

## **Врожденный псевдоартроз голени и лечение методом Илизарова**

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## **Congenital pseudoarthrosis of the tibia and treatment using the Ilizarov technique**

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Врожденный псевдоартроз большеберцовой кости (СРТ) – одна из труднейших проблем в детской ортопедической хирургии. Это редко встречающееся состояние, которое в большинстве своем сочетается с нейрофиброматозом. Варианты лечения чрезвычайно разнообразны. С помощью метода Илизарова можно успешно проводить оперативное лечение псевдоартроза большеберцовой кости. Пациенты и методы: 7 пациентов с СРТ, которых лечили по методу Илизарова в период с 1998 по 2008 г. Результаты: костное заживление произошло во всех случаях кроме одного. (Показатель сращения – 85,7 %). Заключение: Метод Илизарова обеспечивает всесторонний подход ко всем аспектам СРТ. При его применении можно получать очень хорошие результаты.

Ключевые слова: голень, врожденный псевдоартроз, метод Илизарова.

Background: Congenital pseudoarthrosis of the tibia (CPT) is one of the most difficult problems in paediatric orthopaedic surgery. This is a rare condition, which is mostly associated with neurofibromatosis. Treatment options have varied greatly. Successful surgical treatment of tibial pseudoarthrosis is possible by the Ilizarov method. Patients and methods: 7 patients with CPT treated using the Ilizarov technique within the period from 1998 to 2008. RESULTS: Bone healing was achieved in all cases except one. (Union rate – 85,7 %). Conclusion: The Ilizarov technique is a comprehensive approach to all CPT aspects. Its use can end with very good results

Keywords: tibia, congenital pseudoarthrosis, the Ilizarov technique.

### INTRODUCTION

Congenital Pseudoarthrosis of the Tibia (CPT) is a specific type of nonunion; one of the most difficult problems in paediatric orthopaedic surgery; rare condition with incidence 1-200 000, occurring equally in girls and boys. It is characterized by an-

terolateral bowing of the tibia (first physical sign); its cause is unknown (biologic problem and not a mechanical); in 40 % – 80 % of patients it is associated with neurofibromatosis; 10 % of patients with neurofibromatosis have CPT.



Fig. 1 Patient L., 8 y.o.: a, b – before treatment; c, d – X-ray before treatment

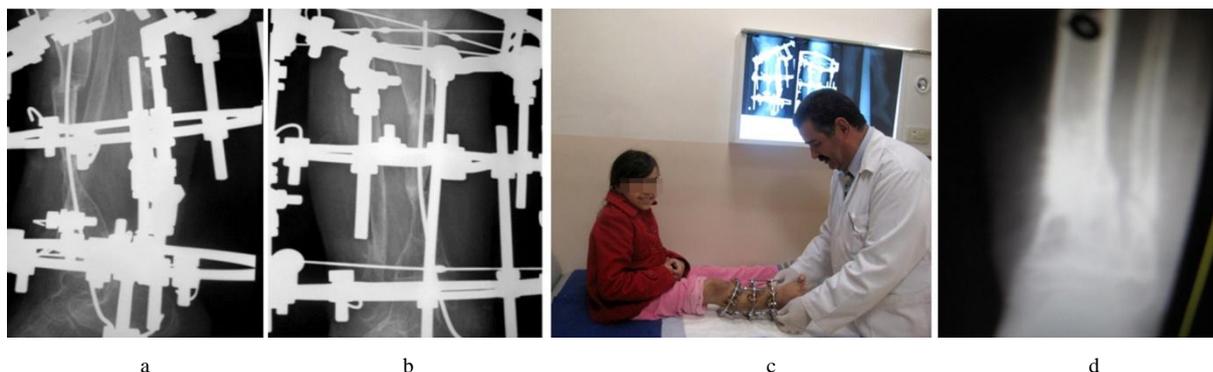


Fig. 2 Patient L., 8 y.o.: a, b – X-ray during treatment; c – patient during treatment; d – X-ray after treatment

### Classification

Several classification systems have been proposed, none provides specific guidelines for management.

Boyd and Andersen are quite similar

**Andersen:** With 5 types:

1. Dysplastic;
2. Cystic;
3. Sclerotic;
4. Fibular;
5. Clubfoot or congenital band type.

**Boyd H.B.** (1982) six types:

*Type I:* Pseudoarthrosis occurs with anterior bowing and a defect in the tibia present at birth.

