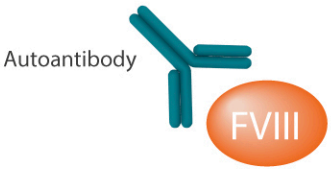
International recommendations on the diagnosis and treatment of acquired hemophilia A



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

If anti-porcine titer low or undetectable

If other options unavailable and low anti-human titer

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

Close monitoring of FVIII activity

- Clinical assesment of efficacy
- Increase dosing interval if no further bleeding
- Swith treatment option if ineffective

Immunosuppressive therapy

FVIII ≥ 1% and ≤ 20 BU/ml	FVIII ≤ 1% and > 20 BU/ml
Steroids alone 3-4 weeks	Steroids + CTX or rituximab 3-4 weeks
Add CTX or rituximab if not responding	Add CTX or rituximab if not responding

Tiede *et al.*, Haematologica, 2020