Northwest Community EMS System Continuing Education

Children with Special Health Care Needs

Student handout KEY – March 2012

Listen to the interview with Amy Julia Becker discussing her daughter, Penny recorded 2-17-12

What would it be like to walk in Amy Julia's shoes or anther parent of a child with special healthcare needs?

What particular challenges do they face?

What was Amy Julia trying to make us feel/see?

What are the limits of your knowledge about caring for children with special healthcare needs?

What do you need to understand better?

How are your views about these children shaped by your experiences, assumptions, or prejudices?

Another testament to the courage and devotion of a parent who just lost her special needs baby:

TUESDAY, FEBRUARY 14, 2012

Love.

Happy St. Valentines Day to all of you.

For me, today is extremely bittersweet. Today makes exactly **ONE month** that I've been without my baby boy. The thought of that physically feels like someone is ripping my heart out of my chest. Knowing that I haven't done a bath, a dressing change, said night-night prayers, or kissed those sweet lips in ONE whole month seems so unreal to me right now. A true nightmare.

But I'll tell you why today also makes me smile- because today is about LOVE. Right? And I know *all about* love. Tripp taught me love. He taught me love like I've never known it before. What I've learned MOST from him is *unconditional* love.

A love so strong that nothing can break it...not even death. A love that shines through pain, anger, and exhaustion, but also through times of complete joy and trust. Tripp taught me that every day counts- and that every minute matters. He loved me with his whole tired little heart every minute he was alive. Never once while he was alive did I think that my job as a mom was hard. I was doing what I was supposed to be doing- all I knew how to do, and all I wanted to do. He led me through every day and every hour by showering me with love like I've never known before. He was wise and holy beyond his years. I believe that with all my heart. I believe that he knew exactly what would happen in his time here on Earth- down to the very last minute. I believe he carried out God's plan for him, God's plan to show true LOVE.

A SPECIAL CHILD

You weren't like other children,
And God was well aware,
You'd need a caring family,
With love enough to share.
And so He sent you to us,
And much to our surprise,
You haven't been a challenge,
But a blessing in disguise.
Your winning smiles and laughter,
The pleasures you impart,
Far outweigh your special needs,
And melt the coldest heart.
We're proud that we've been chosen,
To help you learn and grow,
The job that you have brought us,
Is more than you can know.
A precious gift from Heaven,
A treasure from above,
A child who's taught us many things,
But most of all- "Real Love"

Paramedic National EMS Education Standard

Integrates assessment findings with principles of pathophysiology and knowledge of psychosocial needs to formulate a field impression and implement a comprehensive treatment/disposition plan for patients with special needs.

Knowledge objectives

Upon completion of the class and study questions, each participant will independently do the following with a degree of accuracy that meets or exceeds the standards established for their scope of practice:

1. Define children with special healthcare needs and technology-dependent children.
2. Discuss the general etiology of CSHCN and identify why this select population is rapidly growing.
3. Describe the technology devices that chronically ill children may require for survival.
4. Describe why it is important to listen to the caregivers of CSHCN.
5. Discuss why average age or tape-based weight and vital signs of "normal" children may be an inappropriate expectation for CSHCN.
6. Discuss the etiology, physical features, developmental delays, and common clinical conditions seen in children with Down syndrome.
7. anticipate accommodations that may be needed in order to properly manage a patient with Down syndrome.
8. Explain the purpose, indications, typical parts, types of tubes, elements to assess, and methods of troubleshooting a tracheostomy.
9. Describe the EMS management of a child with a trach who presents in respiratory distress or with one of the common complications related to tracheostomies.
10. Identify reasons why a child would require long-term ventilator support.
11. Describe types of ventilators and list possible causes and the EMS response for ventilator alarms.
12. Compare and contrast the various types of central venous catheters and implantable injection devices.
13. Describe possible indications, complications, and EMS care relative to a central venous line.
14. Explain the purpose, typical parts, and placement of a CSF shunt.
15. Discuss the management for signs and symptoms of increased intracranial pressure in a child with a shunt.
16. Compare and contrast the purpose, time of insertion, and placement of a naso or orogastric tube and G or J tube.
17. Describe aspects to be monitored, possible complications and EMS management relating to feeding tubes.
18. describe the various etiologies and types of hearing and visual impairments; recognize the patient with a hearing or visual impairment, and anticipate accommodations that may be needed in order to properly manage the patient.
19. Assess and manage CSHCN and technology dependent children based on the SOPs.
20. Appreciate the challenges faced by families of CSHCN and technology-dependent children and defend the need for consistent compassionate, competent EMS care.

Children with special health care needs (CSHCN) refers to children who have or are suspected of having a serious or chronic condition of physical, developmental, behavioral, or emotional health that requires health-related services of a type or amount beyond that generally required by children (based on national definition).

Technology-assisted/dependent children refers to those children who depend on medical devices to support bodily functions. These include, but are not limited to, ventilation devices, apnea monitoring/pulse oximetry, long term vascular access devices, dialysis shunts, nutritional support (i.e. gastric tubes), and elimination diversion appliances.

Why is this population increasing for EMS calls?

- Increased survival rates from children suffering from critical injuries or disease.
- Advances in medical technology allow more children into the mainstream.
- More plentiful support services has decreased the time spent in hospitals for these children.

Assessment caveats – General points

- Although the patient assessment is the same as for normal patients, there are special considerations in each part of the assessment that require additional attention by EMS.
- Use Initial Medical Care Assessment Steps

  Patients may have limited expressive speech and a tendency to clinically show development of new disease as a change in behavior. For example, if a patient was previously able to independently feed, dress, and perform personal hygiene but now requires assistance, look for a physical cause.
- Children with chronic illnesses often have different physical development from well children. Their size and developmental level may be different from age-based norms and length based tapes to calculate drug dosages may not be accurate.
- The baseline of these children may be different from others in their age group. Ask the family member or caregiver if there are deviations from this child's normal state with respect to mental status; vital signs, interaction with parents and strangers; verbal abilities; ability to sit, stand, and walk; muscle tone and strength.
- The best source of information about a CSHCN is the person who cares for the child on a daily basis. **Listen to the caregiver and follow their guidance regarding the child's treatment.**
- **Treat the child, not the equipment!** Attempt to determine if the emergency is related to an equipment malfunction and manage child appropriately using EMS equipment.
- Some CSHCN may have sensory deficits (hearing or visually impaired) but may have age-appropriate cognitive abilities. Follow the caregiver's lead in talking to and comforting a child during treatment and transport. Do not assume that a CSHCN is developmentally delayed.
- When moving a CSHCN, a slow, careful transfer with two or more persons is preferable. Do not try to straighten or unnecessarily manipulate contracted extremities as it may cause injury or pain to the child.
- Ask about the child's "Go Bag" that contains additional equipment such as extra trach tubes, adapters for feeding tubes, suction catheters, etc. Bring it with you.
- Ask the caregivers if they have an emergency information form or card or whether the child is enrolled in a medical alert program.
- Caregivers of CSHNC often prefer that the child be transported to the hospital where the child is regularly followed or the "home" hospital. When making the hospital destination decision, take into account travel time, ability to transport to locations other than the nearest hospital, the child's acuity, and family/caregiver wishes. Contact OLMC per usual and customary practice.

