Management of joint dislocations of the lower limb in Larsen syndrome: practical approach

HE Matar, NK Garg

Alder Hey Children's Hospital, UK

ABSTRACT

INTRODUCTION Larsen syndrome is an autosomal-dominant osteochondrodysplasia characterised by large joint dislocations and craniofacial anomalies.

CASE HISTORY We present a rare case of Larsen syndrome with bilateral dislocated hips and knees and severe clubfeet at 7-year follow-up. We undertook bilateral open reduction of both hips at age 8 months. This procedure was preceded by open reduction and left-knee V–Y quadricepsplasty at age 4 months following a failed trial of closed reduction of the left knee. Both feet had a severe deformity (Pirani score of 5.5 and 6.0) and were treated using the Ponseti method, but the left foot relapsed at 24 months and required posteromedial release.

CONCLUSIONS We tried to address the difficult questions on the timing and sequence of surgical interventions by treating clubfeet and dislocated knees early using Ponseti casts which included the knees. Open reduction of hips was done later, and further interventions were guided by functional needs.

KEYWORDS

Larsen syndrome – Congenital knee dislocation – V–Y Quadricepsplasty – Ponseti casts

Accepted 29 February 2016

CORRESPONDENCE TO

Hosam Matar, E: hematar@doctors.org.uk

Described first in 1950,¹ Larsen syndrome (LS) is an autosomal-dominant osteochondrodysplasia characterised by large-joint dislocations and craniofacial anomalies. Mutations in the Filamin B gene have been implicated. We present a case of LS with bilateral dislocated hips and knees and severe clubfeet at 7-year follow-up.

Case History

A child with LS had been under our care for >7 years. Her father is known to have LS and she had been suspected to have LS by week-20 of gestation. She was born by Caesarean section at 39 weeks of gestation with a birth weight of 8lb 11oz, and was the first child in the family. She was found to have abnormalities in the joints of the lower limbs.

When first seen in our clinic aged 10 weeks, she appeared to have some dysmorphic facial features but upper limbs appeared normal. She had bilateral dislocated hips, bilateral dislocated knees and severe bilateral clubfeet. She had no other musculoskeletal involvements. Cardiovascular examination was normal. Subsequent genetic analyses confirmed the diagnosis of LS.

Bilateral hip dislocations

Both hips were clinically irreducible (Fig 1a), so a trial of a Pavlik harness was contraindicated. We proceeded with bilateral open medial reduction at age 8 months once the

dislocated knees had been managed. This strategy was followed by a hip spica cast for 6 months. Serial follow-up (Fig 1b, c) showed concentrically reduced hips that developed normally with symmetrical abduction to >60° at 7-year follow-up.

Bilateral knee dislocations

Initial examination revealed a left knee that was dislocated in extension with no flexion possible. The right knee also dislocated in extension but could be reduced in flexion. A trial of serial casting to the left knee was attempted but was unsuccessful. We proceeded with open reduction of the left knee and a V–Y quadricepsplasty (Fig 2) at age 4 months.² Outcome was successful and uneventful at 7 years with an arc of movement of ≈100–110° (Fig 3).

She continued to progress and achieved her milestones of development. The right knee, however, continued to undergo subluxation at will but, remarkably, she managed to walk unrestricted. Magnetic resonance imaging of the right knee (Fig 4) demonstrated hamstring tendons that did not appear to dislocate, a posterior cruciate ligament that was present but abnormal, and an absent anterior cruciate ligament with anterior subluxation of the tibia. The intracondylar notch was not well developed. Therefore, future reconstruction may be indicated, particularly of the anterior cruciate ligament to control anterior subluxation of the tibia.

Figure 1 a: Anteroposterior radiograph of the pelvis demonstrating bilateral dislocated hips at age 8 months. b: Follow-up anteroposterior radiograph of both hips at age 17 months demonstrating reduced bilateral hips with well-developed ossific nuclei after bilateral open medial reduction at age 10 months. c: Anteroposterior radiograph of the pelvis demonstrating normally developing hips at 5-year follow-up.

Figure 2 a: Anteroposterior plain radiographs of both dislocated knees at age 2 months. The right knee was clinically reducible but the left knee was clinically irreducible (fixed in extension). b: Lateral plain radiograph of the right knee and oblique radiograph of the left knee at age 2 months. c: Anteroposterior and lateral plain radiographs of the left knee 6 months after open reduction and V-Y quadricepsplasty.

