both the affected and unaffected areas of your hands. Think about how the normal sensation of the water feels whilst concentrating on your affected hand. Focus on the action of your affected hand in the water. Where tolerable, use different water temperatures such as tepid, hot and cold. Immerse your unaffected hand first, then your affected hand for short periods. These periods can be lengthened over time.

#### Whilst cooking

When you are making pastry or bread, mix it with both hands. Concentrate on the texture of the mixture and action as you are doing so.

### 2) Use of different textures

Applying different textures to the skin is another way to re-educate the sensory system.

Gather a variety of rough and smooth textures that you can tolerate. Here are some suggestions:

- Smooth – Felt, satin, silk, velvet, make-up or soft paint brushes
- Rough – towelling, netting, scourers, flannel, wool, hook velcro.

Place them on your unaffected limb and apply movements such as light stroking, firm stroking, tapping and circular actions. Note the various normal sensations that you feel. Now on the affected limb, apply the texture in similar movements working from an area that you can tolerate towards the more uncomfortable skin areas, for example from the top of the arm towards the hand. Concentrate on the area by looking at it and thinking about it. Recall the normal sensations that you felt on the unaffected limb.

### **3) Massage**

Massaging the affected limb can also be beneficial. This can be done by either yourself or someone else. Moisturisers or massage oils can be used. Be sure not to use anything which may irritate the skin. Use different pressures such as soft touch and firm massage where tolerable. Apply various movements such as patting, stroking and circular actions. Concentrate on the area being touched by looking at and thinking about it. Massage from your fingers and toes towards the centre of your body.

## Appendix 6 Atkins and Veldman diagnostic criteria for CRPS in an orthopaedic setting

### Veldman criteria<sup>11</sup>

- 1 The patient presents with four or five of the following symptoms:
  - unexplained diffuse pain
  - difference in skin colour relative to other limb
  - diffuse oedema
  - difference in skin temperature relative to other limb
  - limited active range of motion.
- 2 There is occurrence or increase of above signs and symptoms after use.
- 3 Above signs and symptoms present in an area larger than the area of primary injury or operation and including the area distal to the primary injury.

### Atkins criteria<sup>68,72,104</sup>

The diagnosis is made clinically by the finding of the following associated sets of abnormalities:

- 1 neuropathic pain; nondermatomal, without cause, burning, with associated allodynia and hyperpathia
- 2 vasomotor instability and abnormalities of sweating; warm red and dry, cool blue and clammy or an increase in temperature sensitivity; associated with an abnormal temperature difference between the limbs
- 3 swelling
- 4 loss of joint mobility with associated joint and soft-tissue contracture, including skin thinning and hair and nail dystrophy.

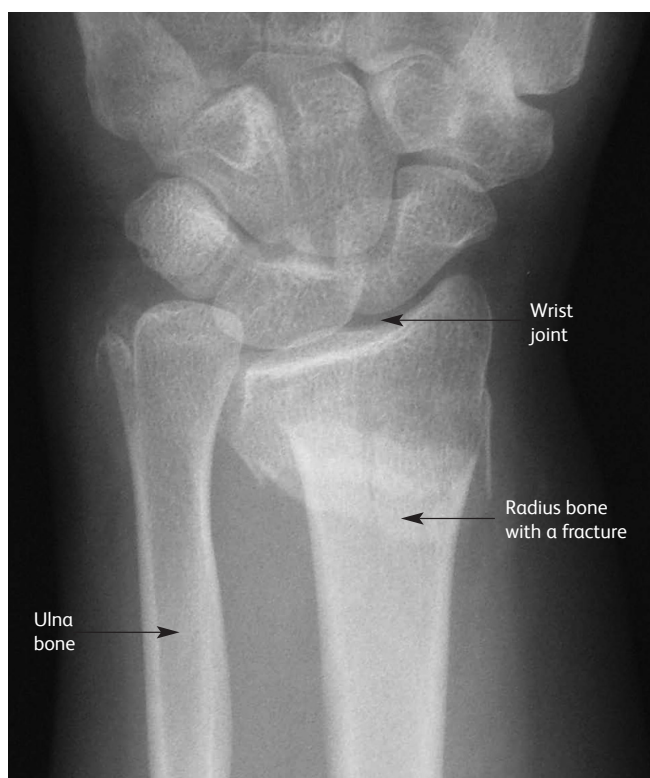
The diagnosis is excluded by the existence of conditions that would otherwise account for the degree of dysfunction.

## Appendix 7 Post-fracture/operation patient information leaflet

This sample leaflet has been kindly provided by professionals at the Royal Liverpool Hospital, Liverpool, and has been adapted for this guidance.

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### Looking after your fractured wrist



**Fig A1** X-ray of a radius fracture

A fracture is a break of the bone. Most wrist fractures are caused by a fall on to an outstretched hand, but a direct blow to the forearm can also cause a fracture. Following a fracture, the wrist is often immobilised in plaster cast usually for up to six weeks. This assists in stabilising the bones to help ensure they heal in a good position. In most simple fracture cases this is sufficient support. The plaster cast can also help to control your pain.

In some cases, an operation may be suggested to improve the position of the bones so that they heal in a more natural position and the soft tissues (muscles, tendons, ligaments and skin) are supported. Sometimes the support from a plaster is not enough to keep the bones in the best position. If this is the case you may have the option of treating this with an operation using pins and/or plates to hold the bone firmly while it heals. This option will be discussed with you in clinic if it is relevant to you. It is not always a clear case as to which option – an operation or the plaster – is the best treatment, and the pros and cons will be discussed with you on an individual basis to help you decide which way you would prefer to be treated.



