MEDICAL NOSEBLEED MANAGEMENT: HEREDITARY HEMORRHAGIC TELANGIECTASIA – ASSOCIATED EPISTAXIS COPYRIGHT NOTICE

Medical Nosebleed Management
Hereditary Hemorrhagic Telangiectasia – Associated Epistaxis

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Hereditary Hemorrhagic Telangiectasia (HHT) is a genetic disorder of the blood vessels, which affects approximately 1 in 5,000 people. It affects males and females from all racial and ethnic groups. The disorder is also sometimes referred to as Osler-Weber-Rendu (OWR) after several doctors who studied HHT about 100 years ago. In 1896 Dr. Rendu first described HHT as a hereditary disorder involving nosebleeds and characteristic red spots that was distinctly different from hemophilia.

Before Dr. Rendu's work, doctors did not understand that individuals with what we now call HHT have abnormalities of their blood vessels, not a clotting problem in the blood itself. Drs. Weber and Osler reported on additional features of HHT in the early 1900s. More than one hundred years later, HHT is still often misdiagnosed in affected individuals and many doctors do not understand all of its manifestations.

The location of **telangiectases and /or AVMs** in the body determines what problem(s) someone with HHT might have. In most locations, and at any size, a telangiectasia or AVM has a greater tendency to rupture and bleed than a normal blood vessel. In the nose, skin, GI tract (stomach and intestines), and brain the primary problem they can cause is bleeding. AVMs in the lungs or liver are less likely to rupture and bleed, but can cause other problems that are less obvious to doctors and patients who lack an in depth understanding of HHT. No one with HHT has all of the signs and symptoms listed below.

Symptoms of HHT vary greatly, even within a family. A parent may have horrible nosebleeds, but no AVM in an internal organ. Yet, the child may have a nosebleed only rarely, but have AVMs in one or more internal organs. We cannot predict how likely someone is to have one of the hidden, internal AVMs based on how many nosebleeds or skin telangiectases they have. In other words, the person with HHT who has minor/mild nosebleeds is as likely to have an AVM in their lung, as the person with HHT who gets severe nosebleeds daily.

Epistaxis

Telangiectases in the nose, along with the **nosebleeds** they cause, are the most common symptom of HHT. About 95% of people with HHT have recurring nosebleeds by the time they reach middle age. The average age at which nosebleeds begin is 12, but they can begin as early as infancy, or as late as adulthood. Nosebleeds can be as infrequent as a couple per year or can occur daily. When a nosebleed occurs, it can last anywhere from a few seconds, up to several hours in some cases. The amount of blood lost may be a few drops, or enough to cause anemia (low blood count). As with most things that are variable in human beings (i.e. height and shoe size), the majority of people with HHT are in between the two extremes in terms of nosebleed frequency and severity.

For many, epistaxis is a major problem that progresses with age. At this time, there is no way to predict who will suffer nosebleeds and no way to prevent their occurrence and progression; therefore, the medical and surgical management is directed at controlling the bleeding. With good management, epistaxis should be kept to a minimum. Blood count, commonly measured as hematocrit, should be maintained at a reasonable level, preferably at a hematocrit greater than 36% and hemoglobin greater than 12%; transfusions should not be required. There is no single best way to manage every patient with HHT, so one must explore and utilize the different techniques and approaches and tailor these for the individual.

The following are the therapies that are most commonly:

1. One should maintain good nasal hygiene. This is done through pulsatile **nasal irrigation** with hypertonic saline. If pulsatile irrigation is not an option, laminar irrigation with any of the many available over-the-counter devices, such as the Netipot, is a second option.

2. Nasal moisture is important. Crusting can be managed with the application of Vaseline. A small amount of Vaseline is applied in each nostril. This is generally performed twice daily, but in times of difficulty, it can be performed more frequently. Anticoagulants, such as aspirin, make management more difficult. For those with medical problems requiring blood thinners, management becomes even more difficult.

3. Avoid Triggers. Anticoagulants, or “blood thinners”, cigarette smoke, environmental allergens, like dust, ragweed, animal dander, and nasal steroid sprays (e.g., Flonase, Beconase, Nasonex) can increase bleeding through one or more ways. Anticoagulants block the ability of the blood to form clots and stop bleeding. Cigarette smoke, environmental allergens, and nasal steroid sprays can dry out the nasal mucosa, which can lead to increase bleeding.

4. Sclerotherapy (Sclerosis). A procedure used to treat blood vessels or blood vessel malformations. A medicine (Sotradeco®l – sodium tetradecyl sulfate 3%) is injected into the vessels, which makes them shrink. Children and young adults - vascular or lymphatic malformations. Adults - varicose veins and hemorrhoids. The process of inflammation resolving by fibrosis takes more than 6 months.

5. The nasal mucosa is a hormone-modulated tissue. Estrogens support the nasal mucosa and decreases bleeding. Estrogen is available as an ointment, and one of the early treatments is to apply a small amount of Estrace® cream or NoBleed® at least once daily. If beneficial, one can apply it twice daily.

6. Anti-fibrinolytics. These drugs, like Amicar® (aminocaproic acid) and Lysteda® (tranexamic acid), are called anti-fibrinolytic drugs and are used for the treatment of HHT patients. These drugs are inhibitors of fibrinolysis and block plasmin formation and can stop bleeding rapidly if other parts of the hemostatisc system are working properly. Increased risk of blood clots should be considered before starting therapy.

7. Anti-Proliferatives. There is increasing evidence to support the use of certain anti- cancer drugs (Avastin® = Bevacizumab and Thalomid® = Thalidomide) to treat bleeding in people with HHT in whom routine medications and interventions have not been effective. Increased risk of blood clots should be considered before starting therapy.

