

## Case Reports

# Idiopathic Juvenile Osteoporosis

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### Abstract

Children with idiopathic juvenile osteoporosis typically presents in the prepubertal period with pain in the back and extremities, multiple fractures and radiological evidence of osteoporosis. Vertebral collapse and metaphyseal compression fractures of long bones are common. The diagnosis is made after excluding other causes of osteoporosis in this age group. The disease is self-limiting with spontaneous resolution after the onset of puberty. Management is directed at minimising permanent deformities. The clinical features, management and outcome of two cases of idiopathic juvenile osteoporosis were reported.

### Key words

Bone pain; Fractures; Juvenile; Osteoporosis

### Introduction

Idiopathic juvenile osteoporosis is an uncommon condition in children. Cardinal features include pain in the back and extremities, multiple fractures and radiological evidence of osteoporosis.<sup>1-4</sup> There is a wide range of severity. No consistent biochemical abnormality is found. Currently the etiology has not been fully elucidated. The diagnosis is made by exclusion of other causes of osteoporosis. The onset of the condition is usually in the prepubertal period between eight and fifteen years of age. Resolution occurs within one to four years after onset of puberty. We report two cases of idiopathic juvenile osteoporosis with the characteristic clinical and radiological features.

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### Case Reports

A ten-year-old girl presented with on and off pain over spine, dorsum of feet, achilles tendons, knees and ankles for three months. The child also complained of on and off ankle swelling, but mother only noticed one episode of ankle swelling. There was no morning stiffness, redness or hotness. During the painful episodes, the girl adopted a limping gait and stooping posture. Her daily activity was affected. The pain was precipitated by walking, prolonged standing or walking. The pain could not be relieved by paracetamol. One episode of ankle pain was relieved by analgesic, but another episode of left ankle and right knee pain required analgesic and steroid treatment by her family physician.

Examination including an ophthalmologic assessment showed no abnormality. The clinical features were compatible with idiopathic juvenile arthritis with enthesitis. However, her radiological examination showed generalised osteopenia. There was also a metaphyseal insufficiency fracture at the right tibia. Radiograph of spine showed compression fractures at multiple mid-thoracic vertebrae. There was no history of trauma. Further evaluation excluded osteogenesis imperfecta, homocystinuria, immobilisation, coeliac disease, Cushing's disease and Turner's syndrome. She had been on carbamazepine for epilepsy but her alkaline phosphatase was normal before the onset of pain.

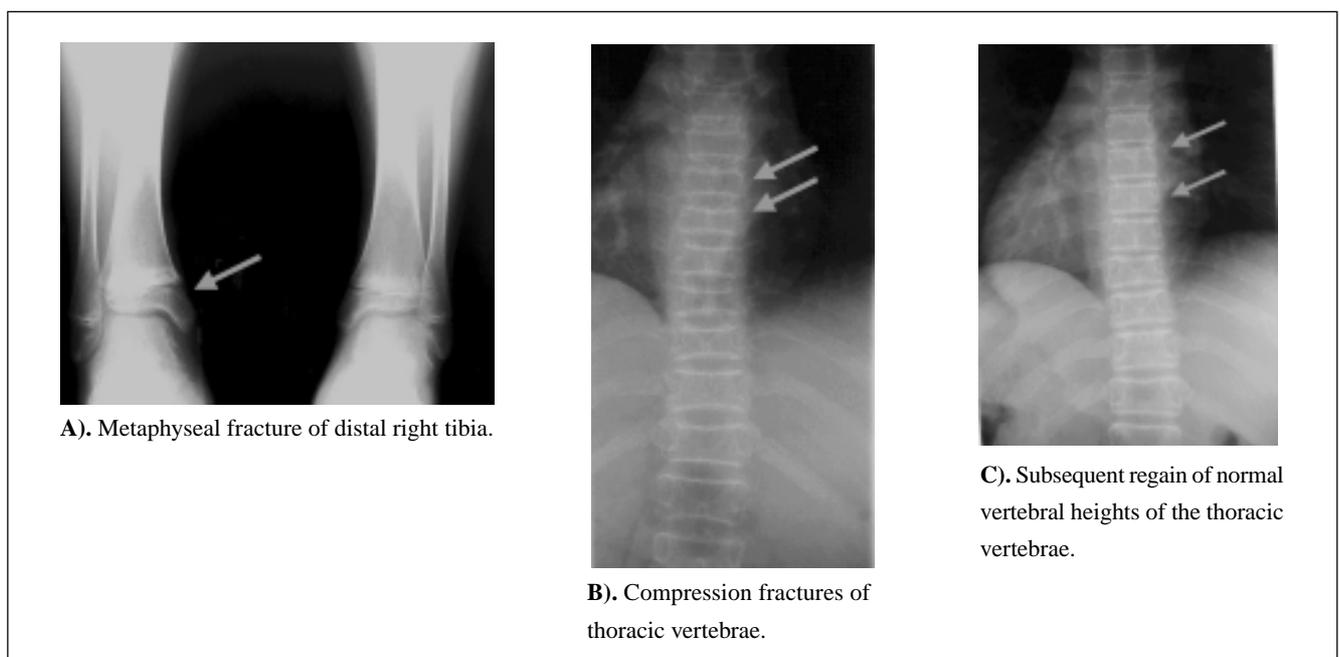
Investigations showed normal complete blood count, erythrocyte sedimentation rate, liver function, renal function, calcium, phosphate and glucose. Bone alkaline phosphatase after the onset of bone pain was 408 IU/L (normal <345 IU/L) and non-bone portion was 78 IU/L (normal <75 IU/L). Antinuclear factor and rheumatoid factor were negative. Twenty-four hours urinary calcium excretion was normal. Urine for metabolic screening including nitroprusside was negative. Thyroid function test, morning cortisol, follicular stimulating hormone, luteinizing hormone, estradiol and parathyroid hormone levels were normal. Bone age was 8 years 10 months to 10 years at a chronological age of 11 years. Idiopathic juvenile osteoporosis was diagnosed. Bone densitometry showed a z-score of -4.54, a level markedly below the fracture threshold. The girl was thus advised to limit vigorous physical activities to protect against fracture and to ensure an adequate intake of milk. She was closely monitored for kyphosis. The child had been followed up for five years since the onset of her bone pain. Her symptoms improved as she entered into puberty. Repeat spine radiograph two years later showed a regain of vertebral body height (Figure 1). Bone densitometry showed improving trend of bone mineral density (Figure 2).

