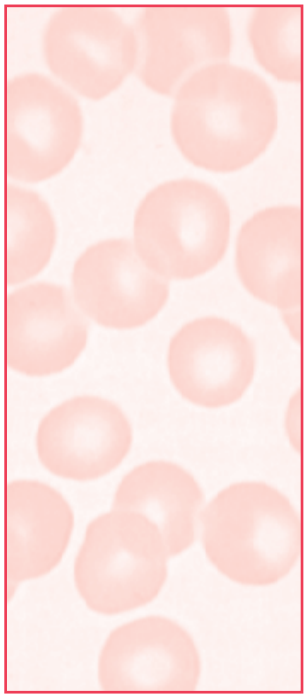
Information for people who carry

Alpha Zero Thalassaemia



People who carry alpha thalassaemia are said to have *alpha thalassaemia trait*. This is often written α thalassaemia trait.

Please keep this booklet handy with your medical card and show it to your doctor or midwife if they discuss thalassaemia with you

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Alpha Zero Thalassaemia

There are several kinds of thalassaemia.

This booklet is for people who have had a blood test that definitely shows they carry ***alpha zero thalassaemia.***

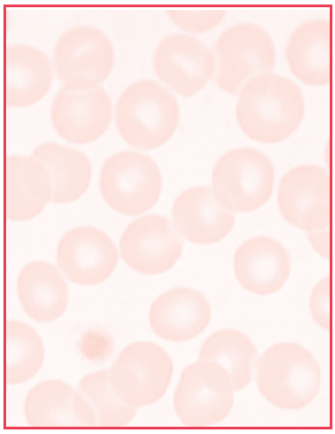
Important points for carriers of alpha zero thalassaemia to remember .

- Carrying alpha zero thalassaemia is not an illness, and does not affect your health.
- Alpha zero thalassaemia can be passed to their children by men ***as well as*** women.
- Alpha (α) zero thalassaemia is ***different*** from beta (β) thalassaemia.
- Although carrying your alpha zero thalassaemia does not affect your health, it could affect the health of some of your pregnancies, but you can avoid this risk with medical advice.
- Do not forget that you carry alpha zero thalassaemia. Keep your blood test results and this booklet, with your medical papers.
- Show this booklet to your doctor when you are thinking of having a family, or when he or she takes blood for any other reason.
- Show this booklet to your GP when you think you may be pregnant.
- Show this booklet to the midwife when you come to hospital for booking.
- If you want more information, ask your doctor to arrange a visit to a special thalassaemia, or genetic counsellor.

Take this booklet with you if you go to see a doctor or genetic counsellor about your alpha thalassaemia.

Blood and Anaemia

To explain about alpha thalassaemia we must talk a little about normal blood and anaemia.



What is blood made of?

Blood is made up of a lot of blood cells in a clear slightly yellow liquid called plasma. Blood is red because the red blood cells contain a substance called haemoglobin, which carries oxygen from your lungs to wherever it is needed in your body. Haemoglobin contains a lot of iron. The main reason why people need iron in their food is to make haemoglobin.

What is anaemia?

Some people have too little haemoglobin in their blood. These people have ***anaemia.*** There are many different kinds of anaemia. The most common kind is ***iron deficiency anaemia.*** This happens when people are not eating enough of the foods that contain iron, or if people lose a lot of blood by bleeding. Some people who carry thalassaemia have a very mild anaemia, but it has nothing to do with the amount of iron you are getting from your food. Thalassaemia is not iron deficiency anaemia, it is an inherited condition. However, people who carry alpha thalassaemia can also get iron deficiency. People who carry alpha zero thalassaemia trait and people with iron deficiency anaemia have smaller red blood cells than usual.

How do you find out if you carry alpha zero thalassaemia?

You have to have a special blood test (a DNA test), which has to be sent to a specialist laboratory. Your results should be written inside the back cover of this booklet.

What is Alpha Zero Thalassaemia?

Alpha zero thalassaemia is a characteristic of the blood. It is inherited, that is, it is passed on from parents to children like hair colour, eye colour or skin colour. It is passed on equally by men and women. It is not catching, and will not turn into an illness.