AIRWAY

- **Children with special needs**: These children should be assessed for airway obstruction from secretions. Many of these children are unable to adequately control their saliva; therefore, this may become an airway obstruction.
- **Technology Assisted Children**: Many of these children have special aids to help them breathe; e.g., tracheotomies, home ventilators, or constant positive airway pressure devices (CPAP) and are at risk for device occlusion, decannulation, malfunction or failure.

BREATHING

- Children with congenital heart disease or chronic illness may be unable to compensate for even mild respiratory distress.
- These children will decompensate rapidly. It is critical for the provider to recognize respiratory distress early and to intervene and transport as soon as possible.

CIRCULATION

- Signs of hypoperfusion may be masked because the baseline HR may be accelerated
- Children with circulatory conditions decompensate faster than healthy children
- Signs of hypoperfusion: worsening mental status, increasing tachycardia, exhaustion and tachycardia that deteriorates to bradycardia

TIME SENSITIVE if any instability at all

Down Syndrome – See Independent Study materials

My face may be different
But my feelings the same
I laugh and I cry
And take pride in my gains.
I was sent here among you
To teach you to love
As God in the heavens
Looks down from above.
To Him I'm no different
His love knows no bounds.
It's those here among you
In cities and towns
That judge me by standards
That man has imparted.
But this family I've chosen
Will help me get started.
For I'm one of the children
So special and few
That came here to learn
The same lessons as you.
That love is acceptance
It must come from the heart
We all have the same purpose
Though not the same start.
The Lord gave me life
To live and embrace
And I'll do it as you do
But at my own pace.

The Creed for Children with Down's Syndrome
Author unknown

What is **Down Syndrome**?
Most frequently occurring genetic chromosomal disorder. Most often, results from an extra chromosome on #21 (Trisomy 21) or 22 thus giving the person 47 chromosomes instead of the usual 46.

What are common physical features of children with Down Syndrome?

Low muscle tone, a single crease across the palm of the hand, almond shaped eyes that slope up at the corners; a large protruding tongue; small face and features; a slightly flattened facial profile with folds of skin at the sides of the nose that may cover the inner corner of the eye; short broad hands; and flattening of the back of the head.

What types of developmental delays should be anticipated & how should EMS obtain a history from these patients?

It causes delays in physical and intellectual development that range in severity from moderate to severe. May have limited expressive speech, and a tendency to manifest development of new disease as a change in behavior.

How can common clinical conditions seen in children with Down Syndrome present and how should EMS intervene?

Airway changes:
Soft tissue and skeletal alterations lead to airway obstruction, up to 50% may experience of obstructive sleep apnea. They also have a greater tendency to be obese than does the general population. Carefully assess airway/breathing.

Congenital heart disease:
Approximately 40% have congenital heart defects. From late adolescence on, there is an increased risk for mitral valve prolapse and valvular regurgitation. Some of the heart conditions require surgery while others only require careful monitoring. Anticipate changes in VS; monitor ECG.

Orthopedic issues/C-spine:
C-spines very susceptible to injury (Atlantoaxial subluxation): evidenced by gait disturbance, weakness, clonus, or spasticity. Careful spine motion restriction for all MOIs.

Neurologic conditions:
S&S of dementia often appear before age 40. Those who have dementia also have a higher rate of seizures. Disorders, such as depression, obsessive-compulsive disorder, abuse, and conduct disorder occur more frequently than other mental health disorders in individuals with Down syndrome. Explore new changes in mental status for a physical cause.

Immune system: *much more susceptible to infectious diseases like pneumonia*. *Careful use of BSI and aseptic technique.*

Vision: *Cataracts 4X more common; may begin to develop in adolescence. Ask about vision disturbances; provide verbal and tactile cues to assessment/care.*

Hearing: *>50% have mild-to-severe hearing loss. Conductive and/or sensorineural loss can begin at any age. Look directly at child, use hand signals.*

GI blockages, hypothyroidism, leukemia

Tracheostomy – See trach handouts

You respond for a child with difficulty breathing at home. On arrival, you find a 4 year old laying on a hospital bed amidst a tangle of tubes and monitors. Your assessment reveals an alert child with rapid breathing rate and increased effort, pale skin and gurgling sounds. The child has a tracheostomy from which the gurgling originates. Mother states that she had suctioned the child with no improvement so called 911.

Mother is visibly upset because the pulse oximeter has been fluctuating between 91 and 94%. Complete the worksheet below. Process through, what additional information is needed? What could be wrong with this child? What intervention is indicated?

