LOWER LIMB LENGTHENING AND DEFORMITY CORRECTION: PHYSIOTHERAPY PATIENT MANAGEMENT - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Patients undergoing limb lengthening and deformity correction of the lower limb require physiotherapy input to commence preoperatively, and this should continue postoperatively until all goals have been achieved.
- The aim of physiotherapy includes:
  - Maintain joint range of motion
  - Minimise contractures
  - Minimise loss of muscle power
  - Education on a home exercise program and splinting
  - Ensure safe mobility and maximise function
- Specific treatment techniques for patients undergoing limb lengthening and deformity correction are outlined in this practice guideline.
- Physiotherapy is an important part of any successful limb lengthening procedure.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
Date Effective:	1st November 2018	**Review Period**: 3 years
Team Leader:	Orthopaedic Physiotherapist	**Area/Dept**: Physiotherapy CHW

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CHANGE SUMMARY

- Literature review performed
- Title changed
- Background
- Referral policy
- Assessment
- Treatment
- Contra-Indications and Precautions
- Prognosis and Outcomes
- References

READ ACKNOWLEDGEMENT

- This document is aimed at physiotherapists working within the physiotherapy Department at the Children's Hospital at Westmead.
- All physiotherapists working primarily in the orthopaedic and outpatient teams are required to read and acknowledge they understand the contents of this document.
- Junior physiotherapists will be supervised by the senior orthopaedic or outpatient physiotherapists prior to independent management of patients undergoing limb lengthening and deformity correction procedures.

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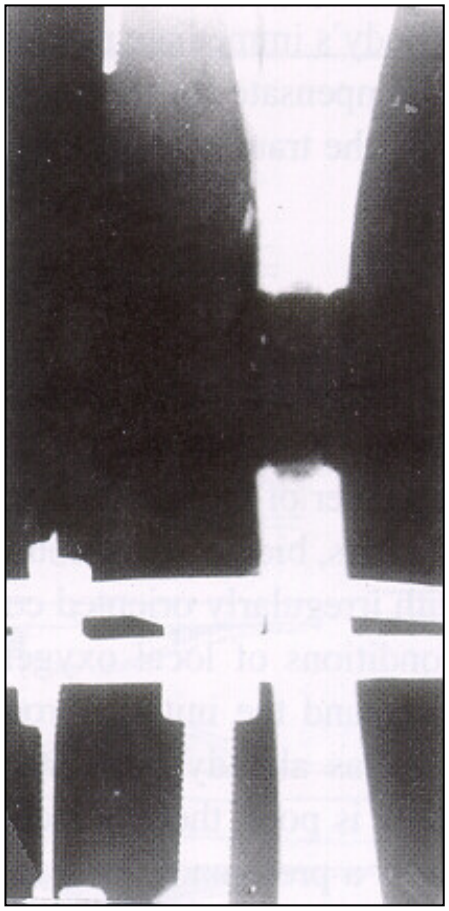
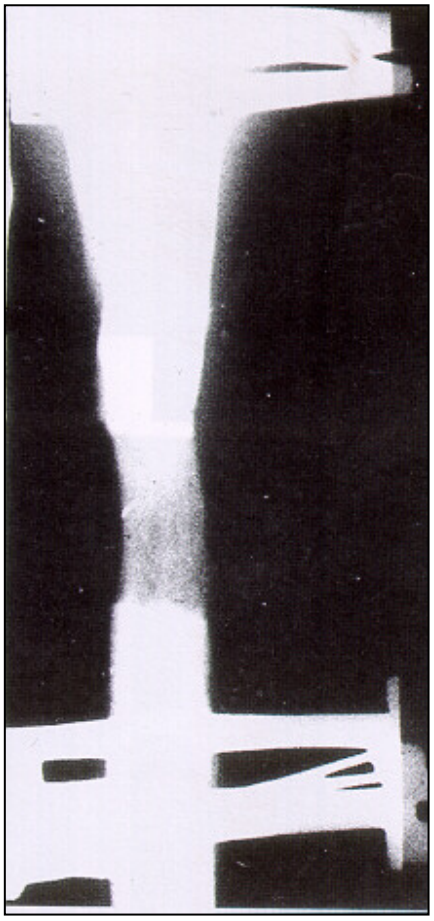
1 Background

Surgical limb lengthening was first reported in 1905 in Italy. Advancements in lengthening and deformity correction have continued since this date however the principle of distraction osteogenesis remains consistent. A lengthening device is used to create an environment in which distraction osteogenesis can take place.

