

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

Robyn Shoemark

Clinical Nurse Consultant Haemophilia/Haematology

Dr Susan Russell

Consultant Paediatric Haematologist/Oncologist



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

What is prophylaxis?

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



The Sydney Children's Hospitals Network
(Randwick and Westmead)

When?

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

2 0 1 1			
January	February	March	April
S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31
May	June	July	August
S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31
September	October	November	December
S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30	S M T W T F S 1 2 3 4 5 6 7 8 9 10 11 12 13 14 15 16 17 18 19 20 21 22 23 24 25 26 27 28 29 30 31



The Sydney Children's Hospitals Network
(Randwick and Westmead)

How?



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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Treatment Record Book: Record!

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

EDUCATION AND SUPPORT



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

VENEPUNCTURE



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Challenges

- Pain



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



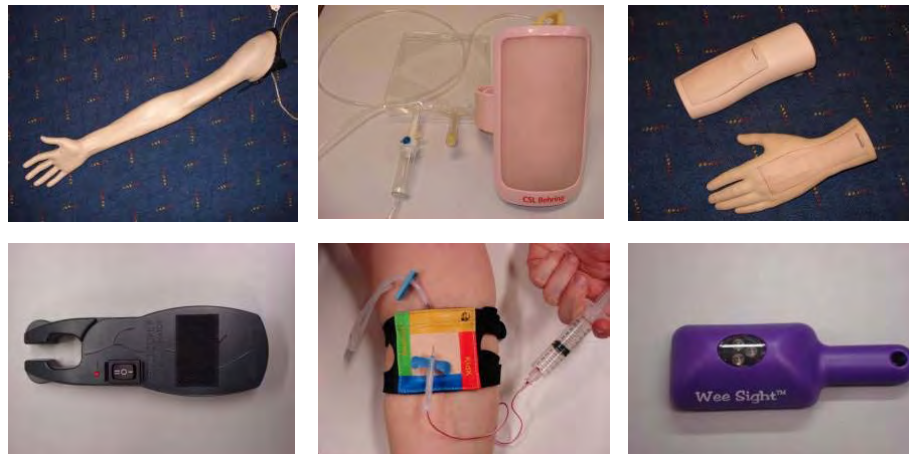
+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Equipment



+



=

The Sydney Children's Hospitals Network (Randwick and Westmead)

Distraction



+



=

The Sydney Children's Hospitals Network (Randwick and Westmead)

Success



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

CVAD

CENTRAL VENOUS ACCESS DEVICE

aka Port-a-cath, Hickman,

Central line



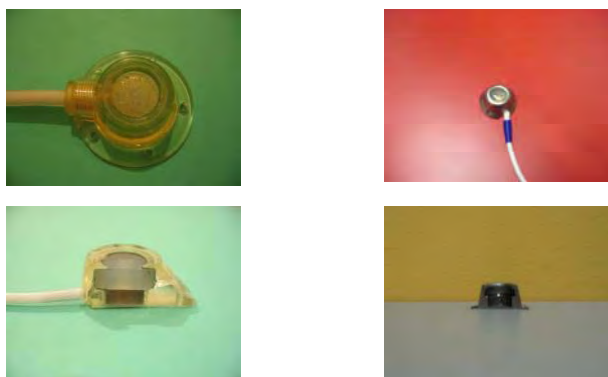
+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Types of Port-a-caths



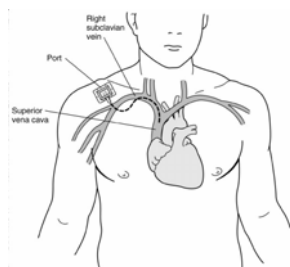
+



=

The Sydney Children's Hospitals Network
(Randwick and Westmead)

Positions



+

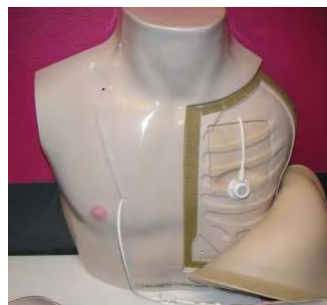


=

The Sydney Children's Hospitals Network
(Randwick and Westmead)