What is a trach?	*A tracheostomy is a vertical or horizontal surgical opening into the trachea that is usually done in the OR under general anesthesia. It forms a temporary or permanent opening that bypasses the nose and mouth for ventilation.*
What is the purpose of a trach?	▪ *To bypass an upper airway obstruction; manage chronic aspiration* ▪ *Allow for long term ventilation*
What are the various indications for a trach?	▪ *Abnormalities of midface area* ▪ *Congenital Craniofacial Syndromes* ▪ *Blockage from polyps, tumors, hemangiomas, cystic hygromas* ▪ *Congenital malacia* ▪ *Upper Airway Obstruction - Vocal cord paralysis* ▪ *Subglottic stenosis from intubation* ▪ *Injuries head/neck that cause airway swelling* ▪ *Chronic aspiration due to muscle or sensory problems in throat* ▪ *Laryngotracheal separation* ▪ *Narrow or obstructed airways, bronchopulmonary dysplasia* ▪ *Neuromuscular diseases*
What are the parts of a typical cuffed trach tube?	***Adaptor**: Portion of the trach tube seen on the outside of the patient's neck* ***Flange**: Place to connect the ties* ***Tube or cannula:** part that goes in the stoma* ***Obturator**: Guide used during trach tube change* *Cuffed-soft **balloon** around the outside of the lower end of the cannula* ***Cuff inflation line -** Tubing that connects the cuff internally and pilot balloon* ***Pilot balloon** –External balloon that shows if cuff is inflated or deflated* ***Valve** – Spring loaded one way valve used to put air into or pull air out of the cuff*
What's the difference between a cuffed and an uncuffed trach tubes in terms of age groups and need?	▪ *Trach tubes may or may not have a cuff: Infants and young children generally have uncuffed tubes. Children > 8 years and those requiring positive pressure ventilation generally have cuffed tubes to prevent an air leak and decrease aspiration* ▪ *Tube selection is dependent on the size of the airway and the child's needs*
What different substances can be used to inflate trach cuffs?	*Air, water, sponge-like foam*
Do most small children have a single or double cannula trach tube?	*Single*
What should EMS do if there are frothy secretions, increased vocalization, Low pressure alarm sounds, and the pilot balloon on the trach is flat?	*These are S&S of a cuff leak* ***Intervention:*** ▪ *Withdraw any air or fluid from the cuff; instill the prescribed volume.* ▪ *Change trach if any of the above signs reoccur*
What should EMS assess about the stoma?	*Presence of granulation tissues, drainage, swelling, color change, skin breakdown.*
What elements of history are important to obtain relative to a child with a trach?	***History of present illness** – What is different today?; interventions taken prior to EMS arrival; child's baseline abilities; why the child has a trach, status of the upper airway; reason for a ventilator; if the child can breathe on their own; baseline VS; amount and route for home O2; suctioning frequency and what their normal secretions are like; devices and medications; medical information forms. Look for MedicAlert® jewelry or health forms.*

What should EMS do if the trach tube is dislodged?	• *Assess child's status and hyperextend the neck to gain access to the trach site* • *Establish a patent airway – always first priority* • ***If partially out**, immediately put tube back in stoma and tighten ties* • ***If completely out**, insert the spare trach tube – If the regular size doesn't fit, use one size smaller to keep the airway patent. If no spare trach tube is available, replace the one that came out.* *If you cannot reinsert the tube, observe child to see if they can breathe through the stoma alone. This may be possible if the stoma is well healed and fairly large. Child may be able to breathe through nose and mouth if there is no severe obstruction above the trach site. If distress persists. Provide manual ventilation as necessary*
What S & S suggest tube obstruction or the need to suction	• *Visible secretion at trach opening; rattling mucus sounds, dry raspy breathing or whistling noise from the trach* • *Respiratory distress, cough, increased RR/work of breathing, increased use of accessory muscles; retractions, flared nostrils; increased inhalation or exhalation time (capnography)* • *Reduced airflow through the tube; decreased/absent air entry/chest rise; decreased/absent or coarse breath sounds* • *Vital sign changes: Tachycardia, change in respiratory rate* • *Restlessness; child asks to be suctioned; child-specific signs that suctioning is needed* • *Change in color/cyanosis; diaphoresis, decreased oxygen saturation; hypercarbia* • *Inability and /or difficulty in passing suction catheter; resistance when trying to suction or ventilate* • *Difficulty or refusing to eat* *Alarms: High pressure alarm on ventilator; cardio-respiratory monitor alarms*
How should EMS determine the size and length of the suction catheter to use?	*Size: Depends on size of trach tube: Use largest catheter that fits the tube size. To estimate size: double the ID of the trach tube size. Ex: ID 3.5 trach tube X 2 = 7. Would lake size 6 suction catheter. Sizes 8 and 10 Fr most commonly used for children.* ***Length:** Pre-measure length using the trach tube obturator as a guide so that distal side holes are just past tip of the trach tube; suction only to end of trach tube.*
How should EMS determine the depth to insert a suction catheter?	*Insert catheter no more than 2 to 3 inches into the trach tube based on premeasured length above*
What is the maximum time that suction should be applied in a child?	*5 seconds at one time*
How should EMS change out a tube	*If cuffed tube: deflate balloon by connecting a syringe to the valve on the pilot balloon. Aspirate air until the pilot balloon collapses. DO NOT cut the pilot balloon or the attached tubing – this will NOT deflate the cuff.* *Cut ties that hold the trach tube in place. Remove tube using a slow, outward and downward movement.* *Gently insert a new same size trach tube with the obturator in place, in a smooth, curving motion directing the tip of the tube towards the back of the neck in a downward arc.*
What if a new tube cannot be inserted? How should EMS troubleshoot this situation?	*Attempt to pass a next smaller size trach tube. If unable to insert - thread a suction catheter through the smaller sized trach tube. Use the catheter tip to probe the opening; slide the tube over the catheter into the opening and then remove the catheter. Attempt to ventilate and check breath sounds.*
What should be in a "Go Bag"?	• *BVM with an appropriate sized mask* • *Same size and size smaller trach tubes; trach ties* • *Suction machine, suction catheters; DeLee suction catheters* • *NS; water soluble lubricant; scissors; tape; medical information*
What could cause respiratory failure requiring a ventilator?	*Increased respiratory load from chronic cardiopulmonary disease; ventilatory muscle weakness; decreased Central Nervous System (CNS) control*
What's the advantage of a volume controlled ventilator?	*Guaranteed amount of gas delivered regardless of lung compliance.*
What action is needed if the DISC/Sense alarm triggers?	*First check the child! Alarm means that the tubing is disconnected; check the trach tube and check all tubing to and from the vent; reconnect; check for kinks or occlusions*
Should EMS assess and communicate with the child based on their develop-mental or chronological age?	*Developmental*

Feeding tubes

Play the video showing a tube feeding

Purpose: Tube inserted to the stomach or duodenum that provides a route for nutritional support and/or medication administration delivered directly to the GI tract by artificial means when a child is unable to take food by mouth for an extended period of time.

Conditions for which gastrostomy tubes are used (EMSC reference card, Aehlert, p. 651)	
Swallowing dysfunction	*Esophageal burns or strictures*
Severe gastroesophageal reflux	*Chronic malabsorption*
Craniofacial abnormalities	*Severe facial injuries secondary to trauma*
Severe failure to thrive	*Neurological or neuromuscular – brain damage, muscular dystrophy*
Esophageal atresia	

Non-surgical tubes are used for short-term support of the child. They are generally used for < 30 days (ave. tube life 10 days). The catheter is inserted through nose or mouth into stomach or small intestine. For more information on NG tubes, see independent study materials.

