



Your Voice. Your Life.



Know Your NUMBER5: Knowledge Is Power



Know Your Numbers:

Knowledge Is Power

20%

Hemophilia cases that are hemophilia B

Foreword

50%

The level at which factor IX activity is said to be normal

Know Your Numbers: Knowledge Is Power discusses the importance of knowing and understanding specific sets of numbers, test values, and other data about living with hemophilia B through all life stages. Healthcare professionals who treat people with hemophilia B recommend ways to help them manage hemophilia B symptoms so they can make informed medical decisions. It's important for all people with hemophilia B to understand their numbers in order to better engage in a conversation with their healthcare professionals. This book offers some ideas and education on how to better understand your numbers.

On behalf of Pfizer Inc, The Coalition for Hemophilia B, and the B2B Consumer Advisory Board, we would like to offer our gratitude to the members of the hemophilia B community who contributed to this book: Paul Brayshaw and Carl Weixler. We appreciate your time and sharing your knowledge about living with hemophilia B.

The views and opinions expressed in this book are those of patients, parents, and hemophilia care specialists who are members of an advisory board within the hemophilia B community and not of Pfizer.

The information in this book should in no way replace the advice of your healthcare professional. Be sure to talk with your healthcare professional, nurse, or hemophilia treatment center (HTC) staff regarding any form of medical advice or treatment.

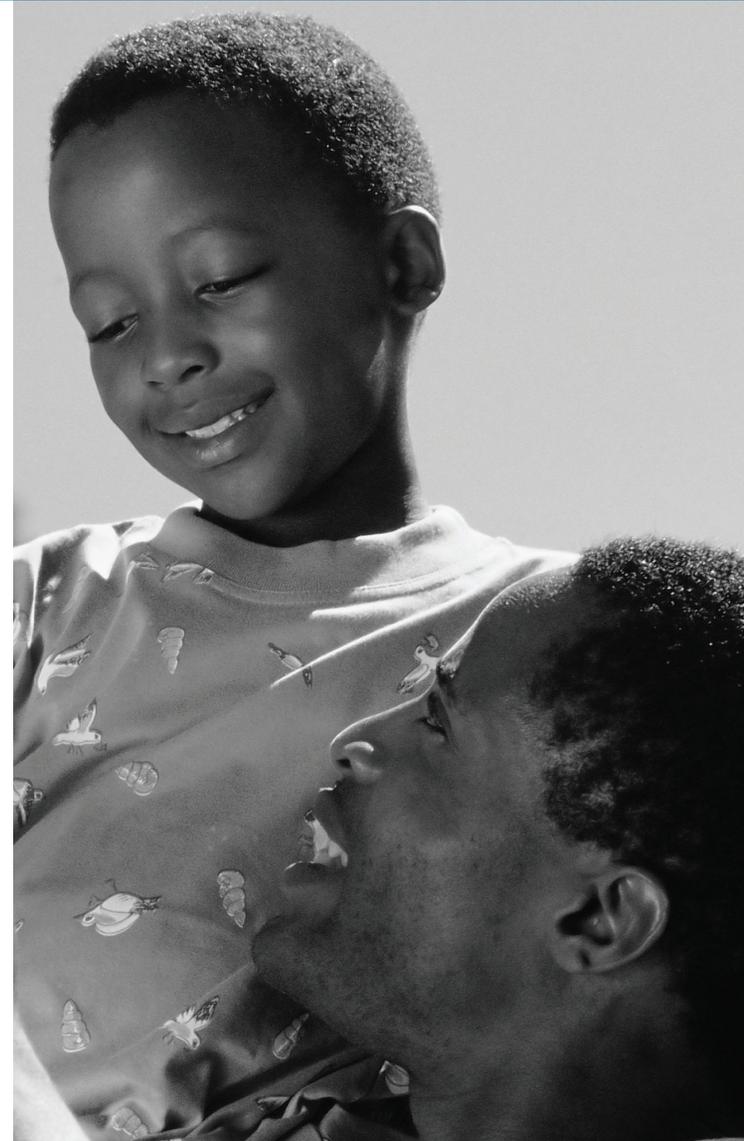


Know Your Numbers:

Knowledge Is Power

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Introduction

The daily management of your hemophilia B is only a part of your overall healthcare. While the management of hemophilia B is sometimes demanding, there are things you can do to help expand your understanding of the disorder and its treatment. You can arm yourself with facts that will help you make informed decisions about your own treatment. There are things you can do to strengthen your overall health.

