Using External Fixation With Pediatric Oncology Patients: How Did We Get Here?

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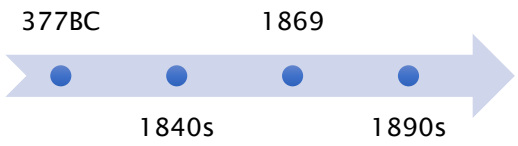
Objectives

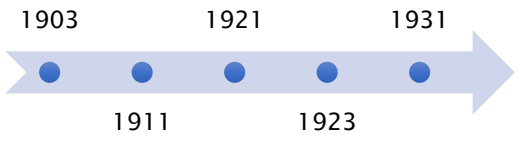
- Detail the history of external fixation and discuss the patient population in which these devices are most commonly used.
- Describe the types of external fixation systems, apply the current evidence regarding external fixators, and understand the pathophysiology of limb lengthening through the use of these devices.
- Identify the importance of physical therapy in optimizing function and quality of life in the pediatric oncology patient with an external fixation system.
- Explain the complex nature of evaluating and treating these patients across the continuum of care.

History of External Fixation

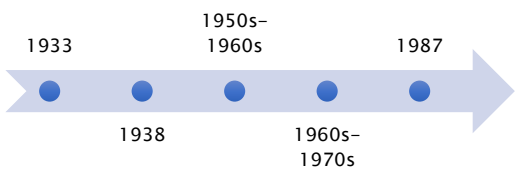
History[1–4]



History[1–4]



History[1–5]



Components of an External Fixation System

Pins[6–8]

- Play an important role in stability of fixation system
- Types of pins
  - Half-pins
  - Transfixion pins
  - Tapered pins
  - Conical pins
  - Hydroxyapatite (HA) coated pins
- Pin diameter should not exceed one-third of the diameter of the bone

Wires[8,9]

- Typically utilized for ring fixators
- Always applied under tension
- Smaller diameter wires allow for controlled micro-motion to promote callus formation

Rods[6,7]

- Act as link between bone and fixator system
- Materials utilized
  - Stainless steel
  - Aluminum alloy
  - Carbon fiber
- Telescopic rods can span longer distances

Rings[6,8]

- Types of rings
  - Full (closed)
  - Partial (open)
  - Arches
- Stability depends on diameter and thickness of ring
- Important to maintain space between ring and skin

Clamps[7,8]

- Connect pins and wires to rods and rings
- Can affect the rigidity of frame based on location and number of pins and wires used
- Types of clamps
  - Simple (single)
  - Modular (universal)

Types of External Fixation Systems

Monolateral Frame[4,10,11]

- Construct: half-pins attach to rod
- Advantages
  - Easier and faster to apply when compared to ring fixator
  - Increased comfort, especially for upper extremity use
- Disadvantages
  - Less versatility than ring fixator
  - When used for fracture management, increased risk of non-union

Bilateral Frame[7]

- Construct: monolateral frame placed on both sides of bone
- Advantages
  - Stiffer than unilateral frames
- Disadvantages
  - More difficult to apply than unilateral frame
  - Increased risk of pin infections

Ring Fixator[4,10,12]

- Construct: tensioned wires and pins connect to rings and rods
- Advantages
  - Offer more versatility and adaptability
  - Can provide greater stability when used in fracture management
  - Can extend across joints as needed
- Disadvantages
  - More complex to assemble and apply
  - Can be more cumbersome or intimidating, especially in pediatric population

Ilizarov Frame[4,6,10]

- Developed by Professor Gavril Abramovich Ilizarov in 1943
  - Transfixion wire ring fixator using metal rings, wires, and rods
- Often used for fracture management and deformity correction
- Attempts to mimic properties of connective tissue

Taylor Spatial Frame[6,13]

- Considered "hexapod" ring fixator
- Uses computer software, which can help with adjustment of frame as needed
- Most useful in treatment of complex and multi-planar deformities

Hybrid Fixator[4,6]

- Construct: combination of monolateral and ring system components
  - Behaves most similarly to monolateral fixator
  - Half-pins replace some tensioned wires
- Most widely used in western countries

Optimal Design[6]

- Rigid enough to manage torsion, bending, and shear
- Flexible enough to allow for axial movement
- Encourage equal loading across bone

Why External Fixation?

