**DH Monash SAQ 16 2023.2 (12 marks)**

A 10 year old boy with known haemophilia A presents to the ED with an atraumatic painful ankle. He is systemically well. His X-Ray shows no fracture. His weight is 30kg and his vital signs are normal.

A.What is Haemophilia A? (1 mark)

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| X-linked Congenital Deficiency of Factor 8  (Some responses too wordy) |

B. Complete the table with the concentration of clotting factor for each level of severity (3 marks)

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| --- | --- |
| Severity | Percentage of residual active factor |
| Mild: | 5-40% |
| Moderate: | 1-5% |
| Severe: | <1% |

Worth taking a guess even if don’t know- good idea to do reasonable ranges

C. What is the likely diagnosis? (1 mark)

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| Haemathrosis (most common presentation of Haemophilia A) |

D. List two supportive managements (2 marks)

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| 1. Analgesia: oral paracetamol 450mg, oxycodone 2.5mg,  -avoid NSAIDs,  -note 0.5 mark if give adult doses,  - IV fentanyl not indicated |
| 2. Splinting/Immobilisation/Crutches/Ice/Rest/compression/Elevation- accept 2 from this list |

Not: antiemetics, fluid/blood resus or drainage

E. State the specific management, including dose(s) and target (3 marks)

Recombinant factor 8

1 unit/kg raises activity by 2%

Target 50% activity in haemarthrosis

Thus 25u/kg = 25x30 = 750 units

RCH:

Day 1: 40 units/kg

Day 2: 40 units/kg

Doses should be rounded up to use whole vials sizes 250, 500, 1000, 1500, 2000, 3000 units

National Blood Authority Guidelines - Haemarthrosis desired level 40–60 (units/dl)

If bleeding does not stop, a second infusion may be required. If so, half the initial loading dose should be repeated in 12 hours

-Analgesia and US not specific Mx(US Ix) and aspiration of jt definitely contraindicated

*Note: Non-factor treatment: Emicizumab (Hemlibra) monoclonal antibody directed toward factor IXa and factor X, and mimics function of factor VIII*

*Creates an equivalent factor VIII level of approx 10%*

*Effective in children with haemophilia A*

*Not intended to treat acute bleeding episodes*

*Desmopressin (DDAVP) use in mild haemophilia A - releases stored factor VIII and von Willebrand factor into circulation*

*It is not adequate as a single agent to achieve haemostasis in major bleeding*

*TXA for epistaxis, not haemarthrosis*

*Special considerations for the treatment of children with inhibitors (antibodies) to factor VIII*

*This Mx is complex and the haemophilia treatment centre should be consulted*

*Treatment and prevention of bleeding in children with inhibitors is managed with bypassing agents eg Novoseven®RT, FEIBA®*

F. How would this dose be altered in the setting of an intracranial haemorrhage, and why? (2 marks)

|  |
| --- |
| Target 100% activity in life-threatening bleeds - Thus 50u/kg = 1,500 units  Or 75 units/kg  Desired level 80–100 units/dl |

References: <https://www.rch.org.au/clinicalguide/guideline_index/haemophilia/>

<https://www.rch.org.au/clinicalguide/guideline_index/Haemophilia_treatments/>

<https://www.blood.gov.au/system/files/HaemophiliaGuidelines-interactive-updated-260317v2.pdf>

Comments

- lots of blank answers, take a guess!

- could achieve pass without specific doses of factor 8

**Results**:

Pass mark >=7, 50 candidates:

19 pass, 31 fail, 38% pass rate

Range 2-11