Type of tube	What are the insertion and end points?
Nasogastric Tube (NGT):	*Runs from the nose to the stomach;*
Nasojejunal Tube (NJT):	*Runs from the nose to the small intestine. This catheter presents less risk of aspiration.*
Orogastric Tube (OGT):	*Runs from the nose to the stomach.*
Orojejunal Tube (OJT):	*Runs from the mouth to the small intestine.*

Long-term feeding catheters

Catheter are implanted through the skin into the stomach or small intestine when nutritional support will be needed for > 30 days. Possible placement techniques:

- Gastrostomy tube (GT) – percutaneous insertion (endoscopic or radiological) into stomach
- Jejunostomy tube (JT) – percutaneous insertion (endoscopic or radiological) into small bowel
- Percutaneous gastrojejunostomy [PEG/J] endoscopic or radiological
- Surgical gastrostomy
- Surgical jejunostomy

Types of devices

- **Gastronomy tubes** – often called by their manufacture's name. For example, the Malencot: has a feeding port and the proximal end of tube is mushroom tipped. All tubes have a mushroom tip or balloon on the inside of the stomach and a disk, clamp or crossbar is on the outside to anchor them in place. These tubes do not have an antireflux valve. They can be vented by uncapping the feeding port. Ex: MIC-G tube – shaft lengths from 0.8 to 4.5 cm with a port that can be closed when not in use. Printed marks on the tube assist in proper positioning.

- **Buttons** – Skin level device. Initially, G tubes are placed and then replaced with a button. Buttons should not be placed until firm adhesions between the gastric and abdominal wall are established to prevent gastric separation (3 months or longer if patient is malnourished or on steroids). Buttons have an antireflux valve that requires an adapter tubing to access. Bring this adapter to the ED with the patient. The button should be rotated 360° once a day to prevent entrapment into gastric mucosa (Adirim & Smith, 89).

The tube feeding is may be delivered into the stomach by bolus, gravity, or pump controlled. The nutrition is usually placed onto a pump device and slowly delivers the feeding over a specified amount of time. The method used depends on patient need and tolerance.

Preparing for transport: If fluids are infusing through the feeding tube, transport pump if there is space and a power source on the ambulance. If pump cannot be transported, stop infusing fluids, have the caregiver flush the tube with water and disconnect from pump prior to transport. The feeding tube should have a clamp that can be used after separating the tube feeding from the feeding tube. Make sure to turn the pump off after disconnecting the patient. Use care not to pull on the feeding tube. If fluid infusion was stopped within 30 minutes of transport time, transport child sitting up if possible.

Complications: What possible causes and actions are indicated to troubleshoot each of the following? (Special Children's Outreach and Pediatric Education, p. 88-90; Aehlert, Comprehensive Peds Emerg Care, p. 649-652; EMSC Reference Card and Prehospital Protocols, p. 21)

Problem	Possible causes – EMS intervention
Child has NG/OG tube and is choking, coughing, or has cyanosis.	*Cause: aspiration* *Action: Suction airway, give oxygen if hypoxic, monitor pulse ox and ECG, repeat primary survey frequently*
NG tube has become partially dislodged	*Assess respiratory and hydration status* *Action: Ask caregiver to check tube position. If position cannot be confirmed, remove tape from child's face; gently pull tube out from the nose or mouth; inform OLMC, bring tube into hospital*
Gastric distention	*Connect syringe to external opening of the feeding tube. Aspirate until resistance is met. If blood seen in aspirated contacts, contact OLMC.*
G/J tube displaced	*Action: Dry, sterile dressing over area* *Transport for reinsertion; Bring old tube to ED for sizing purposes.*
Tube is obstructed	*Cause: blood, crystallized feeding / medications, abdominal tissues* *Action: Transport for physician evaluation*
Nausea/vomiting/abdominal distention, cramping/diarrhea	*Cause: Too rapid feeding; feeding too cold; spoiled formula or change in formula* *Could also indicate peritonitis or perforation of stomach/bowel* *Action: Stop feeding, attach connector to the tube and leave tube open and draining into a container. Transport for physician evaluation* *Give IV (IO) fluids if S&S of dehydration or shock. Transport patient on right side or sitting up if not contraindicated to avoid potential aspiration.*
Leakage of stomach contents	*Cause: Broken or sticking valve; incorrect positioning of tube wedged into gastric mucosa; balloon not inflated properly; increased volume in stomach (feeding or gas); coughing, constipation; bowel obstruction, pneumonia, seizures* *Action: Cover with sterile dressing*
Bleeding from the site	*Cause: Skin irritation, pulling on tube* *Action: Control with direct pressure.*
Stoma site irritation or purulent drainage	*Cause: Maceration due to moisture; gastric acid burn from leakage of gastric contents; infection; excessive granulation tissue* *Action: Apply dry gauze dressing; transport for physician evaluation*
Gastric mucosa coming out of stoma	*Action: Treat like evisceration; transport for physician evaluation*
Feeding tube adaptor breaks	*Clamp tube and transport immediately; tube needs to be replaced*

Long-term Vascular access devices (VAD)

A 7-year old child with acute lymphocytic leukemia (ALL) who is currently undergoing chemotherapy presents with a fever and appears ill. His last chemo was four days ago. He has a Broviac central line though which he gets chemotherapy. (Adirim & Smith, 84).

1. **What are the primary concerns?**

   The child could have a serious systemic infection that could lead to septic shock. Assume that his immune system is compromised due to the chemotherapy. The Broviac catheter is a foreign body that can introduce bacteria directly into the bloodstream.

2. **What other important elements of history and the physical exam to obtain?**

   T 102° F; RR 40; HR 160; BP 100/60

3. **How should EMS care for this patient?**

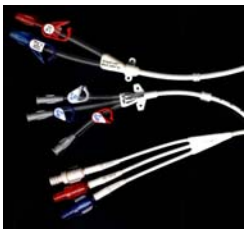
   The child may be anemic and hypoxic; give O2 as needed
   Patient needs IVF 20 mL/kg bolus. If need to access the central line- see below for procedure.

Central line catheters are very common in the home care setting. They are used to administer medications that have to be given over a long period of time such as some antibiotics and chemotherapy, and as a route for hemodialysis, plasmapheresis, frequent transfusions, hydration, and Total Parental Nutrition (TPN).

