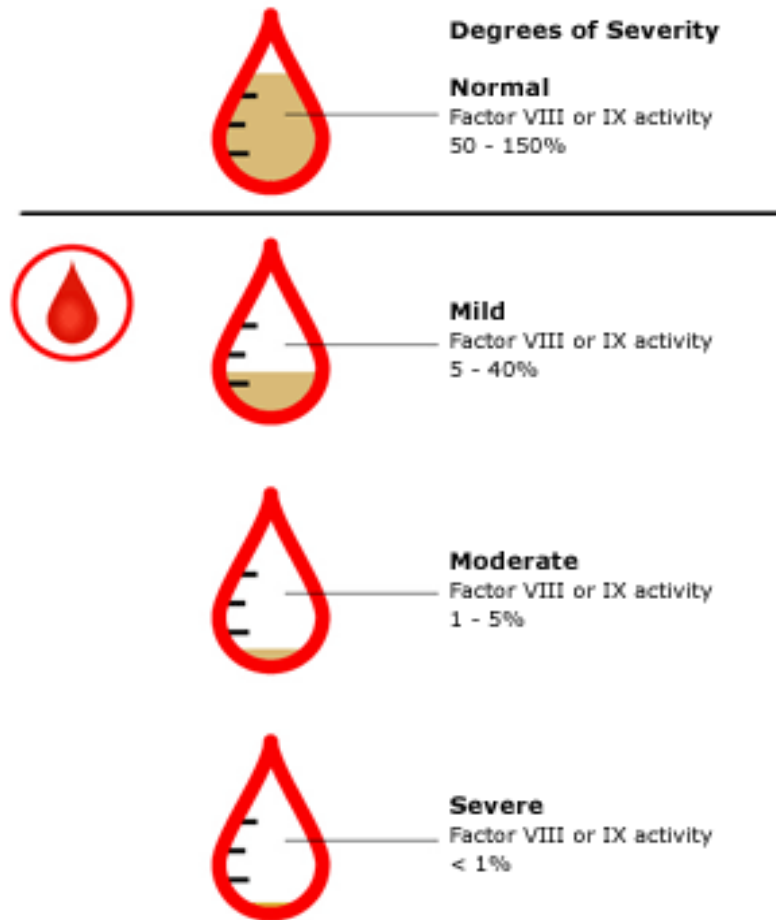


Mild Haemophilia

Penny McCarthy
The Alfred hospital
Melbourne

Haemophilia



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Carriers of Haemophilia may have normal Factor levels. Definitive diagnosis can only be made by DNA gene testing

Brad

Strong family history of mild Haem A

- Mother obligate carrier
- Both brothers tested.
- Brad VIII 5% at 18mths referred to Alfred at request of mother

Age 9 yrs (Sept 1989)

- Presented with knee bleed after falling on concrete AHF 350IU x 2
- Of note both comments requested heat treated to 80deg AHF

Age 15yrs (April 1995)

- Presented with minor ankle bleed climbing a fence
- Mother refusing FVIII
- Sent home with analgesia
- As hep C neg. now offered treatment with recombinant F8. Mother provided written consent

17yrs

- 1997 calf bleed /no memory of trauma
- Kogenate x 3doses (2000IU,2000IU, 1500IU)

23yrs

- bike accident LOC R) shoulder and R) flank pain
- Kogenate 2250IU x2 doses CT NAD discharged

28yrs

- Transferred from private hospital with ? appendicitis. Surgeon wanted to operate with no cover. Mother contacted HTC who negotiated with surgeon for transfer to Alfred.
- USS showed haematoma in r) rectus sheath 22x25 x7 mm
- No evidence of appendicitis or abscess. Discharged no factor given
- FVIII 16%

Total of 7 doses FVIII

(FEB 2011) 31 yrs full time electrician, married 1 daughter

- repair of anal fistula
- FVIII 19%
- BU 0.3
- Struggle maintaining level throughout admission
- CI for 4 days then BD Advate total 10 days discharged with HITH
- Day 10 large PR bleed –Advate FVIII 26% BU 0.2

(MAR 2011) presented with L forearm bleed

- Treated with Advate 42IU /kg at 1230 expected peak 84%
- Pre dose bloods FVIII 1.8% BU 1.6
- 1 hr Peak only 11.4%
- Swapped to 2 hrly Novoseven
- Genetic mutation (pArg 593Cys) (Arg612Cys) CHAMPS data base mild/mod Haem A and known inhibitor risk

| DATES | Hospital days | Rx | Bleed type | | |
|-------------------|---------------|------------|---------------|------------------|-------------------------------------|
| 27/3/11 -30/3/11 | 3 days | 7mg x2/24 | L hand R calf | BU 20 FVIII0.9 | |
| 28/4/11 | | | | | Rituximab 800mg x 4 doses commenced |
| 3/5/11- 9/5/11 | 6 days | 7mg x2 /24 | R Calf bleed | BU 84 FVIII 0.7 | |
| 13/5/11 – 17/5/11 | 5 days | 7mg x2/24 | R calf | BU 80 FVIII0.4 | |
| 26/5/11 – 28/5/11 | 3 days | 7mg x2/24 | R calf L foot | BU 53 FVIII 0.9 | |
| 26/6/11 -12/7/11 | 16 days | 7mg x2/24 | Knee bleed | BU 20 FVIII 0.9 | |
| 24/7/11 – 25/7/11 | 2 days | 7mg x2/24 | Knee bleed | BU 4.3 FVIII 0.9 | |
| 9/8/11 | | | | BU 2. FVIII 2.4 | |
| 23/8/11 | | | | BU 1.4 FVIII5.8 | |
| 2/10/11 – 4/10/11 | 3 days | 7mg x2/24 | R elbow bleed | FVIII 18 | |

