Pathophysiology, clinical, diagnosis and therapeutic aspects of acquired von Willebrand syndrome (AVWS)

Clinical features
- Bleeding diathesis usually occurs rather late in the life of persons with no past and family history of bleeding
- Main symptoms are mild to moderately severe mucocutaneous bleeding
  - ecchymosis
  - epistaxis
  - menorrhagia
  - gastrointestinal tract bleeding*
  *usually associated with the detection of angiodysplasia
- Or excessive bleeding following trauma or surgical procedures particularly when FVIII:C is low

Pathophysiology
- Most cases are due to an increased plasma clearance of VWF caused by such mechanisms as
  - antibodies
  - cell adsorption
  - shear stress
  - increased proteolysis
- Almost always in association with an underlying disorder

Diagnosis
- Usually based on the laboratory measurements used to diagnose inherited VWD (In the absence of a family history of bleeding)
- A defect in primary haemostasis is demonstrated by a prolonged skin bleeding time or prolonged closure time with PFA-100
- VWF multimer electrophoresis is warranted to demonstrate the defect of HMW multimers that helps to distinguish AVWS from type 1 VWD

Therapy
- Three main treatment goals for patients with AVWS
  - control of acute bleeding
  - its prevention in high-risk situations
  - achievement of a stable remission or cure

Hemostatic therapies in the AVWS associated with different underlying diseases

<table>
<thead>
<tr>
<th>Category</th>
<th>Treatment Options</th>
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</thead>
<tbody>
<tr>
<td>Cardiovascular</td>
<td>VWF/FVIII concentrates, antifibrinolytics</td>
</tr>
<tr>
<td>Lymphoproliferative</td>
<td>IgG MGUS</td>
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<td>HDIVlg</td>
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<td>Plasmapheresis, DDAVP, VWF/FVIII concentrates, antifibrinolytics, rFVIIa</td>
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<td>Myeloproliferative</td>
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<tr>
<td>Autoimmune</td>
<td>HDIVlg, DDAVP, VWF/FVIII concentrates</td>
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