

Difference Between Sickle Cell Anemia and Thalassemia

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Key Difference - Sickle Cell Anemia vs Thalassemia

Thalassemia is a heterogeneous group of disorders caused by inherited [mutations](#) that decrease the synthesis of either the α -globin or the β -globin chains. Sickle cell anemia is a severe hereditary form of [anemia](#) in which a mutated form of [hemoglobin](#) distorts the [red blood cells](#) into a crescent shape at low oxygen levels. The key difference between sickle cell anemia and thalassemia is that **in thalassemia both α and β globin chains can be affected but in sickle cell anemia only the β globin chains are affected.**

What is Thalassemia?

Thalassemia is “a heterogeneous group of disorders caused by inherited mutations that decrease the synthesis of either alpha or beta globin chains that compose the adult hemoglobin HbA leading to anemia, tissue hypoxia and red cell hemolysis related to the imbalance in globin chain synthesis”.

Thalassemia can be categorized broadly into two main categories known as alpha thalassemia and beta thalassemia.

Alpha Thalassemia

In alpha thalassemia, some of the genes responsible for the coding of the alpha globin chains are deleted. Usually, the alpha globin gene has four copies. The severity of the disease depends on the number of such copies that are missing.

Hydrops Fetalis

When all four copies of the alpha globin gene are missing, synthesis of alpha globin chains is completely suppressed. Since alpha globin chains are required for the synthesis of both fetal and adult hemoglobin, this condition is not compatible with life and therefore it leads to death in utero.

HbH Disease

The absence of three copies of the alpha globin gene results in moderate to severe hypochromic microcytic anemia with associated splenomegaly.

Alpha Thalassemia Traits

These are due to the absence or the inactivity of one or two copies of the alpha globin gene. Although the alpha thalassemia traits do not cause anemia, they can decrease the mean corpuscular volume and mean corpuscular hemoglobin levels while increasing the red blood cell count over $5.5 \times 10^{12}/L$.

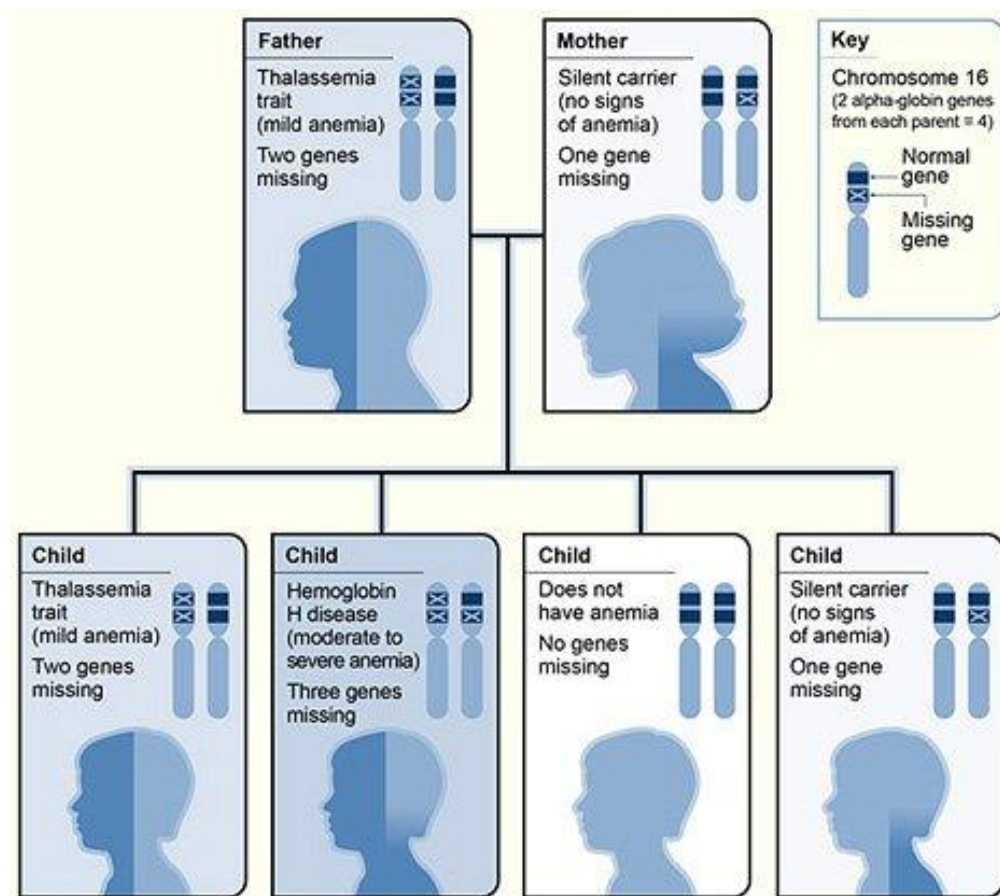


Figure 01: Inheritance of Alpha Thalassemia

Beta Thalassemia Syndromes

Beta Thalassemia Major

If both the parents are carriers of the beta thalassemia trait, the chance of an offspring getting beta thalassemia major is 25%. In this condition, the production