Distraction osteogenesis is a form of direct membranous ossification. Under optimal conditions, the bone regenerates by intramembranous ossification, without a cartilage intermediary. A fibro vascular lattice spans the two ends of bone that are undergoing distraction. Bony trabeculae orient longitudinally in the lattice to form a fibrous-like interzone. This interzone is composed of primitive mesenchymal cells that differentiate into osteoblasts, which form the bone. During the period of neutral fixation, cortex covered with periosteum gradually forms and surrounds the regenerate zone[2].





Picture 1 and 2 – Distraction osteogenesis

The biological and clinical principles, described by Professor Ilizarov[3], that are important to the proper formation of new bone within a widening distraction gap include the following:

1. Preservation of the periosteum, endosteum and bone marrow; this maintains the adequacy of blood supply to the regenerate.

2. Stable external fixation.
3. A delay or latency period before distraction.
4. A slow rate of distraction at a high frequency rhythm, important for maintaining the quality and quantity of osteogenesis, specifically 1mm per day divided into 4 times per day.
5. Functional use of the limb, including functional loading or weight bearing. The combination of muscle pull and weight bearing facilitates the formation of callus and the ossification of the regenerate. In the case of regeneration of bone, this stimulating effect of weight bearing is in accordance with Wolff's law of functional transformation of bone.
6. A period of neutral fixation after distraction to permit the regenerate bone to fully ossify, this period is called the consolidation phase.

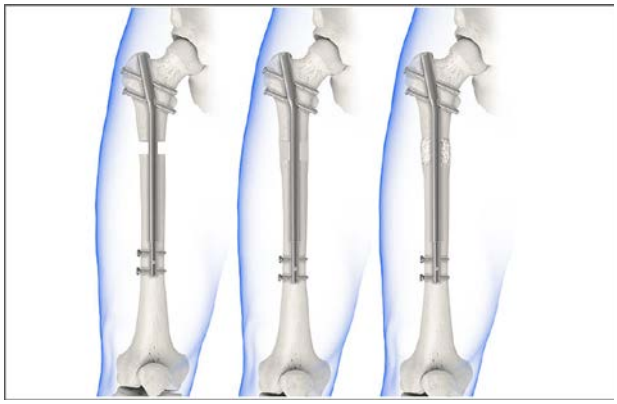
The lengthening process involves three phases:

1. Osteotomy and latency phase - Corticotomy or osteotomy is performed followed by application/insertion of lengthening device. There is then a latency period of approximately 5 to 7 days before distraction starts.

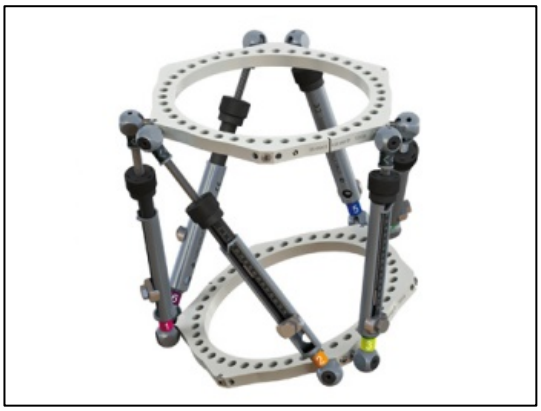
2. Distraction phase - Lengthening is usually carried out at a rate of 1 mm per day at 3 increments of 0.25mm each until the desired length is achieved.

