

Delivery of Prophylactic Factor to Children with Severe Haemophilia: The Challenge of Venous Access

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What is prophylaxis?

- The regular delivery of factor to maintain a baseline $> 1\%$
- Injections of factor given 1, 2 or 3 times a week



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Why?



- To prevent joint damage long term
- To reduce the risk and frequency of other serious bleeds
- To give the family better control over the disorder
- Avoid waiting time in Emergency Departments
- Enable immune tolerance for inhibitor treatment



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When?

- Variable starting times
- After first or second joint bleed
- After major bleed eg intracranial bleed in a baby
- Usual range 18-36 months

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How?



- Factor given intravenously
- Network of (blue) veins carry blood back to the heart



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Treatment Record Book: Record!

DATE	SITE OF INJURY	TREATMENT AND DOSE GIVEN	STICKER(S) FROM FACTOR BOTTLE	COMMENTS



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Venepuncture vs CVAD aka Port-a-cath, Hickman, Central line



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EDUCATION AND SUPPORT



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VENEPUNCTURE



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Challenges

- Patient has terrible veins
- Parent scared or apprehensive about needing their child ± needle phobia
- Which parent will learn first?
- Single parent “doing it all”
- Learning to set up the equipment & draw up the factor



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Challenges

- Pain



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Challenges

- Patient scared or apprehensive or not able to sit still for period of time
- Parent feeling guilty about “missing” the vein & causing bruising
- Anxiety about not being able to stop the bleeding if they “miss” the vein



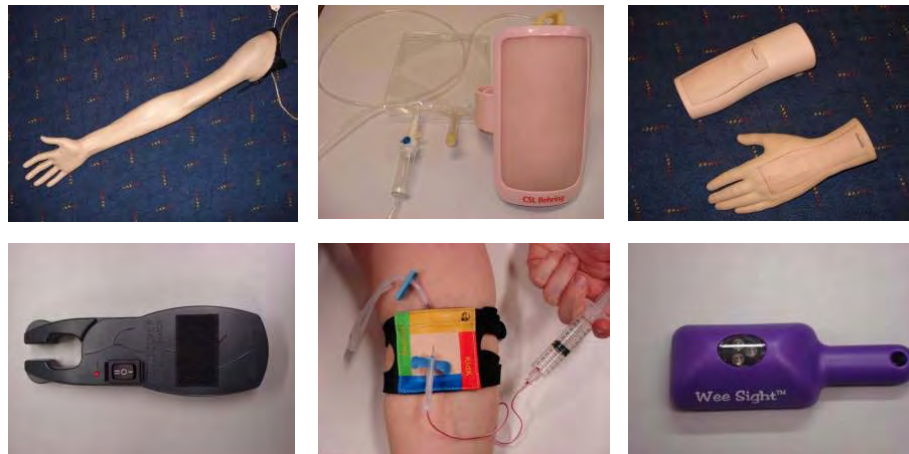
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Equipment



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Distraction



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Success



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CVAD

CENTRAL VENOUS ACCESS DEVICE

aka Port-a-cath, Hickman,

Central line



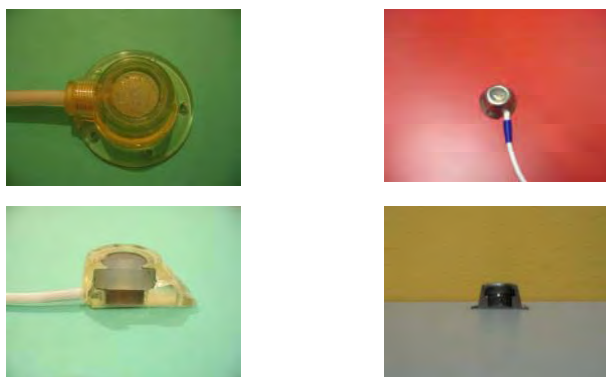
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Types of Port-a-caths



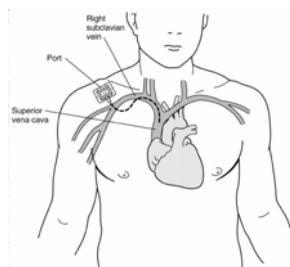
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Positions