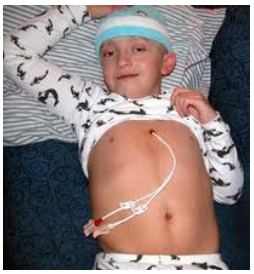
SOP: If peripheral vascular access unsuccessful or not advised, may use central venous access devices already in place based on OLMC.

Types of partially implanted devices

Multilumen central line catheters (Hickman, Broviac)

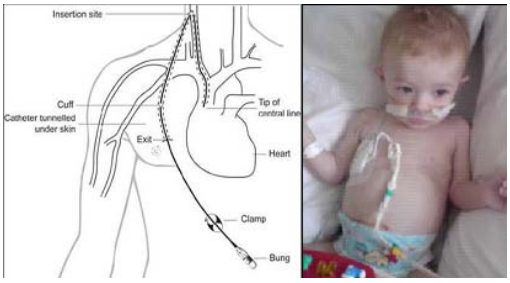


These are blind or closed end devices that have a removable cap on the external end of the catheter and a clamp on the tubing distal to the clamp that closes flow to and from the vein. (Adirim & Smith, 85) The external end of a partially implanted catheter may have a number of ports or lumens. Catheters are usually configured into single, double or triple lumen devices. The number of lumen depends on the intended use of the catheter and the patient's needs. The catheter gauge is marked in millimeters on the side of the lumen.



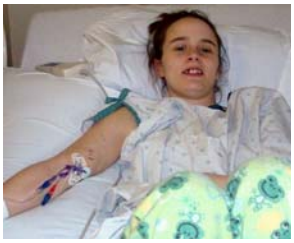
The state of the art in radiology has evolved to the point where image-guided placement of tunneled catheters such as perm-catheters for dialysis, PICCs, Hickman catheters and implantable port catheters is often preferred to surgically placed catheters. Ultrasound and fluoroscopic guidance allows safe, precise placement with less complications than the use of standard surgical landmarks. Also, the same interventional radiological approach eases catheter and port repair, if needed.

The Hickman catheter is made of silastic (a silicone elastomere) that is softer than a simple triple-lumen catheter. The Hickman catheter comes in double-lumen and triple-lumen varieties. These catheters can stay in place for weeks to months; some patients have had the same Hickman catheter for years!

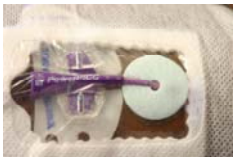


Surgeons often access the subclavian vein under the clavicle and place the distal catheter tip at the SVC/right atrial junction or in the right atrium, but the distal end of the catheter is pulled under the skin for 2-4 inches and comes out of the chest close to the nipple or over the sternum. This creates a "tunnel" which decreases the risk of infection. Interventional radiologists more often select the internal jugular vein for central catheter placement. It runs straight down to the superior vena cava, which reduces the risk of malposition of the catheter and possibly also of central venous obstruction.

Peripherally Inserted Central Catheters (PICC)



These central lines are long, flexible, silicone catheters that are usually inserted into the basilic or cephalic vein via the antecubital space. The catheter is advanced into the superior vena cava. It is the most common external central venous line. Their use is associated with a lower rate of infection than other partially implanted central lines. They can remain in place for up to a year.



EMS providers should not take blood pressures in the arm with a PICC line.

Accessing a central line:

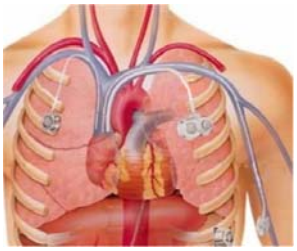
This process needs to be done with strict aseptic technique. Be very careful not to introduce bacteria into the central line, as they can cause a life-threatening infection. Also, if improperly done, the patient is at high risk for an air embolism. Ask the caregiver if the patient received TPN (total parenteral nutrition) through the central lines. TPN is incompatible with a multitude of medications and the line should be flushed first.

If there is an appropriated trained professional, they should be the ones that access the central line.

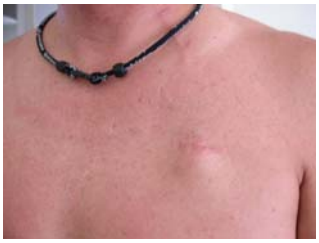
- Wash hands with soap and water or waterless cleaner; apply sterile gloves and a surgical mask. Also have the patient turn away from the access site.
- Cleanse the injector cap with chlorhexidine
- Close the clamp on the tubing 3" from the cap prior to removing the protective injection cap
- Remove the protective cap
- Secure a 10 or 12 mL syringe prefilled with 5 mL NS onto the injection port site of the central line.
- Unclamp the catheter and aspirate 5 mL of blood from the line. Reclamp the catheter and discard the aspirate into an appropriate container for disposal at the hospital.
- If blood can be aspirated, secure a new syringe filled with 10 mL NS, unclamp and slowly infuse 5-7 mL of NS into the catheter to ensure patency. Reclamp the catheter and remove the syringe.
- Attach the well-primed IV line to the injection port – secure with tape and unclamp the line to begin IV flow. Be careful not to introduce any air into the catheter and the IV tubing and bag must NEVER run dry.

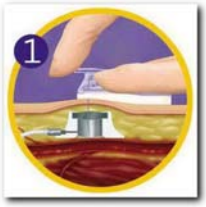
Surgically implanted medication delivery systems

Portacath, Medi-port, LAS Port ®



Portacath is a generic term that describes any implantable venous access device. A small intravenous catheter several millimeters in diameter and 20 – 25 cm in length is inserted into a large vein behind the clavicle. The catheter is connected to a "port" that is shaped like a disc and is about the diameter of a 25 cent coin. It has a flexible silicone membrane injection port that is self-sealing. The port is then totally implanted into the subcutaneous tissue into the subcutaneous tissue in the left or right pectoral regions where it can be easily felt and accessed. Patients receiving chemotherapy will often have a portacath.





Administration of an IV medication or collection of a blood sample is both simplified and made much less uncomfortable when a portacath is available. They must be accessed with a noncoring needle that has a solid tip and side hole opening. It is recommended that this injection needle has a 90 degree angle. A regular IV catheter or metal needle will permanently damage the port and prevent proper resealing of the port when the needle is removed.

