



## CASE REPORT: MANAGEMENT OF THALASSEMIC MOTHER FOR LSCS

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**Abstract:** Thalassemias are genetically determined heterogeneous group of disorders with reduced production of globin. We report a case of LSCS in a second gravida, para one with cephalopelvic disproportion with previous LSCS with anaemia. Screening is recommended as a measure to make an antenatal diagnosis. Anaesthesia management of such a case is also discussed.

**Keywords:** Thalassemia, Anaesthesia, Antenatal screening.

Haemoglobinopathies are of two major types including disorders related to haeme and globin chains and thalassemia being a disorder concerned with  $\alpha$  or  $\beta$  globin biosynthesis.  $\beta$  Thalassemia is an autosomal recessive disease characterized by hypochromic haemolytic anaemia with dependence upon blood transfusions repeatedly to sustain life; overall life expectancy being 25 to 30 years of age.

The disorders of  $\beta$  Thalassemia genes are concentrated in people residing in regions of the world endemic for malaria including Mediterraneans, North Africans, Asian Indians, and South East Asians wherein R.B.C screening reveals reduced MCV, reduced MCH and elevated HbA levels.

In the disorders of  $\alpha$  Thalassemia, HbA synthesis is affected, the clinical presentation being microcytosis, normal serum iron levels, normal HbA levels. Depending upon the extent to which the number of genes on  $\alpha$  chains are deleted, lies the severity of the disease ranging from homozygous Hb Barts,  $\alpha$  Thalassemia major, minor to heterozygous carrier state which is completely asymptomatic. Mating of two carriers may produce a severely affected child.



There is as such no cure for this disease however measures can be taken to prevent conception of an affected foetus. This goal can be achieved only after thorough prenatal screening after a degree of social engineering which might be unacceptable to most people.

**Case Report:** A twenty three year old second gravida, para one with CPD, was admitted to the hospital for safe confinement. She was registered case with moderate to severe anaemia; her Hb level was 6.5 gram% in second trimester and 8 gram% at about the term. She had history of third degree consanguinous marriage. Her obstetric history revealed previous caesarean section done under subarachnoid block three years back, indication was primigravida with contracted pelvis at that time. She was advised complete haemogram, peripheral smear and rest other routine investigations as per the institutional ANC workup guidelines. To find out the cause of her moderate to severe anaemia she was advised to do haemoglobin electrophoresis, sickling, coagulation profile, serum iron levels, Thyroid function tests, platelet count and RDW. She refused to do these tests on account of ill health of her first issue. She also informed to us, incidentally that her first baby, which was a female of three years was required to be given two blood transfusions in her second year of age.

On account of obvious academic interest, the first baby was also examined and her reports verified from our paediatrics department which revealed that Hb of the baby was 3.5 gram%, TLC 15000/ mm<sup>3</sup>, DLC: Polymorphs 54%, Lymphocytes 45%, Reticulocytes 2%, Platelets 2,50,000, Blood indices: MCV 68.9, MCH 21.7, RDW 29%. Hb electrophoresis: HbA 54.3% (96.8 to 98%), HbF 43%(= $\leq$  0.5%), HbA 2.7%(2.2-3.2% in 8 to 12 months and 0 to 3.7% above one year), HbS absent, HbA 54.3 (Above one year 94.3 to 98.5%)

Diagnosis was  $\beta$ -Thalassemia syndrome. The above impression was given if patient had not received blood transfusion for preceding three months. Repeat electrophoresis was advised after three months. Peripheral smear of the baby showed hypochromia +++, microcytes ++, Macro +, Anisopoikilocytosis, polychromatic nRBCs with normal WBCs, platelets and no malarial parasites.

On examination, patient had depressed nasal bridge, malar prominence, pallor ++, no jaundice, liver was 3 cm palpable, soft, firm, non-tender, spleen was 5 cm firm and non tender. Baby was immunized except for hepatitis B, which the parents could not afford. Thus the first issue being a Thalassemia major under follow up of our institution and mother with obvious history of consanguinous marriage with hypochromic anaemia, there was a strong suspicion of her being a Thalassemia trait, expecting a second issue who was also anticipated to be  $\beta$ -Thalassemia.

Counselling was given to her about the expected outcome, the importance of prenatal tests for the diagnosis beforehand and the possibility to detect a Thalassemia foetus in



uterus, to avoid its birth. She was also explained to avoid birth of a child doomed to ill health and early death, with respect to her future pregnancies.

LSCS was done under SAB after giving a preoperative blood transfusion and confirming Hb-9.6 gm%, normal values of bleeding and clotting time and prothrombin time. With use of a fine gauge spinal needle and patient in left lateral position SAB was given atraumatically. Two ml of 0.5% bupivacaine heavy was given after preloading with one pint ringer lactate solution. SpO<sub>2</sub>, pulse, BP were monitored vigilantly after giving inj. Glycopyrrolate 0.2 mg IV. Wedge was inserted for leftward uterine tilt to avoid hypotension. IV fluids were titrated as per BP Inj Pitocin 20 units in drip was started after delivery of the baby. The intra and postoperative course was uneventful.

