CORRESPONDENCE

Hemoglobinopathies: Clinical Manifestations, Diagnosis, and Treatment

by Prof. Dr. med. Elisabeth Kohne in volume 31-32/2011

New Recommendations

In the time interval between completion and submission of the article in April 2010 and its publication on 9 August 2011, the revised guidelines from the Association of the Scientific Medical Societies in Germany for thalassemias (AWMF-Reg No 025/017) and sickle cell disease (AWMF-Reg No 025/016) were published, as well as a new S2 guideline on secondary iron overload in patients with congenital anemias (AWMF-Reg No 025/029) (1–3). Some of these new recommendations deserve to be mentioned here as they were not included in the guideline recommendations accompanying the article. For thalassemia this concerns in particular the recommendation of an annual non-invasive test to measure the hepatic iron concentration by MRI or SQUID (Superconducting Quantum Interference Device) from the start of iron elimination treatment, as well as cardio-MRI for the purposes of a basic functional cardiological examination and assessment of myocardial iron overload from the age of 10 years (2, 3). International studies have found that both tests are essential in the optimal management of thalassemia major. In sickle cell disease, transcranial Doppler ultrasonography is useful for ensuring early detection of an increased risk of neurological infarction, resulting in regular prophylactic transfusion treatment (1). Details can be found in the original guidelines (1–3).

Among the available medications to treat iron overload, the drug deferiprone was not mentioned (4). This oral chelator is approved for the treatment of patients with thalassemia major in whom therapy with deferoxamine is inadequate. Deferiprone is able to bind iron within cells, for example, in cardiomyocytes, and to eliminate it from the cell. The most feared adverse effect is the rare development of severe neutropenia (0.5–2%); for this reason, weekly differential blood counts are recommended (3). In the article, the side effect agranulocytosis was attributed to the drug deferasirox (4). This is not supported by current study data.

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Conflict of interest statement

PD Cario participates in the pediatric advisory board on deferasirox on behalf of Novartis and has received expenses from Novartis, Sobi, and Amgen, as well as third-party funding from Novartis.

The author of the article has chosen not to publish a reply.