*Type II:* with anterior bowing and an hourglass constriction of the tibia present at birth. Spontaneous fracture, or that following minor trauma, occurs before 2 years of age. This type is the most common, it is often associated with neurofibromatosis, and has the poorest prognosis.

*Type III:* Pseudoarthrosis develops in a congenital cyst, usually near the junction of the middle and distal thirds of the tibia.

*Type IV:* Pseudoarthrosis originates in a sclerotic segment of bone, “stress” fracture develops in the cortex of the tibia and gradually extends through the sclerotic bone. With completion of the fracture, healing fails to occur and the fracture widens and pseudoarthrosis develops. The prognosis for this type is generally good.

*Type V:* Pseudoarthrosis of the tibia occurs with a dysplastic fibula. Pseudoarthrosis of the fibula or tibia or both may develop.

*Type VI:* Pseudoarthrosis occurs as an intraosseous neurofibroma or schwannoma that results in pseudoarthrosis. This is extremely rare.

**Crawford A.H.** (1986):

(Radiographic) classification of congenital tibial pseudoarthrosis.

Patients with all types are presented with anterolateral bowing of tibia.

In *type I*, the medullary canal is preserved. Cortical thickening might be observed.

*Type II* is defined by the presence of thinned medullary canal, cortical thickening, and tabulation defect.

*Type III* is a cystic lesion, which may be fractured.

*Type IV*, pseudoarthrosis is present with tibial and possibly fibular nonunion.

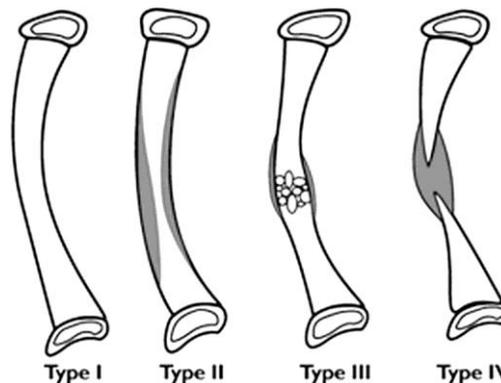


Fig. 3. Classification of congenital pseudoarthroses of the tibia by Crawford A.H.

### EI-Rosasy-Paley Classification

The patient's condition is considered at the time of treatment and is based on three parameters:

- (i) Did the patient have any surgical treatment for the pseudoarthrosis or not?
- (ii) Is pseudoarthrosis mobile or stiff?
- (iii) Plain x-ray examination of the leg affected for CPT radiological typing: atrophic type with narrow bone ends or hypertrophic one with wide bone ends.

### The differential diagnosis of congenital pseudoarthrosis of the tibia.

There are at least four dysplastic lesions of the congenitally bowed tibia (with or without cysts) which are still being confused in the literature:

- 1 – congenital pseudoarthrosis
- 2 – monostotic fibrous dysplasia
- 3 – osteofibrous dysplasia of long bones
- 4 – congenital angulation of long bones

### Treatment

Treatment options have varied greatly, including both operative and non-operative approaches. Although no single method has proven to be ideal, the highest rates of union have been reported with surgery.

The aim of successful treatment is to achieve: *length, union and normal axis of the leg involved.*

The main cause of treatment failure is possibly the abnormal periosteum at the pseudoarthrosis site which has a destruction effect and interferes with bone union achievement (Paley).

### Non Operative Treatment:

– patellar tendon bearing – total contact orthosis, which is used to try preventing fractures or control-

ling development.

The use of a stable knee-ankle-foot orthosis with an anterior shield in the pre-pseudoarthrotic stage might delay fracture and pseudoarthrosis development, 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.

#### Surgical Treatment Options

The superiority of any specific procedure is difficult to determine.

##### Bone graft methods:

1 – *Autologous iliac bone graft*

2 – *Vascularized fibular grafting*

Several authors have reported success in obtaining initial tibial union. However, problems persist. Refracture, non-union. A secondary bone grafting procedure is frequently necessary. Persistent angular deformity of the tibia. 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.

##### Intramedullary stabilization

Recent years the technique of intramedullary rodding described by Anderson is the most common.