Fig A2 Arm in plaster cast after radius fracture

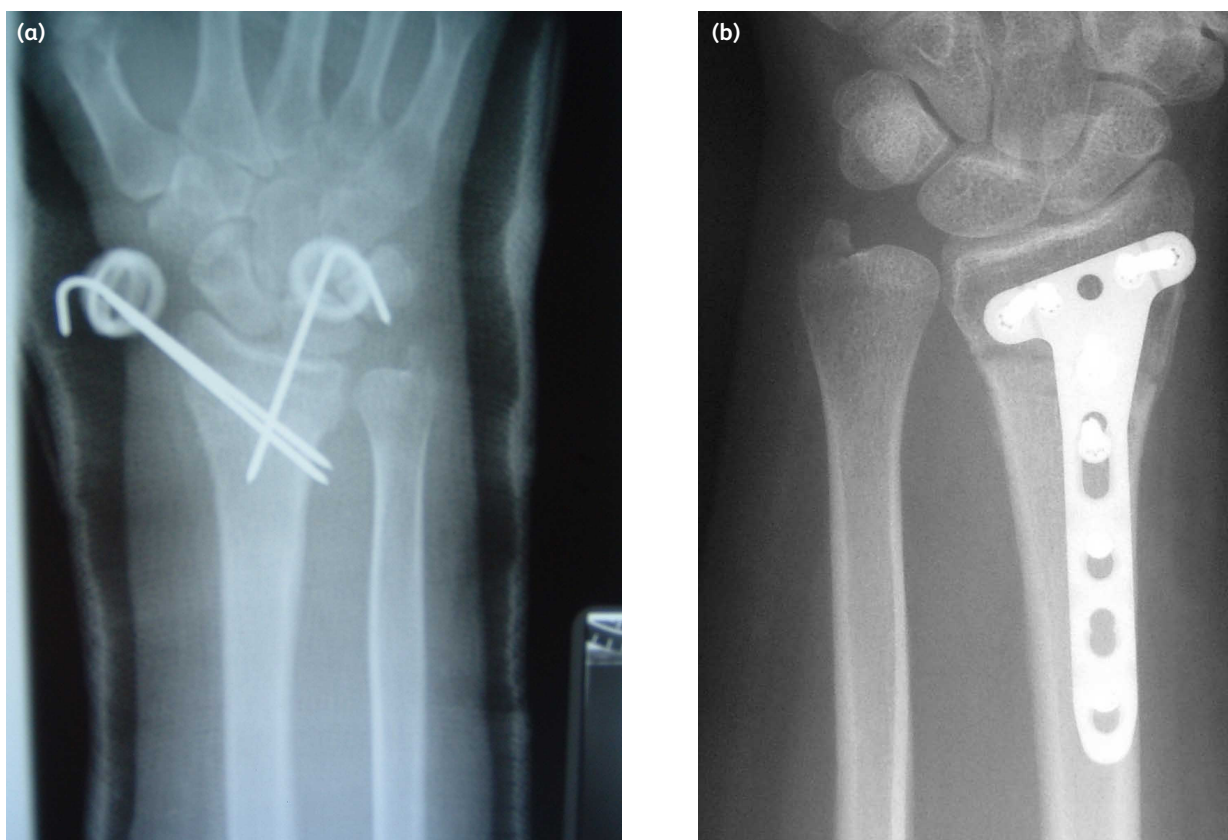


Fig A3 (a) Radius bone with a fracture treated with wires to hold the bones in position while they heal; (b) Radius bone with a fracture treated with screws and a plate to hold the bones in position while they heal

## What can I do now?

### Control your pain

It is important that your pain is minimal to allow the uninjured parts to be kept moving and allow you to sleep well. Ask in clinic or your family doctor (GP) for a prescription if necessary. Your pharmacist may also be able to advise you.

### Reduce swelling

Your hand and arm may swell in because of your injury. This swelling may also increase your pain as it puts increased pressure on the injured parts. If the swelling continues it can cause your joints to become stiff. Any

stiffness of the unaffected joints may delay your return to work or affect your ability to perform activities of daily living.

Swelling can be reduced by raising your arm. You can try the following activities:

- Keep your hand raised above the level of your heart as much as possible.
- If resting/watching television, rest arm out straight, raised on several pillows.
- Every 15 minutes in the hour, raise your hand right up above your head and 'pump' the fingers.



Fig A4 Recommended exercises as described in the text

### Keep fingers, thumb, elbow and shoulder moving

In order to keep your uninjured joints healthy it is important that they are kept moving. This will also encourage the blood supply to your soft tissues and reduce the swelling, as the muscle action helps to squeeze the extra fluid away from the injury. Studies have shown that keeping the uninjured parts moving helps to speed up your recovery once the plaster has been removed.

### Make sure your plaster fits comfortably

A well-fitting plaster will not stop you getting full finger movements, ie making a fist.

### Try to use your hand normally for all light activities (except in water)

For example, brushing hair, dressing, buttons, zips, feeding yourself; use your good hand to help if necessary. Try not to ignore your injured hand. This will help to prevent muscle weakness and abnormal pain responses.

### Eat healthily and avoid smoking

Try to eat a healthy and varied diet, as poor nutrition and smoking are known to slow healing.

### What should I do if I have a problem with my plaster?

- Any problems with your cast need to be reviewed by the medical team.
- Tightness or loosening of the cast may cause further complications. (The cast should not move against your skin but also should not feel tight or cause swelling of your fingers or thumb, or cause pressure on your skin).

If you have either of these problems, contact the plaster room Monday to Friday between 9am and 12.30pm or attend the Emergency Department as soon as possible between 9am and 4pm.

Return to the Emergency Department immediately if any of the following happens:

- increased swelling
- pins and needles/numbness
- inability to move fingers
- unusual colouring eg blue/purple
- increased pain.

A very small group of patients can develop complications after wrist fracture, including a condition called complex regional pain syndrome (CRPS), which requires treatment with early physiotherapy. By following the advice given above you help us to examine you for the possibility that you have developed a rare complication.

Any specific notes for you.



### Further information

Fracture clinic and plaster room

Tel:

Physiotherapy/Occupational Therapy Department

Tel:

Emergency Department

Tel:

A website you may find useful

[www.nhsdirect.nhs.uk](http://www.nhsdirect.nhs.uk)

## Appendix 8 Centres with a special interest in CRPS

### **University College London Hospitals**

Pain Clinic  
Queen Square  
London WC1N 3BG  
Tel: 0845 1555 000

Contact: Mr Paul Nandi or  
Mr Diarmuid Dennerly

### **Manchester Royal Infirmary**

(Central Manchester University Hospitals NHS Trust)  
Rheumatology Department  
Oxford Road  
Manchester M13 9WL  
Tel: 0161 276 1234

Contact: Dr Rachel Gorodkin

### **Hope Hospital**

Anaesthetic Department  
Stott Lane  
Salford  
Manchester M6 8HD  
Tel: 0161 789 7373

Contact: Mrs Alison Dwyer

### **The Walton Centre for Neurology and Neurosurgery NHS Trust**

Lower Lane  
Fazakerley  
Liverpool L9 7LJ  
Tel: 0151 525 3611

Contact: Dr Andreas Goebel

### **St Thomas' Hospital**

Pain Clinic  
Lambeth Palace Road  
London SE1 7EH  
Tel: 020 7188 7188

Contact: Dr Douglas Justins

### **The Royal National Hospital for Rheumatic Diseases NHS Foundation Trust**

Bath Centre for Pain Services  
Upper Borough Walls  
Bath BA1 1RL  
Telephone: 01225 465941

Contact: Professor Candy McCabe

### **Addenbrooke's Hospital**

Cambridge University Hospitals NHS Foundation Trust  
Hills Road  
Cambridge CB2 0QQ  
Tel: 01223 245 151

Contact: Dr Nick Shenker

**New Stobhill Hospital**

Stobhill ACH Chronic Pain Service  
133 Balornock Road  
Town Centre  
Glasgow G21 3UW  
Tel: 0141 355 1490

Contact: Dr Mick Serpell

**West Suffolk NHS Foundation Trust**

Department of Pain Management  
Hardwick Lane  
Bury St Edmunds  
Suffolk IP33 2QZ  
Tel: 01284 713330

Contact: Dr Rajesh Munglani

**Royal Devon and Exeter Hospital NHS Foundation Trust**

Rheumatology Department  
Barrack Road  
Exeter EX2 5DW  
Tel: 01392 411611

Contact: Dr Richard Haigh

**Derriford Hospital**

Derriford Road  
Crownhill  
Plymouth  
Devon PL6 8DH  
Tel: 0845 155 8155 / 01752 202082

Contact: Dr Mark Rockett

**University Hospital of Wales**

Heath Park  
Cardiff CF14 4XW  
Tel: 029 2074 7747

Contact: Mr Brian Simpson

**Royal National Othopaedic Hospital (Stanmore)**

Rheumatology  
Brockley Hill  
Stanmore HA7 4LP  
Tel: 020 8954 2300

Contact: Dr Helen Cohen

Please note that this list is not exhaustive. There are likely to be additional centres and clinicians with an interest and appropriate expertise in the treatment of CRPS.

