Treatment of Congenital Pseudarthrosis of the Tibia with the Ilizarov Technique

Case Report and Review of the Literature

REC Rose, DEP Wright

ABSTRACT

Congenital pseudarthrosis of the tibia continues to pose one of the most difficult problems in paediatric orthopaedic surgery. The surgical procedures most used for treating congenital pseudarthrosis of the tibia are intramedullary nailing associated with bone grafting, vascularized fibular graft and the Ilizarov external circular fixator. Even when union is achieved, the residual deformities in the affected limb often result in significant disability. These deformities include leg-length discrepancy, angular tibial deformities, ankle mortise valgus and fibular non-union. The Ilizarov method allows simultaneous excision of the pseudarthrosis site, correction of the deformity and lengthening. However, refractures, ankle joint stiffness, fibular non-union with progressive ankle valgus are frequent sequelae with the Ilizarov technique. The surgeon should know when to abandon reconstructive procedures and create a more functional patient with an amputation. The authors discuss the indications and results of the Ilizarov external fixator in two patients with this complex problem. In addition, a critical review of the current literature is undertaken.

Tratamiento de la Pseudoartrosis de la Tibia con la Técnica de Ilizarov

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RESUMEN

La pseudoartrosis congénita de la tibia sigue siendo uno de los problemas más agudos a los que se enfrenta la cirugía ortopédica pediátrica. Los procedimientos quirúrgicos más recurridos para tratar la pseudoartrosis congénita de la tibia son el tratamiento con clavo intramedular, asociado con el injerto del hueso, el injerto vascularizado fibular, y el fijador externo circular de Ilizarov. Incluso cuando se logra la unión, las deformidades residuales en el miembro afectado a menudo traen como consecuencia una invalidez significativa. Estas deformidades incluyen la discrepancia en la longitud de las piernas, las deformidades angulares de la tibia, valgus de la mortaja del tobillo, y nounión fibular. El método de Ilizarov permite la escisión simultánea del sitio de la pseudoartrosis, la corrección de la deformidad y el alargamiento. Sin embargo, las refracturas, la rigidez de la articulación del tobillo, y la nounión fibular con el valgus progresivo del tobillo, son secuelas frecuentes con la técnica de Ilizarov. El cirujano debe saber cuándo abandonar los procedimientos de reconstrucción y crear un paciente más funcional con una amputación. Los autores discuten las indicaciones y resultados del fijador externo de Ilizarov en dos pacientes con este problema complejo. Además, se emprende una revisión crítica de la literatura actual.

West Indian Med J 2007; 56 (3): 294

INTRODUCTION

Congenital pseudarthrosis of the tibia (CPT) is one of the most challenging problems in paediatric orthopaedics. The condition exhibits a wide range of severity and the response to treatment is unpredictable. Congenital pseudarthrosis of the tibia is a rare disorder, 1 per 190 000 live births and is of unknown aetiology (1). In 40% – 80% of patients, it is associated with neurofibromatosis which generally does not influence the final outcome (2).

Many researchers consider CPT a biologic problem and not merely a mechanical one (3–5). It has been suggested that the periosteum and subperiosteal tissue act by aggressively constricting the bone. Light and electron microscopy

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indicate that the tissue at the site of the lesion is not a neurofibroma but a fibrous hamartoma (3). Whether the fibrous tissue represents the primary lesion or is caused by the pseudarthrosis is unknown. The hamartomatous tissue could cause the tibial narrowing and pathologic fracture in two possible ways: by actively resorbing the bone or by its fibrous components acting as a passive inexpandable barrier to normal circumferential growth (3, 6, 7).

Treatment options have varied greatly and have included both operative and non-operative approaches (8–11). Although no single method has proven ideal, the highest rates of union have been reported with surgery (12–18). Currently, three different surgical approaches: vascularized fibular graft, intramedullary stabilization and the Ilizarov technique have been relatively successful in obtaining initial union of the pseudarthrosis (17–20). However, despite that success, the quality and longevity of the consolidation and the future function of the involved extremity remain uncertain (16, 20, 21). This uncertainty stems from the fact that surgeons can amass only very limited personal experience due to the rarity of CPT. In addition, the lack of long-term follow-up studies identifying the rates of refracture, limb-length discrepancy and residual angular deformity make it difficult to accurately predict the outcome.

