Factor V is a 330 kDa glycoprotein with several domains that resembles FVIII functionally. It is synthesized in the liver and then released into the blood circulation. Eighty percent of the factor is found in the plasma and the remainder in the platelets. When activated, FV plays a role as the cofactor of activated factor X in prothrombin activation and improves the formation of thrombin (1). Factor V deficiency may lead to mild or severe bleeding and presents with bruising, epistaxis, etc. as well as hemorrhrosis and intracranial hemorrhage (ICH) in severe forms. Due to hemorrhrosis and the prolonged time of bleeding, the disorder is called parahemophilia, too (2, 3). In this study we aimed to evaluate phenotype findings in patients with congenital factor V deficiency and any probable relationship between the factor level in plasma and the severity of symptoms of the patients in order to have earlier and better diagnostic method and prevent the severe complications in these patients.

**Introduction**

Among our patients, the distribution of the clinical presentations were as follows: 30.76% presented with post-traumatic, post-surgery, post-partum and post-circumcision bleeding while 23.10% showed easy bruising. 15.38% of patients manifested post-dental extraction and gingival bleeding and also another 15.38% of them presented with intracranial hemorrhage (ICH). 7.69% of them showed epistaxis and 7.69% were identified post-laboratory evaluation incidentally. The mean age of the onset of clinical signs was 21.52 (Median=18). Also, the mean factor activity level was 3.9 (Median=4). Patients were categorized into two different groups as major and minor bleeding based on their first clinical bleeding symptoms. There was not statistically significant difference between male and females with regards to age of diagnosis or factor activity level. Thus, early detection of the patients and using the prophylaxy in severe cases and therapeutic approaches to prevent the complications is needed.

**Materials and methods**

Phenotypes of the 13 patients who suffered from congenital factor V deficiency were investigated. 7 patients were the offspring of consanguineous marriages. The disorder was diagnosed for them in Shiraz Hematologic Research Center. Also, factor activity level and clinical presentations at the onset of the diagnosis were studied for each patient.

**Results**

Our study identified that there is a discrepancy between plasma FV activity level and severity of clinical presentations and also there is no difference between male and female regard to the age of onset or factor activity level. Thus, early detection of the patients and using the prophylaxy in severe cases and therapeutic approaches to prevent the complications is needed.

**Discussion**

Among our patients, the distribution of the clinical presentations were as follows: 30.76% presented with post-traumatic, post-surgery, post-partum and post-circumcision bleeding while 23.10% showed easy bruising. 15.38% of patients manifested post-dental extraction and gingival bleeding and also another 15.38% of them presented with intracranial hemorrhage (ICH). 7.69% of them showed epistaxis and 7.69% were identified post-laboratory evaluation incidentally. The mean age of the onset of clinical signs was 21.52 (Median=18). Also, the mean factor activity level was 3.9 (Median=4). Patients were categorized into two different groups as major and minor bleeding based on their first clinical bleeding symptoms. There was not statistically significant difference between male and females with regards to age of diagnosis or factor activity level. Thus, early detection of the patients and using the prophylaxy in severe cases and therapeutic approaches to prevent the complications is needed.

**References**