## Appendix 9 Recommendations for the treatment of skin ulcers, skin infection and problematic oedema

Changes in skin innervation, blood flow, interstitial fluid (oedema), the trophic constitution of the skin and skin temperature, in the peripheries increase the risk of skin ulceration.<sup>81,106</sup> Some of these changes are often present in complex regional pain syndrome (CRPS). When ulceration occurs, this allows the entry and multiplication of microorganisms, so that patients are at risk of developing cellulitis and deeper tissue infections.

### Assessment

In a patient with CRPS and skin ulceration in the affected limb, non-invasive doppler studies should be used to exclude peripheral ischaemia.<sup>106</sup> For the lower limb, assessment of the ankle/brachial pressure index (ABPI) is essential to identify any ischaemic element, and should be carried out by someone trained in this technique, usually a nurse in tissue viability. Application of compression without taking into account the ABPI can result in gangrene.

Because all skin ulcers harbour skin microorganisms, swab cultures taken from patients with skin ulceration are usually positive. Positive swab cultures should not be treated unless there are signs of clinical infection.<sup>107,108</sup> Indication of infection includes systemic symptoms (eg fever and leucocytosis) or local signs such as spreading redness, warmth, induration, pain or tenderness. Erythema may be well demarcated or more diffuse. In severe cases, blistering/bullae, superficial haemorrhage into blisters, dermal necrosis, lymphangitis and lymphadenopathy may occur.<sup>107,108</sup> Deep infection (eg necrotising fasciitis or osteomyelitis) has the risk of threatening a limb, and if suspected should be treated aggressively (see under 'Management').

There is often a need to exclude underlying osteomyelitis, which may be suggested by bone destruction or periosteal reaction on plain X-rays, or if probing the wound using a blunt, sterile, stainless-steel probe one encounters bone,<sup>109</sup> but magnetic resonance imaging (MRI) is considered the imaging test of choice when osteomyelitis is suspected.<sup>110</sup> If osteomyelitis is suspected, the early intervention of an orthopaedic surgeon is essential.

### Management

General measures such as adequate diet, ensuring adequate haemoglobin level, diabetic control and cessation of smoking should be emphasised where appropriate.<sup>111</sup>

The management of skin ulceration in CRPS follows general principles established for the management of diabetic foot ulcers. Removal of necrotic tissue, callus, infected or foreign material should be achieved by sharp debridement.<sup>112</sup> For deep or sloughy ulceration, weekly sharp debridement should be considered.<sup>112</sup> Pressure should be relieved using felted foam dressings and low-pressure garments (eg Alcast Walkers boots®, casts, or open shoes).

If infection is diagnosed on clinical grounds, then the choice of antibiotic should be based on the pathogens isolated from swabs, and if possible, tissue culture. The commonly useful broad-spectrum antibiotics are flucloxacillin in mild cases, with clindamycin, cephalexin, ciprofloxacin and amoxicillin-clavulanic acid

(Augmentin) useful in more severe infection.<sup>85</sup> Soft-tissue infections require 10 days' therapy, while osteomyelitis may require more than six weeks of therapy.<sup>106</sup> Antimicrobial therapy in patients who do not improve can be guided by both skin biopsy,<sup>113</sup> which is more reliable than superficial swabs, and early advice from a bacteriologist/microbiologist.

In patients who have had at least two episodes of infection at the same site, prophylaxis with low-dose penicillin V or erythromycin (both typically 250mg bd) for a year should be considered.<sup>114,115</sup>

Dressings that promote a moist wound environment should be the focus of care of chronic wounds.<sup>115</sup> Typically such dressings may include hydrocolloid dressings, or for wounds producing exudate, silver or iodine impregnated dressings, especially when infection is present.<sup>116</sup> Rarely platelet-derived growth factor<sup>117</sup> (Regranex®, Becaplerin gel®) or allogeneic cultured dermis (Dermograft®, Apligraf®) can be used in the wound dressing. While these have been shown in randomised controlled trials to promote wound healing in clean wounds, they are relatively expensive.

Where oedema is present in a patient with skin ulcerations, after full vascular investigation and since the oedema present in CRPS can foster both poor nutrition with consequent ulceration and superinfections when infection has been treated or excluded, appropriate compression bandaging should be used to disperse tissue fluid. The inclusion of the tissue viability and/or lymphoedema teams is crucial. Compression is usually achieved using wool (to even pressure and absorb exudate) and compression bandaging (eg Profore lite®, Profore®, Elset®) in spiral or figure of eight configuration, or graded compression hosiery, depending on vascular status. Other treatment for lymphoedema includes the use of intermittent pump compression (eg Flowpac® pump). Dermatologists should be aware that treatment with spinal cord stimulation (SCS) by pain specialists or neurosurgeons can reduce limb swelling in CRPS.<sup>97</sup>

# Appendix 10 Patient information

## Information leaflet for patients: complex regional pain syndrome

### What is complex regional pain syndrome (CRPS)?

CRPS pain usually develops in an arm or leg after an injury. Only rarely are other areas affected. It can affect people of all ages, including children. There are two types of CRPS:

- CRPS type 1 follows an injury to a limb, such as a broken bone or even a minor sprain.
- CRPS type 2 follows partial damage to a nerve in the limb. The symptoms are very similar. This form is very rare.

Other names: complex regional pain syndrome type 1 (CRPS 1) was known as 'reflex sympathetic dystrophy (RSD)' or 'Sudeck's syndrome', and complex regional pain syndrome type 2 (CRPS 2) was known as 'causalgia'.

### What is it like to have CRPS?

CRPS pain continues after the original injury has healed. It is often severe.

The main symptom is pain in the arm or leg. The pain is often burning, sharp, stabbing or stinging, with tingling and numbness. There are a range of other symptoms which can change over time. The skin may become oversensitive to light touch. Clothes brushing the skin or even air blowing on the skin may be felt as severe pain. This unusual sensitivity is called 'allodynia' and is common in CRPS.

Other symptoms include skin colour change, swelling, stiffness, feelings of hot or cold, less or more sweating and changes to the hair, skin or nails. The pain and other symptoms often spread beyond the site of the original injury. For example, if you hurt a finger, the whole of the hand or forearm can be affected.

Often there is difficulty in moving the limb, together with weakness and sometimes shaking or jerking. Sometimes the muscles in the area can waste and the hand or foot can become twisted.

Many patients say that their limb 'feels strange'. It can feel as if it does not belong to the rest of the body and as if it is not your own limb. Sometimes the limb feels bigger or smaller than the opposite, normal limb.

Some patients have frequent thoughts about wishing to cut off the limb. Unfortunately even surgical amputation does not help the pain (actually, it may make it worse). In extreme pain, some people may consider suicide. If you do feel like this, please see your doctor.

