

Dysplasia spondyloepiphysaria and patella dislocation: a case followed over 10 years

Michael Memminger

Orthopaedic Department, Hospital S. Valentino Montebelluna (Treviso)

Summary. Dysplasia spondyloepiphysaria means altered form and seizure of the vertebral bones and the epiphyseal bone regions. Pathologies related to this disease are: scoliosis, short stature, eye problems, articular deformities. We report a case of tarda form. The main problem was habitual and later fixed dislocation of both patellae together with valgus deformities of the knees. We describe the surgical procedures to gain reduced patellae and correction of the mechanical axis. (www.actabiomedica.it)

Key words: spondyloepiphysal dysplasia, patella dislocation, femur osteotomy, hemiepiphysodesis

Introduction

Spondyloepiphysal dysplasia is a rare disorder with particular deformities of the vertebral bones and epiphysal segments. There are two variants: congenita and tarda form. They differ in the inheritance pattern and the age of onset (4). The patient described in this report suffered from the tarda form. The clinical aspects of the tarda form are: manifestation from 6-12 years, short stature (height <160 cm), short neck, enlarged chest, short arms, scoliosis, kyphosis, lumbar hyperlordosis, reduced hip function, early osteoarthritis of hip and knee. Severe myopia is common (about 50%). About one quarter of people with this condition have hearing loss (1-4).

Case description

Boy born in 2000; at 7 years he started treatment with Cheneau brace for scoliotic deformity and gained good clinical results during the following years. He has short stature and severe myopia. From age 7-10 he presented lateralisation of the patellas with concomitant valgus deformity of both knees. The x-ray at 10

years showed a markedly lateralised right patella (figure 1). During the following 3 years the patellae dislocated completely (figure 2).

At age 13 we applied eight-plates for temporary hemiepiphysodesis on the medial femoral and tibial side of the knees to correct the valgus deformity. The main deformity was on the femoral side, but we applied the plates on the tibial side too, because we wanted to reach faster correction of the main axis accepting an initially oblique joint line. The purpose was to remove the tibial plates former to achieve a correct joint line regarding the mechanical axis. Contemporarily we did a lateral release, a duplication of the medial joint capsule and distalization of the vastus medialis (Madigan procedure). At the end of the procedure the patellae were correctly reduced (figure 3,4). During the following year the mechanical axis improved, was very good on the left side but remained still in valgus on the right side. The patellae were reduced, but the right has tendency to lateralize. After 1,5 years from the first surgical procedure we removed the tibial eight-plates and re-positioned the right femoral plate (figure 4). During the following 6 months the boy had perfect function on the left knee, but subsequently the right patella dislocated and he complained permanent pain.



Figure 1. Age 10 years: valgus of the knee and subluxation of the right patella

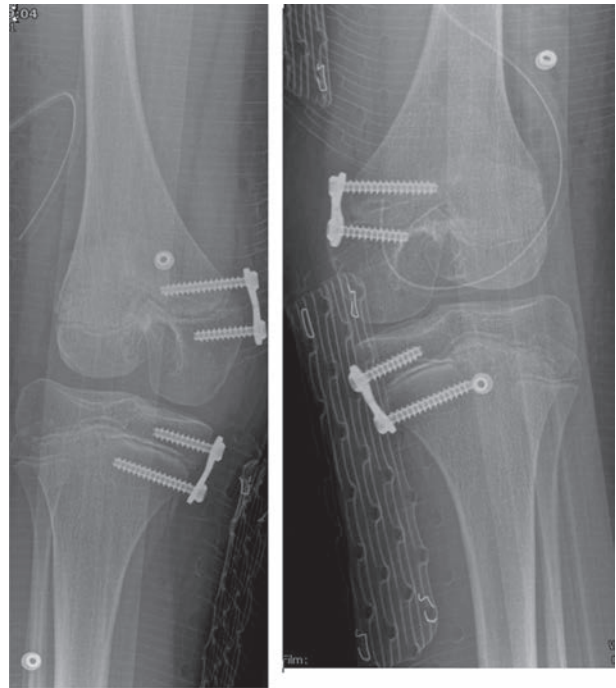


Figure 3. Plates on both knees for temporary hemiepiphysodesis, lateral release and medial capsule duplication con distalization of the vastus medialis (Madigan) and reduction of the patellae. Age 13 years

It was mandatory for us to get a perfectly reduced patella also on the right knee. So at age 15 we re-operated the boy. The procedure was: lateral release of the patella, correction of knee axis with femoral supracondylar varus osteotomy. The patella still remained in a slightly lateral position. So we medialized the tuber-

