Alpha Thalassemia

KANAKIA Health Care

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What is hemoglobin?

Hemoglobin is an iron-rich protein found on red blood cells that carries oxygen throughout the body. It also carries carbon dioxide from the body to the lungs, where it is exhaled. Normal hemoglobin has four protein chains – two alpha globins and two beta globins.

What is thalassemia?

Thalassemia is a genetic (inherited) disorder of the blood where the production of globin chains is reduced. If the alpha globin chain production is reduced, then the person has alpha thalassemia. If the beta globin chain is reduced, the person has beta thalassemia.

What are the types of alpha thalassemia?

Depending on the clinical features, Alpha Thalassemia can be classified as follows:

1. Silent Carrier of alpha thalassemia - 1 alpha gene abnormal
2. Alpha thalassemia trait - 2 alpha genes abnormal
3. Hemoglobin H disease - 3 alpha genes abnormal
4. Hydrops Fetalis - alpha thalassemia major - all 4 alpha genes abnormal

How common is alpha thalassemia in India?

Alpha thalassemia is common in South East Asia, parts of Africa and the Middle East. It is not uncommon in India with an incidence of 10% - 12%. However, it is a mild form that is seen.

How do I know that my child or I have alpha thalassemia?

The symptoms of alpha thalassemia depend on the number of genes involved. The spectrum ranges from an asymptomatic child to a life threatening disease.

1 alpha gene abnormal	**2 alpha genes abnormal**	**3 alpha genes abnormal**	**4 alpha genes abnormal**
Asymptomatic	Very mild symptoms	Hb H disease	Incompatible with life
Silent carrier	Mild anemia Small RBCs Increased RBC count Physicians may mistake carriers for iron deficiency anemia and may give iron which will not correct anemia	Anemia with enlarged liver and spleen May require frequent blood transfusions. This is followed by iron overload and necessity for chelation.	The child may have hydrops fetalis and may die before or shortly after birth. Rarely in-utero blood transfusions have been tried.

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What are the precautions a patient with alpha thalassemia trait needs to take?

Avoid iron tablets (after checking for iron deficiency)
Folic acid to be taken daily
Thalassemia screening for extended family is a must
Foods rich in iron should be avoided and those with poor iron content should be consumed as per the list given below.

	To Be Avoided	**To Be Eaten**
	High Iron Diet	***Low Iron Diet***
Cereals	Kottu, Bajra, Jowar	Rice, Maida, Bread, Tender maize
Pulses	Bengal gram roasted, Lentils (Soya beans, Rajma)	Red gram (masoor), Peas, Green gram
Vegetables	Leafy vegetables, Beans	Cabbage, Cauliflower, Arbi, Sweet potato, Bitter gourd (karela), Cucumber, Pumpkin, Lady Finger, Brinjal
Fruits	Apricots, Pineapple, Pomegranate, Chikoo	Apple, Guava, Banana, Grapes, Papaya, Orange, Cherries
Meats & Products	Beef, Liver, Egg yellow, mutton	Pomfret, Fish (Rahoo)
Miscellaneous	Jaggery, Almonds, Till, Dates, Raisins	Milk and all milk products
	Foods that Increase Iron Absorption	***Foods that Decrease Iron Absorption***
	Pickles, Vinegar, Fermented food, Soy sauce, Carrots, Vitamin C food like citrus fruits	Cereals like wheat bran, maize, oats, rice, and soy
	(Vitamin C food can be taken 3-4 hours after food intake)	Tea and coffee, especially with milk
		Dairy products like milk, cheese and yogurt particularly with meals Spices like oregano

Iron fortified salt as well as wheat flour or infant feeding formulae are to be avoided.
Do not cook with cast iron cookware (e.g. a wok) because iron from the cookware can transfer onto the food.

