PHYSICIAN INFORMATION

Diagnosis and Management of Pulmonary AVMs

AVMs, short for arteriovenous malformations, are direct artery to vein connections in the lung circulation. At least 30% of people with HHT have pulmonary AVMs. People are often unaware that they have pulmonary AVMs until they develop a life-threatening complication, such as stroke, brain abscess or lung hemorrhage. With the right screening and treatment, these life-threatening complications can be prevented.

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

HHT GUIDELINES RECOMMENDATION	COMPLETED
Screen all patients with possible or confirmed HHT for PAVMs Level of Evidence: III GRADE[1] Strength of Recommendation: Strong Expert Agreement: 96%	❑ YES ❑ NO
Use transthoracic contrast echocardiography as initial screening test for PAVMs Level of evidence: II GRADE Strength of Recommendation: Weak Expert Agreement: 96%	❑ YES ❑ NO
Treat PAVMs with transcatheter embolotherapy Level of evidence: II GRADE Strength of Recommendation: Strong Expert Agreement: 96%	❑ YES ❑ NO
Provide long-term advice to patients with documented PAVMs (treated or untreated): • Antibiotic prophylaxis for procedures with risk of bacteremia • When IV access is in place, take extra care to avoid IV air • Avoidance of SCUBA diving Level of evidence: III GRADE Strenth of Recommendation: Weak Expert Agreement: 87%	❑ YES ❑ NO
Provide long-term follow-up for patients who have PAVMs, in order to detect growth of untreated PAVMs and also reperfusion of treated AVMs Level of evidence: II GRADE Strength of Recommendation: Strong Expert Agreement: 100%	❑ YES ❑ NO

1 Schunemann HJ, Jaeschke R, Cook DJ, Bria WF, El-Solh AA, Ernst A, Fahy BF,Gould MK, Horan KL, Krishnan JA, Manthous CA, Maurer JR, McNicholas WT, Oxman AD, Rubenfeld G, Turino GM, Guyatt G. An official ATS statement: grading the quality of evidence and strength of recommendations in ATS guidelines and recommendations. Am J Respir Crit Care Med 2006: 174(5): 605-614.

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 Clinical Guidelines:
http://jmg.bmj.com/content/early/2009/06/29/jmg.2009.069013.long