of beta globin chains is either completely suppressed or drastically reduced. Since there aren't sufficient beta globin chains for them to combine with, the excess alpha globin chains get deposited in both mature and immature red cells. This paves the way for premature hemolysis of red cells and ineffective erythropoiesis.

Clinical Features

1. Severe anemia which becomes apparent at 3- 6 months after birth.
2. Splenomegaly and hepatomegaly
3. Thalassemic facies

Laboratory Diagnosis

The major technique employed in the diagnosis of hematological diseases nowadays is the high-performance liquid chromatography (HPLC). In beta thalassemia major HPLC shows the presence of much-reduced levels of HbA with unusually high levels of HbF. A full blood count will show the existence of hypochromic microcytic anemia while the examination of a blood film will reveal the presence of increased amount of reticulocytes along with basophilic stippling and target cells.

Treatment

- Regular blood transfusions
- Folic acid is given if the dietary intake of folic acid is not satisfactory.
- Iron chelation therapy
- Sometimes splenectomy is also carried out in order to reduce the blood requirement.

Beta Thalassemia Trait/Minor

Beta thalassemia minor is a common condition which is symptomless most of the time. Although the features are similar to those of alpha thalassemia, beta thalassemia is more severe than its counterpart. The diagnosis of beta thalassemia minor is made if the HbA₂ level is more than 3.5%.

Thalassemia Intermedia

Cases of thalassemia of moderate severity that do not need regular transfusions are called thalassemia intermedia.

What is Sickle Cell Anemia?

Sickle cell anemia is a severe hereditary form of anemia in which a mutated form of hemoglobin distorts the red blood cells into a crescent shape at low oxygen levels.

The form of hemoglobin generated due to the genetic mutation in homozygous sickle cell anemia is called the HbSS ([sickle hemoglobin](#)). When the oxygen level drops below a certain critical point, sickle hemoglobin forms crystals through a process of polymerization, producing long fibers, each consisting of seven intertwined double strands with cross-linking. It is this polymerization that distorts the red blood cells into a crescent shape. Since the distorted red cells have lost their compliance, they are no longer capable of passing through tiny blood vessels; inside these tiny vessels, sickle cells block the lumen, compromising the blood supply to the organs which result in multiple infarctions.