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Additional foods recommended	***Rich in Vitamin E***
	Vegetable oils (olive, safflower, palm and soya oil), Ghee, Dairy products, Cereals, Nuts, Eggs
	Rich in Folic Acid
	Lentils, Egg Yolk, Dried Beans, Sweet Potato, Wholegrain Bread, Soya Products, Split Peas, Nuts, Bananas and Peaches
	Rich in Zinc
	Chicken, Fish, Dairy products, Unrefined wheat, maize and rice

Calcium and Vitamin D supplements may be needed.
Note: Silent carriers of alpha thalassemia have no dietary restrictions

SPECIAL PRECAUTION

At the time of marriage, it is imperative to test the partner for alpha thalassemia, to avoid the birth of a child with hemoglobin H disease or hydrops fetalis. This is very rare in India.
(Check for Thalassemia before checking the Kundali)

How does a person get alpha thalassemia?

Since thalassemia is a genetic disease, the patient gets it from the parents and he/she can pass it on to the children. It is an autosomal recessive disease and the following are the possibilities.

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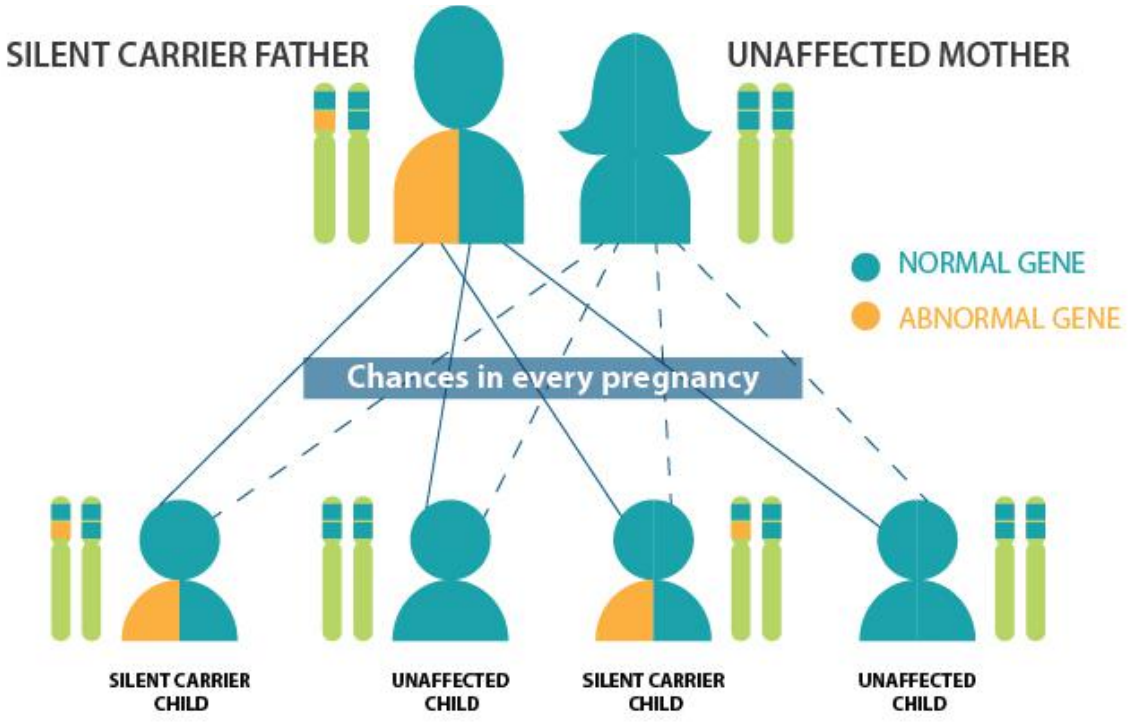

