

GLOBAL NEWS SERIES FOR HEMOPHILIA NURSES



Third Issue

Message from the Editor

The New Year is upon us as you read this edition of the Global News Series for Hemophilia Nurses. Our team would like to wish you, your families and your patients a joyous New Year. We hope that peace and good health will be a blessing for everyone in 2007.

This issue will focus on Venous Access which presents many challenges to the hemophilia community and is an important part of hemophilia care. We have included information about the types of venous access available for the hemophilia patient, and two case studies are provided to illustrate the extreme challenges faced by the patient, their caregiver and the healthcare professionals. I hope you find this information useful.

This information is sponsored by an educational grant from Baxter BioScience. The newsletter's purpose is to share knowledge, experience and current events among hemophilia

nurses around the world. Each edition is developed in conjunction with nurses specializing in the care of persons with hemophilia. Due to the variation in approved therapies and practices, we will not provide specific prescriptive recommendations or treatment guidelines.

We welcome your participation in the newsletter through feedback on the current edition and/or submitting your ideas and topics for upcoming editions. We also invite you to participate as a subject matter expert in the development of this newsletter. To receive previous issues of this newsletter, provide feedback or express your interest as a subject matter expert, please e-mail us at contactus@solutionsight.com or write us at SolutionSight, Inc., 2191 Avalon Drive, Buffalo Grove, Illinois 60089. ❖

Cindy Ping, RN, BSN, MBA, CPP

Advisory Board January 2007

We are pleased to introduce the subject matter experts for the January 2007 edition of the Global News Series for Hemophilia Nurses.

With appreciation, we acknowledge Chris Guelcher, PNP, and Karin Lindvall, RN, for their knowledge and expertise in developing this edition. Their feedback has been invaluable. Chris Guelcher is a Pediatric Nurse Practitioner and Hemophilia Nurse Coordinator in the Pediatric Hematology/Oncology Clinic at Children's National Medical Center, Washington, D.C. Karin Lindvall is the Hemophilia Nurse Coordinator, Department of Coagulation Disorders, University Hospital, Malmö, Sweden. ❖

This Issue's Focus

Venous Access

As you work with your hemophilia patients and families, they look to you for guidance on many issues. Venous access is no exception. Obtaining good access to a vein is a major concern for those with hemophilia. With many intravenous (IV) access options available for delivery of factor replacement, families benefit from your knowledge and expertise regarding venous access.

In this issue, we will discuss peripheral venous access; central venous access which includes central catheters, ports and peripherally inserted central catheter (PICC) lines; and arteriovenous (AV) fistulas. Case studies are presented to share real scenarios involving venous access decisions. ❖

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Types of Access

Overview of Venous Access

Peripheral Venous Access

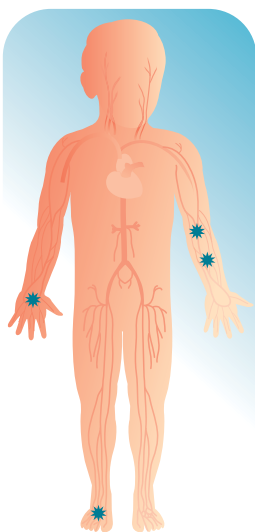
With peripheral venous access, a peripheral vein is used, usually in the hand or arm, although veins in other parts of the body may be accessed if necessary. Some benefits of peripheral venous access are:

- A single needle (such as a butterfly needle) is used for each treatment, and the needle is then removed until the next infusion.
- There are minimal chances of infection with peripheral access.¹

- Minimal site care is required before and after infusions.
- This may be the easiest option for a child once the family masters venipuncture skills.
- This type of access usually takes less time than other options.

Preparing For and Performing a Venipuncture

- Describe the procedure to the patient and caregiver.
- Provide privacy and position the patient comfortably.
- Select the appropriate insertion site, access device and materials.
- Dilate the vein, applying a tourniquet, and prepare the access site.
- Stabilize the vein and position the access device with the bevel side up.
- Insert the needle using a smooth, steady motion.
- Document the process according to your facility's policies.



Complications of Peripheral IVs²

- Local complications may include infiltration, phlebitis, hematoma, venous spasm, thrombosis and damage to tissue at the IV site.
- Systemic complications include air embolism, allergic reactions and septicemia.

