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Living with Hemophilia:

Healthy Living Through the Stages of Life

Introduction

This section provides a brief introduction to the world of hemophilia. If you are not new to hemophilia, you may proceed to the appropriate section using the Table of Contents as your guide.

As an individual, parent or caregiver, hearing the word "hemophilia," a rare bleeding disorder, can cause many different emotions. Even if you have a history of hemophilia in your family, it can still be a shock to hear the diagnosis for you or your child. Anger, denial, and guilt are all normal reactions. Each family member will have his or her own reaction to this news and will process feelings differently.

As an adult, learning you have a bleeding disorder can cause anxiety about how your lifestyle may change. Everyone has a different way of coping with this news.

It is important to understand the disorder and the many resources available to you. Medical professionals are ready to help you and provide the support and guidance you need. Take advantage of this network of support and information — it can help make coping with this adjustment easier.

You may uncover many myths and misunderstandings associated with hemophilia that have been told and retold, often causing incorrect information to be taken as truth. One common misunderstanding is that people with hemophilia can cut themselves and bleed to death. People with hemophilia do not bleed faster than others; they just bleed longer. You may also hear other false statements, such as "you can catch hemophilia" or "a child will outgrow his hemophilia." These statements are simply not true. The following information will help you become an educated person or parent who can, in turn, educate your community to help dispel myths that can circulate when people are not well-informed. Advances in hemophilia care in the past few decades have improved the quality and quantity of life for those living with hemophilia. For parents and caregivers, you have the opportunity to create a supportive and loving environment to help foster physical, emotional and psychological development. As an individual living with hemophilia, you may find great strength and support in surrounding yourself with others familiar with a bleeding disorder in such ways as support groups.

By becoming a well-informed caregiver or individual living with hemophilia, you will have the information needed to safely manage the bleeding disorder through all life stages. As a parent or caregiver, learning what precautions to take as your child develops is a topic your hematologist, board-certified pediatrician or specialty pharmacy can discuss with you. Raising a child with hemophilia requires a delicate balance. Overprotective parents who shelter their child may not allow him or her to develop a healthy sense of self. Parents who provide little guidance or protection for their child risk unnecessary bleeds and setbacks. Parents should strive to help their child develop a normal sense of self-esteem in a physically-safe environment.

You many notice the use of pronouns like "he" or "him" throughout this book. We recognize that bleeding disorders, like hemophilia, are not unique to only males. While the majority of people with hemophilia A and hemophilia B are males, a very small percentage may be females. The information in this book pertains to properly caring for hemophilia and ways to maximize a healthy lifestyle that is not unique to only males with hemophilia.

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Hemophilia Overview

Hemophilia Overview

Hemophilia occurs when one of the clotting factor proteins needed for the blood to form clots is missing or reduced. There are at least 13 different proteins — referred to as clotting factors — in the blood that work together to create a fibrin clot when an injury occurs. Clotting proteins are usually designated by Roman numerals (I, II, III, IV, and so on). People who are missing, or have a low amount of, factor VIII (eight) have hemophilia A, the most common type of hemophilia (with von Willebrand disease being the most common bleeding disorder). It affects about 80 percent of people with hemophilia. People who are missing, or have a low amount of, factor IX (nine) have hemophilia B. Hemophilia B affects about 20 percent of individuals with hemophilia. The incidence (number of new cases) of hemophilia A is approximately 1 in 5,000 males born in the United States.[1] The prevalence (number of existing cases) of hemophilia A is about 16,000 people in the U.S. Hemophilia B affects more than 4,000 individuals.[2] All races and socioeconomic groups are equally affected.

While there are other forms of bleeding disorders like factor XIII (thirteen) deficiency and afibrinogenemia, this booklet will focus on 2 of the more-frequently occurring factor deficiencies of factor VIII and factor IX.

Hemophilia has different levels of severity. The amount of factor in your child's blood will determine the severity level. Severity levels do not change over time in a person with hemophilia.

Classification of severity of factor deficiency[1]

Classification	Factor VIII or factor IX level (compared to normal levels)	What to expect
Severe	Less than 1%	Bleeding after an injury, major trauma or surgery. May have bleeding without a known injury (spontaneous bleeding).
Moderate	1% to 5%	Bleeding after minor injury is possible and after major trauma or surgery. Occasionally spontaneous bleeding may occur and can be associated with prolonged or serious bleeding in a joint.
Mild	5% to 40%	Prolonged bleeding usually only after serious injury, major trauma or surgery.
Normal	50% to 150%	No abnormal bleeding.

How did this happen?

Hemophilia is passed on from parent to child as an inherited, or genetic, trait. About 30 percent of individuals with hemophilia have no family history of a bleeding disorder and develop hemophilia from a genetic mutation.[1]

> Hemophilia used to be called "the royal disease" since Queen Victoria of England was a hemophilia carrier.

A review of Queen Victoria's family tree shows how hemophilia A was passed through the female descendants, but the symptoms of the disease occurred in the males. How does that happen?

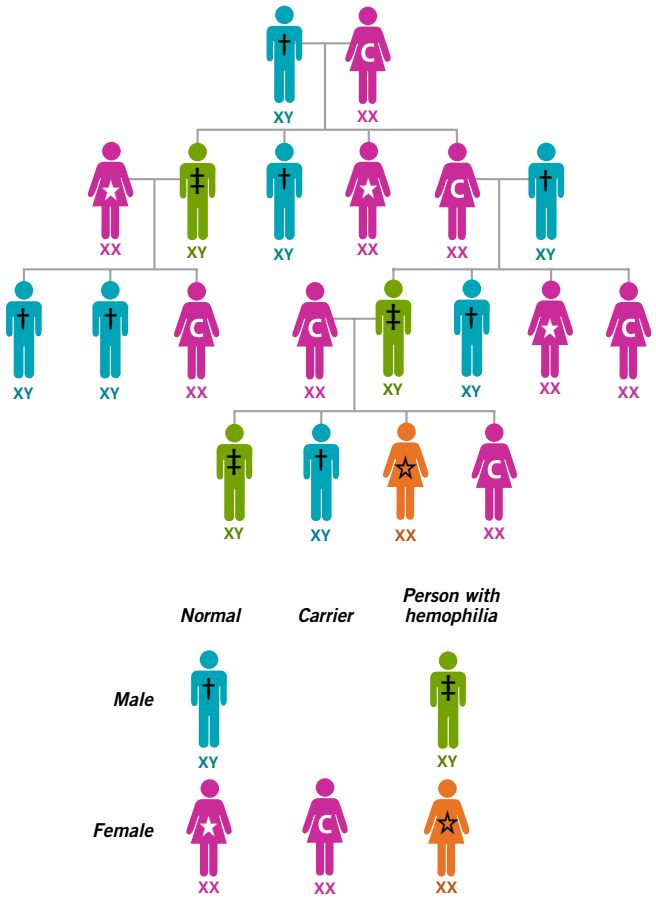
The gene for hemophilia is carried on the X chromosome. Women have two X chromosomes, and men have an X and a Y chromosome. A woman gets pregnant when her egg is fertilized by the man's sperm. The sperm will carry half of the genetic makeup from the father (including an X or a Y sex gene).

The mother's egg will carry half of the mother's genetic makeup and will have an X gene. If the father's X gene fertilizes the egg, the baby will be a girl (XX). If the father's Y gene fertilizes the egg, the baby will be a boy (XY).

For a woman who carries the gene for hemophilia, each pregnancy has four possible outcomes: a boy with hemophilia, a boy without hemophilia, a girl who is a carrier, or a girl who is not a carrier. Some carriers can have low levels of clotting factor. They are sometimes called "symptomatic carriers."

If a man has hemophilia, his daughters will be carriers and his sons will not have hemophilia. Your hematologist will be able to help you better understand this information. A genetic counselor can also help you learn your probability of having a child with hemophilia.

Inheritance of hemophilia gene



Why does an individual with hemophilia bleed?[3]

Nature has given our bodies the ability to stop bleeding at an injury site. There are three major steps in this process:

1. The blood vessel around the area of injury will constrict or become smaller to slow down the amount of blood flowing past the injury.
2. Platelets, which are part of our blood, travel to the injury and lay themselves down over the opening. These platelets stick to each other, forming a "platelet plug," which initially helps stop bleeding.
3. Proteins (clotting factors) in the blood create a substance called fibrin, which weaves through the platelets covering the injury and holds the platelet plug in place. This fibrin clot stops the bleeding until the blood vessel can repair itself.

In hemophilia, the proteins (clotting factors) don't produce enough fibrin. Without the fibrin to weave through the platelets and hold them in place, the platelets wash off the injured area, and the person continues to bleed.

Formation of inhibitors[4]

The immune system is the defense system that normally protects us from illnesses and microscopic invaders that pose harm to our health. As we are exposed to viruses, the body recognizes the illness and creates antibodies to fight against the illness and rid it from the body. Sometimes the immune system does too good of a job trying to protect us. If the body recognizes factor as a foreign invader, it can create antibodies against factor. These antibodies neutralize factor and make it ineffective for clotting or cause an allergic-type reaction. This is why people with inhibitors may need to infuse much more factor to overcome the body's immune response or different products to go around the immune response. About 30% of people with hemophilia A will develop an inhibitor while about 2%-3% of people with hemophilia B will have an inhibitor. If you develop an inhibitor, you and your hematologist will decide if your treatment regimen needs to change.

