Chronic pain problems in children and young people

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Chronic pain has been defined as pain persisting beyond the usual course of an acute disease or reasonable time for injury to heal (usually taken to be about 3 months) or pain that is associated with a chronological pathological process which causes continuous pain or recurrent pain which recurs at intervals for months or years. Contrary to popular belief, it is likely that substantial numbers of children suffer from chronic pain; population surveys in several countries have found that 20–60% of adolescents complain of significant recurrent pain lasting more than a year, although only a small number of these children seek medical advice, and even fewer require specialist care.1,2

Most chronic or recurrent pain in children is easily managed in primary care settings and does not result in significant functional impairment once serious treatable underlying causes have been identified or excluded. However, for a significant minority of children, pain becomes a dominant and refractory symptom needing multidisciplinary input from pain medicine specialists for effective treatment. Although chronic pain in children can arise from well-recognized clinical conditions such as juvenile idiopathic arthritis (JIA) or inflammatory bowel disease, more commonly in those attending chronic pain clinics, the cause of pain cannot be easily linked to a known or even reasonably well-understood underlying pathology. In these children, chronic pain may present in various guises including complex regional pain syndrome (CRPS), widespread musculoskeletal pain (juvenile fibromyalgia), recurrent abdominal pain (RAP), and chronic headache. In addition, although many conditions appear superficially to be similar to those found in adults, there are often important differences in epidemiology, presenting symptoms putative underlying mechanisms, appropriate treatments, and prognosis.3

Pain in childhood can have far-reaching effects, and not just for the child or family. The total financial burden on the UK economy of adolescent chronic pain in 2005 was estimated to be as high as £3840 million in one study.4 Despite this, and although clinical teams dedicated to the management of childhood chronic pain have been established in hospitals in many parts of the world, access to the most up-to-date treatment is often delayed or non-existent for many patients.

General principles

Untreated long-term pain is capable of devastating functional consequences at all ages. In children, it can lead to a recognizable pattern of associated problems (Table 1) that contribute to the morbidity associated with pain.5 Consequently, paediatric chronic pain management is centred on a biopsychosocial model that acknowledges the complex interaction of developmental biological, psychological, educational, family, and other influences on the presentation, intensity, and effects of pain in a developing child. Family functioning and the influence of parental responses to their children’s pain are also important issues that must be considered in the overall management plan.

Pain treatment in children is multimodal and symptom focused, including the pharmacological and non-pharmacological management of pain and the active management of factors contributing to functional impairment such as disturbed sleep, anxiety, low mood, poor pain coping skills, reduced school attendance, and impaired social development. Specialist paediatric pain management therefore requires the skills of a suitably qualified multidisciplinary specialist team working in a coordinated fashion (Table 2).6

Chronic pain assessment

The pattern of pain reporting, symptoms, and types of pain coping strategies vary with age, and therefore, appropriate age-specific evaluation is important. In taking a pain history, the site, intensity, and characteristics of pain are...
the effectiveness of management strategies.

therapies and if repeated at intervals allow tracking of progress and children with chronic pain. Such evaluations provide targets for the PedsQL questionnaire have also been used widely in children but psychological symptoms such as anxiety are documented together with any precipitating or relieving factors. Pain intensity is traditionally evaluated using a 0–10 visual analogue scale. Children as young as 6 yr may be able to understand this concept with suitable explanation and help. In addition, documentation of sleeping pattern, physical functioning, pain coping style, family, and social interactions including school attendance and academic performance are also important.

Chronic pain that is associated with significant functional or emotional impairment such as low mood, high levels of pain-associated anxiety, and poor school attendance is a strong indication for referral to specialist care.

Children who are referred to a paediatric pain management centre will generally undergo an exhaustive developmentally appropriate multidisciplinary assessment. Questionnaire-type evaluations are frequently used to establish baseline levels of important variables. For example, the Bath Adolescent Pain Questionnaire (BAPQ) is a chronic pain assessment tool that incorporates multiple domains including mood, functional status, and school attendance. There is also a parent questionnaire that evaluates parenting behaviour, marital adjustment, and family role functioning. More general assessments such as the Childhood Health Assessment Questionnaire (CHAQ) and the Pediatric Quality of Life (PedsQL) questionnaire have also been used widely in children with chronic pain. Such evaluations provide targets for therapy and if repeated at intervals allow tracking of progress and the effectiveness of management strategies.

