Infusaport insertion in patients with haemophilia

PURPOSE
This guideline is designed to assist medical and nursing staff in the management of children with haemophilia having an infusaport inserted at the Royal Children's Hospital.

DEFINITIONS
Infusaport or portacath is an implantable Central Venous Access Device.

BACKGROUND
Most children with severe haemophilia (<1% Factor VIII or IX) will require prophylactic intravenous clotting factor administration 2-3 times per week to prevent spontaneous bleeding. Accessing peripheral veins can be difficult and traumatic for children and in particular infants/toddlers where veins are often difficult to identify. A number of boys develop significant behavioural issues around treatment after traumatic experiences in their early years.

Approximately 80% of children with severe haemophilia treated at the Royal Children's Hospital will require an infusaport for venous access. Most families report that insertion of a "port" dramatically improves their quality of life in that venous access is no longer fearful and difficult and parents are able to administer clotting factor to their child at home for both prevention and treatment of bleeds. Ports are removed as soon as parents are able to administer clotting factor peripherally. In general ports are removed prior to commencement of primary school.

PROCEDURE
Once the need for a port has been identified and discussed with the family a referral is made. Mr Joe Crameri performs the majority of infusaport surgery in haemophilia patients at the Royal Children's Hospital.
Many families appreciate the opportunity to see a port (there is one in the haemophilia centre) and to speak with a family whose child is established on home prophylaxis via a port. Successful surgery for infusaport insertion involves co-ordination between the surgical team, pre-op admission area, anaesthetic area, post-op ward, clinical haematology and blood bank.

Consent
Informed consent for the procedure will be obtained by the surgeon performing the procedure.

Clotting factor concentrate
Clotting factor concentrate will be provided for the procedure according to the following protocol:

	Factor VIII	Factor IX
Pre-operative (at induction)	75 units / kg	125 units / kg
Post-operative (immediately post procedure, in theatre)	25 units / kg	50 units / kg
Continuous Infusion for 72 hours (commence post-op in recovery)	3 units/kg/hr	3 units/kg/hr
Day 3 post op (on completion of continuous infusion)	25 units / kg	50 units / kg

Clotting factor dosage may need to be reviewed post-op according to swelling/bleeding at the surgical site. If a clotting factor level is clinically indicated, the blood sample should **not** be taken from the same line as an infusion. Notify the lab to expect the specimen.

The continuous infusion is administered via the peripheral IV, however can be given via the port if the IV tissues. Please instruct theatre to leave the port cannulated after surgery. When clotting factor infusions are run via the port an additional line of normal saline should be run at 10 ml/hr to ensure the port does not block.

When the port needle is removed prior to discharge, flush with 20 ml normal saline (heparin is not required). Please instruct nursing staff to support the port and remove the needle with care. Pulling the port away from the muscle wall during decannulation may cause bleeding around the site.

Pain control
For most children panadol and codeine will provide adequate post-operative analgesia.

Follow – up
The patient should be seen by the haemophilia nurse or in Day Medical Unit for a dose of clotting factor (50 units/kg) via the port ~7 days post insertion. If swelling around the port renders it difficult to palpate the day 7 dose of clotting factor should be administered peripherally. (Additional peripheral doses of clotting factor may be required if swelling is significant.) The patient then typically begins weekly prophylaxis in the Day Medical Unit and parents commence training to access the port and administer clotting factor at home. Training is provided by the haemophilia nurse. Nursing staff in the Day Medical Unit are also able to provide infusaport support and training for families.

ATTACHMENTS

N/A

REFERENCES

HA-P-006 “Coordinating surgical procedures in patients with a bleeding disorder”

“Clotting Factor Concentrates” on the RCH Blood Transfusion website http://www.rch.org.au/bloodtrans/about.cfm?doc_id=10611

Ewenstein BM, Valentino LA, Journeycake JM, Tarantino MD, Shapiro AD, Blanchette VS, et al. Consensus recommendations for use of central venous access devices in haemophilia. Haemophilia 2004;10(5):629-48.