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322.DISORDERS OF COAGULATION OR FIBRINOLYSIS: CLINICAL AND EPIDEMIOLOGICAL

Congenital Factor VII Deficiency : A Retrospective Analysis of 14 Cases from One Hospital

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Objective To explore the clinical characteristics, laboratory findings,diagnosis, treatment, and prognosis of congenital factor VII (FVII) deficiency. **Methods** Clinical data of 14 patients of congenial FVII deficiency diagnosed from Jan 2019 to Jun 2022 were retrospectively analyzed. **Results** Of the 14 patients,there were 8 males and 6 females with median age 31.5 (1.3~70) years. Family history was found in 2 cases. There were 7 cases with bleeding symptoms, of which were mucocutaneous bleeding (1 case), gingival bleeding (2 cases), menorrhagia (3 cases), hemoptysis (1 case), post-surgery hemorrhage (1 case), and 1 case with cerebral infarction. Menorrhagia occurred in 3 cases (75% of female patients who were in fertile age). Laboratory findings were characterized by significantly prolonged prothrombin time (PT) (19.2~58.9s) , of which could be corrected by mixed normal plasma, normal partial thromboplastin time (APTT), and decreased FVII activity (FVII:C) (2.4~24.7%). 1 case and his family members received gene mutation analysis and compound Heterozygous mutation (heterozygous deletion mutation and heterozygous missense mutation) was found in the proband. 3 cases were treated with prothrombin complex concentrates (PCC), 1 case with PCC and human recombinant activated FVII (rhFVIIa). 10 patients with no or mild bleeding symptoms did not receive any replacement therapy. **Conclusion** Most patients with congenital FVII deficiency have mild or no bleeding symptom, but have a tendency to excessive bleeding after surgery or trauma. There is no significant correlation between FVII:C and the severity of bleeding symptoms. Prophylaxis could be applied in patients with severe bleeding symptom. Gene mutation test is significant for diagnosis, prognosis prediction of the disease.

Disclosures No relevant conflicts of interest to declare.

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