DDAVP Challenge

PURPOSE

This guideline is designed to assist medical and nursing staff in performing a DDAVP challenge for patients with mild von Willebrand's disease and mild haemophilia A at the Royal Childrens Hospital.

BACKGROUND

DDAVP is an analogue of the natural anti-diuretic vasopressin and induces release of FVIII and von Willebrand factor from endothelial cells into the circulation. DDAVP should be considered in the management or prevention of bleeding episodes in patients with mild haemophilia A and mild von Willebrand's disease avoiding the use of recombinant FVIII (for haemophilia A) or plasma derived FVIII (for von Willebrand's disease).

Common side effects of DDAVP include headache, facial flushing, nausea, decrease in blood pressure and fluid retention leading to hyponatraemia. DDAVP is contraindicated in children less than 2 years and in adult patients with known cardiovascular disease.

Approximately 80% of patients with mild haemophilia A and mild von Willebrand's disease will respond with an adequate rise in FVIII and von Willebrand factor respectively following DDAVP infusion. Demonstration of adequate response to DDAVP is recommended in all children diagnosed with mild haemophilia A and mild von Willebrand's disease. Confirmation of effect will lead to prompt and appropriate treatment of bleeding episodes in these patients. This information can also be used to plan for appropriate management prior to surgical procedures.

DEFINITIONS

- Complete response to DDAVP defined as a greater than 2 fold increase in FVIII / von Willebrand Factor AND an increase in FVIII / von Willebrand Factor to greater than 50%.
- Partial response to DDAVP defined as either a greater than 2 fold increase in FVIII / von Willebrand Factor OR an increase in FVIII / von Willebrand factor to greater than 30%

PROCEDURE

- DDAVP is administered in the Day Medical Unit. The procedure should be explained to the family, noting that an IV cannula will need to be inserted into their child and the procedure will take most of the day
- A booking for DDAVP challenge can be arranged by ringing DMU on ext 55950. Patients should be advised to present to Admissions at approximately 9am on the day of the procedure.
- A medication chart providing the dose of DDAVP (0.3 microgram / kg, maximum dose 20 micrograms, diluted in 50 ml N/Saline, given over 30

minute infusion) needs to be completed prior to admission and sent to the Day Medical Unit. (Note that intranasal DDAVP in concentrations for improving haemostasis is not available in Australia.)

- Investigation of DDAVP effect is calculated based on pre-infusion, 60 minute post infusion and 4 hour post infusion levels of FVIII / von Willebrand Factor Ag (vWF Ag). Collagen Binding assay (CBA) and Ristocetin Cofactor (RiCoF) should also be requested for each time point for patients with von Willebrand disease. Separate pathology forms for each of these time points (clearly marked) need to be completed prior to admission and sent to the Day Medical Unit.
- Blood tubes required:

Test	Tube/volume
Factor VIII	1 x 1.4ml citrate
vWD screen (i.e. vWF Ag, CBA and RiCoF)	2 x 1.4ml citrate (or 1 x 3ml citrate) (Performed at The Alfred Hospital)

Fill exactly to line indicated on collection tube and mix gently. Must arrive in lab within 1 hour of collection (best to send immediately). Note that additional investigations may be requested for some patients.

- Vital signs (TPR, BP) should be taken before and after infusion.
- Patients not experiencing any adverse effects during DDAVP infusion may leave DMU after the 60 minute post infusion blood sample and return for collection of the 4/24 post infusion sample.
- Fluid restriction is recommended in the 24 hour period following DDAVP infusion. As a guide, fluid intake should be restricted to approximately 80% of maintenance fluid intake (refer to RCH Clinical Practice Guidelines, Fluid Therapy)
- Follow – up should be arranged in the Haemophilia clinic four weeks after the challenge in order to review results and document recommendations.
- Please wait for the final specimen to be processed by the lab and found to be suitable for testing before removing the IV and discharging the patient. If the final specimen is clotted/under filled, etc the DDAVP challenge will need to be repeated.

ATTACHMENTS

None

REFERENCES

Das P, Carcao M, Hitzler J. DDAVP-induced hyponatremia in young children. Journal of Pediatric Hematology/Oncology. 2005:330-2.