



Stage 5:

Adulthood (Over 18 Years Old)



Stage 5: Adulthood (Over 18 Years Old)

This section is dedicated to a unique group in the hemophilia community. Many of you have seen hemophilia treatment advance from the days of no factor to the possibility of hemophilia gene therapy. Many of you may have been told you would not live beyond your teens. Yours is a generation that has seen heartbreaking lows and inspiring highs — from losing many to HIV or hepatitis C to the freedom and independence brought about by home infusion.

You have a unique perspective and have overcome many of the constraints hemophilia placed on the community, but you can now pass on skills and wisdom to the younger generation.

Adulthood is a time of great change, where major life decisions are being made. It is a challenging time, as well as an opportunity for personal growth and development. Adulthood is a time to keep learning. Adulthood also encompasses several stages. Some of you may barely be of legal age and embarking on your quest for independence, some of you may be involved in careers and raising families, while others of you may be approaching the retirement years or be retired for a good number of years already.



Physical development

As an adult, it is now up to you to maintain a healthy body. The physical changes you go through are universal for most people, regardless of a diagnosis of hemophilia. You can still enjoy a healthy lifestyle through the years by following a prescribed moderate and regular exercise program, paying close attention to diet and getting regular medical care. The following age-specific characteristics are intended as a guide.

Twenties

You will be at the peak of your physical ability. Actions you take now can either help or haunt you as you age. If you were not active in your teen years, now is the time to learn to strengthen your muscles and joints. Check with your hematologist and a physical therapist, if you have one, who can suggest a sport that fits your abilities. Suggestions may include swimming, tennis, golf, weight training, cardio exercises, personal training or walking.

Thirties

You may find some physical activities require more stamina. Your nutritional needs may change, and you may also start to develop a “spare tire” around the middle. Your body might be adjusting to a slower metabolism, which means your food intake needs to be decreased and your daily exercise increased. Joint problems that began when you were younger might now become severe enough to require surgery or even joint replacement.

Forties

You may notice new aches and pains, along with the regular ones you know from your bleeding disorder. These new discomforts may be related to the aging process rather than to your bleeding disorder. You may need additional medications for other conditions, such as type 2 diabetes, high blood pressure or high cholesterol. Vision changes are a common development at this age, and you may need bifocals, trifocals or reading glasses.

Fifties and up

As you continue to age, you may experience more physical changes:

- Decreased skeletal height
- Decreased bone mass and density
- Gradual slowing of bodily functions
- Development of degenerative changes, such as arthritis
- Slower reflexes
- Decreased physical endurance
- Loss of skin elasticity, dry skin and increased appearance of wrinkles
- Receding hairline
- Progressive hearing loss
- Decreased balance and coordination

Talk with your healthcare providers to find ways that may help address some of these changes.

Sixties to eighties

Individuals with hemophilia are living longer, and children can look forward to a normal life expectancy.²⁵

- Wear hearing devices if necessary to enable understanding of medical instructions and to help with social interactions
- Glasses should be worn as prescribed, especially when driving or moving about
- Balance and coordination may decline, so wear glasses, good shoes and use assistive devices if necessary
- Decrease or abstain from use of alcoholic beverages; kidney function declines as one ages and, because alcohol remains in the body longer, its lingering effects can cause falls
- Joints may be even less mobile, so take care with walking and use assistive devices if necessary or ordered by your physician

Nutrition^{13,14}

Keeping your weight in the proper range is especially important for people with bleeding disorders. Extra weight adds stress on joints, which can increase joint pain and frequency of joint bleeds, makes it harder to find veins for infusion, and requires more factor per infusion, since doses are based on weight.

While genetics can be part of the problem, personal choices also affect one's weight. Activity combined with healthy eating can help maintain appropriate weight.

Here are some helpful tips to get on the path to a healthy lifestyle:

- Eat as a family. You can be a good role model for your family's eating habits.
- Don't eat meals or snacks in front of the TV.
- Read the nutritional label. Ingredients are listed in order of proportion. The label also shows calories and serving size.
- Avoid foods that contain high percentages of sugar, fats, sodium and cholesterol.
- Eat from all food groups every day for a well-balanced diet, and eat more vegetables, fruits and lean meats. Limit sweets, pastries and other foods that are high in calories and low in nutrition.
- Avoid fried foods. Instead, bake or grill meats, and steam or grill vegetables.
- Replace vegetable oil with olive oil.
- Opt for nutritious snacks. Some examples include fresh and dried fruit, vegetables, pretzels and fat-free cookies.
- Drink plenty of water. Choose water, milk or calorie-free beverages instead of soft drinks.
- Don't skip meals; instead, eat smaller meals and healthy snacks.