The surgical technique and preliminary results of the procedure in two patients with CPT treated with the Ilizarov technique are reported. In addition, a review of the other current surgical methods was undertaken.

CASE 1

A ten-year old child was diagnosed at age one year with CPT. Four surgical procedures which included open reduction and plating of the tibia with bone grafting, two separate bone grafting procedures and transposition of the fibula were performed in an attempt to heal the pseudarthrosis. Due to the failure of union, the child was referred to the authors. Examination revealed anterior bowing of the left tibia, a stiff ankle with a valgus deformity and a 9cm leg-length discrepancy (Fig. 1).

Fig. 1: Congenital pseudarthrosis of the tibia with non-union and angular deformity. Inadequate fixation at the pseudarthrosis site. Fibula was transposed and fused to the tibia.

Under tourniquet control, an anterior longitudinal skin incision was made along the distal third of the left tibia. The periosteum was incised proximal and distal to the pseudoarthrosis and a small periosteal elevator was used to elevate the periosteum both proximally and distally. The pseudarthrosis was then dissected from the surrounding muscles extra-periosteally and a segment of bone and periosteum approximately 4cm long were excised. The medullary canal, which should always be visible at the resection site, was drilled. The gap created by the excision osteotomy was not compressed. A thick K-wire (2mm to 3mm diameter) was passed from the osteotomy site through the sole of the foot and then drilled retrograde into the proximal tibia. A circular ring fixator was then applied. The construct consisted of two full circle rings and a 5/8 ring which were attached to the bone with wires and half-pins. The rings were connected to each other by threaded rods. A small anterior skin incision was then made at the level of the proximal tibial metaphysis and the periosteum was elevated. A Gili saw was passed subperiosteally following which the osteotomy was performed. The proximal osteotomy site was compressed for ten days to allow early callous formation. Distraction over the guide wire was then commenced at a rate of 0.25 mm every six hours (Fig. 2). The docking site was grafted with autogenous

Fig. 2: Antero-posterior and lateral views of the tibia with Ilizarov fixator. Middle segment of the tibia was transported distally over the guide wire.

iliac bone. Lengthening of the limb was then started at a rate similar to bone transport (Fig. 3). During the period of lengthening, the docking site was compressed at a rate of 0.25 mm daily for ten days. The frame was removed at twelve months due to complaints by the child of pain in the leg. Solid bone union was evident at the pseudarthrosis site at that time. An above-knee cast was applied to protect the regenerate bone and the pseudarthrosis site since refracture is a common occurrence.

Fig. 3: Patient standing with Ilizarov frame on the left leg. Distraction occurred between the proximal middle rings.

The child fell and fractured through the regenerate bone which was still protected with a cast. An open reduction and internal fixation was performed. Two years following removal of the frame, the pseudarthrosis site remained fully healed (Fig. 4). There still remains a 4cm leg length discrepancy and limited ankle motions. This child will be followed until skeletal maturity.

Fig. 4: Solid union at the pseudarthrosis site. Healed fracture of the regenerate bone. Dynanic compression plate *in situ.*

CASE 2

An eighteen-year old male presented to the authors with a diagnosis of CPT, a 4cm leg-length discrepancy and a stiff ankle. Two previous surgeries which included resection of the pseudarthrosis, fibula osteotomy, insertion of an intramedullary rod (rush rod) into the tibia along with bone grafting and plate fixation also combined with bone grafting failed to achieve bone union (Fig. 5). Treatment consisted of

Fig. 5: Congenital pseudarthrosis of the tibia with anterior bowing of the tibia. The plate has protruded through the skin.