Let's face it: some numbers are more important than others. There are many different numbers that play a part in management of your hemophilia B, such as peak and trough factor levels, current factor level, and even your insurance group number and your insurance company's phone number.

These are only some of the numbers you should know and understand to help you build your own advocacy and help ensure better health management of your hemophilia B. The more you know about managing your hemophilia, the easier it will be for your healthcare professional and care team to help you understand your dosing and treatment regimen, and manage long-term goals such as promoting joint health. Knowing some other numbers may also help you manage your overall health with proper diet and exercise.

The following pages will address the importance of knowing your numbers and what numbers you should know. You will hear from two men with hemophilia B who have developed a personal health management plan. They discuss how knowing and understanding their numbers has helped them to strengthen their own advocacy for their personal healthcare and has increased their engagement in their total wellness.

Everyone has different needs when it comes to advocating for their own healthcare. Your numbers are unique to you. Know your numbers. Become your own advocate. Discuss your needs with your healthcare professional.

1 in
25,000

Occurrences of hemophilia B in live male births

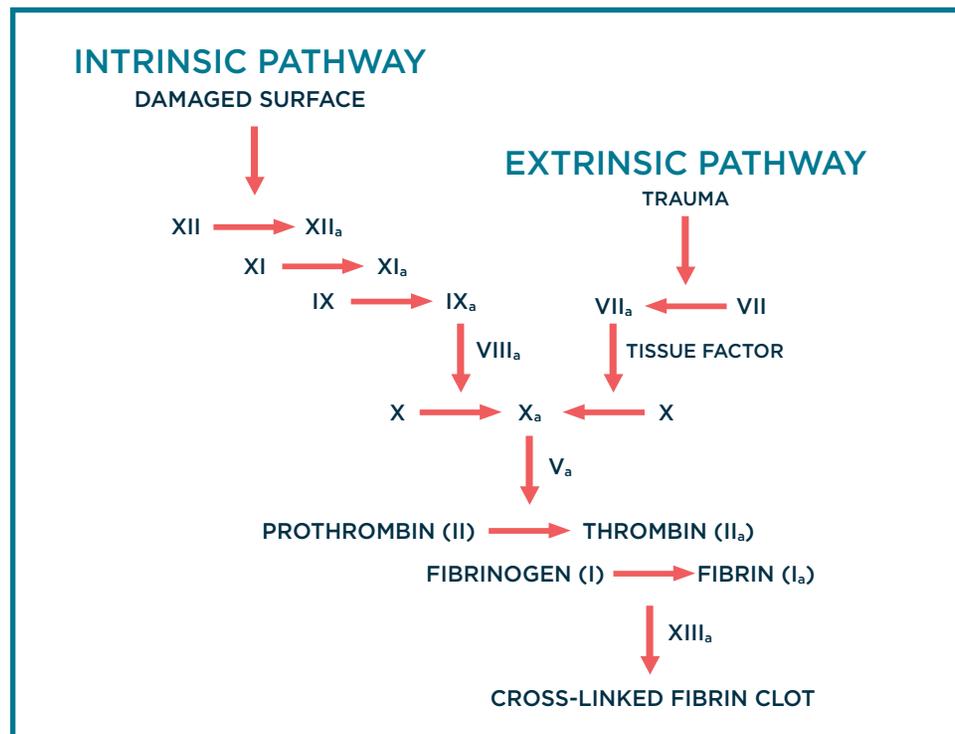


Mild hemophilia factor levels

A Science Refresher: Hemophilia B

A bleeding disorder is an acquired or inherited tendency to bleed excessively. Normally, blood remains in the circulatory system inside the blood vessels. However, if veins or arteries are injured, they will begin to leak blood, either externally or into body tissues. The body stops the blood loss through a complex clotting process called hemostasis. During hemostasis, the injured blood vessel constricts to reduce blood flow, platelets adhere to the injury site and clump together to form a loose platelet plug, and a process of clot formation called the coagulation cascade is initiated.¹