Treatment

Education, including an explanation of pain mechanisms, advice on lifestyle, and pain coping strategies, combined with physical and pharmacological treatment are the mainstays of management. Invasive procedures such as neuraxial injections, complex nerve blocks, spinal cord stimulation, or other neurosurgical techniques have been little used in children and are relatively rarely indicated in comparison with adult pain medicine practice.

There is no evidence for the existence of ‘psychological pain’ in children but psychological symptoms such as anxiety are common and are associated with the intensity and persistence of pain, regardless of diagnosis. In addition, psychological symptoms in parents appear to be strongly related to treatment-seeking behaviour in parents and children; allaying fears and reducing anxiety are therefore important priorities. Formal psychological techniques such as behaviour modification or cognitive behaviour therapy have been shown to be very effective at reducing pain and dysfunction in many types of childhood pain (Table 3). Addressing disordered sleeping patterns and instituting a ‘paced’ incremental increase in normal physical activity with the aid of physiotherapy and occupational therapy aim to restore normal functioning.

Pharmacological treatments are given on the basis of known or inferred pain mechanisms, or directly targeted at pain-associated symptoms such as insomnia or anxiety. For example, tricyclic antidepressants such as amitriptyline can be effective against neuropathic pain (NP) and also have a sedative effect that can be useful to help restore normal sleep patterns. Specific treatment of any known underlying condition can also reduce pain intensity and so close liaison with the relevant primary clinical team is essential; this is particularly true in cancer pain and painful inflammatory conditions such as JIA.

Simple analgesics such as paracetamol and non-steroidal anti-inflammatory drugs (NSAIDs) should not be forgotten, even though they have often already been tried at presentation and found to be ‘ineffective’. They have a role in managing conditions where nociceptive or inflammatory pain is prevalent. As with all analgesics, their effectiveness should be optimized by ensuring correct dosing and timing of treatment.

In contrast, opioids have little to contribute in the long-term management of many conditions that are not self-limiting in children (but may be useful in the treatment of cancer pain and pain in palliative care settings). Mild opioids have frequently been introduced early in pain treatment and many patients who present with chronic pain are already taking significant doses of prescribed or ‘over-the-counter’ preparations. Constipation, lethargy, nausea, and drug tolerance are problematic side-effects seen with even quite small doses and so opioids are usually withdrawn slowly if they are not strongly indicated. Physical treatments such as transcutaneous nerve stimulation (TENS), acupuncture, hot or cold packs, massage, and reflexology are often used in children either alone, or as adjuncts to other analgesics.

Pain management programmes

Failure to progress despite therapy is an indication for multidisciplinary review and reconsideration of the management plan. Some
patients require more intensive rehabilitation with inpatient admission usually for 2–3 weeks in order to improve physical functioning, pain coping skills, increase autonomy, and develop more effective self-management techniques. Such pain management programmes (PMPs) are sometimes intended for children with a particular diagnosis such as headache, abdominal pain, or CRPS and therefore place more emphasis on psychological or physical techniques as appropriate. More commonly, programmes are designed to be flexible and therefore suitable for different types of pain. Unfortunately, PMPs are only available in some specialist centres in a few parts of the developed world.

**Chronic pain conditions**

**Neuropathic pain**

In adults, sensory dysfunction associated with direct, indirect, or inferred nerve damage leads to characteristic symptoms including allodynia, hyperalgesia, and painful dysesthesias described as NP. Examples include diabetic and post-herpetic neuropathy, certain kinds of degenerative back pain, and phantom limb pain. Multiple mechanisms are known to operate in NP leading to changes in nociceptor thresholds and neural excitability. Until relatively recently, NP was rarely, if ever, reported in children, but it is now clear that children can suffer from NP, but there are important differences in epidemiological mechanisms and prognosis between children and adults. Known causes in childhood include inherited neuropathies such as erythromelalgia, direct invasion of nervous tissue, or pressure effects by malignancies; post-traumatic disorders (e.g. phantom limb pain and persistent post-surgical pain) drug toxicity (e.g. chemotherapeutic agents) and HIV infection. Idiopathic NP may be diagnosed on the basis of characteristic symptomatology and can be a component of some types of abdominal and musculoskeletal pain. CRPS is usually considered a variant of NP and is discussed in more detail below.

