Table of Contents

Introduction ................................................................................................. 2

Peripheral Venous Access Devices ....................................................... 3
Where are the peripheral veins? ................................................................. 3
What types of peripheral venous access devices are available? ................. 3
What are the benefits/drawbacks of peripheral venous access devices? .... 4

Central Venous Access Devices (CVADs) ........................................... 5
Where are the central veins? ................................................................. 5
What types of CVADs are available? ...................................................... 5
• Catheters
• Ports
What are the benefits/drawbacks of CVADs? ........................................ 8
• Infection
• Thrombosis
Principles and importance of proper care .............................................. 9

Alternate Access Option ........................................................................ 11
Arteriovenous fistula (AVF) ................................................................. 11
What are the benefits/drawbacks of AVFs? ........................................... 11

At-A-Glance: What You Need To Know About Venous Access Options .... 12

Frequently Asked Questions ..................................................................... 15

Glossary ..................................................................................................... 16

References ................................................................................................. 17
Introduction

Treatment for patients with bleeding disorders many times includes the administration of clotting factor directly into a vein. This is called intravenous administration or IV administration. In order to get factor directly into a vein, there needs to be venous access or a pathway to the bloodstream. Venous access is important for the treatment of hemophilia and the prevention of complications due to bleeding.¹

There are different types of venous access devices with different benefits and drawbacks. For example, you can gain access to a vein simply by using a butterfly needle; however, other venous access devices are made to go deeper into your body and stay inside your body longer.

There are two main categories of venous access: peripheral venous access and central venous access.

- **Peripheral venous access** means infusing factor into a peripheral vein. Peripheral veins are located away from the central part of the body, such as in the hands or arms.
- **Central venous access** refers to infusing factor into a large vein near the heart.²

In general, peripheral venous access is the ideal method for people with bleeding disorders that require factor administration, but it is not always possible.³ This booklet outlines the types of venous access devices available for people with bleeding disorders and highlights some of the major benefits and drawbacks of each device.
Peripheral Venous Access Devices
Peripheral venous access means delivering factor into the body using a peripheral vein. Most experts emphasize that peripheral veins are the route of choice for the delivery of factor in people with bleeding disorders whenever possible.3

Where are the peripheral veins?
• Peripheral veins are the veins that are not in the central area of the body, like the chest or abdomen.
• Common peripheral veins used are in the hands and arms.

What types of peripheral venous access devices are available?
There are two main types of devices used for peripheral venous access: the butterfly needle and the heparin or saline lock.

Butterfly
• The simplest form of IV access is a syringe with a hollow needle attached. The butterfly needle is inserted through the skin into a vein and the syringe with medication is injected through the butterfly needle into the vein.
• A butterfly needle usually contains a small needle (23-gauge, 25-gauge or 27-gauge) with tubing attached to it.
• Butterfly needles are used once and then discarded in a puncture-resistant container. The wings on the needle make it easy to handle.
• Factor is injected from the syringe through the tubing and the needle, where the medication enters the blood.

Heparin or saline lock
• A heparin or saline lock allows peripheral access to a vein for a few days.
• With this type of device, a small, flexible plastic tube called a catheter is inserted into a peripheral vein.
• The outside of the device has a locking hub. The lock, or hub, is flushed with saline or heparin to keep it clean and open. Saline is now preferred over heparin in most cases.2
• A healthcare professional needs to insert the heparin or saline lock for use. This is rarely used in the home environment.
What are the benefits/drawbacks of peripheral venous access devices?

Benefits
It is important to remember that peripheral venous access is the long-term goal for patients with bleeding disorders. With peripheral access there is:
• A lower risk of infection than with central venous access.
• A lower risk of other complications, such as thrombosis, than with central venous access.
• Fewer limitations on lifestyle and activities.

Drawbacks
The drawbacks to peripheral access are:
• The need for frequent needle sticks (children often do not cooperate).
• The potential for the development of scar tissue on veins.
• Difficulties accessing small, immature veins in children.
Central Venous Access Devices (CVADs)

Although peripheral access is recommended for factor delivery, there are situations where a CVAD may be required.\(^4\) Central venous access refers to infusing factor using the large veins of the chest or neck.\(^4\) CVAD is a broad term that includes many types of catheters (thin, flexible hollow tubes) that are placed in large veins.