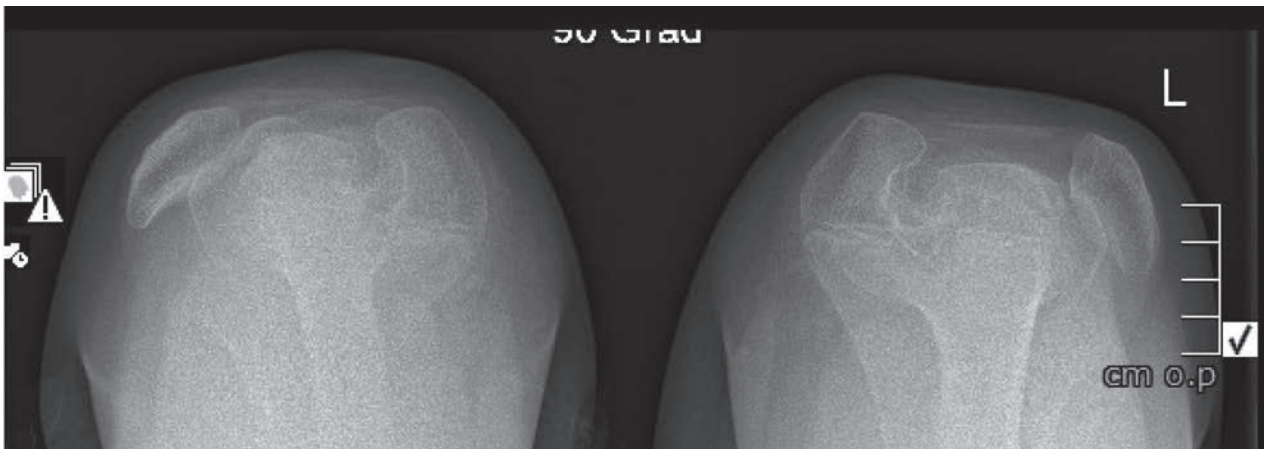


Figure 2. Both patellae are dislocated laterally (age 13 years)