Recognizing Bleeds

Recognizing Bleeds[1,5]

Injuries manifest themselves in different ways, and some individuals are more likely to be injured than others. This is true for individuals with or without hemophilia. Bleeding episodes can be classified by the severity of the bleed, as well as where the bleed occurs. Sometimes, as with cuts, the blood is visible. Other times, the bleeding is internal. Usually, internal bleeds cause the most concern, especially head bleeds.

For parents or caregivers, it is normal to be concerned about your child's well-being and have a baseline concern for their safety. This baseline concern is often heightened in parents whose children have hemophilia. One of the most difficult challenges parents face is recognizing when their child is experiencing a bleed. This can be especially true before your child is able to tell you he hurts.

With infants, it can be difficult to know when your child is having a bleed. He cannot verbalize or tell you if it hurts, where it hurts, or how badly it hurts. The more parents can learn about their child's bleeding disorder, the better prepared a family will be to cope with the everyday experiences they face. The types of bleeds your child experiences will change as he grows and becomes more active. For babies and toddlers, the most common bleeding sites are the head and the mouth.

Teething can cause the baby some discomfort and may make the gums appear swollen or discolored. Do not apply over-the-counter medications to the baby's gums without first discussing this with your pediatrician and hematologist. If bleeding occurs, speak with your hematologist. A damp, soft baby washcloth kept in a clean plastic bag in the refrigerator or freezer can be helpful. Always remove the clean cold baby washcloth from the plastic bag before gently applying it to the gums. Do not rub the gums if there is bleeding or a clot. To avoid the possibility of choking, never leave the baby alone with the washcloth.

Regardless of life stage, bleeds can happen in any area of the body including joints, muscles, the abdomen or the head. Sometimes you may notice an incident that may cause a bleed, such as falling and a head bump. In children, parents or caregivers may notice their child may be fussy and not know why. In these cases, look for signs that a bleed is occurring:

- It hurts to use the affected body part. For example, if a knee joint is affected, your child may stop crawling. If your child is walking, he may revert to crawling or may begin limping.
- You may notice bruising.
- Swelling might be present. If in an extremity, compare the affected body part with the opposite side to determine the extent of the swelling.
- Your child may also be very irritable. This may happen with or without an explanation. Toddlers can be unpredictable. They may be crying because they can't find their favorite toy or because they are having a bleed.
- When your child starts to speak, he may describe bleeds in many ways. The most common description will likely be pain at the bleeding site.

Small cuts and scrapes

Small cuts and scrapes can happen frequently. Most will stop bleeding on their own. Use basic first aid principles to help stop the bleeding, including holding direct pressure on the cut and using ice packs.

Nosebleeds

Nosebleeds are quite common. Ask your hematologist how long you can wait before notifying the office. In the case of a nosebleed, sit with the head tilted slightly forward. For slightly heavier bleeding, allow the blood that may be in the mouth to run out into a cloth or towel or have the older child expectorate (spit) the blood into the sink to avoid swallowing/vomiting. Pinch the bridge of the nose using firm, continuous pressure for 20 minutes. Apply an ice pack to the back of the neck. Large amounts of blood or prolonged bleeding should be reported to the hematologist.

Joint bleeds[3]

Joint bleeds usually start in the lining of the joint, called the synovium. If not treated, bleeding can continue until the joint space is filled with blood. This can happen with or without trauma to the joint. The extra fluid in the joint space creates pressure, swelling, heat and pain. Over time, there could be damage to the joint. If a joint has three or more bleeds in six months, it is considered a "target joint." Prompt treatment is recommended. The sooner factor is infused, the more effective it will be. Follow up a factor infusion by using the RICE (rest, ice, compression, elevation) method (see page 13).

Muscle bleeds

Muscle bleeds can happen in a single muscle or a muscle group. They can happen spontaneously (with no obvious injury) or after receiving a blow or other trauma to the area. Prompt treatment is recommended, and you should consult your hematologist.

Head bleeds

Head bleeds can happen on the inside or the outside of the skull and should always be considered serious bleeds. Head bleeds can occur spontaneously or as the result of an injury. Symptoms can progress slowly and may not be noticed for days after an injury. Prompt treatment is required. Contact your hematologist for more instructions. These bleeds should be considered an emergency situation and a simple bump on the head should not be treated lightly. Visit www.hemophilia.com for materials to aid in a visit to an emergency room.

Early signs and symptoms of a head bleed:

- Irritability
- Nausea and vomiting
- Sensitivity to bright light
- Acting confused
- Being inconsolable

Later signs and symptoms of a head bleed:

- Inability to balance
- Dilated or unequal pupils
- Seizures
- Confusion
- Persistent headache

Neck bleeds

Neck bleeds are also very serious. Swelling can place pressure on the airway, making it difficult to breathe. Prompt treatment is recommended, and you should consult your hematologist. A neck bleed should be considered an emergency situation.

Mouth bleeds

Mouth bleeds can be frightening because the blood and saliva mixed together can make these bleeds look much worse than they might be. Most mouth bleeds are caused by falls or by sharp objects a baby may have placed in the mouth. For those caring for an individual with hemophilia, it is important to inspect the mouth and find the source of the bleeding. Position the individual on their side or stomach to prevent him or her from swallowing blood. Swallowed blood can upset the stomach and may cause nausea and vomiting.

Mouth bleeds will usually stop within a couple of hours. Direct pressure to the area or a cold pacifier for babies can help stop the bleeding. Frozen ice pops can also help stop bleeding for children AND adults. If this does not work to control the bleeding, contact your hematologist. He or she may recommend factor infusions or oral medication. Some medical professionals often prescribe Amicar® (epsilon aminocaproic acid) to help stop the saliva in the mouth from breaking down the clot when it forms.

In some cases, mouth bleeds are not just uncomfortable; they can be dangerous. Sharp corn chips and some types of potato chips can tear or scratch the soft tissues lining the mouth and esophagus when chewing and swallowing. This can cause bleeding that requires a factor infusion and Amicar as prescribed by the hematologist. Chewing gum may lead to accidental tongue and cheek bites. Bleeding that continues longer than several hours or starts and stops for 24 hours should be reported to your hematologist immediately. Ongoing bleeding in the mouth or throat that starts and stops is not controlled, and can be life-threatening if it continues.

Ask your hematologist for more information on dealing with mouth bleeds.

Blood tests and immunizations

Blood tests or immunizations are a potential cause of a bleed in infants in the first few months of life. If you know your child has hemophilia, it is best to consult with your hematologist before any treatment or testing is done. Even the simple drawing of blood from a vein for a routine test can cause a muscle bleed if a tourniquet placed on the arm is too tight. Bleeding from the needle site after a blood test or immunization can occur. Firm pressure is usually enough to stop this from occurring. Bringing a gel ice pack (not frozen) for the nurse to apply over a cloth or folded paper towel after an injection can help prevent bleeding and aid in comfort. While applying pressure to the area of the injection with your fingertips, gently place the cloth-covered gel pack over your fingers to minimize inflammation.

Some immunizations are usually given into the muscle (intramuscular or IM), and some are given under the skin (subcutaneous or Sub-Q). Most immunizations can be given Sub-Q rather than IM to avoid bleeding into the muscle.[6] Consult your hematologist beforehand to determine the best method for you or your child.

Bruises

Bruises are usually considered mild and not a cause for alarm. They first appear dark blue or purple in color and may have a bump in the middle or feel like there are knots. They will turn green and yellow over time as the body clears the blood that caused the bruise. They can take about two weeks to fade and are usually not painful. They do not need to be treated with factor unless your hematologist advises you to do so. You should call your physician immediately if bruises appear on or near the head, throat, joints or groin.

Other bleeds

Other bleeds such as eye, stomach, intestine, kidney, bladder, and hip or thigh muscle bleeds should also be considered serious. Prompt treatment is recommended, and you should consult your hematologist.

Treatment

Treatment[1,7]

Hemophilia is treatable. Missing factor can be replaced by infusion of clotting factor concentrates, elevating factor levels enough to allow the body to form a clot and stop the bleeding. Many parents learn how to infuse their child at home with clotting factor to prevent or treat bleeds. As a child matures, they will learn how to self-infuse and manage their infusions as they move into adulthood.

Clotting factor concentrate comes as a freeze-dried powder that is mixed with sterile water or a diluent (an agent to dilute medications) prior to infusion. It is injected into the body through a vein or a central venous access device. There are two forms of clotting factor concentrates, depending on the source from which it is made: plasma-derived or recombinant.

Plasma-derived factor

Plasma-derived factor is made from donated human blood plasma. Plasma contains clotting proteins. The clotting proteins are separated from other blood components, purified and made into a freeze-dried product. All plasma donated for production of clotting factor concentrates is tested for the presence of viruses.

Recombinant factor

Recombinant factor is made in a laboratory and does not use blood plasma. Because recombinant products contain few or no proteins from human blood, the risk of contracting viruses is minimal or absent. Recombinant factor VIII products were introduced in 1992, and recombinant factor IX products became available in 1997.

All factor products — both plasma-derived and recombinant — are treated for purification and to prevent infection.

Episodic treatment

Episodic treatment (also called on-demand or PRN) occurs when factor is given only when a bleeding episode occurs.