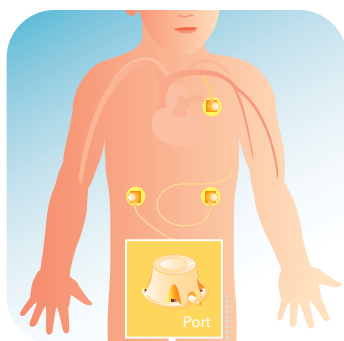
Central Venous Access

Two recent publications shed much light on the field of hemophilia and the usage of central venous access devices (CVADs). Authored by L Valentino, and B Ewenstein, et al., "Central Venous Access Devices in Haemophilia: a Meta-Analysis," presented a systematic review and meta-analysis of complication rates and risk factors associated with CVADs.³ Following the initial study, a second study was undertaken by Valentino and Ewenstein. Working with hemophilia experts, a guide to the best demonstrated practices when using CVADs was developed.⁴

Two major classes of longer-term CVADs (implantable ports and tunneled exterior catheters) are available and require a hospital visit for insertion. Blood samples may be collected through CVADs, however, this practice must be weighed by the medical team. Increased risks of clotting and infection may occur if residual blood remains in the catheter lumen.⁵

Fully Implantable Ports, Such as a Port-a-Cath (PAC)

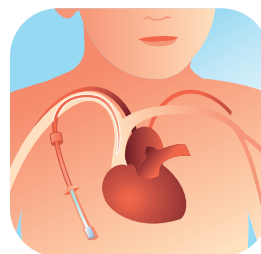
- A port consists of a subcutaneous reservoir with a self-sealing silicon septum. The reservoir is implanted in the chest in the operating room. Using a radiopaque silicone catheter, the tubing is threaded into the superior vena cava.



- A non-coring percutaneous needle (such as a Huber needle) is used to access the septum allowing numerous punctures.
- A port can stay under the skin for months or years, if needed.

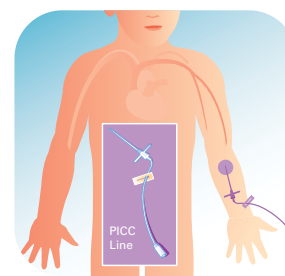
Tunneled Exterior Catheters, such as Hickman® or Broviac® catheters

- Catheters are silastic tubing threaded into the vein by a doctor in the operating room or in interventional radiology.
- One end of the catheter stays inside the vein while the other end comes out through the skin, usually on the chest wall, extending several inches outside the body.
- Factor is given through an injection cap on the end of the catheter.
- Dressings are required to keep the injection cap clean.
- Daily IV flushes are used to maintain patency of the line.



Peripherally-inserted Central Catheter (PICC line)

- A PICC line is inserted into a vessel in the arm with a catheter leading to a large vein emptying into the heart.
- Most often used for short-term therapy (a week to a few months), it does not require surgery to place it or remove it.
- Dressings are required to keep the line clean.
- Daily flushes are used to maintain patency of the line.



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Venous Therapy Complications

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Although these devices are usually implanted by a physician, often times in the operating room, there are certain things you can do for your patient to help ensure their wellbeing.

Before Insertion

- Explain step by step details of the procedure according to your facility's protocols.
- Reinforce the information provided by the physician.
- Respond to any questions or concerns the patient or their family may have.
- Educate the family about use, care and complications of a CVAD.
- Arrange for administration of factor infusion pre- and post-procedure.

During insertion

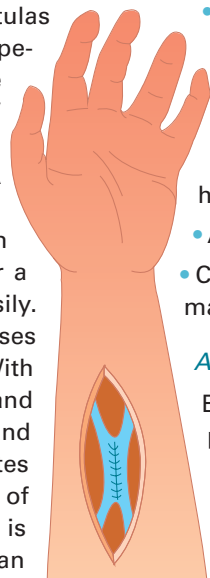
- Provide support to the patient and family.

After insertion

- Monitor the patient and site for complications.
- Follow-up on factor infusions post-procedure.
- Document the insertion according to your facility's protocols.
- Reinforce information provided about use, care and complications of a CVAD.