- CVADs allow frequent access to the bloodstream without deep needle sticks.\(^2\)
- CVADs can remain in place for long periods of time, such as weeks, months or years.\(^2\)
- CVADs can help with repeated administration of factor, fluids and other medications.\(^1\)
- CVADs are a useful bridge to peripheral access in small children until they are old enough to understand the need for treatment and are able to help.\(^3\)

CVADs should remain in place only as medically necessary for the individual patient—they should be used for a few years at the most.\(^3\)

Where are the central veins?
The main veins used for central venous access are:

- Jugular and subclavian veins (used for all types of CVADs)
- Cephalic and basilic veins (used for peripherally inserted CVADs)

What types of CVADs are available?
There are two general types of CVADs:\(^4\)

- Catheters: one end is outside the body
- Ports: surgically placed under the skin and require a special needle (Huber needle) to gain access
Catheters

**Peripherally inserted central catheters (PICCs)**
A peripherally inserted central catheter, or PICC, is inserted into one of the peripheral veins in the upper arm (usually the cephalic or basilic) and advanced into the superior vena cava or the right atrium. There is a shorter version of a PICC known as a midline catheter. Because the tip of the catheter in a midline catheter does not go as far into the central vein as the PICC, some people consider it a peripheral access device.² ⁵

**Non-tunneled catheters**
• A non-tunneled catheter is inserted into the subclavian vein in the chest or the jugular vein in the neck.⁴ ⁵
• Factor is infused into the lumen of the device.⁴ ⁵
• Designed to be used for about five to seven days.⁴ ⁵
• Infection is the most common complication.⁴ ⁵

**Benefits of PICCs² ⁵**
• Easily inserted by a healthcare professional.
• Low rate of complications.
• May remain in place for up to one year with appropriate care.

**Drawbacks of PICCs² ⁵**
• Risk of infection.
• Can become blocked or occluded (thrombosis).
• Can be damaged from movement or squeezing of the arm.
• Require weekly dressing change that can be difficult to manage.
Skin-tunneled catheters

- The Broviac® and Hickman® catheters, which were developed in the mid-1970s, were the first examples of skin-tunneled catheters.
- A catheter is tunneled under the skin and inserted into the subclavian vein or jugular vein and emerges a short distance away. The tunnel reduces the risk of infection compared with non-tunneled catheters.
- Tissue grows into the polyester cuff around the catheter, and scar tissue keeps the catheter in place.
- Factor is infused into a lumen that remains outside of the body.
- Activity is restricted due to the lumen remaining outside the body.
- Skin-tunneled catheters carry a potential risk for serious complications, such as thrombosis.

Ports

Unlike catheters, which exit from the skin, ports are placed completely below the skin to provide access to the central veins.
- The port is usually placed on the chest wall or lower rib cage.
- Ports are surgically placed into a pocket made in the muscles.
- The top of the port, called the septum, is about the size of a quarter or half dollar and can be felt under the skin.
- Factor is delivered by placing a special needle (Huber needle) through the skin over the port reservoir.
- This port can be used for approximately 2,000 sticks into the septum, which seals once the needle is removed.
- The Huber needle can stay in place in the port for five to seven days when covered by a dressing.

Ports offer a number of advantages compared with external CVADs. Some benefits of ports are:
- Ports are less likely to become infected.
- Ports generally last longer.
- Ports require less day-to-day care.
- Ports are subject to less tampering by children or accidental dislodging.
Ports are particularly beneficial in children under two years old, because a catheter can be moved or damaged by a child tugging on it. A catheter may also become infected by soiled diapers.¹

On the other hand, ports still require that a needle be inserted, which can cause discomfort and sometimes skin erosion. In addition, initial costs are higher for a port, and ports are more difficult to put in place and remove than external catheters.¹

Overall, a CVAD frees the patient and family from the need for immediate access to medical care, decreases the pain associated with venipuncture in children and reduces the need for visits to physicians, emergency rooms and treatment centers for infusion therapy.⁶

What are the benefits/drawbacks of CVADs?

Benefits
There are two clear benefits of CVADs in patients with bleeding disorders:⁶

- They are easy to use.
- They reduce the risk of long-term damage to peripheral veins.

Other benefits of CVADs include:¹

- CVADs are particularly useful in children with small, immature veins, when frequent access is required.
- CVADs are helpful in adult patients who are unable to use peripheral veins because the veins have become scarred.
- CVADs are useful in patients who have joint damage and cannot perform the steps required for peripheral venous access.

