



Gynaecological outcomes in women with inherited platelet function disorders: preliminary data from the ISTH REDCap Registry

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INTRODUCTION

The presentation and management of women with inherited platelet function disorders (IPFDs) in the obstetrics and gynaecology settings can be variable due to the rarity and heterogeneity of these disorders with variable bleeding phenotypes. Clinical experience in this area is thus limited. In the literature, most information is scattered in case reports and small case series.

An ISTH RedCap registry was set up to capture international multi-centre data on management of obstetrics and gynaecological complaints in these women. The aim is to build knowledge and experience of the different presentations and management approaches in women with inherited platelet function disorders.

We analyse the preliminary data collected to date pertaining to gynaecology.

METHOD

An ISTH RedCap registry was set up to capture data worldwide. This was publicised through ISTH oral presentation, newsletters and social media outlets. It was also part of a formal publication article in RPTH journal.

The registry can be seen here: <https://redcap.isth.org/surveys/?s=HTHE4TAMTM>

Data from nine centres worldwide were captured through the ISTH Registry completed by healthcare professionals using their patients’ clinical records. Information entered on the registry was anonymised. Preliminary data collected is analysed and statistics are observational.

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RESULTS

- Data from 38 women was analyzed.
- Various aspects of the results have different denominator numbers as not all questions were answered for all the women.