Arteriovenous Fistula

Arteriovenous (AV) fistulas combine the advantages of a peripheral access and the benefits of a CVAD.⁷ The AV fistula connects a selected artery to a nearby vein. A surgical procedure creates an enlarged, natural vessel, often in the arm, which a patient or a caregiver can access more easily. This surgical connection causes the vein to balloon outward. With time, the fistula "matures" and the vein grows tougher and thicker. The connection promotes the growth and enlargement of the selected vein. An AV fistula is natural and does not involve an



implanted port, catheter or synthetic device.

Advantages of AV Fistulas

- Easy to access once patient or caregiver is trained on use
- Rapid access and de-access
- Reliable functioning
- Low risk of infection (natural, no device or foreign body)
- High patient and caregiver satisfaction
- Compliance shown to be excellent⁸
- Allows for preservation of central veins

Disadvantages of AV Fistulas

- Time and effort of training
- Challenges and expensive of surgical procedure to place device
- Specialized vascular access required
- General anesthesia for children
- Need for external catheter placement
- May have cosmetic impact since AVF can bulge and cause local deformities
- Alternative IV access required while AV fistula heals

Possible Complications

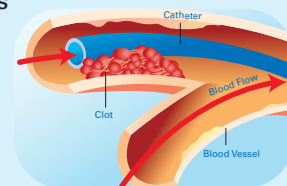
- Failure of surgical site to mature
- Bleeding complications
- Venous thrombosis
- Distal ischemia
- Infection
- Aneurysm (surgery or puncture-related)
- High-output left ventricular heart failure
- Abnormal limb growth
- Cannot be used until maturation (6-8 weeks)

Aftercare for the Patient⁸

Because vascular access problems can lead to treatment failure, the AV fistula requires regular care to make its use easier and to help avoid clots, infection,

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- **Pneumothorax:** Air in the thorax, the most common complication of central venous placement. Signs and symptoms include chest pain, dyspnea, cyanosis, decreased or absent breath sounds on the affected side
- **Hemothorax:** Bleeding into the pleural cavity, treated with the insertion of a chest tube for draining blood
- **Chylothorax:** Puncture of a lymph node with leakage of lymph fluid
- **Hydrothorax:** Infusion of a solution into the chest
- **Air embolism**
- **Thrombosis:** Development of a thrombus (blood clot)
 - Caregivers and healthcare professionals should maintain a high index of suspicion regarding the presence of catheter-related thromboses.¹ "Once a CVAD has been in place for 4 years (or conservatively as early as 2 years), venograms and/or ultrasound to evaluate the central venous architecture should be considered even in the absence of symptoms because of the increased chance of subclinical thrombotic events. Such monitoring should then continue to be performed every 1 to 2 years while the CVAD remains in place."¹



- **Perforation of vessel or adjacent organ**
- **Local infection**
- **Systemic infection**
- **Skin breakdown**
- **Extravasation**
- **Fibrin sheath formation**

Reference

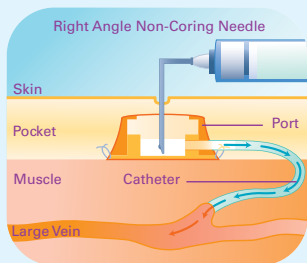
1. Ewenstein B, et al. *Haemophilia* 2004;10:629-648.

For more resources and suggestions, log on to www.SolutionSight.com/January2007

Best Practice Top-Entry PAC

When working with a Port-a-Cath (PAC) or other central venous access devices, sterile technique, including a specific cleansing agent, is required. For these directions, as well as any concerns with the PAC site (such as bruising), follow your institution's policies and procedures. After assembling your equipment, use the following step-by-step guidelines to safely and securely access a top-entry PAC:

1. Palpate the area over the port to locate the septum.
2. Anchor the port between your thumb and the first two fingers of your non-dominant hand. Using your dominant hand, aim the needle at the center of the device in between your thumb and first finger.
3. Insert the needle perpendicular to the port septum. Push the needle through the skin and septum until you reach the bottom of the reservoir. You'll feel the metal back of the port.
4. Check the needle placement by aspirating for a blood return.
5. If you can't obtain blood, remove the needle and repeat the procedure. Inability to obtain blood might indicate that the catheter is malfunctioning. If you can't obtain a blood return, notify the doctor: A fibrin sheath on the distal end of the catheter may be blocking the opening.
6. Flush the device according to the institution's policies and procedures. If you detect swelling or if the patient reports pain at the site, remove the needle and notify the doctor immediately.



