Introduction

Popliteal pterygium syndrome is a rare cause of congenital childhood flexion contracture. The association of joint stiffening and posterior soft parts retraction with a cutaneous
and muscular popliteal band raises specific treatment difficulties [1,2]. After Trélat’s initial description in 1869 [3], Gorlin was the first to introduce the term popliteal pterygium syndrome, in 1968 [4]. There are only a hundred or so cases reported in the literature [5], prevalence being estimated at around 1 per 300,000 births.

The term pterygium comes from the Greek for “wing”, and refers to the web-like posterior popliteal cutaneous and muscular band. Clinically, phenotypic expression is varied [6]. Other occasional associated symptoms are: filiform ankyloblepharon, hair implantation abnormality, inguinal hernia, perigenital hyperpigmentation, gum frenula, hypospadias and micrognathia. The typical three-fold syndrome found at birth comprises popliteal deformity, cleft palate (in 75% of cases) with or without cleft lip, and genital abnormality (in 50% of cases): cryptorchidism, dual scrotum, or labia majora hypoplasia.

The shortness of the sciatic nerve and the fragility of the posterior skin represent the two main difficulties encountered in knee realignment surgery. In limb deformity, the Ilizarov method provides progressive correction, suppling the periarticular structures, and thus seems well-suited for the management of the joint stiffness associated with popliteal pterygium. We here report our experience with the technique, in eight children presenting with popliteal pterygium implicated in severe knee flexion contracture.

Patients and method

Medical records of eight children, successively treated between 1986 and 2007, were reviewed. Three were girls, five boys; 11 knees were treated. Table 1 shows the clinical data.

Knee flexion ranged from 40 to 120°. Contracture (> 90°) was severe in 10 cases. All the children showed popliteal webbing associated with the joint stiffness (Fig. 1). In one case (patient 8), MRI was performed preoperatively to explore popliteal web contents and locate femoral vessels and the sciatic nerve. In nine cases (patients 1, 2, 3, 4, 7 and 8), there was no quadriceps motor activity, with associated patellar agenesis in six cases (patients 1, 2, 3 and 8). Four knees (patients 1 and 7) showed ankle stiffness in equinus. Six of the eight had never had surgery; of the other two, patient 2 had had isolated popliteal band tenotomy at the age of 1.5 months and patient 5 had undergone more complex surgery comprising posterior knee release with inner hamstring transfer to the extensor system and shortening osteotomy of the femur with attempted intraepiphyseal arthrodesis at the age of 20 months.

Progressive correction was by Ilizarov external fixation [1]. Extensive fixation is especially useful, given the joint stiffness. Ring translation with respect to the bone is avoided by fitting olive pins in the plane of the deformity. Superior femoral peg-docking instead of pinning improves assembly stability, reducing the risk of movement. The rings are perpendicular to the corresponding limb segments. The fixator components on either side of the deformity form two cylinders centered on the diaphysis and connected by two

Table 1 Main clinical preoperative data of the series.

<table>
<thead>
<tr>
<th>Side</th>
<th>Age (months)</th>
<th>Flexion contracture (degrees)</th>
<th>Associated deformities</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Left</td>
<td>42</td>
<td>110</td>
</tr>
<tr>
<td></td>
<td>Right</td>
<td>48</td>
<td>90</td>
</tr>
<tr>
<td>2</td>
<td>Left</td>
<td>48</td>
<td>40</td>
</tr>
<tr>
<td>3</td>
<td>Left</td>
<td>36</td>
<td>110</td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>48</td>
<td>110</td>
</tr>
<tr>
<td>5</td>
<td>Left</td>
<td>69</td>
<td>90</td>
</tr>
<tr>
<td>6</td>
<td>Left</td>
<td>96</td>
<td>90</td>
</tr>
<tr>
<td>7</td>
<td>Left</td>
<td>15</td>
<td>100</td>
</tr>
<tr>
<td>8</td>
<td>Right</td>
<td>66</td>
<td>120</td>
</tr>
</tbody>
</table>
sets of three threaded jointed stems. The three joints are perpendicular to the main deviation and are positioned the top of the deformity so that two are lateral to the femoral condyles and the third anterior to the knee in the prolongation of the joint extension. For flexion contracture, the stems are fixed to the ring in the frontal plane, along the femoral diaphysis. Fitting the external fixator never requires periarticular soft-part release.