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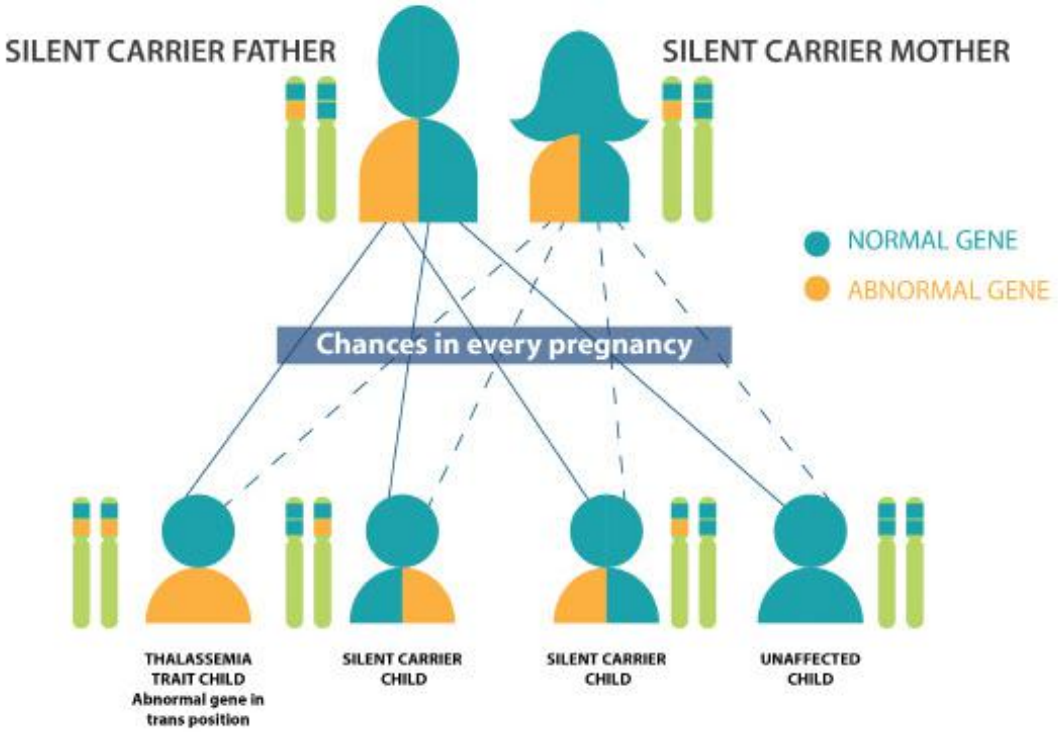

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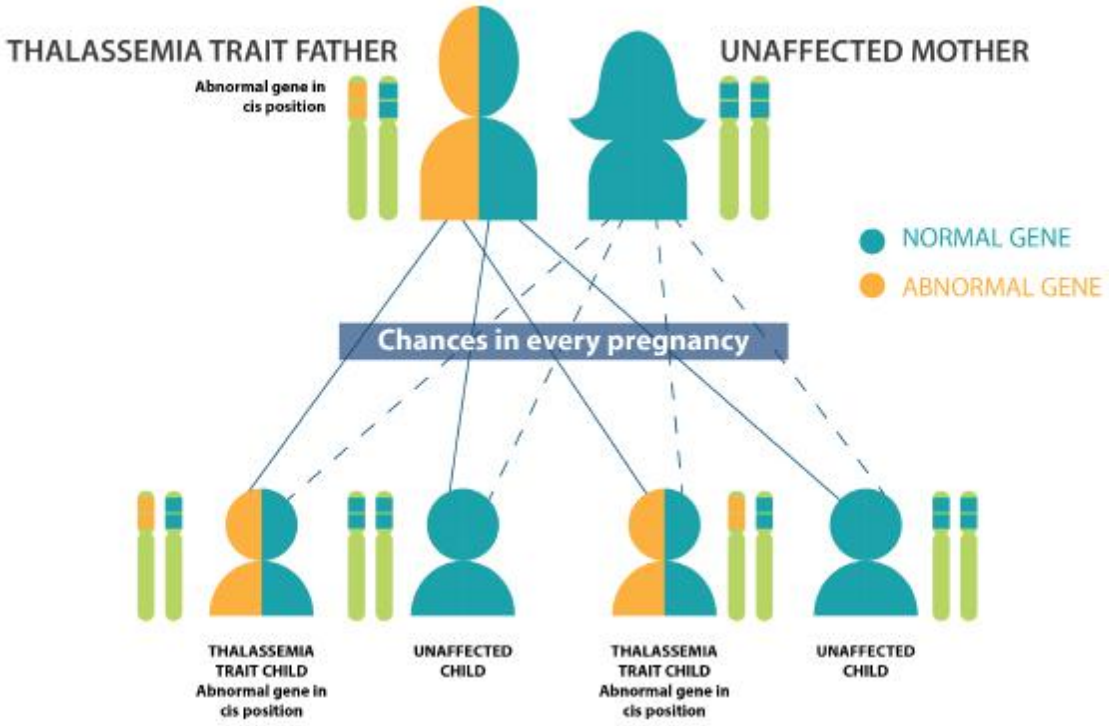

