Congenital anterolateral bowing and pseudarthrosis of the tibia: pathogenesis and treatment with Ilizarov apparatus

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Congenital Anterolateral Bowing and Pseudarthrosis of the Tibia (CPT)

Etiopathogenesis
Pathology
Treatment
Prognosis
Our Strategy of Dealing with Congenital Anterolateral Bowing and Pseudarthrosis of the Tibia

Prevention of fracture or deformity progression with a PTB ankle-foot orthosis

Massive resection of pseudarthrosis site, acute shortening, and fixation with Ilizarov apparatus, with proximal metaphyseal lengthening, at pre-school age

Prevention of re-fracture with an orthosis, preferably until skeletal-maturity

Additional surgery (deformity correction, equalization of length…) if necessary
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STUDY 1
Histological Analysis of the Pseudarthrosis Site

STUDY 2
Retrospective Study on the Efficacy of Preoperative Brace Treatment

STUDY 3
Retrospective Study on the Treatment Course with Ilizarov Apparatus

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STUDY 1

Histological Analysis of the Pseudarthrosis Site
Patients

CPT: 9 cases
(NF1: 6 cases)
Age at Operation:
5y10m

Methods

Histological analysis
Control:
adult posttraumatic pseudarthrosis

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Histology of CPT

Case 1

X-ray

H.E.

Azan

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Fibrous Cartilage in CPT

Case 1

A. H.E.

B. Toluidine blue

C. Toluidine blue

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Osteoclasts in CPT

A. H.E. 
B. TRAP 
C. TRAP

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Osteoclasts not only on Bone Surfaces but also in Fibrous Tissues

Case 2

Case 2

Case 3
Bone Histomorphometry

Osteoclast Surface (%)  
[Oc.S/BS (%)]

Osteoclast Number (/mm)  
[N.Oc/BS (#/mm)]

Osteoclasts in Fibrous Tissues
Enhanced Osteoclastogenesis in CPT

- N.Oc/BS (#/mm)
- Oc.S/BS (%)
- OCs in Fibrous Tissue
Expression of RANK ligand in Fibroblastic Cells and Osteoclasts in CPT

Immunostaining for RANK ligand

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Possible Pathophysiology of CPT

- Enhanced Osteoclastogenesis
- Pseudarthrosis
- Increased Expression of RANK ligand in Fibroblastic Cells
STUDY 2

Retrospective Study on the Efficacy of Preoperative Brace Treatment

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Patients

CPT: 7 patients
(NF1--2, Fibrous Dysplasia--1)
Age at initiation of brace treatment:
4m-14m

Methods

Retrospective radiological analysis:
Measurement of tibial deformity
(coronal and sagittal plane)

PTB ankle-foot orthosis
allowing partial WB

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Status of Tibia & Fibula before Brace Treatment

- Pseudarthrosis at initiation of brace Tx
- Tibia & Fibula: 3
- Fibula only: 1
- None: 3

Measurement of tibial deformity until tibial fracture or operation

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Deformity of Tibia during Brace Treatment

Coronal-plane Deformity

Sagittal-plane Deformity

varus (degrees)  procurvatum (degrees)

age (years)  age (years)
Deformity of Tibia during Brace Treatment

Oblique-plane Deformity

\(\text{=} \arctan \sqrt{\tan^2\text{ap} + \tan^2\text{lat}}\)

The graph shows the relationship between the degrees of oblique-plane deformity and age (years). The graph includes data points labeled (F) and Op, indicating different stages or conditions.
STUDY 3

Retrospective Study on the Treatment Course with Ilizarov Apparatus
Patients

Six patients operated between 1992 and 2002

Sex: 4 boys and 2 girls
Affected Side: 3 in the right and 3 in the left
Associated Problem: NF1--2 patients, Fibrous Dysplasia --1 patient
Age at Operation: 3y4m - 6y1m
Follow-up Period: 1y9m - 12y2m (mean 6y0m)
## Treatment Course during Ilizarov Procedure

