

The Importance of Upper Limb Care Throughout Life

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Objectives

Common complications of the upper limb in haemophilia

Clarify links between upper limb musculoskeletal health, and general health

Physiotherapy interventions

- Methods
- Timing

Cautions and Precautions

Combined management with Orthopedic Surgery

Non-Haemophilic injuries of the elbow

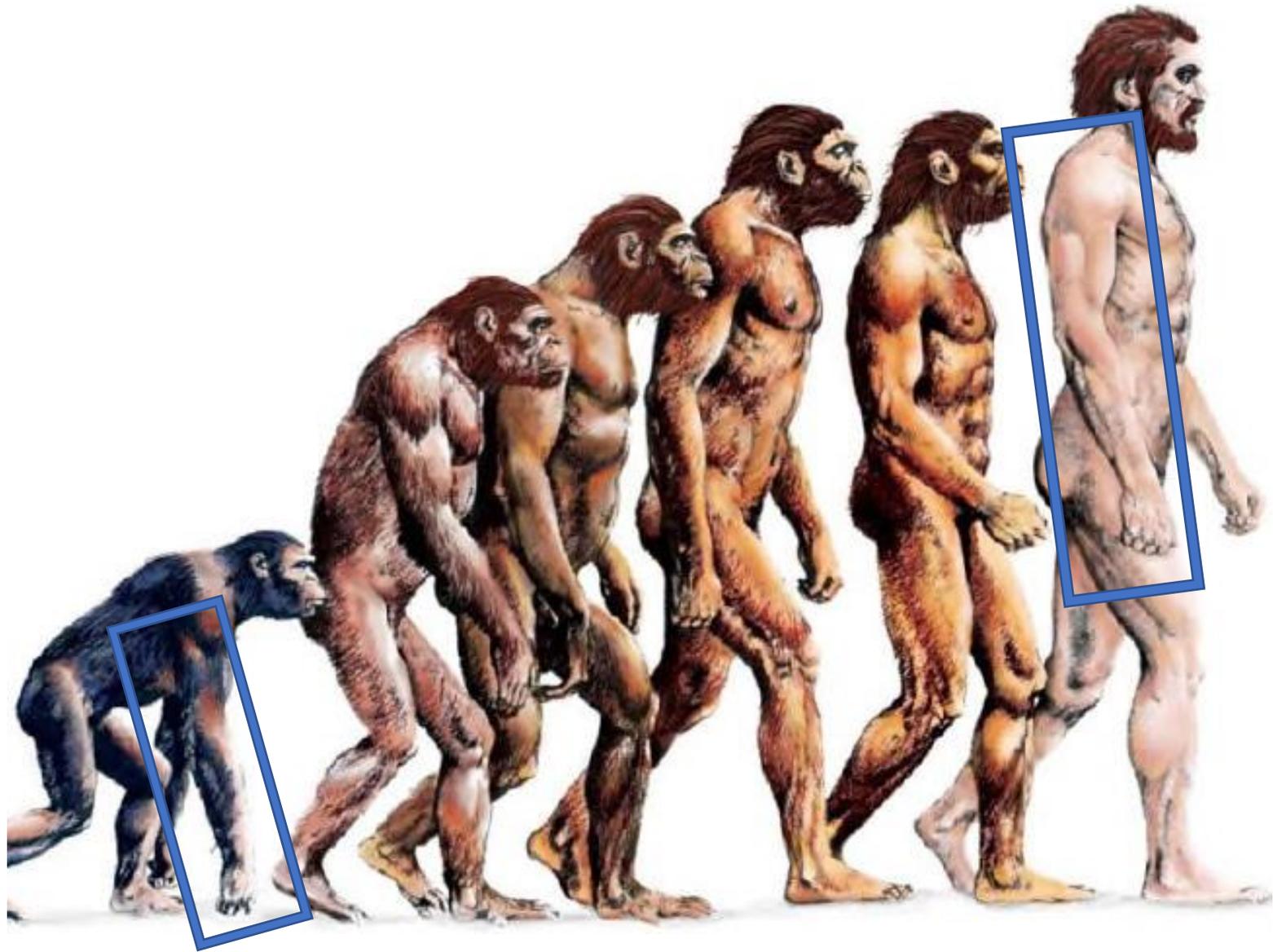
Structure Adapting to Function

As in most things, the manner in which the upper extremity (U/E) is used has affected its development and adaptations

The entire limb serves as a mechanism to allow for placement of the hand in space...the vital end-organ of accomplishment

The limb is oriented such that the hand is almost always under visual control which reinforces the connection of the U/E to vital sensory inputs and motor outputs

Compared to the inherent stability of the ankle and the hip, U/E joints sacrifice structural congruence for adaptive mobility



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The Usual Suspects...

