



## **Hemophilia and Aging**

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### **AGING**

Life expectancy for persons with hemophilia has increased significantly from 11.4 years in 1920 to a potentially normal life span today. The Summary Report from the Centers for Disease Control and Prevention (CDC) Universal Data Collection (UDC) Activity updated February 18, 2011, reports that there are 18,270 participants in the U.S. with hemophilia. [1] Of those participants, 2,278 or 13% are aged 45-64 years, and 530 or 3% are aged 65 years and older. Due to improved hemophilia care and elimination of blood-borne pathogen transmission via clotting factor concentrates, we expect the population of persons with hemophilia over the age of 65 to grow significantly in the coming decade. We must prepare ourselves to care for this unique population.

With advances in medical care and chronic disease management, newer definitions of successful aging have been proposed. Young et al. define successful aging as *“A state wherein an individual is able to invoke adaptive psychological and social mechanisms to compensate for physiological limitations to achieve a sense of well-being, high self-assessed quality of life, and a sense of personal fulfillment even in the context of illness and disability.”* [2] The Hemophilia Treatment Center (HTC) may assist patients in successful aging through assessment of their developmental progression; assessment and prevention of co-morbidities and functional impairments; assessment of cognitive and emotional function; identification of depression and referral for treatment; and reinforcement of social connections while providing opportunities for social interaction through appropriate HTC programming.

### **DEVELOPMENTAL PROGRESSION**

Developmental stages of the aging adult were identified by Erickson as Generativity versus Stagnation and Ego Integrity versus Despair (psychpage.com). [3] Generativity versus Stagnation typically involves the middle-aged adult (25-64 years). Successful progression in this developmental stage leads to a connection to future generations, while struggles may involve a failure to feel a sense of meaning or overextension and lack of self-care. Ego Integrity versus Despair, the final stage of development for adults over 65 years, involves facing our evolution through adulthood and accepting our successes and failures as well as our end of life. Lack of successful progression through these developmental stages will contribute to symptoms of depression and general decrease in quality of life.

## **ASSESSMENT AND PREVENTION OF CO-MORBIDITIES AND FUNCTIONAL IMPAIRMENTS**

As life spans normalize with adequate hemophilia care, people with inherited bleeding disorders are now transitioning from a focus primarily on bleeding and its complications to a focus that includes health promotion and disease prevention. As HTC staff, we are frequently the providers with the most contact and the best ability to educate our patients regarding their general health and well-being. While the primary care provider maintains the responsibility of general health management, HTC staff may serve a vital role in reinforcing the need for our patients to receive age-appropriate screening similar to their healthy peers.

Research shows that people with hemophilia are living longer and are at equal risk as age-related peers for developing complications of smoking, overweight, and insufficient exercise. Hemophilia Data Set (HDS) 2008 reported 16,298 active patients 18 years and older, out of a total of 30,912 patients seen, who were in contact with a Hemophilia Treatment Center (HTC) within the year. [4] Of the total number, 25% did not identify a Primary Care Provider (PCP). Mortality data from April 1997-July 2007 HDS reports show 8.9% of deaths to be cardiac-related, 6.7% due to cancer, 5.8% due to accident/trauma, and 1.6% due to suicide, all amenable to prevention education. [5]

The U. S. Preventive Services Task Force published an updated Guide to Clinical Preventive Services, 2010-2011. [6] Table 1 briefly summarizes current recommendations for screening and testing for healthy individuals without symptoms or significant family history of certain disorders. These recommendations may also be applicable to the aging patient with an inherited bleeding disorder

**See Table 1. The U. S. Preventive Services Task Force Updated Guide to Clinical Preventive Services, 2010-2011.**

Because adults with bleeding disorders have an increased risk of bleeding complications with some health promotion/health screening procedures, it is important to discuss potential procedures with patients during their annual comprehensive visit. Breast, cervical, and colon cancer screenings are sometimes followed by biopsies. Patients may need pretreatment with factor concentrate for these invasive procedures; at the least, they require information about how and when to contact their HTC for treatment recommendations.

### **HEALTH RISKS**

#### **Alcohol misuse/abuse**

Risky drinking has been defined as more than 7 drinks per week or 3 drinks per occasion for women and more than 14 drinks per week or 4 drinks per occasion for men. Additional health risks from alcohol misuse/abuse may occur in patients with liver disease or Hepatitis C. Alcohol misuse/abuse may be reduced through screening and counseling.

## **Smoking**

Tobacco cessation programs are available in most communities and online. A list of resources may be provided during yearly comprehensive visits to those adults using tobacco products.

## **Hypertension**

Hypertension is defined as a persistent elevation of systolic blood pressure over 140 mm Hg or diastolic blood pressure over 90 mm Hg. Hypertension may increase the risk of intracranial hemorrhage in persons with bleeding disorders.

## **Overweight/obesity**

Persons with a body mass index (BMI) between 25 and 29.9 are overweight, and those with a BMI of 30 and above are obese. The UDC found 34.1% of persons with bleeding disorders to be overweight and 22.7% to be obese, compared with age-matched controls. [7] Obesity in persons with hemophilia contributes to poor joint outcomes and impaired healing after orthopedic surgery.

