# Management of a Case of Congenital Afibrinogenemia for Extradural Hematoma Evacuation with Fresh Frozen Plasma

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**ABSTRACT**

*Afibrinogenemia is a rare bleeding disorder with an estimated prevalence of 1:1,000,000. It is an autosomal recessive disease resulting from mutations in any of the 3 genes that encode the 3 polypeptide chains of fibrinogen. Spontaneous bleeding, bleeding after minor trauma and excessive bleeding during interventional procedures are the principal manifestations. Replacement therapy is the mainstay of treatment of bleeding episodes in these patients and plasma-derived fibrinogen concentrate is the agent of choice. Cryoprecipitate and fresh frozen plasma are alternative treatments that should be used only when fibrinogen concentrate is not available. We report a case of congenital afibrinogenemia, a 3 years old child, posted for emergency intracranial hematoma evacuation who was successfully managed by fresh frozen plasma as other alternatives were unavailable.*

**Keywords:** *Afibrinogenemia, Fresh frozen plasma, Cryoprecipitate, Fibrinogen replacement therapy, Extradural hematoma.*

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## INTRODUCTION

Afibrinogenemia is a rare bleeding disorder with an estimated prevalence of 1:1,000,000.(1,2) It is an autosomal recessive disease and most patients are commonly descendent of consanguineous marriages(1). Afibrinogenemia results from mutations in any of the 3 genes (FGA, FGB and FGG) that encode the 3 polypeptide chains of fibrinogen. These mutations affect the synthesis, assembly, intracellular processing, stability or secretion of fibrinogen(3). Its incidence is higher in the Middle East, and in the southern of India, where parental consanguinity is frequent(2).

Spontaneous bleeding after minor trauma, excessive bleeding during interventional procedures are the principle manifestations. We report a case successfully managing a case of congenital afibrinogenemia presenting to us in emergency with extradural hematoma, using fresh frozen plasma (FFP).

## CASE REPORT

A 3yrs old female, weighing 15kg, patient presented to our emergency department with history of head injury after a fall from height and losing consciousness for 5 min. Her GCS was 12/15. On CT SCAN an extradural hematoma of size 4x9 cm was found. The patient was a known case of afibrinogenemia and was diagnosed at the time of birth due to bleeding from umbilical cord. Gene mutations was not done in our patient, however, a diagnosis of congenital afibrinogenemia was made by showing profoundly prolonged PT, aPTT, and an extremely low fibrinogen level. She was born of coansanguienous marriage and her elder sibling was also suffering from same condition. Her current blood investigations showed Hb 10gm/dl, Platlet 1,00000/mcl, TLC 10,000/mm$^3$, bleeding time was 14 min 40sec, PT>150 sec, aPTT> 140 sec and thrombin time =24 sec, INR >4, fibrinogen concentration was 50mg/dl. Her Renal function test and liver function tests were within normal limits (WNL).

Pt was posted for emergency craniectomy but as patient's coagulation profile was deranged surgery was withheld till correction of her profile. Being a poor patient she could not afford cryoprecipitate or fibrinogen concentrate and neither they were available at our government institute at that time at a subsidized rate. Fresh frozen plasma was the only alternative available in the hospital, hence the patient was transfused with four unit of ABO compatible FFP in 8 hours. Repeat coagulation profile after 8 hours showed bleeding time of 3 min, clotting time 4min, and INR 1.5 fibrinogen levels could not be obtained then. Though the patient did not deteoirate, repeat CT scan showed no reduction of the clot and her GCS too did not improve so we took the patient for craniectomy with explaining the high risk to the parents.

The child was taken inside the OT and ECG, Pulseoximeter, noninvasive blood pressure cuff were

attached. After securing an intravenous line under sevoflurane, she was premedicated with IV glycopyrrolate 0.02mg/kg, IV fentanyl 2ug/kg. Induction was done with with IV propofol 2mg/kg and IV rocuronium 0.5mg/kg to aid intubation. Gentle laryngoscopy was done trachea was intubated with 4.5 no. ETT

Craniectomy with evacuation of hematoma was done. The procedure lasted for an hour and blood loss was 250ml which was replaced with packed RBC concentrate. At the end of the procedure patient was reversed with inj neostigmine 0.75mg and inj glycopyrolate 1.2mg IV. After ensuring adequate power and response of the patient she was extubated after thorough oropharyngeal suction. She was shifted to pediatric intensive care for observation. Eventually patient was discharged from hospital after 5 days.

## DISCUSSION

Fibrinogen/ factor I plays an important role in blood coagulation, abscence or functional problem of which results in haemorrhage or thrombosis. Normal volume of fibrinogen in blood is 2-4g/l. These fibrinogen disorders can present as afibrinogenemia or hypofibrinogenemia (quantitative defects) or dysfibrinogenemia (qualitative defects). Afibrinogenemia is a very rare inherited bleeding disorder that affects both male and female of all races and ethenic origin.(3)

The estimated prevalence of afibrinogenemia is one in 10 lakhs. In congenital afibrinogenemia (fibrinogen <0.2g/l) bleeding can vary from slight to severe. Nearly 85% present with umbilical bleeding in the neonatal period(3). The diagnosis of afibrinogenemia is based on the presence of prolonged prothrombin, thrombin, reptilase and activated partial thromboplastin time, undetectable functional fibrinogen and absence or trace amounts of immunoreactive fibrinogen(4). Platelet adhesion and ADP-induced platelet aggregation are also impaired in these patients whereas thrombin- and collagen-stimulated platelet aggregation is normal(5). The exact diagnosis can be done by identifying the molecular defect.

Bleeding that affects the head, neck, chest and abdomen in such patients can be life threatening requiring immediate medical attention. Replacement therapy is the mainstay of treatment in these patients, Fibrinogen concentrates are the choice of replacement therapy in afibrinogenemic patients. Virus inactivation, small-volume infusions, and low risk of allergic reaction are their major advantages over other replacement therapies (6). Cryoprecipitate and FFP shoud be infused only on an emergency basis, when fibrinogen concentrates are unavailable(7).

Transfusing FFP has several drawbacks, such as extended administration time, transfusion-related complications and less efficacy. Blood group matching is required and, as FFP is stored at 20$^{o}$C, thawing time needs to be taken into account. High volumes are needed for effective fibrinogen supplementation as the concentration in FFP is low (typically around 2.5g/l). FFP is not typically subjected to viral inactivation procedures, so there are risks of viral transmission. Treatment with methylene blue or solvent–detergent can be employed, but this can reduce the level of fibrinogen in the end-product (particularly in the case of methylene-blue treatment, where the reduction is around 30%)(8). Current guidelines say virally inactivated FFP should be preferred to non-virally inactivated cryoprecipitate unless volume overload poses unacceptable risks.(6)

Unavailability of fibrinogen concentrate or cryoprecipite led us to use FFP for correction of her deficiency which controlled her bleeding. We assumed that 1lit of FFP was required to give 2.5g of fibrinogen to the patient hence 4U of FFP (1U= 200ml) were transfused. Since there was risk of volume overload transfusion was done slowly over a period of 8 hours. Though we couldn't measure her fibrinogen levels, correction of her other coagulation measures made us go ahead with the surgery and successfully saved the patient.

## CONCLUSION

Even though congenital afibrinogenemia is a rare disorder patient may present to anaesthesiologist for emergency surgery. Fibrinogen concentrate is the choice of replacement therapy. Cryoprecipitate and FFP are suitable alternatives in case of unavailability. Knowledge about the condition and the dose of replacement therapy, which in our case was FFP, is necessary for proper management of the patient.

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