Illustration of coagulation cascade²



There are two main varieties of hemophilia. Hemophilia A, the most common type of hemophilia, occurs when the blood has little or no clotting factor VIII. Hemophilia B, the second most common type of hemophilia, is also known as factor IX deficiency or Christmas disease. About 20% of hemophilia cases are hemophilia B.³

Hemophilia B occurs in about 1 in 25,000 live male births worldwide.⁴ Most cases of hemophilia B are familial (inherited), while spontaneous mutations are rarer.⁵ Persons born with hemophilia have insufficient levels of a protein needed for normal blood clotting. The missing protein is called a clotting factor, which works together with other proteins and platelets to help blood clot. Platelets are small blood cells that are formed in the bone marrow. When blood vessels are injured, clotting factors help the platelets stick together to plug breaks in the blood vessel at the site of the injury to stop the bleeding. Without clotting factors, blood clotting cannot take place. When people with hemophilia are injured and bleeding occurs, the bleeding takes longer to stop.

Classification of hemophilia B based on in vitro clotting activity⁶

Severity	Levels of Factor IX in the Blood
Normal (person who does not have hemophilia)	40% to 150%
Mild hemophilia	Greater than 5% but less than 40%
Moderate hemophilia	1% to 5%
Severe hemophilia	Less than 1%

About two-thirds of hemophilia B cases have factor IX levels greater than 1%.³ The treatment of choice for factor IX deficiency is factor IX replacement. Examples of the levels of factor IX in the blood that your doctor may check include those that are just before a dose to determine the baseline level, soon after a dose to determine the peak level, and immediately before the next dose to determine the trough level.⁷

How is the dose of replacement factor IX determined for an individual?

- The number of factor IX units required = body weight (kg) × desired factor IX increase (%) × product-specific multiplier (varies by factor IX replacement product)^{7,8}
- Because of the wide variation in recovery among individuals, further individualization may be necessary⁸
- For severe bleeding, an appropriate dose of factor IX should yield a factor IX activity level of 80% to 100%^{7,8}
- Your treatment team is in the best position to determine the treatment regimen that is right for you⁸

Why Know Your Numbers?

Knowledge Is Power

“I’m Carl, and I have severe hemophilia B. When I first started treatment in the 1960s, it was with whole blood, not a factor product. Once I started self-infusion, I got more involved in my own care and paid attention to dosing. I began to understand what blood test levels meant. The more I understood about trough levels and half-life, the more I understood about the amount of factor IX I would need at certain times.

“It’s not just knowing your numbers that’s important, it’s understanding the numbers and what they mean that’s really important. People who know me are aware that I am my own advocate, and I work closely with my medical team to determine the right treatment for me. Become your own advocate. I did.”

