HEREDITARY HAEMORRHAGIC TELANGIECTASIA

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RBWH

HHT

What is HHT

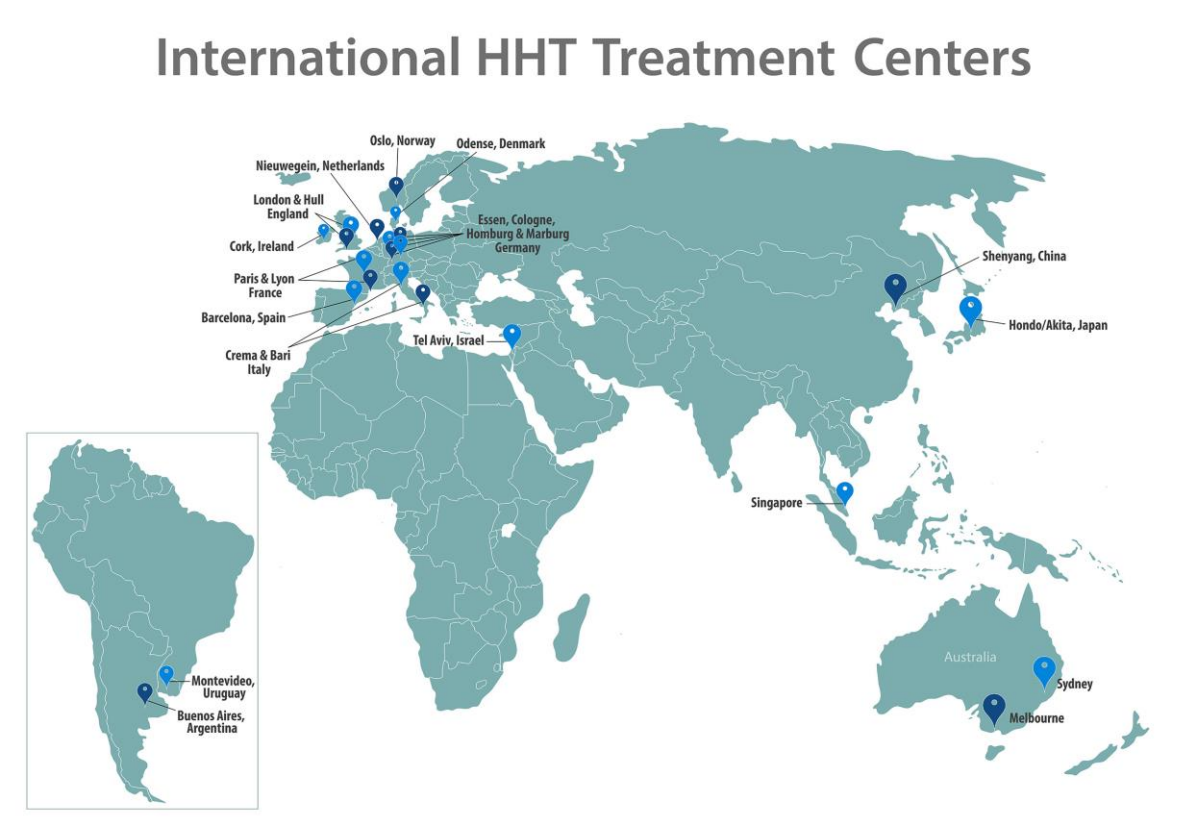
What causes HHT

How is HHT diagnosed

What are common symptoms for patients with HHT

How is HHT managed

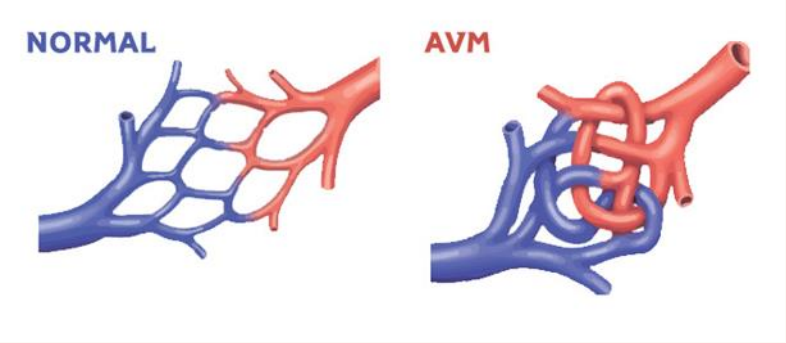
HHT AROUND THE WORLD



WHAT IS HHT

HHT is an uncommon inherited (genetic) disorder that causes the development of abnormal blood vessels