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6. Flush the device according to the institution's policies and procedures. If you detect swelling or if the patient reports pain at the site, remove the needle and notify the doctor immediately.

Reference

1. Dunn M, et al. *IV Therapy Made Incredibly Easy*, 3rd edition. Philadelphia PA: Lippincott Williams & Wilkins; 2006:138.

Venous Access Options Advantages

The following chart lists advantages of the different options for venous access. Some of the advantages pertain to more than one type of access. This chart can be used to help your patients determine what type of venous access is best for them.

Venous Access Advantages	Peripheral Venous Access	Port (PAC)	External Catheter	PICC	AVF
1. May be easiest method with adequate veins.	*				
2. May be best method if veins are inadequate and/or do not tolerate repeated sticks.		*	*		*
3. Little care is required before and after infusion.	*				*
4. Child can usually swim, bathe or shower without limits if needle removed.	*	*			*
5. Lower in cost than other options.	*				
6. Initial cost is usually lower.	*				
7. Lower risk of infection.	*				*
8. Often used if child needs factor on regular basis or over long period of time.		*	*	*	*
9. Blood samples drawn easily (except factor levels and other specific tests).		*	*	*	*
10. Considering CVADs, less likely to become infected.		*			
11. All parts under the skin; low visibility; difficult for young children to tamper with.		*			*
12. Usually not visible when covered with clothing.	*	*			*
13. Considering CVADs, inhibitor patients may have fewer catheter-related infections than with external catheters. ¹		*			
14. Needle stick not required for infusion therapy.			*	*	
15. Easy to access; reliable; excellent way to infuse factor for urgent care.		*	*	*	*
16. Usually easier to insert and remove.	*		*	*	
17. Placed in outpatient department or radiology; surgery not required.				*	

References

1. Valentino L, et al. *Haemophilia* 2004;10:134-136
2. Venous Access Toolkit for Clinicians. Deerfield, IL: Baxter Healthcare Corporation; 2004:15.

Key Term

Port-a-Cath (PAC): A subcutaneous reservoir with a self-sealing silicon septum. The catheter attached to the reservoir is threaded into the superior vena cava.

Extravasation: Infiltration of irritating fluids, resulting in damage to surrounding tissues with potential severe local tissue damage

Thrill: A buzzing sensation felt over an arterio-venous fistula. It indicates that there is good blood flow through the access.

Venous Access Options Disadvantages

The following chart lists disadvantages of the different options for venous access. Some of the disadvantages pertain to more than one type of access. This chart can be used to help your patients determine what type of venous access is best for them.

Venous Access Disadvantages	Peripheral Venous Access	Port (PAC)	External Catheter	PICC	AVF
1. Child's cooperation needed for success.	*				*
2. Requires thorough education and time to learn.	*	*	*	*	*
3. Needle pierces skin each time factor given.	*	*			*
4. Surgery may be required with anesthesia for placement.		*	*	*	*
5. Possibility of infection higher.		*	*		
6. Formation of blood clots/blockages is possible.		*	*	*	*
7. More expensive to place.		*			*
8. Possible malfunctions of the catheter may occur.		*	*		
9. Families may need to depend on treatment centers/medical team for guidance for some time.	*	*	*	*	*
10. More steps required to learn this option.		*	*	*	*
11. Initial and/or on-going education of child and caregiver require willingness/commitment for family to master skills. ¹	*	*	*	*	*
12. Keep in place only as long as is medically necessary. ¹		*	*	*	*
13. Requires more care than other options.			*	*	
14. Has limits for showering, bathing and swimming.			*	*	
15. More time consuming when used properly (20 min for PAC ² , 10 min for pressure application with AVF).		*			*
16. Secure to chest wall to prevent accidental removal during play, sleep and exercise.			*		
17. May be seen under clothing or be more visible; may alter body image.			*	*	*
18. More likely to be displaced or pulled out by young children.			*	*	
19. Hub may become contaminated if close to diaper.			*		
20. Used for short-term therapy (a week to a few months).				*	

References

1. Ewenstein B, et al. *Haemophilia* 2004;10:629-648.
2. Valentino L, et al. Parent Empowerment Newsletter 2003;13:2. LA Kelley Communications, Inc.
3. Venous Access Toolkit for Clinicians. Deerfield, IL: Baxter Healthcare Corporation; 2004:16.

