48 year old male booked for removal tension band wires from elbow. Bikie club member with multiple previous traumas and ICU admission for major trauma. History of Von Willebrand's type 1 – diagnosed 1998, no follow up with haematology. Previously given Factor 8 on induction and 24 hours after, as 'non-responsive to DDAVP' (n.b. Found in about 10% of Type 1). Discussed with bleeding disorders Haematologist at the Mater and referred for follow up.

Info about Von Willebrand's disease:

- The most common inherited bleeding disorder, prevalence 1 in 100 but most asymptomatic
- Clinically significant 1 in 10,000
- Quantitative or qualitative defect in vWF
- vWF is a plasma glycoprotein that is involved in platelet adhesion and aggregation and carrier for factor 8 (decreases clearance from plasma)
- Deficiency leads to easy bruising, epistaxis, gum bleeding and bowel bleeding. Can cause prolonged bleeding post op.

Classification:

  - Type 1 (60-80%) – quantitative defect (mild to moderate bleeding disorder)
  - Type 2 (15-30%) – qualitative defect, with subtype dependent on platelet binding, factor 8 binding and size of vWF
  - Type 3 (5-10%) – absence of vWF (severe bleeding disorder)
  - Acquired vWF – associated with autoimmune disease e.g. lupus

- Autosomal inheritance, dominant or recessive depending on subtype
- Lab diagnosis: factor 8 level, vWF, collagen binding assay, EPG
- Can have thrombocytopaenia, prolonged bleeding time and APTT, but normal PT

Treatment:

  - DDAVP in type 1: stimulates release of vWF (n.b. need fluid restriction)
  - Tranexamic acid for minor procedures (e.g. dental work)
  - vWF/factor 8 concentrates
  - Platelet infusion if bleeding despite factor 8 and vWF replacement

Anaesthetic consideration (from CEACCP):

  - Multidisciplinary management comprising haematologist, surgeon, anaesthetist, physiotherapist, and occupational therapist.
  - Liaison with laboratory services to ensure that appropriate factor concentrates are available and in sufficient quantity. (Locally this is via the Haemophilia nurse at CMH.)
  - Elective surgery scheduled early during the week and preferably in the morning.
  - Preoperative clotting screen and specific factor assays depending on the type of bleeding disorder and preoperative transfusion of recombinant factors 30–60 min before the surgical procedure.
  - Perioperative avoidance of mucosal trauma, i.m. injections, maintenance of normothermia, and pressure point care.
  - Care with vascular access and invasive monitoring. Low threshold for use of ultrasound.
  - Avoidance of tachycardia and hypertension.
  - Risk–benefits for neuraxial block and regional blocks need to be assessed individually and in general avoided.
  - VTE prophylaxis by early mobilization, and mechanical deep venous thrombosis prophylaxis. Risk–benefits of pharmacological methods must be considered and discussed with the surgeon and haematologist.
  - Multimodal pain management, avoid non-steroidal anti-inflammatory drugs.