patient group with arterial thrombosis was compared with the control group, no difference was found in the risk of thrombosis in terms of factor V Leiden, prothrombin, Factor XIII, MTHFR 677, MTHFR 1298, PAI-1 gene mutation (p=0.084, p=0.82, p=1, p=0.65, p=0.064, p=1, respectively). In our study, no significant difference was found in the increased risk of thrombosis in the detection of thrombophilic gene tests in arterial thrombosis compared with the control group. **Conclusion:** In our study, thrombophilia gene panel screening was not considered necessary in patients with arterial thrombosis, and it was observed that factor V Leiden, prothrombin, Factor XIII, MTHFR 677, MTHFR 1298, PAI-1 gene mutations in the hereditary thrombophilia panel did not lead to an increased risk of arterial thrombosis. Hereditary thrombophilia testing is not recommended in patients with arterial thrombosis according to current guidelines.

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OP 05

SURGICAL INTERVENTIONS IN FACTOR VII DEFICIENCY: A SINGLE CENTER EXPERIENCE

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Objective: FVII deficiency is the most common of the rare congenital bleeding disorders with a prevalence of about 1:500,000. Bleeding symptoms are considerably variable in terms of both location and severity, and may have a heterogenous spectrum ranging from asymptomatic conditions to serious/life-threatening bleeds In surgical interventions, the duration of treatment and factor dose should be determined by considering the patient's previous and current bleeding clinic, factor level and comorbidities. **Methodology:** We aimed to share our experience of surgical interventions and bleeding management in individuals with factor VII deficiency between January 2023 and January 2024 who followed up in our outpatient clinic. **Results:** A total of 14 surgical interventions were performed in 12 patients with factor VII deficiency between January 2023 and January 2024 at Ege University Hemophilia Outpatient Clinic. 4 tooth extractions, 2 septorhinoplasties, 1 tympanoplasty, 1 tympanomastoidectomy, 1 lung wedge resection, 1 cataract and 4 orthopedic procedures (arthrodesis, radius fracture repair, total hip replacement and arthroscopy) were performed. The median age was 43 years (20-78 years), 7 of patients were female and 5 were male. 7 patients had ISTH bleeding score below 5 and 4 patients had no bleeding diathesis. Preoperative factor VII levels of the patients varied between 5-36%. Recombinant factor VIIa (rfVIIa) was used in 85% (n=12) and FFP in 15% (n=2) of the procedures. Median duration of treatment was 2.5 days (1-8 days). The median preoperative rfVIIa dose was 15 mcg/kg (10-30 mcg/kg), while the median single dose given in the postoperative period was 16.7 mcg/kg. While a single dose was administered in minor interventions such as tooth extraction, the mean number of total doses administered during treatment in other interventions was 11. In one patient, the procedure was performed with TDP due to the presence of both factor VII deficiency (FVII:36) and hypofibrinogenemia, low bleeding score and no previous history of postoperative bleeding. In another patient who underwent tooth extraction, the procedure was performed with FFP because the factor level was >30% and there was no previous bleeding history. The preoperative FFP dose was 15-20 ml/kg in patients that receiving FFP. Effective bleeding control was achieved and no thrombosis was observed in patients receiving both FFP and rFVIIa. **Conclusion:** The correlation between FVII activity and bleeding tendency is poor, although severe bleeding is most commonly associated between low FVII activity levels and the surgical risk of bleeding.Plasma-derived and recombinant FVII concentrates are currently used for treatment. In countries where access to these products is lacking, fresh frozen plasma and prothrombin complex concentrates are also used, though they contain low amounts of factor FVII. In patients included in the recording system established for patients with FVII deficiency (STER) and who underwent surgical procedures, use of rFVIIa was evaluated in 110 elective surgical procedures performed on 95 patients were examined, and it was shown that neither FVII level nor surgical procedure influenced rFVIIa replacement treatment, and only the patient's phenotype of bleeding was effective in replacement treatment. it was shown that the lowest effective dose of rFVIIafor hemostasis was 13 μg/kg on the day of surgery, and at least three doses were needed.In same study, it was recommended to give a mean total dose of 20 micrograms/kg rFVIIa in invasive interventions and minor surgeries. Furthermore in majör surgeries it is recommended to give rFVIIa at a single dose of 13 mcg/kg in the first 24 hours after operation and at least three administrations needed. Similarly, in our clinic, a median dose of 15 mcg/kg was administered before surgical interventions. Before invasive procedures and minor interventions, rFVIIa was administered in the range of 10-30 mcg/kg. Afterall rFVIIa for factor VII deficiency was well tolerated and maintained effective hemostasis with good clinical outcomes. In factor VII deficiency, surgical intervention and management of spontaneous bleeding may be difficult due to the variability of symptoms and bleeding clinic and the independence of bleeding risk from factor level. However, a road map can be drawn by considering published studies, center experiences and evaluating the clinical characteristics of the patient.

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