Alpha zero thalassaemia is common among people originating from South East Asia. It occurs occasionally among people originating from the Mediterranean area or the Middle East, and very rarely in North Europeans. For example, about 1 in 15 people originating from Southern China, 1 in 50 Cypriots, and about 1 in 1,000 North Europeans carry alpha zero thalassaemia.

There are three forms of alpha zero thalassaemia.

1. **Carriers of Alpha zero thalassaemia.** People who carry alpha zero thalassaemia are perfectly healthy in themselves, but if ***both members*** of a couple carry alpha zero thalassaemia, they may pass ***alpha zero thalassaemia major*** on to their children.

2. **Alpha zero thalassaemia major.** This can happen if a baby inherits alpha zero thalassaemia from ***both*** parents. It is a very severe anaemia that affects the unborn baby in the womb. The baby cannot make enough blood, and dies either before birth, or within a few hours of birth. Alpha zero thalassaemia major is also called ***Hb Barts hydrops fetalis.***

3. **Haemoglobin H disease.** This can happen when one parent has alpha zero thalassaemia trait and the other carries a milder form of alpha thalassaemia called alpha plus thalassaemia. People with Haemoglobin H disease are anaemic, but can usually lead a normal life without the need for any treatment. See page 9 for more details.

What Does Carrying Alpha Zero Thalassaemia Mean For Me?

Is an alpha thalassaemia carrier ill?

No. Thalassaemia carriers are not more likely to get other illnesses. Carrying thalassaemia does not make them weak, and they can do any kind of job they want.

Is there any treatment to get rid of alpha thalassaemia?

No, if you are born carrying thalassaemia trait you will always have it.

Can an alpha thalassaemia carrier develop a more severe form of thalassaemia?

No, it cannot.

Is alpha thalassaemia catching?

No, it is not.

Do alpha thalassaemia carriers ever need iron?

Yes, they sometimes do, because they can also get iron deficiency anaemia like other people. Then they may need iron medicine. But it is important that you only have iron medicine if you really need it. The best way to tell whether a thalassaemia carrier needs iron is by a blood test that measures the amount of iron in your blood. (A serum iron or serum ferritin measurement). If you do not have this test, it may appear that you are short of iron simply because you have small red blood cells and you may keep taking extra iron even when you do not need it. This will do you no good, and in the long–run it could be harmful.

What about pregnant women?

Pregnant women who carry alpha thalassaemia trait may need extra iron in the same way as any other pregnant women.

Why is alpha thalassaemia found in certain countries?

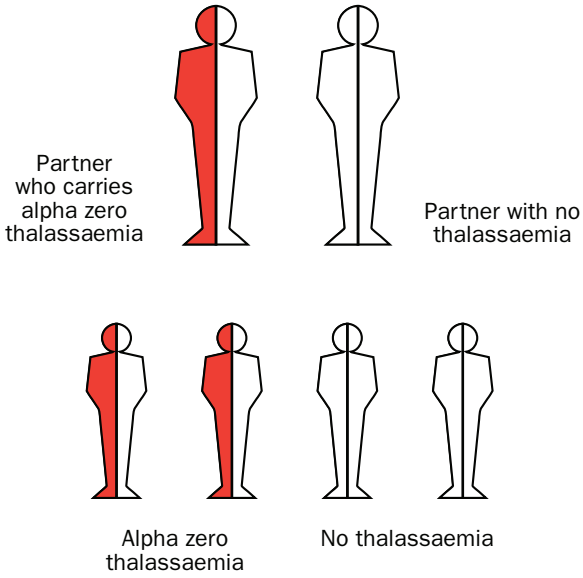
People who carry any kind of thalassaemia are less likely to die if they get malaria. In the past, in countries where malaria was common, thalassaemia survived malaria when other people died, so carrying thalassaemia gave them an important advantage. These people passed thalassaemia on to their children, so as time passed thalassaemia became common in malarious parts of the world. Now we can usually cure or prevent malaria, so carrying thalassaemia is less of an advantage. Because it is inherited, it does not go away from a population when malaria disappears, or from people born in another part of the world.

What Could My Alpha Zero Thalassaemia Mean For My Children?

It is important to know that you carry alpha zero thalassaemia, because sometimes carriers can have children affected by ***alpha zero thalassaemia major.*** This is a serious disease of the unborn baby. If you became pregnant with a baby with alpha zero thalassaemia major, you could get raised blood pressure and have difficulties during pregnancy or at delivery. To explain more about this risk we must see how alpha zero thalassaemia trait is passed on from parents to their children. Let us consider three sorts of couples.

