Audit of a Multidisciplinary Follow-Up Pathway of Acquired Haemophilia A

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INTRODUCTION

- Acquired Haemophilia A (AHA) is a rare bleeding disorder caused by auto-antibody formation against factor VIII.
- Treatment involves managing bleeding and immunosuppression.
- Close follow-up is important to monitor response to immunosuppression and for early relapse detection (Tiede et al., 2020).

AIM

- To audit clinical and laboratory follow-up of patients diagnosed with AHA after starting immunosuppression to inform development of a multi-disciplinary treatment pathway.

METHODS

- Single centre retrospective review of all individuals (n=20) diagnosed with AHA between 2019-2022.
- Audit criteria were based on a local protocol, derived from international guidelines to assess frequency of blood testing (FVIII:C and inhibitor assay) and formal clinical review after starting immunosuppression in patients who achieved remission (n=12).
- Complete Remission: FVIII:C > 50IU/dL. Negative inhibitor titre < 0.6BU. No active bleeding for ≥24 hours post therapy cessation.

CONCLUSIONS

- Improvement in follow-up was seen during the audit. This may relate to the COVID19 pandemic, patient specific factors or publication of international guidance.
- We have developed a multi-disciplinary follow-up pathway to further optimise care for patients with acquired haemophilia A.

FOLLOW-UP PATHWAY AFTER STARTING IMMUNOSUPPRESSION

Pre-remission

Clinical review and testing (FVIII:C and inhibitor titre) **weekly**

1-6 months post-remission

Clinical review and testing **monthly**

6-12 months post-remission

Clinical review & testing every **2-3 months**

RESULTS

DEMOGRAPHICS

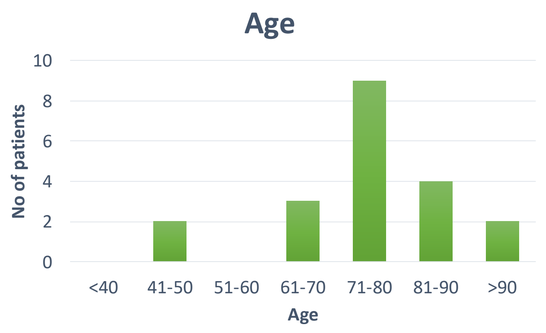

Figure 1: Age at diagnosis (n=20). Mean age at diagnosis: 75 years.

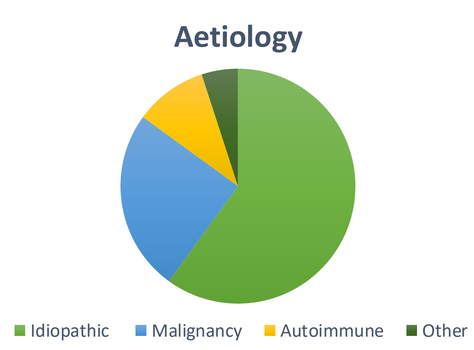

Figure 2: Underlying causes of acquired haemophilia (n=20). 60% cases idiopathic.

IMMUNOSUPPRESION

- Remission in 60% (12/20)
  - Spontaneous remission = 1
- Median time to remission = **121 days**
- Relapse occurred in 3 patients
- 8/20 patients died
  - Infection = 3
  - Malignancy = 3
  - Unknown = 2

Treatment	Mean time to remission (days)
Rituximab (n=3)	191
Rituximab + Cyclophosphamide (n=2)	295
Rituximab + Prednisolone (n=2)	21
Prednisolone (n=2)	77
Prednisolone + Cyclophosphamide (n=2)	40

Table 1: Immunosuppression and time to remission. (n=x): the number of patients on each treatment.