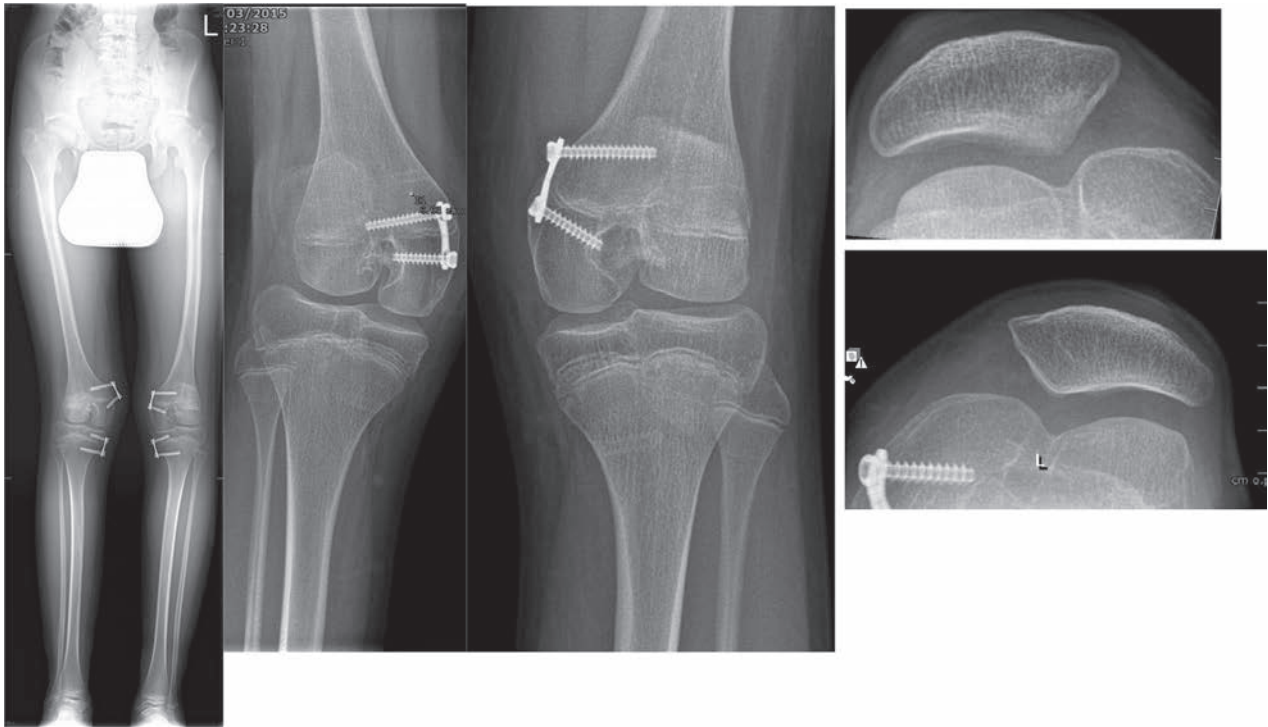


Figure 4. After 1,5 years (age 14,5 years) with hemiepiphysodesis the mechanical axis improved, we removed the tibial eight plates and re-positioned the right femoral eight-plate. Good clinical function, but right patella has still tendency to lateralize

ositas tibiae until the patella movement in extension-flexion was perfectly aligned (figure 5). Clinically the boy had a perfectly centered patella also on the right knee, persistence of light extension deficit and absence of pain. During the last year of growth the boy developed variation of the right knee (figure 6) and at age 16,5 (closed growth plates) we corrected definitely the deformity with femoral valgus and extension osteotomy (3.5 plate) see figure 7.

At 17 years the boy presented a complete knee function, stable knees (mediolateral) and absence of pain. The patellae were perfectly aligned without any tendency to lateralize or medialize during knee function (figure 8).

Discussion

We presented a boy with spondyloepiphyseal dysplasia. The scoliosis was completely controlled by a Cheneau brace. The most difficulties we encountered

were a progressive valgisation of the knees with patella dislocation. On the left knee a soft tissue procedure (modified Madigan) with temporary hemiepiphysodesis produced a correctly aligned knee with reduced patella.

On the right knee the soft tissue correction and growing correction through the hemiepiphysodesis wasn't sufficient to maintain the patella reduced and to correct the valgus. When we tried to re-position the plate on the medial right femur the screw hasn't enough stability in the small condyle, so we corrected the axis by an varus osteotomy of the distal femur. The pathologic traction of the patella tendon pulled the patella still lateral with the risk of dislocation. The boy was 15 years old and with open growing plates: there is contraindication for osteotomy of the tuberositas tibiae. Partial tendon detachment wasn't expected to gain satisfactory medial traction, full detachment would had quite the same damage to the tuberositas tibiae and knee like the osteotomy. Therefore we executed medial sliding osteotomy of the tuberositas tibiae.



Figure 5. Femoral varus osteotomy with 3.5 plate. Medialisation of the tuberositas tibiae (age 15 years)

This produced a stable and correctly aligned patella. Afterwards the boy hasn't anymore patella dislocations and was painfree. However in the remaining year of growth the knee developed a varus deformity and with closed growing plates we brought the knee axis to a more physiologic value by supracondylar valgisation and extension osteotomy. We associated extension correction because the boy had still some degrees of extension deficit on the right knee, not fully corrected after the patella alignment. The clinical results were excellent: full range of movement of the right and left knee, absence of pain, walking without limping. Fortunately the tuberositas osteotomy didn't produce any negative aspect.

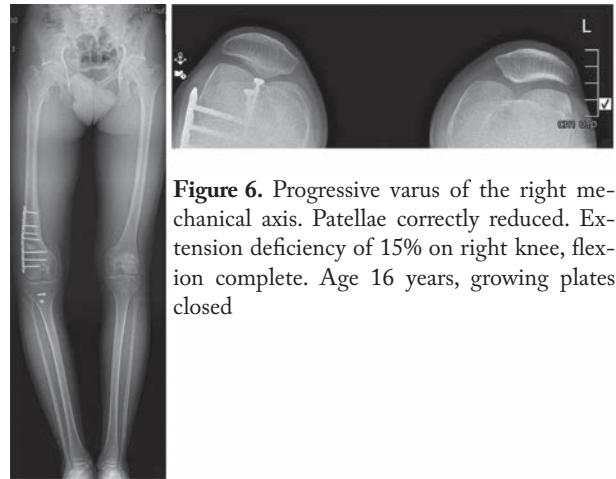


Figure 6. Progressive varus of the right mechanical axis. Patellae correctly reduced. Extension deficiency of 15% on right knee, flexion complete. Age 16 years, growing plates closed