- CARL WEIXLER



Carl began treatment

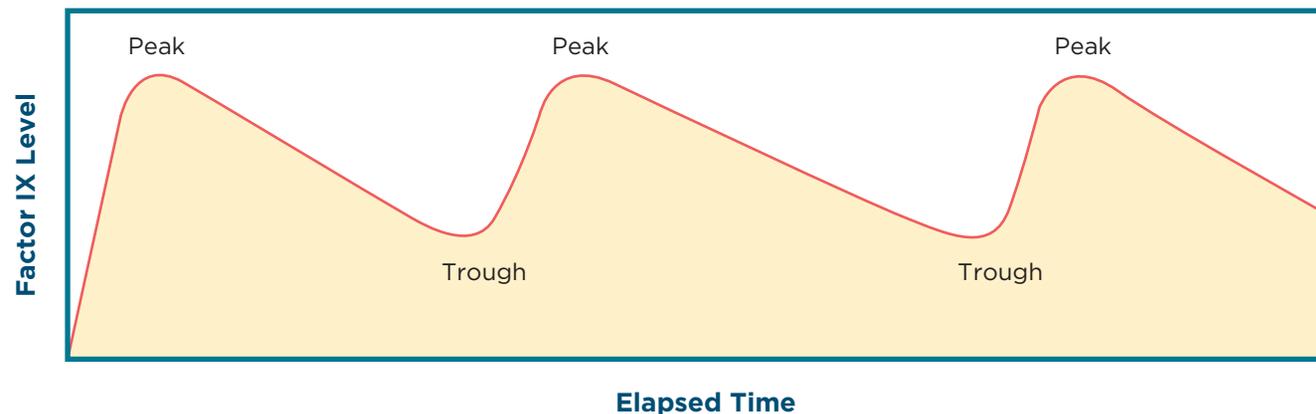
If you have hemophilia B, you should make every effort to learn the significance of laboratory tests that provide information that you and your healthcare professionals need.

What Are the Important Numbers to Know and Understand About Your Bleeding Disorder?

Peak and Trough

- Peak and trough information helps the healthcare professional determine if the dosage of factor IX is appropriate. This information may be especially useful when a new product is prescribed
- To get the peak and trough levels of factor IX activity, plasma samples are collected⁷
 - Peak specimens, representing the highest level, are usually collected soon after a dose is given intravenously
 - Trough specimens, representing the lowest level, are generally collected immediately before the next dose⁷

Illustration of Peak and Trough Levels

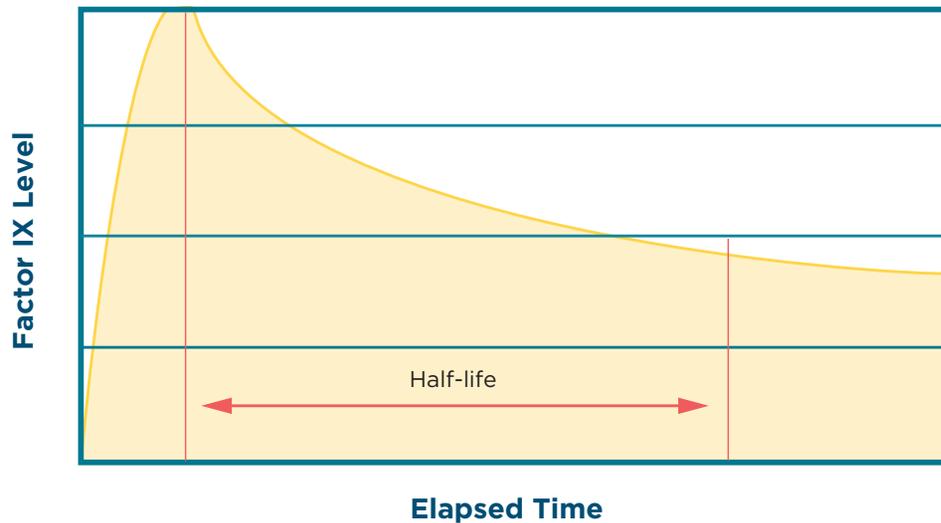


40-150%

Normal factor levels

Drug Half-life

- The amount of factor IX in the bloodstream is highest right after an infusion and then gradually decreases
- The half-life is the length of time it takes for the body to eliminate 50% of the amount of drug in the bloodstream.⁹ The half-life measures how quickly or slowly the factor breaks down in the body
- The half-life is important because it can help guide the appropriate drug dosage⁹
- The drug half-life assists the healthcare professional in recommending a particular dose regimen for an individual patient
- The half-life provides information to help assess how frequently the factor needs to be taken to maintain its therapeutic effect⁹
- Prescribing information is included with prescription drugs and provides information about half-life⁹