BLOOD TESTING & FORMAL CLINICAL REVIEW

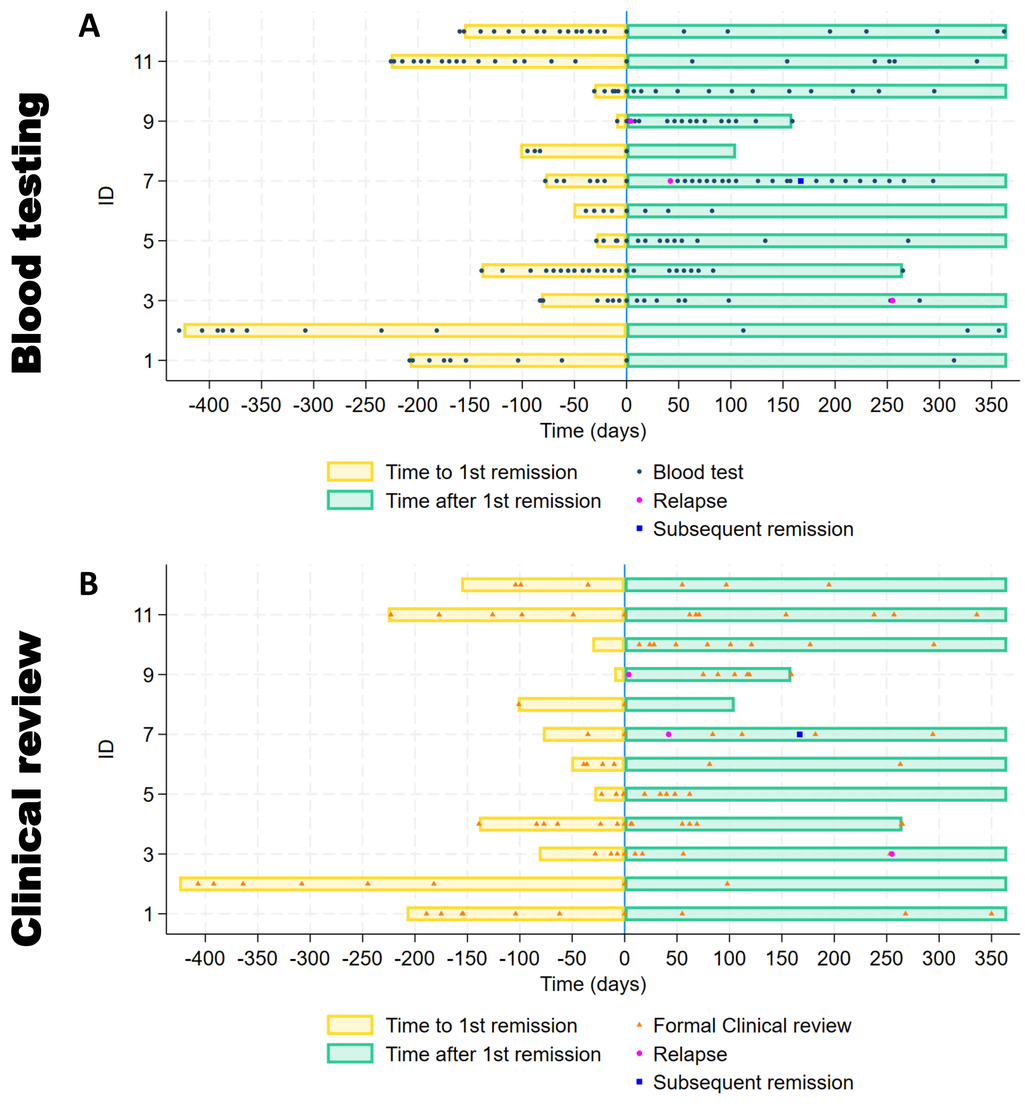

Figure 3: Swimmer plots show the distribution of blood testing (A) and formal clinical reviews (B). Graphs are centred on the date of first remission, with data to the left (yellow bars) showing the timeline from diagnosis to remission and to the right (green bars) the timeline up to one year post first remission.

PRE-REMISSION

Primary outcome

The mean number of days between blood tests or formal clinical reviews on immunosuppression before remission

	Total	G1 2019-2020	G2 2021-2022
Blood test	12	15	8
Clinical review	26	27	25

Table 2: Mean number of days between blood tests & formal clinical review pre-remission. Number of patients in G1 = 7 & G2 = 5.

Days (mean) between review or blood test	Blood test (% pts)	Clinical review (% pts)
< 7	17	0
7 – 14	58	17
> 14	25	83

Table 3: Compliance with audit standards for blood testing pre-remission.

POST-REMISSION

Primary outcome

The mean number of days between blood tests or formal clinical reviews up to 1 year after remission

	Months 1 - 6			Months 6 - 12		
	Total	G1 2019-20	G2 2021-22	Total	G1 2019-20	G2 2021-22
Blood test	71	70	74	95	130	**45.6**
Clinical review	55	60	**48.3**	105	117	**84**

Table 4: Mean number of days between blood testing & formal clinical review in the first-year post-remission. Expected mean for months 1-6 = 30 days & months 6-12 = 60-90 days. Number of patients in G1 = 7 & G2 = 5.

ACKNOWLEDGEMENTS

Acamedics (project sourcing)
Sofia Sardo Infirri (Statistical support)

REFERENCES

Andreas Tiede, Peter Collins, Paul Knoebl, Jerome Teitel, Craig Kessler, Midori Shima, Giovanni Di Minno, Roseline d'Oiron, Peter Salaj, Victor Jiménez-Yuste, Angela Huth-Kühne and Paul Giangrande (2020) "International recommendations on the diagnosis and treatment of acquired hemophilia A", *Haematologica*. Pavia, Italy, 105(7), pp. 1791-1801. doi: 10.3324/haematol.2019.230771.

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