3. Consolidation phase - Neutral fixation and consolidation takes place with lengthening device in situ. The device is removed when a cortex has formed on the new bone[4], three out of four cortices are required before the frame is removed if external fixation is the form of correction being used. If intramedullary lengthening is being used the nail will remain in situ until sufficient bone healing has occurred (usually 12 months post-insertion).

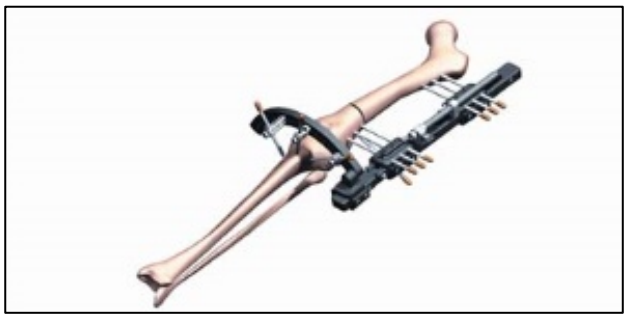
Current limb lengthening and deformity correction devices used at The Children's Hospital at Westmead include Precice intramedullary nail, Hexapod frames (Taylor Spatial Frames, TL-Hex frames) and Monolateral external fixation/rail.



Picture 4 – Hexapod frame



Picture 3 – Precice intramedullary nail



Picture 5 – Monolateral external rail

The frames are capable of correcting deformities in three planes (coronal, sagittal and rotational) simultaneously whilst lengthening.

When lengthening is only required in one plane intramedullary lengthening is used if possible to reduce potential complications from pin site infections and increase patient comfort.

2 Referral policy

A *blanket referral* applies for all patients who undergo limb lengthening and deformity correction. All patients should have a pre-operative physiotherapy assessment when possible and treatment when indicated. Patients should be seen twice daily whilst they are an in-patient beginning day one post operatively.

The patients will be seen by the physiotherapist when they attend follow-up appointments with the orthopaedic surgeon in the limb lengthening clinic and will participate in hydrotherapy sessions once per week. Out-patient physiotherapy should be organised by the treating physiotherapist pre-operatively at the local hospital or private practice in addition to physiotherapy at limb lengthening clinic.

3 Assessment

3.1 Pre-operative Assessment

3.1.1 Subjective Examination

A subjective history should be taken pre-operatively, but can be taken post-operatively if the patient was not seen by the physiotherapist prior to surgery.

The information that should be collected includes:

- Current and past medical history, including the cause of the limb deficiency or bony deformity.
- Previous surgery.
- Current mobility status and any type of walking aid or orthotic previously utilised.
- Social history including home set-up and stairs, school set-up (e.g. stairs, room changes), transport to and from school.

It is important to find out if the patient is already treated by a local physiotherapist. If so contact the therapist and inform them of the upcoming surgery and postoperative protocol. If they do not currently have a local physiotherapist then a **referral must be organised**.

3.1.2 Objective Examination

- Bilateral assessment of each of the following:
    - Active and Passive range of motion (ROM) of the hip, knee and ankle
    - Muscle length[5];
        - Femoral lengthening: Quadriceps, Hamstrings, Hip flexors, Tensor fascia lata / Iliotibial Band and Hip adductors
        - Tibial lengthening: Gastrocnemius/ soleus, Toe flexors, Hamstrings, Tibialis posterior, Tibialis anterior, Quadriceps
    - Muscle strength
    - Gait analysis
    - Leg length
    - Joint stability
    - Sensation

3.2 Post-operative Assessment

3.2.1 Subjective Assessment

A subjective history should be taken if the patient was not seen by the physiotherapist prior to surgery as above.

Confirm the social history for any changes to home or school environment.

Confirm that local outpatient physiotherapy is organised.

