Unsuccessful surgical treatment of hip dislocation in congenital sensory neuropathy with anhidrosis

G. Köster, M. von Knoch, H.-G. Willert
From the University of Göttingen, Göttingen, Germany

A six-year-old girl with congenital sensory neuropathy with anhidrosis (CSNA) presented with bilateral hip dysplasia and subluxation on the right side.

Conservative treatment of the hips by closed reduction and a plaster cast was unsuccessful. When aged seven years the patient had an intertrochanteric varus rotation osteotomy on the right side, but subluxation was again evident after five months. A Salter-type pelvic osteotomy was carried out followed by immobilisation, but one year later subluxation was present in the right hip and dislocation in the left. At the age of nine years, the right femoral head resembled a Charcot joint, although walking ability was preserved.

In patients with CSNA, surgery may not always be advisable.

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Congenital sensory neuropathy with anhidrosis (CSNA) is a rare disease. Classically, patients present with absence of pain, anhidrosis, mental retardation and reduced perception of temperature.1 We describe a patient with CSNA who had bilateral hip dysplasia and subluxation which were treated surgically on one side. Our aim is to alert clinicians that surgical treatment may cause problems and not influence the course of the disease.

Case Report

The hips of a Turkish girl with CSNA were normal radiologically at nine months (Fig. 1). At the age of six years she developed a gait abnormality. Radiographs showed bilateral hip dysplasia and subluxation on the right side (Fig. 2). Since birth she had suffered febrile episodes of unknown origin. Sweating was never observed except in the axilla. She had no response to pain. Cranial nerve function was normal and her muscles had normal strength. Her motor development had been normal, but lingual development was delayed due to the loss of almost all her teeth. Her arms and legs showed multiple dermal scars from previous injuries.

Conservative treatment of the hips by closed reduction and a plaster cast could not stabilise the joints. At the age of seven years an intertrochanteric varus rotation osteotomy was carried out on the right side followed by immobilisation in a plaster cast for six weeks. The osteotomy healed satisfactorily but she developed severe heterotopic ossification around the operated hip (Fig. 3). Five months later the joint space had almost disappeared and the hip had subluxated again. The possibility of septic arthritis during the postoperative period was excluded by normal levels of C-reactive protein and normal white cell counts. There was no delay in wound healing or any wound discharge. A Salter-type pelvic osteotomy was carried out on the right hip followed by immobilisation in a plaster cast for six weeks. Again, there was no delay in the healing of the osteotomy or sign of infection, but heterotopic ossification occurred at the site of the operation (Fig. 4). One year later, the right hip had subluxated and the left hip was dislocated. By the age of nine years, the right femoral head was fragmented, largely resorbed and resembled a Charcot joint (Fig. 5). Despite the severe destruction of the joint, walking ability was still preserved, with complete absence of pain.

Discussion

In CSNA, insensitivity to pain can lead to severe skeletal problems and physical handicap. The principal problems are osteomyelitis, fractures and dislocations, and neuropathic arthropathies.2,3 Hip dysplasia and dislocation have been described twice as a sequel of CSNA. Hasegawa et al4 reported a patient in whom hip subluxation occurred at the age of five years and was treated conservatively. Roberts, Taylor and Burke5 described a 23-month-old patient with CSNA and hip dislocation which was refractory to conservative treatment. In this case, open reduction was suc-
Fig. 1
AP radiograph of the hips of the patient, when aged nine months, showing virtually normal development.

Fig. 2
AP radiograph of the hips of the patient when aged six years showing bilateral acetabular dysplasia and subluxation on the right side.

Fig. 3
AP radiograph of the hips of the patient when aged seven years. A varus rotation osteotomy has been performed. Postoperatively, severe heterotopic ossification developed medial to the osteotomy site. There is progressive subluxation of the left hip.
cessful without complications. To the authors’ knowledge, no previous report of a femoral or pelvic osteotomy in a patient with CSNA has been published.

Osteotomy in patients with neuropathy or CSNA is said to be associated with a high rate of pseudarthrosis. It is recommended that osteotomies should be protected until union has been verified radiologically. Our indication for surgical intervention in this patient was unilateral subluxation which was refractory to conservative measures. The subluxation recurred and a subsequent Salter-type pelvic osteotomy did not give improvement. Nonunion did not occur but severe heterotopic ossification and joint destruction developed. To our knowledge, this complication has not been described before in CSNA.

Surgical intervention (i.e., trauma) on the right hip may have precipitated the development of a neuropathic joint. The non-operated hip on the opposite side dislocated spontaneously but did not show comparable changes. Sensory neuropathy can be the cause of many surgical complications. The development of haematomas, heterotopic ossification, infection, Charcot-like joint destruction and pseudarthroses are associated with the underlying insensitivity to protective pain stimuli.

In our case, the joint destruction is thought to be typical of a Charcot joint since the radiological course showed simultaneous changes in the femoral head and the acetabulum, while osteonecrosis normally starts on the femoral side leading only secondarily to changes in the acetabulum.

Septic arthritis was thought to be highly unlikely in view of the normal white cell count and levels of C-reactive protein throughout the postoperative period.

Our experience shows that surgical improvement of the anatomy may not prevent progressive joint disease and that severe heterotopic ossification can complicate surgery in these patients. Surgical treatment of hip dysplasia and dislocation in patients with CSNA may not always be advisable.
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References