

Heatherwood and Wexham Park Hospitals



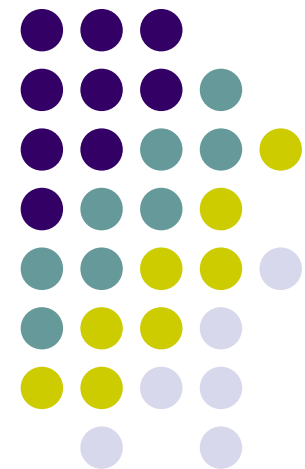
NHS Foundation Trust

Practical Aspects of Delivering Life Long Prophylactic FFP

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History



- 2.12.10: Delivery of first child to parents' who are first cousins: Healthy baby boy
- 12.12.10: Persistent bleeding from circumcision site.
- Results
 - PT: 79.5s (10-14s)
 - APTTs: No clot detected (22-33s)
 - APTT ratio: >8.0 (0.8-1.2)
 - Fib: 3.4g/L (1.5-4)
- Corrected with FFP



Diagnosis

- Factor assay results:
 - Factor II: 56%
 - **Factor V: 4 IU/ml (50-200)**
 - Factor X: 63 IU/ml (50-200)
 - Factor VIII: 109 IU/ml (50-200)
- Consistent with homozygous factor V deficiency
- 1/1 million population



Treatment

- No recombinant factor V available
- Due to spectrum of symptoms no prophylactic treatment was recommended initially
- Feb 2011: Admitted with diagnosis of intracranial bleed
- Patient transferred for craniectomy and surgical evacuation
- Subsequent developmental delay, renal vein thrombosis, renal failure and hypertension and infantile seizures
- Prophylactic treatment with Octaplas started

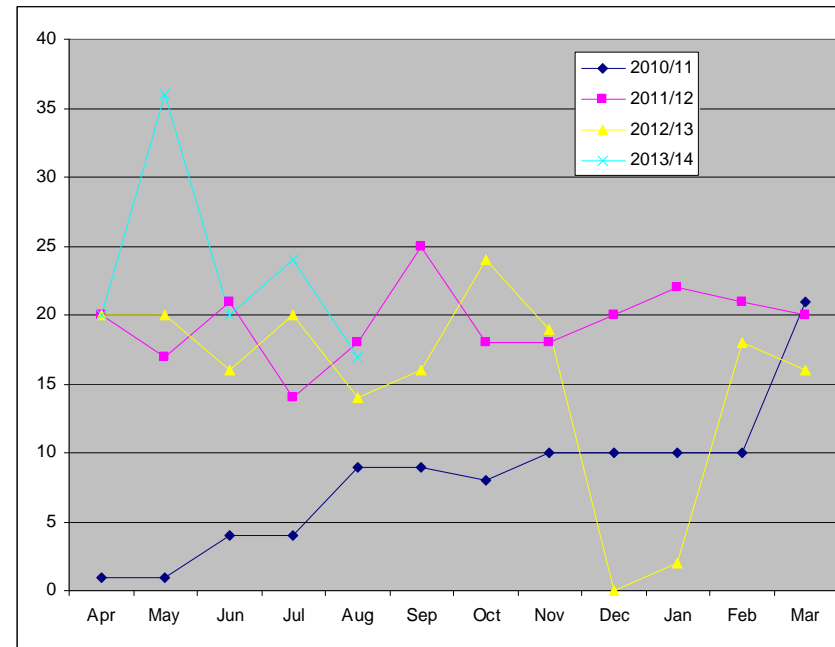
Treatment



- Prophylactic transfusion of plasma commenced every other day (15mL/Kg)
- Patient suffered anaphylactic reaction to Octaplas in Feb 2011
- Moved to Paed MB FFP every M, W, F and Sat
- Carried out in local DGH
- Lab contacted when patient leaves home
- Patient hand held notes

Logistics

- Inadequate storage facilities
- Cost: 250 units per annum at a cost of ~£40,000
- Will increase with age
- Bed space, time spent on ward
- Effectiveness of prophylaxis
- Patient/family quality of life





Continued care

- Home transfusion started in Sept 2012 by Community nursing team
- Policies and procedures written, extensive training undertaken.
- Emergency drugs on site and emergency prescription in place
- Equipment purchased
- Laboratory on board
- Only one transfusion reaction since starting on MB FFP.
- Qualifies for continuing care support at home

What this has meant to the family



- Family took 1 month holiday to Pakistan in Nov 2012 with the help and support of the team
- Home transfusion is arranged around patient/family needs
- Gives valuable time back to the family away from hospital
- Patient can now attend pre-school (with his mum) one day per week soon to be three
- Patient still has developmental delays but seems more alert is making small improvements

With Thanks



None of this would be possible without the dedicated care of:

- The children's community team
- Consultants
- Ward and lab staff
- Multidisciplinary teams both locally and nationally responsible for the care and needs of this child and their family