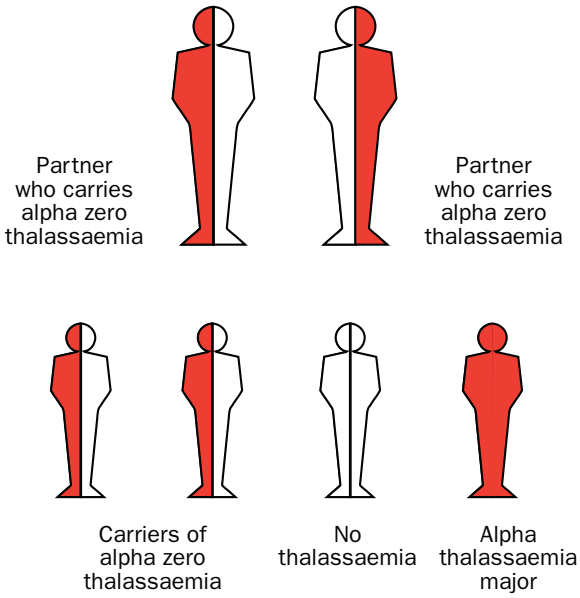
1 A carrier of alpha zero thalassaemia has a partner who ***does not carry any kind of alpha thalassaemia.*** This is the commonest situation. ***There is no problem for an alpha thalassaemia carrier whose partner is not a carrier.***

In each pregnancy there is a 1 in 2 chance that the child will carry alpha zero thalassaemia, and a 1 in 2 chance that it will not. None of the children can have alpha thalassaemia major. The mother will have no particular or additional problems in pregnancy due to the alpha thalassaemia.



2 Occasionally, a carrier of alpha zero thalassaemia chooses a partner ***who also carries alpha zero thalassaemia.***

In each pregnancy there is a 1 in 4 chance of a baby with ***alpha zero thalassaemia major,*** and a three out of four chance that the baby will be healthy.



What is Alpha Zero Thalassaemia Major ?

Other names for alpha zero thalassaemia major are ***alpha thalassaemia hydrops fetalis***, and ***Haemoglobin Barts hydrops fetalis.***

This is a very serious anaemia that develops in the unborn baby. It can only happen when ***both*** parents carry alpha zero thalassaemia.

The unborn baby cannot make enough haemoglobin, and becomes very anaemic. It becomes weak and its heart cannot pump blood around properly. The pregnancy seems to go normally up to about five months, sometimes for longer, but then the baby stops growing normally, and the mother may develop high blood pressure. An ultrasound examination (a scan) may be done. This usually shows that the baby is “oedematous” – which means that it is puffed up with water. Usually the mother starts labour early, between 28 and 36 weeks of pregnancy, and the baby is dead or dying when it is delivered.

There is a one in four chance of the same thing happening in any further pregnancies. This problem can be avoided. This is why it is so important for people who carry alpha zero thalassaemia to find out whether their partner also carries it, before they have a family.

Can alpha zero thalassaemia major be treated ?

Treatment is rarely helpful. Only a few babies with this condition have been known to survive and they have required lifelong transfusions; and some may have lifelong medical problems.

Can alpha zero thalassaemia major be prevented ?

Yes. When both parents carry alpha zero thalassaemia trait, there are several ways to avoid having a stillborn baby.

It is possible to test the unborn baby very early in a pregnancy, to see whether it is healthy, or suffers from alpha zero thalassaemia major. This test is called prenatal diagnosis. It can be done at any time after 10 weeks of pregnancy.

When prenatal diagnosis shows that the developing baby is affected, the parents usually choose to have the pregnancy terminated because the baby has no hope of a normal life. Then can they start another pregnancy, hoping to have a healthy child this time. Remember, there is a three out of four chance of a healthy child in each pregnancy.

There are several other ways to avoid having children with alpha zero thalassaemia major. To find out more, ask your doctor to arrange for you to visit a haemoglobinopathy or genetic counsellor.

3 Sometimes a person who carries ***alpha zero*** thalassaemia chooses a partner who carries another type of alpha thalassaemia called ***alpha plus thalassaemia***.