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 rod end migrates. Finally, the distal tibial growth plate, which already had stunted rate of growth, may be further injured by transfixion,

resulting in limb-length discrepancy.

##### Symes amputation

– indicated for congenital pseudoarthrosis with limb length inequality greater than 5 cm and/or severe foot deformity;

– when treatment fails, Symes amputation and prosthetic fitting can result in a very functional limb;

– the presence of pseudoarthrosis by itself is not an indication for amputation;

– note that amputation thru pseudoarthrosis may leave scars over amputation stumps, and over growth phenomena may end up requiring additional revision amputations;

– in some cases, spontaneous union of pseudoarthrosis will occur (due to total contact of prosthesis and beneficial effects of wt bearing).

##### Ilizarov circular external fixator

The Ilizarov technique is a comprehensive approach to all aspects of CPT. It attends to various aspects of the condition, including:

*Resection of the pseudoarthrosis, deformity correction, shortening defect, infections, articular function, weight-bearing and the valgus ankle.*

To achieve success, a number of basic principles need to be followed.

These principles are:

1. Correction of the axis of the limb.
2. Debridement of the pseudoarthrosis and opening of the intramedullary canal with insertion of one of the ends of the tibia into the other.
3. Lengthening of the tibia with proximal osteotomy and concurrent compression of the fracture site.
4. Absolute correction of the tibial axis.

#### OUR EXPERIENCE

##### Patients and methods

This is a review of the results of 7 patients (4 – boys; 3 – girls) with congenital pseudoarthrosis of the tibia, managed with the Ilizarov technique between 1998 and 2008 in Jordan, in three different institutions: Zarqa governmental hospital (Ministry of health – MOH); King Hussein Medical Center (Royal medical services); and Ibn-Al Haytham Hospital (private hospital).

Patients aged from 3 to 16 years.

The tibia with pseudoarthrosis involved: the right side in 4 cases; left side in 3 cases.

6 patients have been treated several times surgically with failure.

1 patient presented without any previous surgical intervention.

1 case was treated previously twice by the Ilizarov apparatus unsuccessfully.

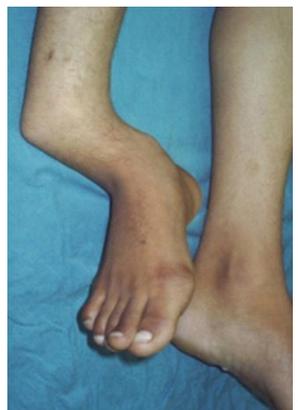
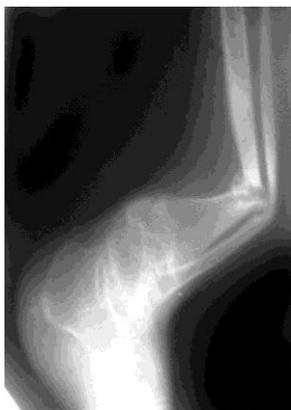


Fig. 4. Patient Y.: leg x-ray (a) and view (b), 14 y.o., before treatment



a



b



c



d

Fig. 5. Patient Y., 14 y.o.: a – during treatment; b – X-ray after treatment; c, d – comparative photo before and after treatment



a



b



c

Fig. 6. X-rays for congenital pseudoarthroses of tibia: a – cystic; b – dysplastic; c – sclerotic

SURGICAL TECHNIQUES

The pseudoarthrosis was approached through a standard anterior approach. The fibrous tissue was dissected and excised, then a small segment of bone and periosteum were excised.

A K-wire was passed and then drilled into the tibia.

The circular Ilizarov ring fixator was then applied.

Osteotomy was performed at the level of the proximal tibial metaphysis.

Then distraction and compression was performed by the rate of 1 mm every day.

The frame was removed at 6-9 months when solid bone union was evident at the pseudoarthrosis site.

An above-knee cast was applied for 2-4 months.

