

*Picture Story*

## Neonatal purpura fulminans caused by protein C deficiency

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Protein C (PC) is a vitamin K-dependent serine protease anticoagulant that plays an essential role in regulating coagulation by degrading activated factors V and VIII in plasma<sup>1</sup>. Homozygosity for PC mutations, producing an absolute deficiency of PC anticoagulant activity is found in only 1/250,000 newborns. The complete lack of plasma PC activity results in neonatal purpura fulminans, characterized by sudden onset of widespread purpuric lesions progressing to gangrenous necrosis, accompanied by disseminated intravascular coagulation (DIC) and shock<sup>1</sup>. In Sri Lanka, neonatal purpura fulminans was first documented in 2000<sup>2</sup>. Definitive diagnosis is by establishing low or absent PC activity in the newborn and heterozygous levels in the parents<sup>1</sup>.

A male newborn, first born to consanguineous parents, was delivered by emergency caesarean section due to meconium staining of liquor and fetal distress. Birth weight was 3130g. Apgar score was normal. Vitamin K 1mg was given intramuscularly (IM) soon after birth. At 2 hours of age he was noted to have purpuric patches on both cheeks.

He was admitted to special care baby unit. No features of sepsis were present. By day two, the purpuric patches on cheeks had become deep blue with new ecchymoses occurring at the perineum, buttocks and left foot.

There was moderate thrombocytopenia with prolonged prothrombin time and activated partial

thromboplastin time. The haemoglobin, white cell count and differential counts were normal. Vitamin K was repeated and fresh frozen plasma (FFP) was transfused. Septic screen was negative. Ultrasound scan of brain was normal. Daily FFP transfusion was continued with intravenous crystalline penicillin and cefotaxime.

Serum d-dimer level was over 4.8mg/l (normal <0.2mg/l). A diagnosis of neonatal purpura fulminans was made. This was supported by detecting heterozygous levels of PC activity (43% and 28%) in mother and father respectively (normal 70-130%).

The baby was started on warfarin on day 18 after birth. Since warfarin can suppress any remaining PC activity, during initial stages daily FFP transfusion was also given.

When 20 days old, the baby was noted to have a sluggish pupillary response to light in the right eye. Ophthalmological examination revealed a vitreous haemorrhage.

During the following two months baby's condition stabilized with healing and scar tissue formation of necrotic sites. Warfarin dose was adjusted with INR monitoring. Repeat ophthalmoscopy revealed that the haemorrhage had cleared by three months of age. On discharge, when 93 days old he was on warfarin 1.4 mg/day. All aspects of development were normal.

Presently the baby is 10 months old on warfarin 2mg/day and clinically stable with no bleeding, good weight gain and normal development. Present concerns include rehabilitation of the auto amputated left sided third finger and fibrotic scarring on face.

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**Figure 1: four hours after birth**



**Figure 2: two days old**



**Figure 3: seven days old**



**Figure 4: seven days old**



**Figure 5: eight months old**



**Figure 5: eight months old**

## References

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