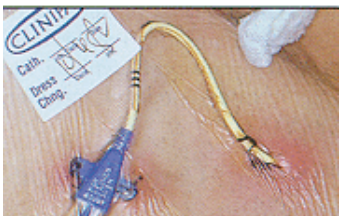
Assessment issues (DOPE) (ESMC reference chart, Aehlert,)

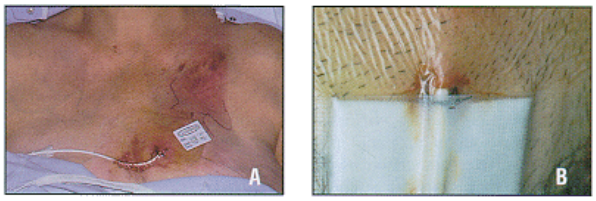
D	*Displaced: Total or partial dislodgement or movement out of the vein into internal tissues*
O	*Obstructed: Blood clot, protein, crystallized medications / IV nutrition*
P	*Pericardial tamponade due to perforation by catheter* *Pulmonary problems: Pneumothorax, pulmonary embolism from clot or catheter shear*
E	*Equipment: tubing kinked or cracked, infusion pump failure*

VAD complications

- **Hemorrhage**: Most patients with a VAD are on some type of anticoagulant therapy to prevent the device from developing obstructing clots. These medications place the patient at risk for increased bleeding (GI and intracranial bleeds). If external bleeding, apply direct pressure to site.
- **Air embolus** - clinical presentation
  - Headache; dyspnea; chest pain
  - Altered mental status; hypoxia

  If air embolus is suspected:
  - Place the patient in left lateral Trendelenburg position.
  - 15 L O_2
  - ECG, pulse ox, capnography monitors
  - Time sensitive patient





Figs 2A and 2B. — Tunnel infections. Note the presence of inflammation along the subcutaneous track of the device greater than 2 cm from the exit site.

- **Infection** of catheter site, catheter tunnel, or system wide sepsis. Assess the access site for redness, swelling, drainage and tenderness. Provide supportive care (IVF, vasopressors) and transport. Tunnel infections occur earlier than device-related bacteremia or fungemia and can be associated with serious local morbidity or death (Figs 2A and 2B). Tunnel infections almost always require catheter removal at the hospital.

  Central venous septic thrombophlebitis is a potentially lethal complication that can be successfully managed at the hospital with prompt catheter removal and IV antimicrobial therapy. The patient can experience a septic pulmonary emboli from seeding of an infected intravascular clot.

Ventricular shunts

Chance's story:

Thursday, September 29, 2011

"There is a sacredness in tears. They are not the mark of weakness, but of power. They speak more eloquently than ten thousand tongues. They are messengers of overwhelming grief…and unspeakable love." ~Washington Irving

"I find myself a little perplexed to find the words to once again write to update you all on my baby boy having another surgery. As I sit here in a silent room hearing nothing but hospital beeps surrounding me and watch Chance recover from another surgery I feel a little overwhelmed with what seems to be a never ending struggle with his shunt. I've sensed before we left the hospital this past weekend that something wasn't quite right but I tried to remind myself that he was recovering still from surgery and should be understandably more fussy at times. As the last few days have passed I've noticed him *more agitated* and that *his soft spot was gradually becoming more full* at times throughout the day. My intuition, unfortunately, has rarely failed me when it comes to my baby boy and for the last few days I could just sense we would be here. I know some might perceive it as me thinking negatively…*but I can't ignore the instincts that something was wrong.* I suppose reflecting on the day might somehow help me…but also help update the many of you who have come to adore my sweet baby boy.

Today along with the fact that he had been *more fussy, lethargic*, and having a more full soft spot, he once again *projectile vomited.* That is usually the one thing that finally sets me off and makes me realize there is no second guessing. I just know there's a *shunt malfunctioning* with all the symptoms I saw today combined.

So I took him to the ER and we waited. It was so busy this afternoon. He underwent a CT scan which showed that his ventricles were larger…essentially that the hydrocephalus (fluid) on his brain was increasing which is typically a sign that his shunt is not working as it should.

Shortly after the CT scan the neurosurgeon came into update me and tapped the shunt. I held him down as they tapped the shunt and valve to see if fluid would flow through either. Unfortunately the fluid was very sluggish which was another sign that his shunt had malfunctioned again. We knew at that moment that Chance would have another surgery. Seeing him a moment after they tapped his shunt left me speechless…*here he is in a daze and totally not himself*.

Everything from that point forward happened so quickly. They bathed him. Put in an IV. And we left the ER fairly quickly as he headed on to get ready for surgery #10.

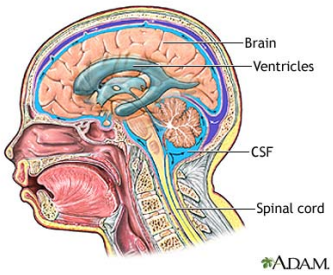
After surgery the doctor came out to update us. Surgery went well but Chance currently does not have a shunt. Yes, you read that correctly. His doctor completely removed his entire shunt. The catheters and the shunt valve were all clogged because *any ounce of blood that gets into the device can cause it to easily clog*. His doctor said he could have tried to put in another shunt but because the CSF came out pink, he has no doubt that this shunt problem would only continue if he didn't give his head time to heal.

Jason and I both are feeling a little overwhelmed. Chance's doctor said we will be here at the hospital at least 4 to 5 days. Though he doesn't have a shunt he does have a tube coming from the back of his head that is currently draining the excess CSF that builds up due to his hydrocephalus that he has due to having Spina Bifida. The hope is that in the next 4 to 5 days his head will have had enough time to recover and that they will be able to have a fresh start in putting in a shunt. Yes, that means he definitely does have to have another surgery.

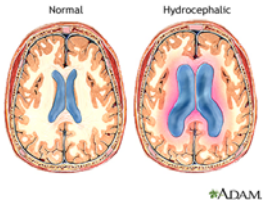
As if the typical medical wires he's connected to aren't enough, he now has an even more important wire that is connected directly to his head draining the fluid. The wire drains the fluid into a device connected to a bag. As you can see, the CSF is definitely pink…likely due to being mixed with small amounts of his blood after surgery. In just the 1½ hours we were in the PICU, 24 ccs of CSF was drained. Thankfully relieving this pressure seems to leave him more comfortable thus far too."

http://www.amandakern.com/blog/2011/09/more-shunt-woes/

Purpose of a shunt



Cerebral spinal fluid is produced in the choroid plexus of the first and second ventricles of the brain. It circulates through the 3rd and 4th ventricles and then around the brain and spinal cord in the subarachnoid space. It is reabsorbed in the dural sinuses of the brain.



