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Osler Weber Rendu Syndrome

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Introduction

Hereditary hemorrhagic telangiectasia (HHT) was first described by Henry Gawen Sutton in 1864. With similar symptoms to hemophilia, the two diseases were differentiated by Henri Jules Louis Marie Rendu in 1896.

HHT is also known as Osler-Weber-Rendu disease. It is an autosomal dominant disorder characterized by multiple mucocutaneous telangiectasias. These telangiectasias represent small arterio-venous malformations that frequently tend to bleeds causing the patient a significant amount of distress in their daily lives.
HHT is a rare genetic vascular disorder affecting about 1 or 2 in 10,000 individuals worldwide.

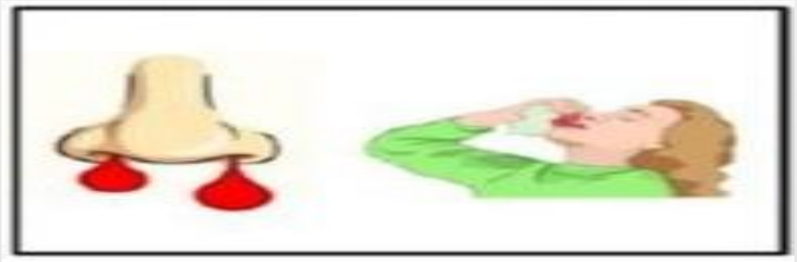
Causes

People with OWR inherit an abnormal gene that causes their blood vessels to form incorrectly. OWR is an autosomal dominant disorder. This means that only one parent needs to have the abnormal gene to pass it on to their children. OWR doesn't skip a generation. However, the signs and symptoms may vary greatly between family members. If you have OWR, it's possible that your child could have a milder or more severe course than you.

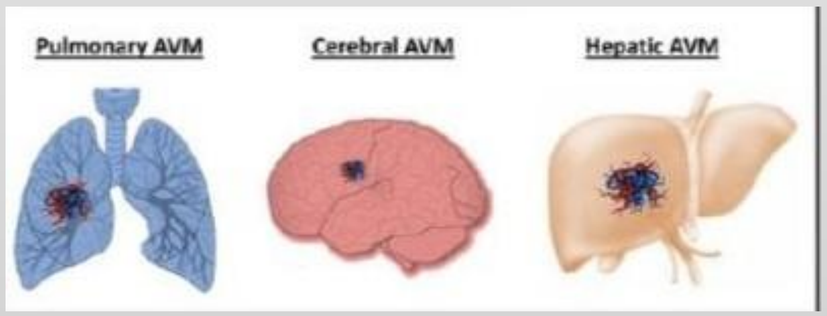
In very rare cases, a child can be born with OWR even when neither parent has the syndrome. This happens when one of the genes that cause OWR mutates in an egg or sperm cell

Symptoms

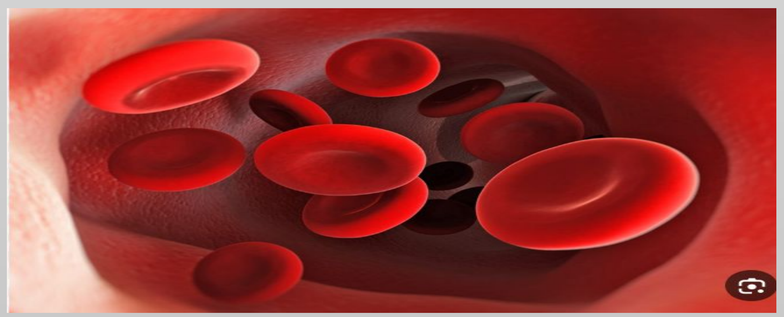
- Frequent nosebleeds in children



- Bleeding in the gastrointestinal tract (GI), including loss of blood in the stool, or dark or black stools.
- Seizures or unexplained, small strokes (from bleeding into the brain)
- Shortness of breath.
- Enlarged liver.
- Heart failure.



. Anemia caused by low iron



Management / Treatment

For most people with Raynaud's phenomenon, avoiding getting cold prevents attacks and keeps symptoms under control. But if this is not enough, medications can be divided into three broad categories:

1. Hormone-related drugs(estrogen)
2. Drugs that block blood vessel growth (Avastin)
3. Drugs that slow the disintegration of clots.(Tranexamic acid)

If you develop iron deficiency anemia, your doctor may also suggest intravenous iron .
in some cases, surgical procedures (nose , lungs , liver and brain) can help .

Summary

Hereditary hemorrhagic telangiectasia (HHT) is also known as Osler-Weber-Rendu disease.
People with OWR inherit an abnormal gene that causes their blood vessels to form incorrectly.
In very rare cases, a child can be born with OWR even when neither parent has the syndrome.
Symptoms of this disease is bleeding in organs(nose GIT and brain)
Some cases used medications and another surgical procedure.

References

https://www.ncbi.nlm.nih.gov/books/NBK482361/

https://www.healthline.com/health/osler-weber-rendu-syndrome#causes