The other child was a twelve years old boy presenting with sudden onset of low back pain after summersault.

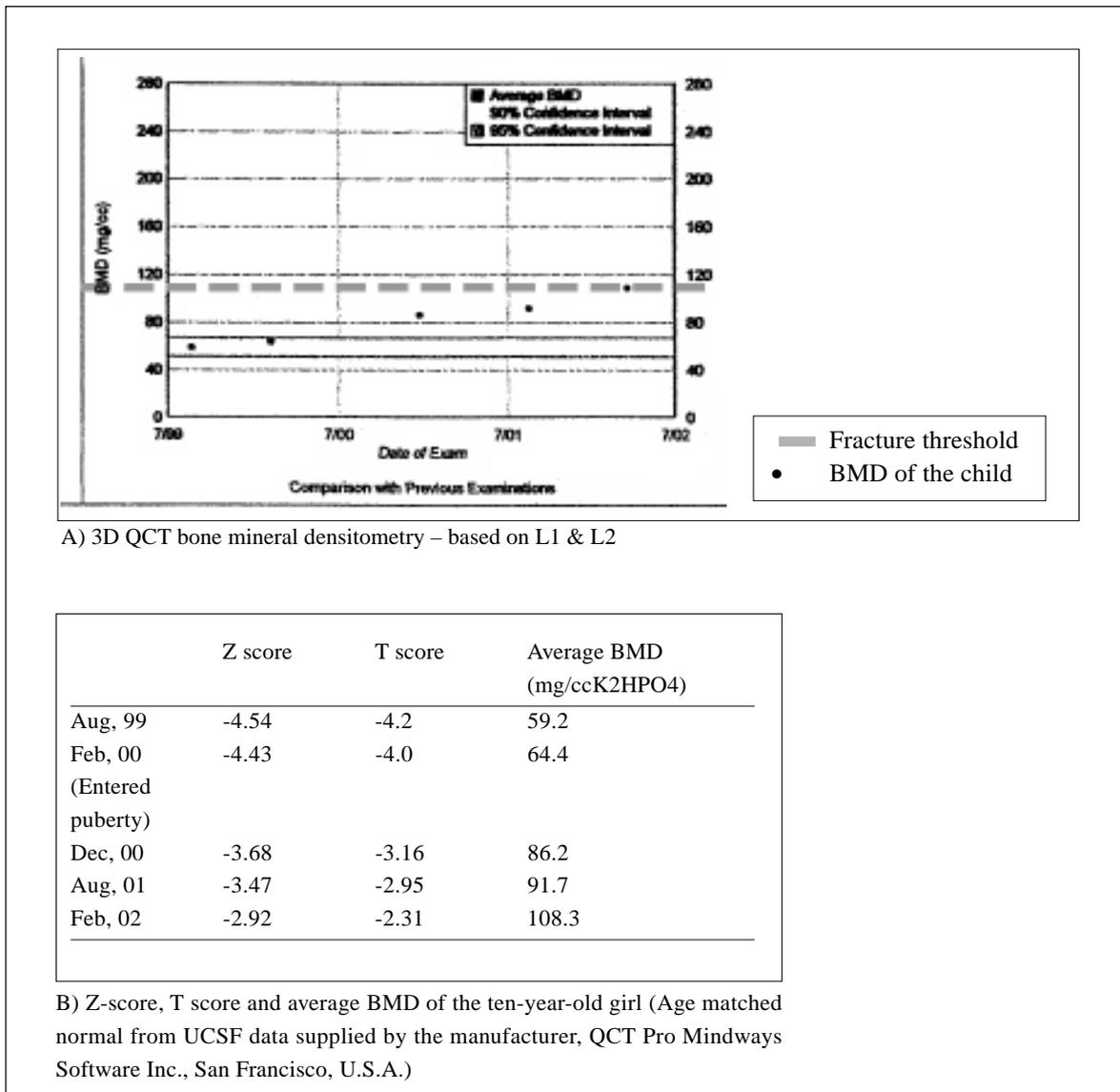
Radiological examination confirmed collapse of T9 and L2 vertebrae. In addition, there was generalised osteopenia. Examination showed that the child was prepubertal. Clinical evaluation ruled out other possible causes for the osteoporosis. Investigations including complete blood count, erythrocyte sedimentation rate, liver function, renal function, calcium, phosphate and alkaline phosphatase, thyroid function test, cortisol and parathyroid hormone were normal. Twenty-four hours urinary calcium excretion was normal. His bone densitometry showed a z-score of -5.1, markedly below the fracture threshold. He was treated with lumbar corset and physiotherapy. Dietary advice was given and the child was advised to avoid vigorous contact sports to prevent further injury. However, he was readmitted four months later for left knee pain after running. Radiological examination confirmed crack fracture over anterolateral aspect of left lateral femoral condyle. The child defaulted follow up after discharge from the hospital.