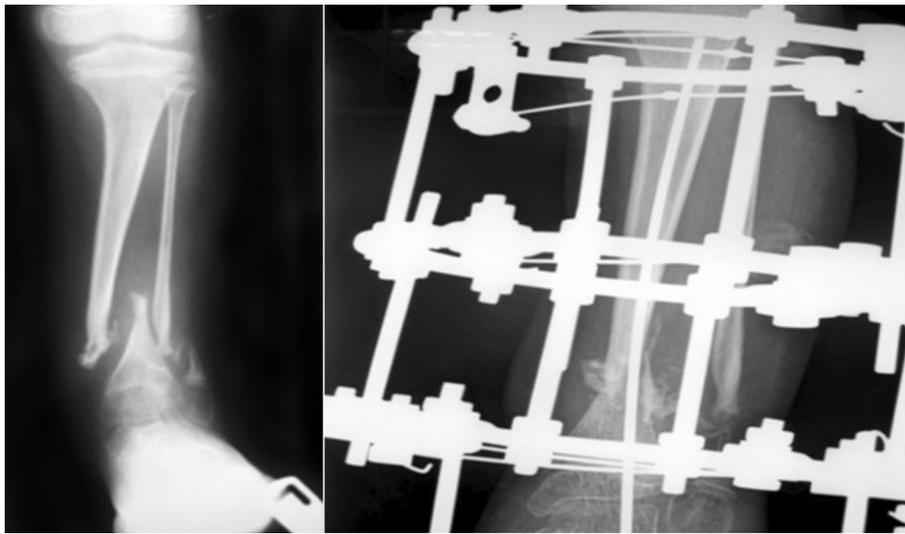


Fig. 7. Case 2

RESULTS

Follow-up ranged from 4 to 11 years.

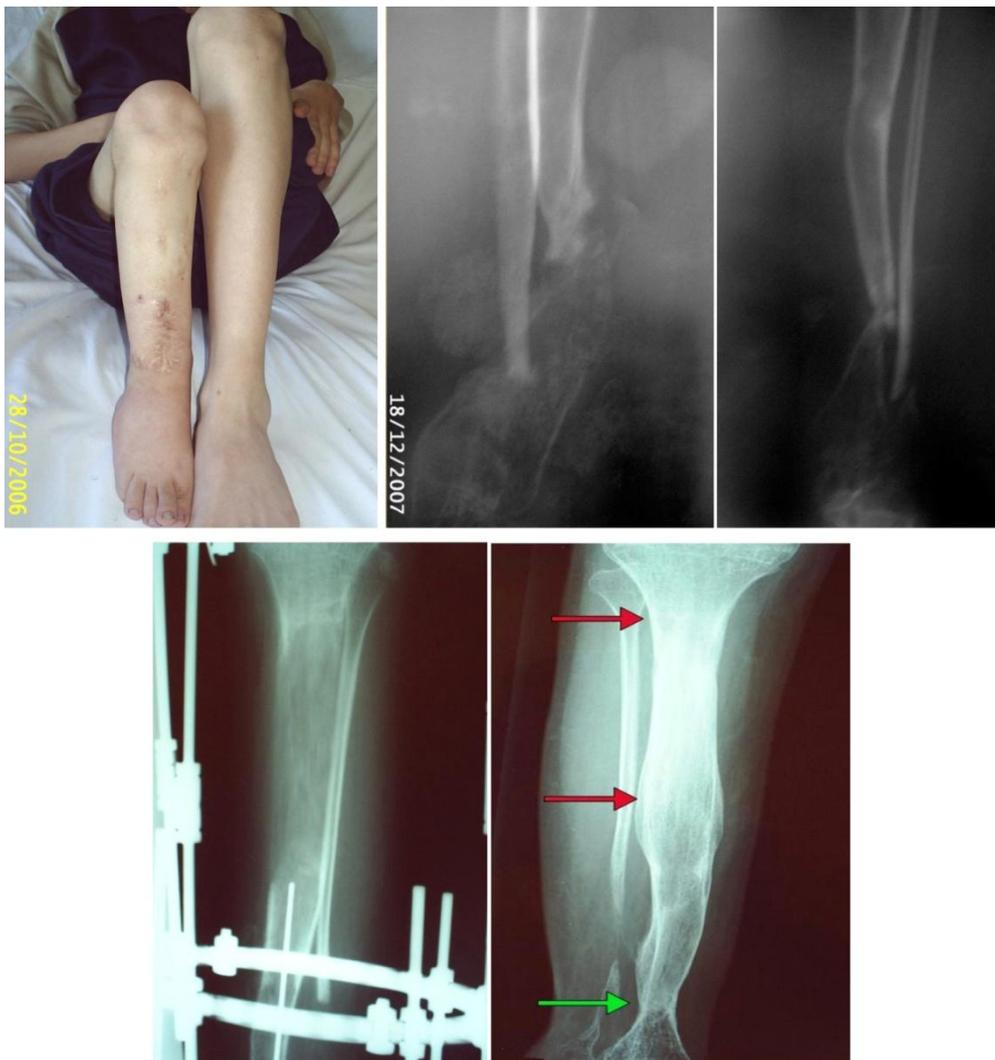
Bone healing was obtained in all 7 patients, one case was complicated by refracture at pseudoarthrosis site after removal of the Ilizarov apparatus by 3 months, and not treated more by this technique (they refused re-application of Ilizarov).

