Limb lengthening in short-stature patients using monolateral and circular external fixators

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Objectives To review the results of distraction osteogenesis in short-stature patients in our centre and analyse outcomes including complications.

Design Retrospective study.

Setting University teaching hospital, Hong Kong.

Patients Eight patients with short stature (three had achondroplasia, three constitutional short stature, and two hypochondroplasia) operated on for limb lengthening using monolateral or circular external fixators between 1995 and 2006 were reviewed.

Results The mean age at the time of surgery was 20 years (range, 9-39 years). The fixators used were either Ilizarov or Orthofix. The average gain in length per bone segment was 5.2 cm (range, 3.2-8.0 cm), and the average percentage lengthening was 21% (range, 7.9-40%). The mean time in frame was 8 months (range, 4-14 months), and the average healing index was 48 days per cm of lengthening (18-110 days per cm). Minor complications (pin tract infection and transient joint stiffness) were common, and after excluding the latter the overall complication rate was 0.6 per bone segment.

Conclusion In our series, limb lengthening of up to 40% of the initial length of the bone segment can be achieved without significant long-term sequelae. However, the procedures were complex and prolonged, and required a special psychological approach directed at both parents and the patients. Complications are quite common, for which patients have to be well prepared before starting the procedures.

Introduction

Limb lengthening aimed at increasing stature\(^1\)\(^-\)\(^3\) is a topic of great current interest and one that triggers debate and controversy, especially in patients with constitutional short stature.\(^3\)\(^1\) A growing number of patients have been requesting this treatment to increase their height, but there is still no consensus on the indications.

In the current study, we reviewed the results of distraction osteogenesis of short-stature patients in our centre and analysed the outcome, including the amount of lengthening, time in the frame, healing index, and complications.

Methods

Between 1995 and 2006, eight patients with short stature underwent distraction osteogenesis using the Ilizarov apparatus or the Orthofix fixator (Orthofix SRL, Verona, Italy) at the Duchess of Kent Children’s Hospital, Hong Kong.

The aetiology of short stature included: achondroplasia (3 cases), constitutional short stature (3 cases), and hypochondroplasia (2 cases). There were six males and two females, with a mean age of 20 years (range, 9-39 years) at the time of operation.

Every patient and their parents were interviewed by the in-charge orthopaedic surgeon before recruitment, and the details of the whole procedure including its duration and possible complications were explained during the interview. Each patient with constitutional short stature was interviewed by the clinical psychologist to ensure that they really needed the procedure. The clinical psychologist was specifically instructed to counsel the patients to live with the short stature and explain that the surgical procedure...
用單邊及環形外固定支架為身材矮小的病人進行肢體延長手術

目的 探討本中心為身材矮小的病人進行牽張成骨術的結果，及分析包括併發症的術後結果。

設計 回顧研究。

安排 香港一所大學教學醫院。

患者 1995年至2006年期間，8位身材矮小的病人接受單邊或環形外固定支架的肢體延長手術。病人中，3例屬軟骨發育不全，3例屬特質性矮小，2例屬軟骨發育過低。

結果 病人接受手術的平均年齡為20歲(介乎9至39歲)。使用的固定支架有Ilizarov或Orthofix兩種。手術後，每個骨段平均有5.2 cm的增長(介乎3.2至8.0 cm)，而長度的增長比例平均為21% (介乎7.9至40%)。施以外固定支架平均需時8個月(介乎4至14個月)，平均癒合指數為每厘米48天(介乎18至110天)。出現輕微併發症(鋼釘感染及短暫關節僵直)的情況很普遍。撇除輕微併發症，總併發率為每個骨段0.6。

結論 本研究顯示肢體延長最高可達至原本骨長度的40%，且沒有長期後遺症。不過，由於手術複雜且需時較長，因此有需要向病人及其親屬施用特別的心理輔導。由於出現併發症的情況相當普遍，病人在進行手術前需有充足的心理準備。

Results

The results of our eight cases are summarised in Table 3. The average gain in length per bone segment was 5.2 cm (range, 3.2-8.0 cm); for the tibia it was 5.2 cm per bone segment and for the femur 5.4 cm per bone segment.

