



## “Hematuria in Hemophilia” - A One Year Study

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### Abstract

Hematuria is the excretion of red blood cells in the urine. There are various causes of hematuria which includes renal, postrenal, hematologic and vascular. These vary with age, sex, race, risk factors, personal and family history. Recently it was noticed that one of the cause of hematuria was coagulopathy like hemophilia. A prospective study of 50 hemophilia cases done from January 2021 to January 2022 on clinically diagnosed cases of hemophilia who came for evaluation of hematuria to Hematology unit, Department of Pathology, JJMMC and Karnataka hemophilia society, Davangere, Karnataka, India. In 50 hemophilia cases, 12 cases having hematuria were studied for aetiology, prevalence and management.

**Keywords:** Hematuria; Hemophilia; Hematoma; Fluids; AHF

**Key Message:** Do not treat with AHF as first line treatment in hemophilia patients having hematuria.

### Background

Hematuria is a problem arising in the genitourinary tract anywhere from the glomerulus to the urethral meatus that can result in RBC's in the urine. Hematuria is defined by the American Urological Association as greater than 3-5 RBC's per HPF. Hematuria is common, can be frightening whether gross or microscopic and sometimes life-threatening. The amount of distress caused to patients who experience hematuria, especially first time hematuria, is often high.

Hematuria can result from several causes. Initially, it is important to determine if it is truly blood in the urine that is observably reddish/brown in appearance. There are many foods and medications that can lead to a blood coloured urine [1]. There are various causes of hematuria which includes renal, postrenal, hematologic and vascular. These vary with age, sex, race, risk

factors and personal/family history. Recently it was noticed that one of cause of hematuria was due to coagulopathy like hemophilia. Now a days its very important to know the management of hematuria in hemophilia [2].

Thus, the aim of the study is to rule out actual cause of hematuria and prevalence of hematuria in hemophilia patients and manage accordingly.

### Material and Methods

A prospective study of 50 hemophilia cases done from January 2021 to January 2022 on clinically diagnosed cases of hemophilia who came for evaluation of hematuria to Hematology unit, Department of Pathology, JJMMC and Karnataka hemophilia society, Davangere, Karnataka, India. Demographic data, diagnosis, USG Abdomen and Pelvis findings and type of management is noted. Requisition forms and registers containing information regarding patients was taken from Karnataka hemophilia society.

**Inclusion criteria**

Patients diagnosed as Hemophilia were included.

**Exclusion criteria**

Acquired causes of bleeding disorders were excluded.

**Statistical analysis**

- Results were subjected for appropriate statistical analysis.
- For categorical variables- Frequency and percentages were used.

**Results**

Out of 50 hemophilia cases, 32(64%) are hemophilia A, 18(36%) are hemophilia B. Among them 12 patients presented with hematuria are subjected for evaluation. Age group ranged from 5-40 years with a mean age of 23 years. Most common affected age group was 0-25 years accounting for 92%, 25-50 was 8% with all affected patients being male.

In these 12 cases of hematuria, factor VIII was deficient in 7(58%) and factor IX in 5(42%) cases. Severe factor deficiency is seen in 5(42%) cases in which 3 of them are of factor VIII and 2 with factor IX. Duration of symptoms ranged from 3-15 days. Urinalysis in all 12 cases showed >3-5 RBC's/HPF. USG abdomen and pelvis was done in all 12 hemophilia cases with hematuria. Most common finding was hematoma in bladder and other causes are hydronephrosis, cystitis, calculus and ureterocele (Table 1). All 12 cases were managed with Oral and IV fluid replacement in which hematuria subsided in 10(83%) cases but persisted in 2(17%) on follow up which needed AHF (Anti hemophilic factor) therapy.

n = 12	
Hematoma in bladder	5
Hydronephrosis	3
Normal	2
Cystitis	2
Calculus	1
Ureterocele	1

**Table 1:** USG abdomen and pelvis findings.

**Discussion**

This study was performed to evaluate the association of hematuria with patient age, race, hemophilia type and severity,

treatment regimen. Out of 50 hemophilia cases all 12 cases having hematuria showed >3-5 red cells in urine, younger age group were more affected and most common imaging finding was hematoma in bladder. Despite fluid replacement there was persistant hematuria in 2(17%) cases which are of hemophilia A category showing high prevalence in them. 2(17%) cases like cystitis needed antibiotic treatment for infection control.

One more similar study done by Siquiera., *et al.* included 179 male patients. Mean age was 26 years, the majority of them were diagnosed with hemophilia A (76%) and 66% presented a severe disease where the prevalence of hematuria was 39%, of these a half presented at least one macroscopic episode [6].

Neil Osterweil study with a total of 93 patients, 67 with hemophilia A and 26 with hemophilia B, were included. In all, 43 patients (47%) were identified as having hematuria, with a median of seven red cells. Hematuria was seen in 37 patients with hemophilia A (55%), and in 6 patients with hemophilia B (23%). Characteristics associated with risk for hematuria included older age and hemophilia A which was similar to our study except younger age group being more affected in our study. Imaging studies available on 24 out of the 93 patients showed renal calculi in 3 patients, minor pelviectasis in 1, and congenital dysplastic left kidney, ureterocele, and right hydroureter in 1 patient each [7].

Study done by Qvigstad C revealed prevalence of macroscopic hematuria was higher among individuals with severe hemophilia similar to our study [8].

Thus management of hematuria in hemophilia patients is different from hematuria due to other causes which includes excess fluid intake, bed rest, urinalysis, USG Abdomen and Pelvis to rule out other causes. First treat with fluid replacement and follow up after 1 week if bleeding didn't subside then AHF therapy should be done under forced diuresis with IV fluids. Antifibrinolytic drugs is strictly contraindicated in management of hematuria in hemophilia in view of impending complications like clot colic even to the extent of causing renal failure.

**Conclusion**

Hematuria is spontaneous but uncommon manifestation in hemophilia necessitating clinical and radiological evaluation to rule out other causes like calculi formation. Most of the cases are

managed by conservative line of management with reassurance, bed rest and fluid replacement therapy. AHF replacement therapy is indicated only in cases of persistent hematuria and decreasing hemoglobin. AHF therapy is indicated but under forced diuresis to prevent clot colic and related complications.

### Bibliography

1. Megan Walsh. "Hematuria: Blood in urine". *National Hemophilia* 193 (2016).
2. Willis G C and Tewelde S Z. "The Approach to the Patient with Hematuria". *Emergency Medicine Clinics of North America* 37 (2019): 755-769.
3. Quon DV and Konkle, BA. "How we treat: haematuria in adults with haemophilia". *Haemophilia* 16 (2010): 683-685.
4. Srivastava A., et al. "WFH Guidelines for the Management of Hemophilia". 3<sup>rd</sup> edition. *Haemophilia* 26 (2020): 1-158.
5. Yeoh., et al. "Macroscopic haematuria: a urological approach". *Australian Family Physician* 42 (2013): 123-126.
6. Siqueira DB., et al. "Prevalence of hematuria in patients with hemophilia A and B and its association with comorbidities: Experience from a Brazilian center". *Hematology, Transfusion and Cell Therapy* 43 (2021): 231.
7. Neil Osterweil. "Hematuria a common finding in paediatric hemophilia". *Paediatric News* (2016).
8. Qvigstad C., et al. "Hematuria in aging men with hemophilia: Association with factor prophylaxis". *Research and Practice in Thrombosis and Haemostasis* 4.2 (2020): 309-317.