

Congenital factor V deficiency, comparison the severity of clinical presentations among patients with rare bleeding disorders

Majid Naderi¹, Shadi Tabibian³, Shaban Alizadeh³, Akbar Dorgalaleh³, Peyman Eshghi⁴, Taregh Bamedi¹, Saeed Dorgaleh⁵

Pediatric department, Alie-bnAbitaleb hospital, Zahedan University of Medical Sciences, Zahedan, Iran
 Department of Hematology, Allied Medical School, Tehran University of Medical Sciences, Tehran, Iran
 Pediatric Congenital Hematologic Disorders ResearchCenter, ShahidBeheshti University of Medical Sciences, Tehran, Iran
 Clinical Laboratory Medical Sciences Department, Zabol University of Medical Sciences, Zabol, Iran

Background

Factor V deficiency (FVD) is a rare bleeding disorder (RBD) mostly present in regions with high rate of consanguinity. After FXIII deficiency is the next more prevalent RBD in Sistan and Baluchistan in south east of Iran. The aim of this study is to evaluate clinical manifestations and severity of bleeding diathesis in patients with FVD. We also compared the severity of clinical manifestation in FVD with other types of RBD.

Methods

This descriptive study was conducted on 23 patients with FVD in this province. FVD was diagnosed by clinical findings and routine laboratory tests. Bleeding diatheses were classified in three grades of I, II and III including (i) asymptomatic (ii) Grade I: bleeding occur due to trauma or drug ingestion, (iii) Grade II: spontaneous minor bleeding (iii) grade III: spontaneous major bleeding.

Results

Based on residual plasma FV activity 6 (26%), 16 (69.5%) and 1(4.5%) patients had mild, moderate and severe deficiency, respectively. Twenty five percent of patients had grade III life threatening bleeding episodes which in comparison with FVII deficiency (17.4%) and FI deficiency (21%) had a higher incidence and in comparison with FX-deficiency (41.7%) and FXIII-deficiency (63.1) a lower incidence. About 72% of bleeding diathesis of Grade III was observed in patients with moderate FVD while these bleeding features was not observed in patient with severe FVD. In FXIII and FX deficiency 100 % of bleeding features of grade III was observed in severe deficiency. In FVII-D 34 % of bleeding diathesis of grade III was observed in patients with severe deficiency and 66 % of them in patients with mild deficiency. Bleeding diathesis in grade II and grade I were observed in 56.2% and 16.7% of patients, respectively. Almost 21.8% of patients with FX deficiency followed by FVII deficiency (16.6%) were asymptomatic while in FVD 2.1% of patients were asymptomatic.

Conclusion

FVD is the second common type of RBDs in this province and grade II bleeding episodes were observed in more than half of the patients. By comparing bleeding episode of Grade III in FV deficiency with other RBDs, FXIII and FX deficiency has the strongest association between bleeding severity and coagulation factor activity level.

References

- Naderi M, Eshghi P, Dorgalaleh A, Tabibian S, editors. Clinical manifestations of rare bleeding disorders in South East of Iran. HAEMOPHILIA; 2013: WILEY-BLACKWELL 111 RIVER ST, HOBOKEN 07030-5774, NJ USA
- Huang JN, Koerper MA. Factor V deficiency: a concise review. Haemophilia. 2008;14(6):1164-9. Epub 2009/01/15.
- Peyvandi F, Palla R, Menegatti M, Siboni S, Halimeh S, Faeser B, et al. Coagulation factor activity and clinical bleeding severity in rare bleeding disorders: results from the European Network of Rare Bleeding Disorders. Journal of Thrombosis and Haemostasis. 2012;10(4):615-21.

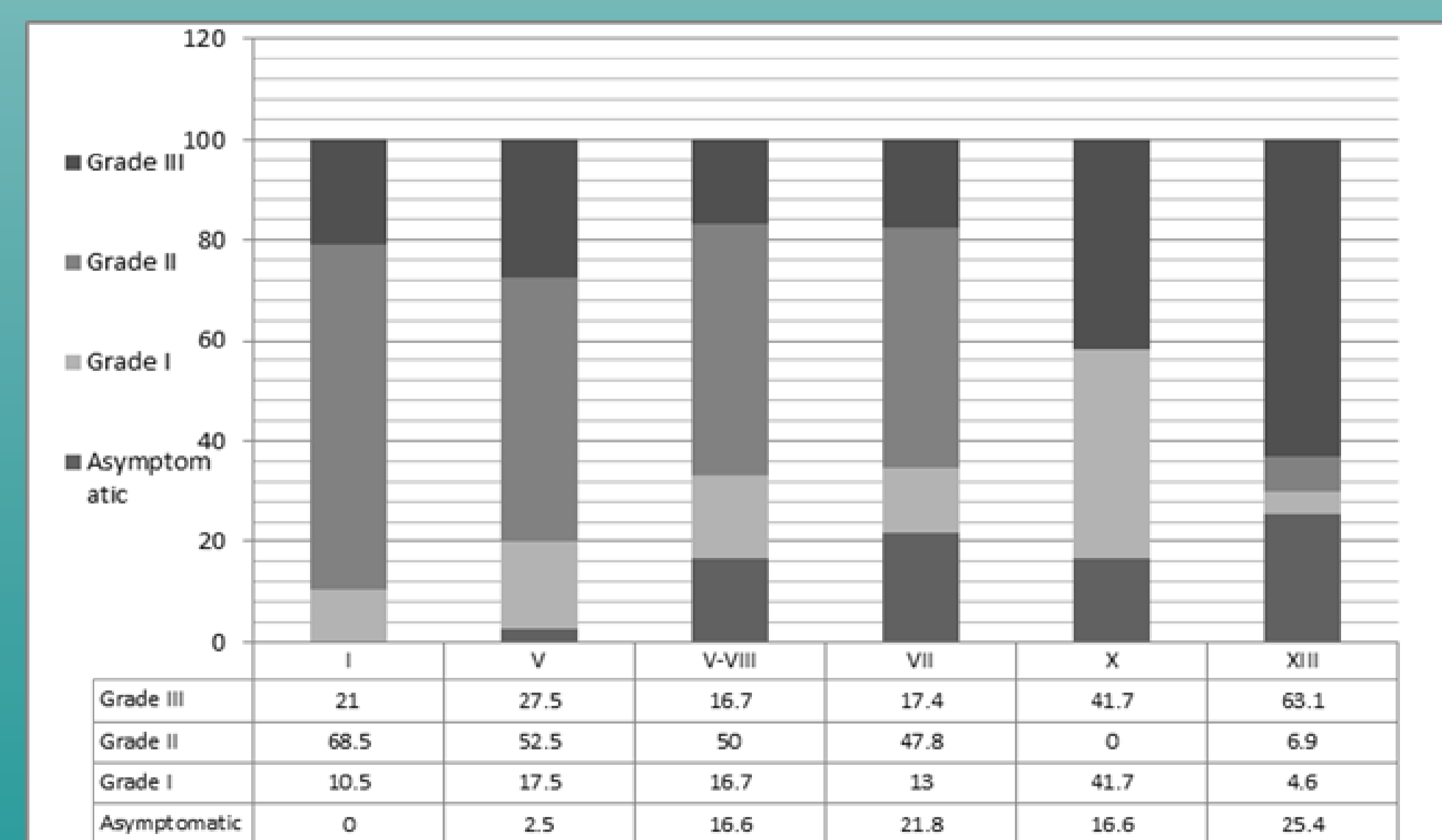


Table 1. Severity of clinical manifestations among different RBDs