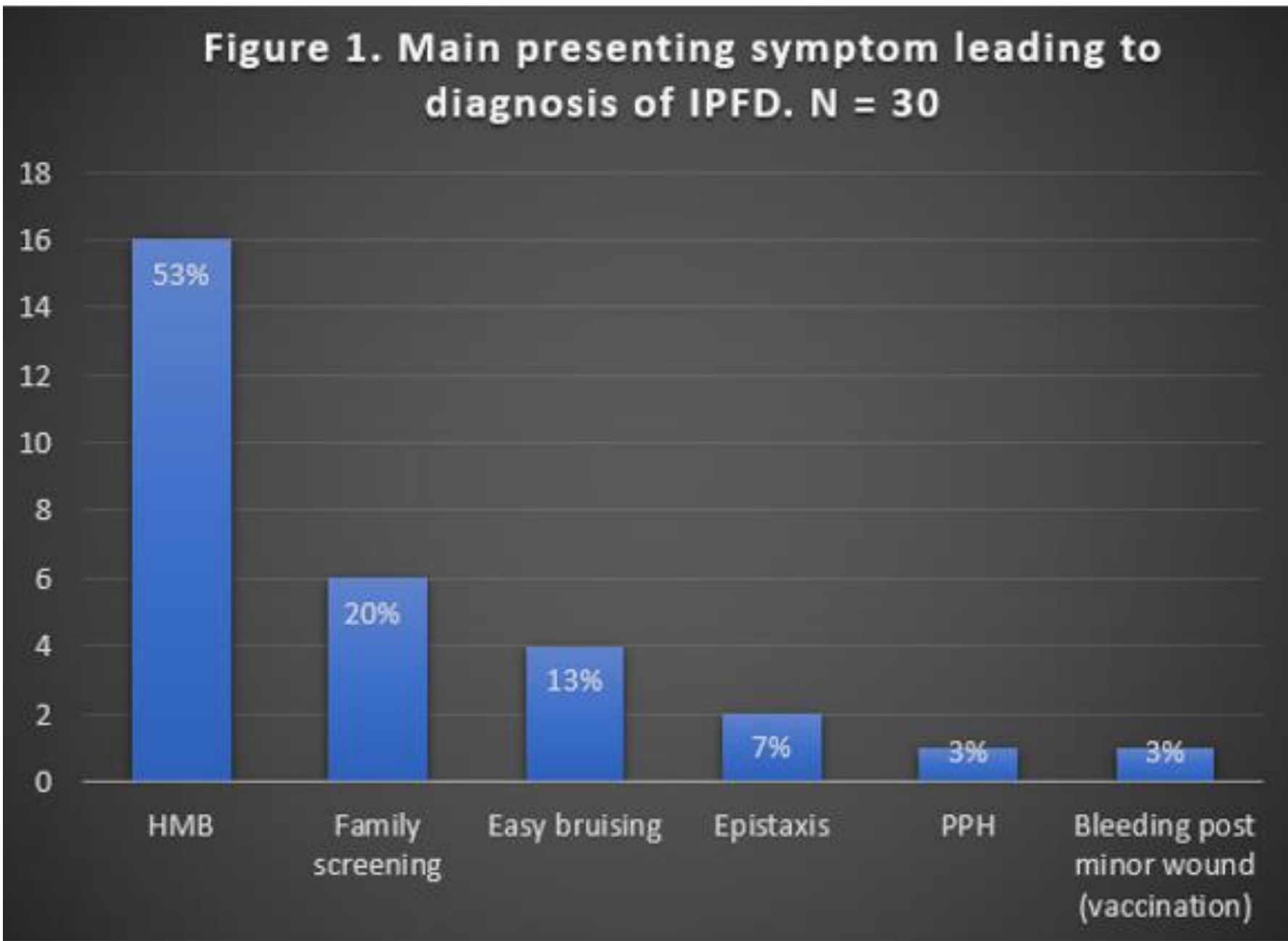


Table 1. Variable characteristics / parameters measured in women with inherited platelet function disorder (IPFD)

Characteristic, measurement unit	Value
Age of woman at data collection point, mean (std) in years	26 (9.4)
Age at diagnosis of IPFD, mean (std) in years	16 (11.1)
ISTH bleeding score, mean (std)	6.3 (4.1)
Pictorial Blood loss Assessment Chart (PBAC) score, mean (std)	459.7 (349.6)
Age at menarche, mean (std) in years	13 (1.5)
HMB started from menarche, proportion (%)	23/36 (64)
Attendance to the emergency department for acute episodes of HMB, proportion (%)	10/36 (28)
Of these women, proportion (%) needing: - packed red cells - Platelet transfusion - Novo Seven (rFVIIa)	7/10 (70%) 2/10 (20%) 2/10 (20%)

DISCUSSION / CONCLUSION

- Data from this international registry show that women with IPFD usually present with HMB as a predominant symptom. This often leads to the initial diagnosis of IPFD.
- An important characteristic of HMB in women with IPFD is:
 - It commonly starts from menarche. As such, a greater proportion of women are diagnosed within a few years after menarche.
 - Over a quarter of women with IPFD presented to emergency department with HMB needing blood and haemostatic products (90%) in addition to routine hormonal and antifibrinolytic therapy.
 - PBAC scores are high, with a mean score of 460 in this study. Most studies use a PBAC score of >150 points to define HMB as it correlates with >80 ml of blood loss.
- Medical management of HMB included tranexamic acid, oral combined pill, progestogens, levonorgestrel-releasing intrauterine system and/or desmopressin (DDAVP, second/third-line only).
- First-line management for HMB used a single medical agent for treatment in 66% and a combination of two medical agents in 34%. This resulted in treatment efficacy of only 17%.
- The proportion of women receiving two or more combination medical therapy increased progressively at second line, then third line of treatment.
- At third-line therapy, endometrial ablation was also used in women who had completed their family.
- Rate of effective treatment increased with line of therapy, reaching 97% in the third-line.
- **CONCLUSION:** Medical management of HMB in women with IPFD can be challenging necessitating a combination of antifibrinolytic agents, hormonal and haemostatic therapy. In retractable cases, endometrial ablation is an option for those who have completed their family. As more data is collected, there may be strength to compare the efficacy of the different combination treatments.

Table 2. Type of treatment for heavy menstrual bleeding (HMB) and their effectiveness in women with inherited platelet function disorders.

Treatment for HMB in women with platelet function disorders	First-line treatment (n=35)	Second-line treatment (n=29)	Third-line treatment (n=14)
Single agent treatment: TXA, COCP, cyclical progestogens or hormonal intrauterine system (e.g. Mirena IUS)	23	2 (all Mirena IUS)	0
Combination of two agent treatment: TXA and COCP, TXA and progesterone, TXA and hormonal intrauterine system, COCP and progesterone, TXA and endometrial ablation [third-line only]	12	18	6
Combination of three agent treatment: TXA and COCP and desmopressin, TXA and COCP and progesterone, TXA and COCP and intrauterine system, COCP and endometrial ablation and intrauterine system [third-line only]	0	9	8
Efficacy of treatment, n (%) Effective Not effective	6 (17) 29 (83)	15 (52) 14 (48)	13 (93) 1 (7)
Cumulative treatment effectiveness	5 (14)	21 (60)	34(97)

COCP, combined oral contraceptive pill; TXA, tranexamic acid.

CONTACT INFORMATION

We encourage more haemophilia centres looking after women with IPFD who have obstetrics and gynaecology complaints to contribute their data to this anonymised registry.

The registry is available at : <https://redcap.isth.org/surveys/?s=HTHE4TAMTM>

Please contact Dr Deborah Obeng-Tuudah on **email: d.obeng-tuudah@nhs.net** for further information.

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