Correspondence

Fetal outcomes in pregnant women with sickle cell disease

To the Editor

I read with interest the study by Al-Farsi et al on the “Fetal outcomes in pregnant women with sickle cell disease”. It is obvious that anemia during pregnancy has generally various adverse maternal and fetal outcomes. The published data of the community-based survey on the genetic blood disorders in Oman has revealed the prevailing of various hemoglobinopathies. The prevalence of sickle cell trait was 6%, and 2% for beta-thalassemia. The prevalence of sickle cell was 0.2%, and homozygous beta-thalassemia was 0.07%. Other abnormal hemoglobins (Hb) were detected in that survey; including HbD (0.6%), HbE (0.3%), and HbC (0.02%). A combination of sickle cell with other abnormal Hb was also detected at low prevalence. However, glucose-6-phosphate dehydrogenase (G6PD) deficiency had a high prevalence in Oman being 25% in males, and 10% in females. The co-inheritance of 2 or more abnormal genes in the same individual is not uncommonly seen, especially in certain ‘closed’ tribes in which the consanguineous marriage is culturally preferred, such as in Oman. Such genetic interactions might modify the clinical presentation and hematological profile of the disease state. Al-Farsi et al did well in considering sickle cell beta-thalassemia, sickle cell C disease, and sickle cell D disease as baseline variables to be evaluated in their studied pregnant population. However, they did not consider the co-inheritance of sickle cell and G6PD deficiency. This is important to be noted, as patients with sickle cell and G6PD deficiency co-inheritance were noticed to have significantly lower mean level of hematocrit with higher level of reticulocyte count as compared with their non-G6PD deficient counter parts. Also, such co-inheritance could reduce the frequency of crises, and improves the prognosis. Accordingly, I presume that that limitation might cast some suspicions on the accuracy of the results addressed by Al-Farsi et al.

Mahmood D. Al-Mendalawi
Department of Pediatrics
Al-Kindy College of Medicine
Baghdad University
Baghdad, Iraq

Reply from the Author

We received with interest a correspondence to the editor by Dr. Al-Mendalawi regarding our publication “Fetal outcomes in pregnant women with sickle cell disease”. We thank the authors for their comments. The letter addressed important issues that enriched the discussion.

Dr. Al-Mendalawi emphasized that co-inheritance of G6PD deficiency in patients with sickle cell disease (SCD) may decrease the severity of complications of SCD, and the lack of inclusion of this variable casts some suspicions on the accuracy of the results of our study. We respectfully disagree. The amelioration of SCD by co-inheritance of G6PD deficiency is controversial at best, and the impact of fetal outcomes has not been examined before. These 2 points are addressed further below.

The G6PD deficiency is a well-known X-linked recessive enzymopathy that is relatively common in our region with percentages reaching 25% in males, and almost 10% in females. It is usually associated with episodic hemolytic events usually during exposure to oxidative agents and infections. There has been some studies that examined the correlation between G6PD status and SCD, to assess whether it has attenuating or exacerbating effect in the SCD course. The cooperative study of sickle cell disease (CSSCD) has shown no impact of G6PD status on acute anemic events, infections, pain episodes, or hemolytic rates as measured by hemoglobin concentration, reticulocytes count, bilirubin, and SGOT level. Two other large studies by Ohene et al and Flanagan et al have shown no correlation between cerebrovascular disease and G6PD deficiency.

Furthermore, the study cited by Dr. Al-Mendalawi has some major limitations that include small sample size, imbalance of the 2 groups compared, and unclear statistical analysis. We therefore still believe that the conclusions from our study are still accurate and valid.

Mohammed N. Al-Hunieni
Murtadha K. Al-Khabori
Department of Hematology
Sharifa H. Al-Farsi
Nihal M. Al-Riyami
Department of Obstetrics and Gynecology
Sultan Qaboos University Hospital
Muscat, Sultanate of Oman
References


Supplements

* Supplements will be considered for work including proceedings of conferences or subject matter covering an important topic

* Material can be in the form of original work or abstracts.

* Material in supplements will be for the purpose of teaching rather than research.

* The Guest Editor will ensure that the financial cost of production of the supplement is covered.

* Supplements will be distributed with the regular issue of the journal but further copies can be ordered upon request.

* Material will be made available on Saudi Medical Journal website