Thalassaemia syndromes and pregnancy

Information for patients

About this information

This information is for you if you want to know more about your thalassaemia and pregnancy. This factsheet covers pre-pregnancy planning, management during pregnancy and the time after delivery in women who either have thalassaemia major or thalassaemia intermedia. If you have thalassaemia trait (carrier) this information does not apply to you. Please refer to our information leaflet for carriers of thalassaemia.

Thalassaemia syndromes

This factsheet is for patients with either transfusion dependent thalassaemia or non-transfusion dependent thalassaemia.

The syndromes are a complex and broad spectrum of genetic disorders. If you are planning to have a family please discuss this with your haematologist so they can talk to you about how thalassaemia is inherited. They can provide clear advice about the possible risk of thalassaemia affecting a future child, with special reference to your and your partner's blood results.

They will also talk to you about how to prepare for a planned pregnancy with reference to your thalassaemia and complications that you may have. It is very important that your partner is tested for a haemoglobin disorder before stopping any contraception so that any concerns are addressed.

Will my baby be affected?

If you have a thalassaemia syndrome, you should find out whether your partner is also affected before getting pregnant.

- If your partner does not have thalassaemia and is not a carrier for sickle or an abnormal haemoglobin, your baby will not have thalassaemia, but will be a carrier of one of your two abnormal haemoglobin genes. Although your child will be healthy you will need to explain this to them when they are older and want to start a family.

- If your partner has thalassaemia or is a carrier of an abnormal haemoglobin or you are unsure what his haemoglobinopathy status is, we can arrange an appointment at short notice for testing plus specialist counselling.

- This will help you decide whether to have tests when you become pregnant to find out whether your baby has the condition, and will help you explore the option of pre-implantation genetic diagnosis (PGD). PGD involves the removal of one or two cells from an embryo to test for genetic diseases. Only unaffected embryos are transferred into the woman's womb for pregnancy. This can be a difficult decision for many couples and further information is available from the NHS Sickle Cell and Thalassaemia Screening Programme at:

- http://sct.screening.nhs.uk
- http://sct.screening.nhs.uk/professional-leaflets

What should I think about before becoming pregnant?

It is important that you let your thalassaemia team know that you are planning to have a baby. They will be able to help you be in the best possible health before you become pregnant. Until that time, they can advise you on which contraception is best for you. We have a leaflet about contraception that goes into more detail.

You should see your thalassaemia team regularly for review, whether you are planning a pregnancy or not. The checks may include:

- A detailed scan of your heart (echocardiogram)
- Blood pressure, urine and blood tests
- MRI scans for measurement of iron
- Bone density scans
- Diabetes tests (oral glucose tolerance test)

Will I need to change my normal treatment before I become pregnant?

We would advise that you are in the best possible health before getting pregnant. One of the most important things is to make sure that the iron in both your liver and heart is at safe levels before pregnancy and if you have diabetes it should be very well controlled. The aim of all this preparation is to ensure both mother and baby remain well during the pregnancy and delivery.

All women with thalassaemia should start on folic acid three months before pregnancy.

For women who are on regular transfusions and iron chelation

It may take several years to become physically fit enough to have a successful pregnancy. Often iron chelation, hormone therapy and diabetes management all need to be modified to ensure that you are as good as can be for a safe outcome.

It is important to remember that iron chelation is normally stopped during pregnancy and the iron has to be well controlled to avoid dangerous complications developing when you are pregnant.

Poor diabetes control and control of other hormone treatments such as thyroid replacement can result in bad outcomes for the baby as well as the mother and can also affect the success of treatment that may be needed to help you conceive.

Also drugs like bisphosphonates are not safe in pregnancy and need to be stopped several months beforehand.

For these reasons we have the following targets:

- A low liver iron (generally less than 5 mg/g/dw)
- Minimal to no cardiac iron loading (T2* above 20ms, if below 20ms needs cardiac review prior to pregnancy.)
- A fructosamine lower than 330 nmol/l in women with diabetes
- Normal thyroid function tests

Chelation treatment with deferasirox and deferiprone should be stopped for three months before you try to conceive or before the treatment for conception.

You should at this time change over to desferrioxamine infusions as these are known to be safe in the very early stages of a pregnancy. Desferioxamine should stop on the day the fertility hormone injection Pregnyl® is administered if you are having treatment to help you have a baby.

If you are naturally fertile you must stop the desferrioxamine as soon as you miss your period and have a positive pregnancy test.

