INTRODUCTION

Emicizumab is a bispecific, humanized, recombinant antibody designed to mimic the cofactor function of coagulation factor (F) VIII. It consists of two monospecific, antigen-binding arms that recognize activated FIX and FX, respectively, placing them into appropriate positions to support the activation of FX. Emicizumab is approved for the prophylaxis of bleeding in congenital hemophilia A (HA) and in HA with inhibitors (HA-I). As compared with on-demand treatment, emicizumab reduced the annualized rate of treated bleeding events in HA and HA-I by >90% and 88%, respectively.

Emicizumab is injected subcutaneously in intervals of 1 to 4 weeks, which is more convenient than conventional intravenous clotting factor replacement.

This article examines the potential role of emicizumab in patients with acquired hemophilia A (AHA), a severe bleeding disorder caused by autoantibodies against FVIII. State-of-the-art management is based on bypassing agents (recombinant factor VIIa, activated prothrombin complex concentrate) and recombinant porcine FVIII; immunosuppressive therapy (corticosteroids, rituximab, cyclophosphamide) is used to suppress autoantibody formation. Case reports and one series suggest that emicizumab can reduce the risk of bleeding and the requirement for hemostatic therapy until remission of AHA is achieved. Further, it may allow to postpone the start of immunosuppressive therapy or to use less intense regimens. However, the risk-benefit assessment of emicizumab in AHA is difficult because demographic and clinical characteristics are different compared with congenital hemophilia. Prospective clinical trials are needed before the use of emicizumab can be recommended in AHA.
situations, review state-of-the-art treatment according to international recommendations,\textsuperscript{5,6} and discuss the potential role of emicizumab.

2 | A PRIMER TO AHA

Inhibitors in AHA are neutralizing autoantibodies against FVIII that develop spontaneously or in association with other autoimmune disorders, malignancy, pregnancy, infection, or certain medications.\textsuperscript{7} The disorder is rare, with an estimated incidence of 1 to 6 cases per million per year.\textsuperscript{8,9} Patients typically present with a sudden onset of bleeding and a prolonged activated partial thromboplastin time (APTT). Large subcutaneous hematomas, occurring spontaneously or after minor trauma, are characteristic of the disease (Figure 1A).\textsuperscript{10} Postoperative bleeding (Figure 1B) and deep muscle hematomas (Figure 1C) are also common. Men and women of all ages can be affected, with a mean age of 74 years at the time of diagnosis and a smaller peak in women of 20 to 40 years because of higher incidence in the postpartum period (Figure 1D).\textsuperscript{11} Owing to the advanced age of many patients, coexisting disorders are common, including cardiovascular risk factors (Figure 1E).

The first and foremost treatment goal in AHA is to control acute bleeds. Bypassing agents (BPAs), including recombinant factor VIIa (rFVIIa) and activated prothrombin complex concentrate (APCC), as well as recombinant porcine FVIII (rpFVIII, susoctocog alfa) are highly efficacious.\textsuperscript{12} However, the risk of bleed recurrence after successful treatment is high,\textsuperscript{13} and prophylactic replacement therapy has not been established. Owing to their short half-life, BPAs are not ideal for prophylaxis, although their use has occasionally been reported.\textsuperscript{14,15} Despite its longer half-life, rpFVIII is not a candidate for prophylaxis either because of cross-reacting inhibitors that can be present at the time of diagnosis\textsuperscript{16} or develop during treatment.\textsuperscript{17,18}

The second treatment goal is therefore to eradicate the autoantibodies, which can be achieved by immunosuppressive therapy (IST) with glucocorticoids, rituximab, cyclophosphamide, or combinations thereof.\textsuperscript{19} Partial remission is achieved by 83% of patients after a median of 31 days but adverse events of IST, most often infection, are very common.\textsuperscript{11} The 1-year survival rates in the prospective French SASHA and Germany-Austrian GTH studies were 62% and 68%, respectively, with fatal IST complications being reported in 12% and 16%.\textsuperscript{11,20}