Correction was initiated as of week 1, the program being entirely carried out in a rehabilitation center on an inpatient basis. Joint uncoupling followed by gradual flexion contracture reduction at joint level sometimes required extending the stems by 4–5 mm per day when they were particularly remote from the mechanical axis of the correction (Fig. 2). Correction speed was variable, adjusted to the patient’s individual tolerance. Correction duration depended on initial lesion severity and on the discrepancy between bone and soft parts, so as to achieve suppleness, and thus varied, while always longer than 6 weeks. The correction was checked on X-ray every 2 weeks (Fig. 3).

Once the angular correction was achieved, it was stabilized by leaving the fixator in compression without change.
in axis, for 1 month. After removal of the external fixator, the correction was maintained by a femoral–pedal orthosis under maximal knee extension [2] (Fig. 4)—prescribed until the end of the patient’s growth period, to ensure against recurrence.

Results

The clinical data are presented in Tables 1 and 2.

Results of initial treatment

Complete extension was obtained in eight out of 11 knees at the end of the initial Ilizarov correction program, with residual contracture of 10–15° in the other three. Correction lasted between 6 weeks and 6 months, with the fixator maintained for 12 weeks to 7 months in all. Three patients (patients 1, 3 and 4) had inner hamstring transfer onto the extensor system, performed 6 months after fixator fitting, once the angular correction had been obtained. In one case (patient 8), femorotibial intraepiphyseal arthrodesis was performed once the external fixator had achieved angular correction. At the end of the immobilization period, ten out of 11 knees were stable.

Recurrence and repeat correction

In six cases, progressive flexion contracture recurrence required repeat correction, using the initial technique, at a mean 3 to 4 years. Postoperative follow-up found four evolutive partial posterior tibial dislocations and one complete posterior tibial dislocation with respect to the femur, in all cases in association with contracture recurrence. Recurrent knee extensor contractions rapidly evolved to above

<table>
<thead>
<tr>
<th>Patient</th>
<th>Side</th>
<th>Flexion contracture after initial correction</th>
<th>Flexion contracture after recurrence</th>
<th>Postoperative stabilisation</th>
<th>Flexion contracture at last follow-up</th>
<th>Age (in years) at last follow-up</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Left</td>
<td>0</td>
<td>Knee dislocation</td>
<td>Yes</td>
<td>Arthrodesis in full extension</td>
<td>14</td>
<td>Femoral fracture, knee dislocation, equinus deformity</td>
</tr>
<tr>
<td></td>
<td>Right</td>
<td>0</td>
<td>No recurrence</td>
<td>Yes</td>
<td>Extension</td>
<td>22</td>
<td>Equinus deformity</td>
</tr>
<tr>
<td>2</td>
<td>Left</td>
<td>0</td>
<td>No recurrence</td>
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<td>Extension</td>
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<tr>
<td>3</td>
<td>Left</td>
<td>10</td>
<td>80</td>
<td>Yes</td>
<td>Extension</td>
<td>15</td>
<td>Tibial and femoral epiphyseal separation equinus deformity</td>
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<tr>
<td>Right</td>
<td>10</td>
<td>85</td>
<td>Yes</td>
<td>Extension</td>
<td>12</td>
<td>Equinus deformity</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>15</td>
<td>50</td>
<td>Yes</td>
<td>Arthrodesis in full extension</td>
<td>8</td>
<td>Equinus deformity</td>
</tr>
<tr>
<td>5</td>
<td>Left</td>
<td>0</td>
<td>No recurrence</td>
<td>Yes</td>
<td>Extension</td>
<td>19</td>
<td>Knee instability, Tibial and femoral epiphyseal separation equinus deformity</td>
</tr>
<tr>
<td>6</td>
<td>Left</td>
<td>0</td>
<td>No recurrence</td>
<td>Yes</td>
<td>Arthrodesis in full extension</td>
<td>12</td>
<td>Equinus deformity</td>
</tr>
<tr>
<td>Right</td>
<td>0</td>
<td>90</td>
<td>No</td>
<td>Arthrodesis in full extension</td>
<td>19</td>
<td>—</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Right</td>
<td>0</td>
<td>No recurrence</td>
<td>Yes</td>
<td>Arthrodesis in full extension</td>
<td>19</td>
<td>—</td>
</tr>
</tbody>
</table>
80°. In four of the six cases, supplementary femorotibial intraepiphyseal arthrodesis was performed after repeat ilizarov correction (Fig. 4).