Prophylaxis

Prophylaxis is the practice of giving factor on a regularly scheduled basis (up to several times a week). The goal of prophylaxis is to raise factor levels to prevent bleeds. Primary prophylaxis is the type of long-term continuous prophylaxis started before a pattern of bleeding has occurred, usually at a young age. Secondary prophylaxis is also long-term continuous prophylaxis but is started after a pattern of bleeding has been demonstrated.

Planned preventive treatment

Planned preventive treatment involves infusing factor before an event or activity that may potentially cause a bleed (for example, infusing before practicing or playing a sport – even when you are not scheduled to infuse). This type of preventive treatment regimen can also be performed for those that already treat prophylactically.

Rest, Ice, Compression, and Elevation (RICE)

No – it's not the stuff you eat. RICE is another treatment option for people with hemophilia. It is often used in conjunction with factor replacement therapy. This treatment can be used when experiencing a bleed away from home until factor is accessible (although it is advised to have factor on-hand whenever possible). The damaging effects of a bleed in a muscle or a joint can be reduced when RICE is used appropriately. Your hematologist will work with you to develop a treatment plan for you or your child.

R = REST

Stop the activity, infuse clotting factor or medication and protect the body part. To rest the injured area, a sling, splint, crutches or a wheelchair may be needed for several days to several weeks after an injury.

I = ICE

Ice or cold packs are useful in slowing the bleeding, reducing inflammation and reducing pain. Heat should not be applied to an area that is bleeding, hot or swollen.

C = COMPRESSION

Compression with soft elastic supports helps reduce swelling and pain.

E = ELEVATION

Elevation above the level of the heart reduces swelling.

Ports and venous access

Factor must be infused into veins. Infants have a high percentage of body fat, which may make finding a vein difficult. For children or adults who require frequent infusions, or those with problematic venous access, your hematologist may recommend a central venous access device (also known as a port) that is implanted under the skin. Factor is infused into the port, which has a tube connected to a large vein. Proper sterile technique when accessing ports is the most important thing to do to avoid infections in your blood.

For Children: As a parent or caregiver, whether using a port or peripheral veins, it is important to involve your child in the infusion process. This can start at an early age. A positive approach is important. Talk to your child during the infusion and speak in terms he can understand. Although toddlers cannot comprehend that their blood lacks a protein necessary for coagulation, they can be taught about the various supplies needed for an infusion: factor, sterile water, needles, gloves, alcohol wipes and so on. These are the seeds you plant for the future, as there will come a time when your child will need to learn to self-infuse. In the early years, keep it simple. First, start by making it the child's job just to get the factor out of the refrigerator or to hold the bandage. These simple actions can be built upon, adding more complex steps as the child matures and can handle more responsibility.

For Adults: As an adult learning to manage your hemophilia, your hematologist and specialty pharmacy can help train and educate you on the infusion process. You can learn to self-infuse as well.

Treatment logs

It is important that you always maintain infusion/treatment logs. They are much more accurate than trying to recall bleed information from memory. They will also serve as a record of infusions and bleeds in the event your child's factor therapy needs to be modified by your hematologist. In the event of a product recall, treatment logs will let you know exactly which lot numbers were infused. **More and more, insurance companies are requiring logs before they will approve a refill of factor products.**

Stage 1:

The Newborn Years (0–12 Months)

Stage 1: The Newborn Years (0–12 Months)[8]

As the parent of a newborn with hemophilia, you may be experiencing a mix of emotions. It may be a shock to hear this diagnosis. This section provides suggestions for ways to safely manage your child's bleeding disorder and precautions you should take during his first year of life. It is important to build a relationship with your child's pediatrician, hematologist and specialty pharmacy in order to develop a care plan that is best for your child.

One of the earliest discussions you'll need to have with your child's pediatrician and hematologist is around the risks and benefits of circumcision. Some babies may have no problem with the procedure, while others may have prolonged bleeding, with some even requiring stitches to help slow the bleeding. In families with no history of hemophilia, the heel stick for infant metabolic screening or bleeding from a circumcision is often the first sign a baby may have hemophilia.

Over the first 12 months of life, your most common challenge with bleeds is likely to be bruises. These are discolorations of the skin due to small blood vessels being broken under the skin surface. Many children with hemophilia develop bruises from normal handling. As they are lifted or carried, children can develop bruises under their arms or on their buttocks. Hugging can cause a circle of bruising around the chest or body. Bath time can cause some bruises when holding children securely in the tub. When using a high chair or walker, bruises can develop on their sides, feet, under their arms and on their chest. Bruises will fade over several weeks' time.

You can use bath time or diaper changes to monitor bruises. If you notice a bruise getting larger over a few hours, you should contact your hematologist. They may want to see your child to evaluate the situation. Some children may need to have ice applied to the bruised area, and others may require a factor infusion. Your hematologist can advise you on the best treatment for your child.

At approximately 6 months or older, your child will become more mobile as a normal part of development. He will be able to roll over, sit up by himself and begin crawling and exploring the world. You may consider using knee and elbow pads to help prevent some bleeds as your child begins to crawl. He will also be pulling himself up and then falling down. This repeated activity may cause bruising on his buttocks and scrotum. Putting an extra diaper on can help prevent some this bruising.

A bigger concern for parents is when he hits his head or neck during a fall. Your child can hit his head as he falls on the floor, because his neck muscles are not yet strong enough to hold his head up to protect it from the fall. Never second-guess a head bump. For a child with hemophilia, any trauma to the head should be considered an emergency as it could have severe damaging effects. Your child should be evaluated as quickly as possible. Bleeding can occur inside the brain without any signs and symptoms initially, while some may experience any variety of the following symptoms: bruising, bump, headache, sleepiness, neck ache, nausea, vomiting, sensitivity to light or loss of consciousness. Always report any head injury to your hematologist immediately to discuss what to do when your child has bumped his head. Always report any head injury to your hematologist or pediatrician as well.

At approximately 9 months or older, children will begin to pull up and learn to walk, creating pressure on joints. It is important to remember that you may not see blood in all bleeding episodes. Sometimes the bleed can be in a joint or muscle. As blood fills the space around the joint, pressure will build up, pain will occur, and the joint will become stiff or swollen and feel warm to the touch. This type of bleed requires factor infusions to prevent joint damage from occurring. When a child bleeds repeatedly into the same joint, it is considered a target joint. Individuals with target joints tend to have more orthopedic problems as they age. Your hematologist will help you understand how to evaluate your child's bleeding and the best treatment option.

Developmental guide[9]

During the first year of life, your child will make enormous developmental gains. On average, a baby will nearly triple his birth weight and grow 10 inches; however, each child will develop at his own pace. The following guidelines show some general development stages your child should experience.

By **3 months**, your child should be able to:

- Raise head and chest
- Grasp and shake toys
- Smile
- Communicate more
- Imitate movements and expressions
- Recognize familiar objects and people

By **4–7 months**, your child should be able to:

- Roll both ways
- Sit without support of hands
- Transfer objects from hand-to-hand
- Respond to expression of emotion
- Find partially hidden objects
- Explore with hands and mouth

By **8–12 months**, your child should be able to:

- Get to sitting position without help
- Assume hands-and-knees position
- Cry when parent leaves
- Eat with fingers
- Begin to use objects correctly
- Prefer certain toys and people

Pain and your infant[10]

Children with bleeding disorders can experience pain as a result of their condition and its treatment. As a parent, you can significantly impact how pain affects your child by understanding what pain is, how to deal with it and how to avoid it.

What is pain?

Pain is how your child's 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." The young infant cannot yet verbally express what he is feeling. This can make it difficult to determine if and where your infant has pain.

How can you tell your infant is in pain?

Your infant may:

- Be irritable or less playful
- Not eat as well as normal
- Bite or squeeze his lips tightly
- Cry with a harsh, high pitch
- Not move out of one position very often, or move around a lot trying to find a comfortable spot
- Frown or squeeze his eyes tightly shut
- Not be comforted by you
- Pull his knees to his chest
- Pull the body part that is hurting away from your touch
- Shudder
- Sleep more or less than usual
- Rub or touch the part of his body that hurts
- Whimper or groan quietly

What to do

- Call your pediatrician and/or hematologist and explain why you think your baby is in pain.
- If medication is ordered, give it exactly as prescribed, and watch your baby to see how well it works. How long was he comfortable after the medication? Does it make him or her sleepy? How many times did you give him the medication? Does his pain seem better or worse?
- Follow up with the hematologist if the pain doesn't go away.
- Apply cool packs to bruises or swelling. Be careful of tender skin! Put a cloth between the pack and your baby's skin. Don't leave the cool pack on for more than 20 minutes at a time. Check the skin frequently to be sure it isn't too cold (evident as white or purplish coloring of the skin). Reapply every two hours.
- Rock and cuddle your infant.
- Play music or sing.
- When a bleeding episode is evident, have factor infused promptly — within three hours of the injury or onset of the bleed. Follow the instructions of your pediatrician or hematologist.
- Plan ahead. Discuss medication options with your child's pediatrician or hematologist, and have those medications available in your home to administer when your child is in pain. **Be sure to avoid aspirin, aspirin-containing products, and nonsteroidal anti-inflammatory drugs (NSAID) like ibuprofen.** Discuss any over-the-counter product with your hematologist or pharmacist prior to giving them to your child.
- During the infusion of factor, play music, sing a song or blow bubbles as a distraction. A favorite toy, stuffed animal or blanket is a must!

