

HEREDITARY HEMORRHAGIC TELANGIECTASIA

(also known as HHT, Osler-Weber-Rendu syndrome, Osler-Weber-Rendu disease, Osler-Rendu disease, Osler's disease, Weber-Osler disease, and Rendu-Osler-Weber syndrome)

Is the initiation of non-invasive dental hygiene procedures* **contra-indicated?** No

- **Is medical consult advised?** Yes, liaison with the patient/client's HHT specialist is advisable regarding the type, severity, and management of the patient/client's HHT before undertaking dental hygiene treatment for the first time. (This includes bleeding risk, if any, for fitting a mouth guard and taking an impression.) Medical consult is also warranted for suspicious, but as yet undiagnosed, bleeding disorder, and if known disease is poorly controlled.

Is the initiation of invasive dental hygiene procedures **contra-indicated?**** Yes. This is a bleeding disorder that may affect appropriateness or safety, and scaling and root planing, including curetting of surrounding tissue, are contraindicated until the patient/client is medically cleared. In some cases, antibiotic prophylaxis is also required.

- **Is medical consult advised?**..... See above. Additionally, bleeding risk (from oral telangiectasias¹) should be ascertained for invasive dental hygiene procedures. There are no reliable laboratory tests to determine bleeding tendency in persons with HHT; clinical findings and history of bleeding problems are used to identify patients/clients at risk.
- **Is medical clearance required?** Yes
- **Is antibiotic prophylaxis required?**..... Yes, if there are lung arteriovenous malformations (AVMs), whether treated or untreated, or if the patient/client has not yet been screened at a Specialized HHT Centre. (If there are lung AVMs, blood-borne bacteria resulting from invasive procedures can pass through the AVMs, lodge in the brain, and cause brain abscesses.) If the patient/client has been screened for lung AVMs and the tests confirm that there are none, then prophylactic antibiotics are not required.
- **Is postponing treatment advised?** Possibly. If the patient/client with HHT has not had the appropriate diagnostic tests for pulmonary AVMs, ideally dental hygiene treatment should be delayed until testing occurs.

Oral management implications

- Because HHT is often unrecognized by patients/clients and healthcare providers, the dental hygienist may be in position of detecting oral lesions (as well as facial, neck, and hand lesions) and a history suspicious of HHT (e.g., frequent nosebleeds and/or gum bleeds) in a previously undiagnosed patient/client. This should prompt medical referral for definitive diagnosis (which may include genetic testing, contrast echocardiography of the lungs, and magnetic resonance imaging - MRI - of the brain) and treatment before serious health consequences (particularly AVM complications) ensue.
- During dental hygiene treatment, certain patients/clients with HHT should remain upright to reduce the risk of nasal bleeding. Others (who have lung AVMs with significant shunting that results in orthodeoxia²) may better tolerate a recumbent position. Oxygen should be readily available, and blood pressure should be monitored before and after treatment.

1 [Telangiectasias](#) are vascular lesions of dilated small blood vessels, typically appearing as red to purplish dots. They consist of focal dilations of post-capillary venules with connections to dilated arterioles, initially through capillaries and later directly. Larger vessel dilations (such as those found in some internal organs) are referred to as [arteriovenous malformations \(AVMs\)](#).

2 Orthodeoxia is a fall in arterial blood oxygen on assuming the upright posture. It is usually caused by right-to-left cardiac or vascular shunting with a posturally induced fall in left-sided pressure permitting a corresponding gradient across the shunt.

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Oral management implications (*cont'd*)

- Gingival bleeding upon scaling is a prime concern. The treatment goals are to prevent and manage hemorrhage.
- Aspirin and non-steroidal anti-inflammatory drugs (NSAIDs), including naproxen and ibuprofen, should be avoided due to the elevated risk of bleeding.

Oral manifestations

- The oropharyngeal cavity (along with the skin and nasal cavity) is the most common area for telangiectasias. 75% of patients/clients with HHT will develop lesions on the buccal mucosa and skin, which are usually visible by age 40 years.
- Telangiectasias are often found on the buccal and labial mucosa; on and under the tongue; on the palate and gingiva; and on the facial skin and perioral region. Intraoral lesions may be macular (flat) or papular (elevated), and are usually “pinpoint-sized” or “pea-sized” (and less commonly linear in appearance). Typically having a red/purple appearance, they may resemble mucocutaneous petechiae, but unlike petechiae, telangiectasias blanch upon applied pressure.
- The lips and tongue are prime areas for small, pinpoint-size telangiectasias.
- While most oral (and skin) telangiectasias are asymptomatic, they are prone to rupture and hemorrhage. Bleeding gums may result from brushing the teeth or other minor oral trauma.
- Chronic oral ulcers and vesicles occur because of diminished vascular wall thickness associated with inflammation.
- Venous lakes³ may be found on the lips and in the oral mucosa (particularly buccal and lower inner lip).
- HHT-related anemia (from chronic bleeding) may manifest as atrophic glossitis (bald tongue), [angular cheilitis](#), and pallor of the lips and oral mucosa.