Orthopedic issues²⁶

Despite advances in medical management of bleeding disorders, target joints can progress to advanced arthropathy (joint disease). This is most commonly seen in people with severe hemophilia. Over time, changes occur in the joint that do not resolve with usual factor infusions. The synovium, the tissue that lines the joint and produces a lubricating fluid, remains thickened.

The joint feels spongy to the touch, may have a limited range of motion and may be painful. Some ways to manage joint disease include:

- Secondary prophylaxis, exercise, bracing or splinting
- Physical therapy
- Synovectomy (surgical or chemical removal of the synovium)
- Hyaluronic acid injection into the joint

The goal is to maximize function by reducing pain and improving range of motion, strength and endurance, as well as joint alignment and stability. Surgical intervention may be necessary to maximize function. A common procedure for chronic arthropathy is total joint replacement. Surgery is coordinated with the care team to manage factor therapy before and after the procedure. Postsurgical rehabilitation protocols will vary by the type of procedure.

Talk to your hematologist about the best plan for you.

Dental care^{5,23}

Regular dental checkups every six months are crucial, but some adults with bleeding disorders may have had difficulty finding a dentist who was willing to treat someone with hemophilia. As a result, you may require major dental work, such as deep cleaning, scaling, root canals, extractions or periodontal surgery. Your dentist should always contact your hematologist prior to scheduled procedures to evaluate if any special pretreatment or posttreatment with factor or other medications is needed.

Your specialty pharmacy can work with your hematologist to ensure you have the right dosing for dental work depending on the procedure.

You need to give your dentist the following information:

- Type and severity of bleeding disorder
- Factor product(s) prescribed
- If you have a venous access device (port)
- If you've had a joint replacement
- If you have a coinfection
- If you have an inhibitor

Mouth bleeds can take a long time to heal because there are so many blood vessels present. General tips to follow include eating soft food at room temperature and avoiding things that can dislodge a clot like use of straws, spitting and smoking. Your hematologist may recommend the use of Amicar (epsilon aminocaproic acid), which can be taken orally and used topically. It neutralizes the enzymes in saliva that often prevent a clot from forming or break it down once it has formed in the mouth. Amicar is available by prescription only, and you should consult with your hematologist for a prescription.

If you wear partial or complete dentures, make sure they fit well. Stress on the gums or jaw from improperly fitted dentures can lead to bleeding. Plan an annual visit so any adjustments can be made if the dentures become loose. Be sure to clean your dentures regularly. Remember: It only takes a few minutes a day to keep your teeth healthy by regular brushing and flossing. Also, it is never too late to take good care of your teeth.



Safety issues²⁷

Some of the leading causes of accidents in the home are slips, trips and falls. These types of accidents can cause serious injuries, and, for people with a bleeding disorder, a fall could result in a bleed. Make safety a family affair. Take time to inspect your home for hazards that could lead to falls:

- Keep walkways clear and free of clutter. Don't store boxes, toys or other obstacles on the stairs or in hallways. Don't run electrical cords under rugs or across walkways.
- Ensure your home is well-lit, especially stairs and landings. What you don't see can hurt you.
- Install and check the handrails on all stairs. Be sure they can support an adult's full weight.
- Wipe up spills immediately, and keep floors dry. A small slick spot can send you flying.
- Install nonslip treads on stairs, as well as in the tub and on the bathroom floor. Water and soap can make surfaces especially slippery.
- Install a grab bar to make it easier to get in and out of the tub, especially for elderly family members.

Psychosocial issues

Just as your body will undergo many physical changes during adulthood, you may experience many emotional, social and psychological changes as you mature.

Depending on what stage of adulthood you are in, you may be finishing your education, initiating a career, finding a mate, developing relationships, getting married, establishing a family or retiring. You will progress from being dependent on your family to being responsible for yourself. You may even become responsible for others, such as children or aging parents.

Dealing with these life stages can be difficult, and having a bleeding disorder can add to complications. Other stressors, such as hepatitis or other medical conditions, job changes, finding and keeping insurance coverage or family and marital responsibilities, affect not only you but also those close to you. One advantage you have is that you are probably well aware of different



aspects of your bleeding disorder by now. You should be able to infuse, calculate your dosage, order and maintain your factor inventory, fill out your treatment log and be familiar with your bleeding patterns. The more you know about your bleeding disorder by keeping accurate records of your bleeds, the better you will be able to manage bleeds, thus reducing stress.