How to avoid pain

- Prevent pain by avoiding injuries. Use protective padding when appropriate or as advised by your pediatrician or hematologist. Keep your baby safe from falls and injuries.
- Avoid pain associated with the infusion process by using a topical anesthetic cream before needle sticks, if approved by your hematologist.
- Discuss treatment options with your pediatrician or hematologist and consider infusions at the first sign of a bleed to minimize the long-term damage that bleeding episodes can cause, while allowing your child to remain active.

Remember, do not give your child any medication containing 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]

Child safety[9]

Keeping your child safe is a big concern for parents. One of the best ways to inspect your home is to get down on the floor and examine your home from your child's viewpoint. Childproofing can eliminate dangers your child could encounter in your home and car.

To prevent falls

- Never leave your baby alone on a changing table, bed or chair.
- Use safety gates if you have stairs in your home.
- Avoid using walkers.
- Use the harness when your child sits in a high chair, stroller or shopping cart.

To prevent burns

- Test bath water before placing your child in the tub.
- Do not smoke, handle hot foods or liquids or cook when holding a baby.
- Place screens around fireplaces and heaters.

To prevent choking

- Inspect toys for any small or broken pieces.
- Remove any drawstrings or ties from clothing.
- Avoid foods children younger than four are most likely to choke on: hot dogs, raw carrots, grapes or raisins, popcorn, nuts and hard candies.
- Don't allow toddlers or young children to wander around the house or play while snacking.

Around the house

- Clean floors so they are free of small objects.
- Cover unused electrical outlets.
- Install safety latches on cabinets containing hazardous materials.
- Make sure appliance cords or drapery/blind cords are out of reach.
- Take a course in infant cardiopulmonary resuscitation (CPR). CPR is a combination of rescue breathing (mouth-to-mouth resuscitation) and chest compressions. Many organizations, such as the American Red Cross, offer classes.
- Have an emergency plan in place. Post the numbers of the poison control center, police and fire departments, ambulance service and your child's hematologist by your telephone.
- Have a stocked first aid kit.

A baby's first year of life is an amazing time. It will seem as if your baby makes a new discovery every day. As parents or caregivers, you will also be making new discoveries and learning about hemophilia. As your child grows, find additional information available in this guide.

Stage 2:

Your Toddler/Preschooler and Hemophilia (1–5 Years Old)

Your Toddler/Preschooler and Hemophilia (1–5 Years Old)

Welcome to the wonderful world of toddlerhood and preschool, a time of rapid change and transformation. As children grow from 1 to 5 years old, they advance from crawling to playing organized sports. This is the time when the mind is ripe for make-believe, ready to accept adventure at every moment, and the time when a child goes from using single words to using over 1,000 words and speaking in complete sentences. In this section, you will learn more ways to help manage your child's hemophilia and how you can help keep your child safe through the very active toddler and preschool years so you can let your child with hemophilia be just that — a child.

Homecare

Comprehensive medical care is extremely important for toddlers and preschoolers with hemophilia. The Centers for Disease Control and Prevention (CDC) has issued statements saying the best outcomes for hemophilia come when people with hemophilia receive homecare services.[12]

Your specialty pharmacy provider should be knowledgeable about hemophilia and many times will be able to provide homecare nursing services that are prescribed by your hematologist. Hemophilia homecare professionals should be able to provide services that give you the education you need to help make living with hemophilia easier and assist you in following the treatment plan determined by your child's hematologist. These services may include:

- Home delivery of factor and infusion supplies
- Help monitoring home factor inventory
- Timely billing and collecting
- Help teaching venous access techniques
- Written educational materials
- Nursing services, when needed

Developmental stages

In addition to your hematologist, it is important to see a board-certified pediatrician and utilize a specialty pharmacy with extensive bleeding disorder experience. A pediatrician must monitor the growth and development of your child and will take care of your toddler's additional health needs. Your pediatrician and hematologist should communicate on an ongoing basis.

Remember, all children are different and will reach these milestones at their own pace. The following chart shows some general development stages your child will experience[9]:

Activity	1 – 2 years	2 – 3 years	3 – 4 years	4 – 5 years
Physical development	• Walks and is learning to run and jump • Becomes more independent	• Is eager to explore and dislikes being confined • Copies adult movements • Helps dress and feed self • Enjoys simple art projects	• Has all baby teeth • Is curious about how body works • Has bladder and bowel control • Can ride a tricycle	• Enjoys dancing and singing • Can handle safety scissors or crayons • Is very energetic
Language development	• Says first words • Uses words to express needs • Combines words with action • Understands simple commands	• Asks about names of things • Expresses thoughts and feelings • Adds hundreds of words to vocabulary	• Likes to hear stories over and over • Asks lots of "why" questions • Speaks in sentences of three to five words	• Carries on long conversations, often involving fantasy • Shows sense of humor and enjoys silly stories • Confuses fact with fiction
Mental development	• Thinks aloud • Learns by trial and error • Develops short-term memory • Enjoys role-playing • Doesn't separate fantasy from real life	• Claims things as "mine" • Learns to share • Begins to sort objects by type • Understands time ("in a little while")	• Can name and sort objects • Understands cause and effect • Shows increased attention span • Makes up stories	• Learns to organize things, such as toys • Is very observant • Loves make-believe play • Grasps the idea of numbers and counting
Emotional development	• Becomes curious and wants attention • Is very self-centered • Begins saying "no" • May react to frustration with temper tantrums	• Wants independence • Imitates other children and adults • Enjoys constant activity (dislikes naps)	• Uses words to express emotion • Likes to make friends • Learns to share, displays fears • Sees him or herself as others do	• Knows difference between right and wrong • Seeks friends' approval • Enjoys being part of a group • May lose control or have mood swings

Nutrition[9,13,14]

Children learn about exercise and proper nutrition at daycare and are often eager to practice what they have learned when they get home. Involvement in selecting healthy foods and exercising as a family will help reinforce these valuable concepts. Instilling the foundation of proper nutrition and exercise from a young age is priceless!

Keeping weight in proper ranges is especially important for people with bleeding disorders. Extra weight can add stress on joints (which can increase joint pain and frequency of joint bleeds), make it harder to find veins for infusion, and require more factor per infusion since doses are based on weight.

The following are a few tips to practice good nutrition and healthy eating habits:

- Eat as a family. Children will follow parents' examples. If children see parents eat properly, chances are they will do the same.
- 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 containing 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.
- Keep nutritious snacks available. Some examples include fresh and dried fruit, vegetables, pretzels, fat-free cookies and sugar-free frozen ice pops.
- 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.

Dental care[15]

A child usually starts teething at approximately 6 months of age. Teething does not usually cause serious bleeding, because it is a slow process. A child should start visiting the dentist at around 1 year of age, and parents should take the responsibility of cleaning their child's gums and teeth until the child can begin to brush on his own. Do not give toddlers a bottle of milk or juice to help put the child to sleep. This can lead to tooth decay.

Allowing the teeth to fall out naturally and not wiggling them — no matter how tempting — can help to avoid additional bleeding.[16] If mouth bleeding does occur, Amicar (epsilon aminocaproic acid), which can be taken orally and used topically, may be recommended by your hematologist. It neutralizes the enzymes in saliva that often prevent a clot from forming in the mouth. Amicar is available by prescription only, and you should consult with your child's hematologist on dosage.

To prevent tooth decay and gum disease, have regular dental checkups, eat a balanced diet and establish a daily routine for brushing teeth (two times per day). Children require adult supervision when brushing teeth.

Immunizations

Immunizations provide protection against a number of infectious diseases. In addition to the standard immunizations, it is recommended that children with hemophilia be immunized against hepatitis A and hepatitis B. Some childhood immunizations are usually given into the muscle (intramuscular or IM), and some are given under the skin (subcutaneous or Sub-Q). Most immunizations can be given subcutaneously rather than intramuscularly to avoid bleeding.[14] If your child is on prophylaxis, schedule immunizations on a treatment day when factor levels are elevated.

Pain and your toddler[10]

Children with bleeding disorders can experience pain as a result of their condition and its treatment. As a parent/caregiver, you can significantly impact how pain affects your child by understanding what pain is and how to deal with and avoid it.

What is pain?

Pain is how your child's 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."

How can you tell if your toddler is in pain?

He may:

- Be less active or less playful
- Cry intensely or whine
- Tell you
- Be unable to sleep
- Eat less
- Exhibit physical resistance by pushing your hand away when you try to touch the spot where it hurts
- Protect where it hurts

What to do

- Ask your toddler directly if he hurts.
- Use a smiley face scale, and have your child point to how much it hurts.

Source: http://wongbakerfaces.org/

- Encourage your child to express what he is feeling. Does it hurt to move?
- Call your pediatrician and/or hematologist and explain why you think your child is in pain.
- If medication is ordered, give it exactly as prescribed, and watch your child to see how well it works. How long was he comfortable after the medication? Does it make him or her sleepy? How many times did you give him or her the medication? Does his pain seem better or worse?
- Follow up with the hematologist if the pain doesn't go away.
- Apply cool packs to bruises or swelling. Be careful of tender skin! Put a cloth between the pack and your child's skin. Don't leave the cool pack on for more than 20 minutes at one time. Check the skin frequently to be sure it isn't too cold (evident as white or purplish coloring of the skin). Reapply every two hours.
- Apply RICE (rest, ice, compression, and elevation) to an injury (see page 13).
- Allow for quiet time. Read, play games or watch a movie to allow the injury to heal.
- Listen to your child. Let him or her tell you what feels best. Your child is developing skills to cope with his pain and to demonstrate self-control and the ability to modify his pain.
- During the infusion of factor, play music, sing a song, or blow bubbles as a distraction. A favorite toy, stuffed animal or blanket is a must!
- Be sure to note any pain on the treatment log.