Joint Bleeding:

- Elbow
- Less frequently, the shoulder, wrist or hand unless by direct trauma

Muscle Bleeding

- Forearm bleeds and possible compartment syndrome
- Upper arm muscles that cross two joints—biceps/triceps

Traumatic orthopaedic injury and repetitive strain injury



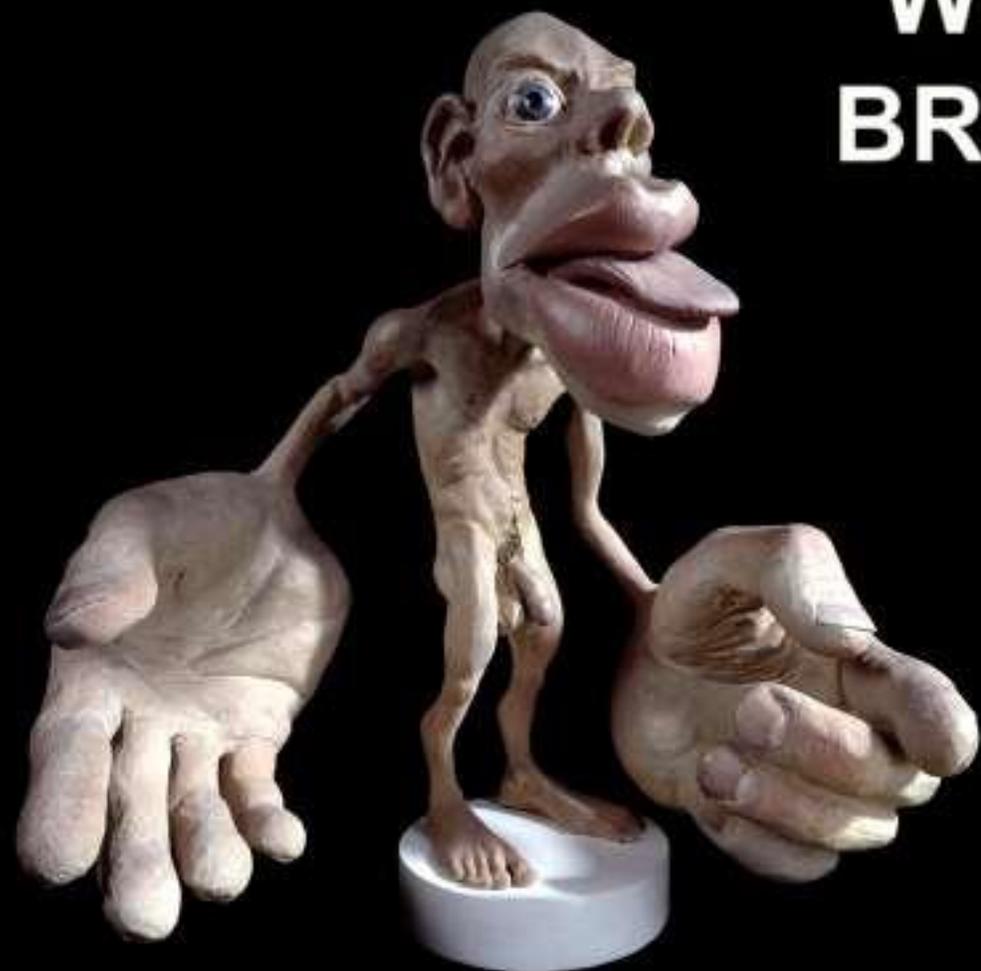
Focus: the Elbow

Structure and Function of the Elbow

- Three articulations enveloped in one joint capsule
 - Humeroulnar; Humeroradial; Proximal Radioulnar
- Any haemarthrosis to the “elbow” joint will therefore affect three joints
- It is impacted by lower limb deficits by having to become an auxiliary joint of ambulation for which it is not ideally designed
- Damage is usually insidious – early losses of ROM are functionally tolerated to a greater extent than lower limb counterparts

- Hinge joints tend to sacrifice full extension as a result of repeated bleeds
 - This compromises the primary function of the knee
 - This minimally disrupts functional use of the elbow
- The functional impact of elbow dysfunction can be devastating
- If 50% of elbow motion is lost, the cost to overall function of the upper limb can approach 80%
- Consider a neuroanatomist's view of the importance of the upper limb...