The overall average percentage of shortening was 21% (range, 7.9-40%). For both the tibia and femur, the average lengthening was 21%. The mean time in the frame was 8 months (range, 4-14 months). We excluded one patient with premature removal of external fixators over the femurs, which resulted in fracture shortly after the removal of implant; the fracture was treated by intramedullary nailing with good healing thereafter. The average healing index was 48 days per cm of lengthening (18-110 days per cm). Again, we excluded the above-mentioned patient with premature implant...
removal when calculating the healing index. The average healing index was lower in patients of 16 years or younger than those who were older (43 vs 53 days per cm, after excluding data from an outlier and a patient with septic arthritis). Patients with bone dysplasia had a lower healing index compared to those with constitutional short stature (37 vs 63 days per cm, after excluding data from the same subjects). Notably, the number of patients in our series was small and those with bone dysplasia were also younger.

After excluding grade 1 complications, there were a total of 17 complications (0.6/segment). Twelve of them were grade 2 (0.4/segment), four were grade 3 (0.1/segment), and one was grade 4. Those who were younger (≤16 years) had fewer complications compared to older patients (0.5 vs 1.0/segment). Constitutional short-stature patients had more complications than those with bone dysplasia (1.0 vs 0.4/segment).

Pin tract infection was the commonest complication. According to the grading system (Table 2), grade 1 infection was noted in almost every bone segment and resolved with local dressings and/or oral antibiotics. Grade 2 infections occurred in four bone segments (0.1/segment), and grade 3 infections in three (0.1/segment). No grade 4 infection or chronic osteomyelitis was encountered.

Joint contracture, another common complication, was defined as loss of motion of more than 10 degrees in the sagittal plane. There were five patients with ankle contracture and four with knee contracture; all but two of these resolved with conservative treatment including stretching exercise and splintage. One of the exceptions had 10 degrees of left ankle equinus (case 4), and another had limited right knee range of motion (ROM) [0–110 degrees] secondary to septic arthritis of the knee (case 1). Both patients were functionally good and without a gait problem. Every patient received physiotherapy for range-of-motion exercise during the procedure, and customised splintage to maintain the ROM and minimise the need for Achilles tendon–lengthening surgery. The liability to joint contracture was greater in patients with greater percentage lengthening.

Four patients developed transient common peroneal nerve neuropraxia (0.2/segment), and presented as toe extensor weakness (1 with grade 0/5, 1 with grade 3/5, 2 with grade 4/5) with or without numbness over the relevant territory. These all occurred during lengthening of the tibia and at least 2 weeks after the procedure. None of these patients

TABLE 1. Classification of complications (Donnan scheme)\textsuperscript{4}

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
<th>Examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>Of no long-term functional or anatomical significance, no surgery or anaesthesia required</td>
<td>Mild contractures which responded to physiotherapy, Fixator problems, Stress fractures, Mild behavioural disturbances</td>
</tr>
<tr>
<td>II</td>
<td>Need anaesthesia or operation to correct, but no long-term significance</td>
<td>Insertion of further wires or screws, Soft-tissue deformity requiring tendon release, Bony deformity requiring manipulation of callus, Displaced or unstable fractures requiring fixation, Open or closed osteoclasis of regenerate, Re-excision of fibula, Angulation of &gt;10º in femur, &gt;5º in tibia</td>
</tr>
<tr>
<td>III</td>
<td>Significant functional or anatomical problem which spontaneously improves or correctable by surgery</td>
<td>Failure of length gain, Reducible joint subluxation, Transient nerve injury, Angulation of &gt;15º in femur, &gt;10º in tibia</td>
</tr>
<tr>
<td>IV</td>
<td>Irremediable by conventional treatment</td>
<td>Osteomyelitis/septic arthritis, Subluxation/dislocation, Permanent nerve injury, Irreversible psychological disturbances</td>
</tr>
</tbody>
</table>

TABLE 2. The grading of pin tract infection

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Responds to cleaning and/or oral antibiotics</td>
</tr>
<tr>
<td>2</td>
<td>Responds to intravenous antibiotics</td>
</tr>
<tr>
<td>3</td>
<td>Debridement and/or removal of pins</td>
</tr>
<tr>
<td>4</td>
<td>Chronic osteomyelitis</td>
</tr>
</tbody>
</table>

