Wedge excision chondroplasty of the knee in dysplasia epiphysealis hemimelica—report of 2 cases

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Case 1

A 2-year-old Caucasian boy suffered from increasing genu valgum of the right leg. He was treated by temporary epiphyseodesis of the medial femoral condyle and the medial proximal tibia, using Blount's staples (Figure 1). The valgus deformity improved only by 5° over a period of 2 years and the staples were removed. Radiographs showed irregularly ossified tissue at the enlarged right medial femoral condyle and the right proximal medial tibia.

At the age of 6 1/2 years, he was referred to us for correction of his marked valgus deformity. Clinical examination showed moderate limitation of knee movement (extension –10° and flexion 135°), and no ligamentous instability. He had no metabolic disorder or an obvious systemic skeletal dysplasia that could account for his deformity. The irregularly ossified mass at the right femoral condyle had extended medially and distally, forming a large exostosis. A similar mass had grown in the proximal tibia (Figure 2). The classic radiographic appearance of an irregular, lobulated, osseous mass protruding from the epiphysis led to the diagnosis of dysplasia epiphysealis hemimelica (DEH).

Surgical treatment was indicated because of the marked valgus deformity. Via a medial longitudinal approach, we divided the medial collateral liga-

Figure 1. Case 1. Treatment of the valgus deformity by temporary epiphyseodesis with Blount's staples (at the age of 3 years, i.e., 1 year after surgery).

Figure 2. The irregularly ossified tissue at the femoral condyle has increased significantly. There are similar changes in the medial tibia.

Figure 3. Surgical procedure.

a. After division of the medial collateral ligament, we performed a wedge-type excision of the lesion.

b. This was followed by a direct suture of the adjacent cartilage.

ment near its proximal insertion. The cartilage covering the enlarged femoral condyle was thin and bluish. We decided to leave the articular cartilage untouched. An incision was made on the medial side of the condyle about 2 cm distal to the growth plate. The enlargement was resected in a manner resembling a closing wedge osteotomy, and the defect was closed by direct suture of the adjacent cartilage (Figure 3). The wedge excision had to be modified slightly to prevent a "dog-ear" deformity. The medial collateral ligament was reinserted during shortening. A plaster cast was applied for 6 weeks. 1 year after surgery the radiographs showed a residual enlargement and irregularity of the medial femoral condyle. We found only a minor valgus deformity. After 4 1/2 years, the boy had an almost normal alignment and moderate restriction of motion in the operated right knee (0° extension and 110° flexion in the right knee and 5° extension and 150° flexion in the left). Radiographs showed more remodeling of the medial condyle (Figure 4). There was a leg-length inequality of 1.5 cm with lengthening of the affected leg. The boy had no pain and was very satisfied.

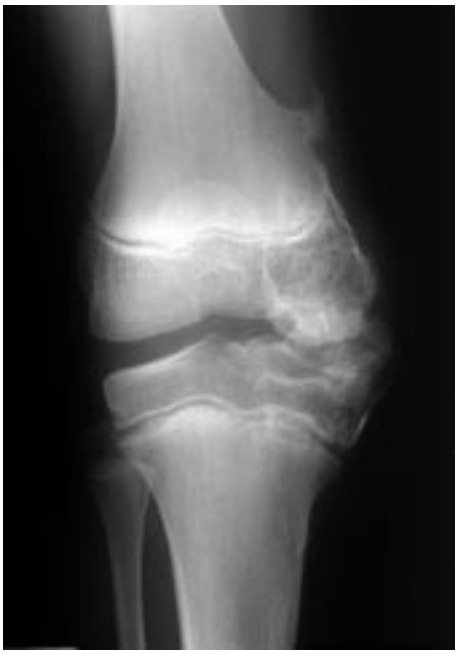


Figure 4. Outcome 4 1/2 years after surgery.

Case 2

A 4-year-old Caucasian boy was referred to us with the diagnosis of DEH. He presented with an irregularly ossified mass in the left medial femoral condyle causing a valgus deformity of about 25°. No other foci of the disease were found. He was also treated by wedge excision chondroplasty of his medial femoral condyle, which almost completely corrected the deformity (Figure 5). Radiographs showed a good reduction of the enlarged condyle (Figure 6).

4 years later, we noted a valgus deformity of 5° in the left knee, a normal range of movement of 5° extension and 150° flexion, and a slight leg-length discrepancy, with the affected side 1 cm

Figure 5. Case 2.

a. The preoperative radiograph shows a valgus deformity of the left leg of about 25°. Only slight valgus of the right leg is present.

b. Good correction of the deformity after surgery.

longer. No ligamentous instability was detected. The radiographic appearance of the left knee was almost normal (Figure 6).