WHAT THE BRAIN SEES



SENSORY



MOTOR

The PRUJ

- The rotatory function of this critical part of the elbow joint complex is essential to normal hand function
- Enlargement of the radial head from repeated bleeds causes impingement based loss of full rotation in the radial notch on the ulna
- Once lost, the only way to restore some motion and reduce the associated pain is to resect the radial head – often with an accompanying synovectomy

- There are key linkages between the musculoskeletal health of the upper limb and overall general health
 - The ability to eat
 - The ability to perform effective oral health care
 - Psycho-Social / Emotional health – communication, sexuality, self-esteem, cultural implications
- Too often we describe functional capacity for a specific task based on how much ROM is required to complete it successfully
 - Significant oversimplification
 - Consider brushing your teeth... or shaving...
 - Requires both ROM AND the ability to sustain it
 - Coordinated movement of the hand, wrist, forearm and shoulder
 - The ability to do it either via a mirror image, or with no visual assistance at all...just by feel

Proprioception

- Coordinated motion of the arm requires balance between motor output and sensory input – feedback and feed-forward mechanisms must be synchronised
- Visual information plays a minor role in the performance of well-trained reaching movements - proprioception is the main source of feedback providing motor control

Kruger et al (2011), Clin Neurophysiol

Progression of Injury and Sequellae

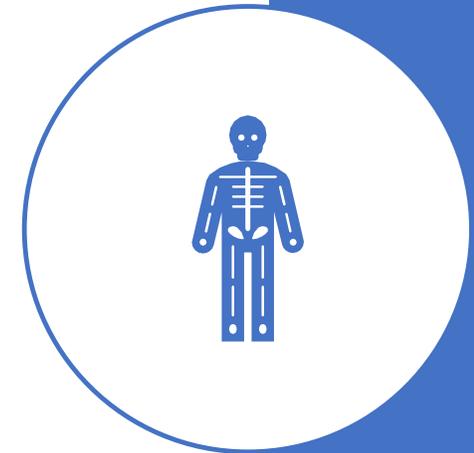
Haemarthrosis

- In a virgin joint may be due to recognised or unrecognised trauma
- In a post-acute or synovitic joint the most susceptible position for causing a re-bleed is end-range extension
- The same effects to joint cartilage are in play for the elbow as for all joints

- Repeated swellings of the joint capsule leave the elbow with reduced medial/lateral stability
- Increases rotational instabilities of the joint leading to greater risk of further injury be it haemophilic or orthopaedic
- Muscular wasting of the upper arm and to a lesser extent the forearm ensues with repeated episodes of bleeding
- Cartilage destruction and subchondral cyst formation precede widening of the epiphysis of the distal humerus

Synovitis of the Elbow

- Impossible to show on a picture when it is in its mild stages...which is part of the problem – and why musculoskeletal examination is so crucial
- Obscuring of the boney landmarks and filling in of the dimples on the posterior elbow are the first visible signs by traditional examination
- Physiotherapy is most amenable to mild - moderate synovitis – with assessment by palpation





What Tools are
Necessary...

What Tools are Complementary...

Batteries of validated outcome measures

- Meaningful to you?
- Meaningful to patients?
- Meaningful to payers?

POCUS

Electromedical devices

Methods of Assessment

- Clinical examination
 - Subjective, Inspection, Objective, Palpation, Functional
- HJHS – the elbow is one of the index joints included within the tool...are you assessing for research? Or clinical management?
- POCUS examination allows for precision assessment of the joint cartilage and synovium as well as angiogenesis; enhanced educational opportunities

Baseline vs. Post-Acute

- Annual assessments including functional and validated tools (eg) HJHS evaluation, allow for a clarified starting point and therefore target for rehab
- Inflammatory signs and symptoms should be assessed and managed first followed by elements of MSK recovery
- ROM – strength – proprioception is a common progression
...but not always the case...

Physiotherapy Management - Acute Bleed

Splinting of the elbow joint:

One of the most common applications for splinting is immobilisation

Duration of application remains debatable and is not an absolute...requiring the human/professional factor

Customisation of splints to the individual situation is critical

Proceed with Caution...

- Consider both short AND long term goals of immobilisation
- 50% decrease in elbow motion limits the function of the entire upper extremity by almost 80%

Sjoberg (1996), Acta Orthop Scand

- Overuse and disuse of a joint both result in degradation of articular cartilage
Leong et al (2011), FASEB J

Effective immobilisation will...

