



# 21<sup>ST</sup> AUSTRALIAN CONFERENCE

ON HAEMOPHILIA, VWD AND RARE BLEEDING DISORDERS

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## Disclaimer

This conference is hosted by Haemophilia Foundation Australia.

Conference speakers include health professionals, researchers, government officials and bleeding disorders community members.

**Some of the treatments discussed may not be registered or funded currently in Australia and should not be considered as a promotion or recommendation. Please discuss this with your health professional.**

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# Mild Haemophilia

Musculoskeletal Issues, Physical activities and Sport across the lifespan

**Abi Polus**

Musculoskeletal Physiotherapist

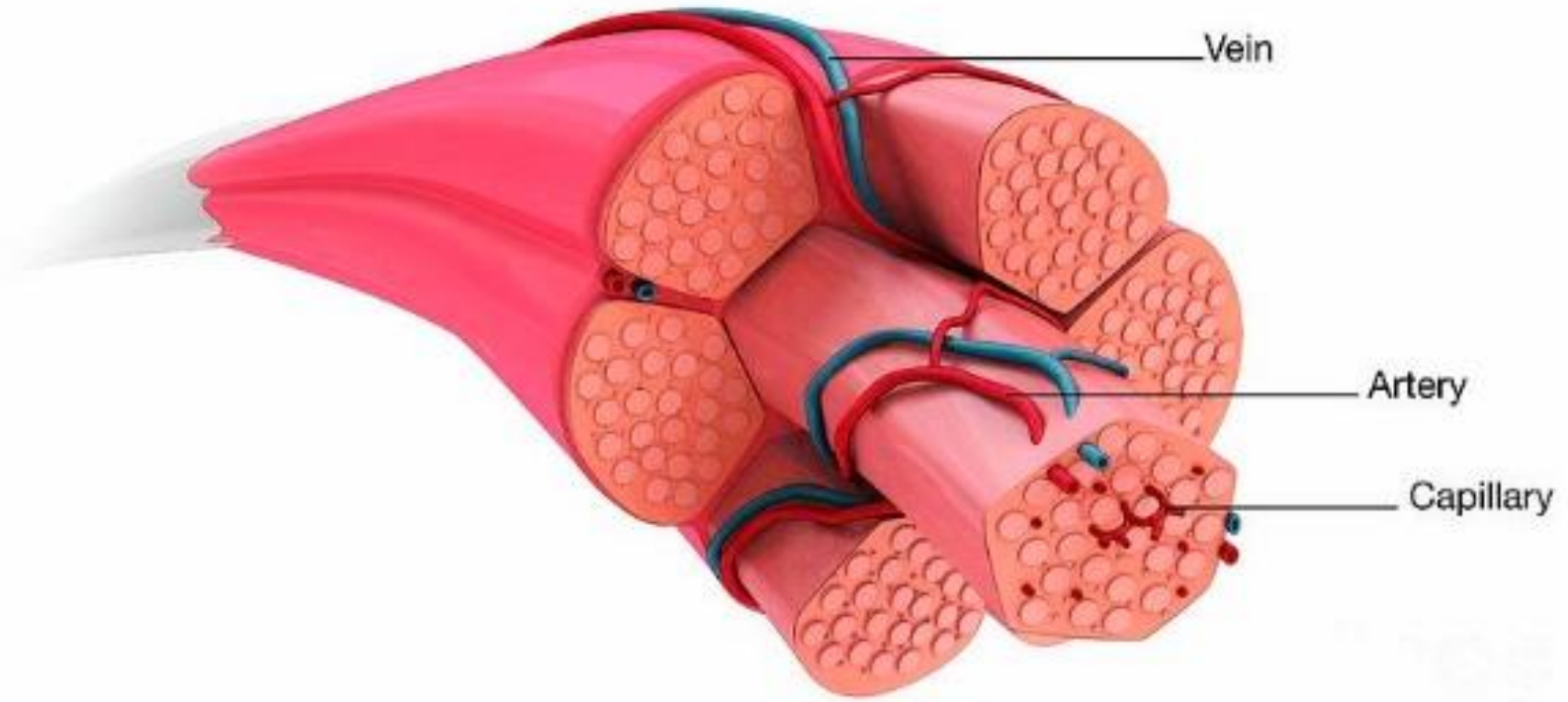
RSHC for Bleeding Disorders, Alfred Health, Vic

