MY PULMONARY AVM CARE CHECKLIST

USING THE HHT GUIDELINES

Pulmonary AVMs= pulmonary arteriovenous malformations (lung AVMs). The HHT Pulmonary AVM Guidelines are detailed on the next pages.

Date: _____

Name: _____

Please check all that apply

☐ I HAVE HHT OR MIGHT HAVE HHT (THE HHT DIAGNOSIS HAS NOT BEEN RULED OUT).

- ☐ Ask my doctor to screen me for lung arteriovenous malformations (AVMs), typically with contrast echocardiography ("bubble echo").
- ☐ Consider having my screening done at an HHT Center of Excellence.

☐ I HAVE LUNG (PULMONARY) AVMs.

- ☐ Request preventative treatment (embolization) of my AVMs, even if I have no symptoms and my oxygen levels are normal.
- ☐ Ask my doctor to refer me to an HHT Center of Excellence for the preventative treatment (embolization) of my AVMs.
- ☐ Talk to my dentist and all my health care professionals about my need for lung AVM precautions, lifelong.
- ☐ Avoid SCUBA diving, lifelong.
- ☐ Plan regular long-term follow-up for the lung AVMs, even once they are treated.

☐ I HAVE TINY LUNG AVMs WHICH HAVE NOT BEEN TREATED.

- ☐ Plan for long-term follow-up to monitor for growth of the lung AVMs.

☐ I HAVE UNDERGONE SCREENING AND I DO NOT HAVE LUNG AVMs.

- ☐ Plan with my doctor for re-screening in 5 years' time.

INTERNATIONAL HHT GUIDELINES

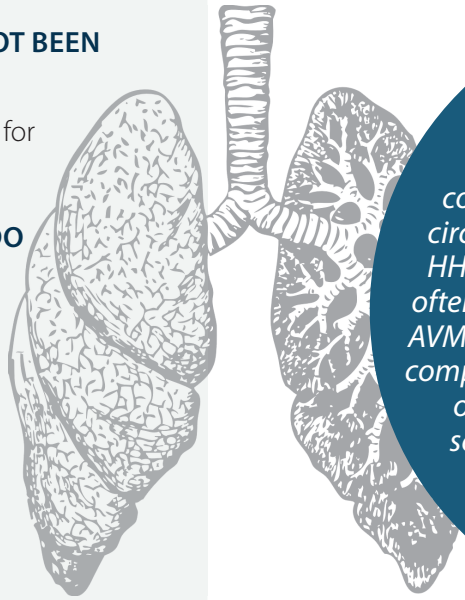
WHAT ARE THE HHT GUIDELINES AND WHY ARE THEY IMPORTANT?

- The HHT Guidelines are recommendations for care based on evidence and expertise from HHT experts from around the world.
- The HHT Guidelines help ensure that people living with HHT get the best care possible.

WHAT IS MY ROLE AS SOMEONE LIVING WITH HHT?

- Be aware of the Guidelines. Share them with your care team. Ideally you should be seen at an HHT Center of Excellence or your care team may want to consult with one.
- Read up on your condition and know what care is available for HHT.
- **Prepare ahead of time for your appointments**: Bring your HHT Care Checklists and a family member or friend. They can help you communicate your questions and priorities, as well as act as a second set of ears. Share your experiences, worries and priorities to help your care team better understand your needs and provide individualized care.

PULMONARY AVMs IN HHT



Arteriovenous malformations (AVMs) are direct artery to vein connections in the lung (pulmonary) circulation. At least 40% of people with HHT have pulmonary AVMs. 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.

HHT GUIDELINES RECOMMENDATIONS

PULMONARY AVMs IN HHT

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The expert panel recommends that:
(all recommendations are from the First HHT Guidelines)

I1 CLINICIANS SCREEN ALL PATIENTS WITH POSSIBLE OR CONFIRMED HHT FOR PULMONARY AVMs.

