Royal College of Physicians

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Complex regional pain syndrome

Concise guidance to good practice series

December 2011

Clinical Medicine 2011, Vol 11, No 6: 596–600

Complex regional pain syndrome in adults: concise guidance

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ABSTRACT – Complex regional pain syndrome (CRPS) is a debilitating, painful condition in a limb associated with sensory, motor, autonomic, skin and bone abnormalities. Pain is typically the leading symptom, but is often associated with limb dysfunction and psychological distress. Prompt diagnosis and early treatment is required to avoid secondary physical problems related to disuse of the affected limb and the psychological consequences of living with undiagnosed chronic pain. UK guidelines have recently been developed for diagnosis and management in the context of primary and secondary care.[1] The purpose of this concise guideline is to draw attention to these guidelines. Information in this article has been extracted from the main document and adapted to inform the management of CRPS as it presents to physicians in the course of their daily practice.

KEY WORDS: clinical guidelines, complex regional pain syndrome

Background

Complex regional pain syndrome (CRPS) is a debilitating condition, characterised by pain in a limb, in association with sensory, vasomotor, sudomotor, motor and dystrophic changes. It commonly arises after injury to that limb. Pain is typically the leading symptom of CRPS, but is often associated with limb dysfunction and psychological distress. Patients frequently report neglect-like symptoms or a feeling that the limb is 'alien'.[2]

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. The subtype of CRPS has no consequences for the general approach to management, but the cause of nerve damage in CRPS 2 should always be clarified – urgently in acute cases (see guideline 1.2).

The diagnosis of CRPS cannot be made on imaging or laboratory tests. The condition is diagnosed on the basis of clinical criteria[3] shown in Box 1. Differential diagnoses are listed in Box 2.

Aetiology and course

The cause of CRPS is currently unknown.

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On behalf of the guideline development group (GDG). Please refer to supplementary electronic documentation for full list of GDG members. The full guideline will be available from the Royal College of Physicians website (www.rcplondon.ac.uk) in 2012.

Series editors: Lynne Turner-Stokes and Bernard Higgens

Box 1. Budapest diagnostic criteria. Adapted from reference 3.

All of the following statements must be met:

- The patient has continuing pain which is disproportionate to any inciting event
- The patient has at least one sign in two or more of the categories below
- The patient reports at least one symptom in three or more of the categories below
- No other diagnosis can better explain the signs and symptoms.

Category	Sign/symptom
1 'Sensory'	Allodynia (pain to light touch and/or temperature sensation and/or Deep somatic pressure and/or joint movement) and/or Hyperalgesia (to pinprick).
2 'Vasomotor'	Temperature asymmetry and/or Skin colour changes and/or Skin colour asymmetry.
3 'Sudomotor/oedema'	Oedema and/or Sweating changes and/or Sweating asymmetry.
4 'Motor/trophic'	Decreased range of motion and/or Motor Dysfunction (weakness, tremor, dystonia) and/or Trophic changes (hair/nail/skin).

- Precipitating factors include injury and surgery. However, there is no relationship to the severity of trauma while in some cases there is no precipitating trauma at all (9%).[4] The development of CRPS does not mean that surgery was suboptimal.[5]
- Transient features of CRPS are much more common than most clinicians realise – occurring in up to 25% of minor limb injuries.[6]
- Approximately 15% of sufferers will have unrelenting pain and physical impairment >5 years after CRPS onset, although more patients will have a lesser degree of ongoing pain and dysfunction[7] impacting on their ability to work and function normally.
- There is no medical cure for CRPS.

It is also now clear that CRPS is *not* significantly associated with a history of pain-preceding psychological problems, or with somatisation or malingering. Patients still report feeling stigmatised by health professionals who do not believe that their condition is 'real'. However, patients may require psychological intervention and support to deal with the particularly distressing nature of CRPS

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Box 2. Differential diagnoses for complex regional pain syndrome (CRPS).