simultaneous removal of the plate and screws, excision of the pseudarthrosis and abnormal periosteum, correction of the angular deformity in the tibia, application of the Ilizarov frame, acute compression of the excision osteotomy site and proximal tibial metaphyseal osteotomy. The limb was shortened a further 3cm following excision of the pseudarthrosis. Autogenous bone graft was placed around the compressed site before the wound was closed. Lengthening of the leg was commenced fourteen days after the osteotomy at a rate of 0.25 mm every six hours (Fig. 6). The pseudarthrosis was

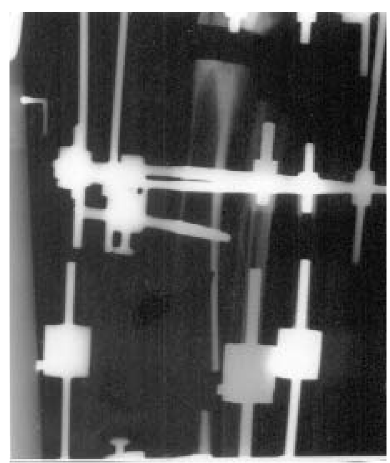

Fig. 6: Illizarov fixator *in situ.* Lengthening of the limb *via* the proximal tibial osteotomy site.

completely healed at ten months after which the frame was removed. An above-knee cast was applied for three months to protect the regenerate bone as well as the union site. Nine months following the removal of the frame, there was solid union at the pseudarthrosis site, consolidation of the regenerate bone, a stiff ankle and a 2cm leg-length discrepancy (Figs. 7, 8).

Fig. 7: Left tibia showing bony union at the pseudarthrosis site. Proximally, the regenerate bone is consolidating well.

Fig. 8: Both lower limbs with mild valgus of the left tibia. There is a 2 cm leg-length discrepancy.

DISCUSSION

Currently, vascularized fibular grafting, intramedullary stabilization and external fixation are being used with relative success for the treatment of CPT (17 – 21). The superiority of any specific procedure is difficult to determine. The different types of pseudarthrosis, the effects of previous treatment, the need for follow-up until skeletal maturity, the timing of the procedure and the definition of what is a successful result all make it difficult to compare the outcome of treatment in different series.

An interesting observation by Grill *et al* (12) was that conservative treatment was not as useless as some authors believed. The use of a stable knee-ankle-foot orthosis with an anterior shield in the pre-pseudarthrotic stage might delay a fracture and the development of a pseudarthrosis, allowing the patient to reach an older age before undergoing surgical treatment. This is of importance as a better healing rate has been shown in most of the procedures in children older than three years of age. Boero *et al* (21) reported an 85.7% consolidation in children who were operated on after five years of age in contrast to a rate of union of 14.2% in those operated on at ages younger than five years.

Vascularized fibular grafting has received a great deal of attention and several authors have reported success in obtaining initial tibial union (15, 16, 19, 20). However, problems persist. Refracture, non-union at one end of the graft site or in the graft itself are not uncommon complications. A secondary bone grafting procedure is frequently necessary. Persistent angular deformity of the tibia which is often progressive is another problem. In addition, many cases are complicated by severe valgus ankle deformities on the donor sides with proximal migration of the distal parts of the fibula following vascularized fibular graft in children (22). To prevent this complication, tibiofibular metaphyseal synostosis (the Langenskiold procedure) has been recommended (19, 22). However, in a recent review, this procedure was found to only delay, not prevent, the development of ankle valgus in children after harvesting of a vascularized fibular graft (22). Toh *et al* (23) reported on one case in which a split vascularized fibular graft was used to reconstruct both the tibia and fibula simultaneously. They postulated that this should prevent secondary valgus deformity at the ankle. In addition, they combined the vascularized fibular graft with an Ilizarov external fixator.

