

Validated HHT “Do’s and Don’ts” from VASCERN HHT Working Group

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1. Physical Activity

What is recommended

- There are no physical activity or sport restrictions, except in the event of acute hypoxia.

What you should not do

- Scuba diving with a diving tank in patients with pulmonary arteriovenous malformations, even if embolised (risk of air embolism).

2. Breast feeding

What is recommended

- Breastfeeding is not contraindicated in women with hereditary hemorrhagic telangiectasia

What you should not do

- No specific recommendations

3. Contraindicated medications

No medication is formally contraindicated.

What is recommended

- Always discuss the risks and benefits with the centre of reference or competence responsible for the care of the patient.
- Adapt the treatment to the patient's clinical condition (epistaxis, gastrointestinal bleeding)

What you should not do

- Prescribe APAs or anticoagulants without having weighed the potential risks and benefits.

4. Antiplatelet agents (APA) and anticoagulants

What is recommended

- Discuss the risks and benefits with the centre of reference or expertise responsible for the patient.
- Adapt the treatment to the patient's clinical condition (epistaxis, gastrointestinal bleeding)
- After an ischemic stroke secondary to pulmonary arteriovenous malformations, there is no indication to continue this type of treatment (APA or anticoagulant)-if all pulmonary arteriovenous malformations have been treated satisfactorily.

What you should not do

- Prescribe APAs or anticoagulants without having weighed the risks and benefits.

5. Deep-vein thrombosis, pulmonary embolism(or venous thromboembolic disease)

There are no coagulation anomalies reported in hereditary hemorrhagic telangiectasia.

See also Statement 1. “Antiplatelet agents and anticoagulants”

What is recommended

- Follow the standard treatment for thrombosis and/or pulmonary embolism (anticoagulant treatment) having weighed the potential risks and benefits
- Adapt the treatment to the patient's clinical condition (epistaxis, gastrointestinal bleeding, blood count)
- In the event of increased epistaxis while taking anticoagulation treatment, arrange for a consultation with an ENT doctor who has knowledge of the disease in order to consider appropriate treatment.
- Outside of an emergency situation, and depending on anti-coagulant tolerance, discuss alternative treatments (thrombectomy, cava filter) with the centre of reference or competence.

What you should not do

- Avoid treating vascular thromboembolic disease because of hereditary hemorrhagic telangiectasia

6. Hemorrhagic stroke

What is recommended

- Emergency management and treatment (as in non-HHT patients)
- Look for underlying brain AVMs to prevent recurrence.
- If the patient's clinical condition requires the insertion of a nasogastric tube, it should be soft, small diameter (unless clinical circumstances demand a large bore tube), and put in place with extreme caution due to the risk of triggering a severe episode of epistaxis related to the presence of mucous telangiectases.

What you should not do

- No specific contraindications

7. Brain abscesses:

A brain abscess is a classic complication of hereditary hemorrhagic telangiectasia. It is related to the right-to-left shunt secondary to pulmonary arteriovenous malformations.

What is recommended

- Emergency management and treatment (as in non-HHT patients)
- Perform a chest CT scan without injection, or contrast echocardiogram to identify pulmonary arteriovenous malformations (the most common cause of brain abscess in hereditary hemorrhagic telangiectasia), and treat the pulmonary arteriovenous malformations to reduce the risk of recurrence.
- If the patient's clinical condition requires the insertion of a nasogastric tube, this should be soft, small diameter (unless clinical circumstances demand a large bore tube), and put in place with extreme caution due to the risk of triggering a severe episode of epistaxis related to the presence of mucous telangiectases

8. Heart failure

Heart failure in hereditary hemorrhagic telangiectasia may be related to the evolution of hepatic AVMs that can entail cardiac chronic overload: both hepatic and cardiac hemodynamics must be investigated.

Medical treatment will be adapted to each particular case: treatment of heart failure, correction of anemia, management of arrhythmia.

What is recommended

- Measure cardiac output and the cardiac index, the filling pressures and the presence or absence of pulmonary arterial hypertension (often post-capillary).
- Search for hepatic arteriovenous malformations (Doppler ultrasound and/or hepatic scan).
- Refer the patient to a centre of reference.
- Correct the anaemia

What you should not do

- Overlook cardiac evaluation (including echocardiography) if severe liver VMs are present
- Treat pulmonary arterial hypertension (PAH) secondary to liver VMs with high output cardiac failure with vasodilators.

9. Kidney failure

What is recommended

- No contraindications to kidney biopsy puncture after exclusion of kidney AVMs by doppler sonography

What you should not do

- No specific recommendations

10. Care for patient with multiple traumatic injuries

It is always necessary to contraindicate nasal manipulations (nasal intubation, aspirations, etc.) due to the significant risk of triggering sometimes very severe episodes of epistaxis linked to mucous telangiectases.

Apart from the risk of bleeding related to the presence of mucous telangiectases (nasal, gastrointestinal), there are no coagulation anomalies associated with hereditary hemorrhagic telangiectasia and no surgical bleeding risk connected with this pathology.

What is recommended

- Check that there is no low SaO₂ that could be related to the presence of undiagnosed pulmonary AVMs, which would warrant treatment.

What you should not do

- Intubate or aspirate through the nose: risk of severe epistaxis

11. Bronchoscopies

Be aware that coughing may be tolerated less well than in other patients due to the risk of haemoptysis from tube abrasion

What is recommended

- Follow the standard fibroscopy technique.
- In case of biopsy, perform with antibiotic prophylaxis if pulmonary AVMs are present, or if pulmonary status unknown

What you should not do

- Nasal manipulations during anaesthesia (nasal intubation, aspirations, etc.) due to the significant risk of triggering sometimes very severe episodes of epistaxis linked to mucous telangiectases.

12. Aortic dissection

Before any surgery it is always necessary to contraindicate nasal manipulations (nasal intubation, aspirations, etc.) due to the significant risk of triggering sometimes very severe episodes of epistaxis linked to mucous telangiectases

Apart from the risk of bleeding related to the presence of mucous telangiectases (nasal, gastrointestinal), there are no coagulation anomalies associated with hereditary hemorrhagic telangiectasia and no surgical bleeding risk connected with this pathology.

What is recommended

- Emergency management and treatment (as in non-HHT patients)
- Follow standard treatment protocols for this pathology

What you should not do

- Intubate or aspirate through the nose: risk of severe epistaxis.

13. Pneumothorax

Nasal manipulations (nasal intubation, aspirations, etc.) should always be contraindicated because of the significant risk of triggering sometimes very severe episodes of epistaxis linked to mucous telangiectases.

Apart from the risk of bleeding related to the presence of mucous telangiectases (nasal, gastrointestinal), there are no coagulation anomalies associated with hereditary hemorrhagic telangiectasia and no surgical bleeding risk connected with this pathology.

What is recommended

- Emergency management and treatment (as in non-HHT patients)
- Follow standard treatment protocols for this pathology
- If the patient's clinical condition requires the insertion of a nasogastric tube, this should be soft, small diameter (unless clinical circumstances demand a large bore tube), and put in place with extreme caution due to the risk of triggering a severe episode of epistaxis related to the presence of mucous telangiectases

What you should not do

- Intubate or aspirate through the nose: risk of severe epistaxis.