August 2023

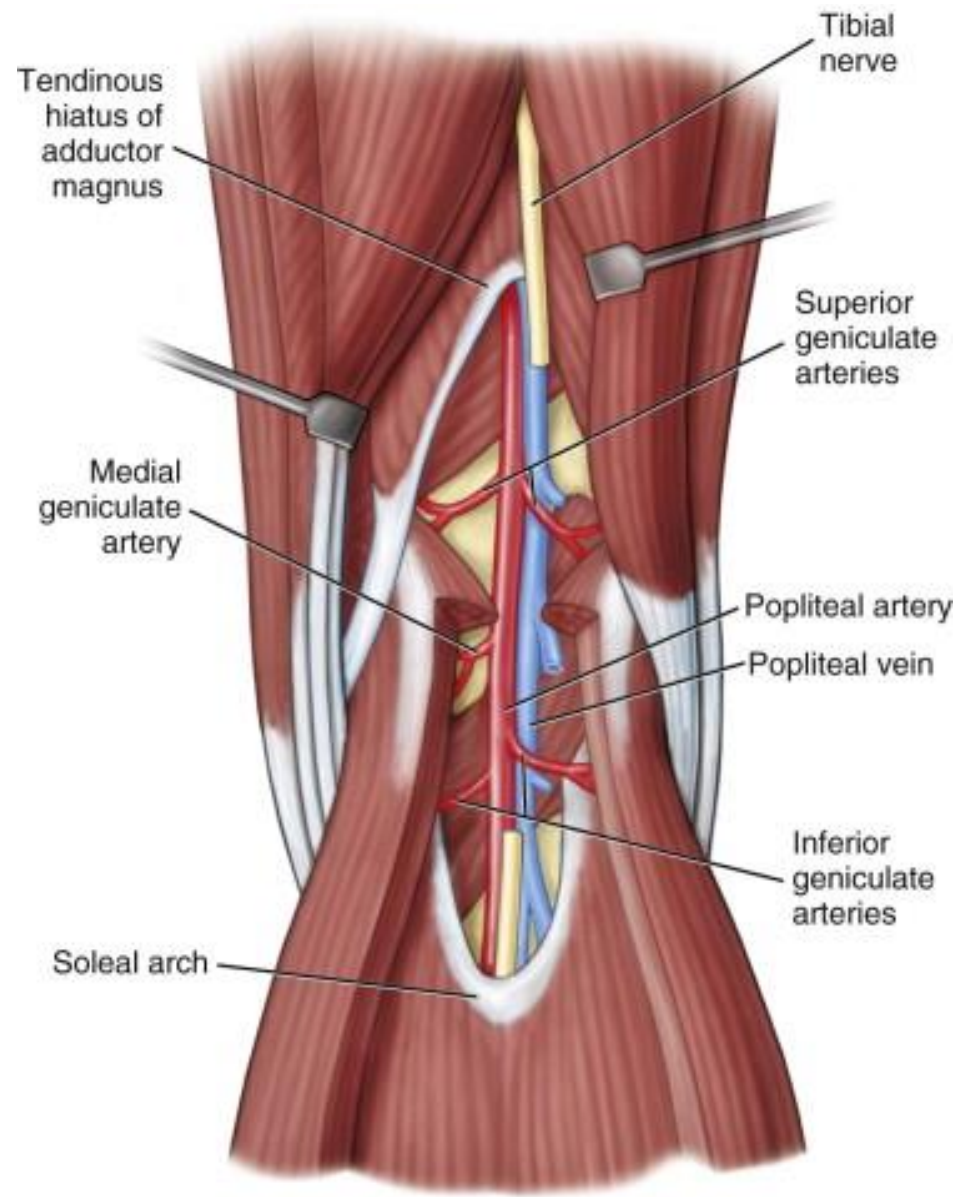
# Mild Haemophilia

- <1% - Severe
- 1-5% Moderate
- 5-40% Mild
  - 5-15%
  - 15-40%
- (Normal levels 40-150%)
  