How adults with hemophilia react to their bleeding disorder is as varied as each individual. The following reactions are not uncommon, and your viewpoint may change over time:

- Denying having a bleeding disorder (to others)
- Becoming self-absorbed, feeling the bleeding disorder rules your life
- Feeling embarrassed or ashamed of your bleeding disorder, or feeling like less than a “whole” person
- Becoming depressed about having a chronic condition
- Feeling isolated, alone, overwhelmed or misunderstood

Some may view seeking help for these scenarios as a sign of weakness, but this line of thinking can lead to poor self-care. There are several avenues of help available to people with hemophilia. Hemophilia chapters and support groups can give people with bleeding disorders a chance to meet and share experiences.



Disclosure^{28,29}

Disclosure is another issue you will face as an adult. Who to tell and when, or how to tell other people about your bleeding disorder, will be major issues for you to decide. This will occur on both personal and professional levels.

Personally, you may be concerned about the impact your bleeding disorder can have on establishing and maintaining relationships. You may question what others think and how you are perceived. You may be more aware of how others accept you in social situations, depending on changes they notice to your health status or mobility. Not being able to keep up physically, the need to always have medications with you, and struggling to find people who will not dismiss you can negatively influence how you feel. Surround yourself with people who support who you are.

Entering the work world will add major concerns regarding disclosure. When interviewing for a job, you may question if telling a potential new employer about your medical condition might keep you from being hired. Once hired, you may wonder if having a bleeding disorder will interfere with opportunities for advancement. Many fear repercussions at their school or workplace due to absenteeism related to a bleeding disorder. Many fear their employer might terminate them due to high usage of company insurance. Federal mandates help protect a person's medical status in the workplace. It is important to keep current with rules and

laws that will protect you from discrimination. Ask to be placed on national and local organizations' mailing lists. Keep in touch with others in your situation by participating in support groups or attending chapter meetings or educational seminars, so you'll be better informed about the latest developments.

Those with coinfections, such as hepatitis or HIV, face multiple challenges as they manage complex treatment regimens. They may also experience fatigue, depression, appetite loss, gastrointestinal problems or sleep disorders that interfere with their work schedule. Unlike school, when absences could be handled with home tutoring, employers are typically not as lenient with repeated absenteeism. This could become a major issue in keeping your job or advancing your career.

The Americans with Disabilities Act (ADA) is a federal law that protects people with disabilities from discrimination. The ADA defines disability as having an impairment that substantially limits "a major life activity." The law applies in various settings, including employment, transportation, education and business services. The law does not apply to employers who have fewer than 15 employees.

Although you may be hesitant about letting others know you have hemophilia, wearing medical identification jewelry (bracelet or necklace) alerts medical professionals to your condition in case of an emergency. It can save your life!

Insurance

Getting and keeping insurance coverage is a major concern for a person with hemophilia. The best time to research insurance options is before you need it. As healthcare coverage regulations change, be sure to stay current on you and your family members' coverage rights. Private insurance can be expensive and may be difficult to obtain. Though complex, you can navigate the insurance maze if you educate yourself on options.

The Affordable Care Act allows young adults to stay on their parents' healthcare plan until age 26. Plans and issuers that offer dependent coverage must offer coverage to enrollees' adult children until age 26, even if the young adult no longer lives with his or her parents, is not a dependent on a parent's tax return, or is no longer a student.²⁴

If you obtain employment with a large corporation or government agency, you will most likely be eligible for coverage because of the large pool of people in their group policy. Smaller companies have different federal regulations. They may enroll you in their group plan but find that the premium for their group coverage may rise once the reports for your medical bills start coming in. You may also be eligible for coverage through a spouse's insurance plan.



Various programs can help you and your family, whether you are still able to work or can no longer work due to a disability. The rules governing eligibility for these programs are often complex and subject to change. Descriptions of some different insurance options and plans are listed below:

Social Security Disability (SSD): This benefit is for younger workers (under retirement age) who become disabled and meet certain criteria. SSD functions like an insurance plan, where you must have contributed by paying Social Security payroll taxes (FICA) over a period of time to meet insured status requirements.

Supplemental Security Income (SSI): This is a combined state and federal program for individuals with limited income and resources who are over 65, blind or disabled. SSI provides a monthly benefit payment and, in most states, includes medical coverage through Medicaid. Beneficiaries may also be eligible for food stamps and other forms of assistance for low-income households.

COBRA: Employers with more than 20 employees who sponsor group health plans must offer employees, retirees and their families the opportunity to temporarily extend health coverage at group rates when coverage under the plan would ordinarily end. The premium for this coverage is totally paid for by you. The 18-month coverage period may be extended in certain circumstances.

