TLC from the PT: Why Inhibitor Patients Need a Physical Therapist

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How Bleeding Affects the Musculoskeletal System

- Bleeds can result in growth of the joint lining (synovium) → more bleeding
- Over time, the body breaks down the blood
- Byproducts of this, (iron, enzymes) damage cartilage and erode joint surfaces
- Cartilage damage leads to:
  - Loss of joint motion
  - Muscle atrophy and weakness
  - Joint arthropathy
  - Pain
  - Impairment in gait
- Clotting factor infusion can stop bleeding or prevent it

What Happens with Inhibitors

- Bleeding is difficult to control
  - Inhibitor interferes with factor VIII or IX working
  - Bypassing agents may be required for treatment
  - Bleeds result in growth of the joint lining (synovium) → more bleeding
  - Overall more frequent bleeding into joints / muscles
  - Leading to more bleeding complications, including joint disease
Bleeding Complications

- More difficulty preventing and stopping bleeding
- Higher risk of developing compartment syndromes with muscle bleeding
- May be more prone to developing target joints
- Typically develop target joints earlier in life due to difficulty with bleed management

Long Term Sequelae of Musculoskeletal Bleeding

- Flexion contractures
- Joint arthritis / arthropathy
- Chronic pain
- Muscle atrophy
- Compartment syndrome
- Neurologic impairment
- Gait impairment

Healthy Joint vs Joint Damage After Repeated Bleeding
Progression of Joint Arthropathy

MRI Right Ankle

Joint Loading & Joint Degeneration

- Injection of blood into right knee of beagles, 3X/week for 4 weeks—forced weightbearing
- Analyzed cartilage and synovium 10 weeks after last injection
  - Rate of proteoglycan synthesis ↑
  - Newly formed proteoglycan release ↑
  - Total loss of proteoglycans from cartilage matrix ↑
  - Cartilage matrix integrity adversely altered
  - Signs of synovial inflammation
- None of this was present in non-weightbearing controls

Evaluation of Acute Joint Bleed

- Swelling
- Temperature
- Pain
- Active motion
- Muscle function
- Circumference
- Gait - in Lower Extremities

Acute Hemarthrosis

- INFUSION – tailored individually / bypassing agents
- RICE
  - Avoid use of ice alone.
  - Should be combined with compression and infusion
- NWB for Lower Extremity bleed
- Avoid putting burden of body weight on other joints
  - May require wheelchair or crutches
  - No weight bearing if blood still in joint
  - Splinting for severe bleeds or in persons with inhibitors

Muscle Bleeding Evaluation

- Description of area of bleeding
  - Size
  - Temperature
  - Degree of swelling - firmness
- ROM assessment at 2 adjacent joints
- Assessment of nerve and vascular integrity
  - Numbness, weakness, pulse
- Pain assessment
Muscle Rehabilitation

- RICE + Infusion therapy
  - Compression may be contra-indicated if risk of compartment syndrome
- Support in position of comfort
- NWB gait
- Begin movement at one joint at a time
- Exercise must concentrate on muscle stretching and strengthening
- MUSCLE BLEEDING EASILY RECURS

Muscle Bleeding Considerations

- Difficult without replacement therapy
- Immobilization risk of further hemorrhages
- Splinting may be used to permit healing by fibrosis to take place in optimum functional position
- Hydrotherapy in early rehab can be useful
- Heel supports can elevate the heel in a calf muscle bleed during healing
Muscle Bleeding

Splints
Alleviate the Pain

Making Splints
Treatment Options For Optimal Therapy Participation

- For best success with a therapy program, ideally the program should be doing with factor coverage
- Tolerized inhibitor: Factor VIII or IX
- For inhibitors that have not been tolerated:
  - Novoseven
  - Feiba
- ALWAYS FOLLOW THE INFUSION RECOMMENDATIONS OF YOUR HEMOPHILIA CARE PROVIDER!
Introduction of Exercise

- If possible, infuse prior to beginning exercise
- Infusion therapy MUST be tailored to the individual (bypassing agents)
- Begin isometrics and movement within pain-free arcs: once pain subsides
- Increase repetitions or time doing exercise slowly / systematically – NO PASSIVE MOTION!
- Goal is to restore muscle balance
- Functional re-training
- Assistance in helping to resume normal ADL without putting stress on the affected joint
- If bleeding is recurring, assessment of movement and activity should be done
  - Extended rest may be required
  - May have to start at lower level

Chronic Synovitis

- Combine structured supervised daily muscle strengthening exercises with splinting to help with muscle weakness and instability
- Can use cold application, but this should be monitored closely: Cold should preferably be used with compression

Hemophilic Arthropathy

- Pain reduction can be achieved with joint relaxation
- Can use slow gradual stretching to help regain motion, especially in extension
- Follow up with active muscle strengthening to maintain new motion
- If joints are still painful, soft orthoses or braces may be helpful
Support Braces