- Reduce pain
- Limit tissue damage
- Enhance the effect of factor or other treatments

Prolonged or unnecessary immobilisation will present the challenge of joint contractures and soft tissue stiffness

Apoptosis of chondrocytes

- Hand path alterations similar to those found in deafferented individuals were observed in subjects who had experienced electrophysical changes induced by twelve hours of upper limb immobilisation
- Upper limb immobilisation induces synaptic remodeling in the sensory and motor cortices

Huber et al (2006), Nature Neuroscience

Addressing Functional Recovery

Adherence

- Up to 67% of patients required to wear an upper extremity splint on a continual basis report non-adherence with the splinting regimen

Sandford (2008), J Hand Ther

- Maximising the convenience and comfort level of the splint is likely to impact the success of the treatment

Modalities – thermal, electrophysical

Systemic – breathing techniques, meditation, relaxation...

PHYSICAL – exercise of the right type at the right time is
capable of producing analgesia

Neuroplastic – graded motor imagery

Pain Management



The Impact of MSK Health Decline

- Many researchers and clinicians are now addressing balance issues with people with haemophilia as they age
- Immobilisation of the shoulder has a negative impact on balance and increases the risk of falling

Coleman et al (2010), J Geriatr Phys Ther

Why is Physiotherapy SO Crucial?

- First and foremost – it is the only form of treatment that in theory if not in practice is available to ALL people with haemophilia (PWH)
- There is a gap in what surgical options can provide for the elbow in comparison to other index joints in haemophilia
- Reportedly over 80% of PWH experience recurrent elbow bleeds but the rate of arthroplasty is a fraction of that of the knee

So why is joint replacement more limited in elbows compared to other joints?

J Shoulder Elbow Surg (2015) 24, 773-778



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Total elbow arthroplasty in bleeding disorders: an additional series of 8 cases

Anne J. Vochteloo, MD, PhD^a,
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Haemophilia

The Official Journal of the
World Federation of Hemophilia,
European Association for Hemophilia and
Allied Disorders and the Hemostasis &
Thrombosis Research Society



Outcomes in total elbow arthroplasty in patients with haemophilia at the
University of California, San Francisco: a retrospective review

Case Series of Elbow Arthroplasties in Haemophilia

HEMOPHILIC ARTHROPATHY OF THE ELBOW TREATED BY TOTAL ELBOW REPLACEMENT

A CASE SERIES

BY SRINATH KAMINENI, FRCS(ORTH), ROBERT A. ADAMS, MA, RPA,
SHAWN W. O'DRISCOLL, MD, PhD, AND BERNARD F. MORREY, MD

Investigation performed at the Department of Orthopedic Surgery, Mayo Clinic, Rochester, Minnesota

Arthroplasty of the elbow in haemophilia

P. J. Chapman-Sheath, P. Giangrande, A. J. Carr

From the Nuffield Orthopaedic Centre, Oxford, England



JOURNAL OF SHOULDER AND
ELBOW SURGERY

Long-term results of total elbow arthroplasty in patients with hemophilia

Lukas Ernstbrunner, MD^{a,b,c}, Andreas Hingsammer, MD^a, Mohamed A. Imam, MD, PhD^{a,c}, Reto
Sutter, MD^d, Brigit Brand, MD^d, Dominik C. Meyer, MD^d, Karl Wieser, MD^a
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40 combined TEA's

58% combined complication rate

COMPLICATION	NUMBER OF ELBOWS	PERCENT (%)
Infection	7	17.5
Ulnar Neuropathy	8	20
Axillary Vein Thrombosis	1	2.5
Persistent Pain	2	5
Compartment Syndrome	1	2.5
Poly Wear	2	5
Loosening	2	5

Normal X-Rays



Haemophilic Arthropathy



End Stage Joint Destruction



Multi-modal Approach to Physiotherapy

Increasingly Physiotherapy practice in haemophilia is drawing from other therapeutic areas...orthopaedics, sport medicine, neuro-rehab...

- Manual Therapy
- Kinesiotaping
- Acupuncture
- PNF
- Muscle energy techniques
- Hand Therapy Model
- Mirror therapy
- Holistic approaches

In addition to more traditional approaches within bleeding disorders care such as hydrotherapy, isometric/tonic/kinetic exercise, static and static progressive splinting, muscle stimulation, CPM...