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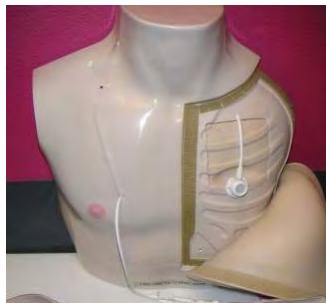


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Equipment and Education

- Chester practice dummy
- Observation
- Practice in hospital to master technique
- Lots of equipment
- Risk of complications



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Success



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Reasons to insert a CVAD

- Doctors (and parents) are unable to put a butterfly or cannula into the child's veins
 - Typically a baby, infant or toddler
- Dangerous situation
 - Unable to treat bleeds
 - Uncontrolled bleeding → even harder to cannulate
 - Patient in remote areas at greater risk
- Child undergoing immune tolerance



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Insertion of the CVAD

- General anaesthetic for insertion
- In theatre, under sterile conditions, performed by a surgeon
- Inserted into a big vein in the neck
 - Eg external jugular vein
- Risk of bleeding so done under Factor cover for minimum 3 days



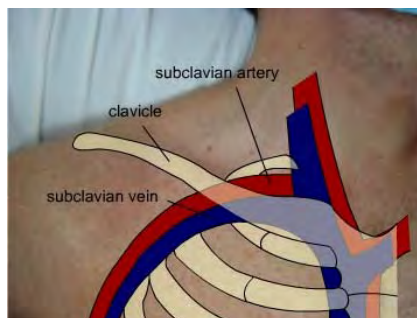
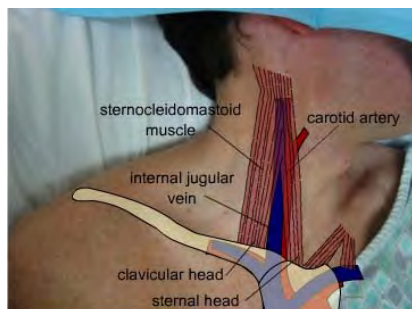
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- Insertion sites for a CVAD



<http://www.google.com.au/imgres?q=PORT-A-CATH+CELLULITIS&hl=en&gbv=2&biw=986&bih=411&tbm=isch&tbnid=fQfLOXUkCaXLVM:&imgrefurl=http://sfgh.medicine.ucsf.edu/education/reesed/procedures/centrallines/&docid=Z5vOwPBHhgOFM&imgurl=http://sfgh.medicine.ucsf.edu/education/reesed/procedures/centrallines/subclavian.jpg&w=360&h=270&ei=79-hTo504GaiQe0-JCS9g&zoom=1&iact=rc&dur=292&sig=114891575858799091724&page=11&tbid=83&tbw=117&start=139&ndsp=13&ved=1t:429,r:4,s:139&tx=77&ty=37>



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Complications

- Bleeding
- Infection
- Thrombosis
- Blockage & other mechanical problems
- Child can outgrow the line



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Bleeding

- At the time of insertion
- At site where needle goes in
- Direct trauma
- Haematoma increases the risk of infection



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Infection

- Commonest complication
 - Organisms: 3 main classes
 - Gram positive cocci eg
 - Staphylococcus aureus
 - Coagulase negative staphylococcus
 - Gram negative organisms
 - Pseudomonas
 - Klebsiella
 - Fungal infection
 - Other bacteria



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Infection

- Up to 40% of CVADs become infected with bacteria
- Sometimes CVAD may need to be removed to bring infection under control
 - Eg cellulitis in surrounding skin
- Sometimes possible to eradicate infection with antibiotics and “save “ the port
- Sometimes it “comes back” and then have to reconsider removing the port



