Pulmonary Arteriovenous Malformations (PAVMs)

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Pulmonary Arteriovenous Malformations (PAVMs)

Pulmonary arteriovenous malformations (or PAVMs for short) are abnormal blood vessels in the lungs. As shown in Figure 1, they let blood bypass or "shunt" past the lung airsacs. As this blood does not receive oxygen, your oxygen levels drop which can make your lips appear blue, and as blood passing through PAVMs is not properly filtered, mini-strokes or brain abscesses can result. Occasionally, the fragile vessels may bleed and make you cough up blood. However, most of the time, people with PAVMs feel entirely well.

Why is it important to be treated for PAVMs?

Patients who have PAVMs are at risk of having a ministroke or brain abscess even if they feel well. In addition, a few women with PAVMs have complications during pregnancy. It is therefore important to check whether or not PAVMs are present, and if so, arrange for a simple treatment to close them off.

How are PAVMs treated?

Although PAVMs used to be treated by surgical removal, it is now recommended that they are treated by blocking off the feeding artery with metal coils or balloons (embolisation treatment). Under local anaesthetic, a small tube is passed into a leg vein through which coils or balloons can be inserted. This procedure usually takes about 2 hours, though afterwards you have to lie flat for 6 hours. The next day you are allowed home, with a letter for your doctor explaining what you have had done. Embolisation treatment is very safe in experienced hands. For instance, Dr James Jackson at the Hammersmith Hospital in London has published results on over 140 patients with PAVMs treated since 1988 – these results are at least as good as any worldwide. Occasionally there may be times when surgery could be the better option, but this is only rarely the case because unlike embolisation, surgery removes good normal lung – this is an important consideration because you may develop further PAVMs or other chest disease later in life.

Once I have had the embolisation treatment for PAVMs, am I cured for life?

Many patients need more than one admission to treat all of their PAVMs. At the Hammersmith Hospital, we let you know if this applies to you before you leave hospital. Only one-third of PAVM patients will be completely cured. In most cases, tiny PAVMs that are impossible to treat remain.
We are not sure how important these tiny vessels are, but it is important that you are followed up regularly in a specialist clinic to check if these vessels are becoming larger, requiring further treatment. If your PAVMs have not been completely cured, you will be advised to take antibiotics before dental treatments or surgery – your GP will need to treat you like a patient with abnormal heart valves, and your PAVM doctors may be able to provide you with a special card. There are also important considerations for your family:

Your family and Hereditary Haemorrhagic Telangiectasia (also known as HHT or Osler-Weber-Rendu syndrome)

It is possible for PAVMs to occur as an isolated finding, but this is unusual. Nine times out of ten, careful questioning and examination picks up features of an inherited condition known as HHT which leads to the development of abnormally wide and fragile blood vessels. This can cause picks up features of an inherited condition known as HHT which leads to the development of abnormally wide and fragile blood vessels. This can cause

- **Nose bleeds**, which often start in childhood, but may develop in later adult life.
- **Blood spots** on lips or fingertips (these are just visible blood vessels).
- **Anaemia**, due to bleeding from the nose, and sometimes the gut. This anaemia may need treatment with iron and even blood transfusions if severe.
- **Pulmonary arteriovenous malformations (PAVMs)** in at least one in five individuals with HHT.
- **Other abnormal blood vessels**: These affect a few people with HHT, but it is not clear that these need to be detected if they are not causing problems already.

Because PAVMs affect one in five people with HHT, **it is very important that we pick up HHT if it is present in your family so that your relatives can be screened and if necessary treated for PAVMs before they have any complications.** HHT may not become apparent until late in life, so it is safer to think about this condition than to ignore it.

Will my children be affected by HHT and PAVMs?

HHT is a hereditary disease, passed on from parent to child. It is caused by an abnormal gene – if one parent has the disease, then half of their children are likely to inherit the abnormal gene. The earliest sign of HHT in children is usually nose bleeds. However, because HHT may not become apparent until late in life, we are unable to say that younger members of a family are definitely unaffected.
It is sensible for their medical records to include the fact that they have a parent with HHT, and for them to be screened for PAVMs.

Who in my family should be tested for PAVMs, and when?

PAVM screening is important for anyone with HHT, but all children of affected family members should be assessed. We recommend screening in adults, to be repeated after five to ten years. We can screen children but this needs to be repeated after puberty. It is particularly important to check young women before they become pregnant.

What should I Do?

- If you have, or think you have, HHT, the best plan is to discuss the matter with your GP. You need to be told the implications for you and your family, and your GP may refer you to your local hospital for a check up. As HHT is a rare disorder and generally not well covered in medical schools, it is possible that your GP or hospital doctor will need further information. This can be obtained from:

  - The Hammersmith Hospital PAVM/HTT Team (headed by Dr Claire Shovlin and Dr James Jackson, http://www.hhtathht.com)
  - The Telangiectasis Self Help Group, 38 Sunny Croft, Downley, High Wycombe, Bucks HP13 5UQ
  - HHT Foundation International, PO Box 8087, New Haven, CT 06530, USA or http://www.hht.org

(Dr Shovlin and Dr Jackson are on the medical board of this international foundation for families with HHT, but point out that the website is directed towards North American patients who like to know everything about their condition, whether or not it will ever cause them problems).
This leaflet is derived from the Hammersmith Hospitals NHS Trust’s leaflet “PAVMs and HHT: What you need to know about Hereditary Haemorrhagic Telangiectasia (HHT) and Pulmonary arteriovenous malformations (PAVMs)” and was prepared by Dr Claire Shovlin, Senior Lecturer and Honorary Consultant in Respiratory Medicine, Hammersmith Hospital.

Approved by Dr M J Cushley July 2008