This graph is for illustrative purposes only

Hemophilia B cases with factor IX levels <1%

≈ 33%



Screening Tests That Your Healthcare Professional May Order for You

- Complete blood count (CBC)⁵
 - Measures the amount of hemoglobin (protein inside red blood cells that carries oxygen)
 - Measures the size and number of red blood cells
 - Measures the numbers of different types of white blood cells and platelets found in blood
- Activated partial thromboplastin time (APTT) test⁵
 - Measures how long it takes for blood to clot
 - Measures the clotting ability of factors VIII, IX, XI, and XII
 - Results of this test will show a longer clotting time in most people with hemophilia A or B
- Fibrinogen test⁵
 - Helps the healthcare professional assess a patient's ability to form a blood clot
 - Fibrinogen is another name for clotting factor I
- Measurement of specific clotting factors (FVIII, FIX, etc.)⁵
 - Measures the function of proteins essential for blood clot formation (also called factors or coagulation factors)
 - Shows type of hemophilia and severity of hemophilia



Factor IX

Make an effort to discuss your numbers with your care team. Let them know you would like to take a part in managing your healthcare by making informed decisions. To do this, you need to learn as much as you can about your important numbers.

Are There Any Other Numbers That Are Important to Know?

“There are many numbers we should know to take an active role in health maintenance and total wellness. As a person with hemophilia, I try to commit to exercising at least three times a week to maintain a healthy body weight. Talk to your doctor about the right level of activity for you.”

- PAUL BRAYSHAW



FIX

Become Your Own Advocate:

Design Your Personal Health Management Plan with Your Healthcare Team

Your health management plan can include anything that impacts your overall wellness. Be sure to understand what your numbers mean and discuss questions and treatment with a healthcare professional.

- Older adults may be asked to monitor their numbers more closely if they have other health issues, such as heart disease or diabetes
- People should be aware of their prescription drug dosing, such as how many times a day to take the dosage

Here are some more numbers that may be important for a person to know

Your factor IX product lot number and expiration date

- Every drug has a lot number on it that identifies the specific batch of medicine from which it came
- Keep this information in your log beside the date and dose of the infusion



Cholesterol and blood sugar levels

- Cholesterol and blood sugar levels are routinely assessed in adults as part of primary care
- Cholesterol levels may relate to cardiovascular health¹⁰ and blood sugar levels are assessed to screen for conditions such as diabetes¹¹

Blood pressure

Blood pressure is the force of blood pushing against the walls of the arteries as the heart pumps blood. If this pressure rises and stays high over time, it can damage the body.¹²

- It is important to know your blood pressure numbers, even when you are feeling fine

Stages of Blood Pressure Levels* in Adults¹²

(People aged 18 and older who don't have a short-term serious illness)

Stage	Systolic (Top Number)		Diastolic (Bottom Number)
Normal	Less than 120	And	Less than 80
Prehypertension	120-139	Or	80-89
High Blood Pressure Stage 1	140-159	Or	90-99
High Blood Pressure Stage 2	160 or higher	Or	100 or higher

*Measured in millimeters of mercury or mm Hg.

- If rising blood pressure is diagnosed early, a healthcare professional can help a person make adjustments in lifestyle or address the problem in other ways in order to avoid bigger problems later



Example of a normal blood pressure

Body mass index (BMI): weight and height¹³

Body weight alone won't give you a total health picture. BMI is the most commonly used measure of body fat.

BMI is calculated by dividing a person's weight in kilograms by the square of his or her height in meters (kg/m^2). Current guidelines from the Centers for Disease Control and Prevention (CDC) and the World Health Organization (WHO) define the following BMI ranges:

Underweight	Below 18.5
Normal	18.5 to 24.9
Overweight	25.0 to 29.9
Obese	Over 30.0

If a person's BMI is too high, there may be an increased risk for many chronic health problems.

People with hemophilia B should speak to their healthcare professional before beginning any weight-loss program.

18.5 to
24.9
Normal body mass index



Design Your Personal Business Card

You may find it's a good idea to prepare a personal business card that you can distribute to relatives, teachers, sports staff, hemophilia treatment center (HTC) personnel, and your closest friends. If you should ever need medical assistance, they will have the numbers to call. Include your parents' and your doctors' phone numbers.



