

THALASSEMIC MOTHERS AND THEIR BABIES

Somchai Insiripong¹, Sunya Prabripitaloong² and Nittaya Wisanuyothin³

¹Department of Medicine, ²Department of Obstetrics and Gynecology, ³Department of Pediatrics, Maharat Nakhon Ratchasima Hospital, Nakhon Ratchasima, Thailand

Abstract. Thirty-two mothers with thalassemia, more than half of whom had hemoglobin H disease with or without other forms of thalassemia or hemoglobinopathies along with their 46 children were evaluated. The mean maternal hematocrit was 22.2% and the mean maternal body mass index was 18.7. The majority of women were not transfused during pregnancy. Eight point seven percent developed toxemia of pregnancy. All had spontaneous labor, 69.5% had a normal vaginal delivery and 23.9% had a cesarean section. The mean infant birth weight was 2,542.2 grams. Low birth weight was found in 34.7%; no extremely low birth weights occurred.

INTRODUCTION

Thalassemia is a chronic genetic disease. Its main clinical manifestation is anemia which can vary from asymptomatic to severe with death occurring during the first few hours of life depending on the type and combination of abnormality. Those with thalassemia major, besides having anemia, have other manifestations of disease, including chronic hypoxia, huge hepatosplenomegaly, bone changes due to hyperplasia of intramedullary hematopoietic tissue, hemochromatosis of the heart, short stature, delayed puberty and a short life span (Benz, 2008). Patients need transfusions. It is not only a health problem for the patient, but an economic burden on the family and the country. When women with thalassemia become pregnant, they may have physical defects, anemia, short height and low body mass index as well as pass along the disease to their children (Deshmukh *et al*, 1998).

Correspondence: Somchai Insiripong, Department of Medicine, Maharat Nakhon Ratchasima Hospital, Nakhon Ratchasima 30000, Thailand. Tel: 081-9556993; Fax 66(0)44 293044 E-mail: chaikorat@gmail.com

Women want to avoid the possibility of having thalassemic babies. However, some women accept the risk and others never knew they had thalassemia.

The aim of this study was to describe certain characteristics regarding thalassemic mothers and their babies.

PATIENTS AND METHODS

Thalassemic mothers who delivered their babies at Maharat Nakhon Ratchasima Hospital between January 2003 and December 2007 were included in the study. They were interviewed using a group of questions consisting of maternal characteristics, such as height and weight before pregnancy, age at first pregnancy. Some data were collected from their medical records, namely, the type of thalassemia, sizes of the liver and spleen, average hematocrit before pregnancy, number antenatal care (ANC) visits, number of transfusions during the pregnancy, mode of delivery and infant characteristics, such as birth weight, sex and the number of other children. Most husbands did not have their blood checked for thalassemia. Hemoglobin typing of the babies was not explored either.

All parameters are expressed as means and percentages.

RESULTS

Thirty-two thalassemic mothers were recruited and interviewed following a questionnaire. Half became pregnant in spite of strong encouragement to use birth control, while the other half were diagnosed with thalassemia at the first ANC visit. The ages of the women at their first pregnancy ranged from 21 to 49 years (mean 31.9 years). The average hematocrit, sizes of liver and/or spleen, height, weight and body mass index before pregnancy are shown in Table 1.

All the women had anemia (mean hematocrit was 22.2%), the majority of them had hepato- and/or splenomegaly, the mean liver and spleen sizes were 1.6 and 3.0 cm, respectively below the costal margin. Five of 32 women (15.6%) had splenectomy during childhood for hypersplenism. No one had regular blood transfusion before or during their pregnancy. A minority of them were transfused because they experienced severe anemic symptoms, such as fainting or severe fatigue.

The types of maternal thalassemia are shown in Table 2. One-third of women (34.3%) had a severe form of beta-thalassemia-hemoglobin E and nearly 2/3 (59.1%) had hemoglobin H or hemoglobin H plus others, such as hemoglobin Constant-Spring, hemoglobin E or beta-thalassemia.

