International recommendations on the diagnosis and treatment of acquired hemophilia A

Acquired hemophilia A (AHA)

- rare bleeding disorder caused by neutralizing autoantibodies against coagulation factor VIII
- interference with coagulation function
- predisposition to severe, potentially life-threatening hemorrhage

Control of acute bleeding and prevention of injury that may provoke bleeding are top priorities in patients with AHA

### Hemostatic therapy

- Clinically relevant bleeding or before invasive procedures
  - rFVIIa 90 µg/kg every 2-3 h
  - APCC 50-100 U/kg every 8-12 h (max 200 per d)
  - rpFVIII 200 U/kg followed by tailored dosing
  - hFVIII 50-100 U/kg followed by tailored dosing

  - Close monitoring of FVIII activity

  - Clinical assessment of efficacy
  - Increase dosing interval if no further bleeding
  - Switch treatment option if ineffective

### Immunosuppressive therapy

<table>
<thead>
<tr>
<th>FVIII ≥ 1% and ≤ 20 BU/ml</th>
<th>FVIII ≤ 1% and &gt; 20 BU/ml</th>
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</thead>
<tbody>
<tr>
<td>Steroids alone 3-4 weeks</td>
<td>Steroids + CTX or rituximab 3-4 weeks</td>
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<tr>
<td>Add CTX or rituximab if not responding</td>
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Tiede et al., Haematologica, 2020