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
Components of an External Fixation System

Pins\textsuperscript{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\textsuperscript{8,9}
- Typically utilized for ring fixators
- Always applied under tension
- Smaller diameter wires allow for controlled micro-motion to promote callus formation
Rods

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

Rings

- 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

- 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\textsuperscript{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\textsuperscript{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\textsuperscript{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\textsuperscript{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\textsuperscript{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\textsuperscript{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\textsuperscript{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\textsuperscript{4,14}

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

Distraction Osteogenesis (DO)\textsuperscript{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\textsuperscript{15-17}

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

Latency Phase
- Osteotomy
- Application of apparatus

Distraction Phase
- 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\textsuperscript{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\textsuperscript{18}

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

Lengthening of the Upper Extremity\textsuperscript{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.)\textsuperscript{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
  - \approx 92\% said “they would do it again”

Complications Related to Limb Lengthening

Muscle Contractures and Joint Stiffness\textsuperscript{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

- 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

- 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

- Can be related to surgery or to distraction
  - Direct vascular damage from corticotomy or osteotomy
  - Fistula formation
  - Compartment syndrome
  - DVT
  - Hypertension
  - Edema
Consolidation Issues\textsuperscript{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\textsuperscript{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 osteomyelitis

Other Complications\textsuperscript{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.)

• 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

• 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\textsuperscript{22–26}

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

Complications of External Fixation\textsuperscript{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.)

- 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

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.)

- 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.)

- 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.)

- 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.)

- 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.)

- 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.)

- 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.)

- 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

- 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

- 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

- 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

- 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.)

- 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.)

- 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

- 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\textsuperscript{31-33}

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

Internal Lengthening with PRECICE\textsuperscript{®} Nail\textsuperscript{33}

- Mechanism activated by external remote control (ERC) device
- ERC placed firmly over magnet of PRECICE\textsuperscript{®} nail implant
- Rotating magnets couple to and rotate magnetic spindle
  - 210 revolutions to achieve 1 mm of lengthening (~7 minutes)
- Direction of ERC is important
  - One direction causes lengthening
  - One direction causes shortening
Benefits of PRECICE® Nail
- 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
- Breakage of nail integrity
- Premature consolidation
- Delayed 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?

References

References