Best Practice Aseptic Technique

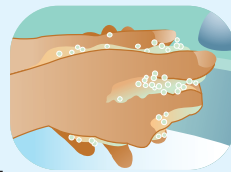
Aseptic technique, including basic hand washing skills, is imperative when working with central venous access devices. If gloves are worn, hand washing should be done before they are put on and after removal.

Steps For Good Hand Washing¹

- Apply antiseptic-containing liquid soap or water-free alcohol-based gel or foam to wet hands. Antimicrobial soaps destroy more germs than a non-antimicrobial product.
- Rub hands together vigorously and scrub all surfaces for 10-15 seconds.
- Rinse well and dry hands with a clean dry towel or paper towels.

CVAD Site Preparation¹

Cleaning of the CVAD site can be accomplished with soap and water, different antiseptics, or combinations of both. It is important to wash the skin with soap and water before using any aseptic agents since topical anesthetics may increase the risk of infection.¹ The Consensus Recommendations state that either povidone iodine or chlorhexidine can be used as a topical skin antiseptic agent.²



Examples of Aseptic Agents

Alcohol

- With vigorous rubbing, removes skin oil and cells
- Helps to kill germs on the skin
- Should remain wet on the skin for 60 seconds
- Germ-killing ability is gone once dry

Topical Antimicrobial (Povidone Iodine Solution)

- Most effective if applied and allowed to dry for two minutes

Chlorhexidine

- Binds to skin's protein
- Kills bacteria for up to six hours
- Should be allowed to dry and not rinsed off
- Can cause skin irritation
- Should not be used for children under two months of age

References

1. Venous Access Toolkit for Clinicians. Deerfield, IL: Baxter Healthcare Corporation; 2004:2, 3-4.
2. Ewenstein B, et al. *Haemophilia* 2004;10:629-648.

For more resources and suggestions, log on to www.SolutionSight.com/January2007

Venous Access

Two Case Studies

Case 1: Severe Hemophilia

The patient initially presented at birth in 1991 with bleeding from his circumcision. His factor VIII level of <1% was consistent with severe hemophilia. He was enrolled on a study with a recombinant factor VIII product. He developed an inhibitor in 1993 and his historical high factor VIII titer level was 20 Bethesda units. In 1994 he received activated complex concentrates (aPCC) to treat a refractory hemarthrosis. He developed some hives with aPCC so he was switched to an alternate aPCC. In April 1995 the patient was admitted to the PICU with a massive subdural bleed on the right with a midline shift and taken to the OR for clot evacuation. He was covered with porcine factor VIII and aPCC (with an antihistamine pre-medication). During admission, the patient developed a cross-reactive inhibitor to the porcine factor VIII product. He subsequently received compassionate activated factor VII from the manufacturer because it was not licensed for use at the time.

The patient had his first Broviac® catheter placed in May 1995 and ultimately discharged to a rehabilitation hospital in June 1995. At that time, his inhibitor level was <10 BU and he started on his immune tolerance regimen with a purified factor VIII (per the family's request). The patient was admitted on multiple occasions with

bacteremia and Broviac® site infections and he line was pulled in March 1997. A PAC was placed in April 1997 and pulled in December 1998 after several admissions for bacteremia. A second PAC was placed in April 1999. Despite immune tolerance, the patient's inhibitor became clinically significant in May 2003 due to non-compliance and required aPCC for management of refractory bleeding. At the same time, the patient's PAC stopped working.

The PAC was replaced in the OR in May 2003. In February 2004, the patient developed a cellulitis over the PAC and was removed after failing to respond to antibiotics.

In April 2004, a Broviac® catheter was placed. After several admissions for bacteremia, the Broviac® was removed in August 2005. For a period of time the patient received his immune tolerance peripherally, which became increasingly difficult for his family and later, homecare nurses.