3.2.2 Objective Assessment

- Bilateral assessment of each of the following:
    - Active and Passive range of motion (ROM) of the hip, knee and ankle
    - Muscle length[5];
        - Femoral lengthening: Quadriceps, Hamstrings, Hip flexors, Tensor fascia lata/Iliotibial Band and Hip adductors
        - Tibial lengthening: Gastrocnemius/ soleus, Toe flexors, Hamstrings, Tibialis posterior, Tibialis anterior, Quadriceps
    - Muscle strength
    - Sensation

4 Treatment

4.1 Preoperative treatment

The patient and family should be taught stretches and strengthening exercises pre-operatively that they will be expected to perform post-operatively. The exercises should commence pre-operatively if the child has a pre-existing muscle contracture or muscle weakness[1,5,6]. The patient should also be taught how to mobilise with crutches or a frame and referred to their local physiotherapist. Measuring for appropriate splints should also be done at this time.

4.2 Post-Operative Treatment

4.2.1 Orthotic Intervention

- The treating physiotherapist will play a key role in ensuring timely orthotic intervention to prevent muscle contracture and optimise mobility.
- The physiotherapist should also monitor the ongoing compliance and appropriateness of these orthotics.

Recommendations for orthotic intervention are listed below but may vary according to patient characteristics and surgeon preference.

Correction Device	Recommended Orthotic Intervention
Tibial External Fixator	Foot Plate fitted day 1 *Knee Extension splint only significant concerns regarding knee ROM.*
Tibial Precice Nail	Hinged AFO with DF straps. (Moulded pre-operatively) *Wraparound for knee extension, only if significant concerns regarding knee ROM/hamstring length.*
Femoral Precice Nail	Wraparound for knee extension (Measured pre-operatively with order sent to Connie), or KAFO (moulded pre-operatively) if significant concerns. *Hinged AFO only if significant Ankle ROM concerns.*

4.2.2 Physiotherapy Aims

The aims of physiotherapy management in this group of patients will change during the treatment phases. Overall the aims of physiotherapy in these patients include[5,6]:

1. To maintain joint range of motion.
2. To minimise contractures/ Maintain muscle length.
3. To maintain muscle power.
4. To educate patients/ carers in a home exercise programme.
5. To educate patients/ carers about the importance and correct use of splints[4].
6. To ensure functional loading and use of the limb as appropriate.

Treatment should start on the first post-operative day. Whilst an in-patient the child should be seen by the physiotherapist twice per day[5,7]. Adequate analgesia should be given prior to treatment.

4.2.3 Physiotherapy Techniques

Treatment techniques to achieve the above aims include[5,7]:

- Active ROM exercises
- Active assisted ROM exercises
- Passive ROM exercises
- Stretches
- Strengthening exercises

- Weight bearing exercises where appropriate
- Gait re-education
- Hydrotherapy (three weeks post-operatively after wound review)

Exercises	Femoral lengthening	Tibial lengthening
Active and Active assisted ROM exercises	Knee extension Hip extension Knee flexion Hip abduction Hip flexion	Knee extension Knee flexion Ankle dorsiflexion Ankle plantarflexion Toe flexion Toe extension
Stretches	Quadriceps Hamstrings Hip flexors Tensor fascia lata/ITB Hip adductors	Gastrocnemius/ soleus Toe flexors Hamstrings Tibialis posterior Tibialis anterior Quadriceps
Strengthening exercises	Hip extensors Hip abductors Quadriceps Hamstrings Back extensors Abdominals	Ankle dorsiflexors Tibialis anterior Quadriceps Hamstrings

- Static stretches held for the duration of 30 seconds have been shown to be the optimum time for increasing flexibility[8].
- Strengthening should begin day one to minimise any loss of strength/power and ROM.
- Out-patient physiotherapy should start within five days of the patient's discharge from hospital[5].
- Outpatient physiotherapy and home exercise programs should focus on aggressive stretching during the distraction phase.

Whilst distracting, the bone lengthens but the muscle and tendons take more time to adjust to a new length. This is the period in which the joints may dislocate secondary to the pull of the shortened muscles. It is of upmost importance to maintain range of motion and stretch muscles during the distraction phase. Close monitoring is needed and the treatment

frequency may need to be increased. Treatments should never be decreased based on pain or pin site infections, unless specified by the doctor.