Other complications

There were no cutaneous or neurologic complications. The preoperative MRI performed on patient 8 confirmed sciatic nerve positioning within the web, with vessels close to the femur. Eight patients developed equinus following joint correction. In three cases, local complications involving the fixation pins required minor reintervention. Six major mechanical complications occurred: two diaphyseal femur fractures, and four Salter-I epiphyseal detachments (two at the distal extremity of the femur and two at the proximal extremity of the tibia), requiring surgical revision involving either extension of the osteosynthesis material or epiphyseal fixation. Following these reinterventions, correction could be resumed without delay. In one case, multidirectional knee instability required continued immobilization by femoral—pedal orthosis.

Results at long-term follow-up

Age at last clinical and radiological follow-up ranged from 8 to 22 years. All patients were able to walk without pain and without canes or frames. Five knees were in arthrodesis, free of residual contracture. The other six showed below 10° residual contracture and 30—45° flexion.

Discussion

Clinical features

The popliteal pathology can be unilateral or bilateral, and tends to be asymmetric. Severity varies from a simple pigmented line on the posterior side of the lower limb to a real popliteal web stretching from ischium to calcaneus. The deformity increases with developmental growth [7—10]. Severe forms show a thick fibrous cord within the subcutaneous tissue on the free edge of the web. The foot is often set in equinus. Unlike the femoral vessels, the sciatic nerve, frequently abnormally short, is in direct contact with the inner side of the fibrous band [10—12], which may sometimes take on the aspect of a genuine muscle, known as the calcaneo—ischiadicus [9,10]. Such an anatomic pattern was found in patient 8’s preoperative MRI scan.

Treatment strategy

Preoperative assessment may include locating the sciatic nerve within the fibrous band. Electrostimulation and CT scan can be useful, but are coming to be superseded by MRI, more effective in detecting vasculonervous structures [13]. MRI was performed in the most recent case in the present series, and perfectly highlighted the close connection between the popliteal band and the sciatic nerve. Other authors have used preoperative arteriography—which, however, turns out to be as pointless as it is invasive, since the femoral vessels are usually positioned quite normally.

In popliteal pterygium syndrome with severe knee flexion contracture, joint mobility is virtually nil, and the aim of treatment is to obtain a stiff but functionally positioned knee to enable walking and facilitate the use of a postural splint [1]. The strategy must take full account of the discrepancy between bone structure and soft parts. Soft tissue retraction, sciatic nerve shortening, popliteal skin fragility, anatomic variations and the presence of the popliteal web all set limits to surgery [14].

For most teams, surgery includes extensive and often destabilizing soft-part release, covering plasty using a local or free flap, shortening osteotomy, and sometimes microsurgical reconstruction of vascular and/or nervous axes [5,15]. There is general agreement that early correction is preferable [5,16]. These techniques usually involve heavy and multistep surgery—which is why we opted rather for progressive correction by external fixator.

Orthopedic methods, using successive casts, can sometimes give good results but have the drawback of lasting for years and being very inconvenient for the patient, and often prove ineffective since joint stiffness tends to be considerable [6,17,15]; correction may fail to be achieved in severe cases, and posterior tibial displacement is a documented associated complication [16,18]. Continuous traction methods, as well as making extreme demands on the patient, can entail severe complications: one case of bilateral amputation has been reported [17].

