European Reference Network

VASCERN

# Do's and Don'ts Factsheets

for Rare Vascular Disease Care in Frequent Situations

*For HCPs*

## Hereditary Hemorrhagic Telangiectasia

*Pregnancy*

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## Do’s and Don’ts for Pregnancy

# Introduction

This factsheet has been prepared by the members of the VASCERN Hereditary Haemorrhagic Telangiectasia (HHT) Working Group to provide guidance for healthcare professionals on the management of pregnancy in patients with HHT. HHT pregnancies are considered high-risk and require close monitoring and medical supervision.

These recommendations have been developed through expert consensus within the HHT Working Group. While this factsheet serves as a helpful guide, individual patient care may vary. It is therefore essential to consult the appropriate medical specialists to ensure care is tailored to each patient's specific needs.

# Before Pregnancy

## WHAT IS RECOMMENDED

- Have a consultation at an expert HHT centre, including:
  - clinical examination with SaO2 measurement
  - screen for pulmonary AVMs (by contrast echocardiography and/or chest CT scan without contrast)
  - screen for anaemia and iron deficiency
  - discuss screening for hepatic AVMs (by Doppler ultrasound )
  - discuss screening for cerebral AVMs
- Whenever possible treat PAVMs before pregnancy.

## WHAT YOU SHOULD NOT DO

- Screening for spinal arteriovenous malformations (AVMs) in asymptomatic patients is not necessary.

# During Pregnancy

## WHAT IS RECOMMENDED

- Close medical supervision by an obstetrician with easy access to an HHT reference centre in case of pregnancy-related complications in patients with HHT.
- Inform the patient that:
  - coughing up blood or experiencing sudden shortness of breath requires immediate specialised consultation and hospitalisation, as this may indicate a ruptured PAVM (haemothorax or haemoptysis).
- Check for anaemia and iron deficiency at least every trimester.
  - In case of anaemia, oral iron intake or intravenous iron therapy should be administered according to the severity of the anaemia.
- At each antenatal visit, check for dyspnoea and $SaO_2$ levels.
  - If $SaO_2$ levels are below 95% and/or the patient is experiencing dyspnoea, seek advice from the nearest HHT centre.
  - If PAVMs are suspected, echocardiography with bubble contrast or a low-dose thoracic CT scan can be proposed.
  - If PAVMs are present, pulmonary embolisation can be performed on a case-by-case basis at an HHT centre.
  - In case of out of proportion dyspnoea heart failure should be excluded

## WHAT YOU SHOULD NOT DO

- Terminate the pregnancy only because of symptomatic PAVMs.

# Delivery

## WHAT IS RECOMMENDED

- In hospital delivery:
  - the usual practices should be maintained, whether it is a vaginal or a caesarean delivery.
  - Antibiotic prophylaxis is recommended unless PAVM has been ruled out.
  - If general anaesthesia is used, avoid PEEP and use a small tidal volume.
  - Epidural analgesia is allowed even without a spinal MRI in patients without neurological symptoms.
  - In the case of an IV infusion or injection, avoid air injection as with all other patients.

## WHAT YOU SHOULD NOT DO

- Intubate or aspirate through the nose.

# After Pregnancy

## WHAT IS RECOMMENDED

- Check for anaemia or iron deficiency.
- Re-screen for PAVMs (by contrast echocardiography and/or a chest CT scan without contrast) after giving birth.
- Continue regular follow-up at the HHT centre.

## WHAT YOU SHOULD NOT DO

- Abstain from breastfeeding due to HHT.

# Editorial Board/ Contributors

## This document was validated by the Hereditary Hemorrhagic Telangiectasia (HHT) Working Group.



## Cooperating Guests



**HCP member before Brexit, January 1st, 2021*

European Reference Network

VASCERN

Gathering the best expertise in Europe
to provide accessible cross-border healthcare
to patients with rare vascular diseases

VASCERN, the European Reference Network on Rare Multisystemic Vascular Diseases, is dedicated to gathering the best expertise in Europe in order to provide accessible cross-border healthcare to patients with rare vascular diseases (an estimated 1.3 million affected). These include arterial diseases (affecting the aorta to small arteries), arterio-venous anomalies, vascular malformations, and lymphatic diseases.

VASCERN currently comprises 48 expert teams from 39 highly specialized multidisciplinary HCPs? coming from 19 EU Member States, as well as various European Patient Organisations, and is coordinated in Paris, France.

Through our 6 Rare Disease Working Groups (RDWGs) as well as several thematic WGs and the ePAG – European Patient Advocacy Group, we aim to improve care, promote best practices and guidelines, reinforce research, empower patients, provide training for healthcare professionals and realize the full potential of European cooperation for specialised healthcare by exploiting the latest innovations in medical science and health technologies.

More information is available at: www.vascern.eu

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European Reference Network on Rare Multisystemic Vascular Diseases (VASCERN)
46, rue Henri Huchard
75018 Paris
France