- **Usually:** Bleed when subject to **trauma** or surgery
  - Can be phenotypically abnormal
  - But 36.5% had arthropathy\*

\*Corte-Rodriguez et al. Arthropathy in people with Mild haemophilia: Exploring the risks. 2022 Thrombosis research. 211. 19-26



All images from google



# Mild Haemophilia



- ‘Old Milds’ Vs ‘New Milds’
  - (Pharmacologically, genetic modification, female)
- Studies comparing ‘(Old)’ Mild to age match controls without haemophilia
  - Joint dysfunction: pain and damage (?resulting from subclinical joint bleeds)
  - Pain and disability increasing with age
  - Haemophilia Joint Health Score (HJHS) worse/higher than age matched controls
  - Missed work 3.4 - 4.7 days/year due to haemophilia
  - QOL reported decrease

# Mild Haemophilia - Musculoskeletal Issues

- Rarely see bleeds in kids with mild haemophilia except sport injuries
- In adults sports and other activities – repetitive tasks, farming incidents
- May lack the ability to identify bleeds
- May lack ability to self infuse
  - >> presenting early vs late
- Not often included in clinical trials
- Many patients diagnosed at an older age AFTER trauma >>1 bleed >> potential arthritis
- Bleed may not be so obvious >> subclinical bleeds >> >> cumulative effect of bleed>> potential arthritis
- Women >> need more research arthritis from bleeds>> plus other female factors eg OA, RA, hypermobility
- No set guidelines in sport and activities due to wide variation in risk of bleeding
- Opinion differs vastly on coagulation levels 9-52% without arthropathy and 12-64% with arthropathy

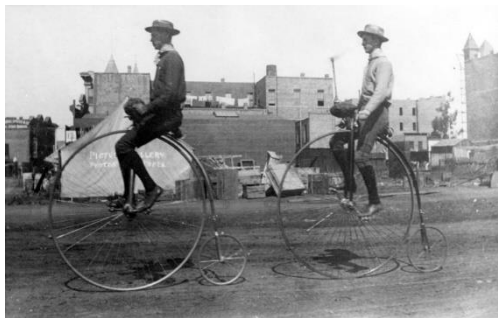


# History of Change

- Management of haemophilia has changed;



- Trends in sport have changed; attitudes to sport have changed



General Benefit of Exercise and Sport	Specific benefit of Exercise and Sport for PWH
<ul style="list-style-type: none"> <li>•<b>Harris and Boggio (2006)</b>: Improved joint ROM in an exercise group vs a non-exercise group and proposed benefits of exercise on cartilage, muscle and peri-articular structures</li> <li>•<b>WHO and US centre for disease control</b>: numerous publications of scientific evidence that regular, appropriate physical activity and sport provide a wide range of physical, social and mental health benefits</li> <li>•<b>Smolander et al (2000), Stewart et al (2005), Bonhauser et al (2005)</b>: Improved body composition and self esteem</li> <li>•<b>Sullivan et al (2005), Warburton et al (2006)</b>: Reduction of Diabetes risk</li> <li>•<b>Woo et al (2004)</b>: Reduction of Cardiovascular risk</li> <li>•<b>Ekeland et al (2005), Brage et al (2004)</b>: Reduction of Metabolic syndromes</li> <li>•<b>Borer (2005), Kohrt et al (2004)</b>: Reduction of Osteoporosis</li> <li>•<b>Harris and Boggio (2006)</b>: Joint disease Management</li> <li>•<b>Gascon et al (2004), Vainio (2006)</b>: Reduction of Certain Cancers</li> <li>• <b>Wittmeier and Mulder (2007)</b> Good motor development (can help pwh)</li> </ul>	<ul style="list-style-type: none"> <li>•<b>Koch et al (1984)</b>: physical activity enhances the concentration of various coagulation factors</li> <li>•<b>Titinsky et al (2002)</b>: marked decrease of bleeding complications after progressive resistance training and decrease of associated bleeding-related pain</li> <li>• <b>van der Net et al (2006), von Mackenson 2007</b>: Increased QOL in PWH</li> <li>•<b>Greene and Strickler (1983)</b>: Increase in muscle strength without increase in bleeding episodes with an isokinetic strengthening program.</li> <li><b>Groen et al (2013)</b>: Strenuous physical exercise increasing level of clotting factor by 2.5 of baseline in mod/mild HA</li> <li>•<b>Buzzard (1997)</b>: synovitic joint management with exercise</li> <li>•<b>Lippi and Maffulli (2009)</b>: Strenuous exercise associated with transiently increased thrombin generation, platelet hyperreactivity, increased activity of coagulation factors including FVIII and vWf</li> <li>•<b>McGee et al (2015)</b>: Strength, balance, joint ROM, decrease in bleeding episodes, obesity management</li> </ul>