HHT patients may develop abnormal blood vessels called telangiectasias and large abnormal blood vessels called arteriovenous malformation (AVM's)

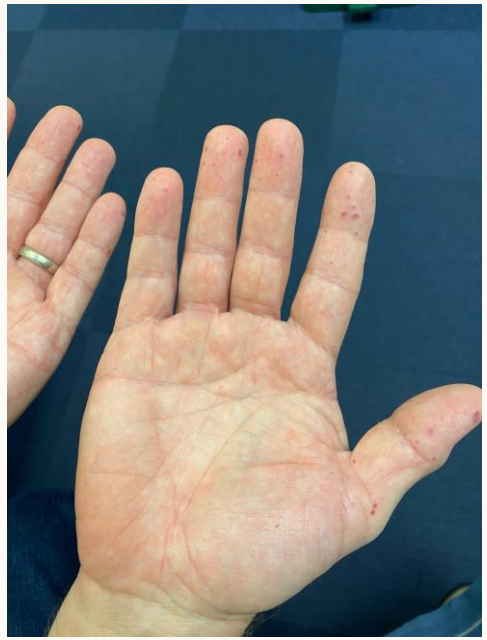


WHAT CAUSES HHT

It’s a genetic disorder passed down from one parent to the child

Autosomal dominant disorder

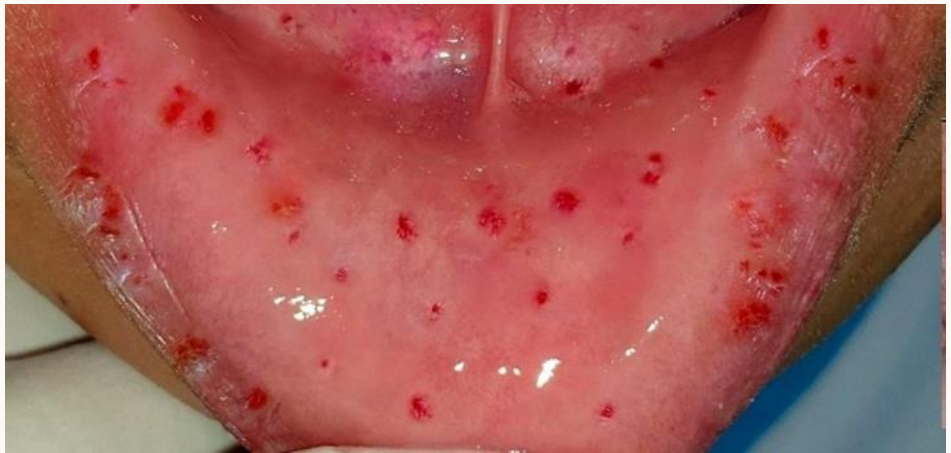
HOW IS HHT DIAGNOSED



Taking clinical history

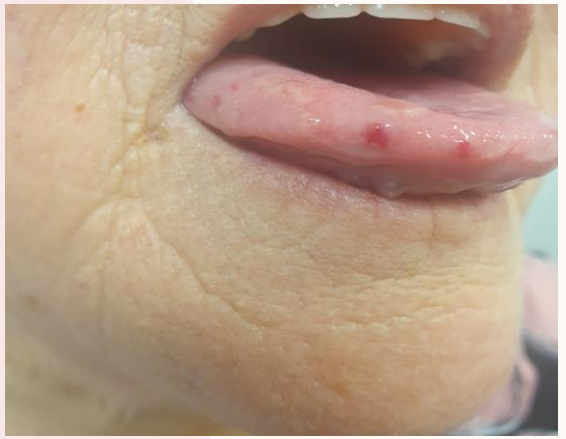
Family history

Genetic testing- helpful to know especially if not matching all clinical which mutation