**FIGURE 1** Spotlight on AHA. (A) Typical subcutaneous and intramuscular hematomas. (B) Severe postoperative bleeding. (C) Computed tomography scan of right iliopectineus muscle hematoma. (D) Age and gender distribution of patients with AHA. (E) Venn diagram of cardiovascular diseases and risk factors in patients with AHA. Data in panels D and E were derived from GTH-AH 01/2010 study, $n = 102\textsuperscript{11}$
TABLE 1  Summary of potential advantages and caveats of using emicizumab in AHA

| Potential advantages                                                                 |
|--------------------------------------------------------------------------------------|
| Shorter time to resolution of initial bleed                                          |
| Reduced dosing requirements of other hemostatic drugs                                |
| Reduced risk of recurrent bleeds                                                     |
| Postpone or displace IST in frail or acutely ill patients                             |
| Long-term prophylaxis in patients with chronic AHA resistant to IST                  |
| Earlier hospital discharge and outpatient management with subcutaneous dosing        |

| Caveats                                                                                   |
|------------------------------------------------------------------------------------------|
| Time to achieve effective plasma concentration                                          |
| Interaction with bypassing agents                                                        |
| Thromboembolic risk                                                                       |
| Risk of anti-drug antibodies                                                              |
| Unknown safety in pregnancy                                                               |
| Interference with laboratory monitoring to detect remission                              |
| Interference with laboratory monitoring of rpFVIII                                       |
| Laboratory monitoring not established                                                     |
| Unknown efficacy during surgery and interventions                                         |

3  | SCENARIO 1: RECURRENT BLEEDING

A 72-year-old man was admitted with a large upper thigh hematoma after a minor bicycle accident 1 month before. He described himself as previously healthy and still very active, and he had never experienced abnormal bleeding earlier in his life. The hematoma had been continuously extending over time, and his hemoglobin (Hb) was 7.2 g/dL at the time of admission. Noting an APTT prolongation to 110 seconds upon admission, laboratory workup revealed a reduced FVIII activity of <1% and a Bethesda inhibitor titer of 80 BU/mL. Clinical and laboratory workup excluded an underlying cause for inhibitor formation. A diagnosis of idiopathic AHA was made and therapy started with rFVIIa at 90 µg/kg body weight every 2 hours. He also received prednisolone 1 mg/kg daily and 4 doses of rituximab 375 mg/m² once weekly. The hematoma initially improved but bleeding recurred several times when reduction of rFVIIa dosing was attempted. After 4 weeks in the hospital, FVIII activity was still <1%, with an inhibitor or 13 BU/mL, and he experienced a new bleed after venipuncture at his right forearm.

Although the efficacy of BPA and rpFVIII is consistently reported as very high in the initial treatment of bleeds, therapy is often needed much longer than in HA-I as indicated by an average bleed duration of >1 week.13 Nearly 60% of patients experienced recurrent bleeds; only achieving remission of AHA protected from bleeding effectively.13 Severe hemorrhage at presentation, frequent bleed recurrences, and the high dosing requirements of BPA and rpFVIII contributed to the frightening image of AHA and established the need for intense IST.

International recommendations would suggest the addition of cyclophosphamide in the described case, in whom prednisolone and rituximab had not resulted in remission of AHA after 4 weeks.5,6 The average time to achieve remission in this patient, considering the baseline FVIII <1% and inhibitor titer >20 BU/mL, would be 6 weeks.11

During this time, prophylaxis with emicizumab could potentially help to shorten the time needed to repair lesions from the initial bleeding event and to reduce the risk of recurrent bleeds. Beneficial effects for the patient could include faster recovery and shortened hospital stay, and for payers decreased consumption of clotting factor concentrates (Table 1).

The bleeding risk in AHA is very high in the first weeks after diagnosis (Figure 2A).12 Using the approved dosing regimen of emicizumab 3 mg/kg weekly for 4 weeks, followed by 1.5 mg/kg weekly, resulted in steady-state plasma concentrations of emicizumab after 4 to 5 weeks in patients with HA-I and HA (Figure 2B).3,4 The minimum plasma concentration to prevent bleeds in AHA is unknown; it is therefore unclear whether the current dosing regimen would be effective during the first weeks of treatment, when the bleeding risk is highest.

If the patient responds to IST, a reactive increase of FVIII activity may occur while emicizumab is still present because of its long half-life (approximately 30 days).21 It is unclear whether the combination of emicizumab with increased FVIII levels over prolonged periods is safe or may pose an additional thromboembolic risk.