Indications for Use[4,14]

- Fractures
  - Open fractures
  - Non-union fractures
- Complex arthrodesis procedures
- Limb deformities
  - Congenital
  - Acquired
- **Distraction osteogenesis (limb lengthening)**

Distraction Osteogenesis (DO)[5,6,15]

- Commonly known as "limb lengthening"
- Corticotomy or osteotomy to allow for formation of new mature bone
- Success of lengthening depends on frame stability or ring block stability (in ring fixators)
  - Number of rings
  - Distance between rings
  - Connections between rings
  - Points of fixation
- Encourages early weight bearing, mobilization, and joint range of motion

Common Diagnoses[15–17]

- Osteomyelitis
- Short stature
- Achondroplasia
- Dwarfism
- Congenital humeral defect
- Acquired deformities with neurologic disorders
- Oncology diagnoses

Phases of Limb Lengthening

Latency Phase[5,18]

- Osteotomy
- Application of apparatus

Distraction Phase[5,18]

- Produces distraction gap between bones
- Typically at a rate of 1mm/day
- Variable duration
  - When optimal lengthening achieved
  - When too many complications or non-compliance issues arise

Consolidation Phase[5,18]

- Newly formed bone (callus) bridges distraction gap and consolidates
- Tends to be longest phase
- Fixator left in place until new bone is determined to be strong enough for removal without bending, fracture, or buckling
- Dynamization is physician-dependent

Dynamization[6,19]

- Gradually increasing load across callus site as bone gains stability
- Destabilization of frame
  - Removing rings, bars, pins, or wires
  - Releasing tension in wires
  - Moves bars away from bone
- May increase risk of delayed union, re-fracture, or deformity development

Dynamization[19]

- Iobst, et al. created an open-ended survey re: use of dynamization before external fixator removal
  - If non-dynamized, 92% used cast post-removal
  - If dynamized, 91% used brace post-removal
- No difference between groups for return to functional activities
- No apparent preference for dynamization versus non-dynamization
  - Variable across surgeons

Removal of Fixator[19]

- Clinical examination and radiographic findings affect decision to remove fixator
  - Clinical: Full weight bearing without pain
  - Radiographic: 2mm thickness across the distraction gap
- Multiple factors affect quality of bone healing

Factors that Affect Bone Healing[18]

- Age
- Underlying etiology
- Comorbidities
- Construct stability
- Amount of bone lengthening
- Length and location of bone
- Location of osteotomy

Lengthening of the Upper Extremity[17]

- Minimal literature
  - Length discrepancies less common and less bothersome
  - Usually performed for cosmetic purposes
- Typically monolateral frame
  - Circular frame requires constant shoulder abduction

Lengthening of the Forearm by the Ilizarov Technique (Villa, et al.)[20]

- 11 out of 12 patients reported the following:
  - Functional improvement, including increased range of motion, improvements in writing, swimming, sports, and play
  - Cosmetic improvement
  - Psychological improvements
- ~92% said "they would do it again"

Complications Related to Limb Lengthening

Muscle Contractures and Joint Stiffness[5,21]

- Muscle contractures
  - Most common in muscles that span two joints
  - Consider splinting to decrease risk of knee or ankle contractures
  - May require surgical intervention following fixator removal (e.g. tendon lengthening)
- Joint Stiffness
  - May be related to persistent muscle contractures, increased pressure on joint surface during lengthening, or scar tissue formation
  - May require surgical intervention (e.g. manipulation under anesthesia)

Joint Instability and Axial Deviation[5,21]

- Joint instability
  - Subluxation may occur due to pre-existing joint instability or imbalanced muscle tension during lengthening process
- Axial deviation
  - Tendency for limb being lengthened to gradually deviate
  - Can adjust current pins or add additional pin

Neurologic Injury[5,21]

- Corticotomy-related nerve injury
- Pin-related nerve injury
  - Ensure pins are placed in safe zones
- Distraction-related nerve injury
  - Decrease frequency or length of distraction to improve symptoms

Vascular Injury[21]

- Can be related to surgery or to distraction
  - Direct vascular damage from corticotomy or osteotomy
  - Fistula formation
  - Compartment syndrome
  - DVT
  - Hypertension
  - Edema

Consolidation Issues[5,21]

- Premature consolidation
  - Early callus formation blocks distraction
  - Most commonly seen in femur or fibula
  - May lead to additional surgery (e.g. re-osteotomy)
- Delayed consolidation
  - Can be due to technical factors or patient factors
  - Shorten distraction gap, then gradually begin distraction again
  - May increase wear time of fixator

Pin Site Problems[5,7,21]

- Can be related to a variety of technical or patient factors
- Excessive motion of skin around pin can result in local inflammation and pin site infection
  - Problems develop from outside to inside
  - Gentle compression and clamps can help to reduce motion
- Pin site infection must be treated without delay to avoid ostemylelitis

Other Complications[5,21]

- Re-fracture, typically following fixator removal
- Pain
- Loss of appetite and weight
- Depression

Lengthening with external fixation is effective in congenital femoral deficiency. (Prince, et al.)[30]

