The Benefits of Physiotherapy: exercise and fitness, maintaining healthy joints, optimizing prophylaxis

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Comprehensive Care

- Hematology
- Nursing
- Physiotherapy
- Psychosocial
- (Orthopedics)
Haemophilia and Physiotherapy

• **1960’s**: cryoprecipitate and then Factor VIII concentrates become available

• P.T. ‘without tears’
  – Factor Pre-physio

• **Post-operative** P.T.
  – Surgical synovectomy
  – Corrective surgery
  – Joint replacement
Physiotherapy

- Education to parents and patients
- Prevent bleeds
- Treat bleeds
Myths

• “Treatment” means “factor”

• People with Mild Hemophilia do not get joint disease

• Joint disease will become extinct because of prophylaxis
Challenges

- Primary prophylaxis
- Home infusion programs
- Sports selection
- Obesity and overweight
- Using factor wisely
Primary Prophylaxis

- Prevent bleeding ‘before it starts’
  - Started young OR after first bleed
  - “Turn severe hemophilia into mild hemophilia”
  - “Allows a ‘normal’ life style”
But.....

- Children on prophylaxis *still get joint damage*
  - Shown on MRI
  - Even with NO clinical history of bleeding! (Manco-Johnson)

- People with Mild Hemophilia also get joint damage (Scully)
WHY??

• Prophylaxis does not make the clotting 100% ‘normal’
  – Raises levels but does not ‘normalize’ levels
  – Severe hemophilia becomes Mild hemophilia

  – *BUT IT IS STILL HEMOPHILIA*
WHY???

• Half life of factor
  – VIII 8-12 hours
  – IX 18-24

• “traditional proph” (alternate days)
  Severe ➔ Mild ➔ Moderate ➔
  Severe again within 48 hours
Half Life

After infusion, 40% FVIII

After 12 hours, half of 40% = 20%
Mild Hemophilia

After 24 hours, half of 20% remains = 10%

After 36 hours, half of 10% remain, = 5%
Moderate hemophilia
Take home point

• Timing is everything
  – Infuse in the morning, not at bed time
  – Participate in sports and activities according to your prophylaxis days
Home Treatment

• Advantages:
  – Convenience
  – EARLY infusion → less blood in joint

• BUT:
  Families make treatment decisions alone

_Treatment is more than just factor replacement._
  – Mark Skinner, President of WFH
Home Treatment

• Bleeds happen.

• Treatment Steps:
  – Ice ?
  – Infusion - yes
WHAT ABOUT REST????
WHY???

• Blood in a joint affects the synovium and the cartilage

• Synovium:
  – Role is to produce fluid for lubrication and nutrition of the cartilage
  – After a bleed, it becomes over-active
  – Enzymes harm cartilage
So what?

- Even though the blood is cleared quickly, the synovium remains inflamed for AT LEAST 2 weeks.
- If a new bleed begins in this period, it ADDS to the inflammation.
- Secondary prophylaxis and/or synovectomy are required early:
  - to de-activate the synovium
  - to slow the progression.
But that’s not all!

• Research in Netherlands has shown that blood harms the cartilage *directly*:
  
  • Blood in a joint is bad for cartilage  
  • More blood is worse than a little blood  
  • *Weight-bearing when there is blood in the joint* is worse still
Wait, there’s more….

• Blood on young cartilage is still worse
• Blood on damaged cartilage is bad too

• Cartilage cells remain disorganized for at least two weeks after a bleed. Cells break down and die and are not replaced.
Take home messages

• The cycle of joint damage is – sadly - set in motion after even one bleed

• The cartilage cells remain in a fragile state for at least two weeks.

• So REST those joints when they bleed!!!!!
  – That means stay off them!!!
  – We’re not really sure for how long.....