Alpha plus thalassaemia trait is also called the harmless form of alpha thalassaemia trait because it can only cause problems when it is combined with alpha zero thalassaemia trait. When one of a couple carries alpha zero thalassaemia trait and the other carries alpha plus thalassaemia trait, most of their children will be completely healthy. But there is a one in four chance for each child to inherit alpha zero thalassaemia from one parent and alpha plus thalassaemia from the other. This leads to ***Haemoglobin H disease*** (Hb H disease).

What is Haemoglobin H disease?

People with haemoglobin H disease are anaemic: their haemoglobin level is considerably lower than normal. All the same, most people with Haemoglobin H disease are quite well, can work and have children like other people, and do not need any special treatment.

Most couples who could have children with Haemoglobin H disease are not really worried. They usually ask to have the baby tested as soon as it is born, so that they can know the situation. If the baby does have Hb H disease, the parents are advised to attend a regular clinic a few times a year, just to check that the baby is growing well, and make sure that there are no additional problems.

Is there anything else I should do now?

You inherited your alpha zero thalassaemia from your father or your mother, so your brothers and sisters, and other blood relatives, could also be carriers. For instance, your brothers and sisters have a 1 in 2 chance of being a carrier. Show them this booklet, and advise them to ask for a blood test for alpha thalassaemia before they have children.

IN CONCLUSION remember that important problems can only arise for an alpha zero thalassaemia carrier if they choose another alpha zero thalassaemia carrier as a partner. Even then, problems are uncommon.

Other Haemoglobin Disorders

Beta thalassaemia

This is also common in most of the populations where alpha thalassaemia occurs.

When people talk about thalassaemia they usually mean beta thalassaemia, because it was known about before alpha thalassaemia and because it causes problems more often than alpha thalassaemia. It is described in a separate booklet "All you need to know about being a carrier of beta thalassaemia", which can be obtained from: The UK Thalassaemia Society, 19 The Broadway, Southgate Circus,London N14 6PH (Tel: 020 8882 0011).

Variant Haemoglobins

There are a very large number of abnormal haemoglobins (more than 500), but there are only four common types. These are:

Hb S, Hb C, Hb D, Hb E

What should I do if my partner carries beta thalassaemia or a variant haemoglobin?

Sometimes a person who carries alpha zero thalassaemia has a partner who carries beta thalassaemia or a variant haemoglobin such as Hb S, C, D or E. It is possible for someone who carries one of these to carry alpha thalassaemia as well. So, if your partner carries one of these, he or she should also be tested for alpha thalassaemia. If he or she does **not** carry alpha thalassaemia, there is practically no risk that your children could have a severe anaemia due to the alpha thalassaemia.

Blood Test Results:
Alpha Zero Thalassaemia

The centre that makes the diagnosis should enter the blood test results for the individual and their partner below and sign the sheet.

To whom it may concern:

The carrier of this booklet has been found to carry alpha zero thalassaemia. Test results are given below.

His or her partner should have a blood test for thalassaemia, preferably before a pregnancy is started and the result should also be entered below. If this blood test shows any unusual red cell finding the couple should be referred for expert assessment and genetic counselling.

Results of Blood Tests

Name	Date of test	Hb g/dl	MCV fl	MCH pg	Hb Electro phoresis	Hb A2 %	DNA studies
Patient							
Partner							

Centre arranging the test: .....

Telephone no: ..... Signed: .....

Name in Caps: .....

Further information and/or testing can be arranged through the following centres:

University College Hospital NHS Trust Regional Haemoglobinopathy Genetics Centre (Perinatal Centre) 86-96 Chenies Mews, London WC1E 6HX. Tel: 020 7388 9246, 0845 155 5000 ext. 5230 Fax: 020 7380 9864.

The SE Thames Regional Centre for Blood Disorders, Department of Haematological Medicine, King's College Hospital, Denmark Hill, London SE5 9RS. Tel: 020 7346 3242.

The National Haemoglobinopathy Reference Laboratory, Oxford Haemophilia Centre, Churchill Hospital, Oxford OX3 7LJ. Tel: 01865 225 329.

The UK Thalassaemia Society
19 The Broadway, Southgate Circus,London N14 6PH
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