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Case Report

Case Report on Thalassemia

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ABSTRACT

Beta thalassemia has a variety of manifestations and complications. Despite of having advanced medical facilities, thalassemia is really a threat and need organized preventive strategies' such as premarital screening and genetic counselling. During pregnancy close observation enhances the women's outcome quality of life. This is a case of 38years old pregnant women presented with shortness of breath and labour pain. She was previously diagnosed with thalassemia major. The case report discusses her presentation, nursing management of thalassemia major.

Key words: thalassemia, nursing management

INTRODUCTION

The thalassemia syndromes are a heterogeneous group of usually inherited chronic disorders that are characterized by an absence or decreased synthesis of one of the normal globin chains of haemoglobin. Thalassemia is an autosomal recessive disorder, although it may also be the result of spontaneous mutation. [1]

It was estimated that Worldwide 56,000 conceptions would have a major thalassemia disorder and among them around 30,000 would have β thalassemia major, the majority of babies being born in middle and low income countries and have severe haemoglobin disorder. [1]

The average prevalence of β thalassemia carriers is 3-4% which translates to 35 to 45 million carriers in our multi-ethnic and culturally and linguistically diverse population of 1.21 billion people which also includes around 8% of tribal groups according to the Census of India 2011. Several ethnic groups have a much higher prevalence. [2]

CASE REPORT

Thalassemia pose a significant health burden, 10,000 to 12,000 thalassemia children born annually in India very few children are optimally manageable. The continuous treatment throughout the life makes them tiring. In developing countries' affordability of health care is challenging to the families with thalassemia major child. The prevalence of beta thalassemia carrier is 3 to 4% which translate 35 to 45 million carriers. Thalassemia is an inherited blood disorder in which the body makes an abnormal form of haemoglobin. Haemoglobin is the protein molecule in red blood cells that carries oxygen. The disorder results in excessive destruction of red blood cells, which leads to anaemia. [3]

A pregnant women 38 years (height 135cm, pre pregnancy weight 35 kg) old primi gravida was diagnosed as thalassemia major, hypothyroidism, type II diabetes mellitus. Her thalassemia was identified at the age of one year. Both the parents were not willing to know about their thalassemia

trait so it was not checked, she is lost child in their family. Her three siblings have normal haemoglobin pattern. The patient had received lifelong blood transfusion at every 30 to 35 days. So her haemoglobin maintained normally. She attained menarche at the age of 13 years. She is having regular menstrual cycle. She was diagnosed as type 2 diabetes mellitus under regular medications from 25 years. She was married for one year. She conceived spontaneously. Her spouse had normal haemoglobin pattern hence the fetus was not subjected to prenatal diagnosis of thalassemia. During pregnancy she received blood transfusion once in 20 days and chelation therapy every 35 days once.

The women had euthyroid before pregnancy. But her TSH during first trimester was above 7.7 and she was on tab. Eltroxin 50µg OD. At the end of the first trimester her TSH levels become normal. Antenatal check-ups were normal till 2nd trimester. She developed bronchitis at 32 weeks of gestation. She started with bronchodilators. In her38 weeks of gestation admitted to labour room with complaints of labour pain, shortness of breath. Her clinical assessment revealed that blood pressure 170/100 mm of Hg, pulse rate 90 beats/minute and fetal assessment demonstrated fetal distress. She delivered under emergency caesarean section. An alive male baby was delivered; birth weight of 2.6kg with APGAR was 8/10 at 1minute, 10/10 at 5 minute. Both mother and baby monitored continuously. At birth infant had hypoglycaemia, hypocalcemia.

Table 1 : Classification of thalassemia

Alpha Thalassemia	Beta thalassemia
• Hydrops foetalis • Haemoglobin H disease • Thalassemia trait	• Beta thalassemia minor • Beta thalassemia intermedia • Beta thalassemia major The mother is a beta thalassemia major. [4]

DISCUSSION

Pregnant women with thalassemia have to avoid routine iron supplements. Counselling is indicated in all people with genetic disorders. Women with thalassemia major or minor have infertility problems, so few pregnancies will occur. 50% of these pregnancies are complicated by stillbirth, IUGR, pre-eclampsia and preterm birth. Medical management consists of ongoing monitoring, aggressive transfusion and iron chelation therapy. Severe thalassemia requires medical treatment. Blood transfusion was the first measure effective for prolonging life. Multiple blood transfusions can result iron overload. The iron overload can be treated by chelation therapy with the medications deferoxamine, deferiprone or deferasirox. Chelation therapy involves administration of chelating agents to remove heavy metals from the body. An oral chelation agent, deferasirox is now available in United States for children older than 2years. Compliance is improved with this orally administered medication. [5]

Nursing Management

For mother.