### What causes CRPS?

CRPS is a stronger-than-normal reaction of the body to injury. We don't know what causes CRPS. What we do know is that the abnormal reaction to injury happens both in the affected limb and in the brain. The nerves in the affected limb are much more sensitive than other nerves and this causes some of the tenderness to touch and pressure. The brain is also involved. The way the brain communicates with the affected limb often changes and this can cause some of the problems with movement.

CRPS is not in your mind. We also know that your mindset cannot cause CRPS, but that some psychological factors such as fear or worry can make the pain worse than it already is.

### Does CRPS run in families?

It may be that genes have something to do with who develops CRPS pain after injury, but they are certainly not the only factor in deciding who gets it. It is also very unlikely that anyone else in your family will ever develop CRPS pain.

### Could it have been prevented?

It is very unlikely that CRPS pain after your injury could have been prevented. The right diagnosis and treatment can reduce suffering from CRPS pain.

### Will it get better?

CRPS usually gets better by itself or with treatment. In some people, CRPS does not get better. We have no way of predicting whether your CRPS will get better and when. Unlike cancer or rheumatoid arthritis, CRPS does not destroy body tissues. Even if you have CRPS for several years, the rest of your body will continue to work as normal.

### Does treatment help?

Treatment aims to improve your quality of life, functioning and reduce pain. It is likely that you can get some pain relief with treatment. The success of some treatments depends on the amount of effort you put into them. There is a range of treatments and your consultant or therapist will discuss these with you.

#### Exercise treatment

Most patients see physiotherapists (PTs) or occupational therapists (OTs). These therapists will work with you in a way which is specially geared towards your CRPS. For example, they may not even touch your limb. It is very important to exercise the limb gently following advice by a PT or OT.

#### Medication treatment

Drugs can sometimes reduce CRPS pain and may also help you to sleep. Your consultant will discuss the correct drug treatment with you. If appropriate, your consultant may also decide to offer you an injection treatment. In this case, you would receive more special information about that.

#### Psychological intervention

Sometimes psychological intervention can be helpful to reduce distress (this does not mean that the pain is in your mind; it is not). Your consultant would be happy to discuss this with you.

## Information leaflet for patients: specialised treatments for CRPS

### What are specialised treatments for CRPS?

Specialised treatments either require a special team of healthcare professionals to deliver them, or these are new treatments, which need to be followed closely to make sure they work.

### Should I be treated with a specialised treatment for CRPS?

The right treatment for CRPS varies from patient to patient. There are two specialised CRPS treatments which need to be given by teams of clinicians. These are: pain management programme (PMP) and spinal cord stimulation (SCS). Research shows that in some patients these treatments can work very well. Your consultant will discuss these treatments with you, if he or she thinks you may need either of them. You may also receive a PMP and/or SCS information leaflet.

The PMP is a programme designed to help you to improve your quality of life and manage your pain better. It is group-based, and lasts between a few days and a few weeks. This is 'multidisciplinary treatment', which means therapists from different professions work together (eg physiotherapists, doctors, occupational therapists and psychologists etc). The PMP is suitable for patients with CRPS, and also for people with other chronic pains. It is designed to improve your quality of life. It is important to understand it is not designed to take your pain away.

The second treatment, SCS, is a fine wire which is placed close to the nerves in your back and connected to a 'stimulator'. The doctor puts the wire in the right place by using a similar technique to putting in an epidural for pain relief during pregnancy. The wire is usually kept in place like this for a short time, and if it works well, an operation is done to make it permanent. The SCS can be taken out in the future when it is not needed anymore.

### Are there any other treatments?

There may be other treatments, but these are not as well researched as the treatments mentioned in your patient information leaflets. Your consultant will discuss with you whether or not other treatments would be suitable in your case.

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## Sources of information for patients on the web

### Arthritis Research UK

[www.arthritisresearchuk.org/arthritis\\_information/arthritis\\_types\\_and\\_symptoms/complex\\_regional\\_pain\\_syndrome.aspx](http://www.arthritisresearchuk.org/arthritis_information/arthritis_types_and_symptoms/complex_regional_pain_syndrome.aspx)

### Reflex Sympathetic Dystrophy Association (RSDSA)

<http://rds.org/index2.html>

### CRPSUK

[www.crpsuk.org](http://www.crpsuk.org)



The following information has been compiled and produced by the CRPS Patients Forum at the Royal National Hospital for Rheumatic Diseases (RNHRD) after adaptation from a version produced by the Reflex Sympathetic Dystrophy Syndrome Association in America.

This following information may be useful if you need to explain your condition to your doctor:

### **I have CRPS (complex regional pain syndrome)**

CRPS is a nerve disorder that usually occurs after an injury or period of immobilisation. The principle symptom is pain which can lead to disability.

I may look healthy but I often suffer from severe, unrelenting, nerve pain. My skin may swell, change colour or temperature, sweat or hurt to the lightest touch.

Often it is difficult for me to sleep, which affects my attention and concentration, or I may be on drugs which do the same.

Chronic pain often leads to depression. Stress increases pain. I have good or bad days, or even hours.

There is no cure at present.

### **Please help me by...**

- believing that the pain is real even though it is invisible and may not be readily apparent by my demeanor or activities
- remembering that it can even hurt to be touched
- remaining positive.

Please email [crpsuk@hotmail.com](mailto:crpsuk@hotmail.com) if you require more information about the CRPS Patients Forum. Please also email any suggestions you may have to the same address.

# Appendix 11 Assessment of CRPS by occupational therapists and physiotherapists

## Assessment tools

There is no standardised battery of assessments for CRPS. It is not within the scope of these guidelines to be prescriptive about these measures, yet validated measures should be used as far as possible.

## What to assess

Assessments should be such that they adequately measure change in signs and symptoms, along with the functional consequences of these. Assessments should therefore cover the following areas:

- pain
- sensation
- swelling
- movement (also consider presence of dystonia)
- function

The functional impact of CRPS can be assessed objectively and subjectively and the assessment should be directly related to patient goals. Recognising patients' criteria for success enables the therapist to individualise treatment.<sup>48</sup> Patients who are autonomous in their goal setting perceive their goals to be more relevant, and express greater satisfaction with the rehabilitation process.<sup>47</sup>

- body perception disturbance (BPD)<sup>118</sup>

Body perception disturbances can be elicited through targeted questions about emotional feelings towards, and the extent of self-ownership of the affected limb. The degree of BPD can be assessed and reviewed using the Bath CRPS BPD scale.<sup>61</sup>

Additional signs/symptoms that may be monitored include:

- skin temperature/colour
- hair/nail growth

## When to assess

Assessment of change during therapy should be ongoing. Outcome measures should be completed at baseline and at the end of treatment. Interim use of outcome measures can be carried out according to the therapists' judgement.