Clinical Considerations: Screening should be performed at the time of initial clinical evaluation for HHT. Although less evidence exists in children, the expert panel included children in the screening recommendation, since they are also at risk of life-threatening complications and treatment appears to be similarly effective. In patients with negative initial screening, repeat screening should be considered after puberty, after pregnancy, within 5 years preceding planned pregnancy and otherwise every 5-10 years.

I2 CLINICIANS USE TRANSTHORACIC CONTRAST ECHOCARDIOGRAPHY AS THE INITIAL SCREENING TEST FOR PULMONARY AVMS.

Clinical Considerations: Screening should be performed by clinicians with significant expertise in HHT, usually in an HHT center of excellence, to achieve the accuracy and low risks reported in the literature. Transthoracic contrast echocardiogrpahy (TTCE) is considered positive if there is detection of any bubbles in the left atrium. Positive screening should be confirmed with unenhanced multidetector thoracic CT with thin-cut (eg. 1-2mm) reconstructions. CT was not recommended as a screening test, due to the associated radiation exposure, but could be considered for screening in centers without expertise in TTCE for pulmonary AVM screening.

I3 CLINICIANS TREAT PULMONARY AVMS WITH TRANSCATHETER EMBOLOTHERAPY.

Clinical Considerations: The selection of pulmonary AVMs for embolization is based on feeding artery diameter, generally 3mm or greater, though targeting AVMs with feeding artery diameter as low as 2 mm may be appropriate. This procedure should be performed by clinicians with significant expertise in embolizing pulmonary AVMs, usually in an HHT center of excellence, to achieve the effectiveness and low-risks reported in the literature. This is particularly relevant when considering embolization in rare or higher risk situations, such as during pregnancy and in patients with mild-moderate pulmonary hypertension. The panel agrees there is no role for surgical management of pulmonary AVMs, other than in the management of life-threatening bleeding in a center where embolization expertise is unavailable.

I4 CLINICIANS PROVIDE THE FOLLOWING LONG-TERM ADVICE TO PATIENTS WITH DOCUMENTED PULMONARY AVMS (TREATED OR UNTREATED):

- Antibiotic prophylaxis for procedures with risk of bacteremia
- When IV access is in place, take extra care to avoid IV air
- Avoidance of SCUBA diving

Clinical Considerations: The rationale for recommending prophylactic antibiotics for bacteremic procedures in people with pulmonary AVMs, is based on expert opinion that cerebral abscess is frequent in these patients (approximately 10% before pulmonary AVM diagnosis), that cerebral abscess in these patients occurs primarily as a complication of bacteremic procedures, the fact that cerebral abscess is associated with considerable morbidity and mortality and that this precaution is low-risk. The AHA guidelines for prevention of bacterial endocarditis should be followed for choice of antibiotics. Similarly, careful avoidance of intravenous air bubbles is recommended to prevent cerebral air embolism, and this could include an in-line filter. There are only theoretical arguments for avoidance of SCUBA suggesting that there may be an increased risk of complications of decompression in patients with pulmonary AVMs. These precautions should be followed life-long, regardless of size of pulmonary AVMs, even once AVMs are treated. These precautions should also be considered in HHT patients in whom pulmonary AVMs have not been excluded or in whom microscopic pulmonary AVMs are suspected (for example, detected on TTCE but not detectable on CT).

I5 CLINICIANS PROVIDE LONG-TERM FOLLOW-UP FOR PATIENTS WHO HAVE PULMONARY AVMS, IN ORDER TO DETECT GROWTH OF UNTREATED PULMONARY AVMS AND ALSO REPERFUSION OF TREATED AVMS.

Clinical Considerations: Follow-up allows the identification of embolized pulmonary AVMs that have re-perfused and other pulmonary AVMs that have grown large enough to be considered for embolization. Multidetector thoracic CT with thin-section reconstruction (1-2mm) should be undertaken within 6-12 months after embolization and then approximately every 3 years after embolization. For patients with only small untreated pulmonary AVMs and in patients with suspected microscopic pulmonary AVMs (for example, detected on TTCE but not detectable on CT), the follow-up period should be determined on a case by case basis (approximately every 1-5 years) with CT (as above), with consideration for limiting radiation exposure.