Haemoglobin H Disease

Information for patients, parents, or carers

This leaflet explains Haemoglobin H disease (HbH disease), the symptoms, how it is diagnosed, how to manage the condition and when to seek help.

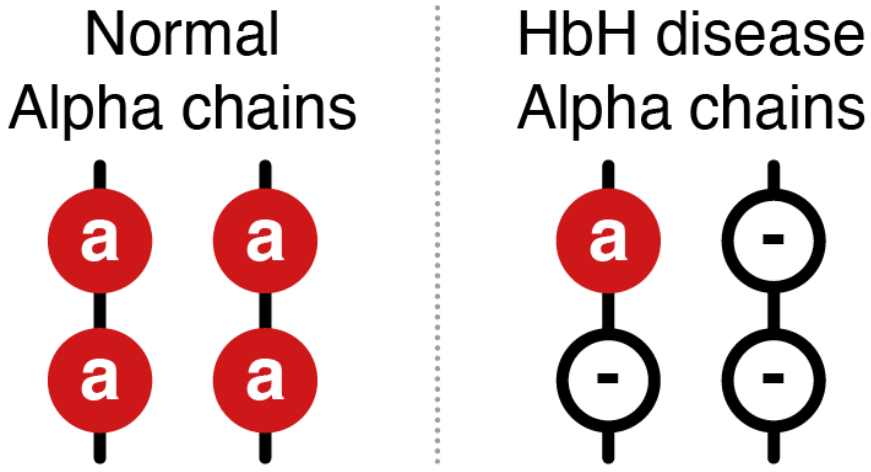
If you have any questions after reading this leaflet, please talk to the specialist team looking after you.

What is a Haemoglobin H disease?

Haemoglobin H disease is a form of alpha thalassaemia. Haemoglobin is the protein in the red blood cells that carries oxygen throughout the body and makes the blood red. Red blood cells in people with HbH disease are smaller and paler than normal because they contain less haemoglobin.

Haemoglobin is made up of different parts. The main parts are called alpha chains and beta chains. Normally there are two alpha chains and two beta chains. The amount and quality of alpha and beta chains you make is controlled by the haemoglobin genes you have inherited from your parents. The most common adult haemoglobin type is called haemoglobin A.

Normally when you inherit haemoglobin A from your parents, you will have:
• four alpha genes (two alpha genes per alpha chain)
• two beta genes (one beta gene per beta chain).



A person with Haemoglobin H (HbH) disease is missing three of the four alpha genes.

Why do I/my child have Haemoglobin H disease?

Haemoglobin H disease is an inherited, genetic condition. A person with haemoglobin H (HbH) disease is missing three of the four alpha genes. Two of the missing genes inherited from one parent, and one gene missing inherited from the other parent.

What are the symptoms of Haemoglobin H disease?

People with HbH disease have smaller, paler red cells than usual, and they are anaemic. Being anaemic means that that the levels of haemoglobin in the blood are low.
Anaemia can cause

- Tiredness and a general lack of energy
- Shortness of breath
- Pounding, fluttering or irregular heartbeats (palpitations)
- Pale skin
- Yellowing of the skin and eyes (jaundice)

Most people with HbH are healthy. They can experience difficulties, such as increased anaemia, when the body is physically stressed, for example in pregnancy, when they are unwell or with intense exercise. Some people may need blood transfusions in these circumstances.

How is Haemoglobin H disease diagnosed?

It is diagnosed by taking a blood test for genetic testing

How can I manage this condition?

It is important to ensure that your diet includes a lot of fresh fruit and vegetables. Sometimes you may be prescribed folic acid, to make sure that the anaemia doesn't get worse. Folic acid is very important vitamin for making red blood cells,

Sometimes when people are anaemic, they are asked to take iron tablets/medicine. You should only take iron medicine if a special blood test (iron profile or ferritin) shows that you are short of iron. Taking iron medicine when you are not short of iron can lead to a build-up of iron in your organs, leading to organ damage.

When should I seek help?

If you become unwell and develop a high temperature this can cause the red cells to break down more quickly. Therefore, any infections need to be treated promptly.

If you become unwell and more anaemic, you should contact your specialist team for assessment as you may require a blood transfusion.

Who can I contact with any queries and concerns?

If you have any questions or concerns, please talk to your Haematology consultant or specialist nurse.

Oxford

Paediatric haemoglobinopathy service	Telephone: 01865 234 212
Dr Amrana Qureshi	**Specialist Nurse** Lesley McCarthy Lesley.mccarthy@ouh.nhs.uk
Adult haemoglobinopathy service	Telephone: 01865 227 907
Dr Wale Atoyebi Dr Kieran Burton Dr Noemi Roy	**Specialist Nurse** Faith Ehigie Faith.Ehigie@ouh.nhs.uk

Southampton

Paediatric haemoglobinopathy service	Telephone: 02381 204567
Dr Nyree Cole Dr Michael Roe	**Specialist Nurses** Ashwini Machado Ashwini.Machado@uhs.nhs.uk Carole Momber Carole.Momber@uhs.nhs.uk
Adult haemoglobinopathy service	Telephone: 02381 204567
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Local haematology service	Telephone:
(Insert details as necessary)	

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Acknowledgements and references:

University College London Hospitals (2019) Patient information: Alpha thalassaemia
Thalassaemia - NHS (www.nhs.uk)