Equipment and Education

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Success



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



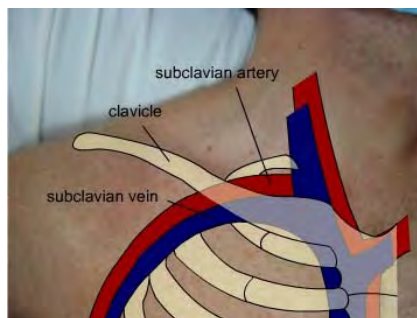
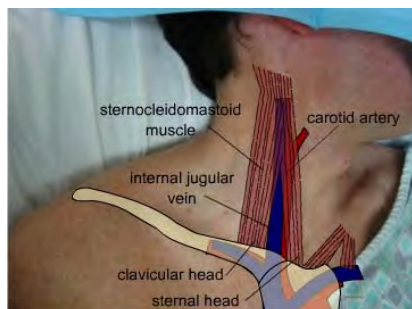
+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

- 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&tbnw=117&start=139&ndsp=13&ved=1t:429,r:4,s:139&tx=77&ty=37>



the
children's
hospital at Westmead

+



SYDNEY
CHILDREN'S
HOSPITAL
RANDWICK

=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Complications

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



the
children's
hospital at Westmead

+



SYDNEY
CHILDREN'S
HOSPITAL
RANDWICK

=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Bleeding

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



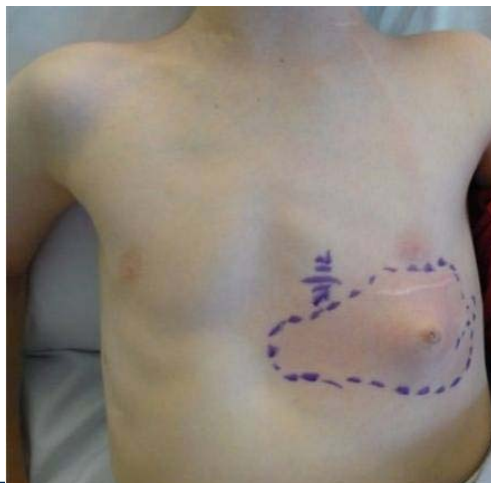
+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Cellulitis



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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**



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Staphylococcus Aureus & Coagulase Negative Staphylococcus



+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

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



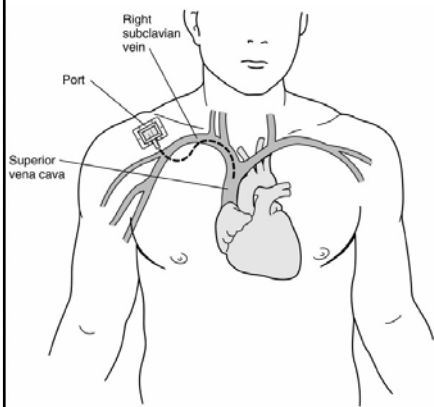
+



=

The Sydney Children's
Hospitals Network
(Randwick and Westmead)

Thrombosis



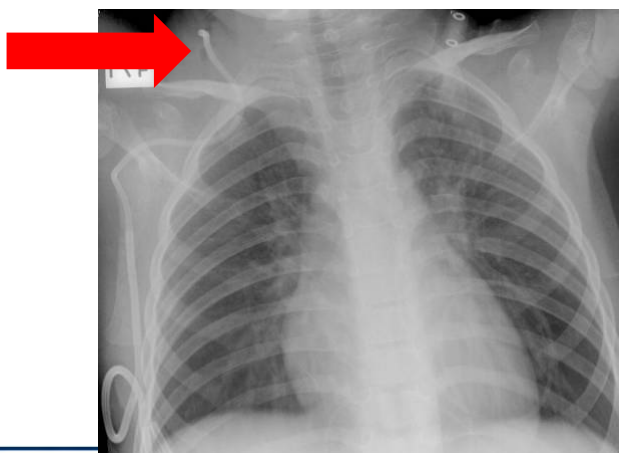
+



=

The Sydney Children's Hospitals Network (Randwick and Westmead)

Mechanical problems



+



=

The Sydney Children's Hospitals Network (Randwick and Westmead)

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