Nine (28.1%) women attended the ANC clinic, though irregularly, and received an occasional blood transfusion during pregnancy (1-4 units of packed red blood cells, per person, a mean of 2 units/person). The rest of the subjects did not attend the ANC clinic and never received a blood transfusion. Four mothers developed full blown toxemia of pregnancy.

There were 46 babies born to these

Table 1
General maternal characteristics.

Maternal characteristics	Range	Mean
Age (years)	21-46	31.5
Hematocrit (%)	13.6-30.3	22.2
Liver size (cm)	0-7	1.65
Spleen size (cm)	2-8	4.0
Height (cm)	146-165	153.1
Weight (kg)	35-55	44.1
BMI (kg/km ²)	15.5-20.8	18.7

Table 2
Types of thalassemia in studied subjects.

Type of thalassemia	No. (%)
Beta-thalassemia-Hb E	11 (34.3)
HbH with Constant-Spring (CS)	7 (21.8)
HbH disease	6 (18.7)
HbAEBart's with CS	3 (9.3)
HbEFBart's	2 (6.2)
HbAEBart's	1 (3.1)
Unknown	2 (6.2)
Total	32

thalassemic women, half the infants were males. Thirty-two (69.5%) delivered their first baby, 13 (28.2%) their second and only 1 (2.1%) their third baby. No hydrops fetalis cases or fetal deaths were recorded. Sixteen babies (34.7%) had a low birth weight (defined as <2,500 grams); of these, one (2.1%) had a very low (1,250 grams) birth weight (defined as <1,500 grams); nobody had an extremely low birth weight (defined as <1,000 grams) (Papageorgiou *et al*, 2005). The birth weights (BW) of the babies born to thalassemic mothers are shown in Table 3.

The majority of women had a blood transfusion during labor. Thirty-two of 46 babies (69.5%) were born by normal vaginal delivery. A minority needed some interven-

Table 3
Birth weights of babies born to thalassemic mothers.

	Range (grams)	Mean(grams)
BW of all babies(N=46)	1,250-3,900	2,542.2
BW of babies of splenectomized mothers (N=5)	1,700-2,900	2,350.0
BW of babies from transfused mothers (N=7)	1,700-3,900	2,592.8
BW of babies from mothers with huge splenomegaly (spleen \geq 8 cm, below the costal margin, N=8)	1,550-3,500	2,400.0

tion: 11 cases had a cesarean section and 3 had a vacuum extraction (Table 4).

The indications for cesarean section were: 3 cases with toxemia, 2 with amnionitis and 1 each for premature rupture of membranes, oligohydramnios, fetal distress and breech position. One woman had repeat cesarean section, the cause for the first cesarean section was not clear.

DISCUSSION

The 32 women delivered 46 infants. The maternal abnormalities documented included anemia, short maternal stature, low body mass index and hepatosplenomegaly, in addition to thalassemia major. Sixteen babies (34.7%) had a low birth weight, which is more common than the general population at our hospital varies between 11.8 and 14.0%. This is presumed to be due to maternal anemia in combination with other maternal factors. In India, low birth weight was found in 30.3% of mothers with anemia, low socioeconomic status, short birth interval, short stature, tobacco exposure, low body mass index (Deshmukh *et al*, 1998) and low non-pregnant weight (<40 kg) (Hirve and Ganatra, 1994). Our findings are consistent with those of a previous study from Ghana (Geelhoed *et al*, 2006) as well as other studies (Bondevik *et al*, 2001; El Guindi *et al*, 2004) which found that maternal anemia and low

Table 4
Modes of delivery in thalassemic mothers.

Mode of delivery	Number (%)
Normal vaginal labor	32 (69.5)
Cesarean section	11 (23.9)
Vacuum extraction	3 (6.5)
Total	46

body mass index contributed to low birth weight infants.

None of the infants had hepatosplenomegaly, pallor or hydrops fetalis in our study.