- Distraction osteogenesis using monolateral external fixator
- Results
  - Average time in external fixator frame was 206 days
  - Most patients with return of normal knee flexion and hip flexion range of motion by 31.5 months
- Overall patients had good functional outcomes at intermediate follow-up
- Authors found that limb should not be lengthened beyond 6 cm or 25% of relative femoral length

External Fixation in the Oncology Setting

Cancers of the Bone[22,23]

- Most common types of bone cancer in pediatrics:
  - **Osteosarcoma**
  - Ewing sarcoma
  - Chondrosarcoma
  - Histiocytoma
  - Benign bone tumors or cysts
- Cure rates among patients with childhood sarcoma are high
  - 5 year survival ranges from 65-86% (localized disease)

Osteosarcoma treatment[22–26]

- Historically, amputations or joint disarticulation
- Now moving towards limb sparing (mainstay of treatment options)
  - Endoprothesis
  - Allografts or allograft expandable composites
  - Expandable prostheses
  - Vascularized bone grafts
  - Arthrodesis
  - Rotationplasty
  - External fixation

Complications of External Fixation[26]

- Length of reconstruction
- Infection
- Tumor cell activation
- Systemic chemotherapy and adjuvant radiotherapy
- Psychological considerations

Review of Evidence for DO in the Oncology Setting

Reconstruction of defects following bone tumor resections by distraction osteogenesis (Erler, et al.)[25]

- Classified distraction osteogenesis into five types
  - Type 1: diaphyseal reconstruction
  - Type 2: metaphyseal reconstruction
  - Type 3: epiphyseal reconstruction
  - Type 4: subarticular reconstruction
  - Type 5: arthrodesis

Reconstruction of defects following bone tumor resections by distraction osteogenesis (Erler, et al.)[25]

- 9 patients with bone tumors
  - Treatment plan: chemotherapy and surgery
  - Epiphysis preserved in all patients
  - Immediate physical therapy intervention
  - Frame dynamized before removal
  - Frame removed, then cast/splint applied for 4-6 weeks
- Most common complications: pin tract infection, reduced range of motion
- No early consolidation in any patients

Ilizarov fixator in the management of benign, malignant and complicated cases of bone tumors (Kamel, et al.)[24]

- 15 patients with bone tumors
  - Treatment plan: chemotherapy and surgery
  - Regular follow-ups until fixator removed
  - Disease free survival: 22.8 months
- Most common complication: pin tract infections
  - More prominent in patients actively receiving chemotherapy during lengthening process

Reconstruction of large tibial bone defects following osteosarcoma resection using bone transport distraction: a report of two cases (Yang, et al.)[27]

- 2 patients with bone tumors
  - Treatment plan: chemotherapy and surgery
  - Low risk of complications show that DO can be beneficial, even during chemotherapy treatment
  - Long term follow up shows that reconstructed bone found to have good biomechanical performance
- Limitations of external fixator use in oncology population
  - Chemotherapy can inhibit bone formation and lead to callus failure/re-fracture
  - Long duration of wear
  - Patient non-compliance

Biological reconstruction for extremity osteosarcoma: distraction osteogenesis technique. (Matsubara, et al.)[22]

- Case report
  - 17-year-old boy with osteosarcoma of proximal tibia
  - Treatment plan: chemotherapy and surgery
  - Gradual lengthening began after operation
  - 26 month follow up: no evidence of disease and patient reports that he can run without pain

Limb salvage using distraction osteogenesis: a classification of the technique. (Tsuchiya, et al.)[28]

- 19 patients with bone tumors
- Distraction osteogenesis involved 3 different procedures:
  - Bone transport (10 patients)
  - Shortening-distraction (3 patients)
  - Either technique combined with intramedullary nail (6 patients)
- Patients receiving chemotherapy with DO
  - Similar results to patients without chemotherapy
  - Callus successfully distracted 0.8mm/day

Removal of metaphyseal bone tumours with preservation of the epiphysis. Physeal distraction before excision.(Cañadell, et al.)[29]

- 20 patients with primary bone tumors
  - Survival rate: 85% (no local recurrence)
  - Limb function varied based on location of tumor
- Complications: infections, dislocation of graft, peroneal nerve palsy, re-fracture

Psychosocial Implications

Clinical implications of psychosocial factors on pediatric external fixation treatment and recommendations (Richard, et al.)[12]

- During procedure, patients and families commonly experienced:
  - Depression
  - Sleep deprivation
  - Poor academic performance
  - Substance abuse
- Important for psychiatrist or child life therapist to be involved before surgical intervention
  - Mental stability
  - Coping skills
  - Home/social environments

Clinical implications of psychosocial factors on pediatric external fixation treatment and recommendations (Richard, et al.)[12]