How to avoid pain

- Prevent pain by avoiding injuries. Use protective devices, such as helmets, knee and elbow pads, or shin guards when appropriate or as advised by your pediatrician or hematologist. Keep your child safe from falls and injuries.
- Avoid pain associated with the infusion process by using a topical anesthetic cream before needle sticks, if advised by your hematologist.
- Discuss treatment options with your pediatrician or hematologist to minimize risks of bleeding episodes while allowing your child to remain active.

Remember, do not give your child any medication containing 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]

Safety[9]

Parents want to keep their children safe, and children depend on their parents for safety. Safety can be challenging during a time when your child wants to play and explore his world. Many common childhood injuries result from preventable causes.

- **Falls:** Stairs, windows, and bicycles represent hazards during these years. Place safety gates in front of all stairs. Make sure windows remain closed and locked. Wearing a helmet when bicycling is recommended for all children.
- **Choking:** Cut finger foods into small, bite-size pieces. Keep your child from putting non-food items in his mouth.
- **Burns:** Inspect your home for fire hazards. Don't overload electrical outlets, and cover outlet plugs. Matches and smoking materials should always be disposed of safely. Keep a fire extinguisher and fire alarms on each level of your home. Do not leave your child unsupervised in the kitchen while cooking.
- **Drowning:** Never leave a small child alone in the bathtub or swimming pool. Use safety devices on toilets. Always use life jackets while boating.
- **Poison:** Keep medication, cleaning products, paints, gasoline, pesticides and other chemicals locked in cabinets and stored in original containers. If poison is swallowed or spilled on skin, follow instructions on the container. Keep the poison hotline number (800.222.1222) posted by the phone.
- **Car crashes:** Always use a properly installed child seat, even for short outings. Some states require the use of booster seats. Children under 12 years should avoid riding in the front seat of a car, and young children should not be placed in a seat equipped with an airbag.
- **Trampolines:** The American Academy of Pediatrics cautions against trampoline use for all children due to ongoing and increasing injuries. Children with bleeding disorders are at even greater risk for permanent damage from fractures and neck injuries.[16]

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

Discipline[17]

The goal of discipline is self-control, and all children need to develop a positive self-image while learning to control their emotions and behaviors. Most experts today agree that physical punishment is the least effective method for accomplishing these ends. In addition, spanking or striking a child with hemophilia could lead to bleeding episodes. When children need help modifying their behavior, experts recommend some basic rules for parents:

- Focus on the child's poor behavior, not on the child.
- Have a plan, and be consistent in your responses.
- Stay calm and in control.
- Allow the child to express his feelings, even those considered negative.
- Be a good role model.
- Try to treat all children in the family equally.
- Realize that change takes time.

Living with your child's hemophilia may be challenging, but it can be managed. Keep its impact to a minimum, and focus on the beautiful child in front of you. With knowledge and a supportive team on your side, watching your toddler/preschooler play, grow, and explore will provide one of life's greatest rewards. As your child matures, find additional information available in this guide.

Stage 3:

School-Aged/Pre-Teen (6–11 Years Old)

Stage 3: School-Aged/Pre-Teen (6–11 Years Old)

Your child is entering an exciting new stage in life and development. Strollers and potty training are a thing of the past, as are the initial encounters of living with hemophilia. As you move ahead to the beginning school years, it is important to understand the next level of development, milestones and issues related to hemophilia. Staying informed and prepared will help you continue this journey with your child successfully!

Growing and letting go

We learn about hemophilia through experience. Some parents become expert "bleed identifiers," while others who have gone to battle with inhibitors (see "Formation of inhibitors" on page 6) can accurately define "Bethesda unit" or "titer." Then there are parents who live with hemophilia as a daily routine of prophylactic treatment and may have children who do not know how to identify a bleed.

Regardless of your experiences with hemophilia thus far, parents experience similar mixed emotions as they begin to let go. Your child is growing closer to becoming an independent individual. During this period, your child can begin to take on real responsibility for this disorder.

Bleeds

Bleeds will still continue, and they may occur outside the home environment. With your child in school for a large part of the day, you can no longer monitor every move he makes and must rely on his ability to communicate whether or not he is experiencing a bleed or has been injured. If your child has severe hemophilia, spontaneous bleeds can also pose a big challenge.

Identifying and treating a bleed promptly are essential to maintaining healthy joints and muscles. Now is the time when emphasis must be placed on helping your child understand what a bleed is, how to identify a bleed and the importance of promptly treating a bleed. Emphasize the importance of prompt treatment, and explain the consequences of delayed treatment.

Involving your child in treatment

Children ages 6 to 11 can understand when and how factor is administered, as well as treatment options. An important step in educating your child about his treatment is to allow him or her to actively participate in his own care by taking on the following responsibilities:

- Taking inventory of medical supplies at home (with supervision)
- Ordering medical supplies and unpacking supplies when they arrive at home
- Scheduling appointments, which enable him or her to establish a relationship with the care team
- Setting up and preparing infusion supplies
- Filling out treatment logs

Treatment logs are important to help keep track of bleeds and to keep notes on infusions. Keeping a log or a journal is not the "coolest" part of learning how to infuse, so added incentives, encouragement and consistency may need to be employed. Remember, learning how to self-infuse is more than a needle stick. It is a process that begins with proper technique and ends in keeping accurate records. Children at this age will be receptive to learning how to keep a log. Instill this in them now before they hit the teen years, when adherence may be challenged.

Note: Be sure you don't make these new responsibilities part of your child's chores. Managing hemophilia care should be a team effort, reinforced with encouragement and praise.

General well-being

Dental care — By the age of 5 or 6, children generally begin to lose their baby teeth, starting with the incisors. By the age of 10, half of their baby teeth will have fallen out.

Allowing the teeth to fall out naturally and not wiggling them — no matter how tempting — can help to avoid additional bleeding.[7] If bleeding does occur, Amicar (epsilon aminocaproic acid) may be recommended by your hematologist. Amicar can be taken orally or used topically. It neutralizes the enzymes in saliva that often prevent a clot from forming in the mouth. Amicar is available by prescription only; consult with your child's hematologist on dosage. Other helpful suggestions to control oral bleeding include using cold liquids or frozen ice pops.

To prevent tooth decay and gum disease, establish a daily routine of brushing teeth two times per day, eating a balanced diet, and getting regular checkups. Although children at this age are quite capable of brushing their own teeth, supervision and assistance is still recommended to ensure proper dental care.

If your child has a port, your hematologist may recommend a dose of antibiotics prior to dental cleaning or dental procedures to avoid the risk of infection in the bloodstream. If your child is on prophylaxis, schedule dental work on a treatment day when factor levels are highest.

During this stage, some children may require braces on their teeth. The actual process of placing the braces on teeth is not a problem and generally does not cause bleeding. Cuts inside the mouth can occur as a result of wearing braces, but bleeding is usually minimal. Covering the metal brackets with a thin film of wax helps avoid injury.[18] If bleeding does occur, contact your hematologist.

Immunizations — Most immunizations are given before entering kindergarten. However, others are typically given during the later grades. Be sure to keep immunization records current. For children with hemophilia, both hepatitis A and hepatitis B vaccines are recommended. Immunizations should be given subcutaneously whenever possible, with ice applied afterwards.[14] If your child is on prophylaxis, schedule immunizations on a treatment day when factor levels are highest.

Sports and exercise — Regular exercise is beneficial for children with hemophilia. Exercise strengthens joints and muscles, which can reduce bleeds. The most common form of exercise for children is sports. Wearing protective gear and infusing factor prior to participation are recommended for certain sports. Stretching is also a good way to avoid injury before participating in an activity or sporting event.[19]

Low-impact sports, such as swimming or golf, are advised for children with hemophilia. Rough contact sports are generally not recommended. As your child becomes interested in a wide variety of sports, it is important to consult your hematologist for advice and treatment options.

Proper nutrition[9,13,14] — Children learn about exercise and proper nutrition in science or health classes at school and are often eager to practice what they have learned when they get home. Involving children in selecting healthy foods and exercising as a family will help reinforce these valuable concepts. Instilling the foundation of proper nutrition and exercise from a young age is priceless!

A report from the Centers for Disease Control and Prevention (CDC) showed that teens with bleeding disorders are almost twice as likely to be overweight as children the same age without a bleeding disorder.[20] Keeping weight in proper ranges 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 your family on the path to a healthy lifestyle:

- Eat as a family. Children will follow parents' examples. If children see parents eat properly, chances are they will do the same.
- 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.
- Don't skip meals; instead, eat smaller meals and healthy snacks.
- Keep nutritious snacks available. Some examples include fresh and dried fruit, vegetables, pretzels, fat-free cookies and sugar-free frozen ice pops.
- Choose healthy beverages. Drink plenty of water, milk or calorie-free beverages instead of soft drinks.

Physical development

Although the growth pace remains steady, a young child's bones at this stage are growing slowly, but involvement in sports and other physical activities helps your child develop balance and coordination.