EMERGENCY CONTACT CARD

Jane Mother

732.272.1234 (cell)
732.272.1234 (home)
732.272.1234 (office)

Joe Father

732.272.1234 (cell)
732.272.1234 (home)
732.272.1234 (office)

Relative

732.272.1234 (cell)
732.272.1234 (home)
732.272.1234 (office)

Dr. Sam Physician
732.272.1234

HTC PERSONNEL

Sam Nurse
732.272.1234 ext. 413

Joe Nurse
732.272.1234 ext. 415

Susan Nurse
732.272.1234 ext. 411

Phone numbers of emergency contacts, healthcare professional, and pharmacy

- Keep these numbers with you at all times, especially when traveling

Reimbursements for medical care — look at your policy for this information

- Insurance policy numbers
- Preauthorization numbers
- Copay amounts for the emergency room and healthcare professional's office

How do I get my numbers?

- Communicate with your healthcare professional and hemophilia treatment center about why you want to know your numbers
- Discuss with your healthcare professional why you wish to know specific blood levels, such as peak and trough
- Explain to them your goals and why knowing your numbers may help you achieve these goals

<1%

Severe hemophilia factor levels

Invest in Yourself: Look at the Whole Picture!

As part of a total self-management program, keep a log that includes all your health information: dates of bleeds, types of bleeds, how bleeds were treated, factor levels when known, and the dosage of any other medications you are taking. Tracking day-to-day information may help your healthcare professional or treatment team develop a treatment plan. Knowing how many bleeds you have had or your child has had will provide important information to your healthcare professional or HTC treatment team.

Note: People with hemophilia should always tell their medical or dental healthcare professionals about their disorder before any surgery or other invasive procedures are performed. No prescription medications or over-the-counter medicines, including herbal products, should be taken without first talking to your healthcare professional.

“I have encouraged my patients with hemophilia to communicate with the hemophilia treatment team about all aspects of their condition. When it comes to test results and what they mean, patients should never hesitate to seek a discussion with the healthcare team.”

- PATRICK F. FOGARTY M.D.