- Authors compared outcomes of single-parent and two-parent households
  - Increased stress on single parent leads to greater reliance on child rather than on parents for post-op care
  - Two-parent household can dedicate one parent to child's daily needs
  - Can lead to narcotic use, unplanned re-admissions, increased length of hospital stay, increased number of outpatient visits, increased number of days in fixator

Clinical implications of psychosocial factors on pediatric external fixation treatment and recommendations (Richard, et al.)[12]

- External fixation and limb lengthening must be a team approach for pediatric patients
  - Children require assistance with rehabilitation and daily fixator maintenance
  - Poor family support will negatively affect overall outcomes
  - Unaddressed psychosocial needs have been proven to complicate rehab
  - Complication rates as high as 97% have been reported
    - Can lead to prolonged disruption of "normal life"
- **Education and social support are crucial to success**

Physical Therapy Intervention

Pre-operative Evaluation[5]

- Manual muscle testing
- Range of motion measurement
- Sensation testing
- Circumferential measurement
- Joint stability assessment
- Postural analysis
- Gait assessment
- Functional mobility analysis
- Assistive device training
- Home exercise program and education for patient/family

Case Study

Inpatient Post-operative Evaluation and Intervention[5]

- Assessment and management of pain is imperative
- Ideal progression of activities
  - Day 1
    - Out of bed activities, active-assisted range of motion, isometric exercises
    - Patient can be weight bearing as tolerated in most situations
  - Day 2-3
    - Gait training (with assistive device)
    - At this time, nursing focus on pin site cleaning and care with patient and family involvement

Inpatient Post-operative Evaluation and Intervention[5]

- Initial hospitalization typically lasts 7-10 days
  - Post-op monitoring
  - Physical therapy to increase mobility
  - Occupational therapy for independence with activities of daily living, including pin-care management

Case Study

Outpatient PT Management (Simard, et al.)[5]

- Ideally, patient will attend outpatient PT 3-5 times per week
  - Range of motion
  - Strengthening
  - Managing splinting needs
  - Providing assistive devices based on progression
  - Further balance and gait training
- Patients with significant range of motion deficits may benefit from dynamic splinting or may require manipulation under anesthesia

Outpatient PT Management (Prince, et al.)[30]

- Frequency: 1 hour of outpatient PT, 5 times per week
  - Main focus: maintain full knee extension and at least 45 degrees of knee flexion
  - Extension bar utilized at night
- Encouraged to perform home exercise program on weekends
- Intensity of physical therapy increased if decrease in knee flexion range of motion or tibiofemoral subluxation
- All children had access to child life therapist

Physical Therapy Goals[5]

- Prevent joint and soft tissue contractures
- Increase range of motion
- Decrease pain and edema
- Increase muscle strength
- Prevent and minimize gait deviations
- Restore functional mobility and independence

Case Study

Where Are We Headed?

Internal lengthening[31–33]

- Types of telescopic nails utilized
  - Motorized nails (FitBone®)
  - Mechanically-activated nails (Albizzia®, ISKD®)
  - Magnetically-driven nails (Phenix®, PRECICE®)
- PRECICE® is commercially available for use and FDA-approved
  - Much research focuses on this type of implantable device

Internal Lengthening with PRECICE® Nail[33]

- Mechanism activated by external remote control (ERC) device
- ERC placed firmly over magnet of PRECICE® nail implant
- Rotating magnets couple to and rotate magnetic spindle
  - 210 revolutions to achieve 1mm of lengthening (~7 minutes)
- Direction of ERC is important
  - One direction causes lengthening
  - One direction causes shortening

Benefits of PRECICE® Nail[32]

- Avoids many "device-related" complications
  - Pin tract infections
  - Joint stiffness
  - Neurovascular injuries related to placement of device
  - Re-fracture after removal of device
  - Less psychological burden
  - Less cumbersome in regards to physical mobility
  - No need for meticulous pin care site management
- Limits "too rapid" distraction (rate of 1.5mm/day)

Complications Related to PRECICE® Nail[32]

- Breakage of nail integrity
- Premature consolidation
- Delated and failed bone formation
- Nerve stretch injury
- Joint subluxations

Case Study

Conclusion

- External fixation can be utilized in the oncology population for limb lengthening following salvage procedure rather than previous gold standard of amputation.
- It is crucial to manage patients undergoing limb lengthening via external fixation with a multi-disciplinary team approach, including psychology and/or child life therapy, family involvement, and physical and occupational therapy.
- Physical therapy can play an important role in managing many complications associated with external fixator placement and limb lengthening.
- Internal limb lengthening may become a more popular option in the future.
- More research on limb lengthening (both external and internal methods) are needed to demonstrate functional outcomes following surgical intervention.

Questions?

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