Effects on standard laboratory tests also need to be considered in this scenario. In the presence of emicizumab, the APTT is artificially shortened and APTT-based clotting factor assays and inhibitor tests cannot be used.22,23 This is important during follow-up to detect remission and potential recurrence of AHA. FVIII activity could be monitored with chromogenic assays using bovine components, and the inhibitor titer be followed by modified Bethesda assay based on such chromogenic assays,24 or by ELISA,25 but few centers currently have such assays routinely available.

4  | SCENARIO 2: CATASTROPHIC BLEEDING AND SURGERY

A 29-year-old woman was diagnosed with AHA 2 days after cesarean section, when she continued to bleed from the surgical wound despite tranexamic acid. Hematologists was involved because of diffuse bleeding during surgical revision and prolonged APTT FVIII activity was 8%. She received rpFVIII 200 IU/kg every 4 hours, resulting in FVIII activity increased to 50% to 80%, but bleeding continued, and 14 units of packed red blood cells were required in the next 18 hours to keep her Hb >8 g/dL. During a second revision, hysterectomy was performed, and 2 L of hematoma evacuated. She needed vasopressor support and resuscitation with packed red blood cells, fibrinogen, and prothrombin complex concentrate. The surgical site was packed, and she was transferred to the intensive care unit, where she continued to need vasopressors and did not stabilize her Hb. FVIII activity was 20%, 1 hour after the last dose of rpFVIII 200 U/kg, and therapy was switched to rFVIIa 90 µg/kg every 2 hours.
This patient was in a life-threatening condition. Although rpFVIII resulted in an increase of FVIII activity, the patient could not be stabilized. This may not necessarily be seen as a failure of rpFVIII because severe postpartum bleeding can have catastrophic dynamics on its own resulting from local tissue damage, blood loss, factor consumption, dilution, disseminated intravascular coagulation, or hyperfibrinolysis. Nevertheless, switching the treatment of AHA appeared justified because the poor recovery after rpFVIII dosing (postinfusion activity 20%) suggested cross-reacting inhibitors. International recommendations recommend switching between rpFVIII and BPA if clinical or laboratory monitoring indicates insufficient efficacy.

Is there any role for emicizumab in this setting? Probably not in the setting of acute, catastrophic bleeding. First of all, because of its slow onset of action emicizumab is not useful for on-demand therapy of acute bleeds. Second, hemostatic resuscitation in massive bleeding emergencies is multifaceted and difficult enough to monitor. It is not unlikely that emicizumab would add more risk than benefit in this setting.

Even under less dramatic circumstances, starting emicizumab at the time of the initial bleed would probably require combination with BPA or rpFVIII in most cases. In HA-I, the combination of emicizumab with APCC > 100 U/kg for >24 hours resulted in thrombotic microangiopathy and should avoided, probably also in AHA. Combining emicizumab with rFVIIa has been reported as safe in HA-I; however, owing to the more frequent cardiovascular risk factors in elderly patients with AHA, caution should be exercised.

FIGURE 2  Possible gap between bleeding risk in AHA and emicizumab plasma concentrations with dosing regimens established in congenital hemophilia A. (A) Risk of newly occurring bleeds in patients with AHA. Data were derived from GTH-AH 01/2010 study, n = 102. (B) Mean emicizumab trough concentration in patients with congenital hemophilia A and inhibitors receiving 3 mg/kg once weekly for 4 wk, followed by 1.5 mg/kg once weekly. Modified after Oldenburg et al.
| First Author, Publication Date, Reference | Patient Age, Sex | Reason for Emicizumab | Dosing of Emicizumab | Concomitant Use of CFC | IST | CVRF | Efficacy | Safety |
|------------------------------------------|------------------|-----------------------|----------------------|------------------------|-----|------|----------|--------|
| Möhnle 14/06/19 36                       | N = 1 83 y, M    | Severe bleeding       | 1× 3 mg/kg, 2× 1.5 mg/kg | rFVIIa                 |     |      | Bleeding stopped, no bleeding | Died 36 d after last dose (unknown cause) |
| Dane 12/07/19 37                         | N = 1 72 y, M    | Prophylaxis for planned PCI | 4× 3 mg/kg weekly, 5 mo 1.5 mg/kg weekly | -                       |     |      | No bleeding | No AE   |
| Al-Banaa 19/07/19 38                     | N = 1 87 y, F    | Prophylaxis            | 4× 3 mg/kg weekly, 2 mo 1.5 mg/kg weekly | -                       |     |      | No bleeding | No AE   |
| Hess 08/05/20 39                         | N = 1 91 y, M    | Recurrent bleeding     | 4× 3 mg/kg weekly, 6 mo 1.5 mg/kg weekly | Glucocorticoids, CTX   |     |      | No bleeding | No AE   |
| Knöbl 09/08/20 40                        | N = 12 51–87 y (range), 6 F, 6 m | Various                | 1.7-3.5 mg/kg for 3-9 doses (range) | rFVIIa, human FVIII     |     |      | Initial bleeding stopped after median 3 d (range 2-15) | Minor stroke in 1 patient on concomitant rFVIIa, 1 patient died of peritonitis |

