Chondrodiatasis in a patient with spondyloepimetaphyseal dysplasia using the Ilizarov technique: successful correction of an angular deformity with ensuing ossification of a large metaphyseal lesion

A case report

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Accepted: 26 March 1998

Summary. *Distraction through the physis (chondrodiatasis) is a controversial technique with unpredictable results. However, it has been used in the past for the lengthening and correction of angular deformities of long bones. We report the case of an 11-year-old patient with spondyloepimetaphyseal dysplasia (SEMD) who presented with a severe recurvatum deformity of the left proximal tibia secondary to collapse of the tibial plateau into a large metaphyseal cystic lesion. Using the chondrodiatasis technique with a percutaneously applied Ilizarov circular frame, we were able to correct this deformity. Surprisingly, healing and ossification of the metaphyseal lesion was simultaneously observed at the end of the treatment, a finding which, to the best of our knowledge, has not been previously reported.*

Résumé. *La distraction physaire (chondrodiatasis) est une technique controversée qui a été néanmoins utilisée pour l'allongement et la correction des déformations angulaires des os longs. Le cas présenté ici est celui d'un garçon d'onze ans atteint de dysplasie spondylo-épi-métaphysaire (S.E.M.) qui s'est présentée avec un genu recurvatum sévère au niveau d'une grande lésion kystique métaphysaire du tibia proximal. La correction de la déformation fut faite au moyen d'un fixateur externe d'Ilizarov permettant une distraction physaire progressive. La correction s'est accompagnée d'une disparition complète de la lésion dysplasique, par ossification progressive. A notre connaissance, cette observation n'a pas été rapportée auparavant.*

Case history

The patient was born at term following a normal pregnancy. His family history was unremarkable, and all his anthropometric parameters at birth were normal, except for a height of 43 cm (below 3rd percentile). Psychomotor development was adequate for his age. Chromosomal and metabolic studies were normal. By the age of 5 years, extensive involvement of the proximal femora was evident, and the diagnosis of spondyloepimetaphyseal dysplasia was made. He underwent bilateral valgus derotational osteotomies of the proximal femur for the correction of severe coxa vara deformities.

At 11 years of age he developed a progressive left genu recurvatum deformity of 30° (Fig. 1). Radiographs showed anterior collapse of the tibial plateau into a large, cystic radiolucent lesion of the proximal tibial metaphysis which lay adjacent to the physis and measured 3.0×3.2×1.2 cm (Fig. 2). It was decided to correct the deformity by slow, controlled distraction through the physis. An Ilizarov external fixator was percutaneously applied to the left tibia, and no osteotomy was performed. One ring was secured to the proximal tibial epiphysis oriented in the plane of the tibial plateau. The distal rings were secured to the proximal tibial shaft and then crosslinked. The proximal ring was connected to the distal ones by three hinged distracting rods, two anterior and one posterior. The distraction rate did not exceed 0.5 mm/day (Figs. 3 and 4). A simultaneous lengthening of 3 cm was performed to compensate for the expected growth arrest of the proximal tibial metaphysis following the physeal distraction. The fixator was removed 10 months after its application. The patient then remained in a cast for a period of 6 weeks followed by a brace for an additional 9 weeks. The only complication that occurred during treatment was a pin-tract infection which was successfully treated with oral antibiotics. Clinical and radiologic examination at the end of treatment and 8 months after showed complete correction of the angular deformity. The patient maintained a full range of motion of the affected knee. Surprisingly, the large metaphyseal cystic lesion ossified completely. At the latest follow-up, 3 years after the end of treatment, the patient was functioning well. The 3-cm lengthening performed was judicious, as there was no limb-length discrepancy at the end of growth (Figs. 5 and 6).

