Perioperative management of Haemophilia B: A HaemSTAR UK-wide audit

Poster 3126

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INTRODUCTION AND AIM

Perioperative management of people living with haemophilia is part of comprehensive care. UK adherence to World Federation of Haemophilia (WFH) guidelines is unknown. Data on the perioperative haemostatic management of people living with haemophilia B (PLWHB) is generally scarce [1].

Aim: To compare UK practice with WFH guidelines [2].

METHODS

Inclusion: PLWHB of all ages who had had an invasive procedure between 1 October 2019 and 01 October 2024. Standards (table 2) were based on WFH guidelines. Procedures were classified as minor or major bleeding risk by the data analysis team, with major procedures defined as having a >1.5% major bleed rate or a risk of bleeding that is difficult to diagnose or treat [3]. Approval was obtained from individual centres' audit departments.

CONCLUSIONS

This UK-wide real-world study represents the largest evaluation of perioperative haemostatic management in haemophilia B. Whilst the rates of compliance with WFH guidelines appear low in some areas, this reflects individualised treatment in UK practice and there are excellent rates of haemostatic efficacy. Areas for improvement may include tranexamic acid, inhibitor screening. Management of mild haemophilia B in women needs consideration. A further recommendation may be to review WFH guidelines on minor procedures as 3 days of cover may not be required in the context of EHL products.

REFERENCES

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RESULTS

Haemophilia centre directors were emailed about the study via the UK Haemophilia Centre Doctors' Organisation. Fifteen haemophilia comprehensive care centres (CCC), 7 haemophilia treatment centres and one orthopaedic hospital provided data on 160 patients who underwent 218 procedures (Table 1).

The highest rates of alignment with the guidelines (Table 2) relate to PWHB being managed at or in consultation with a CCC.

There were two episodes of unexpected blood loss during procedures affecting one male having a revision hip replacement, and one female with a baseline factor level of 38 iU/dL who was given tranexamic acid only to cover an egg harvest. This patient required transfusion of packed red cells and had a delayed discharge. Post-operatively, 13 procedures were associated with unexpected blood loss including 3 major and 10 minor bleeding risk procedures. One was a colposcopy with significant postoperative bleeding related to the patient with a baseline level of 38 iU/dL. It was not covered with factor IX or tranexamic acid. All other patients were given factor IX cover with the exception of the patient having an egg harvest (TXA only). Three further patients had significant haemoglobin reductions or further transfusions for post-operative bleeding.

Standard	Major bleeding risk procedure (n=31)	Minor bleeding risk procedure (n=187) †
PWHB treatment duration (sequential days post-surgery) of ≥7 days for major and ≥3 days for minor procedures.	18/31 (58.1%)	22/174 (12.6%)
Pre-operative FIX level measured	22/31 (71.0%)	87/187 (46.5%)
Pre-operative FIX level >60 IU/dL for major, >50% IU/dL for minor procedures.	6/22 (27.2%)	7/87 (8.0%)
Inhibitor screen done post-operatively at 4-12 weeks (major procedures only).	6/31 (19.3%)	N/A
PWHB receive tranexamic acid alongside FIX (with the exception of genitourinary surgery or bleeding (n=3 major, n=12 minor).	24/28 (85.7%)	135/175 (77.1%)
Personalised plan for haemostasis management in consultation with a haematologist.	25/31 (80.6%)	175/187 (93.6%)
Managed at, or in consultation with a haemophilia care centre.		
Surgery took place in a haemophilia CCC	22/31 (71.0%)	139/187 (74.3%)
Surgery took place in consultation with a haemophilia CCC	9/31 (29.0%)	28/187 (15.0%)

	Major bleeding risk procedure (n=28 patients, n=31 procedures)	Minor bleeding risk procedure (n=132 patients, n=187 procedures)
Age (range, median) (years)	15-70, 58.5	0-87, 36
< 18y	26/28 (92.9%)	41/132 (31.1%)
Male sex (n, % of patients)	26/28 (92.9%)	127/132 (96.2%)
Haemophilia severity		
Mild (n, % of patients)	15 (53.6%)	56 (40.1%)
Moderate (n, %)	5 (17.9%)	37 (26.8%)
Severe (n, %)	8 (28.6%)	45 (32.6%)
Factor replacement (% of procedures)		
SHL (n, %)	11/31 (35.4%)	81/187 (43.3%)
EHL (n, %)	17/31 (54.8%)	90/187 (48.1%)
Other (novoseven)	1/31 (3.2%)	1/187 (0.5%)
Factor type unknown (n,%)	1/31 (3.2%)	3/187 (1.6%)
Tranexamic acid only	1/31 (3.2%) §	6/187 (3.2%)
No factor (n,%)	0/31 (0.0%)	6/187 (3.2%)

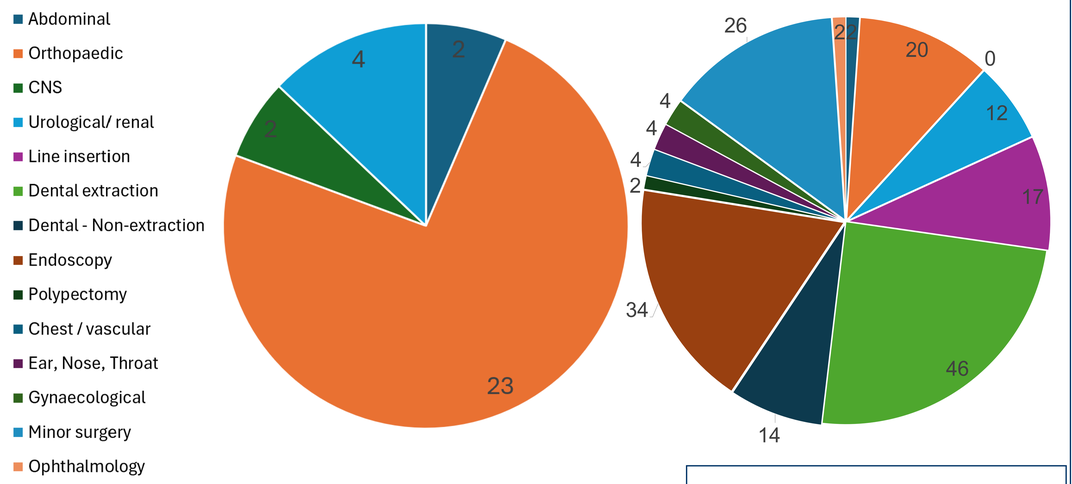

Table 1 (above) . Demographics and factor use according to procedural bleeding risk. SHL: standard half-life factor IX; EHL: extended half-life factor IX. § Ventriculoperitoneal shunt not requiring factor. **Table 2 (right). Compliance with standards.** †Number of procedures covered with factor IX=174. FIX: factor IX. **Graph 1: Procedures in major (left) and (minor) categories**.

DECLARATION OF INTERESTS
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