

HEREDITARY HEMORRHAGIC TELANGIECTASIA (HHT)

Osler-Weber-Rendu Syndrome

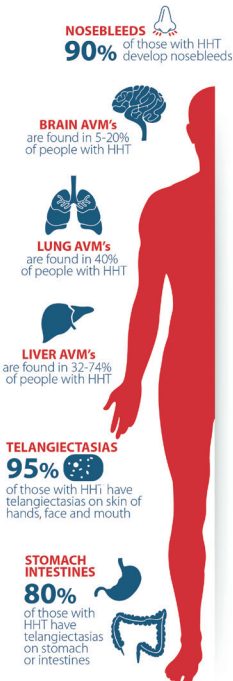
HHT is an inherited, genetic disorder that causes malformed blood vessels in multiple organs of the body, and typically begins with nosebleeds during childhood.

Approximately **1 in 5,000** people are affected by HHT, totaling **1.4 million** worldwide. However, HHT is often misdiagnosed and its symptoms are often mistaken for other conditions. The disease affects men, women and children at any age, as well as all ethnicities and races.

DIAGNOSIS

It's important to recognize that HHT does not affect every individual in the same way. Some people have symptoms in several different parts of the body (detailed below), while some may show hardly any symptoms at all.

AFFECTED ORGANS



COMMON MISDIAGNOSES

SKIN: Birthmarks or cherry angioma

NOSE: Allergies, drug addiction, hemophilia, or Von Willebrand Disease

BRAIN: Aneurysm, cerebral hemorrhage, epilepsy, abscess, migraine, or stroke

LUNGS: Asthma, Alzheimer's disease, lung disease, tumor, stroke, Polycythemia, Patent Foramen Ovale (PFO)

SPINE: Scoliosis or stroke

LIVER: Cirrhosis, heart failure, shortness of breath

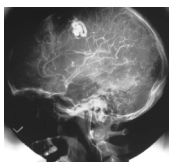
GI TRACT: Angiodysplasia, anemia, black stool or cancer

visit www.curehht.org for more info.

Diagnostic Criteria*

- » **Recurrent and spontaneous nosebleeds (epistaxis)**, which may be mild to severe.
- » Multiple **telangiectasias** on the skin of the hands, lips or face, or inside the nose or mouth.
- » **Arteriovenous malformations (AVMs)** or telangiectasias in the internal organs including the lungs, brain, liver, intestines, stomach, and spinal cord.
- » A **family history** (i.e., first-degree relative such as brother, sister, parent or child) who meets these same criteria for definite HHT or has been genetically diagnosed.

*Definite HHT = 3+ criteria; Possible HHT = 2 criteria



BRAIN AVM



LUNG AVM



TELANGIECTASIA

ICD-10 / Insurance Codes

HHT	178.0	Iron deficiency anemia	D50,D50.0
Cerebral AVM	Q28.2	Pulmonary AVM	Q25.72
Colon AVM	K55.21	Stomach AVM	537.83
Epistaxis	R04.0	Telangiectasia	448.9,178.8
GI or Liver AVM	Q27.33		

Complications & Statistics

- » **Untreated AVMs** can cause strokes, seizures, brain abscesses, numbness of the arms or legs, heart failure and in the worst of cases, death.
- » **50%** of HHT patients suffer from **anemia and iron deficiency** which causes symptoms such as fatigue and migraines.
- » **Dental care:** The direct connection of blood vessels and lack of capillary filtration in the lungs places patients at-risk to develop a brain abscess if bacteria enters the blood stream. 5-10% of patients with pulmonary AVMs develop a brain abscess. This may be preventable with the administration of a single dose of prophylactic antibiotics approx. one hour prior to dental work.



RESOURCES

- » **Locate HHT Treatment Centers of Excellence (CoEs)**
North America & Int'l: curehht.org/hht-treatment-centers
- » **Access the free Cure HHT Resource Library**
Review Clinical Guidelines: curehht.org/resource-library
- » **Review Physician Directory**
Add yourself or find a local doctor: directory.curehht.org