Carrying Hereditary Persistence of Fetal Haemoglobin (HPFH)

A carrier can use this booklet to…

- *help explain carrying HPFH to their partner, blood relatives and others.*
- *show to any health professional (doctor, nurse or midwife) they see about having a family , or pregnancy, or carrying HPFH.*

Contents

Introduction

HPFH is one of a range of variations in the blood called *haemoglobin gene variants*. Here we call them *haemoglobin variants* for short.

Carriers of HPFH are also sometimes said to

HPFH is inherited. That is, it is passed on from parents to their children, like height, hair colour or eye colour. It is passed on equally by men and women. It is present at birth, and it remains the same for life.

It is important to distinguish clearly between people who *carry HPFH* and people who *carry delta-beta thalassaemia.*

Carriers of HPFH…

… inherited HPFH from one of their parents. Carrying HPFH does not affect their own health. It is also extremely unlikely to affect the health of their children, even if their partner is also a carrier. HPFH is therefore said to be a *harmless haemoglobin variant*.

People who carry delta-beta thalassaemia…

… have inherited a haemoglobin variant that could affect the health of their children if their partner is also a carrier.

This document is about carrying HPFH, a harmless haemoglobin variant.

What does it mean to carry HPFH?

Blood is made up of millions of *red blood cells* floating in a fluid called *plasma*. Red blood cells are full of haemoglobin, which is red. This is why blood is red. The heart pumps blood round the body through the blood vessels. The body needs oxygen to function. Haemoglobin picks up oxygen as blood passes through the lungs, and carries it to the rest of the body as the blood circulates.

The usual haemoglobin is adult haemoglobin or *haemoglobin A.* Unborn babies and new-born babies have fetal haemoglobin, or *haemoglobin F* instead of haemoglobin A. Normally, haemoglobin F is gradually replaced by haemoglobin A during in the first six months of life. Hereditary persistence of fetal haemoglobin (HPFH) means that this change-over was not completed, so a carrier of HPFH still has some haemoglobin F in their blood.

HPFH is *inherited* - it is handed on from parents to their children, it is present at birth, and it remains the same for life.

How is HPFH inherited?

It is inherited through *genes.* Every human characteristic, such as eye colour, or height, or type of haemoglobin is controlled by genes that parents pass on to their children. A child inherits two genes for every characteristic, one from each parent. Most people inherit two genes for the usual type of haemoglobin, *haemoglobin A.* Each gene is responsible for making half of the haemoglobin A in each red blood cell.

A carrier of HPFH has inherited a gene for haemoglobin A from one parent and a gene for HPFH from the other, and still has some haemoglobin F in their blood. Their blood functions normally, and carrying HPFH does not cause them any health problems.

How do people find out that they carry HPFH?

By having a special blood test “for haemoglobin disorders”. This usually involves two steps.

- The first step is to measure the size of their red blood cells. This shows nothing unusual.
- The second step is to analyse the types of haemoglobin in their blood. This shows that their blood contains some fetal haemoglobin (haemoglobin F) as well as haemoglobin A.

A special "Kleihauer" test shows that the haemoglobin F is present in equal amounts in all their red blood cells.

Can carrying HPFH affect your health?

Carriers of HPFH are not ill, and are no more likely to get ill than other people. Carrying HPFH does not make them weak, and they can do any kind of work they choose.

It does not cause health problems. However, it can sometimes lead to misinformation, and uneccessary investigations.

Is there any treatment to get rid of HPFH?

No, a person who is born carrying HPFH will always carry it.

Can carrying HPFH turn into a serious haemoglobin disorder?

It cannot.

Can people catch HPFH from a carrier?