Final Thoughts



- Early and comprehensive intervention at the elbow may be more important to maintenance of long term function than in any other major joint
- In factor-available environments the incidence of orthopaedic versus haemophilic elbow pathology is on the rise

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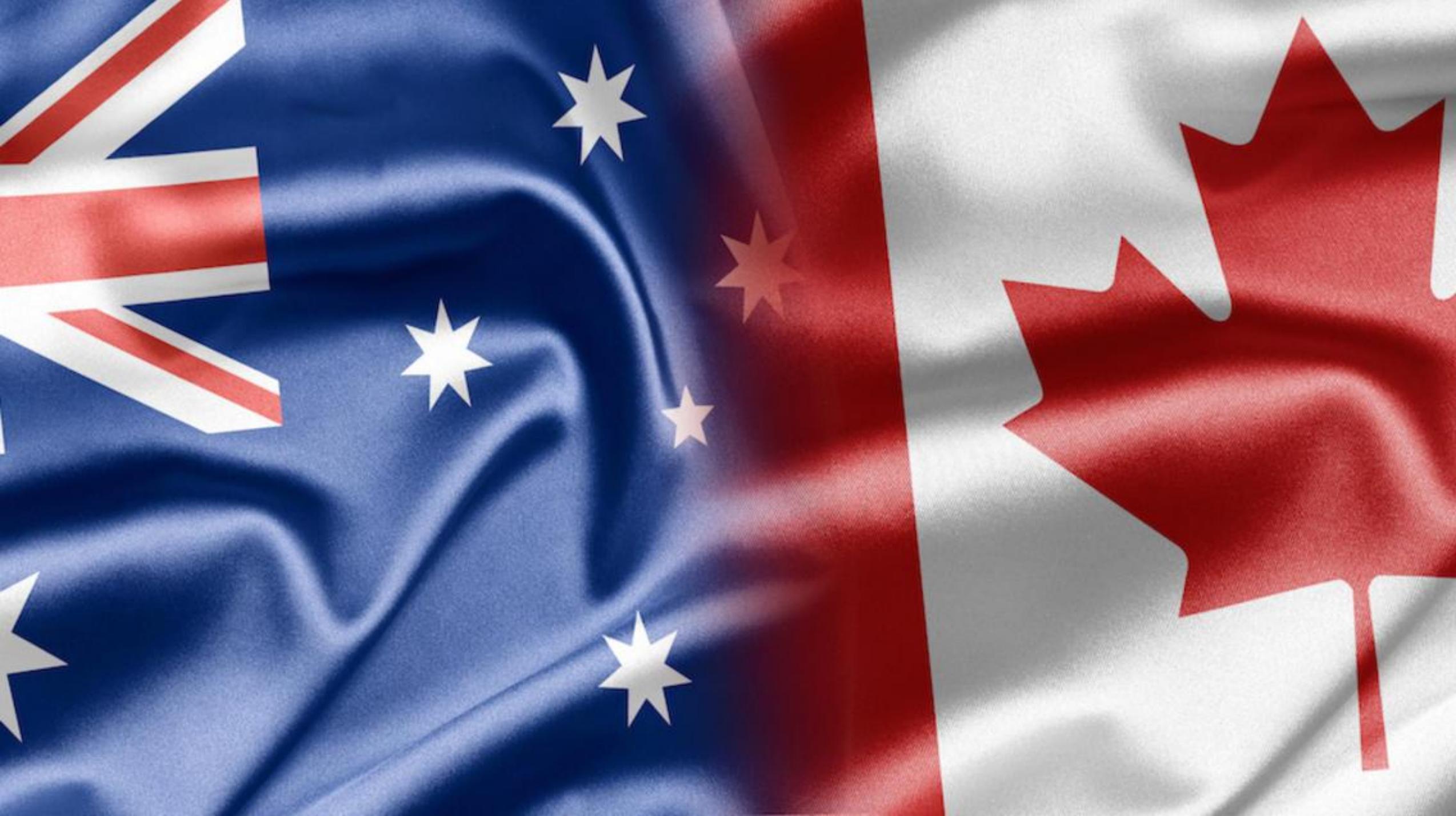
Muscle Bleeding

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The Unusual Suspects...

Congratulations Haemophiliacs!

- Rotator Cuff Pathology
- Radial Tunnel Syndrome
- Median Nerve Compression at the elbow
- Cubital Tunnel Syndrome
- Medial and Lateral Epicondylitis
- Radial Head fractures
- Intersection Syndrome
- DeQuervain's Tenosynovitis
- Scapholunate pathologies
- TFCC Injuries
- Wartenberg Syndrome ...!!!





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