RETROSPECTIVE ANALYSIS OF 115 PATİENTS WİTH HAEMOPHILIA A IN A SINGLE MEDICAL CENTRE

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AIM:

Prophylactic treatment for severe haemophilia A is likely to be more effective than treatment after bleeding occurs, however prophilaxis is costly. Whereas the benefits of primary prophylaxis are well documented, data related to secondary prophylaxis are limited. Our aim is to share our data regarding to secondary prophylaxis and to point out the benefits of secondary prophylaxis.

MATERIALS AND METHODS:

Patients were grouped in to four according to bleeding frequencies:

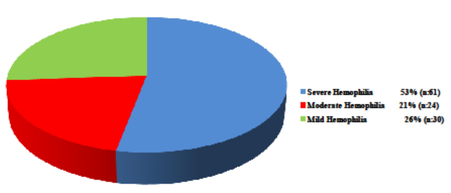
Group 1: Bleeding of 3-4 times in a month
Group 2: Bleeding of 1-2 times in a month
Group 3: Bleeding of less than once a month
Group 4: No bleeding

The target joint involvements were recorded. Patients response to prophylaxis treatment were also recorded by comparing the frequencies of bleedings before and after prophylaxis by chi-square test . Patients with radioactive synoviectomy were evaluated for both response and treatment complications.

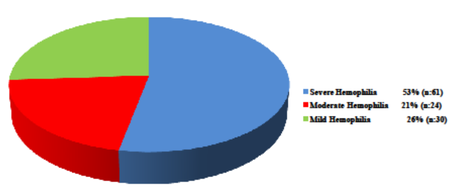
RESULTS:

We are presenting the data of 115 male with the diagnosis of haemophilia A, aged between 11-338 months (mean is 178 months). Mean age of diagnosis was 28 months (1-144 months).

The classification of patients according to factor levels



The classification of patients according to factor levels



In 56 (48.7%) of 115 patients there were target joint involvement and in 29 patients 2 joints were affected. Among patients with target joints, 7 had grade I, 14 had grade II, 18 had grade III, 10 had grade IV arthropathy. In 4 patients there were target joints but no arthropathy had developed.

Table 1: Target joint involvement according to hemophilia severity:.

Severe hemophilia	49 of 61 patients (80%)
Moderate hemophilia	5 of the 24 patients (20.8%)
Mild hemophilia	One of 30 patients (3.3%)

Table 2: The distribution of patients with 2 target joint involvement:

Severe hemophilia	28 of 61 patients (45.9%)
Moderate hemophilia	One of 24 patients (4%)

Table 1: The localization of target joints:

	Patients with one target joint	Patients with two target joints
Knee joint	37 (67.3%)	44 (53%)
Ankle joint	13 (23.7%)	27 (32%)
Elbow joint	5 (9%)	11 (13%)
Hip joints	-	2 (2%)

When the results were evaluated, it was seen that if patients had one target joint, involved joint was knee in the first order, followed by ankle and elbow joints. If two joints were involved, the order of involvement did not change but the percentage of knee joint involvement was decreasing while ankle and elbow joints were increasing. Hips were also involved if two target joints were present.

Of all the patients, 42 (36.5%) were under secondary, 3 (2.6%) were under primary prophylaxis.

By secondary prophylaxis bleeding episodes decreased by 78% (32). Nine had no change in bleeding (16%). By comparison of bleeding frequencies before and after prophylaxis by chi-square test, it was shown that the difference was statistically significant ($p<0,01$).

Two patients under primary prophylaxis had no significant bleeding.

Twenty eight patients that had radioactive synoviectomy, of whom 17 had more than one joint and/or more than one radiosynovectomy to the same joint. Twenty four had no complications, 2 had extraarticular leakage and 2 had intraarticular hemmorhage. Nineteen patients (68%) had controlled bleeding after procedure, but 5 were accepted as unresponsive (18%). Four patients were lost from follow-up after procedure (14%).

Bleeding control

CONCLUSION:

Compared with reports from industrialized countries, our experience with TR ALL- BFM 2000 protocol showed a similar remission rate, but a lower 5-year EFS, and a higher mortality result from infectious complications and insufficient supportive care.