4. Serial Cautery. In some patients, the HHT-associated epistaxis may progress and some form of sclerosing injection (Sotradecol) or cautery becomes necessary. At first, this can be managed with spot injection of Sotradecol or spot welding. The welding or coagulation can be performed with silver nitrate sticks, electric cautery, or even laser.

5. **Emergency Department.** If bleeding is profuse and doesn't stop, the HHT patient may elect to seek care from an Emergency Department physician. Because HHT is a relatively rare disease, the ED physician may not be familiar with the unique aspects of HHT-associated epistaxis. Unless the bleed is massive and prolonged, most nosebleeds will not require packing. The HHT patient usually coagulates adequately unless they are on an anti-coagulant or blood-thinner medicine. Blood transfusion may be required. **Nasal packing is to be avoided, except in the most extreme situations, as this will only lead to more bleeding once the pack is removed.** Only a well-moistened nasal pack (e.g., Rapid Rhino) should be inserted. Because of concern over bleeding, nasal packs are often removed in the operating room.

6. Patients with a deviated or crooked nasal septum may require septoplasty; this has benefit for breathing but is also required so the surgeon can gain easy access to the nasal cavity for repeat laser procedures. The septoplasty must be performed conservatively. It is important to preserve as much nasal septal cartilage as possible. One of the troublesome complications of epistaxis and repeat cautery is a nasal septal perforation. This occurs when the mucosa is cauterized on both sides. If the cautery and injury are full thickness, a hole may develop in the septum. This hole is called a septal perforation; while it is not impossible to manage, it increases crusting, nasal discomfort, and bleeding. Nasal septal perforations are typically managed with twice daily hypertonic saline nasal irrigation.

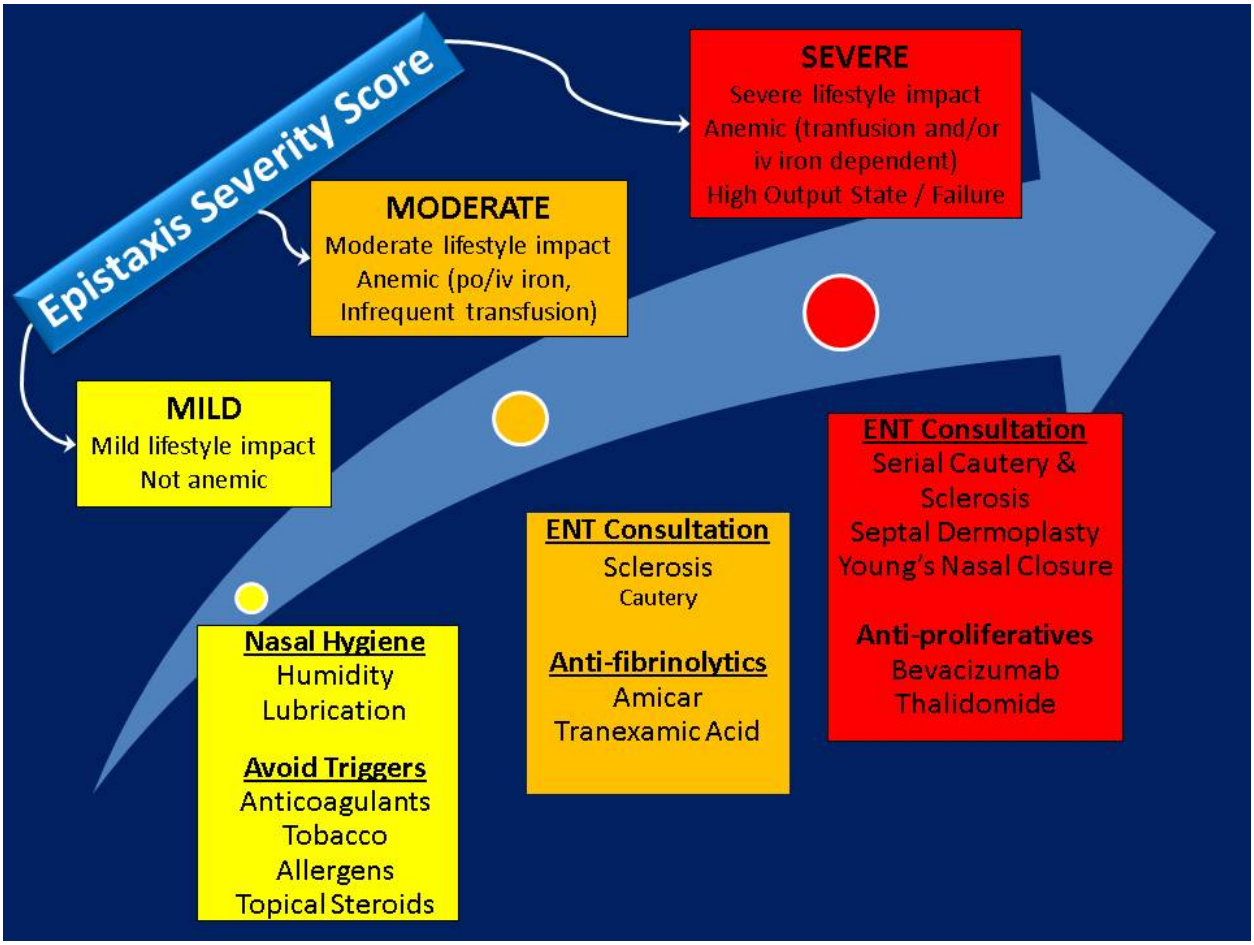

Figure. Standard Medical Management for HHT-Associated Epistaxis Based on Epistaxis Severity Score at Washington University.