Hydrocephalus (too much CSF) develops when there is an interruption of this normal circulation due to an increase in CSF production, obstruction of CSF flow, or a decrease in CSF absorption. The ventricles become enlarged with cerebrospinal fluid. This condition causes the brain tissue to become compressed against the skull, causing serious neurological problems. It often begins in infancy and may co-exist with many congenital and acquired disorders such as myelomeningocele, intraventricular hemorrhage, and infection (Aehlert, 645). Other indications include post- meningitis, brain injury, surgery, or tumors.

Untreated, hydrocephalus is fatal. Shunting is necessary to drain the excess fluid and relieve the pressure in the brain. This should be done as soon as hydrocephalus is recognized to give the child the best possible neurological outlook.

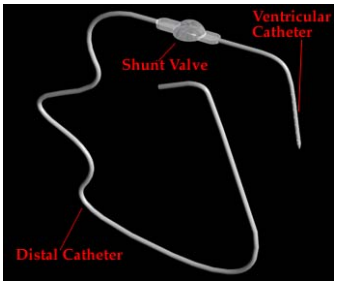
What are the S&S of hydrocephalus based on age (Aehlert, 645)

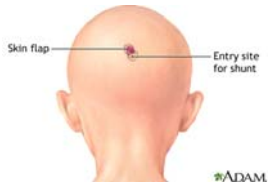
Infants	*Irritability, lethargy, vomiting, full fontanelle, head circumference that is larger than normal*
>1 year; toddler	*Head circumferences changes more slowly; irritability, vomiting, lethargy, large ventricles seen on brain imaging*
Older children	*Headache before other S&S of increased ICP*

Purpose: Shunts are inserted to drain excess CSF and reduce ICP.

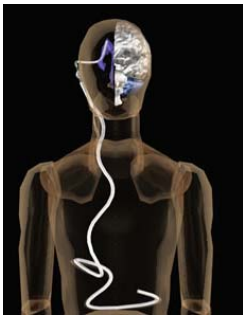
Location: They are surgically implanted into the brain to drain CSF from the ventricles into another part of the body.

Typical device: All shunts have three basic parts: a Ventricular (proximal) Catheter, a Shunt Valve (one-way valve system) and a Distal Catheter. They may have on-off valves, an antisiphon device, and a reservoir.





The shunt is inserted in the OR under general anesthesia. A flap is cut in the scalp and a small ("Burr") hole is drilled in the skull. The Ventricular Catheter may enter from various positions of the skull, most commonly from the right front top of the skull or from the right back side of the skull just above and behind the ear. This allows the catheter to pass through a relatively silent portion of the brain which minimizes risk of complications.



The ventricular catheter attaches to the one way valve which is placed under the skin on the outside of the skull. From the valve, the distal shunt tubing is tunneled underneath the skin down to a body cavity. The fluid is released into this cavity and absorbed.

Shunts are named for the position of their proximal and distal catheters. The proximal catheter is usually in one of the ventricles. The distal catheter is tunneled under the skin and is most often in the peritoneal cavity (ventriculoperitoneal [VP] shunt). It could be placed in the right atrium (ventriculoatrial [VA] shunt); pleural cavity (v-pleural shunt), gall bladder, ureter, urinary bladder, or thoracic duct.

In VP shunts, an extra length of distal catheter is placed into the peritoneal cavity to allow for growth (Aehlert, 646). Some patients have more than one shunt which may or may not be connected; others may have old nonfunctioning shunts that have not been removed.

VP shunts are prone to complications

The most common causes of shunts not working correctly are mechanical complications (shunt hardware malfunction), over drainage or under drainage of CSF fluid, bleeding and infection.

S&S malfunctioning shunt:

In general, when a patient's shunt is not working properly, they will develop the same or similar complaints that they had prior to having the shunt placed. Family members will usually notice these problems before health care providers can pick them up since initially, they can be intermittent and very subtle.

What S&S suggest a malfunctioning shunt with an increase in intracranial pressure? (Aehlert, 646) Ask caregiver for changes from normal behavior.

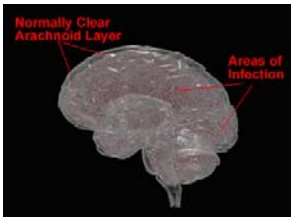
Irritability; lethargy, behavioral changes, "just not acting right"; Memory deficits
Headache, neck pain; apnea or change in respiratory pattern
Nausea, vomiting (projectile)
Bulging or full fontanelle in infants
New onset seizures or a change in the child's seizure pattern
Downward deviation of eyes (sunsetting)

First few months after surgery: Misplacement, disconnection, migration (identified on x-rays -skull, chest, abdominal films).

Causes of CSF shunt malfunction (DOPE) (Aehlert, 648)

D	*Displacement (catheter migration), disconnection of shunt components, drainage – over or inadequate*
O	*Obstructed or fractured catheter, kinking of distal catheter*
P	*Perforated abdominal viscus, peritonitis, pseudocyst*
E	*Erosion of the equipment through the skin*

Infection: "Shunts can become infected at the time of surgery or via the body fluids such as the blood stream. Infections can be very serious and can lead to strokes, paralysis, or even death. Many times the shunt will stop flowing due to debris from the infection. One may see redness, swelling, or tenderness along the path of the shunt. If the abdominal end becomes infected one may develop peritonitis with severe abdominal pain and fever. If the shunt is in the lung, the patient can develop pneumonia. If the shunt is in the heart, the patient may develop endocarditis. If the brain end becomes infected, an infection of the brain can lead to meningitis as depicted on the left. The patient may present with fever, confusion, headache, a stiff neck, some or all of these." http://neuroanimations.com/Hydrocephalus/Shunts/shunt_malfunction.html)



Malfunction: Any of the shunt parts may develop sub-optimal or incomplete function. If you look at the ventricular catheter closely, there are tiny holes in it which can become obstructed with tissue growing into the holes. This results in obstruction of CSF flow. Some tissue debris may flow into the valve and obstruct flow as well. The valve is a mechanical device and it is subject to malfunction just like any other mechanical

device. Finally, the distal catheter end may become obstructed by tissue. It may also migrate out of its cavity due to movement of the surrounding tissue environment (lungs, heart, bowel). (http://neuroanimations.com/Hydrocephalus/Shunts/shunt_malfunction.html)

After several years, failure often results from damaged tubing, overdrainage, or erosion of the equipment through the skin or into an abdominal organ. Perforation of the stomach or intestine may present as peritonitis. Surgery will be necessary.