Young children may experience "growing pains." The concept of growing pains is half-truth and half-myth. Growing children do have normal pains, particularly in their legs and feet. These pains, however, are caused not by growing but by excessive use of young muscles and joints that are not yet completely developed. Young children are extremely active, and this extra activity places stress on their still-developing muscles and joints.

Signs and symptoms: Growing pains generally occur in different parts of the thighs, calves and feet. The pains can be severe enough to awaken a child from sleep. A key symptom of growing pains is that they occur only when the child is at rest, usually at night or during naps. They never occur when the child is active. This fact distinguishes growing pains from pains caused by diseases or injuries, which are typically worse when the child is active. Growing pains do not interfere with or interrupt a child's daily play or routine and are never accompanied by fever or other symptoms of general illness.[21]

To help young children take proper care of their joints and muscles, encourage the following behaviors:

- Exercise and wear protective gear.
- Have the child wear sturdy shoes.
- Keep "in tune" with his body. Look for signs of a bleeding joint, which include limping, favoring a limb or swelling. The extent of swelling can be determined by comparing the extremity in question with the body part measurement on the opposite side using a paper or cloth tape measure.
- Respond to bleeds promptly and accurately with RICE and factor replacement therapy (with proper dosing).

Cognitive and psychosocial development

Children ages 6 to 11 are becoming more independent by exploring their neighborhood with friends, joining sports teams or clubs and engaging in social media and texting. This group of children is more aware of how peers, teachers and family members perceive hemophilia. The children may be more self-conscious about their hemophilia and not know how to explain it to others. Additionally, they do not want to be perceived as "different." A child with a port may be hesitant to take off his shirt before going swimming. A child experiencing a bleed may delay his treatment in order to finish a sports event or may just think the bleed will go away on its own.

Emphasize the importance of prompt treatment, and explain the consequences of delayed treatment.

Children at this stage of development have an increased attention span and the ability to reason and process information and may enjoy reading. School-age children are often computer savvy, and many are becoming proficient at using the internet. This combination of skills and access to information is very valuable for a child with hemophilia. When questions about hemophilia arise, parents can teach their children how to search for answers. Be sure to focus on websites from well-respected medical centers and communities, and avoid reading blogs which provide more opinions than facts about hemophilia. It is important to teach children that living with hemophilia means learning for life. Giving them essential educational tools will help pave the way for social and emotional development.

School issues[22]

As a parent, you have had several years to become accustomed to hemophilia, but, chances are, your child's school or teachers may never have had a student with hemophilia before. This is an opportunity to be proactive and educate school personnel. Plan a meeting before the school year to give them the information they need. There are resources available for you to share, and your Customer Relations Specialist may be available to assist in scheduling education for school personnel. Visit www.hemophilia.com to download a copy of the Coach and Educator's Guide to Bleeding Disorders.

Absences

Academic growth, peer relationships and self-esteem begin to come together as a result of positive school experiences. Absence from school can affect academic achievement and impact social relationships. If a child needs to be absent due to a bleed, it is vital he return to school and catch up on any missed school assignments as quickly as possible.

If your child has trouble attending school, he may qualify for a Section 504 plan. As part of the Rehabilitation Act of 1973, school districts are required to reasonably accommodate students whose disability limits one or more major life activities. These include walking, seeing, hearing, speaking, breathing, learning, working, caring for oneself and performing manual tasks. You may not think of having hemophilia as a disability, but hemophilia is recognized as one of the qualifying conditions covered by Section 504.

Section 504 plans are created for students who require some accommodations to place them on the same level as other students. For some people with hemophilia, certain activities included in a mainstream physical education class can be too strenuous. Your child may be able to participate in alternative activities or take a different class to meet that educational requirement. This accommodation may be as simple as the teacher knowing your child has hemophilia and being familiar with signs and symptoms of a bleeding episode. Please note that each state or school district will have specific procedures for implementing a Section 504 plan.

Privacy

Since fitting in is now paramount to your child, he does not want to be perceived as different. For children with hemophilia, disclosure of their medical condition can become a sensitive issue. Some children elect to tell their closer friends but are more discreet with other peers. Of course, a select few don't mind telling everyone about hemophilia.

Although your child may be shy about letting others know he has hemophilia, wearing medical identification jewelry (bracelet or necklace) alerts medical professionals to his condition in case of an emergency. It can save your child's life!

Keep in mind that, although it is important to respect your child's privacy, you will need to reinforce the need for school personnel to be informed of your child's condition.

Bullying

Bullying is a challenging issue that all children face. Unfortunately, we cannot protect our children from all the browbeaters of the world. Instead, we must educate them on how to handle such situations and, most importantly, to tell an adult if someone hurts him or her.

Pain and your school-age child[10]

Children with bleeding disorders experience pain as a result of their condition and its treatment. As a parent, you can significantly impact how pain affects your child by understanding what pain is and how to deal with and avoid it.

What is pain?

Pain is how your child's 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."

How can you tell if your child is in pain?

He may:

- Tell you
- Decrease his activity level
- Eat less, sleep more
- Limp or not use the injured area
- Protect where it hurts
- Be unable to sleep
- Try to conceal the injury or pain from his family, teacher or nurse

What to do

- Ask your child directly if he is in pain. Talk over what you have observed and what your child is experiencing.
- Determine if the pain is related to hemophilia.
- Use a scale of 0 to 10 (with 10 being the highest) to have your child point out how much he hurts.
- Encourage your child to express what he is feeling. Ask questions: Where does it hurt? Does the pain move from one area to another? How does the pain feel — sharp, dull, throbbing? Does anything make it feel better?
- Call your pediatrician and/or hematologist, and explain why you think your child is in pain.
- If medication is ordered, give it exactly as prescribed, and watch your child to see how well it works. How long was he comfortable after the medication? Does it make him or her sleepy? How many times did you give him or her the medication? Does his pain seem better or worse?
- Follow up with the hematologist if the pain doesn't go away.
- Apply cool packs to bruises.
- Apply RICE (rest, ice, compression, and elevation) to an injury (see page 13).

- Encourage your child to participate in his pain management. Talk with each other, and make a plan for how to best control the pain.
- Provide positive reinforcement when your child expresses pain promptly and participates in his treatment.
- Play music or sing.
- During the infusion of factor, encourage your child to select a vein, gather supplies and complete his treatment log. Playing a favorite video or music will help him or her relax.
- Be sure to note any pain on the treatment log.

How to avoid pain

- Prevent pain by avoiding injuries. Use protective devices, such as helmets, knee and elbow pads or shin guards.
- Treat bleeding episodes immediately or at least within 2-3 hours to prevent long-term damage and resulting chronic pain. Follow up with infusions as required or ordered.
- Avoid pain associated with the infusion process by using a topical anesthetic cream before needle sticks, if advised by your hematologist.

Remember, do not give your child any medication containing 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]

Safety[9]

Parents want to keep their children safe, and children depend on their parents for safety. Safety can be challenging during a time when your child wants to play and explore his world. Many common childhood injuries result from preventable causes:

- **Sports safety:** Wear a helmet and protective pads when bicycling, inline skating or riding a scooter. Use all protective gear suggested for any sport or activity.
- **Auto safety:** Always use seat belts. Children under the age of 12 should avoid riding in the front seat of a car, and children should not be placed in a seat equipped with an airbag.

As a parent, you will be amazed at all the things your child will learn, experience and accomplish during this brief five-year span — from mastering cursive writing and multiplication tables to gaining independence and becoming more involved in his care.

This time provides a chance to bond with your child before heading into adolescence, a stage that has its own milestones and challenges. As your child matures, you will find additional information available in this guide.

Stage 4:

The Teen Years (12–18 Years Old)

Stage 4: The Teen Years (12–18 Years Old)[8]

Now that you're a teenager, the management of your hemophilia will be different than before. You may notice things about living with hemophilia that you never noticed before. For example, have you seen adults with hemophilia with stiff, arthritic joints who need a cane or a wheelchair to get around? They are living examples of what uncontrolled bleeding can do to your joints.

Moving into the teen years is a critical turning point for both you, as a teenager, and your parent(s)/caregiver(s). You will now be given some more freedom to make choices for yourself. You are now responsible for making the right choices to keep your body working for you in the many years to come. If you make good choices now, your body will function in tip-top shape when it comes time to give the same advice to your own children. While you take on this newfound freedom and responsibility, your parents will be going through a major change as well. They will begin passing some responsibility to you – responsibilities that they have been managing for you your entire life. You may have been feeling that your parents/caregivers have been bossy when they ask you to brush your teeth, eat your vegetables and take your factor. Bossy or not, they've been trying to get you into the habit of taking care of yourself.

This is a tough stage in your life when you are expected to act like a grownup, but some still see you as a kid. Teenagers face a lot of pressures, and having hemophilia can certainly make things more complicated. The goal of this section is to help guide you to a realistic approach to taking care of your body. You only have one body, so it's a good idea to take care of it.

Taking control of your treatment[8]

Have you heard the joke "How many teens with hemophilia does it take to change a light bulb?" The punch line is "None, their parents do it for them." It's kind of funny, and it's also kind of true. As a teenager, it's important to know all about your treatment and how to administer it. Remember: It's your body and your hemophilia.