For women who have non transfusion dependant thalassaemia

The majority of women with this type of thalassaemia are naturally fertile and often aren’t on any treatment for the thalassaemia.

Before planning a pregnancy all your medications should be reviewed and any drugs that may be harmful to the baby stopped.

Hydroxycarbamide is often used to help improve the haemoglobin values in some types of thalassaemia and this should be stopped three months before trying to conceive.

Your thalassaemia team will also review any other medicines you are taking and check your blood for antibodies that may have arisen after blood transfusions. You should make sure all your vaccinations are up to date.

What if I am finding it difficult to get pregnant?

Some people with thalassaemia will find it difficult to get pregnant particularly if they had high iron levels as a child.

Sometimes people find it difficult to get pregnant for reasons unrelated to thalassaemia. If you have any concerns regarding your fertility please discuss it with your thalassaemia specialist. We have a lot of experience in this area and are well connected to the teams that can help you

What are the risks to me and my baby in pregnancy?

Being pregnant is physically demanding and may require an increased frequency of blood transfusions. You will be seen by many healthcare professionals and have more frequent visits and scans during the pregnancy. This is to ensure that you and your baby thrive and problems are identified early and managed properly.

Women with transfusion dependent thalassaemia

Most women with thalassaemia major will have a straightforward pregnancy without serious problems. However if you have diabetes, hypothyroidism or cardiac iron loading you will need even more specialist support and monitoring.

If the level of iron was less optimal at the start of pregnancy (cardiac iron loading with a T2* less than 10ms or severe hepatic iron loading), chelation with desferrioxamine may be restarted. This will take place sometime during the second trimester of pregnancy by your thalassaemia specialist in conjunction with the cardiology team.

They may also recommend an MRI of your heart during the pregnancy. All women with thalassaemia major will start intravenous desferrioxamine when they arrive in hospital for the delivery.

We would expect the delivery to be a natural delivery unless there are maternal or obstetric reasons to recommend a caesarean delivery. There is however a slightly higher incidence of caesarean deliveries in women with thalassaemia.

Women who have non transfusion dependent thalassaemia

Most women have straightforward pregnancies and no serious problems. In some women the haemoglobin can drop down to significantly lower levels than before pregnancy and they may need to go onto regular transfusions for the duration of the pregnancy.

Sometimes the anaemia may not be problematic for the mother but the baby may be growing poorly. The mother should then start regular transfusions to help ensure that the baby gets a good supply of oxygen and nutrients.

Transfusion during pregnancy is not harmful to the mother or the baby and although some iron overload may develop this can be managed after the delivery of the baby.

Additional treatments that may be offered

As you have a medical problem you will have consultant-led care, though often you will be seeing the midwife too. You will be seeing specialist teams throughout the pregnancy. If you have not had the recommended tests in the previous year, they should be carried out. Your vaccinations for hepatitis B, flu and pneumonia should be updated if necessary. These vaccinations are safe in pregnancy.

Other treatments may also be recommended during the pregnancy by obstetricians, haematologists, endocrinologists or cardiologists and these will be discussed with you in detail. These treatments are specific to each person and dependent on their health both before and during the pregnancy.

Pregnant women have a higher risk of developing blood clots in the legs (venous thrombosis) compared with women who are not pregnant. Thalassaemia increases the risk of developing a venous thrombosis as well.

A risk assessment will be done during your pregnancy to help decide if we need to give you any treatment to prevent venous thrombosis.

Women who have had a splenectomy and have a high platelet count may need to use heparin during pregnancy.

We may also recommend the use of heparin and aspirin if the baby is not growing well, in addition to the use of insulin if diabetes develops.

If you have any other concerns, contact your maternity unit and thalassaemia team as soon as possible.

You should be seen at the antenatal clinic at least every four weeks until your 24th week, and then every one to two weeks until you have had your baby. At each visit you will have your blood pressure checked and your urine tested. As well as the routine scans, you should have extra scans to check that your baby is growing normally.

Your blood transfusions will be routinely given though you may find that you have symptoms of higher haemoglobin more than usual and therefore you need more blood or need to be transfused more often than you are used to.

What medicines or treatment should I take in pregnancy?

You will be advised to continue your folic acid (5 mg) once daily and if your spleen has been removed, penicillin V 250mg twice daily.

You should also continue on all your vitamin D and calcium supplementation. If you are on thyroxine or insulins these should be continued but will need frequent monitoring and appropriate dose adjustments to make sure the control is good.