<table>
<thead>
<tr>
<th>Case</th>
<th>Age at Initial Surgery</th>
<th>Preop. Pseudarthrosis of Tibia</th>
<th>Preop. Pseudarthrosis of Fibula</th>
<th>Resection Length of Tibial Lesion</th>
<th>Proximal Metaphyseal Lengthening</th>
<th>Duration of Ilizarov Application</th>
<th>Complications during Initial Treatment</th>
<th>Fusion of Pseudarthrosis at Ilizarov Removal</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>3y4m</td>
<td>yes</td>
<td>yes</td>
<td>15mm</td>
<td>no</td>
<td>214days</td>
<td>superficial infection</td>
<td>yes</td>
</tr>
<tr>
<td>2</td>
<td>5y6m</td>
<td>yes</td>
<td>yes</td>
<td>20mm</td>
<td>50mm</td>
<td>399days</td>
<td>deformity at pseudarthrosis site, equinus foot, deep infection</td>
<td>no</td>
</tr>
<tr>
<td>3</td>
<td>5y0m</td>
<td>yes</td>
<td>yes</td>
<td>25mm</td>
<td>35mm</td>
<td>364days</td>
<td>delayed union at pseudarthrosis site, premature consolidation of fibula, deformity of elongated callus, knee &amp; ankle contracture</td>
<td>yes</td>
</tr>
<tr>
<td>4</td>
<td>5y2m</td>
<td>no</td>
<td>yes</td>
<td>40mm</td>
<td>50mm</td>
<td>210days</td>
<td>knee &amp; ankle contracture, superficial infection</td>
<td>yes</td>
</tr>
<tr>
<td>5</td>
<td>6y1m</td>
<td>yes</td>
<td>yes</td>
<td>50mm</td>
<td>62mm</td>
<td>315days</td>
<td>fracture of ipsilateral distal femur</td>
<td>yes</td>
</tr>
<tr>
<td>6</td>
<td>5y2m</td>
<td>yes</td>
<td>yes</td>
<td>40mm</td>
<td>70mm</td>
<td>186days</td>
<td>premature consolidation of fibula, breakage of Ilizarov ring, knee &amp; ankle contracture</td>
<td>yes</td>
</tr>
</tbody>
</table>

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## Treatment Course after Ilizarov Removal

<table>
<thead>
<tr>
<th>Case</th>
<th>Procedures at the time of Ilizarov Removal</th>
<th>Complications after Ilizarov Removal</th>
<th>Overall No. of Operations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>no</td>
<td>re-fracture (10 years after removal, Orthofix external fixation)</td>
<td>3</td>
</tr>
<tr>
<td>2</td>
<td>intramedullary rodding, ABMI</td>
<td>failure of fusion (17 months after removal, intramedullary rodding, led to fusion), ankle valgus (5 years after removal, PETS)</td>
<td>8</td>
</tr>
<tr>
<td>3</td>
<td>bone graft for elongated callus, ABMI</td>
<td>re-fracture (3 months after removal, intramedullary rodding), ankle valgus (17 months after removal, PETS), breakage of intramedullary rod (4 years after removal)</td>
<td>8</td>
</tr>
<tr>
<td>4</td>
<td>distal tibio-fibular bone graft</td>
<td>re-fracture (17 months after removal, Ilizarov external fixation)</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>bone graft for elongated callus and compression site</td>
<td>tibia valga (5 months after removal, PETS) re-fracture (21 months after removal, Ilizarov external fixation)</td>
<td>6</td>
</tr>
<tr>
<td>6</td>
<td>no</td>
<td>no</td>
<td>3</td>
</tr>
</tbody>
</table>

**AMBI** : autologous bone marrow injection  
**PETS** : percutaneous epiphyseodesis using transphyseal screws  

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Deformity of Tibia after Removal of Ilizarov Apparatus

Coronal-plane Deformity

Sagittal-plane Deformity

varus (degrees)

procurvatum (degrees)

valgus

Ilizarov
Pre-Fx
Final

Ilizarov
Pre-Fx
Final

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Deformity of Tibia after Removal of Ilizarov Apparatus

Oblique-plane Deformity

degrees

Removal  Ilizarov  Pre-Fx  Final

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Patient 6

Pre-Op (5y2m)

Initial Operation

Before Ilizarov Removal

Final (5y10m)
Patient 3 with re-fracture and ankle valgus

Pre-Op (5y0m)            Before Ilizarov Removal            Re-fx 3m after Removal
IM for Re-fx            PETS for Ankle Valgus            Final (10y9m)
Discussion

Osteoclastogenesis even in the fibrous area apart from bone surfaces

Massive Resection of Pseudarthrosis Site and Acute Shortening to obtain wide contact area and opening of the medullary canal 5/6 fusion

Simultaneous proximal metaphyseal lengthening Ext. Fixation for 6m-12m

High Re-fracture Rate (4/6)

probably MALALIGNMENT

Solution???
Residual Challenges after Healing of Congenital Pseudarthrosis in the Tibia

Kristiansen LP et al.: Clin Orthop 2003

5/7 re-fracture
7/7 axial deformity

to avoid MALALIGNMENT

Precise alignment check during external fixation
PETS for malalignment during follow-up period
Intramedullary rodding after Ilizarov removal ??

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Efficacy of Preoperative Brace Treatment

EPOS Multicenter Study (2000, JPO-B)
14 patients with intact tibia & anterior bowing procurvatum 2-53 degrees (at presentation) (plaster or splints) ↓ 25-54 degrees (pseudarthrosis -) 30-80 degrees (pseudarthrosis +)
Conclusion

1. In CPT, osteoclastogenesis is enhanced on the surface of the bone and cartilage and even in the fibrous area apart from bone surfaces.

2. Such possible pathogenesis leads to the necessity of massive resection of the pseudarthrosis site during surgery. In 5 patients out of 6, fusion was obtained after 6 to 12 months of treatment. But re-fracture occurred in 4 patients. Malalignment may be one of the factors of re-fracture.

3. Brace treatment before surgery was beneficial at least until fracture of the tibia.

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