NP in children appears to have a better prognosis for recovery than NP in adults; this is probably related to differences in underlying mechanisms such as lower capacity to activate spinal microglial pain maintaining responses and the greater capacity of the developing nervous system to adapt to injury.

The evidence base for the use of medications and interventions in childhood NP is weak and largely based on case reports or extrapolations from adult data. Systemic treatment with anti-depressants (amitriptyline) or anticonvulsants (gabapentin and pregabalin) is sometimes effective and usually given on a ‘trial’ basis for 3 or 4 weeks. Local anaesthesia can block pain in NP, and indeed, i.v. lidocaine is sometimes used as a diagnostic test. Localized NP can usually be temporarily (hours or days) relieved by regional nerve blocks, but this approach is limited by the short duration and toxicity of most agents and problems associated with indwelling catheters for long-term use. Topical ‘lidocaine patch’ treatment may be an alternative. Specific treatment of NP is difficult, empirical, and often unsatisfactory and so measures to improve pain coping behaviours and lifestyle change to minimize the impact of pain are also very important.

Physical treatments such as desensitization, massage, and paced exercise may also be of benefit in restoring normal function in NP; this is particularly true in CRPS where physiotherapy is the mainstay of treatment.

**Musculoskeletal pain**

Children with widespread musculoskeletal pain, sometimes called juvenile fibromyalgia or diffuse idiopathic pain (DIP) syndrome, are increasingly presenting for treatment. Characteristically, they have pain at multiple sites, disturbed sleep, fatigue, and low mood. A significant number also have a degree of joint laxity known as benign joint hypermobility syndrome (BJHS), but the relationship between BJHS and pain is sometimes disputed. Management is symptomatic, again involving multidisciplinary assessment and treatment. Exercise therapy, simple analgesics, and psychological strategies are most often used. Physiotherapy is also essential in improving joint stability in BJHS, thereby reducing the incidence of sprains and other injuries that may be important precipitants of more widespread pain. The prognosis of widespread musculoskeletal pain in children appears to be better than fibromyalgia syndrome in adults.

**Complex regional pain syndrome**

Formerly known as reflex sympathetic dystrophy or causalgia, CRPS is characterized by pain and swelling at a distal site accompanied by discoloration, abnormal sweating, and dysmorphic changes to hair and nails. Most commonly, there is no precipitating factor, but CRPS may also follow mild trauma or frank nerve damage. Pain in CRPS has similar characteristics to NP; other changes are attributed to autonomic involvement, which has also been demonstrated in laboratory models of the condition. CRPS in adults has been the object of intense scrutiny and it is clear that multiple mechanisms operate such that it is better to view the condition as a local manifestation of a systemic pathological process. CRPS in children is less well studied, but there are notable differences in its epidemiology and characteristics; it is rare below the age of 5 yr, with a peak incidence at 14 yr, it is much more prevalent in girls, and in contrast to adults, the lower limb is more commonly affected. The prognosis of childhood CRPS is good, although relapses are common (25–40%), but they can often be aborted or treated by correct self-management. Mobilization of the affected limb is important, and so patients presenting with CRPS need early referral to an appropriately experienced physiotherapist. Delay in diagnosis and inappropriate treatment may adversely affect prognosis.

Amitriptyline, gabapentin, and other agents active in NP are often used, and although conventional analgesics, including opioids, are not usually directly therapeutic, they may help the tolerability of painful exercises. Severe CRPS can be accompanied by
dystonia with strikingly abnormal posture or limb position and sometimes associated movement disorders. A few refractory cases have been treated with invasive therapies including adjunctive sympathetic blocks and even spinal cord stimulation.

**Headache**

Headaches are common during childhood. Most are benign, do not lead to long-term problems, and are managed by simple measures. However, a minority of children report daily headaches that are associated with significant disability. Headache in childhood is usually investigated and treated in consultation with a paediatric neurologist.