More common conditions:

- bony or soft tissue injury (including stress fracture, ligament damage, instability)
- neuropathic pain (eg due to peripheral nerve damage or central nervous system/spinal lesion)
- arthritis/arthrosis
- infection (bony, joint, soft tissue, skin)
- compartment syndrome
- arterial insufficiency (usually due to atherosclerosis in the elderly, trauma or thromangiitis obliterans (Burger's disease))
- Raynaud's disease
- lymphatic or venous obstruction
- thoracic outlet syndrome (due to either nerve compression or vascular compression).

Rare conditions:

- Gardner-Diamond syndrome*
- erythromelalgia (may include all limbs)
- self-harm/malingering.

**Psychogenic purpura (Gardner-Diamond syndrome, autoerythrocyte sensitization, painful bruising syndrome) is a rare and poorly understood clinical presentation of unexplained painful ecchymotic lesions, mostly on the extremities and/or the face.*

symptoms. Psychosocial risk factors that may predict chronicity[8] are shown in Box 3.

Treatment approach

- Prompt diagnosis and early treatment is required to avoid secondary physical problems associated with disuse of the affected limb and the psychological consequences of living with undiagnosed chronic pain.[10]
- Early referral to physiotherapy and encouraging gentle movement as early as possible, may potentially prevent progression of symptoms.[11]
- Except in mild cases, patients with CRPS are generally best managed in specialist pain management or rehabilitation programmes.
- An integrated interdisciplinary treatment approach is required, including the four 'pillars of intervention' (see Box 4).

This guidance

UK guidelines have recently been developed for diagnosis and management in the context of primary and secondary care.[1] Information in this concise guidance document has been extracted from the main document and adapted to inform the management of CRPS as it presents to physicians in the course of their daily practice.

The guidelines were drawn up in accordance with the principles recommended by the AGREE Collaboration.[12] The methodology table is available online, and a full description of the methodology and systematic literature review may be found in the main guideline document.[1] The original 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. However, in the absence of RCT-based evidence to inform specific guidance with respect to diagnosis and management, the majority of recommendations in this document are based on the expert opinion of service users (E1) and professionals (E2) (according to the classification used by the National Service Framework for Long Term Conditions).[13]

Box 3. Identified psychosocial risk factors (yellow flags) which may predict chronicity. Adapted from reference 9.

Yellow flags

- Iatrogenic factors, ie previous negative experiences with health professionals.
- Excessive illness behaviour.
- Poor coping strategies, eg ongoing 'guarding ' of the limb despite education.
- Involvement in litigation is affecting willingness to progress in treatment.
- Overuse of appliances.
- Distress.
- Anxiety/depression.
- Lack of willingness to set goals.
- Passive in treatment sessions.
- Inappropriate beliefs despite education.
- Negative family influences.

Implications for implementation

The implications for implementation primarily relate to training requirements for clinical staff to recognise CRPS and make the appropriate referrals for specialist management.

Currently, access to specialist pain management and rehabilitation for CRPS is patchy across the UK. For the subgroup of patients with complex needs, there is an additional requirement for closer liaison between these services to share their expertise and resources, as well as ongoing facilities (including self-help and support groups) to assist patients to manage their own symptoms and optimise their level of physical psychological and social function.

References

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2 Frettlöh J, Hüppe M, Maier C. Severity and specificity of neglect-like symptoms in patients with complex regional pain syndrome (CRPS) compared to chronic limb pain of other origins. *Pain* 2006;124: 184–9.

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Box 4. Four pillars of treatment for complex regional pain syndrome (CRPS) – an integrated interdisciplinary approach.

1 Patient information and education

Patients should be provided with appropriate education about CRPS to support self-management:

- a sample patient information sheet is available (see full guidelines).

Patients should be:

- reassured that physical and occupational therapy are safe and appropriate
- engaged in the process of goal-setting and review.

2 Pain relief (medication and procedures)

Pain specialists should be aware of the evidence for efficacy of pain interventions in the context of CRPS (see full guideline for systematic review).

