FOR YOUR PHYSICIAN

HHT Guidelines for Management of Nosebleeds

Recurrent spontaneous nosebleeds (epistaxis) are the most common symptom of HHT and often lead to iron-deficiency anemia and reduced quality of life. Nosebleeds appear before the age of 20 in about 50% of people with HHT, with 90 % of all HHT patients developing it eventually.

Please review the checklist below with your physician to make sure your HHT is being treated properly.

HHT GUIDELINES RECOMMENDATIONS: EPISTAXIS	COMPLETED
The expert panel recommends that physicians advise patients with HHT-related epistaxis to use agents that humidify the nasal mucosa to prevent epistaxis. Level of evidence: III Strength of recommendation: Weak Agreement: 94%	❑ YES ❑ NO
The expert panel recommends that for HHT-related epistaxis requiring surgical intervention, clinicians consider endonasal coagulation as a first line treatment option Level of evidence: III Strength of recommendation: Weak Agreement: 93%	❑ YES ❑ NO
The expert panel recommends that clinicians refer HHT patients with epistaxis and who desire treatment to otorhinolaryngologists with HHT expertise for evaluation and treatment. Level of evidence: III Strength of recommendation: Weak Agreement: 87%	❑ YES ❑ NO
The expert panel recommends that when considering nasal surgery for reasons other than epistaxis, the patient and clinician obtain consultation from an otorhinolaryngologist with expertise in HHT-related epistaxis. Level of evidence: III Strength of recommendation: Weak Agreement: 93%	❑ YES ❑ NO
The expert panel recommends that the treatment for acute epistaxis requiring intervention include packing with material or products that have a low likelihood of causing re-bleeding with removal (e.g., lubricated low-pressure pneumatic packing). Level of evidence: III Strength of recommendation: Weak Agreement: 93%	❑ YES ❑ NO
The expert panel recommends that HHT-related epistaxis is not an absolute contraindication to anticoagulant/antiplatelet therapy. Anticoagulant/antiplatelet therapy can increase the risk of epistaxis and the decision to use these agents should be based on the individual patient risk and benefits. Level of evidence: III Strength of recommendation: Strong Agreement: 100%	❑ YES ❑ NO

FOR YOUR PHYSICIAN

International Guidelines for the Diagnosis and Management of Hereditary Hemorrhagic Telangiectasia

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Abstract

HHT is an autosomal dominant disease with an estimated prevalence of at least 1/5000 which can frequently be complicated by the presence of clinically significant arteriovenous malformations in the brain, lung, gastrointestinal tract and liver. HHT is under-diagnosed and families may be unaware of the available screening and treatment, leading to unnecessary stroke and life-threatening hemorrhage in children and adults. The goal of this international HHT guidelines process was to develop evidence-informed consensus guidelines regarding the diagnosis of HHT and the prevention of HHT-related complications and treatment of symptomatic disease. The overall guidelines process was developed using the AGREE framework, using a systematic search strategy and literature retrieval with incorporation of expert evidence in a structured consensus process where published literature was lacking. The Guidelines Working Group included experts (clinical and genetic) from eleven countries, in all aspects of HHT, guidelines methodologists, health care workers, health care administrators, HHT clinic staff, medical trainees, patient advocacy representatives and patients with HHT. The Working Group determined clinically relevant questions during the pre-conference process. The literature search was conducted using the OVID MEDLINE database, from 1966 to October 2006. The Working Group subsequently convened at the Guidelines Conference to partake in a structured consensus process using the evidence tables generated from the systematic searches. The outcome of the conference was the generation of 33 recommendations for the diagnosis and management of HHT, with at least 80% agreement amongst the expert panel for 30 of the 33 recommendations.

Open Access Link to International HHT Guidelines:
http://jmg.bmj.com/content/early/2009/06/29/jmg.2009.069013.long

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The HHT Foundation International, Inc. was formed to aid and support families with the genetic disorder Hereditary Hemorrhagic Telangiectasia (Osler-Weber-Rendu Syndrome).