



# Successful Anaesthetic Management Of Moderate Haemophiliaa Patient With Acute Subdural Hematoma Posted For Emergency Craniotomy

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## ABSTRACT:

Intracranial hemorrhage in patients with haemophilia is associated with high mortality and sequelae. We report the case of 35 year male k/c/o Haemophilia A, who presented with acute subdural hematoma and was posted foremergency craniotomy with clot evacuation. Main concerns for the anaesthesiologist in such patients are the associated excessive & prolonged bleeding intraoperatively,ensuring adequate restoration of clotting deficiency by Factor VIII replacement as well as theneuroprotection protocols which have to be taken care of.Perioperative management of patient of hemophilia with intracranial bleeding is very challenging and requires a multi and interdisciplinary team approach in the operating room and intensive care unit.

**KEYWORDS:** anaesthesia, haemophilia, subdural hematoma, emergency craniotomy.

## INTRODUCTION:

Haemophilia is an X-linked recessive hereditary disorder characterised by deficient or defective Factor VIII coagulant<sup>1</sup>. Intracranial hemorrhage is a life threatening complication of haemophilia. The site of bleeding is equally distributed between subdural hematoma, intracerebral and subarchnoid hemorrhage. Acute subdural hematoma is one of most lethal form of intracranial injury. Prompt surgical evacuation, when indicated has better prognosis.Prior to the age of modern blood banking mortality and morbidity of hemophiliac patient was much higher<sup>3</sup>. Development of factorVIII concentrates has greatly revolutionized the management of haemophiliac



patients<sup>3</sup>. Mortality from intracranial hemorrhage in haemophilia, however is still high.

**CASE REPORT:** A 35 year male non-hypertensive and non-diabetic presented in emergency room with headache, vomiting since 2 days and altered sensorium since 1 day following trivial fall at home 2 days back. There was no history of fever, any drug or alcohol intake. He was a diagnosed case of Haemophilia A with past history of repeated hemarthroses and also factor VIII infusions four times. There was no h/o bleeding from other sites. On examination in emergency unit patient was unconscious with stable vital signs (HR of 56/min and BP 118/78mmHg). His Glasgow coma scale (GCS) was 5/15 (E1M2V1) with bilaterally mid-dilated pupils not reacting to light. In view of poor GCS trachea was intubated with cuffed endotracheal tube no. 8.0 with extra care in manipulation and intubation of airway in view to avoid submucosal hemorrhage and shifted to intensive care unit for elective mechanical ventilation. The patient had no significant medical problems and family history

was not contributory. Computed tomography revealed a left fronto-temporo-parietal subdural hemorrhage with midline shift of 10mm to right and effacement of cisterns. Investigations revealed haemoglobin of 8.0g/dl with a normal bleeding, clotting time, thrombin and prothrombin time. Platelet count was 1,80,000/mm<sup>3</sup>. The activated prothrombin time (APTT) was 150 sec. Plasma factor level done preoperatively it was 0.02 IU/ml (2%). Preoperatively Factor VIII transfusion was done with 50U/kg followed by 25-30U/kg every 8 to 12 hourly. Neuroprotection measures in the form of inj. Mannitol (1gm/kg) IV 8 hourly, hyperventilation (to a PaCO<sub>2</sub> of 32 mmHg) and seizure prophylaxis (inj. Levetiracetam 1 gm loading dose followed by 500mg IV 12 hourly) were initiated. Patient was scheduled for emergency craniotomy with decompression of SDH. General anaesthesia with endotracheal intubation was planned with neuro-anaesthesia protocols. Inside the operation theatre two wide bore (16 G) peripheral lines were secured. Monitoring included Pulse Oximetry, Electrocardiogram, NIBP, EtCO<sub>2</sub> and urine output.



Patient was given antisialogogue inj. Glycopyrrolate 0.2mg IV and anaesthesia induced with Inj.fentanyl 100ug + inj.Propofol 80mg. Muscle relaxation was achieved with inj. Vecuronium 4mg. Anaesthesia was maintained with oxygen + air + sevoflurane and intermittent inj. Vecuronium. Also intraoperatively 1gm of tranexamic acid was given. Blood loss was 1000ml. Two unit PRBC and eight unit FFP were infused. Patient remained hemodynamically stable throughout the surgery and was shifted to ICU for postoperative elective ventilation. Postoperative analgesia was provided with inj. Fentanyl and inj. Paracetamol (1gm 8 hourly). Factor VIII assay done in post-op period and was found 20% of normal. In order to keep Factor VIII level of 100% till first 5 days and then 50% for next 7 days Factor VIII transfusion was given 50U/kg 12 hourly followed by 25U/kg for 14 days. Factor VIII and APTT monitoring was done on alternate days. On day five patient was electively tracheostomized. Patient showed signs of neurological improvement (GCS E2M3Vt) and was weaned off the ventilator on 10<sup>th</sup> post-op day.