The following problems will require an increase in the frequency of out-patient treatment[5]:

- Moderate / major loss of ROM
- Increased muscle contraction
- Poor compliance with out-patient physiotherapy/ home exercise program
- Difficulty in weight bearing if patient is allowed to weight bear.

4.3 Consolidation phase

- Emphasis is on regaining any loss of range of movement and increasing functional activities.
- Weight bearing should be maximised when external fixation insitu. When Precice nail lengthening weight bearing is as per doctors instructions, according to manufacturer guidelines.

Precice Nail Diameter	Recommended load limit
8.5mm	25kg
10.7mm	50kg

- Patients should continue with progressive strengthening exercises using resistance, body weight and functional activities[5] as appropriate.
- During the consolidation phase, emphasis is now on regaining full range of motion and strengthening.
- During this phase activities should focus on the following areas[5,6]:
    - when external fixation lengthening has been performed:
        - Full weight bearing activities
        - Single stance activities
        - Proprioception
        - Sport (non-contact)
        - Cardiovascular fitness
        - Increasing endurance
    - when Precice nail lengthening has been performed:
        - Partial weight bearing as per doctors instructions
        - Re-gain any loss of range of motion
        - Strengthening

- Cardiovascular fitness (upper limb activities/non-weight-bearing)

5 Contra- Indications and Precautions

Hydrotherapy is contra-indicated if the patient has a deep pin site infection.

5.1 Complications

Reports in literature range from 3-200% depending on the criteria used for what is a complication and the type of device used. Prince et al[13] found no differences in demographic, radiographic or patient-based outcome measures were found between patients who had complications or problems compared to those without complications.

The most acceptable criteria separate the complications into three categories depending on the severity[9].

1. **Problem** – Difficulties that do not require operative interventions to resolve (e.g. Pin site infection, wound infection)
2. **Obstacle** – Difficulties that do require operative interventions (e.g. Pin loosening requiring exchange of pin, loose strut that can be tightened in clinic, fixator adjustment under GA, nerve decompression or bone grafting.)
3. **Complication** – True complications represent intra-operative injuries and or problems that do not resolve before the end of treatment (e.g. Nerve injury, joint dislocation or stiffness, fracture of regenerate after removal of fixator.)

5.1.1 Muscle contractures

A result of the tension generated on the muscle due to distraction. This tends to occur to the overpowering muscles. Most frequently involved muscles are those that cross two joints as these have fibres of various lengths.

Significant loss in range of motion has been shown to occur in the latent period prior to lengthening. Therefore the importance of stretches should be emphasised in this phase in order to reduce muscle related complications that could happen during limb lengthening[10,11].

Treatment for muscle contractures can include stretching, splinting, serial casting, botox injection, manipulation under anaesthetic and in extreme circumstances surgical release[13]

5.1.2 Joint subluxation/ dislocation

The most common predisposing factor is pre-existing joint instability usually due to congenital causes. The imbalanced muscle tension caused by distraction may lead to subluxation[12]. Correction may be slowed or ceased until this is resolved. Maintaining range of motion is of upmost importance in this group of patients.

5.1.3 Axial deviation

The limb segment being lengthened may gradually deviate due to the imbalance of muscle forces on different sides of the bone. The direction of deviation depends on the bone involved and the level of the osteotomy. It can also be caused by instability from inadequate construct, loss of tension or loosening of the pins[12].

5.1.4 Nerve Injury

There is a risk of nerve injury due to the following reasons[9,12]:

- Placement of the pins when external fixation has been used.
- As a result of the corticotomy and the surgical technique e.g. direct injury from the surgical instruments.
- Compartment syndrome.
- Rate of distraction.
- Correction of deformity, e.g. when correcting a patient from valgus into varus, the peroneal nerve will undergo stretching during the correction and may need to be decompressed.

5.1.5 Vascular Injury

There is a risk of vascular injury due to numerous reasons[12]:

- Damage can occur from pin insertion at the time of surgery when external fixation is used. If an artery and vein are perforated simultaneously an arteriovenous fistula can form, but this is extremely rare.
- From the surgical instruments.
- Rapid or excessive distraction can lead to hypertension.