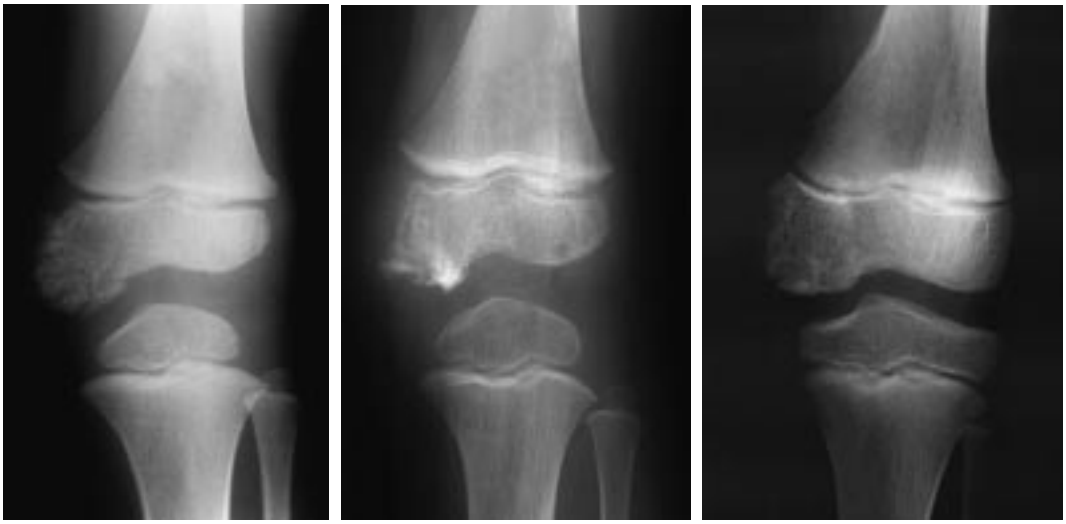
Discussion

Dysplasia epiphysealis hemimelica, also known as Trevor's disease, is a rare skeletal disorder with abnormal growth of bony tissue mainly in the epiphyses of long bones. Trevor (1950) described 8 children, whose knee or ankle joint was most often involved. He believed that the lesion was congenital and that the etiology was related to damage during the formation of the limb bud. Fairbank (1956) reviewed Trevor's cases, added 14 of his own, and coined the term dysplasia epiphysialis hemimelica to describe the findings better. He believed the process to be "a true dysplasia or faulty growth of part of the epiphysis itself". So far as we know, the first case was described by Mouchet and Belot (1926).

The index lesion is intra-articular and is usually confined to only half of a single limb (hemimelic), either medial or lateral. There has been a report, however, of asymmetrical involvement (Heilbronner 1988). Initially the lesion is solitary, although a portion may break off to form an intra-articular body (Fasting and Bjerkreim 1976, Kuo et al. 1998). It is typically found in the joints of the lower extremity, especially the medial femoral condyle, the distal aspect of the tibia, and the talus (Trevor 1950, Fairbank 1956, Kettelkamp et al. 1966, Fasting and Bjerkreim 1976, Azouz et al. 1985, Graves et al. 1991, Kuo et al 1998). The osseous portions make it easy to distinguish it from synovial chondromatosis, which always consists of multiple lesions usually dispersed throughout the joint.

The histological appearance, a well-defined cartilage cap over projecting bone that is continuous with the underlying normal bone, is indistinguishable from that of an osteochondroma (Fairbank 1956, Kettelkamp et al. 1966, Graves et al. 1991, Takagi et al. 2000).

Figure 6. Case 2. Note the reduction of the ossified mass and the gradual remodeling of the medial femoral condyle.



a. Before surgery. b. After 2 months. c. After 4 years.

The radiographic aspect has been well described in many reports (Trevor 1950, Fairbank 1956, Kettelkamp et al. 1966, Fasting and Bjerkreim 1976, Azouz et al. 1985, Keret et al. 1992, Kuo et al. 1998). The first characteristic finding is the development of secondary ossification centers in the epiphysis. The lesion can not be seen on plain radiographs, if ossification has not begun. Therefore, in "idiopathic" cases of increasing axial deformity, MR imaging is indicated. We used MRI routinely for evaluation in both of our cases. This greatly helps detection of the site and extent of the osteocartilaginous overgrowth (Keret et al. 1992, Kuo et al. 1998). However, these findings were not of value for our decision to perform the new surgical procedure.

In case 1, careful examination of the radiographs would have enabled to make the correct diagnosis.

The treatment of dysplasia epiphysealis hemimelica is disputed and it depends on the location of the lesion. According to the literature, simple surgical excision of the articular epiphyseal enlargement, including the joint cartilage, has given satisfactory results (Trevor 1950, Fairbank 1956, Kettelkamp et al. 1966, Fasting and Bjerkreim 1976). Keret et al. (1992) and Kuo et al. (1998) recommended that juxtaarticular lesions should be excised. They cautioned against resection of articular lesions, which may further compromise the joint and predispose to degenerative joint disease. In the area of the knee, a supracondylar osteotomy may be necessary to correct (residual) genu valgum or varum (Trevor 1950, Fairbank 1956, Keret et al. 1992). In a few cases, an arthrodesis or epiphyseodesis had to be performed (Trevor 1950, Kettelkamp et al. 1966).

We obtained a more than satisfactory result by lateral wedge excision and closure of the defect by direct suture. Thus, correction of the axial deformity was achieved with preservation of the surface of the articular cartilage—reducing the risk of late osteoarthritis. We think that early surgery is preferable because of the body's better remolding capacity and agree with Fasting and Bjerkreim (1976) who suggest surgery before 4 years of age.

No competing interests declared.

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