Drawbacks
There are also a number of drawbacks to the use of CVADs.¹⁻³

- CVADs must be placed under sterile conditions by a surgeon using a special x-ray machine.
- CVADs require careful management by the patient and/or caregiver.
- All CVADs must be flushed with saline or heparin to keep them clean and open.
- Aseptic techniques must be used to access the CVADs in order to prevent infection.
- Blockages can occur in CVADs.

The table on pages 12 and 13 lists many of the risks associated with CVADs. The two major risks are infection and thrombosis.

Infection
CVADs are foreign bodies that can become infected.⁶ Because CVADs are in direct contact with the blood, infections can be serious and sometimes life-threatening. Infections seem to be more common in children under two years old, children with inhibitors, children with a past history of CVAD infection or thrombosis and children whose lines need to be accessed frequently. Other factors that increase the risk of infection are poor oral hygiene and dental cavities and/or abscesses.⁶ The most important measure to prevent the development of infection is strict adherence to handwashing and aseptic techniques when accessing the device.

Thrombosis
Occlusions within the tubing, or blocks, are a common complication associated with CVADs. Although many clots can be dissolved with medications, some clots cannot be dissolved and the device may need to be removed or replaced.⁶
Principles and importance of proper care

Most patients and families can be taught appropriate care of CVADs to provide safe administration of factor at home.\(^1\), \(^3\), \(^6\)

- Children should be taught the purpose of the device, but should not be taught how to access the CVAD.
- Children can be allowed to assist with gathering supplies and set-up procedures if they are able and want to help.
- Transition of CVAD care should occur when the child is physically and emotionally able to accept responsibility for home care, usually at 12 or 13 years old.

There are three main principles of managing a CVAD: Prevent infection; maintain patency; and prevent damage to the device.\(^3\), \(^5\)

- **Prevent infection:** Aseptic technique and compliance with recommendations for equipment and dressing changes are essential to prevent infection. “Aseptic” means to complete an action without microorganisms or germs present. It is important that you keep the equipment and dressing free from germs that can cause infection. Proper handwashing is an important part of maintaining an aseptic environment. Symptoms of infection such as swelling around the site or catheter track, pain, redness, pus or drainage around the CVAD or fever and/or chills should be immediately reported to the hemophilia treatment center or hematologist.

- **Maintain patency:** Patency means that the device is open and not blocked or obstructed. If the device is blocked, it may be damaged and disrupt the delivery of factor and other medications. The most common cause of a block is a blood clot or thrombosis. Symptoms such as redness, pain or swelling should be immediately reported to a healthcare professional.

- **Prevent damage to the device:** A number of mechanical complications can also occur with a CVAD, such as catheter breakage/rupture and pinch-off syndrome. Most catheters are made of silicone, which can crack or split if handled incorrectly. Pinch-off syndrome means that the catheter becomes trapped between the collarbone and the first rib and is squeezed, or “pinched-off.” If pinch-off syndrome occurs, the catheter can fracture. In addition, catheter migration, or movement to another area, can occur as children grow.

Patients and families should review appropriate care of CVADs on a regular basis with their healthcare prescribers and should bring any questions regarding the CVAD to the prompt attention of their hemophilia treatment center or hematologist.\(^6\)
Alternate Access Option

Arteriovenous fistula (AVF)

- An arteriovenous fistula, or AVF, is an alternate access option for bleeding disorders patients who have lost the use of peripheral access and/or have had repeated mechanical failures or infections with a CVAD.\(^1,7\)
- An AVF surgically connects an artery to a nearby vein that allows venous access. A vascular surgeon creates an AVF by connecting an artery directly to a vein, usually above the elbow.\(^1,7\) Connecting the artery to the vein causes the vein to balloon outward and ultimately grow tougher and thicker.
- An AVF takes six to eight weeks to mature after surgery.\(^1,7\) While the fistula is developing, peripheral access or a CVAD is used for infusion.\(^8\)
- The main complications associated with AVF in people with bleeding disorders are thrombosis and failure of the fistula to mature. Sometimes the arm with the AVF can be a little shorter or longer than the other arm.\(^8\)
- Although AVFs have been used for people with bleeding disorders for more than 20 years,\(^9\) only a limited number of hemophilia treatment centers routinely use AVFs.\(^10\)

What are the benefits/drawbacks of AVFs?