No individual drugs or pain intervention procedures can be recommended at the current time due to lack of evidence (see full guideline for systematic review).

The following may be considered:

- neuropathic pain medication delivered according to National Institute for Health and Clinical Excellence guidelines[14]
- pamidronate (60 mg intravenous single dose) for suitable patients with CRPS of <6 months duration as a one-off treatment
- spinal cord stimulator treatment for patients with CRPS who have not responded to appropriate integrated management.

3 Physical and vocational rehabilitation

Physical rehabilitation should be delivered by therapists competent in treating patients with chronic pain and/or CRPS.

Emphasis should be on restoration of normal function and activities through acquisition of self-management skills, with patients and their families actively engaged in goal setting.

The programme may include elements of chronic pain management including:

- general body re-conditioning through graded exercise, gait re-education, postural control
- restoration of normal activities, including self-care, recreational physical exercise and social and leisure activities
- pacing and relaxation strategies, to support self-management of the condition
- vocational support.

It may also include specialised techniques to address altered perception and awareness of the limb, for example:

- self-administered desensitisation with tactile and thermal stimuli
- functional movement to improve motor control and limb position awareness
- graded motor imagery, mirror visual feedback, mental visualisation
- management of CRPS-related dystonia.

4 Psychological intervention

Psychological intervention is based on individualised assessment, to identify and proactively manage any factors which may perpetuate pain or disability/dependency including:

- mood evaluation – management of anxiety/depression
- internal factors, eg counter-productive behaviour patterns
- any external influences or perverse incentives.

It usually follows principles of cognitive behavioural therapy delivering:

- coping skills and positive thought patterns
- support for family/carers to manage their own needs and to maintain relationships.

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11 Oerlemans HM, Oostendorp RAB, de Boo T *et al.* Pain and reduced mobility in complex regional pain syndrome I: outcome of a prospective randomised controlled clinical trial of adjuvant physical therapy versus occupational therapy. *Pain* 1999;83:77–83.

12 Guideline development in Europe: an international comparison. *J Technol Assess Health Care* 2000;16:1036–46.

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The guidelines

1 Diagnosis and exclusion of remediable conditions — Grade

1.1 **Complex regional pain syndrome (CRPS)** should be considered in the presence of **persistent or disproportionate limb pain,** particularly in the context of trauma or surgery. — **E2**

All clinicians should be:

- aware of the condition
- familiar with the **Budapest criteria** for diagnosis (see Box 1).

1.2 **Exclusion of reversible causes** — **E2**

Where **disproportionate pain persists for longer than expected:**

1. differential diagnoses should be considered and investigated as a matter of urgency, to **exclude remediable conditions** (see Box 2)
2. where pain occurs in the distribution of a peripheral nerve following a surgical procedure, the patient should be referred back to the responsible surgeon for urgent re-evaluation and possible surgical exploration.

1.3. **Patient information**

- General post-fracture/operation **patient information leaflets** should include advice to observe and report **CRPS warning signs.** — **E1/2**
- **Management should include reassurance** that:
  - CRPS is a recognised condition although its causes are poorly understood
  - its development should not be considered evidence of suboptimal surgical management
  - symptoms are transient in the majority of cases, and usually respond well to desensitisation (see 2.3), movement and resumption of normal activities.

2 Early management of CRPS – analgesia and exercise

2.1 Medical management should include: — **E2**

- **regular review,** with advice about the use of simple analgesics, eg paracetamol, co-codamol (particularly for trauma-related pain)
- **non-steroidal anti-inflammatory drugs** (in the presence of bony or soft tissue trauma, and in the absence of contraindications).

If simple medication does not reduce pain to a mild level after 3–4 weeks, consider using **medication for neuropathic pain** according to the National Institute for Health and Clinical Excellence guidelines for neuropathic pain.[14] Earlier use may be appropiate.