## Appendix 12 Key contents of an interdisciplinary specialist rehabilitation programme

### Engagement: Education and information for the patient and his/her family

- Active engagement of the patient/family in goal setting, goal review
- Control – the patient remains responsible for their own rate of progress
- Understanding and insight:
  - how emotional stress, muscle tension and de-conditioning can increase pain experience
  - how their own behaviours may serve to exacerbate pain experience
- Learning:
  - self-management approach, including goal setting and pacing
  - the right balance between doing too much and too little
  - relaxation techniques, breathing exercises etc to reverse sympathetic arousal
- Empowering the family:
  - encouraging individual to keep active and to do more for themselves

### Medical management

- Investigation and confirmation of diagnosis
- Pharmacological intervention (in conjunction with pain team wherever appropriate) to provide a window of pain relief
- Reassurance that physical and occupational therapy are safe and appropriate
- Provide medical follow-up to prevent iatrogenic damage through inappropriate referral
- Support any litigation/compensation claim to its resolution and conclusion

### Psychosocial and behavioural management

- Identify any psychological factors contributing to pain and disability
- Treat anxiety and depression
- Identify, explore and proactively address any internal factors (eg counter-productive behaviour patterns) or external influences (eg perverse incentives, family dynamics etc) which may perpetuate disability/dependency
- Consider needs of family/carers – provide psychological intervention/counselling where appropriate
- Provide a practical problem-solving, goal-orientated approach (involving both the patient and their family) to reduce barriers and promote healthy functioning

### Physical management

- Retrain normal body posture
- Desensitisation – handling the affected part following by passive stretching/isometric exercise
- Progression to active isotonic exercise and then strength training
- General body re-conditioning – cardiovascular fitness
- Encourage recreational physical exercise and functional goals
- Techniques to address altered perception and awareness of the limb, eg mirror visual feedback training<sup>50</sup> or graded motor imagery\*

*continued*

\* Note for pain relief, the evidence for the efficacy of GMI is conflicting (Johnson *et al. Eu J Pain*, 2012, in press),

### Activities of daily living and societal participation

- Support graded return to independence in activities of daily living with clear functional goals
- Assessment and provision of appropriate specialist equipment/adaptations to support independence<sup>119</sup>
- Removal of inappropriate/unnecessary equipment
- Adaptation of environment
- Extend social and recreational activities in and outside the home
- Workplace assessment/vocational re-training

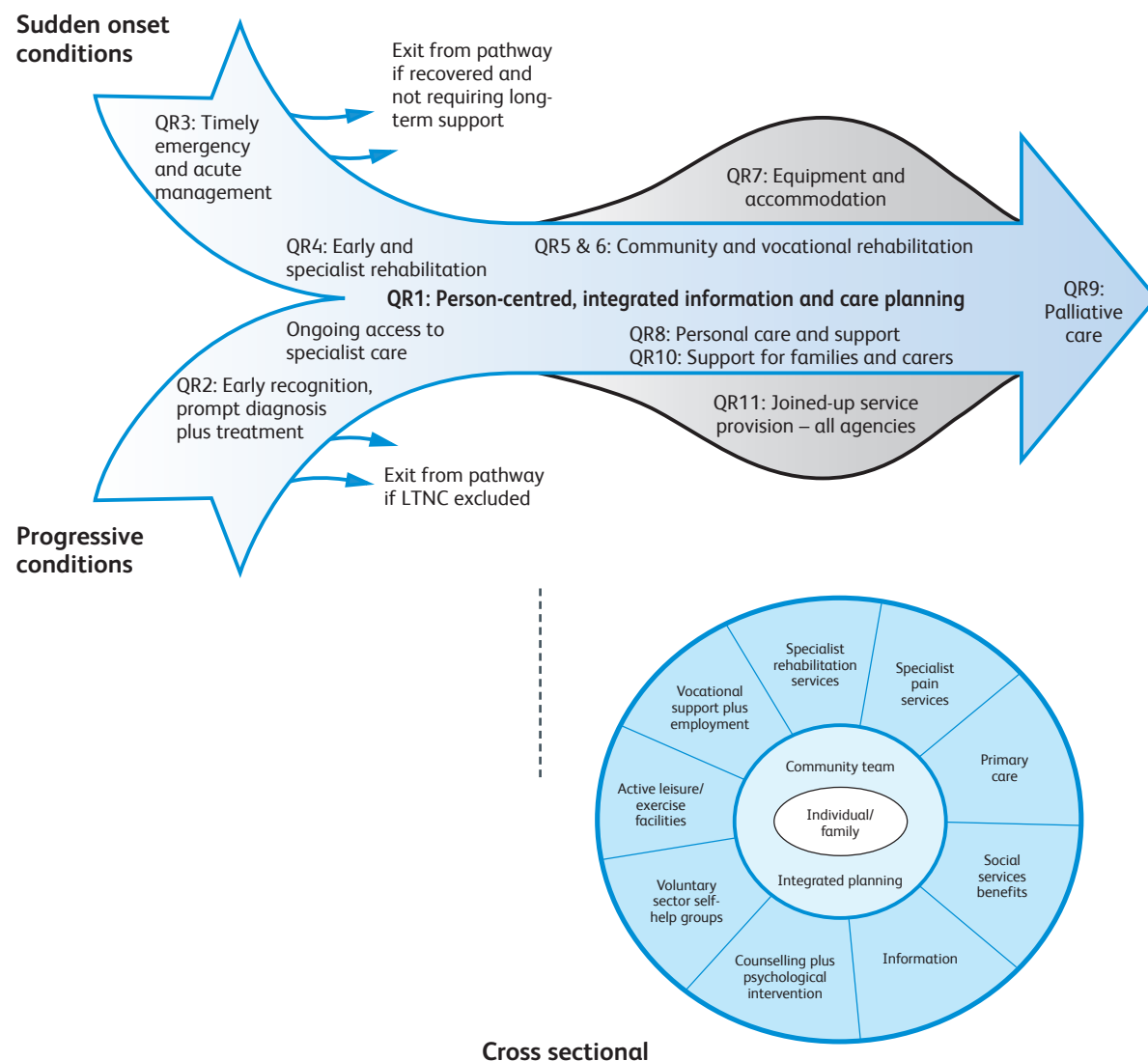
Adapted from the BSRM report on musculoskeletal rehabilitation<sup>100</sup>

# Appendix 13 The National Service Framework for long-term conditions

The National Service Framework (NSF) for long-term conditions (2005) provides a useful framework on which to underpin the longer-term management of patients and their families with long-term CRPS.<sup>120</sup> Although originally developed around six exemplar neurological conditions, the NSF standards (or ‘quality requirements’) were designed to be applicable across the broader spectrum of long-term conditions.

The NSF emphasises the need for life-long care and integrated service provision. It therefore covers care at all stages along the pathway from diagnosis throughout an individual’s life span, and encompasses both health and social care needs across primary secondary and tertiary services. It also emphasises the need to provide flexible support that maximises personal choice and empowers patients to manage their own condition. Patients with CRPS form a diverse group and their needs will vary over time. The fish diagram (Fig A5) illustrates the 11 quality requirements (QRs) of the NSF, and the type of services that may need to be coordinated during integrated care planning for long-term care in CRPS patients.