He was admitted again in January 2006 for a new Broviac® catheter to facilitate the immune tolerance infusions every other day. At this time the patient (now 15) was instructed on infusing factor using sterile technique and he was able to infuse independently every other day without com-

plications until September 2006 when he presented with bacteremia (gram positive cocci) and was treated with a full course of IV antibiotics. During this time his Broviac® exit site was erythematous. A site culture grew pseudomonas and he was started on oral antibiotics. Unfortunately, the line fell out. An MRV performed during the admission failed to show any options for central venous access due to multiple previous central

venous access devices.

The patient has been continuing to receive his factor VIII every other day via peripheral IVs that are placed in clinic weekly and maintained by the patient at home. He was admitted for anastomosis of an artery and vein in the forearm (AV fistula).



Pediatric surgeons requested venous mapping, which showed that the right cephalic vein was too small to follow into the bicep. The right basilic vein was noted to branch from the brachial vein at the mid bicep. The left cephalic was too small to follow from origin to proximal forearm. The left basilic vein branched at mid-bicep. Ultimately, the patient was taken to the OR and the end of the brachial vein was anastomosed to the side of the brachial artery. A good thrill (a buzzing

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Case 1 – Venous Access Placement Summary

Date Placed	Type of Venous Access	Date and Reason Removed
May 1995	Broviac® Catheter	March 1997; Bacteremia and site infection
April 1997	Port-a-Cath	December 1998; Bacteremia
April 1999	Port-a-Cath	May 2003; PAC stopped working; development of clinically significant inhibitor
May 2003	Port-a-Cath	February 2004; Cellulitis at access site
April 2004	Broviac® Catheter	August 2005; Bacteremia
August 2005	Peripheral Access	January 2006; Experienced difficulty
January 2006	Broviac® Catheter	September 2006; Bacteremia; Catheter fell out
September 2006	AV Fistula	To present

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sensation felt over an AV fistula; indicates good blood flow through the access) was noted and no active bleeding was appreciated. Despite the findings on the MRV, a Broviac® catheter was successfully placed in the right subclavian on the second attempt. The Broviac® was intended to facilitate the immune tolerance infusions every other day while the AV fistula matured over 6-8 weeks.

The patient received 100 u/kg of factor VIII IVP pre-op, then he was to receive 50 u/kg Q8 hours post-op. His first post-op factor VIII was suboptimal at 34%. His subsequent dose was increased to 75 u/kg and a follow-up factor VIII was 186%. He was then decreased to 50 u/kg again and he continued to receive that dose Q8 hours until post-op day 3 when he was decreased to daily doses of 50 u/kg for 2 days and then to resume regular immune tolerance schedule with 50 u/kg every other day. He is scheduled to follow-up with surgery in 2 weeks.

Case 2: Moderate Hemophilia

A 59-year old man with a family history of hemophilia was diagnosed in 1956 with moderate hemophilia A and a factor level of 1%. Because he was diagnosed with moderate hemophilia, he did not receive regular prophylactic treatment. The patient has developed hemophilia arthropathy after repeated joint bleedings. In 1999, he underwent a left knee replacement. He was put on prophylactic treatment after this procedure with bad compliance. He is hypertensive, overweight and increased difficulties



with venous access lead to admission for an AV-fistula. He underwent necessary pre-investigations with ultrasound of the right and left arms for assessment of vein and artery morphology. Brachial, radial and ulnar arteries were without atherosclerotic changes. The patient is left-handed so the contemplated fistula was on the right arm.

Thirty minutes before the surgery he was given 5000 units of factor concentrate (49 u/kg). An anastomosis was established between right cephalic vein and radial artery on the right forearm. Good venous pulsations were achieved with a blood flow measured to 115 ml/minutes. Three hours from first injection another 2000 units of factor concentrate was given. Seven hours post surgery the AV-fistula occluded, no thrill was noted and no obvious intruding sound of the fistula was found. The patient was re-admitted for surgery where a thrombosis in cephalic vein was found and several fresh clots were collected 50 cm in proximal direction of cephalic vein.

A second attempt was made but during the procedure, the blood flow was rapidly falling and a new occlusion occurred. The clots were removed and the surgeon managed to achieve a new anastomosis. A couple of hours later a renewed occlusion occurred. There was a suspicion of a clinical hyper-coagulable condition. The patient was discharged from the hospital the following day and a PAC was successfully implanted a month later. ❖

Did you know?