Related signs and symptoms

- HHT is an autosomal dominant genetic vascular disorder, occurring in one in 5,000 North Americans and about 1.4 million persons worldwide. Nearly all cases are familial (and hence the importance of family history), with spontaneous mutations being rare.
- HHT is multi-system vascular dysplasia. The arterioles, capillaries, and venules are affected, causing the vessels to become engorged and dilated. Telangiectatic lesions occur in the skin (usually appearing as macular red/purple spots ranging from 1 mm to more than 7 mm in diameter), mucous membranes, and internal organs; bleeding may occur because of the inherent mechanical fragility of affected vessels. Telangiectasias usually appear in affected persons before 40 years of age (with cutaneous changes usually beginning at puberty), and they tend to increase in number with aging. At least a few telangiectasias or spiderlike skin lesions will develop on the facial skin and/or hands in most persons with HHT by middle age; the nasal mucosa, fingertips, toes, and trunk also tend to be affected.

3 A venous lake is a bluish or red/blue, soft, discrete, painless bleb which is somewhat movable beneath the epithelium. It results from dilation of small blood vessels.

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Related signs and symptoms

- Some forms of HHT also manifest with arteriovenous malformations (AVMs, which are essentially larger forms of abnormal blood vessels), whose location and severity of symptoms are highly variable. The AVMs can be congenital, but they usually develop over time. Similar to telangiectasias, AVMs are usually asymptomatic but prone to rupture.
- The most commonly affected organs with HHT lesions are the skin, nose, lungs, gastrointestinal tract, liver, and brain. The spleen, pancreas, urinary tract, and spinal cord may also be affected.
- Bleeding from telangiectasia and/or AVMs can occur in virtually every organ, particularly from gastrointestinal and urogenital sites.
- Epistaxis (nosebleed) occurs in nearly 95% of persons with HHT by adulthood, with varying frequency and severity. The severity of HHT can often be gauged by the age at which epistaxis begins, with the most severely affected patients/clients experiencing recurrent nosebleeds in childhood.
- Significant gastrointestinal bleeding (more so in the stomach and small bowel than the colon) develops in 20% to 25% of persons with HHT, but rarely before age 50 years. Melena⁴ and iron deficiency anemia are sequelae, and the anemia can result in fatigue, shortness of breath, chest pain or light-headedness.
- Lung AVMs occur in 40% of persons with HHT, and pose a significant risk of bleeding. Pulmonary arteriovenous shunts are associated with oxygen desaturation, hemoptysis, hemothorax, brain abscess, and cerebral ischemia due to emboli. Patients/clients may show signs of hypoxemia or orthodeoxia with dyspnea, cyanosis, clubbing of the fingers and toes, and polycythemia.
- At least one cerebral AVM or aneurysm occurs in 5% to 20% of persons with HHT. These manifest at any time in life, including at birth, causing headaches and seizures, as well as serious complications such as hemorrhagic stroke, paralysis, and death.
- Liver AVMs are common, but they are mostly asymptomatic. However, large AVMs in the liver occasionally cause heart and liver failure, usually later in life.
- Cirrhosis of the liver occurs in some familial types of HHT.
- Hepatic and splenic arteriovenous shunts, as well as aortic and splenic aneurysms, may occur. Heart failure (associated with fatigue, shortness of breath, and swelling of the lower extremities, amongst other signs/symptoms) may result if the heart has been overworked for years, pumping extra blood through the low resistance pathway of an AVM (which in this context is called a “shunt”).
- Spinal vasculature involvement can cause pain in the back over the spine or loss of feeling or function in the arms or legs.
- Treatment of HHT includes laser therapy for nasal and cutaneous lesions; lubricants, antifibrinolytics, split-thickness skin grafting, embolization of arteriovenous communications, or hormonal therapy for epistaxis; pulmonary embolization or resection for lung AVMs; and hormonal therapy and laser coagulation (as well as iron therapy if anemia is present) for gastrointestinal lesions. Thalidomide may reduce the severity and frequency of nosebleeds in some persons with HHT.
- Chronic bleeding often requires iron supplements and sometimes blood transfusions.
- There are four Canadian Specialized HHT Centres, in Toronto (www.hhttoronto.com), Montreal, Edmonton and Vancouver.
- About 10% of persons with HHT die prematurely or are disabled due to complications of their vascular malformations.

4 black tarry stool resulting from digestion of blood from upper gastrointestinal bleeding

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References and sources of more detailed information

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* Includes oral hygiene instruction, fitting a mouth guard, taking an impression, etc.

** Ontario Regulation 501/07 made under *Dental Hygiene Act, 1991*. Invasive dental hygiene procedures are scaling teeth and root planing, including curetting surrounding tissue.

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