

# Case Report on Thalassemia in Children

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## Abstract

**Introduction:** Thalassemia which is also called as Cooley's anemia or Mediterranean anemia is a group of hereditary hemolytic anemia characterized by reduction in the synthesis of hemoglobin. The word 'thalassemia' is derived from the Greek word 'Thalassa' which means the great sea.

**Clinical Findings:** Abdominal pain, fever (Temperature - 101°F), Anemia (Hb – 6.4gm%).

**Diagnostic Evaluation:** Blood test: Hb – 6.4gm%, Total RBC count – 2.3 millions/cu mm, RDW – 18.2%, HCT – 20.2%, Total WBC count – 3200/cu mm, Monocytes – 02%, Granulocytes – 20%, Lymphocytes – 77%, AST(SGOT) – 112 U/L.

**Peripheral Smear:** RBC – mild hypochromic with mild cytosis which show few microcytic and mildly hypochromic. Platelets – Reduced on smear, APC – 62,000 cells.

**Ultrasonography:** Splenomegaly.

**Therapeutic Intervention:** Blood transfusion, Inj. Cefotaxime 750mg IV x BD, Syr. Azee 4ml x OD, Tab. Folic Acid 5mg x OD, Tab. Udiliv 150mg x BD, Cap. Hydra 500mg x OD, Tab. Prednisolone 10mg x BD.

**Outcome:** After treatment, the child show improvement. His fever and abdominal pain were relieved and his Hb% increased from 6.4gm% to 11gm% after blood transfusion.

**Conclusion:** My patient was admitted to Pediatric Ward No – 14, AVBRH with a known case of Thalassemia and he had complaint of fever and abdominal pain. After getting appropriate treatment his condition was improved.

**Keywords:** *Thalassemia, Cooley's anemia, Mediterranean anemia, hereditary, hepatosplenomegaly.*

## Introduction

Thalassemia produces hypochromic microcytic anemia due to defective hemoglobinization of RBCs, hemolysis and ineffective erythropoiesis. It can be considered as hemolytic and hypo - proliferative anemia related to abnormal hemoglobin.

It was first noticed in patients, originating from the littoral countries of the Mediterranean Sea. At present the disease has been found in several countries all over the world. The prevalence of the disease in India is high among Gujaratis, Sindhis and Punjabis. Millions of

people are found to be carriers of thalassemia gene and every year thousands of thalassemic children are born in our country.<sup>1</sup>

Progressive pallor, anemia, fatigue or weakness, jaundice, hepatosplenomegaly, recurrent respiratory infection, failure to thrive, facial bone deformities are common symptoms of thalassemia.<sup>2</sup> Blood examination, Bone marrow study, Osmotic fragility test, radiological screening are common investigations done in thalassemia.<sup>1</sup>

Mild thalassemia might not be needed to be

treated but severe thalassemia required regular blood transfusions along with symptomatic treatment.<sup>2</sup>

Thalassemia is classified into three main types, the classification depends on the number of gene mutation and the part of hemoglobin molecules which are affected either alpha or beta.<sup>3</sup> The different types are: *Thalassemia major* – Here thalassemia genes (beta) are inherited from both the parents and it is associated with homozygous state. Synthesis of beta chain is markedly reduced. *Thalassemia intermedia*: It is a chronic hemolytic anemia caused by alpha or beta chain synthesis. It is also a homozygous form. *Thalassemia minor*: It is a mild form of illness and produced by heterozygosity of either alpha or beta chain.<sup>1</sup>

Management of thalassemia is mostly done by Repeated blood transfusion, iron chelation therapy, splenectomy, folic acid supplementation, bone marrow transplantation, gene therapy and gene mutation, supportive management.<sup>1</sup>

**Patient Identification:** A male child of 8 years from Shirajgam admitted to pediatric ward no 14, AVBRH on 23<sup>rd</sup> January 2020 with a known case of Thalassemia major. He is 25kg and his height is 126cm.

**Present medical history:** A male child of 8 years old was brought to AVBRH on 23<sup>rd</sup> January 2020 by his parents with a complaint of abdominal pain (left hypochondriac region) and fever and he was admitted to Pediatric ward no 14. He is a known case of Thalassemia Major and his Hemoglobin level at the time of admission was 6.4gm%. The child is weak and inactive on admission.

**Past medical history:** My patient was diagnosed to have thalassemia at the age of 8 months when he was admitted to hospital due to fever. Till then, he was admitted to hospital time to time for the purpose of blood transfusion.

**Family history:** There are four members in the family. My patient was diagnosed to have Thalassemia major and his parents were diagnosed to be carrier of thalassemia. Type of marriage of the parents is non – consanguineous marriage. All other members of the family were not having complaints in their health except for my patient who was being admitted in the hospital.