Primary headache disorders in children are predominantly migraine and tension-type headaches. Secondary headaches such as those related to central nervous infection, mass lesions, or other pathology should be excluded in cases of new-onset headache. Migraine headaches in children differ from those in adults; they usually start without an aura, and migraine variants are more common, for example, ophthalmoplegic migraine associated with third nerve paresis or complex migraine with other transient neurological abnormalities. More conventional migraine with unilateral headache frequently develops in later childhood.

Primary headache is usually amenable to pharmacological and cognitive-behavioural interventions. Patient and family education, removal of possible triggers, and lifestyle changes, for example, school attendance, physical activity, sleep, and dietary habits including adequate hydration are frequently effective. Simple analgesics are tried first; specific therapies and prophylaxis for migraine such as triptans and β-blockers are also used but have not been subjected to rigorous evaluation in children.

**Abdominal pain**

Abdominal pain in childhood is frequently mild, transient, and although often unexplained is generally managed conservatively. Significant ongoing pain is usually medically investigated in order to identify directly treatable causes such as inflammatory bowel disease, but the cause often remains obscure. Functional abdominal pain (FAP) otherwise known as RAP also includes diagnoses such as functional dyspepsia, irritable bowel syndrome (IBS), and abdominal migraine. The incidence of FAP peaks between ages 4 and 6 yr and again in early adolescence (particularly in girls aged >12 yr). The continuation of pain in children with FAP is associated with psychological distress, a family history of IBS, and an increased use of health-care services. Stressful life events such as school examinations, family, or social problems can trigger symptoms in FAP, IBS, and inflammatory bowel disease.

The treatment of FAP is focused on psychological treatments that are designed to improve coping skills and relaxation; self-hypnosis is often used. Child and parent education to reinforce healthy functional behaviour is also important. FAP symptoms are sometimes associated with specific foods, and eating is frequently cited as a pain trigger.

Peppermint oil supplements reduced symptoms in children with IBS in one randomized controlled trial. Amitriptyline, lamotrigine, and other drugs are sometimes prescribed, but a recent systematic review concluded that there is little evidence to support the use of analgesics or other drug treatments in FAP.

**Cancer pain and pain in palliative care**

There is a wide range of life-limiting conditions in children which may require pain management (Table 4). Children can suffer from a variety of types of pain due to both the underlying condition and treatments such as cytotoxic or antiretroviral therapy. Nociceptive pain, NP, musculoskeletal pain, headache, and other distressing symptoms may co-exist. For many children, pain management will include treatment of the underlying cause, for example, bony metastases may be treated with radiotherapy and painful muscle spasms can be treated with neuromuscular blocking agents, for example, diazepam, baclofen, or dantrolene. In children with malignant disease, oral chemotherapy may be helpful in reducing tumour-related musculoskeletal pain, and headaches from central nervous system leukaemia respond well to intrathecal methotrexate. In children with long-term conditions associated with immobility, such as osteogenesis imperfecta, musculoskeletal pain may respond to bisphosphonates.

Where such disease-modifying treatments are not appropriate or do not relieve pain sufficiently, analgesia should be used according to the underlying pain mechanism (if known) alongside psychological and practical approaches to pain management. The WHO ladder of analgesia, although now more than 20 yr old, is still widely adopted for children. Opioids are generally used more liberally and relatively early in life-limiting conditions, but recent evidence has shed some doubt on the routine use of codeine. This is because a significant number of children may be ‘poor metabolizers’ of the drug (to morphine) rendering it of little or no benefit, while low efficacy in infants may also be due to immaturity of hepatic enzyme systems.

Opioid management in opioid-sensitive pain can be challenging and combinations of rapid onset and long-acting preparations are often used; buccal, transmucosal, and transdermal formulations are very useful. Opioid rotation or the addition of adjuvant drugs, for example, ketamine or clonidine to control dose escalation due to tolerance may be required. Opioid side-effects must also be controlled. Nausea and vomiting are surprisingly rare and routine antiemetics are not needed often, but constipation is always
troublesome and so laxatives should be prescribed. NP is treated as outlined above and is often helped by antiepileptic and antidepressant drugs. For severe NP unresponsive to these treatments, the ratio of benefit–risk is often sufficient to warrant placement of long-term intrathecal or epidural catheters or to justify complex nerve block procedures.

Conflict of interest
None declared.

References

Please see multiple choice questions 17–20.