## Discussion

The classical presentation of pain in the back or extremities in idiopathic juvenile osteoporosis may be related to fractures in the vertebrae and long bones.<sup>1-4</sup> Pain



**Figure 1** Classical radiological findings in the ten-year-old girl.



**Figure 2** Bone densitometry of the ten-year-old girl.

may occur spontaneously or after minor trauma. Metaphyseal fracture at bones of the lower limb may cause difficulty in walking.<sup>1,4</sup> Multiple fractures in the long bones may occur as overt fractures, but more commonly as compression fractures. The axial skeleton is often involved resulting in wedge collapse, loss in vertebral height or biconcave vertebrae.<sup>1</sup> Osteoporosis should be confirmed radiologically by the presence of generalised osteopenia and by the decrease in bone mineral density in bone densitometry. Cortical discontinuities and transverse dense lines at the metaphyses of long bones resulting from compression fracture of the osteoporotic new bone at this

region (neo-osseous osteoporosis) are characteristic for this condition.<sup>2</sup>

Back pain, limb pain and difficulty in walking were present in our children. Their radiological examination also showed the typical abnormalities in the vertebrae and long bones. However it is important to exclude other causes of osteoporosis since idiopathic juvenile osteoporosis is a diagnosis by exclusion. Common differential diagnoses for osteoporosis in children include leukaemia, osteogenesis imperfecta, homocystinuria, immobilisation, coeliac disease, Cushing's disease and Turner's syndrome. Clinical evaluation often gives clues to most of these conditions

with the exception of acute leukaemia and mild forms of osteogenesis imperfecta. Acute leukaemia is common in children and back pain is not an uncommon presentation. However, subtle abnormalities such as leukocytosis, leucopenia or thrombocytopenia are often present in the complete blood counts.<sup>5,6</sup> In osteogenesis imperfecta, fractures in the long bones may occur but metaphyseal fractures are unusual.<sup>2</sup> The diagnosis of idiopathic juvenile osteoporosis was made in the children in our report after extensive investigation and follow up evaluation.

Controversies exist as to the effectiveness of treatment by calcium, vitamin D, biphosphonates, calcitonin and sex hormones.<sup>1,7-9</sup> Conservative expectancy with appropriate intake of calcium, prompt rehabilitation of fracture and avoidance of prolonged immobilisation is usually adequate. Although improvement is expected after the child enters into puberty, it is important to recognise idiopathic juvenile osteoporosis because of the risk of acquiring permanent deformity during the vulnerable period. Close monitoring of the spine and early use of orthotic brace may be necessary to prevent significant kyphosis.<sup>2</sup> However the girl in our report showed that milder compression fractures may be reversible with complete regain of height of vertebral bodies. Her bone densitometry also confirmed the improvement in bone mineral density as she entered into puberty.

Finally it is important to note that the new bone formation at the metaphyses of long bones is osteoporotic in idiopathic juvenile osteoporosis. Normal bone can be cortical or cancellous. Cortical bone (compact bone) makes up most of the skeleton and is composed of tightly packed osteons. On the other hand cancellous bone (spongy or trabecular bone) is less dense, has a higher turnover rate and undergoes more remodelling according to lines of stress. The abnormality in idiopathic juvenile osteoporosis has been

reported to be related to the impaired osteoblast performance at the cancellous bone (typically at the metaphyses of long bones) resulting in low bone volume, low bone formation rate and a propensity towards fracture.<sup>10</sup> Thus in contrary to the general advice of increasing weight bearing exercise to improve bone mineral density in children with osteoporosis, proper restriction of weight bearing exercise to avoid recurrent metaphyseal fracture and permanent deformity in idiopathic juvenile osteoporosis may be needed.

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