

# 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)



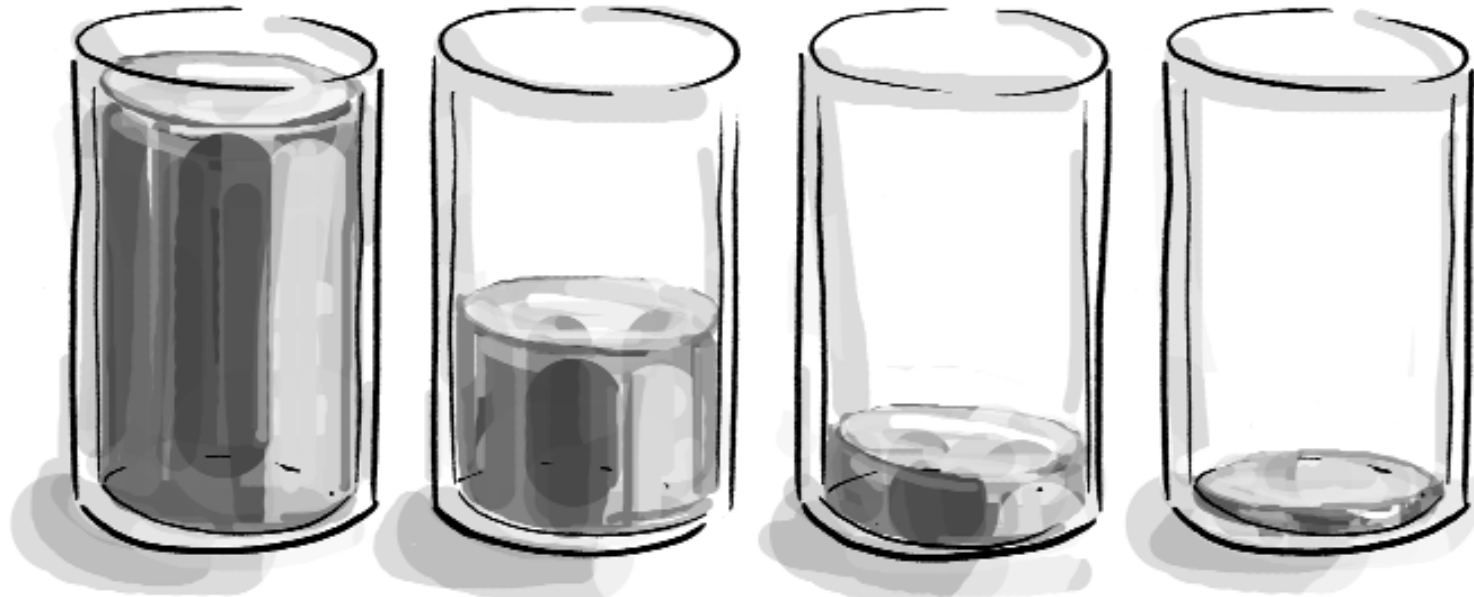
# WHY??

- 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.....

# References

- Jansen, Roosendaal et al : Exposure of human cartilage tissue to low concentrations of blood for a short period of time leads to prolonged cartilage damage: an in vitro study. *Arth and Rheum* 2007
- Roosendaal et al: Articular cartilage is more susceptible to blood induced damage at young than at old age. *Journal of Rheumatology* 27, 1740-44. 2000.
- 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 → stronger bones

## Non-weight-bearing:

- Less stress on cartilage
- Very important after bleeds
- But → weaker bones
  - Osteoporosis is common in PWH

# References

Wallny et al. Osteoporosis in haemophilia - an underestimated comorbidity? Haemophilia 2007

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

# osteoarthritis.about.com

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

