

SINGLE CENTRE EXPERIENCE IN PREVIOUSLY UNTREATED PATIENTS WITH HEMOPHILIA

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INTRODUCTION

- Hemophilia A is an X-linked inherited bleeding disorder. Prophylactic treatment of children with hemophilia increased the quality of life.
- However, about 25% of the children with severe hemophilia A develop inhibitors against factor VIII (FVIII).
- Inhibitor development is a complex and multifactorial process influenced by a number of factors.
- We report our PUP patients who admitted to our centre in last 3 years.

MATERIAL-METHOD

- An analysis of PUP patients between 2013-2015 was carried out.
- Demographic characteristics, first factor exposure day (ED), prophylaxis age, exposure day were recorded.
- All patients were screened for inhibitor development after 3-4 ED in the 20 ED and 10-15 ED in the 20-50 ED.

RESULTS

- Totally, 15 PUP patients were admitted to our centre and median age at diagnosis was 9 months (range:0.5-19).
- All of them were severe Hemophilia A and 10 had family history.
- One patient was excluded because of irregular follow-up.
- In 13 patients median factor exposure age was 10 months (range:0,1-14), 1 patient haven't still have any factor exposure.
- Thirteen patients were began primary prophylaxis and median age was 12 months (range:1-20). The reasons for prophylaxis were hemarthrosis for 4 patients and 1 had deep muscle hemorrhagia.
- The patient who received prophylaxis at 1 month had intracranial hemorrhagia.
- **Three patients developed inhibitor.**
- **One patient who was treated with plasma derived FVIII had low titer inhibitor (4 BU)** after 8th ED and had no family history of hemophilia. This patient continued FVIII prophylaxis at the dose of 50U/kg three times a week and around 1 year his inhibitor was negative, recovery/half life of FVIII was adequate. **The other two who received recombinant FVIII had developed high titre** after 19 ED and 9 ED and had family history of hemophilia and also one had family history of inhibitor.
- Nine of 11 patients continued 50 ED and 1 continued 40 ED.

CONCLUSION

- Inhibitors presents certain challenges in people with hemophilia.
- In our PUP patients who were treated prophylaxis had inhibitor 23%. Inhibitor was developed in 20 ED.
- Although limited number of patients, our study is important for creating treatment guidelines at the emerging countries..