1. **Nursing diagnosis:** Acute pain in the abdomen related to trauma to the tissue as evidenced by pain scale score 8/10

Expected outcome: mother will verbalizes reduction of pain.

Intervention

Position the mother with adequate devices such as extra pillow, back rest etc.

Provide calm and quite environment.

Provide wrinkle free bed and comfortable position.

Allow the mother to take adequate rest by providing comfortable bed.

Assist the mother while breast feeding

Provide extra comfort devices for small pillow to support the surgical site while moving.

Advise the patient to follow diversion therapy like reading newspaper.

Administer controlled epidural analgesia as per physician order.

Administer analgesic as per physician oreder.

Evaluvation: mother verbalized that her pain was reduced to 4/10.

2. **Nursing diagnosis:** Ineffective airway clearance related to increased

tracheobronchial secretion as evidenced by thick mucus production and cough.

Expected outcome: mother will maintain normal breathing pattern

Intervention

Provide fowler's position to the patient.

Encourage patient to perform deep breathing and coughing exercise.

Encourage patient to drink luke warm water.

Advise patient to do spirometry three times in a day.

Encourage patient to take steam inhalation.

Administer nebulisation three times a day as per physician order.

Evaluvation: mother maintained normal breathing pattern as evidenced by reduced cough and thin mucus expectorant.

3. **Nursing diagnosis:** Imbalanced nutritional status more than body requirement related to less intake of food as evidenced by hyperglycemia.

Expected outcome: mother will maintain normal blood glucose level.

Intervention

Monitor the blood sugar three times in a day.

Advise the patient to eat high carbohydrate, normal calorie food and more amount of water .

Encourage the mother to eat cereals, green leafy vegetables.

Advise the mother to avoid sweet and sugar products.

Advise mother to ambulate and resume to regular activities.

Explain the mother about signs of hypoglycaemia, hyperglycemia.

Inform the mother to follow the medicine as per doctor's order.

Advise the mother to come for regular check up.

Evaluvation: mother maintained normal random blood glucose level of 150mg/dl.

For newborn

4. **Nursing diagnosis:** Imbalanced nutritional status less than body requirement related to inadequate intake of feed as evidenced by hypoglycaemia at birth blood glucose 30mg/dl.

Expected outcome: Newborn will maintain normal blood glucose level.

Intervention

Encourage initiation of feeding as soon as possible after birth.

Encourage second hourly feeding to newborn.

Provide neutral thermal environment

Monitor serum calcium and blood glucose by heal stick as per protocol to ensure adequate feeding.

Monitor strict intake of scheduled feeding and number of urine output.

Administer intravenous glucose through large peripheral vein via syringe pump.

Monitor strict intake output chart.

Evaluvation: Newborn maintained normal blood glucose level.

5. **Nursing diagnosis:** Ineffective thermoregulation related to inadequate subcutaneous tissue and history of maternal hypothyroidism as evidenced by temperature 98F.

Expected Outcome: New born will maintain normal body temperature.

Dry newborn thoroughly, quickly and discard the wet blanket.

Wrap infant in warm blanket and carry to mother. May place infant skin to skin with mother and place blanket over mother and baby.

Teach family about the infant's need for warmth and to keep the infant's head covered.

Avoid placing infant on cool surfaces or using cold instruments in assessment.

Monitor temperature as per protocol.

Place cribs away from windows, avoid drafts or air conditioning blowing on the sides of the crib or on the infant.

Teach family to adjust infant's covering after discharge to the room temperature based on how they are feeling.

Keep the baby under radiant warmer to maintain normal temperature.

Evaluvation: Newborn maintained normal thermoregulation by wrapped on warm blanket.

CONCLUSION

Thalassemia major is also known as Cooley's anaemia. This case report discusses the issues associated pregnant women with thalassemia major and focused on nursing management. Careful monitoring and treatment has improved life expectancy and fertility, enabled women with thalassemia major to accomplish successful pregnancy.

Conflict of interest

There is no conflict of interest.

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