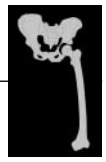
Strat Traum Limb Recon (2007) 2:55–56
DOI 10.1007/s11751-007-0014-8



CASE REPORT

C.C. Verheyen • R.M Castelein

A life without pain: a case report

Received: 1 January 2007 / Accepted in revised form: 18 March 2007

Abstract Congenital insensitivity to pain is a rare condition with an abnormality of interpretation of painful stimuli. This case report illustrates how a sequence of injuries after no or trivial trauma incapacitated a young boy. Especially the bilateral collapse and dislocation of the hip is an unusual sequela of this disorder.

Key words Congenital • Insensitivity • Pain • Charcot

C.C. Verheyen (✉)
Department of Orthopaedic Surgery and Traumatology
Isala Clinics (de Weezenlanden Hospital)
P.O. Box 10500, 8000 GM Zwolle, The Netherlands
e-mail: c.c.p.m.verheyen@isala.nl

R.M Castelein
Department of Orthopaedic Surgery
University Medical Center
Utrecht, The Netherlands

Introduction

'Pain is not the enemy, but the loyal scout announcing the enemy' [1].

A Caucasian boy was first seen at our department at the age of 12 years with a red and swollen left ankle. He was previously diagnosed with congenital insensitivity to pain with a high activity level, behavioural disturbances and admissions for self-mutilation. Initially for the ankle the diagnosis of septic arthritis was entertained but later it was established that in fact it concerned a Salter-Harris type IV physeal fracture of the left distal tibia probably sustained while playing soccer some weeks earlier. He was treated in a below-the-knee plaster of Paris cast for several months ultimately resulting in a Charcot arthropathy of the ankle.

Two years later open reduction and fixation of a displaced fracture of the left proximal humerus healed uneventfully. This fracture was diagnosed only several days after a fall. Shortly thereafter his parents had noted a swelling on the right thigh and radiographs demonstrated an avulsion of the anteroinferior iliac spine (Fig. 1). Two months later, at age 15, an acute impairment of the left hip turned out to be a slipped capital femoral epiphysis (Fig. 2). Despite an attempt at surgical fixation and spica cast immobilisation, not only was reposition of the involved hip not secured but also the right hip dramatically deteriorated to the point of collapse and dislocation (Fig. 3), leaving this previously active boy free of pain as before but severely restricted in his activities even to the point that he now, two years later, frequently uses a wheelchair.

Discussion

In the absence of a normal interpretation of painful stimuli the boy in this case report continued to be the victim of

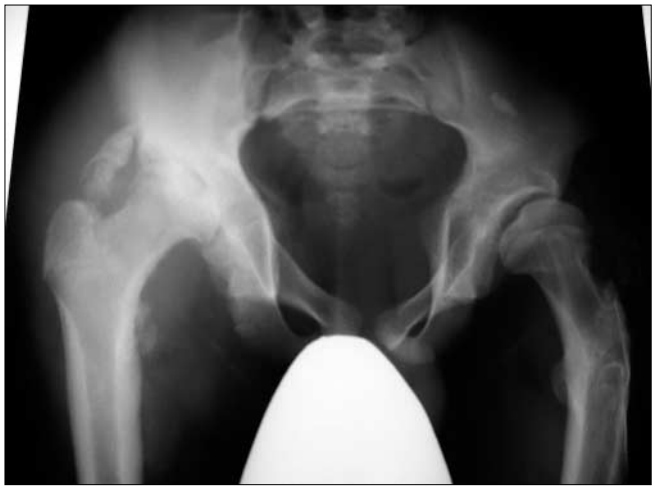


Fig. 1 Pelvic AP radiograph demonstrating avulsion of the anteroinferior iliac spine (*right*) and incongruity of both femoral heads where the left head is not centred in the acetabulum