Benefits\(^7,8\)
- AVFs are easy to access and reliable.
- There is a low risk of infection compared with other access devices.
- Because there is little change in how the body looks after the AVF is placed, there is no impact on body image.
- In general, patients and parents are very satisfied following placement of an AVF and treatment compliance improves.
- AVFs preserve central veins.

Drawbacks\(^7,8\)
- AVFs are expensive to place.
- Placement requires a vascular surgeon who has experience placing AVFs.
- General anesthesia is required.
- AVFs cannot be used for 6 to 8 weeks after placement.
- Peripheral catheter or CVAD is needed until the AVF matures.
At-A-Glance: What You Need To Know About Venous Access Options

Peripheral Venous Access Devices: Butterfly needle/Saline or Heparin lock

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hands and arms</td>
<td>• Long-term goal in bleeding disorders&lt;br&gt;• Lower risk of infection and complications than CVADs&lt;br&gt;• Fewer lifestyle limitations</td>
<td>• Frequent needle sticks&lt;br&gt;• Scar tissue can develop over time&lt;br&gt;• Difficulty mastering administration technique&lt;br&gt;• One-time (like a butterfly needle) or short-term use only (applies to Saline/Heparin lock only)</td>
</tr>
</tbody>
</table>

Central Venous Access Devices (CVADs)

**Peripherally inserted central catheters (PICC)**

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inserted in peripheral vein (cephalic or basilic) and advanced into superior vena cava</td>
<td>• Easily inserted&lt;br&gt;• Avoids painful needle sticks&lt;br&gt;• Low rate of complications&lt;br&gt;• Can remain in place for about one year&lt;br&gt;• Relatively safe and inexpensive</td>
<td>• Require weekly dressing changes&lt;br&gt;• Occlusions can occur&lt;br&gt;• Can be damaged from movement or squeezing of the arm</td>
</tr>
</tbody>
</table>

Non-tunneled catheters

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inserted in subclavian vein in the chest or jugular vein in the neck</td>
<td>• Avoids painful needle sticks</td>
<td>• Infection is main complication&lt;br&gt;• Designed to be used for five to seven days</td>
</tr>
</tbody>
</table>

Skin-tunneled catheters

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inserted in subclavian vein in the chest or jugular vein in the neck</td>
<td>• Avoids painful needle sticks&lt;br&gt;• Designed for long-term use&lt;br&gt;• Decreased infection rate compared with non-tunneled catheters</td>
<td>• Require weekly dressing changes&lt;br&gt;• Occlusions can occur</td>
</tr>
</tbody>
</table>
## Ports

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
</table>
| Inserted in central vein via muscle of chest wall or lower rib cage | • Easy to use — particularly in children with small veins  
• Reduced risk of damage to peripheral veins  
• Low visibility, improved body image  
  **Compared with external CVADs:**  
  • Reduced risk of infection  
  • Less day-to-day care  
  • More resistant to dislodging | • Needle puncture that can cause discomfort and skin erosion  
• Higher initial costs  
• Difficult to place and remove; must be placed under sterile conditions by a surgeon  
• Require careful management by patients and families  
• Risk of infection and thrombosis |

## Arteriovenous Fistula (AVF)

<table>
<thead>
<tr>
<th>Access</th>
<th>Benefits</th>
<th>Drawbacks</th>
</tr>
</thead>
</table>
| Artery and vein are connected by surgeon | • Easy to access  
• Quick and reliable  
• Low risk of infection  
• No relevant impact on body image  
• High patient and parent satisfaction  
• Improved treatment adherence  
• Spares central veins | • Expensive to place  
• Requires a vascular surgeon who has experience with AVFs  
• Requires anesthesia which can be problematic in children  
• Takes six to eight weeks to mature  
• Need for peripheral catheter or CVAD until AVF matures  
• Risk of thrombosis and failure to mature |
Frequently Asked Questions

Q: How do I determine which type of venous access device is best for my child?
A: Your healthcare providers will help you select the most appropriate device for your child based on your child’s clinical situation, past history with these devices, age and size.³

Q: Can a CVAD be used for blood sampling as well as factor infusion?
A: CVADs can be used for blood sampling. However, the 2004 Consensus Recommendations note that this practice should be limited or avoided because it increases the risk of infection and clotting.³

Q: Can an AVF be used for blood sampling as well as factor infusion?
A: It is not recommended that an AVF be used for routine blood sampling.⁷, ⁸

Q: How can infection be avoided in a patient with a CVAD?
A: To avoid contamination and infection, it is important that you use aseptic technique and comply with the recommendations for equipment and dressing changes provided by your treatment center.⁵ Thorough handwashing is a required component of proper aseptic technique.