The mean birth weight of the babies born to mothers who received occasional transfusion during pregnancy was not different from those born to non-transfused mothers. This may be because this women did not receive adequate transfusions to keep maintain adequate oxygenation of the fetus. The hemoglobin level should have been kept above 11 g% (Singla *et al*, 1997). Over half (69.8%) of the studied women had a normal vaginal delivery and 23.2% had a cesarean section, which is a much lower sesarean section rate than in the women without thalassemia who deliver at our hospital (41.6-43.3%). The indications for cesarean section in our study were toxemia of pregnancy, amnionitis, premature rupture of

membranes, oligohydramnios, fetal distress and breech presentation, which are different from those of the normal population which mainly consist of a delayed second stage of labor and previous cesarean section. In a previous study (Jensen *et al*, 1995), the cesarean section in mothers with thalassemia was primarily due to cephalopelvic disproportion (10 out of 16, 62.5%). In our study, in spite of a low body mass index in mothers, transfusions were not adequate, resulting in a decrease in low birth weights, therefore no cases of CPD were diagnosed.

Regarding the women who had a splenectomy, the birth weights of their infants were similar to those without a history of splenectomy, although the sample size was too small to draw any conclusions. It appears a history of splenomegaly had no effect on infant birth weight.

Four of 46 pregnancies (8.7%) were complicated by toxemia of pregnancy which is slightly higher than (3.1-7.1%) the general obstetric population at our hospital. The causes(s) of this are unclear, but the anemia common in this group may have contributed to placental ischemia, one pathophysiological mechanism for toxemia of pregnancy. This association needs to be explored further. Women with more severe thalassemia babies, such as found with alpha thalassemia or Bart's hemoglobin who developed hydrops fetalis during pregnancy had a higher incidence of toxemia. There were no cases of hydrops fetalis in our study in spite of the majority of mothers having hemoglobin H, including some with hemoglobin Constant-Spring.

In our study, the hemoglobin type of the fathers was not determined, therefore the expected frequency of hydrops fetalis or he-

moglobin H could not be calculated. These infants will have hemoglobin typing carried out at 6 months after birth.

REFERENCES

- Benz EJ. Clinical manifestations and diagnosis of thalassemsias 2008. [Cited 2008 Oct 20]. Available from: URL: www.uptodate.com
- Bondevik GT, Lie RT, Ulstein M, Kvåle G. Maternal hematological status and risk of low birth weight and preterm delivery in Nepal. *Acta Obstet Gynecol Scand* 2001; 80: 402-8.
- Deshmukh JS, Motghare DD, Zodpey SP, Wadhwa SK. Low birth weight and associated maternal factors in an urban area. *Indian Pediatr* 1998; 35: 33-6.
- El Guindi W, Pronost J, Carles G, *et al*. Severe maternal anemia and pregnancy outcome. *Gynecol Obstet Biol Reprod (Paris)* 2004; 33 (6 Pt 1): 506-9.
- Geelhoed D, Agadzi F, Visser L, *et al*. Maternal and fetal outcome after severe anemia in pregnancy in rural Ghana. *Acta Obstet Gynecol Scand* 2006; 85: 49-55.
- Hirve SS, Ganatra BR. Determinants of low birth weight: a community based prospective cohort study. *Indian Pediatr* 1994; 31: 1221-5.
- Jensen CE, Tuck SM, Wonke B. Fertility in beta thalassemia major: a report of 16 pregnancies, preconceptual evaluation and a review of the literature. *Br J Obstet Gynaecol* 1995; 102: 625-9.
- Papageorgiou A, Pelousa E, Kovacs L. The extremely low-birth-weight infant. In: MacDonald MG, Mullett MD, Seshia MMK, eds. *Avery's neonatology pathophysiology and management of the newborn*. 6th ed. Philadelphia: Lippincott Williams and Wilkins, 2005: 459-89.
- Singla PN, Tyagi M, Kumar A, Dash D, Shankar R. Fetal growth in maternal anemia. *J Trop Ped* 1997; 43: 89-92 [doi:10.1093/tropej/43.2.89].