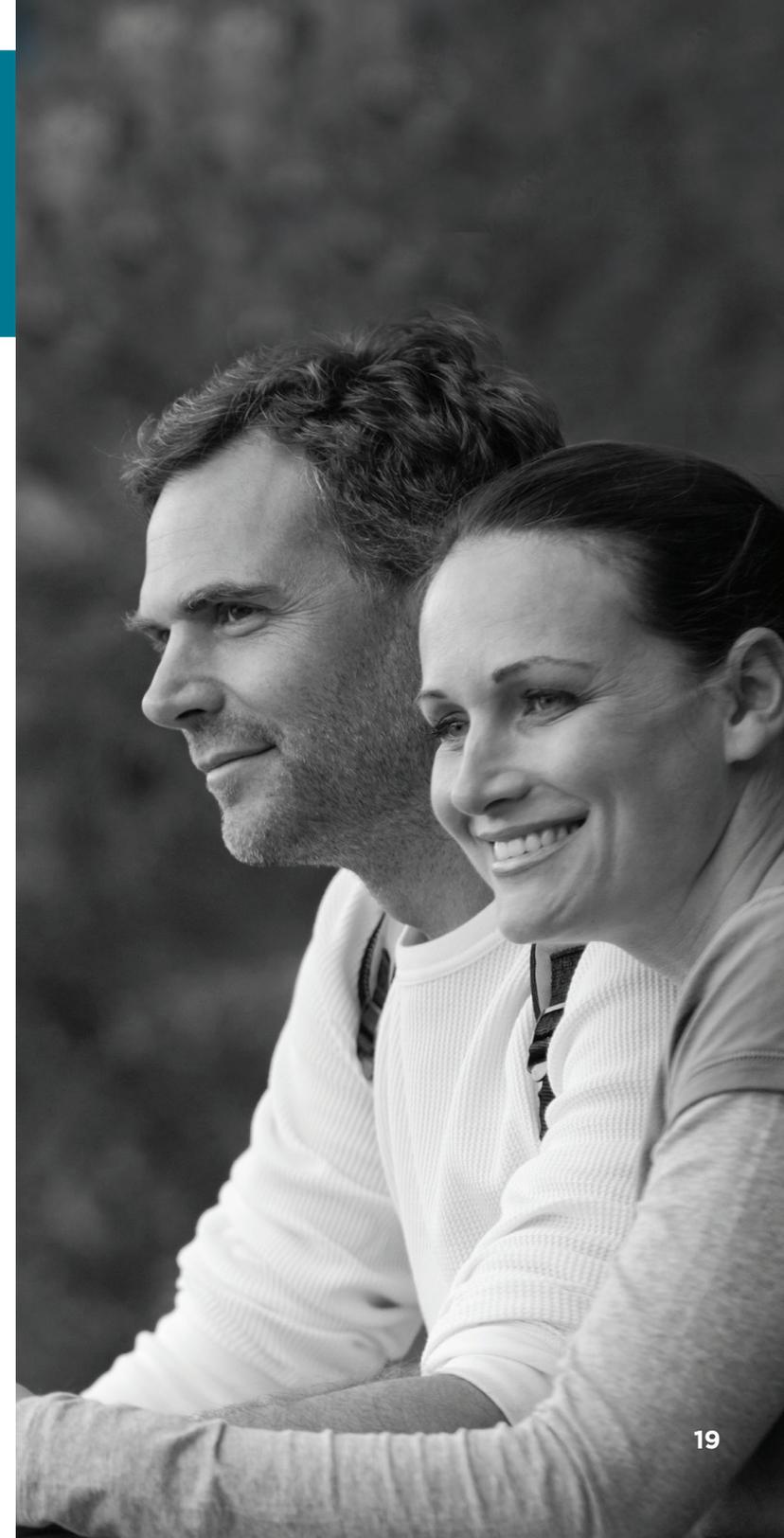


Conclusion:

Know Your Numbers — Know Yourself

Become an active advocate for your own healthcare; make this the new norm for the rest of your life.

- Empower yourself to make informed decisions and focus on healthy behaviors
- Communicate with your healthcare professional and your treatment team; become a partner in your healthcare
- Get up and get out: get on a regular exercise program (make sure you have the go-ahead from your treatment team)
- Know your numbers, understand your numbers, and use this knowledge to play an active role in managing your own healthcare
- Trust your numbers
 - Keep an active log of your numbers, as advised by your treatment team, and understand what these numbers mean
- Share your numbers
 - Make sure your family members know and understand your numbers in the event that your healthcare professional needs to speak to someone else



Resources and Support



Pfizer Hemophilia Connect

We're committed to helping the hemophilia community.

For the past two decades, Pfizer has been a part of the hemophilia community. We recognize it can be difficult to understand what support is available to help families with hemophilia, and to help address this challenge, we created Pfizer Hemophilia Connect.

Pfizer Hemophilia Connect is a one-stop destination to access all of our resources for eligible patients.



Soozie Courter Hemophilia Scholarship Program

Pfizer provides scholarships to students with hemophilia A or hemophilia B who are high school seniors, have a graduate equivalency diploma (GED), or are currently enrolled in an accredited junior college, college (undergraduate or graduate), or vocational school. Awards are based on academics, recommendations, and a personal statement from the student. Visit www.HemophiliaVillage.com to download an application.



HemophiliaVillage.com

The Pfizer-sponsored website, www.HemophiliaVillage.com, provides information for the hemophilia community. Consumers and professionals alike can find product information and learn about programs and services.



B2Byourvoice.com

The Pfizer B2B Consumer Advisory Board was developed to directly connect Pfizer to hemophilia B patients and caregivers in order to gain firsthand feedback from the hemophilia B community. As a result, the B2B program has created tools and resources to support the community and continues to evolve to address needs as they change. Visit www.b2byourvoice.com to check out a series of books, videos, patient stories, and other resources for people living with hemophilia B.

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