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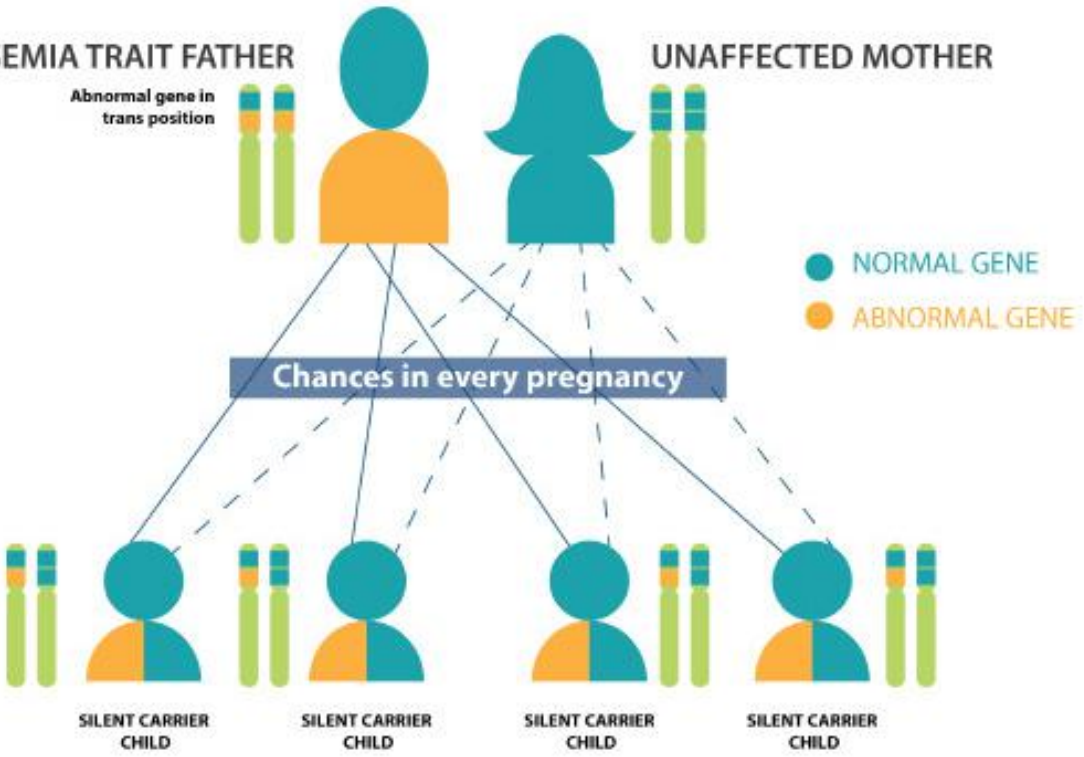

