

SPECTRUM OF RARE BLEEDING DISORDERS IN PAKISTAN: CLINICAL PRESENTATION AND PRIMARY CARE

Sehar Khaliq, Lubna Zafar, Muhammad Iqbal, Tahira Zafar

Blood Diseases and Hemophilia Treatment Centre, Rawalpindi

Introduction

- Rare inherited bleeding disorders are autosomal recessive disorder that includes deficiencies of coagulation factors; fibrinogen, factor (F)II, FV, combined FV and FVIII, FVII, FX, FXI and FXIII. ¹
- Despite the worldwide prevalence of rare bleeding disorders knowledge their management is suboptimal. ²
- Patients affected with rare bleeding disorder present with wide spectrum of clinical manifestation ranging from mild-to-life threatening bleeding diathesis. ³
- The identification, diagnosis and management of these disorders is a big challenge in developing countries like Pakistan where there is lack of proper diagnostic facilities and referral centres.

Objectives

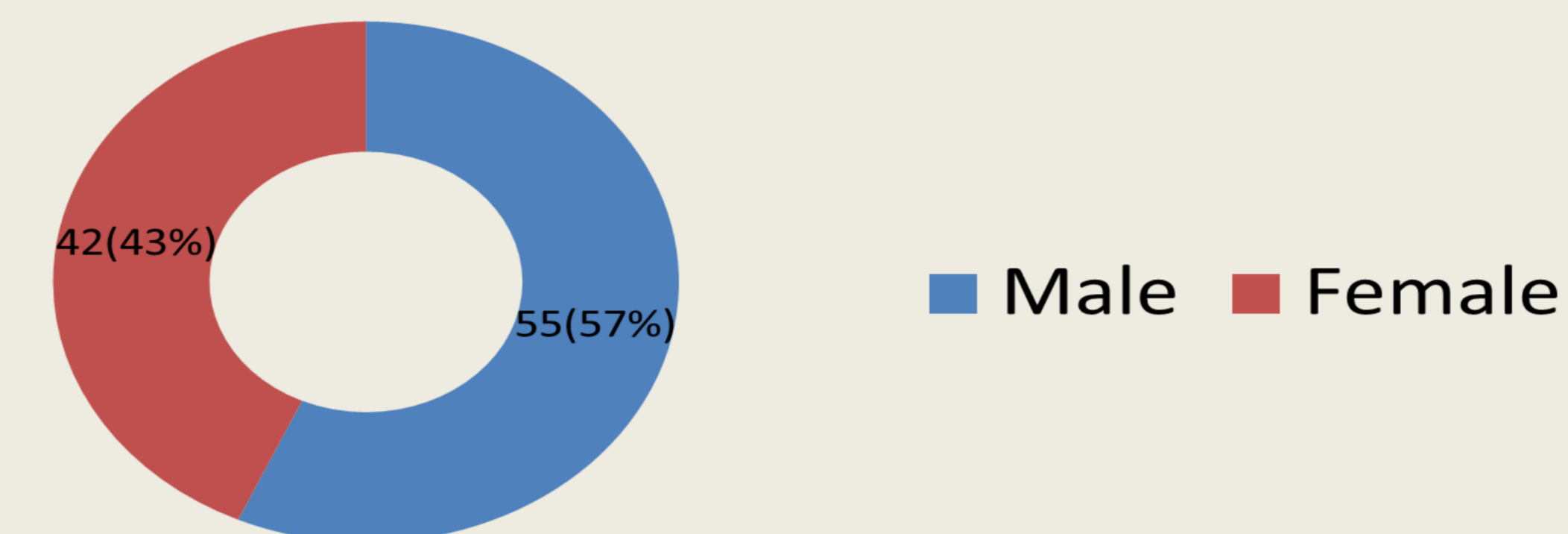
This study was conducted to find out the prevalence, clinical presentations and primary care of rare bleeding disorders.

Methodology

A cross-sectional study was conducted at Blood Diseases and Hemophilia Treatment Center, Rawalpindi from January 2008-January 2016. Detailed history was taken and physical examination was done. CBC, bleeding time, PT, APTT, coagulation factor assays was done. FXIII screening was done by urea clot solubility test. Platelet function analysis was done on platelet aggregometer.