2.2 The following treatments **are not generally recommended** and should be used with caution by appropriately experienced clinicians: — **E2**

- **opiate analgesia** – except with specific advice from a specialist pain clinic
- **intravenous regional sympathetic block**s – these have no proven efficacy in prospectively randomised studies
- **amputation** – this does not provide pain relief in CRPS but may be necessary for life threatening symptoms, such as infection. The patient should be warned that the CRPS may recur in the stump and the pain may be worsened
- **surgery on a CRPS-affected limb** – surgery represents a painful stimulus and may worsen CRPS. It should be avoided where possible. Where surgery is unavoidable, it should be carried out by a surgical team experienced in this area and an anesthetist with experience in the management of patients with chronic pain.

2.3 **Analgesia should not be given in isolation**, but should be prescribed with the aim of supporting an **exercise/therapy programme.** — **RB/E2**

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

2.4 Unless contraindicated, **physiotherapy and/or occupational therapy** should be initiated **immediately when CRPS is suspected**. — **RB/E2**

- Early physical therapy should include:
  - encouragement to look at the affected limb
  - **gentle movement** and **desensitisation** (eg gentle stroking with textured fabrics)
  - early incorporation into **functional activity** and weight-bearing when tolerated.
- Slings, splints and other **immobilising devices should be avoide**d wherever possible
- Orthotic devices (such as insoles) can support weight bearing and function, but should only be provided under a physiotherapist's supervision.

3 Monitoring and escalating referral

3.1 Patient should be kept under **regular monitoring** to: — **E2**

- assess the effectiveness of treatment and pain control
- assess and monitor mood, in particular for signs of developing distress and/or evidence of depression/anxiety.

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The guidelines (*continued*)

In the **presence of persisting or worsening pain**:

- **timely onward referral** should be made to **specialist pain and/or rehabilitation** services (see below)
- escalation should be proportionate to the severity/impact of pain symptoms
- the presence of certain **psychosocial risk factors identified as 'yellow flags'** (see Box 3) may predict chronicity[7] and should **prompt early referral** to multidisciplinary services with access to psychological interventions.

4 Specialist pain management and rehabilitation

4.1 Patients with moderate or severe persisting CRPS should hav**e timely access to specialist pain clinics** able to offer an **integrated multidisciplinary approach to treatment** (the four pillars of intervention, see Box 4 for details). **RC/E2**

4.2 Specialist pain management services and rehabilitation teams should work together to **share their expertise and resources** for patients with severe complex presentations of CRPS. **E2**

5 Patients with complex disabling CRPS **E2**

5.1 Patients with **complex disabling CRPS** should have access to specialist interdisciplinary **rehabilitation programmes.** **E2**

5.2 Specialised rehabilitation programmes led/supported by a consultant in rehabilitation, may be required in the following situations:

- CRPS presenting in the context of:
  - o another existing disabling condition (eg stroke, severe multiple trauma)
  - o complex psychological or psychiatric co-morbidities – either pre-or post-dating the onset of CRPS
- severe physical disability/dependency requiring treatment in an inpatient setting
- specialist facilities, equipment or adaptations are required, or need review
- unable to work – requiring specialist vocational rehabilitation/support
  - o eg supported work schemes, employer liaison for job modification
  - o or support for work withdrawal where appropriate.
- litigation is ongoing – requiring support to facilitate an early conclusion.

6 Long-term ongoing support **E1/2**

6.1 People with CRPS should have access to **appropriate information, advice, education and support** to allow them to understand and manage their condition optimally. **E1/2**

6.2 People with CRPS should have access to a range of facilities to maintain their independence, and levels of activity and societal participation, which may include:

- **self-help and peer support** groups
- facilities for **self-directed exercise** (eg adapted gym, swimming/hydrotherapy pool)
- support for **vocational, social and leisure** activities **E1/2**
- **counselling/psychological support where indicated.**

These services are often appropriately run by voluntary organisations with input from professionals as required.

6.3 **Families and carers** of people with CRPS should have access to **advice, support and information** to manage their own needs and to **maintain relationships.**

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