Radboud Repository



PDF hosted at the Radboud Repository of the Radboud University Nijmegen

The following full text is a publisher's version.

For additional information about this publication click this link. http://hdl.handle.net/2066/202652

Please be advised that this information was generated on 2020-09-10 and may be subject to change.

CORRECTION



Correction to: Eculizumab in atypical hemolytic uremic syndrome: strategies toward restrictive use

Kioa L. Wijnsma 1 · Caroline Duineveld 1,2 · Jack F. M. Wetzels 2 · Nicole C. A. J. van de Kar 1

Published online: 15 January 2019

© The Author(s) 2019

Pediatric Nephrology

https://doi.org/10.1007/s00467-018-4091-3

The original version of this article unfortunately contained two mistakes. The presentation of Table 1 and Fig. 1 was incorrect. The corrected versions are given below.

Table 1 Eculizumab dosage regimen, standard therapy according to EMA/FDA

Weight category	Induction phase	Maintenance phase
Above 40 kg	900 mg, every week, for 4 weeks	1200 mg, in fifth week, every 14 days thereafter
30 to < 40 kg	600 mg, every week, for 2 weeks	900 mg, in third week, every 14 days thereafter
20 to < 30 kg	600 mg every week, for 2 weeks	600 mg, in third week, every 14 days thereafter
10 to < 20 kg	300 mg once	300 mg, in second week, every 14 days thereafter
5 to < 10 kg	300 mg once	300 mg, in second week, every 21 days thereafter

Eculizumab has to be administrated intravenously

EMA European Medicines Agency, FDA Food and Drug Administration

The online version of the original article can be found at https://doi.org/ 10.1007/s00467-018-4091-3

- Radboud Institute for Molecular Life Sciences, Amalia Children's Hospital, Department of Pediatric Nephrology, Radboud University Medical Center, P.O. Box 9101, 6500, HB Nijmegen, The Netherlands
- Department of Nephrology, Radboud University Medical Center, Nijmegen, The Netherlands



Patient with TMA

In anticipation of diagnostics to rule out other causes of TMA^a &
Strong suspicion aHUS

Eculizumab treatmentb

Standard protocol, dose therapy depending on body weight.



Evaluation therapy after 3 months^c

In case the patient is in remission (normalized hematological parameters, normalized and/or stabilized kidney function and proteinuria, normal blood) adjustment of eculizumab therapy can be considered



Withdrawal or taperingd:

Scenario 1-5 figure 2



Monitoring after therapy adjustment

Regular workup at every hospital visit is required or in case of discontinuation after 1,2,3,6,9 &12 months after withdrawal. Between hospital visits, dipstick analysis and/or blood pressure (aim for p50 in children or below 130/80mmHg in adults) monitoring 3 times per week at home is advised.

Fig. 1 Treatment algorithm. After adequate exclusion of other causes of thrombotic microangiopathy (TMA) such as thrombocytopenic purpura (TTP), Shiga toxin-producing *Escherichia coli*-hemolytic uremic syndrome (STEC-HUS), or secondary TMA and in patients with strong suspicion of atypical hemolytic uremic syndrome (aHUS), eculizumab treatment should be started within 24 h after presentation. When the patient is stable and in remission, withdrawal or tapering can be considered, depending on patient characteristics (see Fig. 2). After therapy adjustment, strict monitoring is essential. NB in case of antibodies against complement factor H, a different treatment protocol has to be initiated as described by Loirat et al. [1]. a, For extensive overview of practical diagnostics approach for TMA, see Fakhouri et al. [3], b, Treatment should preferably be started within 24 h after presentation.

In adults with first episode of aHUS in native kidney, treatment with plasma exchange (PE) for 4 days (high volume PE with 1.5 plasma volume) is advised to allow diagnosis of secondary causes of aHUS. Adolescents may be considered adults [33]. After exclusion of secondary causes of aHUS and if the patient does not show a favorable response after 4 days of PE, treatment should be switched to eculizumab. Starting treatment with eculizumab within 7 days after presentation in PE-resistant patients was effective in the clinical trials [32]. In case the patient is PE sensitive, PE should be tapered and discontinued in the course of 1 month [9, 10]. c, Improvement of platelets and lactate dehydrogenase (LDH) is expected within 2–4 weeks. If no response, consider alternative diagnosis or inefficacy of eculizumab (C5 polymorphism

Open Access This article is distributed under the terms of the Creative Commons Attribution 4.0 International License (http://creativecommons.org/licenses/by/4.0/), which permits unrestricted use, distribution, and reproduction in any medium, provided you give appropriate credit to the original author(s) and the source, provide a link to the Creative Commons license, and indicate if changes were made.

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

