

## CASE REPORT

### Factor VII deficiency in a Home Delivered Neonate

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#### ABSTRACT:

Factor VII deficiency is a rare inherited autosomal recessive bleeding disorders, and is a chameleon disease that hides behind the wall of Hemorrhagic diseases of newborn that are usually Vit K dependent . The clinical phenotypes range from asymptomatic condition—even in homozygous subjects—to severe life-threatening bleedings (central nervous system, gastrointestinal bleeding) and hence presenting a case of a 9 day old neonate delivered at home who presented with oro nasal bleeds followed by seizures due a intra ventricular haemorrhage and chest X ray showing features of pulmonary haemorrhage. Child was initially treated symptomatically with blood transfusions, FFP and cryo- precipitate but did not improve until the factor VII values were sent and found to be deficient. Following which replacement therapy was given and all bleeding manifestations settled. Child later was followed up and found to be hemodynamically stable with adequate weight gain and no further bleeding manifestation.

**Key words:** Cryo- precipitate, Factor VII deficiency,

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#### BACKGROUND:

Congenital factor VII deficiency is rare, affecting an estimated 1 in every 500,000 people. Males and females are equally affected.

Despite Vit K deficiency being the most common cause of bleeding in a home delivered neonate, we must also rule out factor deficiency to be a viable diagnosis as seen in the above case.

#### PRESENTING FEATURES:

9 day old, male, preterm neonate delivered at home, with a birth weight of 1.515 kg, presented to department with complaints of oro-nasal bleeding and increased work of breathing. Child developed seizures during hospital stay. No relevant medical, social or family history.

#### INVESTIGATIONS:

- Hb - 8.1g/dl

- Sepsis screen – negative
- PT - 22.6/INR 1.72
- APTT- Prolonged
- Factor VII levels -30% of optimal value (deficient )
- Chest XRAY - suggestive of pulmonary haemorrhage
- USG skull - features suggestive of intra ventricular haemorrhage
- 2d ECHO - TR jet >55% hence suggestive of PPHN( persistent pulmonary hypertension of newborn )

#### DIFFERENTIAL DIAGNOSIS:

- Hemorrhagic disease of newborn
- Sepsis

## TREATMENT

- Intubated and symptomatically treated.
- Inj PHENOBARBITAL given in view of seizures.
- 5 days of Vit K , FFP, Cryo-precipitate given but no relief.
- Inj NOVO 7 (Factor VII ) given over 3 days.
- Inj SILDENAFIL given in view of PPHN.

## OUTCOME AND FOLLOW-UP

After administration of Factor VII child's bleeding manifestations settled , child's saturation maintained and we were able to extubated within 4-5 days. Child's general condition improved and was discharged after adequate weight gain.

On follow up the child had adequate weight gain, tolerating feeds well and had no further episodes of bleeding. Parents were advised to watch for any bleeding manifestation and to follow up for replacement therapy if required.

## DISCUSSION

Factor VII deficiency is a rare autosomal recessive bleeding disorder usually detected in the homozygous state. It may present with mild to severe bleeding manifestations like hemarthroses, muco-cutaneous bleeds and intra cranial haemorrhages.

Factor VII levels in the blood can be measured with a factor VII assay test. Normal factor VII levels are between 50% and 200%.

## LEARNING POINTS/TAKE HOME MESSAGES

All cases of bleeding in the neonatal age warrants for a detailed investigation for disorders like Factor deficiency along with work up for hemorrhagic diseases of newborn

## REFERENCES

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