Supracondylar femoral extension osteotomy is most suited to cases where some mobility is to be conserved and flexion contracture predominates in the bony structure rather than in the joint [18].

Supracondylar femoral shortening osteotomy is a logical attitude for correcting the discrepancy between soft parts and bone tissue [14,16]. It relaxes the soft parts and especially the sciatic nerve, but at the cost of weakening the quadriceps, with consequent risk of recurrence [14].

External fixators and the ilizarov technique in particular are close in conception to orthopedic management [1]. This technique may be indicated in severe periartricular retraction when the aim is to provide the child with a limb in extension to enable walking [2]. The progressive nature of the reduction enables continuous distension of capsule-ligamentary, tendinous and vasculonervous components [2]. The cutaneous retraction—bands and webs, however extensive—are also supplanted, circumventing the need for covering or cutaneous expansion plasty. No cutaneous complications occurred in the present series of 16 corrections. Nor did any sciatic nerve lesion occur during treatment. Uncoupling the joint while correcting the deformity saves the growth cartilage from stress caused by the fixator, thus conserving bone growth potential in these young patients and reducing the risk of disabling difference in limb length.

We found it logical to associate inner hamstring transfer onto the extensor system in patients with extensor system agenesis, expecting it to limit the risk of flexion contracture recurrence; however, not only was the operation difficult to perform, but it failed to prove effective, and we have since abandoned it.
Complications

The fixator exerts considerable force, entailing a risk of fracture either beyond the synthesis zone (femoral or tibial diaphysis) or within it (notably, epiphyseal detachment), as we found in some of the present patients. Such complications occurred in young patients (3–4 years old), raising the question of the optimal age for correction. Correction is easier in young children, but the ideal age is hard to determine taking account of the fragility of the bone and the force of the external fixator’s action. This treatment is not risk-free, and is contraindicated in patients under 3 years of age.

Ankle equinus, found in one in three cases on initial consultation, may occur or worsen during correction. In the most severe cases, Achilles tendon lengthening may be indicated, which will either reduce the force of the external fixator or require extending it to the ankle—although this was not needed in the present series.

More than half of the cases in the present series showed recurrence following the initial correction program. Popliteal pterygium syndrome’s natural evolution almost systematically entails flexion contracture recurrence, due to muscular imbalance between more-or-less retracted posterior components and anterior components that are weak or absent. The quadriceps is unused and stretched beyond capacity, leaving it almost nonfunctional. Posterior component retraction tends to worsen with growth. Our hamstring-transfer muscular rebalancing strategy, applied in four cases, failed to avoid or even to delay recurrence in three cases.

Recurrences were managed with the same progressive correction technique, supplemented according to age by intrajoint arthrodesis between the epiphyses. Five such arthrodeses were performed at a mean age of 64.6 months, including three during repeat Ilizarov correction, conserving growth potential by conserving conjunctive cartilage; at last follow-up, they showed consolidation in extension.

Mobility after Ilizarov correction remained poor, with residual malalignment. The technique is known to cause stiffening, which was turned to advantage in the present case. The loss in mobility can be explained by the often severe deformation of the epiphysis under correction strain and by the presence of pins in the muscles for a period of weeks to months, causing adherences. This stiffness does not, however, protect against recurrence, and splints or orthoses and rehabilitation are mandatory to maintain the correction.

In conclusion, Ilizarov’s circular external fixator technique corrects significant joint stiffness while supplying soft tissue. It does not preclude any subsequent surgery to soft tissue or bone. The technique needs to be fully mastered, and treatment and follow-up need to be long. Despite stiffness, the functional results are satisfactory, enabling walking—which is impossible in severe cases of popliteal pterygium syndrome. It does not, however, protect against recurrence, which is virtually systematic during developmental growth. Repeat correction can always be performed, ahead of possible definitive stabilization by arthrodesis in a functional position.

Conflict of interest

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.otsr.2009.01.004.

References