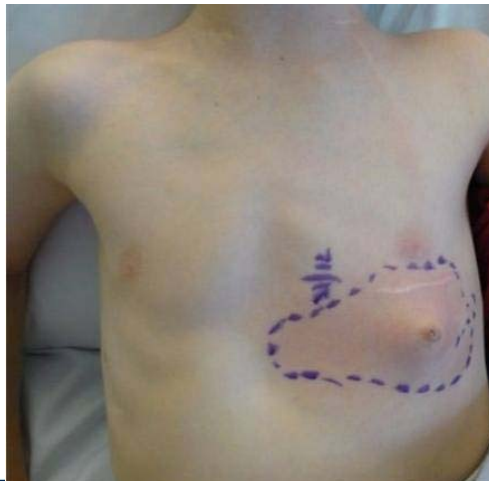
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Cellulitis



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Infection

- If your child has a fever $\geq 38.5^{\circ}\text{C}$
- \pm redness, swelling & pain around port
 - Contact HTC medical staff immediately
 - Go to nearest emergency department
 - Port accessed, FBC & blood cultures will be taken
- **Infection in a CVAD can be life-threatening**



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Infection

- Broad spectrum intravenous antibiotics will be started and given for 48 hrs until blood culture results known
- If organisms found in blood the correct antibiotics will continue for a minimum of 1 week
- A decision will be made about possible removal of the port



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Staphylococcus Aureus & Coagulase Negative Staphylococcus



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Thrombosis

- It is possible for a child with a bleeding disorder to form a clot within and at the end of a CVAD
- Pulsating flush and positive locking of the port help prevent this
- Significant thrombosis may require removal of the port and sometimes anticoagulants



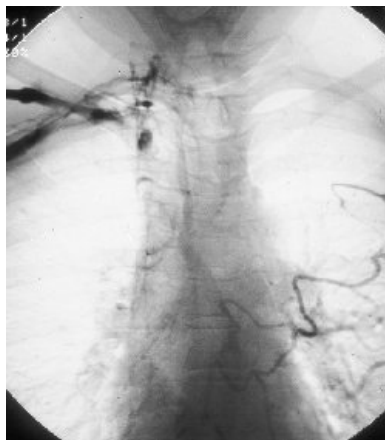
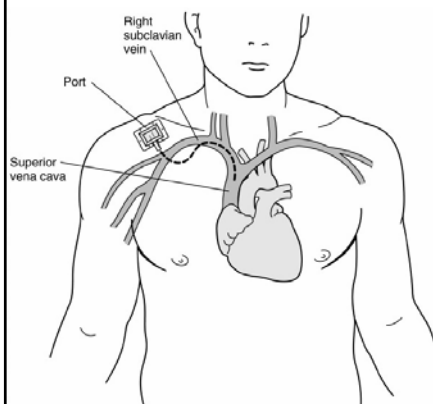
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Thrombosis



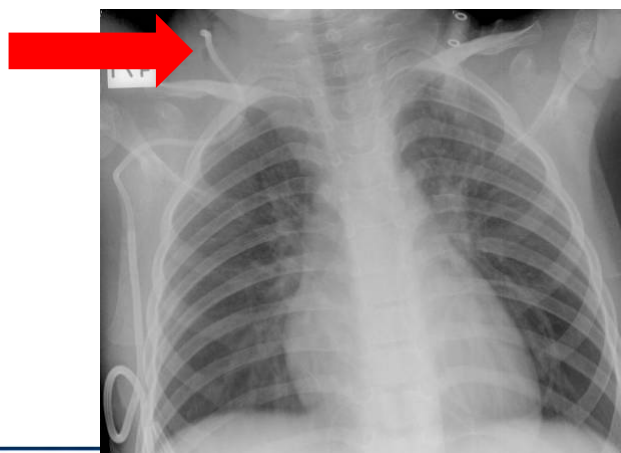
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Mechanical problems



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In summary

- Venepuncture first choice for delivery of prophylaxis
- CVAD if venepuncture not feasible
 - Despite problems can be life saving
- Removal of CVAD
 - Complications
 - Veins able to be accessed; parents able to be taught



When do you come to your HTC or ED?

- Child bleeding and you have missed the vein
- Port-a-cath accessed but cannot infuse
- Fever $\geq 38.5^{\circ}\text{C}$, even if you think your child only has a cold
- Skin changes around the port ie redness, swelling & pain