Umber *et al* popularized the use of the two-part Williams intramedullary rod in North America (24). Anderson et al (25) obtained 100% union in ten patients with this technique. Although union was achieved, many problems were observed, including stiffness of the ankle and subtalar joints caused by insertion of the nail through the calcaneus, talus and tibia, transfixing the ankle and subtalar joints and the distal tibial physis. Refractures in the unprotected region of the tibia may occur with growth of the leg as the end of the rod migrates. Finally, the distal tibial growth plate, which already had a stunted rate of growth, may be further injured by transfixion, resulting in limb-length discrepancy. In an attempt to decrease the prevalence of ankle stiffness, custom interlocking intramedullary nails that do not transfix the ankle joint were developed. In a long term follow-up study by Dobbs *et al* (17), temporary transfixion of the tibiotalar and subtalar joints did not negatively affect the long-term functional result. Valgus deformity of the ankle can compromise the functional result in patients treated successfully with an intramedullary rod. To prevent this complication, Dobbs *et al* (17) recommended placement of a syndesmosis screw once valgus correction had been achieved.

Numerous authors have reported on the use of the Ilizarov circular external fixator in the treatment of CPT (18, 21, 26–28). The Ilizarov technique is a comprehensive approach to all aspects of CPT. It simultaneously attends to various aspects of the condition, including resection of the pseudarthrosis, deformity correction, shortening defect, infections, articular function, weight bearing and the valgus ankle. The disadvantages of this technique are the duration of treatment, its relative complexity, pin tract infections, ankle valgus and refractures. Paley *et al* (18) were the first Western surgeons to report results of circular external fixation for CPT. Their series consisted of 16 cases with a union rate of 94% with one treatment and 100% with two treatments. There were five refractures.

Grill reported results of circumferential removal of pathologic periosteum, excision of the non-union site, immediate correction of angular deformity and compression of the fragments, bone grafting and proximal metaphyseal lengthening using the circular external fixation (28). The pseudarthrosis healed in all nine patients, but six had ankle osteoarthritis, six had residual leg or foot deformity, three had limb-length discrepancy and one sustained a refracture. Boero *et al* (21) reported results of 21 patients treated with the Ilizarov device. Primary consolidation was achieved in 17 of 21 patients in an average of 13.7 months. Refractures, deformity and re-operation were frequent sequalae.

Grill *et al*, in a multicentre study, analyzed the different therapeutic methods used by the European Paediatric Orthopaedic Society (12). The treatment data of 340 patients who underwent 1287 procedures for CPT were analyzed. The therapeutic modalities which were reviewed included the McFarland bypass graft, plating, rodding and grafting, the Ilizarov fixator as well as conservative measures. The findings of that study demonstrated that plating and rodding seemed to afford inadequate stability to allow the pseudarthrosis to heal, and that surgeons who used that kind of fixation resected too little of the pseudarthrotic bone in an attempt to avoid shortening. The results of that study also showed that the Ilizarov technique was the method of choice in the treatment of CPT. In addition to success in correction of the other deformities, this method achieved the highest rate of union (75.5%).

In the indexed cases, the Ilizarov method allowed resection of the pseudarthrosis, immediate correction of the angular deformities, bone transport, compression of the fragments and limb lengthening. In both cases, union was achieved with one treatment. However, the follow-up period is short, less than three years, and in Case I, the patient is still skeletally immature. Refracture is a strong possibility.

Refracture is common in patients with CPT despite apparently solid clinical and radiological union (12–28). There is consensus that surgery should be avoided before the third year of life, and if possible, it should be postponed until the age of five years (12, 17). Late-onset fracture and pseudarthrosis is a more benign form of the lesion (29, 30).

The Ilizarov technique is useful in many cases of CPT in which union failed to occur inspite of many previous surgeries. The use of this method does not preclude the use of other procedures. The Ilizarov method takes considerable time and effort to obtain good results. The surgeon must know when to abandon this procedure and perform an amputation which will make the patient more functional.

The true success of treatment of CPT in a growing child can be known only by following the patient until skeletal maturity. Whatever the technique selected for its treatment, CPT remains one of the most difficult problems in paediatric orthopaedics.

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