Fig. 1. Clinical picture showing the severity of the left genu recurvatum deformity prior to treatment

Fig. 2. Tomogram of the left proximal tibial metaphysis. The cystic lesion and the collapse of the tibial plateau can be appreciated

Fig. 3. Lateral radiograph of the left tibia with the Ilizarov frame at the beginning of treatment

Fig. 4. Lateral radiograph of the left tibia with the Ilizarov frame at the end of correction

Fig. 5. Lateral radiograph of the left tibia 3 years after the end of treatment

Fig. 6. Clinical picture of the patient's lower limbs 3 years after the end of treatment

Discussion

The spondyloepimetaphyseal dysplasias (SEMD) constitute a rare and complex group of disorders characterized by anomalies of the vertebrae and a dis-

turbance in the ossification of the metaphyses and epiphyses of the tubular bones. Histopathological studies have demonstrated a fundamental disturbance in growth-plate development [4, 22]. These disorders often manifest themselves in early childhood by ab-

normal height and gradual development of angular and rotational deformities that are often disabling. Traditionally, these deformities have been addressed by conventional osteotomies. However, with the introduction in the Western world in the 1980s of the Ilizarov methods and principles of distraction osteosynthesis, many centres today are using this technique in the management of complex limb deformities associated with skeletal dysplasias [6, 8, 9, 17, 24].

While the application of the Ilizarov technique of gradual and slow distraction through an osteotomy in the metaphysis or diaphysis is a well-accepted method, the use of this technique for slow (0.5 mm/day) distraction through the physis, chondrodiatasis, remains very controversial. Originally described by De Bastiani et al., this technique was developed in an attempt to prevent the premature epiphysiodesis observed with rapid distraction rates [3, 11, 14, 18, 23].

The mechanism by which chondrodiatasis lengthens the physis, as well as the short- and long-term effects on its physiological activity, has been extensively studied in animal experiments. De Bastiani speculated that the elongation in the hypertrophic zone was due to cellular hyperplasia and a higher turnover rate of cellular components in rabbits [10, 11]. However, Elmer et al. suggested that elongation was due to passive stretching of the matrix rather than by stimulating cell division or synthetic function [13]. Using a sheep model, de Pablos and Canadell reported that regardless of the rate of distraction, the basic mechanism of lengthening appeared to be the production of a physal fracture between the degenerative and calcified layers [12]. De Pablos also noted that the lower the rate of distraction employed, the greater was the short- and long-term viability of the physis. Other reports have confirmed de Pablos' observations [1, 2, 5, 19]. In addition, these studies have shown that the apparent thickening of the physis with distraction is caused by a decrease in the enchondral ossification rate with an ensuing accumulation and disorganization of hypertrophic chondrocytes. With regard to the residual activity of the physis after distraction in animal models, the literature remains controversial [1, 5, 21]. All these seemingly conflicting reports reflect the inherent difficulty in predicting growth-plate behaviour after chondrodiatasis in experimental models. They appear to suggest that a single rate of distraction may not be appropriate for all situations.

In the clinical setting, a significant number of complications, including early growth arrest, have been reported with the use of this method [7, 15]. At the present time, the use of chondrodiatasis can only be recommended in children nearing skeletal maturity. Added caution seems in order when applying this technique to an already abnormal physis.

In our case, chondrodiatasis was successfully used for the correction of an angular deformity in a patient with SEMD. The effect of the expected early closure of the proximal tibial physis was counterbalanced by the simultaneous lengthening of the tibia by 3 cm, an

amount estimated to be equal to that of the remaining growth. In addition, and to our great surprise, chondrodiatasis led to the healing and complete ossification of the proximal metaphyseal cystic lesion that was the cause of the deformity. To the best of our knowledge, this has not been previously reported for such a large cystic lesion. We can only speculate that distraction through the matrix of this cyst stimulated its ossification. Moreover, we do not know the long-term effects of distraction through abnormal bone and cartilage [20]. Only one case of osteogenic sarcoma originating in bony regenerate in a patient with fibrous dysplasia has been reported [16].

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