Congenital factor VII deficiency: Multidisciplinary approach is the key to successful perioperative outcome

Sir

Factor VII (FVII) is a vitamin K-dependent coagulation factor with short half-life (3-4h), responsible for the activation of the coagulation pathway. Congenital deficiency of FVII is a rare disorder with an incidence of 1:500000.[1] It is usually diagnosed in infancy. Brain and gastrointestinal haemorrhage is the most common presentation, seen in 60%-70% of patients. They are at an increased risk of post-traumatic and post-operative bleeding.[1] Infusion of Recombinant factor VIIa (rFVIIa) is a safe and effective therapy in the management of haemorrhage and prophylaxis before surgery.

A 2 month old male child, born of consanguineous marriage, weighing 2.7 kg, and a diagnosed case of FVII deficiency, was posted for right herniotomy. FVII deficiency was diagnosed 10 hours after birth during the diagnostic work up for brownish aspirate through orogastric tube. At that time, plasma FVII level was <1% (70-150 biological reference value). The child was managed with discontinuation of orogastric feeding followed by fresh frozen plasma (FFP) transfusion till normalisation of coagulation profile. Family history was unremarkable.

Pre-operative laboratory test showed elevated prothrombin time (PT)/international normalized ratio (INR) (PT-39.6 s/INR-3.4) with normal activated partial thromboplastin time (aPTT), liver enzymes and platelet count. The child received 30 μg of rFVIIa five hourly with PT/INR monitoring. Values were normalised after 90 μg of rFVIIa over 15 h with the last dose being 1 h before the surgery. Standard non invasive monitoring was done. Anaesthesia was induced with thiopentone 15 mg, succinyl choline 5mg followed by endotracheal intubation by an experienced anaesthesiologist and maintained with oxygen, nitrous oxide, isoflurane and controlled ventilation. Ketamine 3 mg, paracetamol 75 mg and wound infiltration with 0.125% bupivacaine through hypodermic needle were given for adequate analgesia. The surgery was uneventful with minimal blood loss. Incision site bleeding or oozing was not seen in the postoperative period. rFVIIa (30 μg)was repeated prophylactically 2 h after surgery. PT/INR monitoring was done till postoperative day 2 and was normal. The patient was discharged without any episode of bleeding or thrombosis on postoperative day 6.

Multidisciplinary approach involving haematologists, anaesthesiologists and surgeons to prevent life threatening haemorrhage is necessary. We planned general anaesthesia without caudal block to avoid any bleeding associated with the procedure. Care was taken to avoid any sort of trauma to the child during laryngoscopy, suctioning and positioning.

Inherited FVII deficiency is an autosomal recessive disorder, commonly seen in births following

consanguineous marriage.[2] Type I deficiencies result from decreased biosynthesis or accelerated clearance. Type 2 deficiencies arise due to mutation in the FVII gene located on chromosome 13[2]. Clinical bleeding can vary widely and does not correlate with FVII activity in plasma.[3] Elevated PT/INR with normal aPTT indicates FVII deficiency.[2] FVII activity was not monitored perioperatively because of inconsistent relation with clinical bleeding. The recommended dose of of rFVIIa is 15-30 μg/kg and must be re dosed every 4-6 hourly until haemostasis is achieved, with timely monitoring of coagulation parameters. [1]Complications of replacement therapy with rFVIIa include thrombosis and production of antibodies against FVII, usually seen with the frequent and large dose infusion.[4] Measurement of FVII activity and PT-INR is helpful for preventing overdose administration. Maintaining FVII level of at least 15%-25% provides adequate haemostatic level for most surgical procedures.[2] Recently, continuous infusion of rFVIIa has been used to maintain appropriate plasma concentration with a good safety profile.[5] Other treatment alternatives include FFP and prothrombin complex concentrate containing factors II, VII, IX and X.[2]

Successful perioperative management of an infant with FVII deficiency necessitates timely administration of rFVIIa with PT/INR monitoring.

We conclude that the management of infants with FVII deficiency necessitates timely administration of rFVIIa with PT/INR monitoring to avoid life threatening haemorrhage perioperatively.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_2_17

How to cite this article: Tandale SR, Khedkar SM, Yadav AH, Ajatshatru S. Congenital factor VII deficiency: Multidisciplinary approach is the key to successful perioperative outcome. Indian J Anaesth 2017;61:275-6.