October 10th 2011 FVIII finally started to increase and bleeds stopped
 December 20th FVIII 13% BU 0.3

No further challenges or bleeding

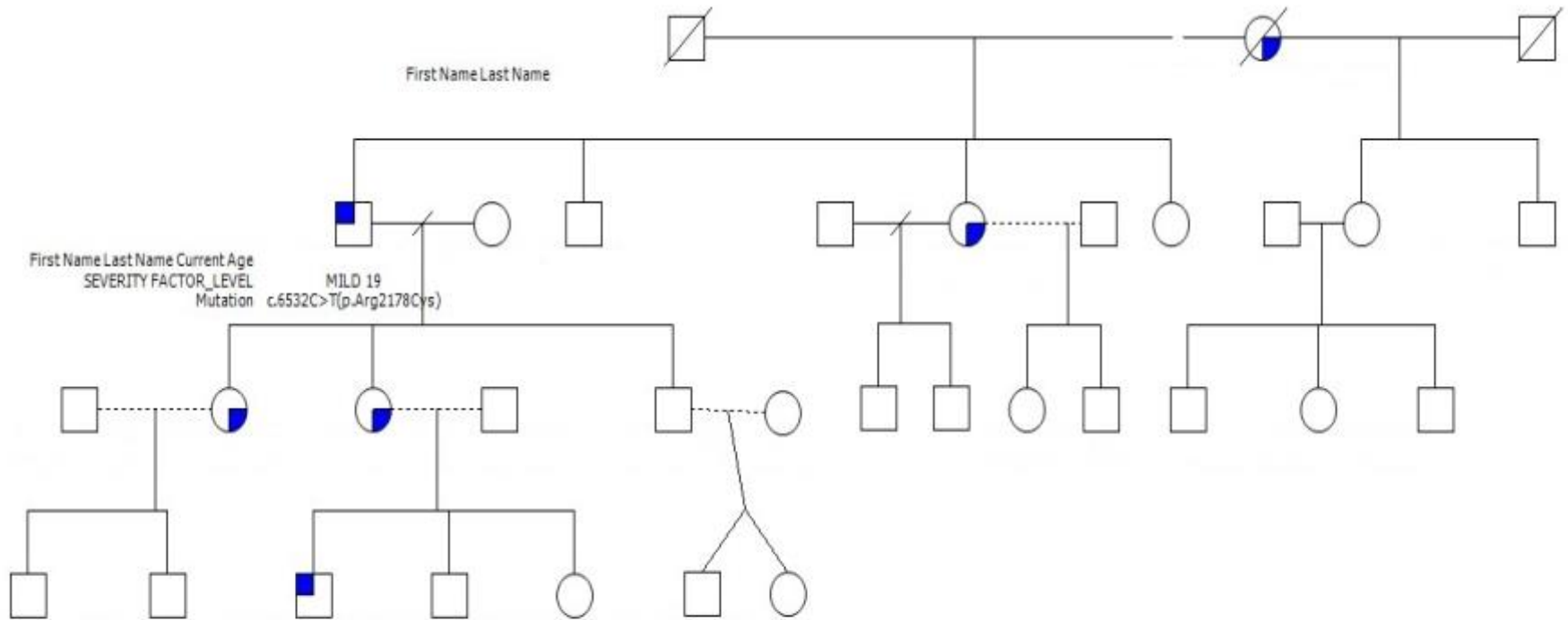
MR L

- April 2014 64yo man referred to HTC from the Royal Eye and Ear hospital for surgery - pterygium repair
- Type 2 diabetes; obesity; Hypertension; Dyslipidemia; Smoker;
diagnosed with haemophilia “B” in Spain as a child following tonsillectomy. No bleeds since
- RV in HTC
- History of bleeds as a child in Chile
- Significant bleed following tonsillectomy aged 30
- Teeth removed managed with TA
- Previous cataract surgery no cover
- FIX 106%
- FVIII 19.4% **Haemophilia A**

1st July 2014 GP faxed a referral asking for ‘opinion and management of `R lower leg bleeding’”

- 2nd July 2014 rang pt
- Hx 2 days lower leg pain
- GP U/S lower leg haematoma
- Pain +++ unable to walk
- Altered sensation and discolouration.
- Advised to call ambulance
- Arrived in ED
- Compartment syndrome
- Advate
- Went directly to theatre for a fasciotomy.

 DIAGNOSIS = HAEMOPHILIA A
  DIAGNOSIS = HAEMOPHILIA A CARRIER



SR (18yo Grandson of Mr L)

- September 2014 presented to MMC with R thigh haematoma from soccer injury
- FVIII tested 18% treated with Biostate referred to HTC
- Difficult time
- New diagnosis played high level soccer going trying for scholarship to the states.
- On crutches off work for weeks
- Unable to play soccer
- Felt life ruined!