Abbreviations: AF, atrial fibrillation; CFC, clotting factor concentrate; CTX, cyclophosphamide; CVRF, cardiovascular risk factors; FVIII, factor VIII; IST, immunosuppressive therapy; NSTEMI, non-ST elevation myocardial infarction; PCI, percutaneous coronary intervention; rFVIIa, recombinant factor VIIa.
same might be true for rFVIII, in particular if high peak activity occurs during treatment as reported in the pivotal clinical trial of this drug.

The use of emicizumab would create some difficulties in laboratory monitoring, which is important during treatment with rFVIII. One-stage clotting assays of FVIII activity yield artificially high results in the presence of emicizumab. Chromogenic FVIII tests using bovine components are insensitive to emicizumab but underestimate the activity of rFVIII by about 50%. Thus, monitoring of rFVIII would not be possible in patients treated with emicizumab.

5 | SCENARIO 3: THE FRAIL PATIENT

An 83-year-old woman developed bruising and gastrointestinal bleeds. She was admitted to the hospital because of progressive anemia with an Hb of 8.2 g/dL. Diagnostic workup revealed a FVIII of 6% and an inhibitor of 4 BU/mL. She had a long history of rheumatoid arthritis, lived in a day-care facility, and was wheelchair bound. She also had chronic obstructive pulmonary disease, arterial hypertension, and reduced renal function. Over the past 2 years, several episodes of exacerbation of her pulmonary disorder had occurred. Her medication included prednisolone 20 mg per day, torsemide, enalapril, and inhalative glycopyrronium bromide/formoterol.

Although IST is recommended in all patients with AHA, particular caution should be exercised in frail patients. Disability, pulmonary disorder, and chronic glucocorticoid use are risk factors for IST complications, in particular infection, in this patient. Mortality was 54% in patients with infections in the GTH study.

Provided emicizumab was sufficiently effective to prevent bleeds in this patient, the need for inducing remission of AHA with IST would be reduced. At the least, IST could be postponed until she recovered from the acute situation, or a milder regimen could be chosen, accepting a longer time to remission while being protected from bleeding by emicizumab. Of note, Lottenberg et al observed spontaneous remission of AHA in a small series of patients, who never received IST.

6 | SCENARIO 4: CHRONIC AHA

A 60-year-old woman had developed AHA 20 years ago. Several attempts to induce remission with IST were unsuccessful, including glucocorticoids, rituximab, and mycophenolate mofetil. FVIII activity had been <1% in recent years, with an inhibitor titer undulating between 5 and 10 BU/mL. Concomitant disorders included obesity, arterial hypertension, atrial fibrillation, and heart failure New York Heart Association grade II. She had experienced several severe bleeding episodes, including iliopsoas muscle hematoma, which had been treated successfully with rFVIIa.

Although most patients achieve remission of AHA after IST, the disease can be refractory or chronically recurrent in some. The bleeding tendency in these patients is highly variable but may involve frequent or severe symptoms such as in this case. Short-term prophylaxis with APCC has been described but may not be fully effective and is associated with high cost and treatment burden. This situation resembles that of HA-I, in which emicizumab has been considered a breakthrough that changed the life of many patients.