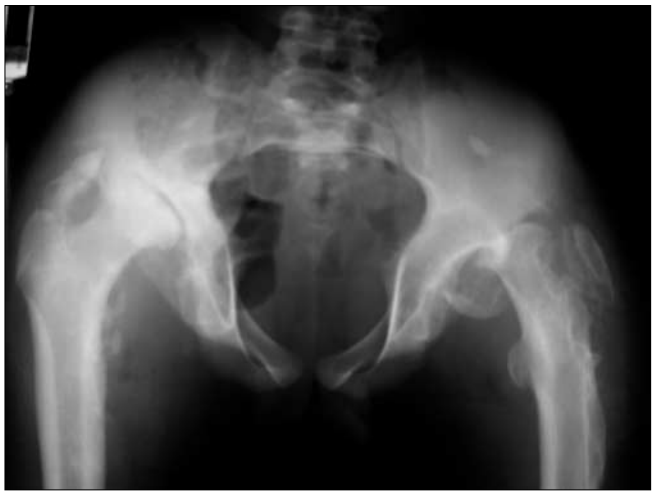


Fig. 2 Two months later: slipped capital femoral epiphysis and anteroinferior iliac spine avulsion (*left*)

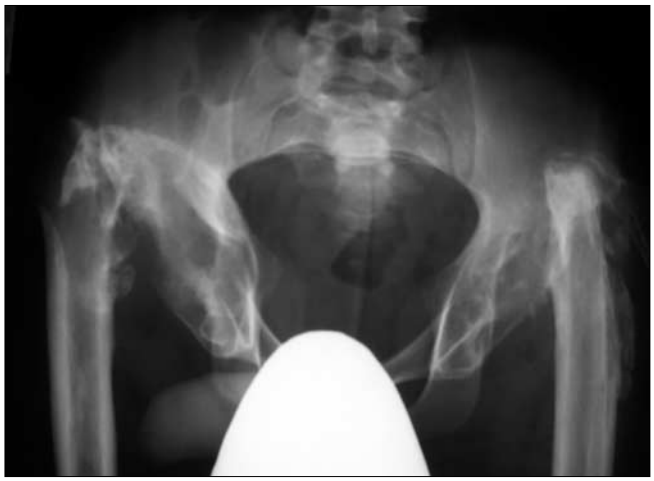


Fig. 3 Another 9 months later: severe destruction and dislocation of both hips

several skeletal injuries while playing. It illustrates that pain is essential in preserving physical integrity. The destruction and dislocation of both hips is a rare finding in congenital insensitivity to pain and has only once been previously reported [2].

Congenital insensitivity to pain is a rare neurological entity that was first described by Dearborn [3]. Its pathophysiology has not been established. It seems that patients have a life span close to normal although unnoticed visceral problems are a major concern. The incidence of skeletal and joint injuries decreases with age because as they mature patients become more aware of their limitations. In the differential diagnosis of the absence of perceived pain in children four other categories should be considered [4]: congenital sensory neuropathy, distal sensory neuropathy, familial dysautonomia (Riley-Day syndrome) and a group of miscellaneous disorders including syringomyelia, hysteria, mental deficiency and leprosy.

The first report of neuroarthropathy by Charcot [5] was described in tabes dorsalis (ataxic arthropathy). Charcot joints are very rare before closure of the epiphyseal plate but if present in children they are frequently caused by congenital insensitivity to pain [6]. Currently two theories are believed to play a significant part in the pathogenesis of Charcot arthropathy [7]. The first theory is neurotraumatic where joint destruction is the result of unrecognised (repetitive) trauma, the second is neurovascular referring to a sympathetic dysfunction (autosympathectomy) with joint dissolution from hyperaemia and bone resorption. In this case report where the joints are unable to sense pain, the former theory seems to be the obvious explanation for the Charcot joints of ankle and hips. The rapid deterioration of the more or less uninvolved right hip while in a protective spica however may also lend support to the neurovascular theory.

References

1. Brand PW, Yancy P (1993) Pain: the gift nobody wants. A surgeon's journey of discovery. Marshall Pickering, London
2. MacEwen GD, Floyd GC (1970) Congenital insensitivity to pain and its orthopedic implications. Clin Orthop Relat Res 68:100–107
3. Dearborn GV (1932) A case of congenital pure analgesia. J Nerv Ment Dis 75:612–615
4. Comings DE, Amromin GD (1974) Autosomal dominant insensitivity to pain with hyperplastic myelinopathy and autosomal dominant indifference to pain. Neurology 24:838–848
5. Charcot JM (1868) Sur quelques arthropathies qui paraissent dépendre d'une lésion du cerveau ou de la moelle épinière. Arch Physiol Norm Pathol 1:161–178
6. Greider TD (1983) Orthopedic aspects of congenital insensitivity to pain. Clin Orthop Relat Res 172:177–185
7. Brower AC, Allman RM (1981) Pathogenesis of the neurotrophic joint: neurotraumatic vs. neurovascular. Radiology 139:349–354