Fig A5 The ‘fish’ diagram



## Appendix 14 Summary of results from systematic reviews of RCTs for treatment of pain in CRPS with a summary of NICE guidelines and IASP recommendations for neuropathic pain

<p><b>Drugs for pain relief</b> <i>Evidence of efficacy in CRPS</i></p> <p><b>Strong</b> Bisphosphonates: L<sup>†</sup> IV Pamidronate (60mg iv single dose) E<sup>†NA</sup> IV Alendronate (7.5mg OD iv × 3days) E<sup>†</sup> IV Clodronate (300mg OD iv × 10 days) E<sup>†NA</sup> Oral alendronate (40mg OD po × 8 weeks)</p> <p><b>Moderate:</b> LL<sup>††</sup> IV Ketamine (low-dose, 2 trials, 10 days outpatient or 4.5 day cont)</p> <p><b>Limited:</b> L<sup>††</sup> P<sup>c</sup> Tadalafil (20mg OD po × 12 weeks) L<sup>†</sup> IV Immunoglobulin (0.5g/kg) U<sup>†NA</sup> Bretylium (1.5mg/kg) IVRSB with lidocaine (0.5%) L<sup>†</sup> Epidural clonidine</p> <p><b>Classed as no evidence:</b> E<sup>↓</sup> Oral prednisolone (10mg TDS po up to 12 weeks) L<sup>↓x</sup> Botulinum toxin (75U) + LSB with bupivacaine (10ml 0.5%)</p>	<p><b>Procedures for pain relief</b> <i>Evidence of efficacy in CRPS</i></p> <p><b>Limited:</b> L<sup>†</sup> Spinal cord stimulation L<sup>†</sup> Transcranial magnetic stimulation</p> <p><b>Psychological treatments</b> No trials conducted in CRPS <i>Treatments with efficacy in chronic pain</i> Cognitive behavioural therapy</p>	<p><b>Rehabilitation/Physiotherapy</b> <i>Evidence of efficacy in CRPS</i></p> <p><b>Strong</b> LL<sup>††</sup> Graded motor imagery (2 trials)</p> <p><b>Moderate</b> L<sup>↓</sup>E<sup>†ps</sup> Mirror therapy (2 trials)</p> <p><b>Limited</b> E<sup>†</sup> 'Traditional' physiotherapy* and 'Traditional' occupational therapy U<sup>†</sup> Qigong</p>	<p><b>Evidence of non-efficacy or where the evidence is conflicting in CRPS</b></p> <p><b>Strong:</b> UELL<sup>††††</sup> Guanethidine IVRSB (4 negative trials)</p> <p><b>Moderate:</b> EE<sup>↓</sup> Manual lymphatic drainage (2 negative trials)</p> <p><b>Limited:</b> L<sup>d</sup> Gabapentin (1800mg) L<sup>R</sup> Parecoxib (5mg) IVRSB with clonidine 30ug, lidocaine 1g/kg LL<sup>††NA</sup> Reserpine IVRSB (2 negative trials) – same trials as 2 of the guanethidine trials L<sup>†</sup> IVRSB droperidol with heparin U<sup>†</sup> IV phenolamine (0.5g/kg) E<sup>†</sup> IV mannitol L<sup>†</sup> IT glycine E<sup>†</sup> 50% DMSO cream L<sup>††</sup> P<sup>c</sup> Topical transdermal isosorbide dinitrate L<sup>†</sup> Occlusal splints E<sup>†</sup> Electromagnetic field therapy EEE<sup>↓↓↓</sup> Acupuncture (3 trials)</p> <p><b>Classed as no evidence:</b> L<sup>↓R</sup> Iohexol before LSB with lidocaine E<sup>↓</sup> Methylprednisolone with lidocaine IVRSB</p> <p><b>Conflicting</b> UL<sup>NA</sup> Ketanserin: IV(10mg) 1 trial, IVRSB 1 trial EE<sup>††pf</sup> Intranasal calcitonin (2 trials) L<sup>†</sup> Lidocaine sympathetic ganglion block</p>
<p><b>Drugs with efficacy in neuropathic pain</b> according to the NICE Clinical Guideline<sup>31</sup> for neuropathic pain and the recommendations of the IASP Neuropathic Pain Special Interest Group<sup>88,89</sup></p> <p><b>Neuropathic pain first line treatments:</b> TCAs: amitriptyline, nortriptyline, imipramine gabapentin (see column 4), pregabalin SSNRIs: duloxetine; venlafaxine Topical lidocaine</p> <p><b>Neuropathic pain second line treatments:</b> Morphine, oxycodone, tramadol, methodone,<sup>††</sup> NA Levorphanol <sup>†</sup>Not originally developed for CRPS. <sup>††</sup>Methodone should be used only by experienced specialists.</p> <p>Third line treatments for neuropathic pain, and NSAIDs and paracetamol, are not listed as there is substantially less evidence.</p>			

† High-quality trial (methodology score >=50) ↓ Low-quality trial (methodology score <50). The methodological score for a trial is in no way linked to the level of evidence for the efficacy of a treatment. (The scoring methodology and levels of evidence are detailed in appendix 2.)

L<sup>†</sup> Treatments with efficacy in longstanding CRPS (>6mths cut off or >14mths median duration or >19mths mean duration);

E<sup>†</sup> Early CRPS. U<sup>†</sup> Disease duration unknown. NA Not available in some countries (UK) or at doses used in this trial.

R<sup>†</sup> Quite separately from the methodology score, the panel had some reservations about the protocol used in the study (described in detail in Cossins *et al.*<sup>22</sup> In review) † Primary outcome – temperature change in the limb. Secondary outcome – VAS for pain.

P<sup>s</sup> Evidence for both trials is from a subgroup with CRPS post stroke. P<sup>f</sup> 2 weeks post fracture – an unusually early patient group – no CRPS diagnosis. P<sup>c</sup> subgroup with cold CRPS. <sup>d</sup> This trial was for lower dose gabapentin than recommended in commonly accepted NP guidelines.<sup>27</sup> x Primary outcome was whether the effect of sympathetic block was prolonged

\*Includes information, practical advice and support; relaxation exercises; rest; connective tissue massage; transcutaneous electrical nerve stimulation; exercises to reduce pain; compensatory activities; instructions about body position

Abbreviations: LSB – lumbar sympathetic blockade, IT – intrathecal, IV – intraVenous, IVRSB – intraVenous regional sympathetic blockade, NP – neuropathic pain, TCA – tricyclic antidepressants, NSAIDs – non steroidal anti-inflammatory drugs, SSNRI – selective serotonin noradrenaline reuptake inhibitors, VAS – visual analogue scale, DMSO – dimethyl sulphoxide

Review methodology details are given in Appendix 2. There is no evidence for the efficacy of paracetamol or non-steroidals in CRPS

NB. Some previously published research from SS Reuben has been withdrawn and no publications from this author have been used as evidence in the development of this guidance.

## Appendix 15 Experimental treatments for CRPS – published research

Treatments for CRPS based on publications from June 2000 until April 2010 of trials which were not RCTs. These treatments cannot currently be recommended because although there may be some evidence, there is not sufficient evidence available to support their efficacy.