FIG 2. X-rays showing bilateral tibial lengthening using (a) Ilizarov method and (b) its result
had a valgus deformity of the knee, which is a known risk factor for this nerve injury that seems to be associated with extensive lengthening rather than the subject’s age or diagnosis. Distraction was withheld once neuropaxia was discovered, and in two patients it was resumed after nerve recovery. In one patient, neuropaxia was related to close proximity of the pin to the common peroneal nerve, and the problem was resolved after the pin site was changed. In the remaining three cases, there was no obvious identifiable technical error or clinical suspicion of an anterior compartment syndrome. None of these patients underwent surgical decompression and all recovered with conservative treatment.

Three patients having lengthening (1 for a femur, 1 for a tibia, and 1 for a fibula) developed premature consolidation (0.11/segment), despite the fact that all patients started distraction at day 7 and at a rate of 1 mm/day. One occurred at week 3, one at week 4, and one at month 3 after the start of the procedure. When we reviewed the cases, one of them appeared related to ceasing distraction for a few days after discovery of common peroneal nerve neuropaxia. The other two cases had no identifiable cause, there being no failure of the construct or compression at the osteotomy site. All of them underwent a second operation (an osteotomy). This problem did not seem to be related to the patient’s age or diagnosis.

Two patients developed proximal migration of the distal fibula (0.07/segment) which was addressed by a second operation involving insertion of syndesmotic screw. This occurred in the first two patients in which the syndesmotic screw was not inserted routinely for tibial lengthening.

One patient (case 1) developed leg length discrepancy (0.04/segment) of 1.15 cm. That patient also developed septic arthritis of his right knee. We therefore ceased the distraction on the right side, but continued distraction on the other side for 2 more weeks, since there was uncertainty as to how much more we could achieve on the affected side. Also, no premature consolidation was evident on the unaffected side. The septic arthritis subsided after arthroscopic lavage and antibiotics. On reviewing the case, it may have been related to the close proximity of one distal femur pin to the knee joint, although there was no gross infection or loosening over that pin site when the septic arthritis was diagnosed. During follow-up, the patient was asymptomatic and walked with a normal gait.

One patient (case 2) had bilateral femoral fractures (0.07/segment) due to premature removal of external fixator, and later underwent intramedullary nailing. At the time the fixators were removed, that patient’s healing over three cortices was borderline (as shown in X-rays), but he insisted on early removal despite the risk of fracture having been explained. Also, his femoral braces were ineffective, and finally he fractured both femurs after a fall injury in school.

One patient (case 6) had slow healing over right femur due to concurrent treatment with bone grafting; marked difference in time in frame for the two sides was resulted.

