



CLINICAL, OBSTETRIC, AND PERINATAL COMPLICATIONS IN PREGNANCIES OF A PATIENT WITH β -THALASSEMIA INTERMEDIA

COMPLICAÇÕES CLÍNICAS, OBSTÉTRICAS E PERINATAIS EM GESTAÇÕES DE UMA PACIENTE COM β -TALASSEMIA INTERMÉDIA

COMPLICACIONES CLÍNICAS, OBSTÉTRICAS Y PERINATALES EN EMBARAZOS DE UNA PACIENTE CON β -TALASEMIA INTERMEDIA

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ABSTRACT

Objective: to describe the clinical, obstetric, and perinatal complications during pregnancies of a patient with β -thalassemia intermedia. **Method:** this is a clinical case report with retrospective follow-up of four pregnancies of a patient with β -thalassemia intermedia. The study was approved by the Research Ethics Committee of Universidade Federal de Mato Grosso do Sul (UFMS), under the Protocol 1,781/2010. **Results:** as clinical complications, the patient showed heart failure due to iron overload, pain crisis, prostration, frequent dyspnea, and repeated urinary infections. As obstetric complications, there was preterm labor, intrauterine growth restriction, and oligohydramnios. Regarding fetuses, all were classified as small for gestational age and having low weight. **Conclusion:** in β -thalassemia intermedia, pregnancy can contribute to the onset of complications, requiring follow-up by a multidisciplinary team during the gestational and perinatal process for reducing maternal and neonatal morbimortality. **Descriptors:** β -Thalassemia Intermedia; Hematological Complications In Pregnancy; Perinatal Care.

RESUMO

Objetivo: descrever as complicações clínicas, obstétricas e perinatais durante gestações de paciente com β -talassemia intermédia. **Método:** trata-se de relato de caso clínico com acompanhamento retrospectivo de quatro gestações de uma paciente com β -talassemia intermédia. O estudo foi aprovado pelo Comitê de Ética em Pesquisa da Universidade Federal de Mato Grosso do Sul (UFMS), sob o Protocolo n. 1.781/2010. **Resultados:** como complicações clínicas, a paciente apresentou insuficiência cardíaca devido à sobrecarga de ferro, crise algica, prostração, dispneia frequente e infecções urinárias de repetição. Como intercorrências obstétricas, houve trabalho de parto prematuro, restrição de crescimento intrauterino e oligo-hidrânio. Em relação aos conceitos, todos foram classificados como pequenos para a idade gestacional e com baixo peso. **Conclusão:** na β -talassemia intermédia, a gestação pode contribuir para o surgimento de complicações, sendo necessário o acompanhamento por uma equipe multidisciplinar durante o processo gestacional e perinatal para redução da morbimortalidade materna e neonatal. **Descritores:** β -Talassemia Intermédia; Complicações Hematológicas na Gravidez; Assistência Perinatal.

RESUMEN

Objetivo: describir las complicaciones clínicas, obstétricas y perinatales durante los embarazos de una paciente con β -talasemia intermedia. **Método:** esto es un reporte de caso clínico con seguimiento retrospectivo de cuatro embarazos de una paciente con β -talasemia intermedia. El estudio fue aprobado por el Comité de Ética en Investigación de la Universidade Federal de Mato Grosso do Sul (UFMS), bajo el Protocolo 1.781/2010. **Resultados:** como complicaciones clínicas, la paciente presentó insuficiencia cardíaca debido a la sobrecarga de hierro, crisis de dolor, postración, disnea frecuente y repetidas infecciones urinarias. Como complicaciones obstétricas, hubo trabajo de parto prematuro, restricción de crecimiento intrauterino y oligohidramnios. Con relación a los fetos, todos fueron clasificados como pequeños para la edad gestacional y con bajo peso. **Conclusión:** en la β -talasemia intermedia, el embarazo puede contribuir a la aparición de complicaciones y se muestra necesario el seguimiento por un equipo multidisciplinario durante el proceso gestacional y perinatal para reducción de la morbimortalidad materna y neonatal. **Descriptor:** β -Talasemia Intermedia; Complicaciones Hematológicas en el Embarazo; Atención Perinatal.

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INTRODUCTION

The hemoglobinopathies are hereditary disorders involving genes responsible for the synthesis of normal hemoglobin. Sickle cell disease, hemoglobin C, and the alpha and beta-thalassemias are common hemoglobinopathies in Brazil.¹ The thalassemias are characterized by a decreased production of one or more polypeptide chains which, often, result in the development of a microcytic and hypochromic anemia, and they are classified as thalassemia major, thalassemia intermedia, and thalassemia minor.¹⁻³

A β -thalassemia intermedia is rarer, with clinical manifestations such as anemia and splenomegaly, less severe and non-dependent on transfusion. This is a different group of thalassemias, in which the red blood cells have a short half-life, with promotion of anemia, but without requiring regular blood transfusion.^{1,4,5}

β -thalassemia intermedia has a broad and variable clinical spectrum. It can characterize both a mild expression, completely asymptomatic throughout life, and the most severe expression, usually from 2 to 6 years of age, with growth and development retardation.^{3,5-7}

It is believed that as the frequency of pregnancies increases in women with β -thalassemia intermedia, complications become more frequent and more complex.¹⁻⁴ In this context, the aim is:

- To describe the clinical, obstetric, and perinatal complications during pregnancies of a patient with β -thalassemia intermedia.

METHOD

Paper prepared through the dissertation << *Intercurrent complications in pregnancies and the perinatal outcomes of women with hemoglobinopathies followed up by a high-risk pregnancy service* >>, presented to Universidade Federal de Mato Grosso do Sul. Campo Grande, Mato Grosso do Sul, Brazil. 2012.