Q: How long can a CVAD remain in place?
A: The 2004 Consensus Recommendations state that a CVAD should only remain in place for as long as medically needed. The maximum length of time that a CVAD can safely stay in place has not been determined, but there have been reports of CVADs remaining in place for five to ten years. Due to the risk of complications, CVADs are best used for only a few years at the most. The goal is to transition to peripheral venous access as soon as possible.³

Q: Does my child need to take antibiotics if he has a CVAD?
A: Not on a routine basis. There is no evidence that routine antibiotic therapy reduces the risk of infections; however, oral antibiotics will be necessary when having dental work or other invasive procedures.⁵

Q: Does a port require less care than an external catheter?
A: Yes. External catheters require more care than ports. With external catheters, the exit site must be regularly cleansed with an antiseptic agent and covered with an occlusive dressing, which places restrictions on showering, bathing and swimming. The external hub and catheter must be secured to the chest to prevent accidentally moving it during play, sleep and exercise.³

Q: Can a port be seen from the outside?
A: The entire port is inside the body and nothing can be seen except for a small bulge under the skin.²

Q: Can my child resume normal activities immediately after a procedure to place a CVAD?
A: After your child is discharged, you should follow the directions of your doctor. Usually your child can resume most activities the next day, but will be advised to avoid lifting five to ten pounds and other strenuous activities for a period of time after the insertion procedure. Avoid getting it wet by covering it with plastic wrap after placement. After a tunneled catheter or port is placed, you should expect to see some bruising and swelling, and tenderness in the chest, neck or shoulder. However, these symptoms usually go away after about three to five days. Pain medicine may help relieve these symptoms. It takes about 10–14 days for the port to heal after the procedure. It is important that you closely follow the instructions given to you by your healthcare providers.¹¹
Glossary

Arteriovenous fistula (AVF) — Connects a vein to a nearby artery allowing infusion of factor.

Aseptic — Without microorganisms or germs.

Catheter — Flexible plastic tube used in many venous access devices.

Central venous access device (CVAD) — A broad term used to describe a number of venous access devices that allow infusion of factor into the large veins near the heart.

Huber needle — A special bent or curved needle used to inject factor into the reservoir of a port.

Lumen — The cavity or channel within a tube or tubular vessel (like a vein or an artery).

Midline catheter — A shorter version of a PICC that does not go as far into the central vein as a PICC line.

Non-tunneled catheter — Flexible plastic tube that is inserted into the subclavian vein in the chest or jugular vein in the neck to allow the infusion of factor.

Patency — The openness (lack of obstruction) of a passage or a duct, such as a catheter.

Peripheral venous access device — A term used to describe devices that allow infusion of factor into veins located away from the central part of the body, such as veins of the arms or hands.

Peripherally inserted central catheter (PICC) — A tube that is inserted into one of the peripheral veins in the upper arm and advanced into the superior vena cava or the right atrium to allow the infusion of factor.

Pinch-off syndrome — The situation that occurs when a catheter is trapped between the collarbone and the first rib and is compressed.

Port — A device that is placed completely under the skin to allow the infusion of factor. The port is usually placed on the chest wall or lower rib cage.

Reservoir — The part of a port where the factor is injected.

Right atrium — One of the four chambers of the human heart. It pumps blood returning to the heart into the right ventricle.

Septum — Top layer of a port that seals once the needle is removed.

Skin erosion — The eating away of skin as can happen when a port is accessed many times.

Skin-tunneled catheter — Flexible plastic tube that is tunneled under the skin and inserted into the subclavian vein in the chest or jugular vein in the neck and then emerges a short distance away to allow the infusion of factor.

Superior vena cava — A large, short vein that carries blood from the upper half of the body to the heart’s right atrium.

Thrombosis — The development of a blood clot in a venous access device that can block the catheter.

Venous access — A pathway to the bloodstream.
References


