Summary
30 patients of hemophilia were admitted in pediatric surgical unit in Sir Salimullah Medical College, Mitford Hospital, Dhaka, Bangladesh from September, 1995 to December, 2001. Of which 15 (50%) patients had severe hemorrhage following circumcision, 25% (9) patients of this group had their circumcision by Hazams (quacks only perform circumcision), rest 25% (3) cases were done by registered doctors (MBBS) at their chambers, upazilla health complex and even in district hospital without any examination of bleeding and clotting time. 7 (23.35%) patients presented with uncontrolled bleeding from different sites of the body due to injury. 3 (10%) patients were the patients of hemarthrosis of knee and elbow joints. Another 3 (10%) patients presented with history of fresh bleeding per rectum without any pathology in anal canal, rectum and large intestine. 2 (6.65%) patients had hematuria. Prolonged clotting time was the diagnostic criteria. Fresh blood transfusion was needed in 5 (16.66%) cases due to delayed hospital attendance with severe anaemia. Only in 2 (6.65%) cases factor VIII was given. Aminocaproic acid and tranexamic acid was applied in every cases in intravenous or oral route. Surgical treatment was done in few cases. There were 2 (6.66%) death in this study.

Introduction
Hemophilia is a keen caring disease in case of male pediatric surgical patients. There is no statistical data of hemophilic patients in Bangladesh. In India 1 in 5000 male is hemophilic and each year 1300 children are born with this hemorrhagic disorder. There are nearly 50,000 patients with severe hemophilia A at present. Data will be more or less same in our country. There are 20,000 hemophilic in the United States. People of our country not at all aware of this disease. Manifestation starts after the circumcision when hemorrhage could not be stopped by usual procedures. So, early detection is possible in our country. But picture is quite different in other developing country where circumcision is not a compulsory religious custom. Ti: is fact that only 3 to 5% of the affected people of this disease have the adequate resources and access to the medical treatment in South East asia. In developed country greater availability and use of cryoprecipitates and factor concentrates has greatly improved and the management of hemophilia with an overall reduction in the morbidity and mortality. Combined development of comprehensive hemophilia treatment centers there has been a remarkable change in the outlook for these patients. So, these patients have near normal life but in developing countries the management of these patients continues to be a major problem.

Table -1. Types of different presentations of hemophilic patients.

<table>
<thead>
<tr>
<th>Types of presentations</th>
<th>Number (n = 30)</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bleeding after circumcision</td>
<td>15</td>
<td>50%</td>
</tr>
<tr>
<td>Bleeding from different sides of the body</td>
<td>7</td>
<td>23.65</td>
</tr>
<tr>
<td>Hemoarthrosis</td>
<td>3</td>
<td>7</td>
</tr>
<tr>
<td>Fresh bleeding per rectum</td>
<td>3</td>
<td>10</td>
</tr>
<tr>
<td>Persistent bleeding from the teeth</td>
<td>2</td>
<td>6.35</td>
</tr>
</tbody>
</table>

Methods, materials and managements
It was a retrospective study. 30 patients of...
Hemophilia with hemorrhage on the 2nd post operative day

Post circumcision hemorrhage due to hemophilia.

7 Cases presented with nonstop bleeding from the different sites of the body due to trauma. Trauma were mainly from fall, injury by the hard toy, sharp edge of the furniture’s, betting of the tongue and lips. These injuries were more in case of toddlers and children. Surgical trauma was also an important factor.

In blunt trauma, one boy presented with a big hematoma in the left thigh. It was increasing day by day and responding to conservative treatment. In surgical trauma and punctured wound bleeding was not stopped. No investigations even BT, CT were done in any patients. As soon as the patients were admitted in hospital, hemophilia was suspected 3 patients presented with hemarthrosis. This occurred spontaneously in one case and in 2 cases following a minimal unforgettable trauma.

Table II: Types of trauma

<table>
<thead>
<tr>
<th>Types of trauma</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgical trauma</td>
<td>2</td>
</tr>
<tr>
<td>Blunt trauma</td>
<td>3</td>
</tr>
<tr>
<td>Punctured wound</td>
<td>2</td>
</tr>
</tbody>
</table>

Table III: hemarthrosis

<table>
<thead>
<tr>
<th>Types of trauma</th>
<th>No</th>
<th>Joint involved</th>
</tr>
</thead>
<tbody>
<tr>
<td>Trauma</td>
<td>2</td>
<td>Knee and elbow</td>
</tr>
<tr>
<td>No trauma</td>
<td>1</td>
<td>Knee</td>
</tr>
</tbody>
</table>

The joints were swollen and warm and had restricted movement. Irritability, guarding and limitation of movement of the affected joints were present in a 3 years boy.

3 patients presented with fresh bleeding per rectum which was painless. Digital examination revealed nothing. Proctoscopic findings were normal. Medical treatment for different types of dysentery were failed. Colonoscopy done but it was completely normal. Blood examination confirmed the diagnosis. 2 boys were presented with continuous bleeding from teeth.
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Results and Management

Patients with post circumcisonal bleeding usually admitted average 2 to 5 days after the procedure. None of the patients had any investigations even BT, CT. The surgeons tried to stop the bleeding in various procedures such as tight dressings, applying gauge tourniquet around the root of the penis, re-suturing, ligation of blood vessels. When all attempts failed, patients were referred to the hospital. All the patients were anaemic, just after receiving patients, blood was sent for grouping and cross matching. At the same time BT, CTs were done. Factor VIII estimation was not possible. Tight dressings, dirty clothes were urgently removed. Clean dressings were applied. Where there was any clot formation and dressings were very much adherent to the wound, we didn't try to make it clean. Intravenous fluid was started. First we gave aminocaproic acid and tranexamic acid. They inhibited plasminogen and thus prevented the lysis of the clot.

