Letter to editor

Major thalassemia and pregnancy

β-thalassemia (Cooley anemia) initially was described by Dr Cooley.[1] It is recognized that various types of thalassemia are inherited anemia caused by mutations at the globin gene, affecting the production of α- or β-globin protein. The anemia interferes with red cell maturation.[2] β-globin gene mutations give rise to β-thalassemia.[3] Patients have abnormal growth, altered pubertal development, and immune function and deficits in bone mineral acquisition.[4] Thalassemia major (TM) refers to a disease requiring more than eight red blood cell transfusions per year. The combination of early diagnosis, improvements in monitoring for organ complications, and advances in supportive care, however, have enabled many patients who have severe thalassemia syndromes to live productive and active lives well into adulthood.[3]

A 26-year-old Iranian primigravida with severe form of β°/β^{+} thalassemia major experienced reduced fetal movement at 35th week of gestation. The patient had received lifelong blood transfusions and been regularly treated with chelation therapy since early childhood that was continued in pregnancy. Her developmental milestones had been normal, with spontaneous menarche at the age of 12 years followed by regular menstrual periods. Pregnancy was spontaneously conceived. Her partner had a normal hemoglobin pattern and the fetus was not judged to be at risk for a major hemoglobinopathy. There was moderate evidence of thalassemic facies and moderate hepatosplenomegaly. Folic acid supplementation (5 mg daily) was taken throughout pregnancy. Her pregnancy was uncomplicated with no evidence of intrauterine growth retardation. Two units of packed cells were transfused to the patient every 10 days during pregnancy. Maternal ferritin levels in pregnancy ranged from 1800 to 1500 μg/L (normal: 10 to 120) and her serum iron ranged from 35 to 37 μmol/L (normal: 14–30). She had a non-reactive non-stress test, and oxytocin challenge test was positive. Pregnancy was terminated by cesarean section with spinal anesthesia. A live infant born with birth weight 2300 gram, Apgar scores of 8 at 1 minute and 10 at 5 minute, and the neck was encircled by three coils of the umbilical cord. They were discharged after 2 days.

Successful pregnancy and delivery of healthy babies is possible without hormonal assistance among women who have major thalassemia.[5,6] They need psychological supporting prior and during pregnancy.[7] The endocrine organs are exquisitely sensitive to the toxic effects of iron and this may result in hypogonadotropic hypogonadism, pituitary damage, diabetes, osteopenia, and osteoporosis.[8,9] Hypogonadotropic hypogonadism is common in young thalassemic adults that is believed to contribute to low fertility.[10] The effect of chelation on preservation of gonadal function currently is being investigated.[3]

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