Vascular damage may cause displacement of the osteotomies and a haematoma may lead to compartment syndrome. Deep Vein Thrombosis is possible but rare. Oedema can occur which can take several months after removal of the frame to disappear.

5.1.6 Premature consolidation

Failure of the osteotomy to open after initial distraction can occur due to an excessive latency period allowing significant callus healing to block the distraction of the osteotomy. It can also be due to incomplete osteotomy at the time of the surgery. Continued distraction can be carried out until the consolidation bridge of bone ruptures, a painful pop is usually experienced by the patients and then the lengthening will begin. Alternatively a closed rotational osteoclasis or a repeat corticotomy may need to be performed[12].

Premature consolidation is common in Ollier disease and the speed of lengthening is increased to avoid this occurring.

5.1.7 Delayed consolidation

Delayed consolidation can result from[9,12]:

- A traumatic corticotomy
- Frame instability
- Rapid distraction
- Infection

- Malnutrition
- Vitamin D deficiency

This is treated by either reversing the lengthening and then going forwards one or more times, adding more pins, bone grafting or BMP.

Delayed regenerate healing is usually treated by a prolonged use of the external fixator. Occasionally, the regenerate will be bone grafted with consideration for the use of bisphosphonate treatment.

5.1.8 Pin site problems (External fixation lengthening only)

This relates to pin-skin motion, the amount of soft tissue between skin and bone and the diameter of the pin used. Pin–skin motion and pin-bone motion can be minimised by adequate wire tension. Recalcitrant infections, infections around the wires that pass through joints and cellulitis around a pin site are treated with antibiotics or removal of the offending wire with or without replacement. Infections usually start as superficial and then can develop into deep tissue infections[9,12] or osteomyelitis[13].

Pin site infections are a common complication and will likely occur several times whilst the patient has the fixator on. A pin infection does not mean that the patient or the therapist should alter the physiotherapy program. The parents are instructed to begin antibiotics and the child can benefit from some mild analgesia, but should not stop physiotherapy.

5.1.9 Refracture

Timing of the removal of the lengthening device is extremely important in preventing fracture and subsequent deformity. It should only be removed when a complete cortex on both sides of the lengthened callus is evident on radiographs, three of the four cortices seen on 2 orthogonal x-rays[12]. Refracture after removal of device can result in loss of length previously gained with device [13].

Where external fixation has been utilised a cast brace is often applied for six weeks following external fixator removal to prevent refracture.

5.1.10 Joint stiffness

Joint stiffness is often a late complication. This is reported to increase when bone is lengthened by 15% or greater[12].

5.1.11 Pain

Pain is the most common complaint but should subside in the first couple of weeks. It often increases with the number of osteotomies[9].

Should the patient have a lot of pain during therapy, the therapist should contact the doctor to inquire about muscle relaxants and mild analgesia.

The following problems should be checked for and if present will require referral back to the consultant[12]:

- Significantly decreased Range of Motion (i.e. loss of full knee extension or loss of knee flexion to less than 45-50 degrees)

- Joint subluxation
- Loss of sensation
- Neurogenic pain
- Pin/wire breakage or problems with the frame
- Sudden increase in pain on weight-bearing
- Loss of extensor hallucis longus
- Severe pin site infection

6 Prognosis and Outcomes

Discharge from hospital post-operatively should occur once the patient can carry out the following activities[5]:

- Safe with functional transfers.
- Mobilising safely with an appropriate aid.
- Safely negotiate (up and down) stairs.
- Have a reasonable amount of ROM.
- Understand and be able to complete home exercise program and have outpatient therapy organised for follow up.
- Correctly apply appropriate splints.

Patients should not be discharged from out-patient physiotherapy until the following criteria has been fulfilled[5]:

- Maximal ROM has been achieved.
- Maximal weight-bearing is occurring.
- Maximal function has been attained.
- Optimal gait has been achieved.

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