Notwithstanding the medical need in this setting, the efficacy and safety of emicizumab needs to be established in clinical studies and long-term observation. The patient had atrial fibrillation and did not receive anticoagulation because of AHA. Whether withholding anticoagulant treatment would be safe while on emicizumab remains speculative, in particular if she would need additional hemostatic drugs in case of breakthrough bleeding. Other potential caveats of using emicizumab in AHA are summarized in Table 1.

7 | REPORTED USE OF EMICIZUMAB IN AHA

Since emicizumab was launched in the European union in 2018, four single-case reports and one retrospective series of patients have been published (Table 2). At least one clinical trial is currently planned and estimated to be completed in 2022 (NCT04188639).

7.1 | Safety

Most of the 16 patients were of advanced age and had cardiovascular risk factors. In some cases, rFVIIa or FVIII concentrates were concomitantly given because of bleeding or for surgical prophylaxis. One thromboembolic event was reported in one of the 16 patients. This case, as part of the series by Knöbl et al, required repetitive rFVIIa because of surgical procedures while on emicizumab, developed a minor stroke and recovered without sequelae. No other cases of thromboembolic events were reported. However, the follow-up duration after achieving remission was usually short or not specified.

One death was reported in the Knöbl series resulting from peritonitis 45 days after the last dose of emicizumab. The case reported by Möhnle et al died 36 days after the last of three doses of emicizumab. In the report by Dane et al, emicizumab was started in a patient with recurrent acute coronary syndrome from stenosis of a bare-metal stent. This patient with chronic IST-resistant AHA had previously been bleeding while on APCC and dual antiplatelet therapy after stenting. Planned drug-eluting stent implantation was attempted after loading with emicizumab rather than using BPA. No bleeding or thromboembolic event occurred after the procedure. In summary, these reports appear to support that emicizumab can be used safely in patients with AHA, including those of advanced age and with cardiovascular risk factors, but prospective clinical trials with longer follow-up are needed to establish its safety.
in the scenario of concomitant use of BPA and after achieving remission of AHA.

7.2 | Efficacy

In the single-case reports, recurrent bleeding was not described after starting emicizumab. In the Knöbl et al series, bleeding stopped a median of 3 days after starting emicizumab. It was further noted that no bleeding occurred after the FVIII activity (measured with a chromogenic assay containing human components) had increased to >5% in these patients. In summary, these data speak for a good hemostatic efficacy of emicizumab in patients with AHA.

This is in line with the observed ex vivo activity of emicizumab in plasma from AHA patients. Takeyama et al reported that adding emicizumab at 30 µg/mL improved peak thrombin generation to median 49.7% (range 25.8%-94.6%) of pooled normal plasma. Even low concentrations of emicizumab 2.5 µg/mL resulted in notable improvements of median 13.9% (range 7.9%-20.7%). Nevertheless, a suitable dosing regimen of emicizumab in patients with AHA remains to be established.

8 | CONCLUSIONS

Emicizumab may address unmet clinical needs in the treatment of AHA. The drug has the potential to shorten the duration of initial bleeds, prevent recurrent bleeding, and reduce the need for other hemostatic therapy. Moreover, efficacious prophylaxis with emicizumab may alleviate the need for early and intense IST, in particular in frail patients. Ex vivo studies, case reports, and a small retrospective series suggest that emicizumab is safe and efficacious in AHA. However, the reported experience may suffer from confounding factors and should be confirmed in prospective clinical trials. Given the difficult risk profile of patients with AHA, international recommendations argued against its use outside clinical trials. Until such trials will have been reported, the risk-benefit assessment of emicizumab in AHA remains speculative.

CONFLICT OF INTEREST

Dr. Tiede received research funding, honoraria for lectures, and fees for consultancy from Chugai, Novo Nordisk, Roche, and Shire/Takeda. Dr. Kemkes-Matthes received consultancy honoraria and speaker fees from Bayer, Novo Nordisk, Roche, and Shire/Takeda. Dr. Knöbl received consultancy honoraria, speaker fees or travel grants from Novo Nordisk, Shire/Takeda, CSL Behring, Roche, Sanofi, and Technoclone.

AUTHOR CONTRIBUTIONS

All authors developed and discussed the clinical scenarios. Andreas Tiede performed the literature search and wrote the manuscript. All authors critically revised the manuscript and approved of its final version.
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