As a young adult with hemophilia, you should be able to answer these questions: Do you have hemophilia A or B? Is your hemophilia mild, moderate or severe? Do you know what kind of factor you use? Do you know if your factor is plasma-derived or recombinant? Do you know when and how many units to infuse? Do you have a target joint? Do you know what inhibitors are and do you have inhibitors?

Talk to your parents, your hematologist or medical staff to become familiar with this information. You can even start by paying closer attention to the box of factor you're opening and the pharmacy label on it. One day, sooner than you think, you'll be out in the world on your own. You'll be a step ahead by knowing this information.

Now that people can receive factor at home, treatment is much more convenient. But it's important to know how much factor you have at home. Paying attention to your factor and supply inventory will help avoid times when you run out and have to go to the emergency room for treatment, where you may have to wait a long time before you get treated and released. Your hematologist can help you decide how many doses of factor need to be kept at home.

You should talk to your parents and put together a plan to take a more active role in managing your factor and your hemophilia. It could be as simple as writing your parents a note that says, "I need more factor," or picking up the phone and ordering from your pharmacy or pharmacy care provider.

As a teenager, you may want to start doing your own infusions. This means doing all the steps necessary for the infusion, including the needle stick. If you're not self-infusing already, let your parents and medical staff know when you feel ready to learn. Self-infusion is easy to learn and can be important for your independence. Once you're independently infusing your medication, you don't have to rely on other people and can go anywhere as long as you take your factor and supplies with you.

A lot of people complain about having to keep treatment logs, which record and document details about their bleeds, how much factor they used for each infusion and factor lot numbers and expiration dates. It is important to keep track of this information so you can see at a glance what types of bleeding episodes you are having, how often you treat and if there are any target joints developing. More and more insurance companies are making people keep treatment logs and turn them in as part of approving payment for the next shipment of factor. Treatment logs vary and can come from your hematologist, pharmacy, insurance company or factor manufacturer. Find a system that works for you, and stick with it.

General well-being

Dental care[23]

It's important to take good care of your teeth so they, too, can last a lifetime. As you grow, you might need braces. Your wisdom teeth may need to be removed. Consult with your family dentist about this, and always make sure your dentist has the number of your hematologist. If you're on prophylaxis, schedule dental appointments on treatment days.

Immunizations

Even though you're finished with elementary school, your school may require additional vaccinations, immunizations or booster shots. Check with your school or its website to make sure that you are up to date.

Nutrition[9,13,14]

A report from the Centers for Disease Control and Prevention (CDC) showed that teens with hemophilia are almost twice as likely to be overweight as children the same age in the general population.[20] Keeping weight in best range for your body 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 with your family whenever possible.
- 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 containing 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, milk or calorie-free beverages instead of soft drinks.
- Don't skip meals; instead, eat smaller meals and healthy snacks.

Exercise[19]

Exercise is also important for a person with hemophilia. Strong muscles mean healthy joints. Healthy joints usually mean fewer bleeding episodes. You can talk to your medical staff, including the physical therapist, to customize an exercise plan to help you get the exercise you need without aggravating a target joint or a bleeding episode. Regardless of which sport or activity you decide on, make sure you wear any recommended safety equipment.

As hemophilia care has advanced, kids with hemophilia can grow up living an active lifestyle. For many, this includes playing organized sports, such as soccer, swimming, basketball or even baseball. It's important to be comfortable with the level of competitiveness on your team and to make sure your coach is aware of your hemophilia. Most importantly, discuss your sports participation with your medical staff to determine whether you will treat hemophilia on a prophylactic (preventive) or an as-needed basis. Visit www.hemophilia.com to download a copy of the Coach and Educator's Guide to Bleeding Disorders.

School issues[22]

Some teenagers make a conscious effort to blend in with other people at their school, while others get unusual haircuts or wear different clothes to stand out. Having hemophilia can make blending in a difficult task. Many teenagers try to avoid the subject by hiding or "forgetting" they have hemophilia and ignoring the symptoms of a bleed. This can lead to delayed treatment for a bleed or missed prophylaxis doses and result in more bleeding. Most people would agree that the cost of fitting in isn't worth the pain and joint damage an untreated bleeding episode can cause. As hard as being different from your peers may seem, it is very important that key people at your school know about your hemophilia, such as your school nurse. Since you've lived with your condition all of your life, it is likely that you and your family know more about your hemophilia than your school nurse does. Take this opportunity to share information about your hemophilia and how it's treated with school personnel. Your hematologist or specialty pharmacy provider can help you. Telling some of your closest friends about your condition can also be a benefit. If you are injured and they know about your hemophilia, then they can help you get the medical assistance you need.

Although you may be shy 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!

If you have trouble attending school, you may qualify for a Section 504 plan. The Rehabilitation Act of 1973 requires school districts to reasonably accommodate students whose health or disability limits one or more major life activities. These include walking, seeing, hearing, speaking, breathing, learning, working, caring for oneself and performing manual tasks. You may not think of having hemophilia as a disability, but hemophilia is recognized as one of the qualifying conditions covered by Section 504.

Section 504 plans are given to students who require some accommodations to place them on the same level as other students. For some people with hemophilia, certain activities in a physical education class can be too strenuous. You may be able to participate in alternative activities or take a different class in order to meet that educational requirement. It may be as simple as the teacher knowing you have hemophilia and being familiar with signs and symptoms of a bleeding episode.

Please note that each state or school district will have specific procedures for implementing a Section 504 plan.

Psychosocial issues

Dating and intimacy are subjects few teenagers like to discuss, but they are important for every young person. Many teenagers struggle with telling a boyfriend or girlfriend about their hemophilia. Talk about it with your closest friends, parents or other people whose advice you trust. In any relationship, honesty and open communication are essential. Treat the people you date with respect, and expect the same from them. If you choose to be intimate, choose to prevent pregnancy and sexually transmitted diseases. For more advice on this topic, speak with your parents or your hematologist.

A teenager's desire to fit in can sometimes have negative effects. Some teens choose to experiment with alcohol and/or drugs. In addition to being illegal, drugs and alcohol impair judgment and can cause you to engage in risky behavior. Think about where you stand on issues such as drug use, underage alcohol use or other behaviors that may have negative consequences. Make your decision and stand firm, despite what your friends may say. If they are your friends, they will respect your decision. If they do not respect your decision, then perhaps you should reevaluate some of your friendships.

As you approach 18, you will want to become more independent and do things by yourself or with your friends. You may still want to spend time with your parents, and you may want to have nothing to do with them, all at the same time! This is a natural part of growing up and testing your independence. Even though your parents may embarrass you, it's important to treat them with respect and to try not to criticize them. It's good to keep lines of communication open with your parents.

Driving skills

Some people can't wait to drive. Others are scared to death. As a teenager, you'll get to be in the driver's seat and step on the gas! It's important to get as much supervised behind-the-wheel training as possible so your driving experience is a positive one. Always wear your seat belt. It not only keeps you safe, it's the law. If your hemophilia limits your mobility in any way, you may want to discuss accommodations that can be made in order for you to drive safely. Refer to your state laws and regulations for teen driving.

It is important to understand that taking pain medications, whether over-the-counter or prescribed, may cause drowsiness, and driving while taking them may be considered "driving under the influence." This can cause fatal or life-threatening accidents. Do not drive while taking pain medications.

Developmental Guide

As you grow into a young adult, your body will change. Puberty will continue, and you will have to deal with issues like body hair, getting a deeper voice, acne, body odor and further development of your reproductive organs. Your testosterone level will increase, and you may find yourself getting angry more easily or getting more competitive on the sports field.

Teenagers typically experience a rapid rate of growth. Some teens grow so fast that activities they once did with ease become awkward to them, and they may notice their coordination isn't the same. Sometimes they trip over their own feet or lose their balance easily. The majority of the time, these issues can be chalked up to growing too fast for your other senses to catch up.

As a result, you may find that your long bones (legs and arms) sometimes ache, seemingly for no reason. These are often called "growing pains." Growing pains generally occur in different parts of the thighs, calves and feet. The pains can be severe enough to wake you from sleep. A key symptom of growing pains is that they occur only when you are at rest, usually at night and rarely during activity. This distinguishes growing pains from pains caused by diseases or abnormalities, which are typically worse when you are active.[8]

During your teen years, you may experience growth spurts. As a result, your factor dose may increase since factor is dosed by weight. Update your weight regularly with your medical team to ensure you have the right dose of factor. You should let your hematologist and pharmacy know if you gain or lose more than 10 pounds.

Pain management[10]

As a teen with a bleeding disorder, you may experience pain as a result of the 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."

How can you tell you are in pain?

- Activity level decreases
- Eating less, sleeping more
- Limping or avoiding use of the injured area
- Protecting where it hurts
- Being unable to sleep

How to manage your pain

You may try to hide your pain from your friends and family members, but don't let peer pressure interfere with your need to have pain evaluated promptly. You can be an active participant in your pain management plan by following these tips:

- Discuss factor treatment options with your hematologist if you participate in sports.
- If pain medication is ordered, take it exactly as prescribed, and observe how well it works.
  - How long are you comfortable after the medication?
  - Does it make you sleepy? How many times did you take the medication?
  - Does your pain seem better or worse?
  - Never take more pain medication than your physician prescribed.
- Follow up with your hematologist if the pain doesn't go away.
- Apply cool packs to bruises.
- Apply RICE (rest, ice, compression, and elevation) to an injury (see page 13).