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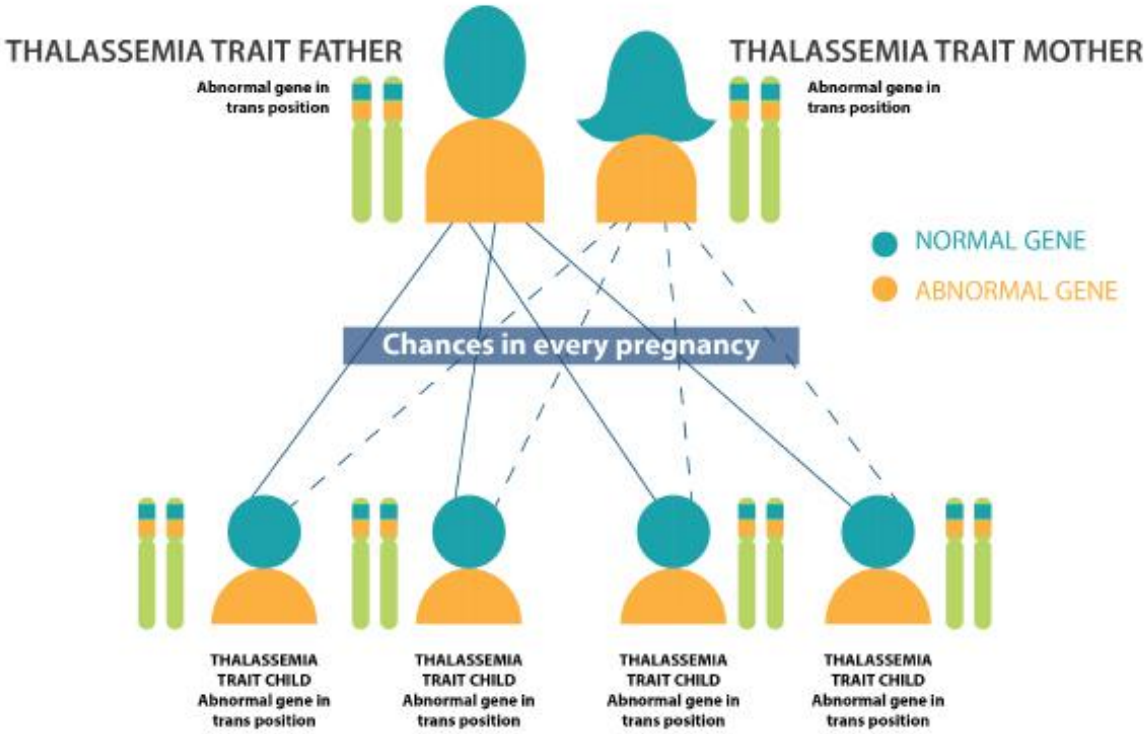

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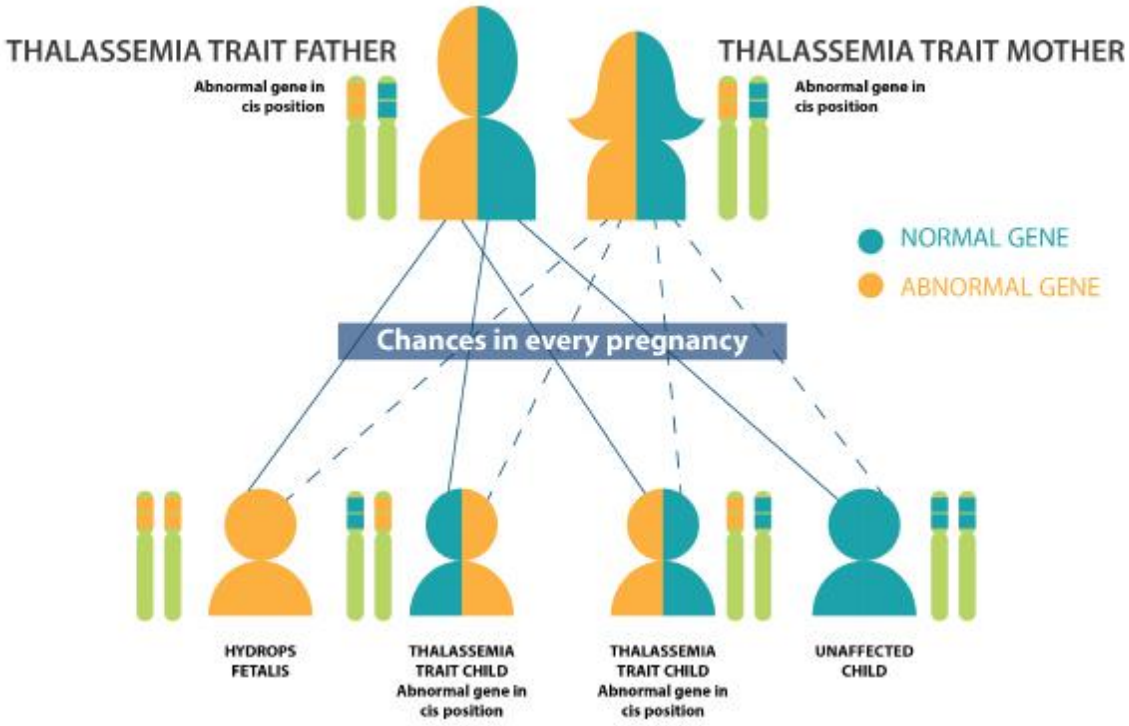

