800

HEMOPHILIA: A SINGLE-CENTRE EXPERİENCE OF 12 YEARS

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Introduction:

- Hemophilia A and B (HA,HB) are the most common X-linked inherited bleeding disorders.
- Development of neutralizing inhibitory antibodies to FVIII and FIX is a challenging treatment-related complication of hemophilia.
- Aim of this study is to evaluate demographic and clinical characteristics and treatment outcomes of patients treated in a tertiary unit in Turkey between 2003 and 2015.

Material and Method:

- We retrospectively reviewed the medical records of 92 haemophilia A, and 17 haemophilia B patients.
- There were 34 mild, 17 moderate, 41 severe haemophilia A and 6 mild, 2 moderate, 9 severe haemophilia B patients.

Results:

- Twenty five of the 41 severe HA patients were under primary prophylaxis, where as 14 patients were on-demand treatment.
- 3 of 25 patients on primary prophylaxis were using plasma derived factors, 22 were treated with recombinant factors.
- Two severe HA patients with inhibitors are on prophylaxis with bypassing agents.
- One moderate HA patient was on primary prophylaxis (recombinant factor) and others were on demand therapy.
- 5 of 9 severe HB patients were on primary prophylaxis with a plasma derived factor and 1 was on prophylaxis with a recombinant factor.
- Radioactive synovectomy (RAS) was performed in 7 patients and was applied a second time in the same joints in 2 severe HA patients.
- 5 severe HA patients developed inhibitors to FVIII. In 2 patients, inhibitors were low titer and transient. Prophylaxis with recombinant factors were continued in these patients and antibodies disappeared in 6 and 8 months, respectively.
- In 3 severe HA patients who are on prophylaxis with recombinant factors, high titer inhibitors developed (6 BU, 32-43 BU, 45-51 BU). Immune tolerance treatment was started at 20, 36, 12 months in these patients, respectively. ITI was successfull in 1 (complete remisson) at 3 months. Inhibitor titers decreased however did not disappear in 2 patients at 21 and 20 months, respectively.
- None of the hemophilia B patients developed inhibtors.

Conclusion:

- Hemophilia is a serious congenital bleeding disorder that requires early diagnosis, intensive family and patient education, and regular comprehensive care to prevent life-threatening complications and potentially lifelong disability.
- Radioactive synovectomy is a treatment option in patients with chronic artropathy especially in countries like ours in which primary prophlaxis is still not widely practiced as it should.