## **Hyperlipidemia**

The National Cholesterol Education Program guidelines describe elevated Cholesterol as greater than 160 mg/dl and a high Total Cholesterol as greater than 240 mg/dl. There are many considerations involved in determining appropriate treatment of hyperlipidemia. There are several oral agents such as statins that, combined with dietary changes, can lower cholesterol to acceptable levels and decrease the risk of cardiovascular disease.

## **Arthropathy**

Patients over the age of 65 often have hemophilic arthropathy in multiple joints. Joint arthropathy is associated with reduced bone mineral density [8]. Functional impairments related to obesity, joint arthropathy, neurologic impairment, or other consequences of hemophilia may lead to increased risk of falls, fractures, bleeding episodes, and other injuries. Assessment for sensory changes, balance, mobility changes, and circulatory impairment will lead to practical recommendations to ensure healthy aging and longevity. Surgical management, i.e. joint replacement, for this age population must include assessment of functional challenges as well as the added risks of thromboembolic disease due to immobility and frequent factor administration and inhibitor formation in patients who have previously had limited exposure to factor concentrates. The special needs of this population, i.e. factor administration, often make it difficult to obtain care in a skilled nursing facility for post-operative management.

## **Importance of regular physical activity**

Regular physical activity helps prevent cardiovascular disease, hypertension, type 2 diabetes, obesity, cancer, and osteoporosis. Screening for hypertension, lipid disorders, and obesity at each visit; education regarding healthy eating and exercise habits; and referral for treatment may be provided during a comprehensive visit to help improve the health outcomes of the aging hemophilic population.

## **ASSESSMENT OF COGNITIVE AND EMOTIONAL FUNCTION**

People with hemophilia have the same risk for cognitive or emotional impairment as the general population. HTC's need to be vigilant for cognitive and emotional dysfunction and assess how these dysfunctions may affect patients' ability to continue with routine hemophilia care such as home infusion. Screening tools that are validated and commonly used in primary care offices may be easily added to the HTC assessment of the geriatric patient. Tools such as the PHQ9 are effective in screening for depression. The Mini-Mental State Examination tests a person's attention span, counting skills, memory, and ability to problem-solve. A system to assure accurate diagnosis, treatment, and follow-up of cognitive or emotional dysfunction must be in place before screening is initiated. Hemophilic co-morbidities must be taken into account when accessing for cognitive dysfunction and depression. For example, narcotics, acetaminophen, and NSAIDS used for pain control of chronic hemophilic arthropathy may contribute to cognitive impairment, and Hepatitis C treatment may contribute to symptoms of depression.

## **REINFORCING SOCIAL CONNECTIONS**

According to a study by Iannone et al., the presence of social support provides an 80% decrease in the risk for depression, and employment provides an 84% decrease in the risk for depression in adults with hemophilia. [9] HTC's, in coordination with their National Hemophilia Foundation chapter partners, may provide opportunities for social interaction and promote involvement in local community activities. Providing treatment recommendations in coordination with employment obligations may assist in maintaining employment. Prophylaxis may contribute to good work attendance and decreased morbidity.

## **CONCLUSION**

The biggest challenge we as healthcare providers of this aging population face is the assessment, prevention, and management of the unknown. Our patients are having fewer hemophilia-related complications, more aging-related morbidities, and an almost normal lifespan. There is insufficient research to predict risk of thromboembolic disease, risk of cardiovascular disease, risk of cancer treatment, issues of dementia, and appropriate management with concentrated clotting factor for various bleeding episodes and surgical interventions. However, ongoing vigilance regarding safety and coordination with our peers regarding unique situations will continue to promote healthy aging and longevity in this at-risk population.

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**Table 1. THE U. S. PREVENTIVE SERVICES TASK FORCE UPDATED GUIDE TO CLINICAL PREVENTIVE SERVICES, 2010-2011. [6]**

<b>Disease State</b>	<b>Population Targeted</b>	<b>Recommendation</b>
<i>*** Recommendations for asymptomatic, healthy adults.</i>		
Abdominal Aortic Aneurysm	Men 65-75 years who were smokers	Ultrasound
Breast Cancer	Women 50-74	Mammography every two years
Cervical Cancer	Women who have a cervix and are sexually active	Cytologic screening (PAP smear) every 3 years
Colorectal Cancer	Men and Women 50-75 yrs	Screening using Fecal occult blood testing yearly, sigmoidoscopy, or colonoscopy every 5-10 years
Cardiovascular Disease	Men 45-79	Aspirin prophylaxis <b><i>IF benefit of reduction in MI outweighs risk of harm from GI hemorrhage</i></b>
Hypertension BP > 140/90 sustained	Adults 18 and older	Screening blood pressure every visit
Lipid Disorders	Men 35 and older; men 20-35 yrs and women 45 and older who are at increased risk for coronary heart disease	Screening for elevations in total cholesterol, LDL-C, or triglycerides (TG), or deficiencies of HDL-C
Alcohol Misuse	All Adults	Screen and provide behavioral counseling for those in need.
Depression	All Adults	Screen ONLY if staff available for accurate diagnosis, effective treatment and follow up.
Tobacco use	All Adults	Screen for use and provide cessation interventions
Obesity (BMI >30)	All Adults	Screen and offer counseling and behavioral interventions to promote sustained weight loss
Type 2 Diabetes	Adults with blood pressure > 135/80	Screen for Fasting Blood Sugar (FBS)>125.