International recommendations on the diagnosis and treatment of patients with acquired hemophilia A

Angela Huth-Kühne, Francesco Baudo, Peter Collins, Jørgen Ingerslev, Craig M. Kessler, Hervé Lévesque, Maria Eva Mingot Castellano, Midori Shima, and Jean St-Louis

¹SRH Kurpfalzkrankenhaus Heidelberg gGmbH and Hemophilia Center, Heidelberg, Germany; ²Thrombosis and Hemostasis Unit, Niguarda Hospital, Milan, Italy; ³Arthur Bloom Haemophilia Centre, University Hospital of Wales School of Medicine, Cardiff University, Cardiff, UK; ⁴Center for Hemophilia and Thrombosis, Skejby University Hospital, Department of Clinical Biochemistry, Aarhus, Denmark; ⁵Georgetown University Hospital, Lombardi Cancer Center, Division of Hem/Onc, Washington, DC, USA; ⁶Department of Internal Medicine, Centre Hospitalier Universitaire de Rouen-Boisguillaume, Rouen, France; ⁷Regional Universitary Hospital Carlos Haya, Division of Hematology, Málaga, Spain; ⁸Department of Pediatrics, Nara Medical University, Nara, Japan, and ⁹Hématologie-Oncologie, Hôpital Maisonneuve-Rosemont, Montréal, QC, Canada

Citation: Huth-Kühne A, Baudo F, Collins P, Ingerslev J, Kessler CM, Lévesque H, Mingot Castellano ME, Shima M, and St-Louis J. International recommendations on the diagnosis and treatment of patients with acquired hemophilia A. Haematologica 2009; doi:10.3324/haematol.2008.001743

Online Supplementary Table A. Questions addressed in this survey.

Diagnosis

- 1. Which clinical symptoms or laboratory parameters should trigger a consideration that acquired hemophilia may be present?
- 2. Which clinical symptoms or laboratory parameters may be employed to establish a differential diagnosis of acquired hemophilia?
- 3. Which aPTT level should trigger additional investigation?
- 4. Is FVIII:C level relevant for the diagnosis, management or prediction of bleeding in patients with acquired hemophilia?
- 5. Is inhibitor titer relevant for the diagnosis, management or prediction of bleeding in patients with acquired hemophilia?
- 6. How should a patient with suspected or confirmed acquired hemophilia be managed?
- 7. Under what circumstances should a patient be referred to a specialized hemophilia center?
- 8. Which procedures should be avoided in a patient with suspected or confirmed acquired hemophilia?
- 9. Which clinical parameters should receive special consideration in the management of elderly patients with unexplained bleeding?

Anti-hemorrhagic treatment

- 1. Which criteria can be used to define severe bleeding in the patient with acquired hemophilia?
- 2. Which acquired hemophilia patients require anti-hemorrhagic therapy?
- 3. Which anti-hemorrhagic treatment is appropriate in the bleeding patient with acquired hemophilia?
- 4. Which dosage is appropriate for the treatment of bleeding in acquired hemophilia?
- 5. Which combination therapies are appropriate for the treatment of bleeding in acquired hemophilia?
- 6. How long should different types of bleeding be treated in the patient with acquired hemophilia?
- 7. How can bleeding and response to anti-hemorrhagic therapy be monitored in the patient with acquired hemophilia?
- 8. Which approaches can be employed to prevent bleeding?
- 9. How can re-bleeding be identified in the acquired hemophilia patient?
- 10. How does the management of bleeding in the acquired hemophilia patient differ in the presence of significant co-morbidities?
- 11. Which adverse events are likely to be associated with the treatment of hemorrhagic events in acquired hemophilia patients?

Inhibitor eradication

- $1. \quad Should all \ patients \ with inhibitor \ and \ a \ diagnosis \ of \ acquired \ hemophilia \ receive \ immunosuppressive \ the \ rapy?$
- 2. Is the inhibitor level a criterion that determines whether immunosuppressive therapy should be initiated?
- 3. At what time point should immunosuppressive therapy be initiated?
- 4. Which criteria can be used to assess the effectiveness of immunosuppressive therapy?
- 5. How can response to immunosuppressive therapy be monitored in the patient with acquired hemophilia?
- 6. Which criteria should be used to determine when treatment regimen should be altered?
- 7. At what time point should alternative regimens be considered?
- 8. Which immunosuppressive regimens should be considered in which patient groups?
- 9. How should acquired hemophilia patients be monitored following remission?
- 10. How should patients with excessive FVIII:C levels following eradication be managed?
- 11. Should ITT play a role in treating patients with acquired hemophilia?