**Discussion:** All forms of thalassemia are inherited as autosomal recessive trait. The lethal homozygous state results only when all of an individual's genes for  $\alpha$  and  $\beta$  chains are affected. Mating of two heterozygous individuals with same condition results in twenty five percent chance of conceiving an affected foetus and fifty percent chance of the foetus being heterozygous.

Our patient was a suspected carrier but there were no substantial investigations available with us. From history she had undergone previous LSCS under SAB uneventfully three years back. Findings of routine investigations were favourable for SAB namely Hb 9.6 gm% after one blood transfusion, haematocrit 30.8, MCV 59.9, MCH 18.7, MCHC 31.2, platelet count 3.2 lac/mm<sup>3</sup>, Prothrombin time 17/16 sec. We decided to give spinal anaesthesia as we desired complete surety of analgesia and patient was also comfortable as she was aware of the intervention from her previous experience. In this procedure we thought of having less chance of anticipated drug interactions as GPD levels and thyroid function tests were not available. Risk of intubation and aspiration was also avoided by not giving GA. Drug induced decreased cardiac output, leftward shift of oxyhaemoglobin dissociation curve, owing to respiratory alkalosis from iatrogenic hyperventilation of lungs is known to interfere with tissue oxygen delivery. Volatile anaesthetics are less soluble in plasma of anaemic patients, thus an arterial pressure gradient is established. So there is likelihood of more rapid induction and anaesthetic overdose in anaemic patients. Postoperatively measures were taken to minimize shivering and the resulting increased total body oxygen requirement.

According to Gregory S Voyagis and KP Kyriakis, homozygous Thalassemias accompanied with maxillary deformities constitute an aggravating factor for difficult intubation. They have proved this to be of statistically equal strength when compared to traditionally recognized risk factors. Haemoglobin level was found to be inversely correlated with maxillary size and severity of difficult intubation was judged by view on laryngoscopy (arytenoid &/or glottis seen=easy, epiglottis seen / not seen =difficult).



Edward T. Riley, Sheila E. Cohen compared epidural versus spinal anaesthesia for non – emergent LSCS. According to them, to administer epidural block, the anaesthesiologist must progress more slowly with epidural needle to avoid dural puncture, the catheter must be threaded, taped, test dose given and patient observed for 3-5 minutes to exclude IV or intrathecal placement and the entire local anaesthetic dose needs to be delivered incrementally. Onset is slower than spinal. Despite the fact that spinal anaesthesia may result in a denser motor block, it did not increase time spent in post anaesthesia care unit.

Epidural anaesthesia however remains the most preferable technique of choice in medical conditions like PIH, or cardiac disease where a slower onset of sympathetic block is desirable or in cases in which prolonged anaesthesia may be required. In a survey performed at 1993 meeting of the society for Obstetric anaesthesia & perinatology(SOAP), 52% of conference attendees reported using spinal anaesthesia as a technique of choice for elective caesarean delivery. Data was summarized by Michael H Plumer in 1993 SOAP programme abstracts and membership directory.

The role of managed care in health services is increasing. Detailed investigations always help us to deliver effective and efficient medical care. Proper resource utilization can enable us to overcome the documented associated complications.

Sylvie Langlois, Jason C Ford et al have suggested carrier screening program for Thalassaemia and Haemoglobinopathies. They recommend early screening of every pregnant mother along with her partner. CBC and Hb electrophoresis to quantify HbA2 and HbF levels is done. The results are interpreted as follows-

- MCV<80fL (microcytosis) &/or (hypochromia), MCH<27pg with Normal Hb electrophoresis—then blood smear testing is done to identify H-bodies. Serum iron levels are done to exclude iron deficiency anemia.
- If microcytosis + hypochromia is associated with elevated HbA2 levels or any other variant then partner screening is recommended in a similar way.
- if both are detected carriers then preconception genetic counselling is of importance, otherwise, Molecular studies to clarify risk in the foetus are done.
- Prenatal diagnosis is done by cells of chorionic villi, obtained by amniocentesis in those who consent for invasive testing. Confirmatory studies by DNA analysis of amniocytes should be done if MTP is considered.
- If prenatal testing is declined, then diagnosis of child as early as possible and referral to a Paediatric Haematology clinic for treatment is indicated.



**Conclusion:** Thalassemia leads to life-long medical, obstetric, anaesthetic and paediatric problems. These problems may be minor or life-threatening. A close cooperation and communication with the expectant mother and her family is helpful at each step.

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