# Mild haemophilia

- Prior studies consistently exclude people with mild haemophilia (PWMH)
- ‘While their risk of bleeding may be less, participation in higher risk sports could place them at increased risk for injury and subsequent bleeding’
- ‘There is concern that patients with mild haemophilia may underestimate that risk of bleeding with injuries which may result in delayed treatment and significant consequences’
- Not more injuries than those who did not participate in organized sport

- Various classifications lists in existence for recommendations of which sport is best for PWH to participate in
- These may be based on:
  - The probability of contact or collision
  - Others are based on incidence of injuries (high, medium, low, risk)
  - The frequency of recorded injuries in this sport
- More complex parameters are now being proposed to classify sports including:
  - biomechanical aspects
  - level of difficulty
  - risk of injury



# Playing It Safe

Table 5. Sports Ratings by Activity

Activities have been divided into five ratings:



Activity	Category
Aerobics	2
Archery	1
Aquatics	1
Baseball	2.5
Basketball	2.5
Bicycling	1.5
BMX Racing	3
Bowling	2
Boxing	3
Canoeing	2.5
Cardiovascular Training Equipment	
Elliptical Machine	1
Rowing Machine	1.5
Ski machine	1.5
Stationary Bike	1
Stepper	2
Treadmill	1.5
Cheerleading	2.5
Circuit Training	1.5
Dance	2
Diving/Competitive	3
Diving/Recreational	2
Exercise Classes	
Body Sculpting	1.5
Cardio Kick-Boxing	2
Physioball	1.5
Spinning	1.5
Fishing	1
Football	3
Frisbee	1
Frisbee Golf	1.5
Ultimate Frisbee	2
Golf	1
Gymnastics	2.5
Hiking	1
Hockey (Field, Ice, Street)	3
Horseback Riding	2.5
Ice-Skating	2.5

Activity	Category
Inline Skating	2.5
Jet Skiing	2.5
Jumping Rope	2
Kayaking	2.5
Lacrosse	3
Martial Arts – Karate/ Kung Fu/Tae Kwon Do	2.5
Martial Arts/Tai Chi	1
Motorcycling/ Motor Cross Racing	3
Mountain Biking	2.5
Pilates	1.5
Power Lifting	3
Racquetball	2.5
River Rafting	2.5
Rock Climbing (Indoor/Challenge Course)	2
Rock Climbing (Natural Setting)	3
Rodeo	3
Roller-skating	2
Rowing/Crew	2
Rugby	3
Running and Jogging	2
Scooter (motorized)	3
Scooter (non-motorized)	2.5
Scuba Diving	2.5
Skateboarding	2.5
Skiing/Cross Country	2
Skiing/Downhill	2.5
Skiing/Telemark	2.5
Snorkeling	1
Snowboarding	2.5
Snowmobiling	3
Soccer	2.5
Softball	2.5
Surfing	2.5
Swimming	1
T-Ball	2
Tennis	2
Track and Field	2.5
Trampoline	3
Volleyball	2.5
Walking	1
Water-skiing	2.5
Weight Lifting/Resistance Training	1.5
Weight Lifting/Power Lifting	3
Wrestling	3
Yoga	2