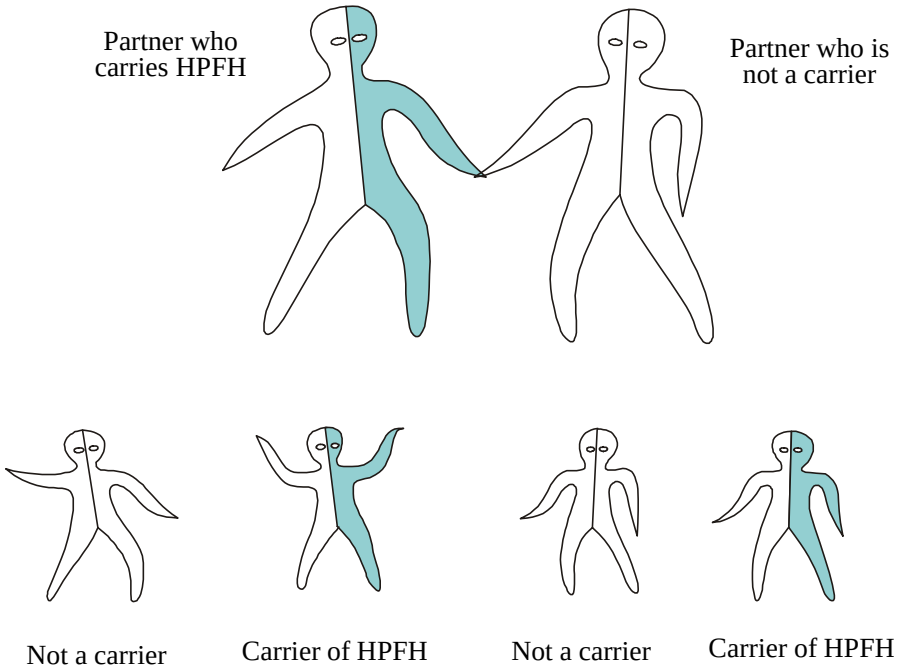
They cannot.

Can a carrier of HPFH be a blood donor?

They can give blood like other people, provided they are not anaemic (do not have a lower haemoglobin level than usual).

Implications for a carrier's children…

If one partner carries HPFH and the other does not carry any haemoglobin variant, their children could not have a serious haemoglobin disorder



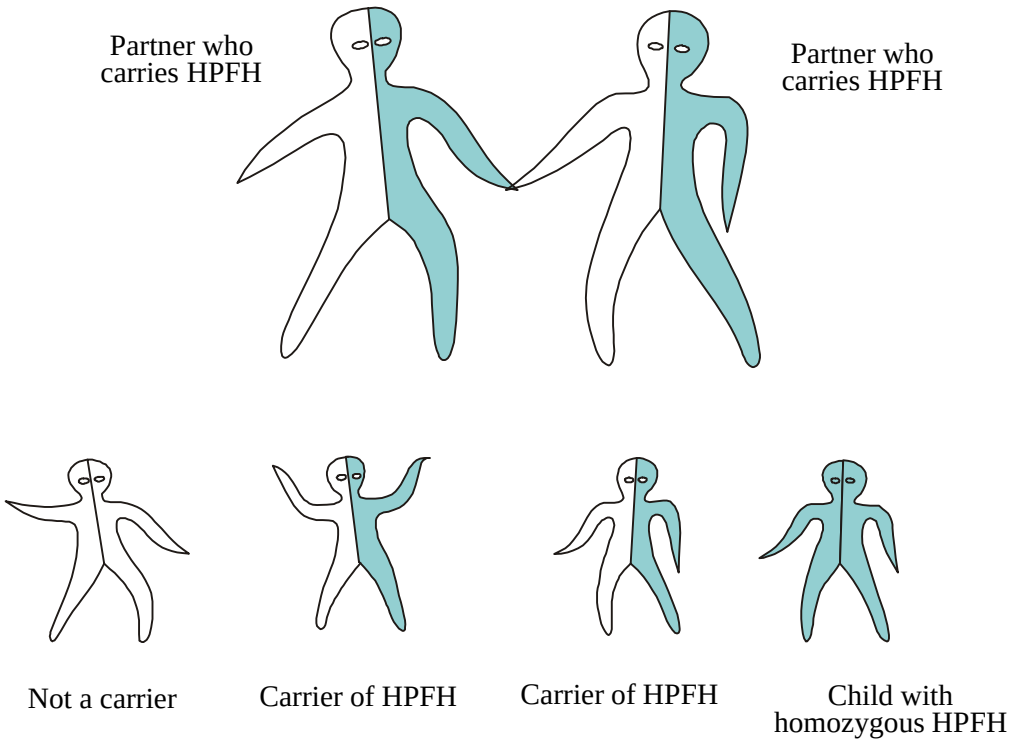
In each pregnancy there are two possibilities:

- The child may not carry any haemoglobin variant.
- The child may carry HPFH. This is harmless.