This is a clinical case report, conducted through retrospective follow-up of secondary data from medical records, of four pregnancies of a patient with β -thalassemia intermedia, assisted at the high-risk pregnant women outpatient unit of a university hospital in Campo Grande, Mato Grosso do Sul, Brazil, within the period from November 2010 to August 2011. The case was analyzed when conducting a cross-sectional study for investigation of intercurrent complications in

pregnancies and the perinatal outcomes of women with hemoglobinopathies followed up by a high-risk pregnancy service.

The study was approved by the Research Ethics Committee of Universidade Federal de Mato Grosso do Sul (UFMS), under the Protocol 1,781/2010.

CASE REPORT

T.R.M., 30 years of age, living in Campo Grande. She gestates 4, gives birth to 3, with 1 abortion, having an obstetric history with a report of abortion in the third pregnancy, preterm labor, and two newborn infants small for gestational age. The diagnosis was performed by means of hemoglobin electrophoresis at 15 years of age after a hospitalization for renal crisis and acute anemia, followed by reports of weakness, dizziness, and painful crises.

The patient started an outpatient follow-up by a hematologist. We conducted a family survey and found out that her mother, two aunts, and three cousins, all of Italian descent, were also β -thalassemia carriers.

Pregnancy 1: At 23 years of age. When she noticed symptoms, such as amenorrhea, for about a month, she sought the basic health unit (BHU) close to her home, when pregnancy was confirmed and referred to the high-risk pregnancy service of the university hospital. Pregnancy went on without clinical or obstetric complications, except for an anemia with hemoglobin of 8.8 g/dL and an episode of inhibited preterm labor around 33 gestation weeks. As for perinatal outcome, the newborn infant was full term, male, characterized as small for gestational age, and weighing 2,880 g. She was discharged from hospital without puerperal or perinatal complications.

Pregnancy 2: At 25 years of age. She attended the high-risk pregnant women outpatient unit directly when symptoms were identified, and the pregnancy diagnosis was confirmed. Care for her was provided along with the hematology team, with a total of 12 obstetric consultations. As for clinical complications, a severe anemia had 4 episodes, when a blood transfusion was required. There was a report of extreme fatigue and prostration associated to the severe anemia conditions. The blood tests confirmed the diagnosis, with hypochromia, microcytosis, poikilocytosis, teardrop and target red blood cells. She showed an episode of urinary tract infection (UTI) at the 20th gestation week, with urine culture confirmed for *Escherichia coli*, treated with antibiotic agents. Regarding the obstetric complications,

there were no reported complications. As for perinatal outcome, delivery occurred at term, after 39 gestation weeks, vaginally and with no complications. The female newborn infant was small for gestational age, weighing 2,305 g, 47.5 cm high, with a head circumference (HC) of 31 cm, chest circumference (CC) of 29 cm, and abdominal circumference (AC) of 29 cm. The Apgar score showed normal parameters at the 1st and 5th minutes of life.

Pregnancy 3: At 27 years of age. She sought the high-risk pregnancy outpatient unit with 9 weeks and 5 days of pregnancy, with pain in lower abdomen and vaginal bleeding in small amounts. She was referred to the maternity hospital in order to conduct an ultrasonography (USG) examination, with diagnosed non-embryonic pregnancy, referred to the obstetric center for curettage and transfusion of two units of packed red blood cells.

Gestation 4: At 28 years of age. She sought the high-risk pregnancy outpatient unit and the pregnancy diagnosis was confirmed by means of human chorionic gonadotropin (HCG) β -hormone, with 4 gestation weeks. Regarding clinical complications, she showed heart murmurs and split second heart sounds and the diagnostic hypothesis of intra-atrial communication (IAC) was suggested. She had an episode of pain, persistent headache, dyspnea, and frequent fatigue. She underwent 3 blood transfusions, with 29, 30, and 36 weeks. She had 2 episodes of UTI, one with 4 and another with 36 gestation weeks. As for obstetric complications, she complained of pain in lower abdomen and USG showed changes in resistance of the umbilical artery and thin placenta, indicative of fetal distress. Regarding perinatal complications, newborn infant small for gestational age, weighing 2,870 g.

DISCUSSION

In this case of patient with β -thalassemia intermedia, we observe that the diagnosis is often obtained in adolescence and early adult reproductive phase, because the disease progresses in a oligo-asymptomatic way.⁸ She showed hypochromic anemia, followed by reports of dizziness and muscle weakness throughout childhood and adolescence, when she was treated for iron deficiency anemia and diagnosed only after renal crisis and acute anemia, at 15 years of age. This fact draws attention to the importance of early diagnosis of hemoglobinopathies.

The patient showed an increased morbidity as the number of pregnancies increased, with more frequent clinical complications,

probably related to the physiopathology and the normal hematological changes of pregnancy, as well as to the high frequency of blood transfusions, which culminated in iron overload and cardiovascular exhaustion. Chronic anemia can cause increased miscarriages, preterm labor, and intrauterine growth retardation, besides endocrine complications due to iron overload.^{5,6,8}

The pregnant woman was monitored by the multidisciplinary team throughout pregnancy, something which contributed to the low number of obstetric complications and the perinatal outcomes. In perinatal outcomes, we found out that low birth weight was observed in all pregnancies.

CONCLUSION

In β -thalassemia intermedia, certain stress conditions, such as pregnancy, may contribute to the crisis worsening. There is a need, therefore, for follow-up by a multidisciplinary team during the gestational and perinatal period.

The occurrence of clinical, obstetric, and perinatal complications, in 4 pregnancies of the case under analysis, confirms the hypothesis that as the frequency of pregnancies increases in women with β -thalassemia intermedia, the complications also become more frequent and more complex. However, an adequately monitored prenatal care proved that, despite the increased gestational complications, the morbimortality of the fetus was decreased.

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