Hospital Policies and Procedures: When performing patient care interventions, such as inserting intravenous lines, hospitals and healthcare institutions may have specific policies and procedures in place to protect the clinician and the patient. Be sure to investigate these before you engage in procedural interactions with patients. These policies and procedures may include, but are not limited to, direction for:

- Clinician hand washing
- Flushing ports or catheters
- Cleaning the patient's skin
- Changing dressings
- Starting peripheral or central lines
- Patient identification, teaching and documentation

Upcoming Events Conferences

2-4 February 2007

2nd International Symposium on Women's Health Issues in Thrombosis and Haemostasis

Vienna, Austria

E-mail: whith2007@palex.co.il

www.palexconventions.co.il/whith2007

17 April 2007

World Hemophilia Day

E-mail: wfh@wfh.org

www.wfh.org

19-20 April 2007

Canadian Hematology Meeting

Buenos Aires, Argentina

E-mail:

direccion@hematologia.anm.edu.ar

www.ish-world.org/canadian_meeting.htm

3-6 May 2007

WFH 10th Musculoskeletal Congress

Stresa, Italy

E-mail: msk2007@wfh.org

www.wfh.org

Call for Abstracts deadline:

31 January, 2007

13 May 2007

Mysore Half Marathon

Mysore, India

E-mail: varsha@hfindia.org

www.mysorehalfmarathon2007.com

6-12 July 2007

XXI ISTH Congress

Geneva, Switzerland

E-mail: isth2007@mci-group.com

www.isth2007.com

14-16 September 2007

40th Biannual Congress of ESPHI (European Society of Pediatric Hematology and Immunology)

Athens, Greece

E-mail: info@esphi.org

www.esphi.org



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and other complications. Patients can help protect the access by:

- Making sure the access is checked before each treatment.
- Not allowing blood pressure to be taken on the access arm.
- Checking the pulse in the access every day.
- Always keeping the access clean.
- Using the access site only as instructed.
- Being careful not to bump or cut the access.
- Not wearing tight jewelry or clothing near or over the access site.
- Not lifting heavy objects or putting pressure on the access arm.
- Sleeping with the access arm free, not under the head or body.

An AV Fistula Study

Recently, interest in the use of AV fistulae have grown. Dr. Elena Santagostino reported her experience with 27 patients who had 31 AV fistulae placed due to difficulties with venipuncture.⁹ In the study (1999-2002), 23 people studied were children and four were adults. The complication rate was low. Mild hematomas formed at the surgical site in five patients, three of whom had inhibitors. A clot in an AV fistula occurred in one child with an inhibitor, but resolved spontaneously and the AV fistula remained function-

al. Of the 31 AV fistulae placed, four (13%) failed to mature or develop fully to allow adequate blood flow. In spite of these failures, high patient and parent satisfaction, excellent compliance with the prescribed treatments and acceptable complication rates were enough that Dr. Santagostino's group now offers the AV fistula as a first option for venous access in children with hemophilia when peripheral access is not feasible.

The types of anastomosis used in the study were brachiomedian or cubital (19), brachiocephalic (8), brachio basilic (3) and radiocephalic (1). The surgical evaluations were completed by a vascular surgeon, experienced in hemodialysis access. Patients were excluded if they were less than one year of age, or if they had cardiac or vascular disease. Suitable vascular sites had vessel size greater than 2mm, blood flow with arterial pulses, elasticity of the vessel walls and signs of poor venous drainage. The non-dominant arm was preferred and it was recommended that the vessels not be used for venipuncture until creation of the AVF. Distal vascular sites are preferred for adults; however, proximal vessels are more successful in children.

With surgery, an external catheter was placed and used for 6-8 weeks

while the fistula matured. Children received general anesthesia while adults received local anesthesia for the surgery. Prophylactic antibiotics were prescribed. The hospital stay was approximately 3-4 days and 6-7 days for an inhibitor patient.

Each AV fistula was evaluated monthly by physical exam and by Doppler ultrasound until maturation. Successful maturation was defined as sufficient dilatation and arterialization to allow vascular access for concentrate infusion. At 29 months of follow-up, 26 patients (96%) are regularly using their AV fistula at home. ❖

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