**Past interventions and outcome:** My patient was diagnosed with thalassemia when he was 8 months old,

from that time onwards he was admitted to hospital time to time for treatment of the disease mostly blood transfusion. It was found effective as the patient does not develop complications till then.

**Clinical findings:** Abdominal pain, fever (Temperature - 101°F), Anemia (Hb – 6.4gm%)

**Etiology:** Thalassemia occurs when there is mutation or any abnormality in anyone of the gene which take part in producing hemoglobin. When there is genetic abnormality, it can be pass – on from parents to children. When only one of the parents is a carrier, there is chance of developing a mild form of thalassemia that is thalassemia minor where symptoms might not be seen but the person will be a carrier. When both the parents are carrier, there is a great chance of developing severe form of thalassemia like thalassemia major or thalassemia intermedia.<sup>4</sup>

**Physical examination:** There is not much abnormality found in head to toe examination, the child is lean and thin and having dull look. He is weak and not so cooperative. Though it is found that the child is having splenomegaly from ultrasonography, it is not palpable.

**Diagnostic assessment: Blood test:** Hb – 6.4gm%, Total RBC count – 2.3 millions/cumm, RDW – 18.2%, HCT – 20.2%, Total WBC count – 3200/cumm, Monocytes – 02%, Granulocytes – 20%, Lymphocytes – 77%, AST(SGOT) – 112 U/L. **Peripheral Smear:** RBC – mild hypochromic with mild cystosis which show few microcytic and mildly hypochromic. Platelets – Reduced on smear, APC – 62,000 cells. **Ultrasonography:** Splenomegaly.

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## Discussion

A male child of 8 years old from Shirajgam was admitted to pediatric ward no 14, AVBRH on 23<sup>rd</sup> January 2020 with a complaint of abdominal pain, fever and Hb% less than normal limit. He is a known case of thalassemia major which was diagnosed when he was 8 months old. As soon as he was admitted to hospital investigations were done and appropriate treatment were started. After getting treatment, he shows great

improvement and the treatment was still going on till my last date of care.

A study was done on, “A clinico - epidemiological study of thalassemia cases in India”. It was done to assess the clinical presentation and managements practices in thalassemia. For the study, patients case sheets were collected between 2005 - 2014 which is 10 years. These case sheets were being examined and recorded in a specially made proforma for the study. The result from the study were - a total of 183 cases are recorded and among those, 179 (97.8%) were beta thalassemia major, 3 (1.6%) were beta thalassemia intermedia and 1 (0.6%) was beta thalassemia minor. Most of the case were diagnosed at age of 1 year, and one fourth of the case were diagnosed in first 6 months. Fever present in 34 patients (18.6%), pallor found in 179 patients (97.8%), hepatomegaly seen in 172 patients (94%) and bone deformity found in 13 patients (7.1%). One third of the under five patients were found to be underweight and more than half of the patients were found to be stunted. Mean post - transfusion value of hemoglobin after 1 year of transfusion among cases was  $10 \pm 1.6\text{g\%}$ , 51 patients (27.9%) were given desferrioxamine as iron chelation therapy and the mean age of starting this therapy was  $11.1 \pm 8.2$  years. In 4 cases of beta thalassemia major splenectomy was done at a mean age of  $10.7 \pm 4.8$  years. On treatment of thalassemia with desferrioxamine lenticular opacity was found in greater proportion. The study concluded that among thalassemic patient different kinds of complications were found so it is important to involve different specialization in care of thalassemia patient to control the problems.<sup>5</sup> Baliga et al studied about the Malondialdehyde Levels in Serum and Saliva of Children Affected with Sickle Cell Anemia<sup>6</sup>. Jaiswal et al conducted a comparative study on peripapillary retinal nerve fiber layer thickness in patients with iron-deficiency anemia to normal population<sup>7</sup>. Garg et al studied about Dengue serotype-specific seroprevalence among 5- to 10-year-old children in India<sup>8</sup>. Puri et al<sup>9</sup> and Uddin et al<sup>10</sup> analyzed on infants and young child nutrition. Taksande et al reported on 3 cases of children with rare diseases<sup>11</sup>. Tendolkar et al reported about physical abuse of children whereas Khatib et al reviewed the effect of electronic media on children. All these studies have some aspects related to thalassemia, anemia and other nutritional disorders.

### Conclusion

Thalassemia is one of the most common case found

among children, its is very important to diagnose in early stage so that the child will not develop complications from the disease. It is also very important to take preventive measures like antenatal screening and giving genetic counseling are very important. My patient show great improvement after getting the treatment and the treatment was still going on till my last date of care.

**Ethical Clearance:** Taken from institutional ethics committee.

**Source of Funding:** Self.

**Conflict of Interest:** Nil.

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