Figure 3 a: Anteroposterior plain radiographs of both knees at age 2 years. The range of motion of the left knee upon flexion was $>100^\circ$. The right knee continued to undergo subluxation. b: Lateral radiographs of both knees at age 2 years.

Bilateral syndromic congenital talipes equinovarus (CTEV)

Initial assessment (at age 10 weeks) using the Pirani score revealed a more severe score for the left foot (6.0) than for the right (5.5). Treatment commenced using the Ponseti method.⁵ She required casts every 7 weeks followed by bilateral tendoachilles tenotomies, and then the wearing of orthotics as per the Ponseti protocol.

At 12-month follow-up when she started walking, dorsiflexion of the right foot was 25° and abduction 45° , whereas those of the left foot were 15° and 50° , respectively. The right foot also had a mild deformity of the metatarsus adductus that was passively correctable to valgus.

Figure 4 Magnetic resonance images of the right knee demonstrating anterior subluxation of the tibia and an absent anterior cruciate ligament.

Figure 5 Clinical photographs of both knees and feet at 7-year follow-up.

At 24-month follow-up, the left foot had relapsed into an equinovarus deformity in a fixed equinus of 10° . The right foot had developed a skewed foot deformity with metatarsus adductus and hind foot valgus, but was quite flexible and highly functional. She underwent a left posteromedial soft-tissue release. Outcome at 7-year follow-up was satisfactory with dorsiflexion of 15° and abduction of 40° in the left foot, although she has mild residual metatarsus adductus but walks with full weight-bearing with both feet in plantigrade (Fig 5).

Discussion

Only a few studies have addressed the difficult questions of the timing and sequence of surgical interventions for LS management.⁴

Our patient presented first at age 10 weeks. We commenced management of talipes equinovarus using the Ponseti method, which carries a high risk of recurrence if used for LS-associated talipes.⁵ This strategy also allowed inclusion of the dislocated knees in casts as an initial attempt at closed reduction. Once serial casting had been completed, bilateral percutaneous tendoachilles were done with bilateral hip arthrography. This approach permitted

planning for an open medial reduction of both hips once the dislocated knees had been managed.

Congenital knee dislocations are managed conservatively in the first instance with closed reduction and casting. If congenital knee dislocations are associated with a hip dislocation, the knee should be treated first to facilitate use of a harness or hip spica cast when treating the dislocated hip. However, failure to achieve progress warrants surgical reduction and soft-tissue release with the goal of obtaining: 90° of flexion with a V-Y quadricepsplasty or Z-lengthening; release of the anterior joint capsule; posterior transposition of the hamstring tendons; mobilisation of collateral ligaments. The left knee was not amenable to closed reduction, so we undertook open reduction, soft-tissue release and V-Y quadricepsplasty. The right knee, however, remains unaddressed but highly functional. Surgical reconstruction of the right anterior cruciate ligament may be necessary, but her functional demands/needs will dictate further surgical intervention.

Acknowledgements

We thank Peter Beirne (senior physiotherapist), Dave Simms (appliance officer) as well as John Franklin, Julie Mottaram, Val Domnax and Anthony Holden (plaster technicians) at Alder Hey Children's Hospital.

References

1. Larsen LJ, Schottstaedt ER, Bost FC. Multiple congenital dislocations associated with characteristic facial abnormality. *J Pediatr* 1950; **37**: 574–581.
2. Curtis BH, Fisher RL. Congenital hyperextension with anterior subluxation of the knee. Surgical treatment and long-term observations. *J Bone Joint Surg Am* 1969; **51**: 255–269.
3. Morcuende JA, Dolan LA, Dietz FR, Ponseti IV. Radical reduction in the rate of extensive corrective surgery for clubfoot using the Ponseti method. *Pediatrics* 2004; **113**: 376–380.
4. Rodríguez A, Asenjo B, Domínguez R, Lemaire R. Larsen syndrome: multicenter study of 12 new cases. Diagnosis, planning and results of treatment. *Acta Orthop Belg* 1994; **60**: 259–273.
5. Janicki JA, Narayanan UG, Harvey B *et al*. Treatment of neuromuscular and syndrome-associated (nonidiopathic) clubfeet using the Ponseti method. *J Pediatr Orthop* 2009; **29**: 393–397.