Overdrainage:. Shunts may drain too much fluid or too little fluid. If they drain too little, the patient suffers with the hydrocephalus symptoms that person had initially to various degrees. If the shunt drains too much CSF, the patient may develop "spinal headaches". These are headaches which occur when the patient is sitting up, or moving about and resolve fairly quickly when the patient lies flat. Over drainage of shunts can lead to subdural hematomas. As child spends more time in an upright position, a siphoning effect on the distal tubing can put negative pressure across the valve, resulting in excessive drainage

Pseudocyst: May develop when bacteria enter the peritoneal cavity via the shunt catheter. This can cause an inflammatory response that involves the greater omentum that wraps around the distal tip of the shunt catheter and seals off the catheter outlet. The resulting cyst fills with CSF, causing abdominal pain and recurrence of the hydrocephalus (Aehlert, 648).

What EMS actions are indicated for a malfunctioning or infected shunt? (Aehlert, 648)

• *Vomiting precautions; ensure suction equipment readily available*
• *Give O2 as needed, assist ventilation as necessary, be prepared to intubate*
• *If shock or hypotension present: IVF 20 mL/kg bolus; may need epi*
• *Check blood glucose; give dextrose as needed*
• *Treat seizures if indicated*
• *May need to briefly hyperventilate (EtCO2 30-35) if S&S of brain herniation: unconscious, unequal pupils, fixed, dilated, non-reactive pupils; increased SBP, slow pulse*

Children with SPECIAL HEALTHCARE NEEDS (CSHN)

- Track CSHN in your service area and become familiar with the child and their anticipated emergency care needs.
- Refer to the child's emergency care plan, if available. Is the current presentation significantly worse than their baseline? Caregivers are best sources of info on meds, normal baselines, functional levels/normal mentation, usual color and RA SpO_2 readings, likely medical complications, equipment operation and troubleshooting, and emergency procedures.
- **Assess in a systematic** and thorough **manner**. Observe for ↑ or ↓ RR, use of accessory muscles, retractions, cyanosis, extremity edema, hydration status; palpate for ↑ or ↓ HR, decreased peripheral pulses, cool extremities, poor cap refill; listen carefully for crackles or wheezes. If child has known paralysis carefully examine extremities for injury.
- Be prepared for differences in anatomy, physical & cognitive development, and possible surgical alterations or mechanical adjuncts. **Common home therapies**: respiratory support (O_2, apnea monitors, pulse oximeters, BiPAP/CPAP, mechanical ventilators, chest physical therapy vest), IV therapy (central venous catheters), multiple meds, nebulizer machines, feeding tubes and pumps, urinary catheters or dialysis (continuous ambulatory peritoneal dialysis), biotelemetry, ostomy care, orthotic devices, communication or mobility devices, or hospice care.
- Communicate in an age-appropriate manner. Maintain communication with and remain sensitive to parents/caregivers & child.
- Ask parents for child's daily medical record notebook or medical information form to take to hospital. Ask caregiver to accompany EMS to hospital to continue assisting w/ child's care if possible.

BLS Interventions:

1. **Assess and support ABCDs**: Closely monitor airway, RR, HR & mental status. Support airway of those who have difficulty handling oral secretions (severe cerebral palsy, mental retardation). Provide O_2 (or manual resuscitation) when indicated. If child normally has a bluish color or SpO_2 <90%, use extreme caution in giving O_2. Give just enough to return to normal baseline.
2. Suction the nose, mouth, or tracheostomy tube as needed.
3. **Positioning**: place in position of comfort. If "*tet spell*" from tetralogy of Fallot, may put in knee chest position to increase systemic resistance. If shunt failure; sit up if possible to ↓ICP. Protect weak or paralyzed limbs. Do not attempt to straighten contracted extremities. Support with pillows/ towels in a position of comfort. Most respond best to slower movements & secure contact.
4. Flashing ambulance strobe lights can trigger a seizure in a child w/ known seizure disorder. Cover their eyes or turn off lights, if safety allows, when moving child in and out of the ambulance.
5. Technology-assisted children may experience an emergency if equipment fails to function. Use EMS equipment to support child.

ALS Interventions

6. Consider need for **intubation** if in respiratory failure
7. **Vascular access** if IV meds or fluids needed. If chronic cardiac condition: IVF only per OLMC. NS 20 mL/kg IVF bolus if hypoperfused. If on anticoagulant like Coumadin (warfarin), use caution when starting IV or when handling the child. They bruise easily and may have difficulty clotting.
8. Avoid placing **defib pads** over internal pacemaker generator (usually found in upper chest).
9. Consider use of inotropes (**dopamine 5 mcg/kg/min**) w/ severe hypotension unresolved with fluid boluses.
10. Rx **seizures** per SOP; monitor ECG as arrhythmias may be present in CSF shunt failure.
11. **Decompress stomach** by venting (opening) feeding tube if abdomen is distended.

Chronic respiratory or cardiac problem notes:

- If older than 6 yrs and has a peak flow monitor at home, ask child to blow into monitor to determine current reading. If < 50% "personal best" or unable to blow into the meter, child is in severe distress.
- Ask caregiver if any meds have been given in last 2 hrs to reverse respiratory distress. If yes, monitor for med effects. Base further management on therapies already given at home.
- If infant receives home O_2 therapy of 2 L or less by NC and presents in respiratory distress, do not give more than 2 L/NC. Increase O_2 delivery with blow-by O_2 or placing a facemask at no less than 6 L/min over child's nose & mouth.
- Take appropriate steps so child does not inhale noxious fumes from running ambulance.

Osteogenesis Imperfecta: Use extreme caution when moving child or taking BP. Use a draw sheet. Hare traction contraindicated. Pad between stretcher straps and child. Drive cautiously. Avoid sudden jolts that could cause a fracture.

Sickle cell disease:

- Vaso-occlusive crisis is very painful. Place warm compresses over swollen joints. Request OLMC orders for pain med.
- Very susceptible to infection d/t malfunctioning spleen. Fever, abd pain , S&S of stroke suggest a medical emergency.
- Vascular access challenging d/t frequent sticks. Give 20 mL/kg IVF bolus if signs of shock.

Hemophilia: Bleeding will not stop w/ conventional methods. Needs missing clotting factors at hospital.

Leukemia: Fever is an emergency; immune system is suppressed. Wear masks and gloves when caring for pt.