Jansen et al: Degenerated cartilage is as vulnerable to blood induced damage as healthy cartilage is. Ann Rheum Dis 2007

Jansen et al: Very rapid clearance after a joint bleed in the canine knee can not prevent adverse effects on cartilage and synovial tissue. Osteoarthritis and Cartilage- 2008
Sports and activities

- Physical Activity is good.
  - Fun
  - Muscle strength and coordination
  - Joint mobility
  - Weight control
  - Cardiovascular fitness
  - Bone density
However….

PWH in the developed world have begun participating in more ‘risky’ sports:
(Might not be a new phenomenon)

• Because they can *pre-*treat with factor
• Because they can *treat injuries* with factor
Sports and Activities

• Australia
  – Aussie rules, rugby
  – surfing

• Canada
  – Ice hockey
  – snowboarding

Boys will always be boys
Choosing sports and activities: things to consider

The Sport:
- Contact or collision?
- Speed?
- Type of injuries?
  - Head, neck, abdomen
- Equipment needed?
- Facilities & coaching?
- Weight-bearing or non-weight-bearing?*

Your own body:
- Target joint(s)
- Strength?
- Coordination?
- Flexibility?
- Response to factor
The activity should match the person (and vice versa)

Seuser et al (Germany)- computerized assessments to assess sports and patients.

Physiotherapy can also do this.....
Physiotherapy can:

• Assess joint health, strength, coordination, flexibility, etc
• Assess the requirements of the sport
• Find solutions to any mis-matches:
  – Exercises
  – Protective gear
  – Modify the sport
  – Work with the coach
Weight-bearing or not?

Weight-bearing:
• Stress on stressed cartilage may speed up damage
• But will $\rightarrow$ stronger bones

Non-weight-bearing:
• Less stress on cartilage
• Very important after bleeds
• But $\rightarrow$ weaker bones
  – Osteoporosis is common in PWH

Kovacs CS. Hemophilia, low bone mass, and osteopenia/osteoporosis. Transfus Apher Sci 2008
Overweight and Obesity

• Common throughout the world
• Almost 60% of the adult population in Australia is overweight or obese
• Increasing quickly in children too:
  – 25% of Australian children are currently overweight or obese
  – one of the highest rates amongst developed nations
Obesity is linked to many health problems –
• osteoarthritis is just one of them
• overweight/obesity directly affects weight-bearing joints, especially the knees
• knee osteoarthritis is 4 to 5 times more common in overweight people than in people who are of normal body weight (and that is without hemophilia)
osteoarthritis

• During walking, a force of 3 to 6 times a person's body weight is exerted across the knee joint
  – being 10 pounds overweight increases the force on the knee by 30 to 60 pounds with each step taken….

• What if the joint is already damaged by blood?
Take home message

• Maintain a healthy body weight
  – *Protect your joints*
Using factor wisely

• Is it really a bleed?
• Is there a pattern?
Is it really a bleed?

Stephensen et al- UK
MSK Congress in Stresa, 2007

45 patients with acute ‘spontaneous’ joint pain and stiffness- assessed with Ultrasound

- Acute joint bleed 31%
- Muscle bleeding 18%
- Subcutaneous tissues 15.6%
- Synovitis diagnosed in 22%
Is it really a bleed?

- 11% pathology unrelated to hemophilia
- *No evidence of bleeding in 33%*

- If in doubt, treat?
- Maybe not:
  - Pain from arthritis will not respond to factor
  - The challenge: Improve our assessment skills: patients AND treaters
Is there a pattern?

- Does one joint bleed more than others?
- Do bleeds occur after particular activities?
- Is there synovitis or early degeneration?

- Bleeding logs!
- Annual assessment!

- Maybe something needs to change.....
Take home message

• Complete bleeding logs regularly
• Attend your annual assessments
• Allows EARLY recognition and intervention for small problems
• Opportunity for reviewing prevention, assessment, treatment
Cure is a long way off

- In the meantime:
  - Use prophylaxis appropriately
  - Choose activities wisely
  - Treat all bleeds completely
    - Make sure it’s really a bleed
  - Maintain a healthy body weight
  - Don’t let little problems get out of control
and be glad you live in Australia