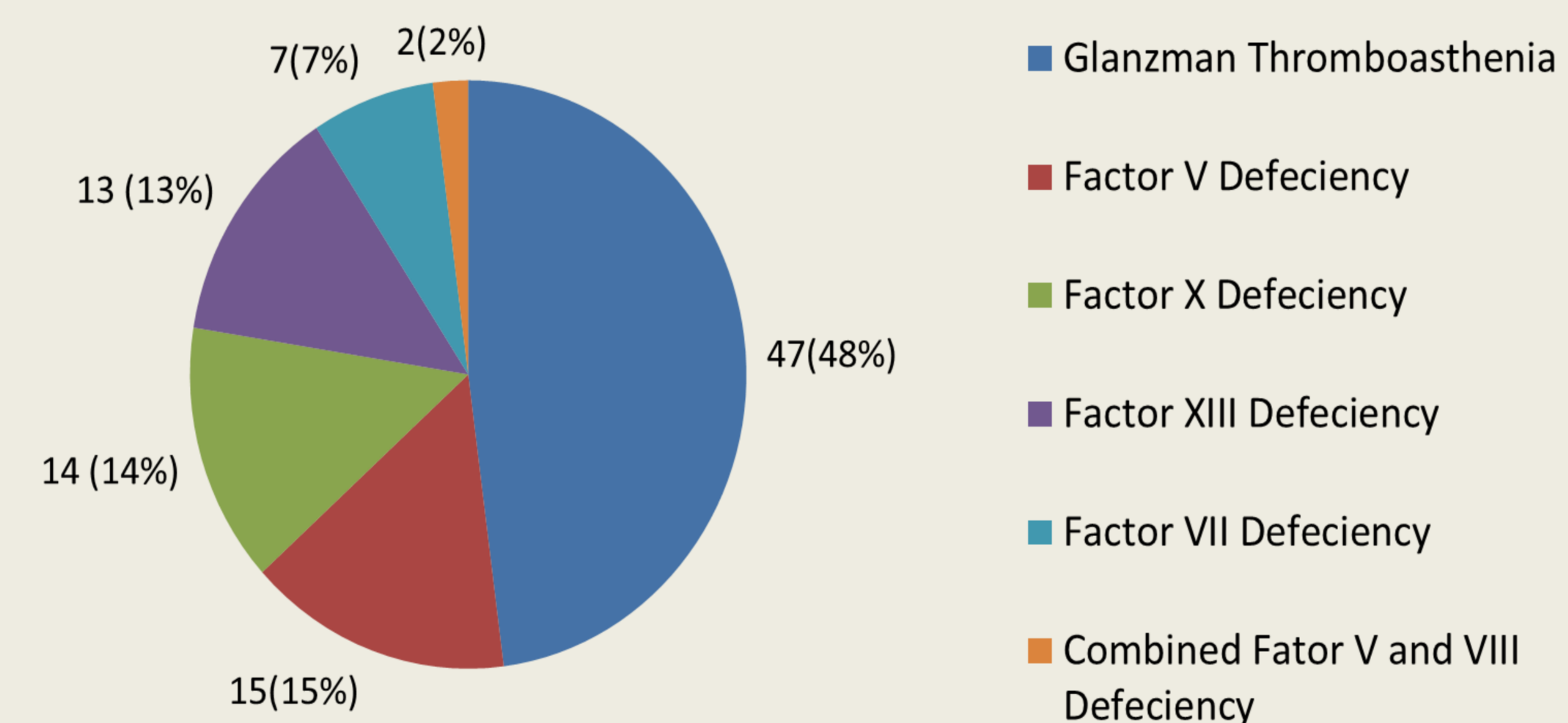
- A total of 97 patients were diagnosed to have rare bleeding disorder.

GENDER DISTRIBUTION OF PATIENTS WITH RARE BLEEDING DISORDERS



The mean age was 15 years.

BREAKDOWN OF PATIENTS WITH RARE BLEEDING DISORDERS



PRESENTING SYMPTOMS	PERCENTAGE OF PATIENTS
Petechia/Bruises	38%
Epistaxis	18%
Menorrhagia	20%
Post-Traumatic Bleed	08%
Post-Circumcision Bleed	09%
Umbilical Cord Bleed	06%
Intracranial Bleed	01%

- The mainstay of management was fresh frozen plasma and cryoprecipitate.
- Factor XIII concentrates were also given when available.

Conclusion

- There is a lack of information on the prevalence of rare bleeding disorder. In spite of their apparent rarity, Pakistan has substantial number of patients with rare bleeding disorders but lack of proper diagnostic facilities and management leads to inadequate treatment and life-threatening complications.

Bibliography

- Palla R, Peyvandi F, Shapiro AD. Review Series: Inherited Bleeding Disorders
Rare bleeding disorders: diagnosis and treatment . Blood 2015;125(13)
- Peyvandi F, Palla R, Menegatti M, Mannucci PM. Introduction. Rare bleeding disorders: general aspects of clinical features, diagnosis, and management. Semin Thromb Hemost 2009;35(4):349-355
- Schroeder V, Kohler HP. Factor XIII deficiency: an update. Semin Thromb Hemost 2013;39(6):632-641.



Poster Presented at:

DOI: 10.3252/jso.ed.V17F01.2016

Rare bleeding disorders
Tahira Zafar

207--PP-M

9T0ZHM