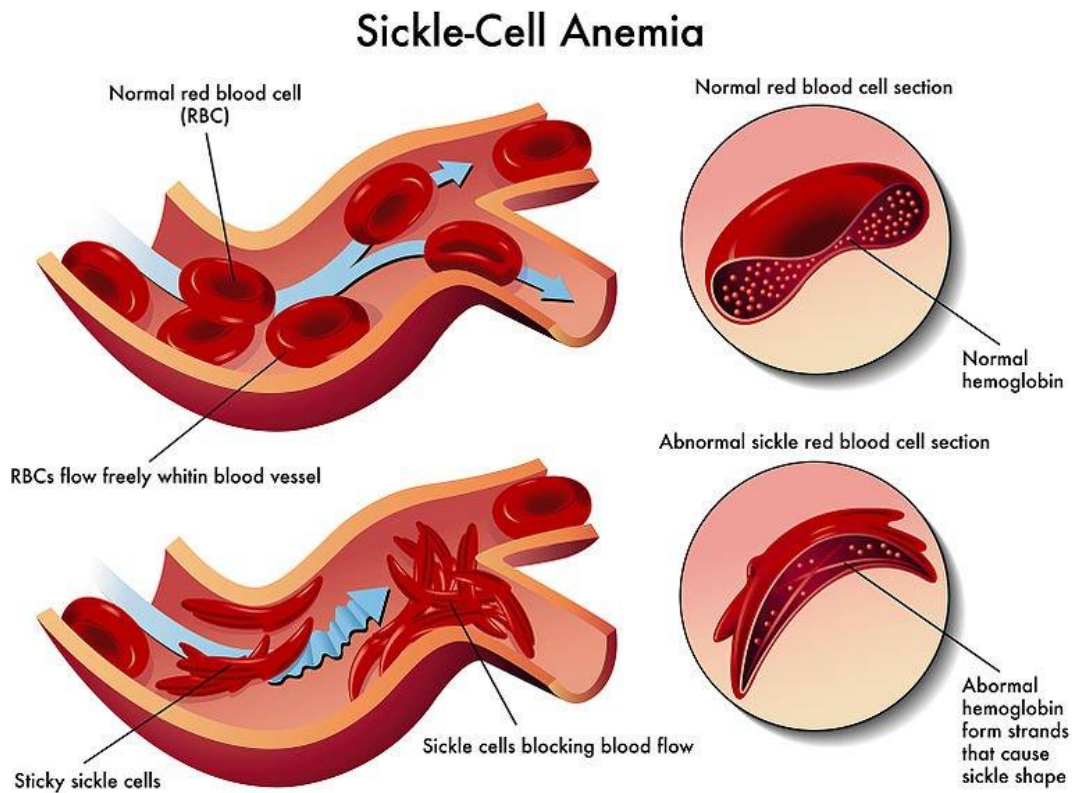


Figure 02: Sickle Cell Anemia

Clinical Features

Severe hemolytic anemia punctuated by crises can be observed.

Vaso Occlusive Crises

Factors such as infection, dehydration, acidosis, and deoxygenation predispose to the vaso occlusive crises. The patient complains of severe pain in the extremities. This is because of the infarctions in the small bones of the limbs.

Visceral Sequestration Crises

These are because of the pooling of blood and sickling, taking place inside the organs. The most fatal complication of this sort is the sickle chest syndrome where the patient presents with dyspnea and chest pain. Pulmonary infiltrates can be seen on chest x-ray.

Aplastic crises

Aplastic crises are characterized by the sudden drop in hemoglobin level most often following a parvo virus infection. Folate deficiency can also be a contributory factor.

Other Clinical Features

- Ulcers in the lower limbs
- Pulmonary hypertension
- Gallstones

Laboratory Diagnosis

- Hemoglobin level is usually 6-9g/dL.
- Presence of sickle cells and target cells in the blood film.
- Screening tests for sickling with chemicals such as dithionate are positive when the blood is deoxygenated.
- In HPLC, HbA is not detected.

Treatment

- Avoiding the factors known to precipitate the crises.
- Folic acid.
- Good nutrition and hygiene.
- Pneumococcal, Haemophilus and meningococcal vaccination.
- Crises should be treated according to the condition, age and drug compliance of the patient.

What are the similarities between Sickle Cell Anemia and Thalassemia?

- Both thalassemia and sickle cell anemia are genetic diseases which affect the structure and function of hemoglobin.

What is the difference between Sickle Cell Anemia and Thalassemia?

Sickle Cell Anemia and Thalassemia	
Thalassemia is a heterogeneous group of disorders caused by inherited mutations that decrease the synthesis of either α or β globin chains.	Sickle cell anemia is a severe hereditary form of anemia in which a mutated form of hemoglobin distorts the red blood cells into a crescent shape at low oxygen levels.
Type of Globin Chain Affected	
Both alpha and beta chains can be affected in thalassemia.	In sickle cell anemia only beta chains are affected.
Nature of Genetic Mutation	
Thalassemia is caused either by a point mutation or by a gene deletion.	The genetic defect causing sickle cell anemia is a gene substitution.
Resistance Against Malaria	
Genetic defect in thalassemia does not provide a resistance against malaria.	The genetic defect causing sickle cell anemia is known to have a protective action against malaria.
Genetic Origin	
Thalassemia is an autosomal codominant disorder.	Sickle cell anemia is an autosomal recessive disorder.

Summary - Sickle Cell Anemia vs Thalassemia

Thalassemia and sickle cell anemia are two serious hematological disorders mostly encountered in the pediatric practice. Therefore, it is important to clearly understand the difference between sickle cell anemia and thalassemia. Increasing the community awareness on these diseases will be helpful in minimizing the incidence of them since the consanguineous marriages play a major role in the transmission of these genetic disorders from generation to generation.

References:

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2. Kumar, Vinay, Stanley Leonard Robbins, Ramzi S. Cotran, Abul K. Abbas, and Nelson Fausto. Robbins and Cotran pathologic basis of disease. Philadelphia, Pa: Elsevier Saunders, 2010. Print.

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