# 'Football'





# Contact and 'non-contact' sports



# Contact sport

**Contact may not just be with another player but also the ground or equipment.**







# Limitations

- There are limitations to all guidelines
- In most cases lists are generated on **theoretical** risk experience and opinions of haemophilia treaters who may have limited knowledge of the specific complexities and demands of the sport.
- Some sports are classified as high-risk due to high *frequency* of injury, despite the injuries being mild.
- Others, may be classified as medium risk, where injuries sustained may be catastrophic.
- Direct contact may not only be the cause of bleeds in haemophilia; muscle strains and overloaded joints can be due to many non-contact sports eg weightlifting
- Other factors such as rest and recovery periods, duration, muscle mass, expertise may all play a part

Full Length Article

## Arthropathy in people with mild haemophilia: Exploring risk factors

Hortensia De la Corte-Rodriguez <sup>a</sup>, E. Carlos Rodriguez-Merchan <sup>b</sup>  ,  
M. Teresa Alvarez-Roman <sup>c</sup>, Monica Martin-Salces <sup>c</sup>, Isabel Rivas-Pollmar <sup>c</sup>, Victor Jimenez-Yuste <sup>c</sup>

ORIGINAL ARTICLE |  Full Access

## Understanding minimum and ideal factor levels for participation in physical activities by people with haemophilia: An expert elicitation exercise

Antony P. Martin , Tom Burke, Sohaib Asghar, Declan Noone, Gabriel Pedra, Jamie O'Hara

- Investigate factors associated with development of arthropathy in PWMH (85)
  - Findings: Age, clotting factor levels, activity levels
- No statistical significance between level of physical activity and arthropathy
- Striking that those who did not participate in sport or activity had similar scores to those performing level 2 activities
- 42% participated in level 3 activities with no joint arthropathy
- In PWMH ?Acute exercise transient hypercoagulability
- Higher rate of arthropathy than general population; may not been seen until 2nd decade
- 'Risk of arthropathy increases 7.9 % for each additional year of age and decreases 7.7% with each additional 1 IU/dL increase of clotting factor'
- Suggest factor levels of 5-15% suggested during physical activity (excluding bleeding trauma) and up to 64% in patients with joint arthropathy
- All pts <17% have regular joint assessments



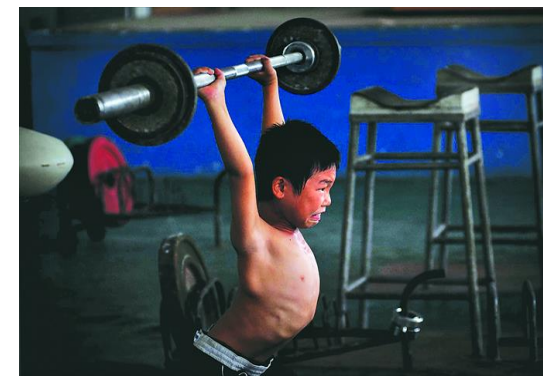
# Other factors to consider when selecting sports in Mild haemophilia– Everyone is not equal??

- Countries and areas where there is a scarcity of factor replacement
- Considerations for patients with inhibitors
- Previous joint status
- Previous fitness status
- Impact on job capacity





# Other factors to consider when selecting sports - Age



- Progression of the sport from non-contact to contact.
- Avoidance of repetitive motion in sport that may cause muscle imbalance; especially on the developing skeleton.
- Weight-lifting is contraindicated on those who are not skeletally mature.
- During growth spurt times adolescents are particularly vulnerable to injury due to imbalance between strength and flexibility.





Majority of the literature now advises that the decision making process is **individual** to each pt and should be done **collaboratively**, taking into account:

- Clinical status
- PWMH interests and desires
- PWMH abilities, capabilities and existing joint damage

**EDUCATE to EMPOWER**

# A note on peri-articular injuries



- PWMH sustain **non-bleeding** injuries just as people without bleeding disorders
- Factor replacement usage to treat sprains, strains, overuse injuries, sore muscles after sport
  - Not only a costly waste of money
  - Conditions that go untreated cause pain and may increase further injury risk to muscle and joints
  - Good injury assessment is imperative to confirm or rule out a bleed and manage the correct issue



# Take Home Message:

**PWMH should be supported in making informed choices.**

- This includes being educated about potential benefits and risks**
- And awareness of signs of bleeding and the need for prompt treatment and consequences**