- When factor is needed, treat immediately to prevent ongoing bleeding.
- 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.

How to avoid pain

- Prevent pain by avoiding injuries. Use protective devices, such as helmets, knee and elbow pads or shin guards.
- Avoid pain associated with the infusion process by using a topical anesthetic cream before needle sticks, if advised by your hematologist.

Remember, do not take any medication containing 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]

Transitional tools

Once you turn 18, you are legally considered an adult. You'll be given all sorts of responsibilities, whether you want them or not. At 18, you will need to become the primary contact for your medical needs. You will have to give permission for others to have access to your medical information, even your parents. You may want to start making your own medical appointments and calling your specialty pharmacy to order your factor. It's important to start preparing yourself now so the responsibilities don't come as a surprise to you.

For a person with hemophilia, health insurance coverage is a key concern when you become an adult. If you have a social worker, he or she can help you and your family navigate the path from being on your parents' insurance to being on your own employer-provided insurance.

Currently, 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] Talk with your family about how long you qualify to be on your parent's insurance.

Use this opportunity to get a good education. This can translate into a well-paying job with health insurance benefits.

Choosing a career

Your teen years are not too early to think about a career choice. Once you graduate from high school, some of the options you have may include:

- Attending a 4-year college
- Finding a job
- Attending a community college
- Attending a trade school
- Attending school part-time and working part-time

There are several scholarships available to people with hemophilia to pursue their higher education. Online research can easily provide more information about available options. Reliable organizations to start with are the National Hemophilia Foundation (NHF) and Hemophilia Federation of America (HFA). Don't forget to look for scholarships for individuals with your job interests and from service organizations in which you or your parents may be members.

Regardless of the path you choose, you will need to have a plan.

This is a time to explore which careers are attractive to you. Your school guidance counselor can tell you about different careers and the training they require. He or she can help you find a career matching your abilities and interests.

Having a career is an important step, as it can offer financial independence and give you a positive self-image.

When considering a career, it's important to consider if a job offers flexibility in case you do have a bleed. Other considerations include salary, hours, ease of commute, activity level and benefits package. Remember, eventually you will need to get your own insurance coverage.

By age 18, hopefully you'll be an independent person who can make medical decisions for yourself. That includes ordering your own factor, infusing your factor, following your hematologist's orders, filling out treatment logs and taking care of your body. Remember, it's your body and your hemophilia. As you continue to mature, you can find additional information later in this guide.

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 issues[26]

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 issues[27]

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]

Glossary of Hemophilia-Related Terms

Glossary of Hemophilia-Related Terms

Antibody — A protein made by the body's immune system to fight and destroy foreign substances, such as viruses, not normally found in the body. In some people with hemophilia, the immune system may make an antibody in response to infused clotting factor proteins. This is called an inhibitor.

Arthropathy — Joint disease. Repeated bleeding into the joints of a person with hemophilia may cause arthropathy.

Assay — A laboratory test that measures the amount of clotting factor activity. In the manufacture of clotting factor concentrates, assay is indicated on each vial of product to indicate the number of clotting factor units (listed as i.u., or international units).

Bleeding episode (bleed) — A collection of blood at a site inside or on the surface of the body that occurs when a person with a bleeding disorder is actively bleeding.

Blood vessel — The part of the body that carries blood to and from all body areas. An artery carries oxygen-rich blood from the heart to the body's tissues; a vein carries blood from the body's tissues back to the heart and lungs to receive more oxygen. **Capillaries** are the small blood vessels that get oxygen into the tissues and connect veins and arteries.

Butterfly needle — A type of needle used to infuse factor or other medications into a vein. It has a small metal needle connected to plastic tubing and wings used to hold the needle during insertion.

Chronic — Long-lasting or lifelong.

Clot — A jelly-like substance made of a protein called fibrin interacting with blood cells called platelets. A clot forms to stop bleeding from a damaged blood vessel.

Coagulation — The process by which liquid blood changes into a jelly-like semisolid to seal an injured blood vessel and stop bleeding.

Factor concentrate — A freeze-dried powder product that contains measured amounts of a particular clotting factor, such as factor VIII or factor IX.

Factor level — The amount of clotting factor in the blood, referred to as a percentage of normal.

Gene therapy — A method of treating hemophilia by replacing the defective gene with normal factor VIII or factor IX genes so the body will produce factor. This is currently an experimental process that is being researched. Studies are currently underway for both hemophilia A and hemophilia B.

Half-life — The amount of time it takes the body to use up one-half of the factor level circulating in the blood. Factor VIII concentrates have a half-life of approximately 8 to 12 hours. Extended half-life factor VIII products range from 12 to 20 hours. Factor IX concentrates have a half-life of approximately 18 to 24 hours. Extended half-life factor IX products range from 66 to 118 hours.

Hemophilia treatment center (HTC) — A specialized treatment facility where a team of healthcare providers (hematologists, nurses, social workers, physical therapists and other disciplines) work together to deliver comprehensive care for people with bleeding disorders and their families.

Home therapy — Administration of treatment products in the home by a nurse, the caregiver or the patient.

Immune tolerance induction (ITI) — Treatment used to overcome an inhibitor that may require high daily doses of clotting factor for an extended period of time with a goal of suppressing inhibitors.

Inflammation — The body's defensive reaction to injury or irritation. Symptoms include swelling, heat, redness and pain. In people with hemophilia, inflammation may occur in response to a bleeding episode.

Infusion — Giving a medication or solution into a vein. In hemophilia, infusion refers to giving factor into a vein or central venous access device.

Inhibitor — A protein produced by the body that attaches to the factor VIII and neutralizes it, so the infused factor does not work to stop a bleed in hemophilia A, or binds to factor IX and produces an allergic, often anaphylactic-type reaction in hemophilia B. The body produces this protein or antibody when the immune system recognizes infused factor as foreign protein and prevents it from working.

Joint — A place in the body where two or more bones come together.

Joint bleed — When a bleed occurs in a joint. Also called hemarthrosis.

Plasma — Liquid portion of the blood containing clotting proteins.

Platelet — Disc-shaped blood cells needed for clotting.

Port — A type of central venous access device surgically implanted under the skin to allow easy access to a vein for factor infusion. A port is made up of a small reservoir with a rubber-like top. The reservoir is attached to a plastic tube that is inserted into a large vein, usually in the upper chest. A special needle is used to inject factor through the skin into the top of the port. The factor travels through the tube into the vein.

RICE — Complementary treatment for bleeding episodes used in addition to factor therapy. The letters stand for rest, ice, compression and elevation of the injured area.

Specialty pharmacy — A company that provides factor products and infusion supplies in the home setting to support home treatment of hemophilia.

Spontaneous bleed — A bleeding episode that occurs with no obvious cause.

Synovium — The tissue lining a joint that produces a fluid that lubricates the joint. Bleeding into a joint occurs from blood vessels in the synovium.

Synovitis — Inflammation of the synovium of the joint that may result from joint bleeds. Left untreated, synovitis can lead to joint damage.

Target joint — A joint that bleeds repeatedly, at least three times within a 6-month period.

Trauma — Injury.

Unit — A measurement of factor activity referring to that found in 1 milliliter of normal plasma. Prescriptions to treat hemophilia are usually written as a target dose number of units based on kilogram of body weight, plus or minus 10% to allow for variances in factor production. Unit of factor concentrates (see Assay) refers to a standardized "international unit" (i.u.) of factor activity per vial of product.

Venipuncture — Insertion of a needle into a vein.


Aa Bb
1 2 3 4 5
11 12 13 14
21 22 23 24
31 32 33 34
93 94

Recalling What You Read

Recalling What You Read

Hopefully you have learned quite a bit about managing hemophilia throughout various life stages. See how well you know the answers to some of these frequently asked questions based on the section you just read.

The Newborn Years: Ages 0–12 Months
Which chromosome carries the gene for hemophilia?
See page 5.

What is prophylaxis treatment?
See page 12.

What are the two most common sites for a bleed in babies and toddlers?
See page 8.

What is a target joint?
See page 16.

Name four ways you can tell that your infant is in pain.
See page 18.

List three child safety tips.
See page 20.

Your Toddler/Preschooler and Hemophilia: Ages 1–5 Years Old
What can you do to involve your toddler in the infusion process?
See page 14.

What are three signs a bleed is occurring?
See page 35.

List three early signs of a head bleed.
See page 9.

What are three ways you can help your child develop healthy eating habits?
See page 24.

School-Aged/Pre-Teen: Ages 6–11 Years Old
Describe the RICE treatment option.
See page 13.

What are three ways to control oral bleeding?
See page 31.

Name two sports recommended for children with hemophilia.
See page 31.

Do you know what a Section 504 plan is?
See page 34.

List four actions to take if your child is in pain.
See page 35.

The Teen Years: Ages 12–18 Years Old

List three ways you can take more control of your treatment.
See page 38.

What information should your treatment log contain?
See page 39.

What are the three major consequences of extra weight relating to bleeding disorders?
See page 39.

What is a Section 504 plan?
See page 40.

Name three criteria to consider when choosing a career.
See page 44.

Adulthood

What are some ways to manage advanced arthropathy?
See page 47.

Name three ways to prevent accidents in your home.
See page 49.

List three of your insurance options/plans.
See page 51.

What is HIPAA?
See page 51.

What are four methods to deal with pain?
See page 53.

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