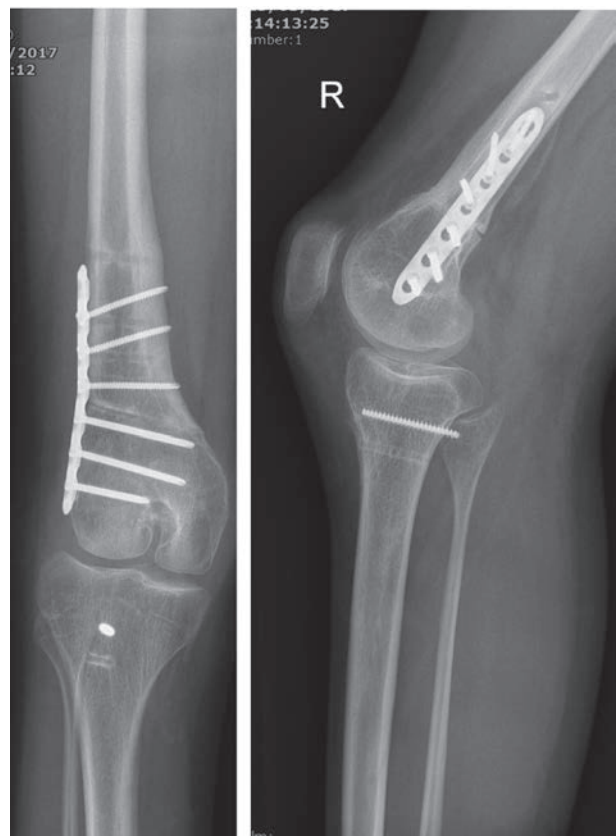


Figure 7. Femoral valgus and extension osteotomy. Age 16,5 years

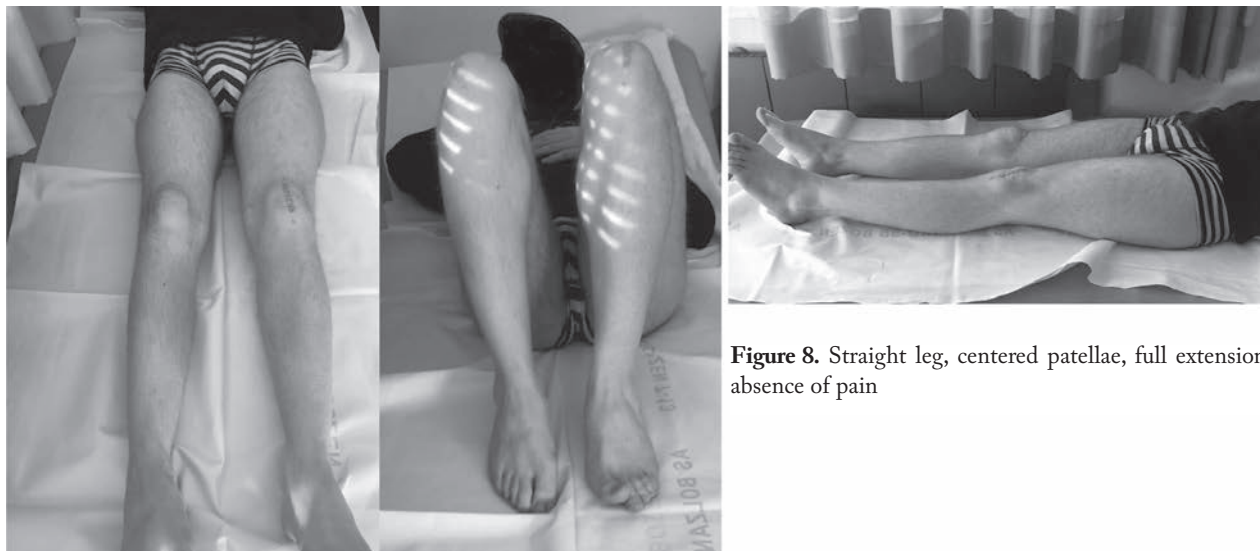


Figure 8. Straight leg, centered patellae, full extension, absence of pain

Conclusion

The patella dislocation and valgus deformity in this case of spondyloepiphyseal dysplasia required on the left knee a soft tissue reconstruction and simple hemiepiphysodesis. The right knee was completely different: soft tissue procedures and hemiepiphysodesis weren't sufficient and bony procedures were necessary. The osteotomy of the tuberositas tibiae produced a stable and correctly aligned patella.

Conflict of interest: Each author declares that he or she has no commercial associations (e.g. consultancies, stock ownership, equity interest, patent/licensing arrangement etc.) that might pose a conflict of interest in connection with the submitted article

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Correspondence:

Michael Memminger MD

Orthopaedic Department

Hospital S. Valentino Montebelluna (Treviso)

E-mail: memminger@hotmail.com