### Brachial plexus analgesia

- 1 Azad SC, Beyer A, Romer AW *et al.* Continuous axillary brachial plexus analgesia with low dose morphine in patients with complex regional pain syndromes. *Eur J Anaesthesiol* 2000;17(3):185–8.
- 2 Day M, Pasupuleti R, and Jacobs S. Infraclavicular brachial plexus block and infusion for treatment of long-standing complex regional syndrome type 1: A case report. *Pain Physician* 2004;7(2):265–8.

### Surgical sympathectomy

- 3 Bandyk DF, Johnson BL, Kirkpatrick AF *et al.* Surgical sympathectomy for reflex sympathetic dystrophy syndromes. *J Vasc Surg* 2002;35(2):269–77.
- 4 Bosco Vieira DJ, Kux P, Duarte DF. Endoscopic thoracic sympathectomy for the treatment of complex regional pain syndrome. *Clin Auton Res* 2003;13 (Suppl 1):I58–I62.
- 5 Golubev VG, Krupatkin AI, Zeinalov VT, Merkulov MV, Kuz'michev VA. New facilities in management of complex regional pain upper limb syndrome with thoracoscopic sympathectomy. *Vestn Ross Akad Med Nauk* 2008;(8):52–5.
- 6 Kargar S, Parizi FS. Thoracoscopic sympathectomy in causalgia. *Ann Chir Gynaecol* 2001;90(3):193–4.

### Anaesthetic blockade with specific agents

- 7 Chen LC, Wong CS, Huh BK *et al.* Repeated lumbar sympathetic blockade with lidocaine and clonidine attenuates pain in complex regional pain syndrome type 1 patients—a report of two cases. *Acta Anaesthesiol Taiwan* 2006;44(2):113–7.
- 8 Hord ED, Stojanovic MP, Vallejo R *et al.* Multiple Bier blocks with labetalol for complex regional pain syndrome refractory to other treatments. *J Pain Symptom Manage* 2003;25(4):299–302.

### Local anaesthetic infusion with physiotherapy

- 9 Mak PH, Irwin MG, Tsui SL. Functional improvement after physiotherapy with a continuous infusion of local anaesthetics in patients with complex regional pain syndrome. *Acta Anaesthesiol Scand* 2003;47(1):94–7.

### Electroconvulsive therapy (ECT)

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**Lycra pressure garments ( eg ‘second skin’ devices)**

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## Appendix 16 Systematic review update – RCTs published from April 2010 – December 2011

Further to the previous review period, we reviewed randomised controlled trials published on the treatment of CRPS from April 2010 to December 2011.

Medline (PubMed), SCOPUS, CINAHL, and AMED bibliographic databases and the Cochrane Central Register of Controlled Trials were searched electronically using combinations of the following search terms: complex regional pain syndromes; with therapy; drug therapy; rehabilitation; randomised controlled trial; clinical trial. All foreign language papers were included.

One reviewer filtered the resulting 16 studies. Four studies were found to be appropriately randomised and thus further assessed by another reviewer (see Appendix 2 for full description of methodology).

The filtered studies were not scored for their methodological quality as in the main review but were assessed as follows:

	<b>van der Plas 2011</b> <sup>121</sup>
Randomisation	suitable, appropriate
Outcome measure	NRS pain, NRS dystonia
Early or late	late (Median 12.5 years)
Diagnosis	IASP criteria
Intervention	intrathecal baclofen for dystonia
Design	active v active in two concentrations/infusion rates double-blind cross-over
Outcome	increased infusion rate of more dilute solution does not improve control of dystonia
Population	14 patients already receiving ITB for dystonia with unsatisfactory response
	<b>Eckmann 2011</b> <sup>122</sup>
Randomisation	suitable, appropriate
Outcome measure	NRS pain, short-term pain NRS, limb volume difference, joint pain score, ROM
Early or Late	1 to 29 months
Diagnosis	IASP, lower limb
Intervention	intravenous regional block with lignocaine (50mls 0.5 %) and ketorolac (0,30,60,120mg)
Design	active v active – lignocaine plus various ketorolac dose double-blind cross-over
Outcome	negative (only 1 day of pain relief after ketorolac)
Population	10 patients with lower limb CRPS
	<b>Gustin 2010</b> <sup>123</sup>
Randomisation	randomised, method not explained
Outcome measure	VAS rest pain, VAS movement pain, disability score, functional MRI
Early or Late	6 to 36 months
Diagnosis	IASP, mixed type I and II
Intervention	morphine alone v morphine with memantine*
Design	active v active double-blind
Outcome	positive; combination was more effective than morphine alone, from 1 to at least 8 weeks into the intervention
Population	20 patients with CRPS

\* These results were not replicated in a later study. Goebel A. Morphine and memantine treatment of long-standing complex regional pain syndrome. *Pain Med* 2012;13(3):357–8.

	<b>Picarelli 2010</b> <sup>124</sup>
Randomisation	randomised, method not explained
Outcome measure	VAS pain, McGill questionnaire, Depression and Anxiety, Disability
Early or Late	10–180 months
Diagnosis	IASP CRPS type I upper limb
Intervention	repetitive transcranial magnetic stimulation v. sham (1x per day for 10 days)
Design	active v placebo/sham (best medical treatment was continued in both groups) double blind
Outcome	repetitive transcranial magnetic stimulation relieves pain better than sham (greatest pain relief at day 10, not persisting after one week or three months)
Population	23 'refractory' patients completed the protocol
Comment	figure 2 incorrectly labelled? Shows sham better than active

## Glossary of terms

Allodynia	Meaning 'other pain', this is a pain due to a stimulus which does not normally provoke pain, and which can be either thermal or mechanical.
Ankylosis	A stiffness of a joint due to abnormal adhesion and rigidity of the bones of the joint, which may be the result of injury or disease. The rigidity may be complete or partial and may be due to inflammation of the tendons or muscular structures outside the joint, or of the tissues of the joint itself.
Anticonvulsants	A diverse group of pharmaceuticals used in the treatment of epileptic seizures. Anticonvulsants are also increasingly being used in the treatment of bipolar disorder, since many seem to act as mood stabilisers. Anticonvulsants are more accurately called antiepileptic drugs (AEDs).
Antidepressants	Medication used to alleviate mood disorders, such as major depression and dysthymia, and anxiety disorders such as social anxiety disorder. Drugs including the monoamine oxidase inhibitors (MAOIs), tricyclic antidepressants (TCAs), tetracyclic antidepressants (TeCAs), selective serotonin reuptake inhibitors (SSRIs), and serotonin-norepinephrine reuptake inhibitors (SNRIs) are most commonly associated with the term.
Beau's lines	Deep transverse grooves of nails. They can be on any or all nails. They may look like indentations or ridges in the nail plate.
Bullae	Large vesicles.
CBT	Cognitive behavioural therapy. This is not a single therapy or even a single set of standardised interventions. Rather CBT is a broad category of different treatment regimens. Almost always, however, CBT regimens 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/activity-rest cycling, exercise and activity management, and/or pleasant activity scheduling
Compartment syndrome	The compression of nerves, blood vessels, and muscle inside a closed space (compartment) within the body. This leads to tissue death from lack of oxygenation; the blood vessels being compressed by the raised pressure within the compartment. Compartment syndrome most often involves the forearm and lower leg.
Contracture	In a muscle or muscle fiber, usually refers to a continuous contraction in the absence of a stimulus, such as an action potential. A muscle contracture is a shortening of a muscle or joint.
Debridement	Debridement is the medical removal of a patient's dead, damaged, or infected tissue to improve the healing potential of the remaining healthy tissue. Removal may be surgical, mechanical, chemical, autolytic (self-digestion), and by maggot therapy, where certain species of live maggots selectively eat only necrotic tissue.
Dystonia	A neurological movement disorder, in which sustained muscle contractions cause twisting and repetitive movements or abnormal postures.