TABLE 3. Summary of demographic and clinical features of the eight patients

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex/age at diagnosis</th>
<th>Segment</th>
<th>External fixator</th>
<th>Gain in length (cm)</th>
<th>% of lengthening</th>
<th>Time in frame (months)</th>
<th>Healing index (days/cm)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1*</td>
<td>M/36 (constitutional)</td>
<td>Femur</td>
<td>Orthofix</td>
<td>3.2 (R) / 3.8 (L)</td>
<td>11.8 (R) / 8.3 (L)</td>
<td>11.7 (R) / 7.2 (L)</td>
<td>110 (R) / 57 (L)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tibia</td>
<td>Orthofix</td>
<td>4.6 (R) / 5.2 (L)</td>
<td>13.5 (R) / 15.5 (L)</td>
<td>7.8 (R) / 9.0 (L)</td>
<td>51 (R) / 22 (L)</td>
</tr>
<tr>
<td>2*</td>
<td>M/14 (constitutional)</td>
<td>Femur</td>
<td>Orthofix</td>
<td>3.5</td>
<td>7.9</td>
<td>9.8 (R) / 11.0 (L)</td>
<td>49</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tibia</td>
<td>Orthofix</td>
<td>3.6</td>
<td>11.4</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>M/11 (achondroplasia)</td>
<td>Femur</td>
<td>Orthofix</td>
<td>6.1</td>
<td>30.5</td>
<td>13.6 (R) / 13.0 (L)</td>
<td>67 (R) / 64 (L)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tibia</td>
<td>Orthofix</td>
<td>5.3</td>
<td>22.8</td>
<td>5.0 (R) / 8.0 (L)</td>
<td>28 (R) / 34 (L)</td>
</tr>
<tr>
<td>4</td>
<td>F/39 (constitutional)</td>
<td>Tibia</td>
<td>Orthofix</td>
<td>6.5</td>
<td>19.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>M/15 (achondroplasia)</td>
<td>Tibia</td>
<td>Ilizarov</td>
<td>4.8</td>
<td>27.4</td>
<td>7.8 (R) / 6.4 (L)</td>
<td>49 (R) / 40 (L)</td>
</tr>
<tr>
<td>6†</td>
<td>M/11 (achondroplasia)</td>
<td>Femur</td>
<td>Orthofix</td>
<td>8.0</td>
<td>40.0</td>
<td>14.5 (R) / 6.5 (L)</td>
<td>54 (R) / 24 (L)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tibia</td>
<td>Orthofix</td>
<td>5.8</td>
<td>36.4</td>
<td>4.4 (R) / 4.4 (L)</td>
<td>23 (R) / 23 (L)</td>
</tr>
<tr>
<td>7</td>
<td>M/11 (hypochondroplasia)</td>
<td>Tibia</td>
<td>Orthofix</td>
<td>6.7</td>
<td>31.5</td>
<td>4.1</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Femur</td>
<td>Orthofix</td>
<td>7.7</td>
<td>25.6</td>
<td>7.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tibia</td>
<td>Orthofix</td>
<td>6.3</td>
<td>20.1</td>
<td>5.4</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>F/27 (hypochondroplasia)</td>
<td>Tibia</td>
<td>Ilizarov</td>
<td>3.3</td>
<td>12.1</td>
<td>6.5</td>
<td></td>
</tr>
</tbody>
</table>

* He developed right-knee septic arthritis; difference in gain in length and time in frame on the two sides
† He had premature removal of the external fixator of both femurs, which resulted in fracture afterwards and required intramedullary nailing
‡ He had slow healing over right femur due to concurrent treatment with bone grafting; marked difference in time in frame for the two sides was resulted
Discussion

Short stature generates difficulties and problems in three respects: physical, emotional, and social. Daily activities such as conducting business at counters and using a public toilet become affected. Such individuals often feel ‘different’ within their family or social circle, resulting in emotional disturbance. The social factors associated with short stature lead to development of an inferiority complex. Furthermore, in many countries dominance is often associated with tall stature, which is seen as an expression of strength, power, good health, and success.

On the other hand, a limb lengthening procedure is invasive, complex and prolonged, and requires a special psychological approach on the part of both the patients and their families. Complications are not uncommon, for which each patient should be well prepared before starting the procedure.10-22

From our experience, distraction osteogenesis is a useful and effective method of limb lengthening in short-stature patients. Some authorities report lengthening as much as 40 to 70% of the initial bone length, but most current studies report an average gain in length of 4 to 6 cm or 8 to 22% of the initial bone length.10-12 In our series, lengthening of up to 40% of the initial length of the bone segment could be achieved without significant long-term sequelae.

The average time in the frame was 8 months (4-14 months) and the average healing index was 48 days per cm of lengthening (18-110 days per cm), which was faster in younger subjects.

In our series, minor complications like pin site infection and transient joint stiffness were common, and after excluding the minor Donnan grade 1 complications, the overall complication rate was 0.6 per bone segment. Patients undergoing limb lengthening for short stature experience more complications than those with leg length discrepancy, especially those undertaking bilateral femoral and tibial lengthening (actually involving four procedures performed simultaneously). Older patients (>16 years), constitutionally short patients, and persons undertaking extensive lengthening were more prone to complications. Complications such as proximal migration of the distal fibula can be avoided by insertion of a syndesmotic screw during the lengthening procedure. Despite all our patients eventually enjoying an improved quality of life at the end of the procedure, the high complication rate was clearly explained to all of them and their parents, and the decision to have the procedure was not undertaken lightly.

References