If you are on oral diabetes medication the diabetes doctors or nurse specialist should review this urgently and you may need to go onto insulin. Some oral diabetes drugs cannot be used during pregnancy. All cardiac and antihypertensive drugs need to be reviewed before pregnancy to ensure they are safe for use in pregnancy.

Drugs that are stopped

- If you became pregnant unexpectedly and had not stopped your hydroxyurea, this should be stopped.

- Chelation is stopped in pregnancy, although desferrioxamine may be started during the second trimester if iron levels are not safe. Deferiprone and deferasirox should not be used during pregnancy.
- Bisphosphonates such as pamidronate and alendronic acid should be stopped before pregnancy.

You can take painkillers such as paracetamol and codeine. Like all pregnant women, you should not take painkillers such as ibuprofen before 12 weeks or after 28 weeks of pregnancy without talking to your doctor as this could cause problems for your baby.

What if I become unwell during pregnancy?

If you become unwell, contact the Whittington Health haematology team (see contacts below.) You will have been given other contact numbers at your “booking” appointment (first visit to the antenatal clinic) that you should also call, especially if it is clear that the issue is pregnancy related.

You will be checked for other causes of your symptoms and you may be given antibiotics. You should also be given heparin injections to reduce the risk of blood clots. You will be monitored closely – often in a high-dependency area of the hospital. Your baby’s wellbeing will be checked.

What happens in labour?

When you arrive in the hospital to have your baby, you will be started on a drip which is giving desferrioxamine over 24 hours at a low dose. The reason for this is to control the free iron in the blood during the stress of labour (which puts a lot of stress on the heart). Desferioxamine infusions given to you will not cause any problems for the baby. Your baby’s heartbeat will be closely monitored in labour.

You should be able to have a vaginal birth if there are no complications and you are in good health.

What about pain relief?

You should see an anaesthetist before you go into labour to discuss pain relief; this is often possible in the high risk obstetric clinic. All the usual methods should be suitable for you.

What happens after my baby is born?

You will be encouraged to get up and about to help stop blood clots forming in your legs. You should be offered special stockings and daily heparin injections (to help thin the blood) for at least a week to reduce the risks further. You may be advised to continue heparin for six weeks if you have any other risk factors, for example caesarean section.

Chelation will start soon after delivery if it has not been started already. If you are going to breast feed it will need to be desferrioxamine (desferal).

Breastfeeding is recommended, and you will be given the support you need.

You will be able to go on to the oral chelation drugs once you stop breast feeding.

It is very important after delivery that you take your chelation medication regularly and you may need higher doses to bring the iron burden back into control. You must be seen by the haematology doctors at around six weeks after delivery to check that you are back to normal after the delivery and taking iron chelation treatment at the right doses.

If there is a chance the baby could have sickle cell disease or thalassaemia major the haematology team can organise for the baby to have an in-hospital blood test so you can have the results back the same working day (or day after).

What about contraception?

Progesterone-only pills, injections (Depo-Provera®), implants (Nexplanon®), the Mirena® coil and barrier methods (sheaths and caps) are safe and effective. The combined oestrogen/progesterone oral contraceptive ('the pill') and copper coil can be used too. You can talk to your GP or family planning specialist.

Key points

Thalassaemia is one of the most common inherited single gene disorders in the world. Most women with thalassaemia will have a relatively straightforward pregnancy and a healthy baby.

Chelation is usually stopped in pregnancy; transfusions may need to happen more frequently. If you are planning a pregnancy, let your thalassaemia team know so that they can review your medicines and vaccinations and make sure your checks are up to date.

Further information

NHS Sickle Cell and Thalassaemia Screening Programme:
http://sct.screening.nhs.uk and http://sct.screening.nhs.uk/professional-leaflets

Specialist nurses

Matty Asante-Owusu (community matron, adults)	07920 711210
Edith Aimiuwu (nurse specialist, children)	07799 347161
Emma Prescott (nurse specialist, thalassaemia)	020 7288 5225

Haematology consultants

Dr Bernard Davis
Dr Farrukh Shah
Dr Ali Rismani

Paediatric consultants

Dr Andrew Robins
Dr Sara Hamilton

Obstetric consultants

Miss Amma Kyei-Mensah
Mr Ashokkumar Oliparambil

If you or your family have any other questions please do not hesitate to contact any of the above medical team at The Whittington Hospital

Patient advice and liaison service (PALS)

If you have a question, compliment, comment or concern please contact our PALS team on 020 7288 5551 or whh-tr.whitthealthPALS@nhs.net
If you need a large print, audio or translated copy of this leaflet please contact us on 020 7288 3081. We will try our best to meet your needs.

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