Epithelioid haemangioma	Angiolymphoid hyperplasia with eosinophilia (also known as epithelioid hemangioma), usually presents with pink to red-brown, dome-shaped, dermal papules or nodules of the head or neck, especially about the ears and on the scalp.
Erythromelalgia	Also known as Mitchell's disease (after Silas Weir Mitchell), acromelalgia, red neuralgia, or erythermalgia, is a rare neurovascular peripheral pain disorder. There is severe burning pain (in the small fibre sensory nerves) and skin redness. The attacks are periodic and are commonly triggered by heat, pressure, mild activity, exertion, insomnia or stress.
Erythema	Erythema is redness of the skin, caused by hyperemia of the capillaries in the lower layers of the skin. It occurs with any skin injury, infection, or inflammation.
Fascia	A layer of fibrous tissue that permeates the human body. A fascia is a connective tissue that surrounds muscles, groups of muscles, blood vessels, and nerves, binding those structures together. It consists of several layers: a superficial fascia, a deep fascia, and a subserous (or visceral) fascia and extends uninterrupted from the head to the tip of the toes.
Hyperaesthesia	A condition that involves an abnormal increase in sensitivity to stimuli of the senses. Stimuli of the senses can include sound that one hears, foods that one tastes, textures that one feels, and so forth.
Hyperalgesia	A condition where normally painful stimuli (eg a pinprick) are more painful than usual
Hyperhidrosis	Condition characterised by abnormally increased perspiration, in excess of that required for regulation of body temperature.
Hypoxia	A pathological condition in which the body as a whole, or a region of the body is deprived of adequate oxygen supply.
Hypertrichosis	Hair growth on the body in an amount considered abnormal. There are two distinct types of hypertrichosis: generalised hypertrichosis, which occurs over the entire body, and localised hypertrichosis, which is restricted to a certain area.
Hypotrichosis	Condition of abnormal hair patterns – predominantly loss or reduction. It occurs, most frequently, by the growth of vellus hair in areas of the body that normally produce terminal hair.
Intrathecal	Something introduced into or occurring in the space under the arachnoid membrane of the brain or spinal cord.
Litigation	A lawsuit or a civil action brought in a court of law in which a plaintiff, a party who claims to have incurred damages as a result of a defendant's actions, demands a legal or equitable remedy.
Leukonychia	Increased whiteness and opacity of the nails.
Motor dyspraxia	Motor skills disorder (also known as motor coordination disorder or motor dyspraxia) is a human developmental disorder that impairs motor coordination in daily activities. It is neurological in origin.
Myoclonus	Brief, involuntary twitching of a muscle or a group of muscles. It is a medical sign. The myoclonic twitches are usually caused by sudden muscle contractions; they also can result from brief lapses of contraction.

Neuromodulation	In neuromodulation, several classes of neurotransmitters regulate diverse populations of central nervous system neurons (one neuron uses different neurotransmitters to connect to several neurons). This is in contrast to direct synaptic transmission, in which one presynaptic neuron directly influences a postsynaptic partner (one neuron reaching one other neuron), neuromodulatory transmitters secreted by a small group of neurons diffuse through large areas of the nervous system. Neuromodulation also refers to the effect of neurostimulation such as spinal cord stimulation.
Neuropathic pain	‘Pain arising as a consequence of a lesion or disease affecting the somatosensory system’. Neuropathic pain may have continuous and/or episodic (paroxysmal) components. Common qualities include burning or coldness, ‘pins and needles’ sensations, numbness and itching
Osteoporosis	A disease of bones that leads to an increased risk of fracture. In osteoporosis the bone mineral density (BMD) is reduced, bone microarchitecture is deteriorating, and the amount and variety of proteins in bone is altered.
Onychodystrophy	Nail disease. A deformation of the nails.
Pathophysiology	The study of the changes of normal mechanical, physical, and biochemical functions, either caused by a disease, or resulting from an abnormal syndrome.
Placebo	A placebo is a sham or simulated medical intervention that can produce a (perceived or actual) improvement, called a placebo effect.
Prophylaxis	Any medical or public health procedure whose purpose is to prevent, rather than treat or cure a disease.
Qigong (Tai Chi)	A set of exercises, originally a Chinese martial art. Qi and gong – the two words are combined to describe systems and methods of ‘energy cultivation’ and the manipulation of intrinsic energy within living organisms.
Randomisation	Randomisation is the process of making something random.
Somatisation	A tendency to experience and communicate somatic distress in response to psychosocial stress and to seek medical help for it.
Spinal cord stimulation	A device is used to send pulsed electrical signals to the spinal cord to control chronic pain. It consists of stimulating electrodes implanted in the epidural space (space within the spinal canal lying outside the dura mater), an electrical pulse generator implanted in the lower abdominal area or gluteal region, conducting wires connecting the electrodes to the generator internally, and the generator remote control. Spinal cord stimulation (SCS) has notable analgesic properties.
Sudomotor	A medical term used to describe something that stimulates the sweat glands.
Systematic review	A literature review focused on a research question that tries to identify, appraise, select and synthesise all high-quality research evidence relevant to that question. Systematic reviews of high-quality randomised controlled trials are crucial to evidence-based medicine.
Thoracic outlet syndrome	A syndrome involving compression at the superior thoracic outlet, of a neurovascular bundle passing between the anterior scalene and middle scalene muscles. It can affect the brachial plexus (nerves that pass into the arms from the neck), and/or the subclavian artery or vein (blood vessels that pass between the chest and upper extremity).

Vasculitis	A heterogeneous group of disorders, that are characterised by inflammatory destruction of blood vessels. Both arteries and veins are affected. Lymphangitis is sometimes considered a type of vasculitis. Vasculitis is primarily due to leukocyte migration and resultant damage.
Vasoconstriction	Narrowing of the blood vessels resulting from contraction of the muscular wall of the vessels, particularly the large arteries, small arterioles and veins. The process is the opposite of vasodilation, the widening of blood vessels.
Vasodilation	Widening of blood vessels resulting from relaxation of smooth muscle cells within the vessel walls, particularly in the large arteries, smaller arterioles and large veins. The process is essentially the opposite of vasoconstriction, or the narrowing of blood vessels. When vessels dilate, the flow of blood is increased due to a decrease in vascular resistance. Therefore, dilation of arterial blood vessels (mainly arterioles) leads to a decrease in blood pressure.
Weber-Christian syndrome	Also known as relapsing febrile non-suppurative panniculitis, is a cutaneous condition characterised by recurrent subcutaneous nodules that heal with depression of the overlying skin. It is a type of panniculitis.

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