On 25<sup>th</sup> postoperative day GCS improved to E3V5Vt and Factor VIII level of 60% was attained.

#### DISCUSSION:

Haemophilia is the most common hereditary bleeding disorder, with an incidence of 0.7-0.8/10,000. Classic haemophilia A is characterized by an inherited defect in Factor VIII while Factor IX deficiency leads to haemophilia B (Christmas disease)<sup>4</sup>. It is transmitted as an X-linked trait with variable expression; it is ordinarily carried by females who are unaffected. Thus it is a disease of males<sup>3</sup>. Haemophilia A represents almost 90% of haemophilia cases<sup>1</sup>. It is a bleeding disorder that has a spectrum of manifestations ranging from persistent bleeding after minor trauma to spontaneous haemorrhage<sup>2</sup>. Intracranial hemorrhage is a rare complication of hemophilia, with frequency of about 2.2-7.8%, and a mortality of 34%<sup>4</sup>. Diagnosis of hemophilia A is made on the basis of Factor VIII assay and depending on the activity, may be classified as "mild", "moderate"



or "severe"<sup>4</sup>. Factor VIII activity levels are reported in units, with 1IU/ml corresponding to 100% of factor found in 1ml of plasma. Normal plasma activity levels usually range between 0.5-1.5 IU/ml (50%-150%)<sup>1</sup>. Severe cases have less than 1% of the Factor and bleed spontaneously into joints and muscles. Moderately deficient patients have levels of 2% to 5%, while mildly affected patients have 7% to 15%. The moderate hemophiliacs have no spontaneous bleeding, but usually bleed after minor or major trauma. In mild hemophiliacs greater trauma is necessary for bleeding<sup>4</sup>. Our patient complained of episodes of bleeding into knee joint. Levels of 3-5% may adequately protect the patient from spontaneous bleeding, whereas patients with 10-15% of normal activity may be entirely asymptomatic. Haemophilia lowers the blood clotting factors, thus when a vessel is injured a temporary scab does form but the missing coagulation factors prevent fibrin formation and stabilization of clot. Thus a haemophiliac does not bleed more than a normal person, but always carries a risk of

prolonged bleeding and re-bleeding<sup>1</sup>. In areas such as brain or inside joints this can be fatal or permanently debilitating<sup>1</sup>. The diagnosis of haemophilia is often made from family history, laboratory finding such as greatly reduced factor VIII, and elevated PTT. The PT, bleeding time, platelet count and clot retraction will be normal in these patients, since none of these tests are dependent upon factor VIII<sup>1,3</sup>. The patient's haemoglobin may be low because of acute or chronic blood loss. When large haematomas are present, the serum bilirubin may be increased<sup>1</sup>. In preparing patients with haemophilia A for surgery, factor VIII levels are routinely raised to approach 100% of normal activity. It should be maintained for the first 3 postoperative days from the day 4 onwards it should be maintained at 80%, from 7th day onwards it is allowed to decline to 40% of normal activity<sup>1</sup>. The formula used to calculate the factor VIII dose is -  $N = \text{plasma volume (ml/Kg)} \times \text{weight (Kg)} \times \text{Percent activity increase}$  where N is the number of units required. Plasma volume is 40 ml/Kg for adults. Since half-life of factor VIII is



about 12 hours, it must be administered twice daily<sup>3</sup>. Factor VIII, in varying concentrations, is found in FFP, cryoprecipitate and Factor VIII concentrate. One ml of FFP contains 1 IU of clotting factor activity, while cryoprecipitate contains between 80 and 100 IU in a volume of 25 ml. Factor VIII concentrate contains between 250 and 2000 IU of activity in a reconstituted volume of 25 ml<sup>4</sup>. Anaesthesia considerations in these patients include 1) Avoiding intramuscular premedication 2) Vascular access itself does not cause excessive bleeding and should be appropriate for the proposed procedure 3) CVP line should ideally be placed guided by ultrasound 4) After induction of anaesthesia, extra care should be taken in manipulation or intubation of the airway as it can cause submucosal haemorrhages, which can prove life threatening. Nasal intubation should be avoided, as it can prove traumatic and bleeding from the site can lead to aspiration. 5) Care be taken during positioning of the extremities and pressure points should be padded to prevent intramuscular haematomas or haemarthrosis. 6) Post operatively

analgesics such as apirin and other NSAIDs should not be given as it can predispose to gastrointestinal haemorrhage. Inj. Paracetamol and patient controlled analgesia are found to be safe options. 7) The fibrinolytic inhibitors, epsilon amino-caproic acid (EACA) or tranexamic acid are commonly administered to reduce requirement of factor VIII as in our case we administered 1gm of inj. Tranexamic acid IV intraoperatively.

#### CONCLUSION:

Haemophiliacs are at risk of life threatening haemorrhages during surgery. Early intervention, careful management, and normalization of haemostatic defects with aggressive Factor VIII concentrates are essential. Adequate preoperative work-up, a team approach and blood bank support is essential in improving the outcome of these patients.

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