HHT SYMPTOMS

- Epistaxis (Nose Bleeds)
- Gastrointestinal Bleeding
- Anaemia
- Arteriovenous Malformations (AVM's)



EPISTAXIS SCORE EXAMPLE

How often do you TYPICALLY have nose bleeding?
Less than once per month

How long does your TYPICAL nose bleeding last?
> 30 minutes

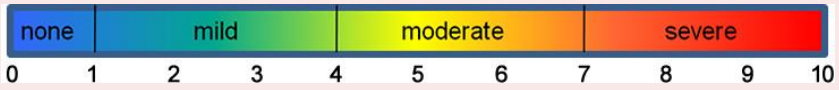
How would you describe your TYPICAL nose bleeding intensity?
Not Typically Gushing or Pouring

Have you sought medical attention for your nose bleeding?
Yes

Are you anemic (low blood counts) currently?
I don't know

Have you received a red blood cell transfusion SPECIFICALLY for nose bleeding?
No

Your normalized epistaxis severity score is ***4.71***, meaning your current risk is ***Moderate***



HHT MANAGEMENT

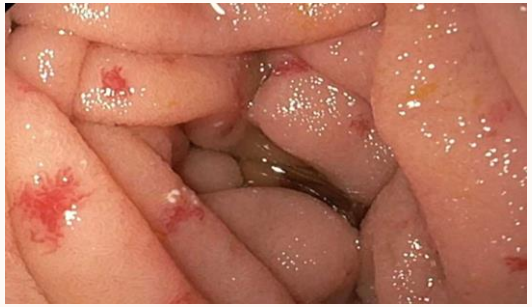
- **Epistaxis management**

* Humidification, nasal spray and/or ointment, ice packs to back of neck

* Severe and prolonged nosebleeds refer to ENT specialist who can consider ligation of bleeding vessels, intranasal therapies, tranexamic acid, nasal packing

- **Gastrointestinal bleeding**

* Monitor for black or dark stool if not on iron supplements
* May need iron infusion or blood transfusion in anaemic
* Endoscopy to determine if active bleeding telangiectasia
* Capsule Endoscopy if no active bleeding telangiectasias found but remain symptomatic



HHT MANAGEMENT

- **Anaemia Management**

* Iron supplements, iron infusions, blood transfusions, iron rich foods

- **AVM screening**

* MRI/CT sc ans of brain, lungs and ultrasound doppler Liver

HHT TREATMENT

Nosebleed treatments

AVM treatments- multiple options

Drug therapy- Tranexamic Acid and more recently Bevacizumab (Avastin)

HHT TREATMENT

What has changed with newer treatments?

ARTICLES

An international, multicenter study of intravenous bevacizumab for bleeding in hereditary hemorrhagic telangiectasia: the InHIBIT-Bleed study

Hanny Al-Samkari, Raj S. Kasthuri, Joseph G. Parambil, Hasan A. Albitar, Yahya A. Almodallal, Carolina Vázquez, Marcelo M. Serra, Sophie Dupuis-Girod, Craig B. Wilsen, Justin P. McWilliams, Evan H. Fountain, James R. Gossage, Clifford R. Weiss, Muhammad A. Latif, Assaf Issachar, Meir Mei-Zahav, Mary E. Meek, Miles Conrad, Josanna Rodriguez-Lopez David J. Kuter, Vivek N. Iyer

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THANK YOU

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