Management of Contractures
Low Weight - Long Duration Stretch

Management of Contractures
- Dynasplint™
- Ultratex™
- Increase the resistance very slowly to prevent bleeding
- If a bleed occurs, return to minimal resistance and increase resistance slowly again
- It may take 6-12 months before obtaining more movement

Bracing
Carbon Fiber AFO
Hinged, Locking AFO
**Massage**

- Has been used to help mobilize muscle tissue, but needs to be used guardedly.
- NEVER use massage during an acute bleed.
- Do NOT massage over an area of muscle bleeding.
- If possible, it is better to treat with clotting factor before massage to prevent bleeding.

- If you are NOT able to treat with factor:
  - Use less pressure.
  - Work around any area of bleeding – if in joint or muscle.
  - Avoid deep pressure strokes – pressure should NEVER cause bruising.

**Functionally-based Treatment**

- Childhood living skills.
- Encourage family games:
  - Playing with softer balls.
  - Playing on grass vs harder surfaces.
  - Swimming.
  - Cycling with protective gear.
  - Use adult supervision.
- Age-appropriate program planning to assure correct instruction and teaching to assess risks of the activity.
- Evaluation and documentation:
  - In clinical environment.
  - In school.

**What Physical Activities or Sports are Safe?**

- Decisions?
- Decisions?
- Decisions?
Identifying Risky Sports and Fitness Exercises
National Hemophilia Foundation Categories

1: Low Risk
- Bicycling
- Fishing
- Frisbee
- Golf
- Hiking
- Tai Chi
- Swimming
- Walking

2: Some Risk: Must "Play Smart!"
- Baseball
- Basketball
- Bowling
- Horseback riding
- Martial arts
- Mountain biking
- Rollerblading
- Running and jogging
- Skiing
- Soccer
- Tennis
- Weight lifting

3: High Risk: NOT Safe!
- Boxing
- Contact karate
- Football or Rugby
- Hockey
- Lacrosse
- Motorcycling
- Racquetball or Squash
- Rock climbing
- Wrestling
- Snowboarding

Preparation for the Sport / Activity?
- Approval of hemophilia team
- Have a management plan
- Proper equipment
- Age or risk-appropriate risk activity
  - Consider music and the arts
- Non-competitive environment supporting skill development
- Well-maintained facilities or fields
- Proper instruction / coaching
- Never “play through pain”, either from bleeding or arthritic pain
Ready for Activity?

“My Game Plan”
Do each time prior to doing activities:
- Is he walking / running normally?
- Is he using his arms appropriately?
- Is he complaining of, or showing, any signs of pain?
  - Irritability, fussiness, withdrawn behavior?
- Is pain, if present, different from baseline?
- Will the activity contribute to an already compromised joint?

When to Re-evaluate the Sport

Is This Working?

- If level of activity ↑’s risk
- Child reports pain in any joints or muscles
- If continued trauma causes recurrent bleeding ↑ joint damage
- If child develops swelling in any joints (inflammation or bleeding?)
- If bleeding occurs
  - When playing or soon afterwards
  - Without history of injury or trauma
- Is he enjoying the activity?

Adapted Sports

- In some cases sports may be too risky for someone with an inhibitor
- Adaptations can be made to encourage participation in the activity
  - Team management
  - Record keeping, playing music etc.
- If lower extremity joints are affected, can use a wheelchair for participation in activities requiring more upper body use
  - Wheelchair tennis, basketball, volleyball
  - Archery
- ENCOURAGE THE ARTS AND MUSIC !!
Modifying Equipment

Aquatic Therapy

- Buoyancy of the water supports joints and muscles
- There is no risk of falling
- Water ↑ resistance to the muscles
- ↓ the force of the weight of the body
  - Allows walking on weak legs
  - Alleviates pain in joints and muscles
  - ↑ Range of Motion (ROM)
- Begin in deep water → move to shallower water

Aquatic Equipment

- Hydraulic lifts
- Flotation jacket (life jacket)
- Kickboard
- Flippers
- Weights
- Chair in the water

- Water temperature: 35.5-36.6°C (96-98°F)
Prognostic Factors for Success

- Age
- Activity level
- Factor VIII / IX level Inhibitor
- Physical Therapy
- Self-Esteem
- Motivation
- Level of Joint Disease / Weakness
- Family Support
- Bleeding History
Conclusion

- Boys with inhibitors need exercise and activity as much as any other children
- The challenge is to provide it safely
- The treatment center team, and specifically the therapist, can be a resource for keeping muscles strong and joints flexible
- Physical therapy is needed more in those with inhibitors but it may need to be modified to meet their special needs
- Therapy may need to be on-going and progressed slowly, but should always be considered

References

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Thank You!