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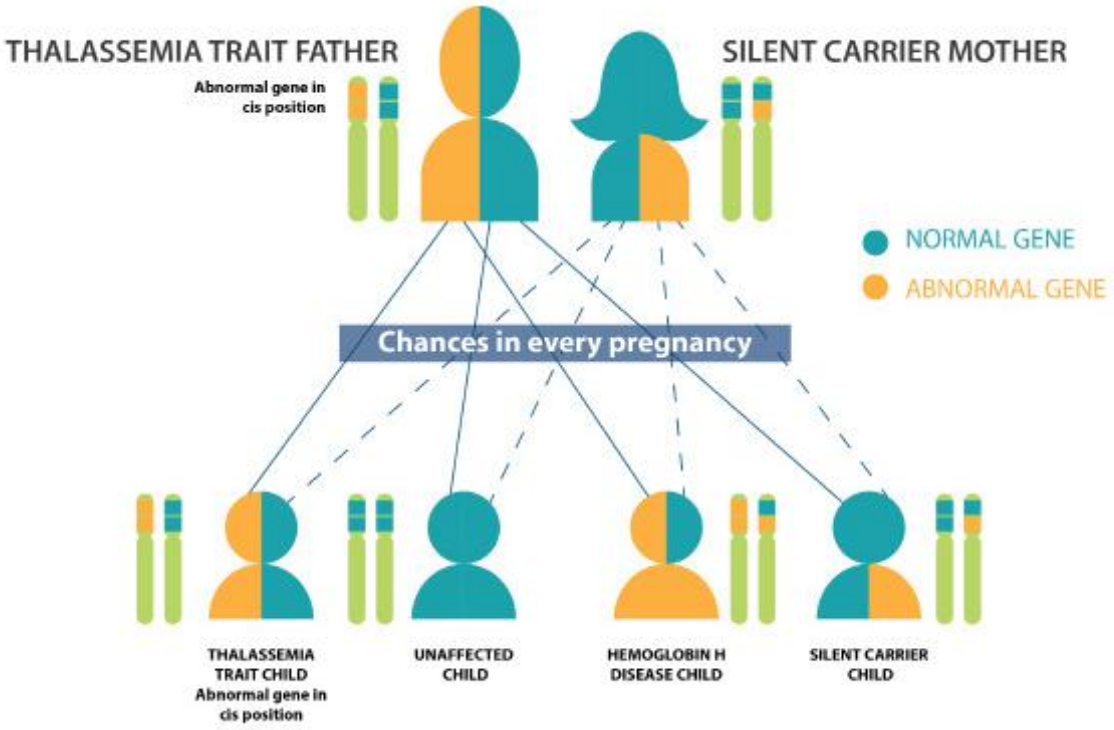

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How is alpha thalassemia diagnosed?

A complete blood count and special blood tests for alpha thalassemia are now available to diagnose the condition. Now molecular and genetic tests are available that can diagnose the condition even before birth.