Correspondence

The survival analysis of beta thalassemia major patients in the South East of Iran

To the editor

I read with interest the article by Roudbari et al[1] on the survival analysis of beta thalassemia major (BTM) patients in the South East of Iran. It is of no doubt that the longevity of patients with BTM has remarkably improved in recent years as a result of better understanding the genetic, clinical, diagnostic, and therapeutic contexts of the disease. However, complications secondary to the disease itself, or its treatment are still cumbersome. I have comments concerning the aforementioned article.

The authors stated that the proportion of thalassemic patients in the South East of Iran were until 5 (97.9%), 10 (97%), 15 (92.1%), 20 (81.2%), and 25 (59.3%) years. These figures are actually astonishing. They probably reflect the better compliance of patients on one side, and the adequacy of applied medical care to the patients on the other. Indeed, we have no similar data in Iraq. However, I found through daily practice that most patients with BTM were unlikely to pass the second decade of life, even before catching puberty. The main reasons of death are related to: hemosiderosis with all its aftermaths due to the paucity of chelating therapy, and anemia due to the unavailability of pooled blood.

Growth faltering is characteristic in badly or poorly managed patients with BTM.[2] It is an important foreseeable limitation for life expectancy. I wonder whether the authors have any idea about the growth parameters of the studied patients. In Iraq, it was noticed that the growth deficits were statistically significant in female thalassemics, compared to male thalassemics that started from the second year of age onward for weight for age, and weight for height/length indices, and from the third year of age onward for height/length for age index. This might be attributed to the presence of more members affected in the family, male preference in the society, and parental pessimistic future view towards the disease.[3] This gender difference in anthropometric indices will probably undermine the quality of life of thalassemic patients, and hence, their survival.

Enhancing the longevity of patients with BTM requires multi-disciplinary team cooperation. Regular blood transfusion, planned chelating therapy, improving nutritional status, psychosocial support, parental education, adopting proper antibiotic coverage along with the prophylactic vaccines in splenectomized patients, screening donated blood for potential pathogens, initiation of neonatal screening programme for hemoglobinopathies, and prenatal diagnosis are requested campaigns to be followed. These campaigns will obviously ameliorate the severity, or abort the birth of new cases of BTM. Finally, bone marrow transplantation, and peripheral blood stem cell transplantation[4,5] remains the radical curative option for patients with BTM.

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Reply from the Author

The authors had thoroughly checked the data collected from Zahedan Thalassemia Center, and the results of the research once more, and they are correct, especially in the patient's survival proportions. Also, these proportions are the same as other researches performed in different parts of Iran, and on patients with different ages.[6,7] The increased survival proportions that are seen in the thalassemia patients are due to the early diagnosis of the disease at the Zahedan Center of Special Disease. In this center, the treatment of diseases such as thalassemia, diabetes, and other diseases, is free for all.

The thalassemia patients are visited twice a week by the doctors in the center, and those with hemoglobin under 9 gram/dl will be transfused with blood that was screened against different diseases before transfusion. Transfusions in newborns are started at the age of 4-6 months, which can increase the patients' survival, and life quality. Furthermore, these early transfusions can prevent the facial bone changes occurring in patients with BTM, and also decreases the comorbidities, especially the diseases of the heart and lungs.

Using Desferal (desferrioxamine) to reduce the level of ferritin, is also one of the center's protocols, and is applied for patients over 2.5 years of age, those who had ferritin level higher than 1000 gram/dl, or those also have been transfused at least 10 times. Desferal is given to patients with the aid of a pump. The patient's hormone levels is checked at the time of diagnosis, and repeated after the age of 10, and the level of iron is checked every 3-6 months. Also, patients' are regularly screened for infection with hepatitis virus, and if necessary, treatment will begin. Checking the heart iron volume is also carried out regularly with magnetic resonance arteriography in second phase (MRAT2), and all patients' benefit from complementary treatments such as vitamin E, and C. Finally, the routine protocol of thalassemia treatment is also applied for all patients.

The patients will be followed up by the center staff for transfusion, treatment, and other necessary services throughout their life.

The survived patients who have ages of more than 25 years, and also the presence of a 43-year-old male patient show that the patients' survival is more than a decade, which is in contrast with the letter of Dr. Al-Mendalawi. This high survival, and good quality of life is due to early diagnosis, and a suitable treatment protocol for the patients in Zahedan Thalassemia Center.

The growth indexes of the patients have not been investigated in this study,[1] and it will be investigated in later research.

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