HIPAA: The Health Insurance Portability and Accountability Act (HIPAA) provides that individuals eligible for group health plan coverage cannot be denied coverage or have their coverage terminated based on their health status or any preexisting conditions. HIPAA also ensures that you are not charged a premium greater than that charged to a similarly situated individual under the same plan. Due to HIPAA guidelines, we ask a series of questions when you contact us with questions or for a reorder to ensure we are following proper protocol.

Medicare: The federal government offers Medicare for people over 65 and, in some cases, for those who are disabled. You must meet specific requirements for inclusion in this government insurance program. It is not uncommon to be rejected at the first application for coverage.

Medicaid: This is another government program administered by your state of residence. It is co-funded by the federal and state governments but administered by state agencies. This program requires that you have a low income or be dependent on other agencies serving low-income or poor individuals. Your Customer Relations Specialist (CRS) can help direct you to state-specific information about eligibility.

Factor assistance programs: Most companies that manufacture clotting factor have assistance programs. Some companies give certificates that you earn for each month of purchase or for a certain amount of medication used. The certificates can be redeemed for factor when you no longer have third-party insurance. These programs can help fill the gap while you search for other coverage options. Others have compassionate care programs that have few, if any, requirements.

Social services: Your local county government usually offers some help through its assistance programs (often called Department of Human Services or Social Services). It can advise you of other medical assistance programs available in your area.





Pain management^{30,31}

Adults with bleeding disorders can experience pain as a result of their condition and its treatment. You can significantly impact how pain affects you by understanding what pain is and how to deal with and avoid it.

What is pain?

Pain is how your body reacts to an injury or an illness. It is a sign that something is wrong. Pain can be protective — it's the body's way of saying, "Pay attention to where you hurt." Pain can have different sensations, such as dull, throbbing, sharp, intermittent, continuous or burning.

How to manage pain

Different people need different methods to resolve pain. This means some techniques will work for you, but others may not. Some methods you can do for yourself, while others require help from health professionals and other experts. Some methods include:

- **Medications:** Follow your hematologist's advice about how much and when to take your medication. Your prescription may have labels that warn you not take it on an empty stomach, for example. Take these warnings seriously. Follow both your hematologist's and pharmacist's advice when you consider using

over-the-counter medication. If medication is ordered, record how well it works. How long were you comfortable after taking the medication? Does it make you sleepy? How many times did you take the medication? Does your pain seem better or worse? Do not take pain medication more often than it is prescribed.

- **Treat bleeds immediately:** Delayed treatment can result in chronic or ongoing pain in your teens and later years which may require surgery and/or narcotic prescriptions. Narcotic use can easily become habit-forming or addictive and is a very serious problem in the U.S., affecting individuals with hemophilia as well.
- **Exercise:** An exercise program designed by your hematologist or physical therapist can help you cope with pain. Exercise helps relieve stiffness and gives you an improved sense of well-being. Be sure to warm up first.
- **Heat or cold treatments:** Use of hot or cold treatments can reduce the pain and stiffness associated with joint damage. Cold packs numb the sore area and can reduce swelling. Heat treatments relax your muscles. Use either heat or cold for only 10–15 minutes at a time. Always put a towel between your skin and any type of hot or cold pack.
- **Relaxation/stress reduction:** Relaxing can help reverse the physical and emotional effects of pain. The best time to use relaxation skills to manage your pain is before the pain becomes too intense. Many people find the following techniques helpful: guided imagery, prayer, breathing exercises, hypnosis or relaxation audio tapes.
- **Keep a record:** Many people find it helps to keep a pain journal. You can record where you hurt, what the pain feels like, how long you had pain, steps you took to alleviate the pain and if these steps made you feel better or worse. Reviewing your journal will make it easier to see patterns and help you when you discuss your pain with your hematologist. You can even add this information to your infusion logs.
- **Take control:** Be an active participant in your pain management plan.

How to avoid pain

- Prevent pain by avoiding injuries. Use protective devices, such as helmets, knee and elbow pads or shin guards.
- Infuse preventive doses prior to activities as directed by your hematologist.
- Treat bleeding episodes immediately to prevent pain from increasing and to prevent chronic damage which can result in ongoing pain.

Remember: Do not take any medication that contains aspirin or nonsteroidal anti-inflammatory drugs (NSAID), such as ibuprofen, as it can cause bleeding. Be sure to check with your pharmacist and read the ingredients list on any medication to see if aspirin is included. It can also be listed as “acetylsalicylic acid” or “ASA.”¹¹

