LUNG *(Pulmonary)* ARTERIOVENOUS MALFORMATIONS & HHT

COMPANION FACTSHEET TO
MY HHT CARE CHECKLISTS

SIGNS AND SYMPTOMS

- SHORTNESS OF BREATH
- LOW OXYGEN SATURATION OR DECREASED OXYGEN IN BLOOD
- DECREASED EXERCISE TOLERANCE
- MIGRAINES
- BLUISH OR PALE LIPS OR FINGERS
- COUGHING UP BLOOD

AFFILIATED ISSUES

- MIGRAINES
- STROKE
- BRAIN ABSCESS
- LUNG HEMORRHAGE

FACTSHEET **FS**

SOME IMPORTANT FACTS TO REMEMBER ABOUT HHT ARE:

At least 40% of people with **HHT (Hereditary Hemorrhagic Telangiectasia)** have pulmonary AVMs (about 50% with HHT1 and about 10% with HHT2).

Pulmonary AVMs often present early in life and are found in children of all ages with all types of HHT.

People are often unaware that they have pulmonary AVMs until they develop a life-threatening complication, such as stroke, brain abscess or lung hemorrhage.

With the right screening and treatment, these life-threatening complications can be prevented.

During pregnancy pulmonary AVMs can be especially dangerous as the volume of blood flowing through the body significantly increases and make complications more likely.

cure hht
The Cornerstone of the HHT Community

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Pulmonary (lung) arteriovenous malformations (AVMs) are direct artery to vein connections in the pulmonary circulation. Normally, the lung arteries get smaller and smaller as they go deeper into the lungs, similar to the branches of a tree. At the ends of these artery branches, hair-like blood vessels called **capillaries** join the **arteries** and **veins**. These capillaries perform many important functions including allowing passage of oxygen into the blood as well as filtering the blood of **impurities (clots, bacteria, air bubbles)** before the blood circulates to the brain and other organs. With a pulmonary AVM, these capillaries are missing, and the artery connects directly to the vein. If the artery leading to the pulmonary AVM is larger than two to three millimeters in diameter, small **blood clots** can travel through the pulmonary AVM and go into the brain, causing a **stroke**. **Bacteria** can also travel through AVMs and result in **brain abscesses (a brain infection)**. Stroke and brain abscess can be life-threatening.

HERE ARE SOME THINGS TO DISCUSS WITH YOUR PHYSICIAN:

Whether you are due for routine screening for pulmonary AVMs.

If you have been previously diagnosed with and/or treated for pulmonary AVMs.

If you have any of the listed signs and symptoms.

Getting screening and/or treatment at an HHT Center of Excellence.

Talk to your dentist and other health care professionals about the need for pulmonary AVM precautions, such as prophylactic antibiotics.

HOW IT IS DIAGNOSED

- **Transthoracic contrast echocardiography (echo bubble study)**: The recommended study for initial screening. This test uses sound waves (ultrasound) to determine if injected saline bubbles can get through the lung circulation and be seen back in the heart, on the left side. This is called a **shunt**. An **IV** will need to be started for saline bubbles to be given. No radiation is used during this study.
- **CT (computed tomography) scan**: If the echo bubble study is positive, diagnosis should be confirmed with a CT scan. This a high-resolution X-ray of your lungs. If **contrast** (dye) is used, an IV will need to be started.
- **Screening** should be performed at the time of initial clinical evaluation for HHT.
- **Repeat screening** should be performed every 5-10 years AND after puberty, after pregnancy, within 5 years preceding planned pregnancy.

TREATMENT

- **Pulmonary embolization**: A procedure performed to block the blood flow to the abnormal vessels. The patient is given **sedation** or general anesthesia for this procedure. In an angiography suite, a **catheter** (small tube) is inserted into a **vein** in the top of the thigh and directed through the blood vessels in the body to the pulmonary arteries. A small **coil** or plug is then inserted to block off the artery that leads into or "feeds" the **pulmonary AVM**. This stops the blood flow to the pulmonary AVM which eliminates the occurrence of a potentially life-threatening complication. After the procedure, the patient is observed for several hours or overnight before being discharged home.
- **Surgical Removal**: A surgical procedure to remove the part of the lung that contains the pulmonary AVM. Because of the success of embolization, surgery is rarely necessary.
- Pulmonary AVMs of a certain size should be treated (those feeding an artery diameter of **2-3 mm or greater**).

IMPORTANT PRECAUTIONS FOR PATIENTS WHO HAVE BEEN DIAGNOSED WITH A PULMONARY AVM OR WHO HAVE NOT YET BEEN SCREENED FOR THEM:

- **Antibiotic Prophylaxis**: Recommended for **dental** and other procedures that can introduce **bacteria** in the blood.
- **IV Filter**: An **IV air filter (bubble trap)** should be used when possible, if a patient has an intravenous **(IV)** line. This is to prevent any large air bubble from entering the bloodstream, going through a **lung AVM**, and then causing a temporary **stroke**. This is most effectively done by using a filter in the IV line as close to the patient as possible. A **0.22 micron filter** is best if available, but a **blood filter** is also acceptable (about 260 microns) and will stop all large air bubbles. During a **blood transfusion**, a standard blood filter is all that is needed. Please note that filters often cannot be used for IV contrast injections like you might get for CT or MRI scans.
- Avoidance of **SCUBA diving** is recommended.

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