Living with HHT: Understanding and Managing Your Hereditary Hemorrhagic Telangiectasia

Sara Palmer PhD
Price: $29.95
160 Pages
ISBN-10: 1421423901
Johns Hopkins University Press

It was a big task when author Sara Palmer PhD set out to write a book entitled "Living with HHT". Hereditary Hemorrhagic Telangiectasia (HHT) is a complex, multisystem disorder that presents in many disparate ways that are not intuitive or easily understood by many physicians. The hemorrhagic risks and consequences related to the vascular lesions that characterize the disorder (arteriovenous malformations as well as telangiectases) are well recognized but tend to be overestimated in terms of morbidity and mortality. That is particularly the case with regards to the relative significance of bleeding in the gastrointestinal tract. On the other hand, physicians are frequently unaware or under appreciative of the risks associated with the effects of vascular shunting per se that results from solid organ arteriovenous malformations - particularly of the lung and liver.

HHT, like most dominant genetic disorders, is extraordinarily variable. There are about as many "forms" of HHT as there are patients living with it. The clinical spectrum of HHT can include the patient with a three-drop-of blood-nosebleed every month and a few oral/dermal telangiectases unnoticed by a patient to the patient with epistaxis causing iron deficiency anemia and right heart failure at 40 years of age due to shunting secondary to hepatic vascular malformations.

Dr. Palmer is a psychologist who herself has HHT. The book excels at explaining for an affected patient, what HHT is, how it is diagnosed, and the overall medical management considerations. In a relative sense, it sometimes falters when detailing complex genetics concepts or particular interventions. However, the message comes across that HHT is largely a manageable disorder with the most severe complications being preventable with appropriate life-long surveillance. The book accomplishes the author's stated purpose of bringing together all of the basic information an HHT that a patient or family member needs to understand including how HHT can affect the body, what to do about various symptoms, and where to get treatment. Equally important, it addresses the emotional impacts for an individual and families living with this particular chronic, hereditary disorder.

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