The dose of aminocaproic acid was 100mg/kg body wt every 6 hours intravenously and tranexamic acid 25mg/kg body wt orally or 10mg/kg body wt Intravenously every 6-8 hours. At the same time, when bleeding didn't stop within one hour, we started fresh blood transfusion. 75% of the patients responded with this treatment. Rest 25% patients were sent to operation theatre for exploration of the wound. Under general anesthesia dressings were taken out and stitches were off to see any bleeding. These were ligated. But where we didn't get any bleeding vessel, we usually applied continuous suture between skin and the mucous membrane of the penis. The patients were kept 4 to 7 days for observation. We didn't use any diathermy. In 2 patients of post circumcision bleeding, factor VIII was given keeping in mind that 80% of all hemophilia patients have factor VIII deficiency which was a classical hemophilia.

Dosage calculation of factor VIII

One unit of factor VIII per kilogram of body weight increases the factor VII activity by 2%. It has biological half life of 8 hours. The amount of factor required is calculated as:

\[
\text{Factor VIII dose} = \frac{\% \text{ rise required} \times \text{body weight}}{2}
\]

Skin laceration in hemophilia was treated on same line as normal persons. We sutured the gap when it was necessary by silk or catgut. Sometimes we applied sterile adhesive like Duoderm or Nishipore. When there was no gap and stitches were not required, ice wrapped in thick cloth was applied immediately at the local site to control bleeding. Gauge soaked in dilute adrenaline (1:10,000 dilution) used for tropical hemostasis. At the same time aminocaproic acid was also tried. Result was good. Hemarthrosis cases were managed as follows:

1) By keeping the joint in position of least pain along with application of ice pack
2) Elevation of affected limb
3) Immobilization of the joint in functioning position
4) To relieve the pain, analgesics like paracetamol was given.
5) Aspirin was never given, because it absolutely contraindicated.
6) Physiotherapy was initiated as soon as the pain subsided to strengthen the muscles, stabilized the joint and to prevent contracture.

In hematuria aminocaproic acid or ranexamic acid should not be given as it stabilizes the clots which may result in clot colic. During discharge we give advises to the guardians of all the patients as follows:

1) Life of the children needs to be adjusted to
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minimize the risk of trauma
2) Soft plastic toy should be given for playing
3) All intramuscular injections are contraindicated and injection should be given intravenously or subcutaneously
4) Child should not be left unattended in an infant seat or on a raised unprotected beds
5) Protective elbow and knee pads can be used.
6) Furniture with sharp edges must be avoided or the edges be padded. Minimum furniture should be kept in house.
7) Sports like swimming, badminton, fishing and walking have minimal risk while basketball, cricket and jogging have moderate risk, football, high and long jump have the greater risk. The risk of injury can be reduced with proper training, protective clothes and equipments.

Discussion

Hemophilia A and B are the sex-linked recessive bleeding disorders caused by decreased levels of functional procoagulant factors VIII and IX, respectively. Approximately, 80% of all hemophilia patients have factor VIII deficiency. The remaining 20% have factor IX deficiency which is called Christmas disease. But in our country it is very difficult to measure both the factors, for that reason we must depend on the simple test of clotting time.

In our religion, it is a rule to perform circumcision in the children. The most of circumcisions are performed by village quack without knowing coagulation status of blood. Even the registered practitioner who usually performs circumcision doesn’t do this investigation. Problem starts with them. (Fig - II &III) Hemophilia is diagnosed in our country after manifestation of bleeding after circumcision which can’t be controlled by usual procedure of hemostesis. (Fig-IV, V) Bleeding problems during operation were not observed but 23% of all serious operations were complicated by post-operative hemorrhage. 4

Hemophilia patients are classified into three categories based on their level of circulating procoagulant.
1) Severe hemophilic - circulating procoagulant factor VIII is less than that of 1% high risk hemorrhage. Usually needs replacement therapy. 5,6
2) Moderate hemophilic- circulating procoagulant level is 1 to 5% . Spontaneous hemorrhage occurs infrequently.
3) Mild hemophilic - procoagulant levels more than 5%, rarely have bleeding problems and most have problems only major trauma or surgery. 5,7

In this study mostly the patients were of mild and moderate type of hemophilic (Fig-VI &VII). But confirmation could not be done due to lack of investigation facilities, financial constrains, devoid of knowledge of seriousness of the disease. Death was 2. Because those were the cases of moderate and severe hemophilia. These are also very difficult to manage even in the most developed country like USA or UK.

The management of this type of patients requires close co-operation among the surgeons, hematologists, pediatricians, and blood bank personnel. 8 Regarding any surgery careful pre-operative planning is essential. Fresh blood is to be kept ready. If possible adequate supply of clotting factor must be there. Patient must be admitted in the hospital a day before surgery. A bolus dose of factor VIII must be given. If there is per-operative hemorrhage fresh blood to be transfused. If there is any drainage the amount of collection must be monitored. In hemophilia after a minor surgery like circumcision, patient should be kept in hospital at least for a week. But for major surgery in our country, it is very difficult to manage all these patients. Because facilities are very poor. Factor VIII is very costly. Drugs like titre inhibitor porcine factor VIII, prothrombin complex concentrates (PCC) or activated PCCs such as autoplex or FEIBA are not available here. In the field of management of hemophilic patients especially moderate and severe type we are still in the primitive stage.
References

1. ICMR Task Force. Collaborative study of hemophilia, New Delhi, Council of Medical Research, 1990