This couple has the same chance of a healthy family as other couples do.

There is also no known risk of a serious haemoglobin disorder if their partner carries any other common haemoglobin variant.

If both partners carry HPFH, their children could have homozygous HPFH



In each pregnancy there are three possibilities:

- The child may not carry any haemoglobin variant.
- The child may carry HPFH. This is harmless.
- The child may inherit HPFH from both parents. Such a child would have homozygous HPFH. This is also known as having only fetal haemoglobin (haemoglobin F). It is practically harmless.

This couple has the same chance of a healthy family as other couples do.

Why is it important for a person to know that they carry HPFH?

There are three main reasons.

- Firstly, if they have a blood test for any reason it may show that they carry HPFH. If HPFH is mistaken for a different haemoglobin variant, this can lead to confusion, misinformation and uneccessary tests. A person who carries HPFH and understands that it is harmless can explain it to any health workers they see, and these problems can be avoided.
- Secondly, if they are thinking of having children, they should ask their partner to have a blood test “for haemoglobin disorders”. If their partner is not a carrier they can be sure that there is no risk to the health of their children. However, if their partner is also a carrier they should see an expert in haemoglobin disorders to confirm that there is no risk to the health of their children.
- Thirdly, their blood relatives, for example their children or brothers or sisters, may also carry HPFH and may also have problems with misinformation and uneccessary tests. They should tell them about carrying HPFH, and advise them to have a blood test “or haemoglobin disorders”.

Asking a partner to have a blood test

A carrier who is thinking of having children needs to tell their partner that they carry HPFH, and ask him or her to have a blood test "for haemoglobin disorders".

Is it difficult for a carrier to ask their partner to have a blood test?

It can be easy in some cases and difficult in others. It is easier if both the carrier and their partner know that:

- Carrying a haemoglobin variant is common.
- The test will probably show that the partner does not carry a haemoglobin variant.
- If they do carry a haemoglobin variant it will not affect their health or lifestyle in any way.
- A couple who are both carriers can have a healthy family.
- Expert counselling is available.
- The results of blood tests, and the couples' decisions, are completely confidential.

If a couple are both carriers, can it interfere with their relationship?

It is unusual for a relationship to suffer because one or both of a couple carry a haemoglobin variant.

When is the best time for a carrier to ask their partner to have a blood test?

Ideally as early as possible, because it can take time to arrange a blood test, and for a couple who are both carriers to see an expert. Of course, the best timing depends on the couple's relationship with each other. It may also be influenced by their families' views, and the culture they belong to.

In practice a carrier might suggest a blood test to their partner at one of the following points.

- Before they settle down together.
- After they settle down together but before they start a pregnancy.
- As soon as they have started a pregnancy.

Telling the family about HPFH

A carrier inherited HPFH from one of their parents, so their brothers and sisters and other blood relatives could also be carriers. For example a brother or sister has a 1 in 2 chance of being a carrier.

If a carrier has brothers or sisters, or already has children, they should advise them to ask their GP or practice nurse for a blood test "for haemoglobin disorders".

Carrying HPFH

(hereditary persistence of fetal haemoglobin)…

… is not an illness, and will never become an illness. It was passed to you by one of your parents and you could pass it on to your children.

It is a variation of the blood.

It will not affect the health of your children, even if your partner is also a carrier.

- If you are thinking of having children, your partner should have a blood test “for haemoglobin disorders”, to confirm that there is nothing to worry about.
- If you have children or brothers and sisters, they could carry HPFH like you.

To find out more, see your GP or a specialist counsellor.