Superficial pin tract infection occurred in most cases (6-7), and they were managed simply and conservatively.

Transient decreased motion of the joint also occurred in most cases.

As noted, one complication can be considered as major which necessitated an additional operative procedure (case of refracture).

Others are minor complications that responded to non-operative treatment and did not cause lasting sequelae.



DISCUSSION AND REVIEW OF THE LITERATURE

Congenital pseudoarthrosis of the tibia is a rare disease with variable history, that is difficult to treat successfully.

Conventional treatment, such as bone grafting and plating, has the reported failure rate approaching 100 %.

New treatment modalities, such as free fibular grafting and the Ilizarov technique, are able to achieve union in most cases.

Although the refracture rate continues to be high, in our experience it is 1 of 7 (14 %).

The union rate using the Ilizarov method is very high.

The potential advantages of this technique include: Full weight-bearing during treatment.

The ability to resect abnormal bone and periosteum.

Equalization of limb lengths.

Elimination of bone deformity.

REVIEW OF THE LITERATURE

Study	Date	Number of cases	Final result and conclusions
1	2	3	4
Journal of the Formosan Medical Association = Taiwan yi zhi 1997;96(5):359-65. Treatment of congenital pseudoarthrosis of the tibia with the Ilizarov method.	1997	8	Union at the pseudoarthrosis site was achieved in 7 of 8 patients (union rate, 87.5 %)
Journal of Pediatric Orthopaedics: September/October 1997 - Volume 17 Ilizarov Treatment of Congenital Pseudarthroses of the Tibia	1997	11	Successful in 9 of 11 patients with an average of 322 days
Journal of Children's Orthopaedics Volume 2, Number 6 / December, 2008 Association of Ilizarov's technique and intramedullary rodding	2008	10	Tibial union was achieved and maintained by combining the Ilizarov technique and intramedullary nailing in 9 of

1	2	3	4
in the treatment of congenital pseudoarthrosis of the tibia			10 patients at final follow-up
Plawecki S, Carpentier E, Lascombes P, Prevot J, Robb JE Department of Orthopaedic Surgery, Hopital A. Michallon, Grenoble, France. Treatment of congenital pseudoarthrosis of the tibia by the Ilizarov method	1990	3	Consolidation was obtained by 3.5 months in three patients who had under- gone several previous surgical procedures
Paley D, Catagni M, Argnani F, Prevot J, Bell D, Armstrong P. University of Maryland Hospital, Baltimore. Treatment of congenital pseudoarthrosis of the tibia using the Ilizarov technique.	1993	16	The union rate was 94 % with one treat- ment and 100 % with two treatments
Journal of Bone and Joint Surgery - British Vol, 84-B, Issue SUPPL. III, 307-308. The surgical management of congenital pseudoarthrosis of the tibia Y. Odeski	2003	10	All patients had eventual successful out- come
Ashraf A .Khanfour, MD Pan Arabic J. Orth. Trauma- vol.12, no (2)   july 2008 Rule of the Ilizarov method in treatment of congenital pseu- doarthrosis of the tibia	2008	9	9 cases were treated by the Ilizarov me- thod, consolidation was obtained in all of them
J Pediatr Orthop B 2000; 9(1):1-2 European pediatric orthopedic society multicenter study of congenital pseudoarthrosis. (members from 13 countries) Treatment approaches for congenital pseudoarthrosis of tibia	2000	340	Ilizarov Tx, + ICBG, 75 % healing; Vascularized fibula, 61 % healing

As noted in the table, many authors have reported good to excellent results in treating congenital pseudoarthrosis of the tibia by the Ilizarov technique, and

many agree that it is the method of choice.

We recommend this type of treatment for this difficult and challenging problem.

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