To the Editors:

Neonatal purpura fulminans due to protein C deficiency

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A 5-day old baby boy was admitted to the Lady Ridgeway Hospital in July 1998 with multiple gangrenous lesions on both feet, legs, thighs and buttocks, that were first noticed by the mother 2 days before admission. The baby was sucking well from the breast, active and afebrile. The antenatal, natal and immediate postnatal histories were normal. There was marital consanguinity.

The baby was given phototherapy and repeated transfusions of fresh frozen plasma, but continued to deteriorate, and died after 2 weeks.

Purpura fulminans describes the acute development of haemorrhagic necrosis of the skin accompanied by thrombosis of the cutaneous vasculature (1). Although initially identified as a rare post-infectious sequel in children, purpura fulminans in neonates can have a familial tendency and may occur without antecedent infection (2). Protein C is a vitamin K-dependent glycoprotein circulating in plasma as an inactive zymogen. It is converted to the serine protease-activated protein C, a natural anticoagulant that inactivates factors V and VIII, after they are activated by thrombin during the coagulation process. The homozygous form of congenital protein C deficiency can result in life-threatening neonatal thrombosis and purpura fulminans, diseases that are frequently associated with disseminated intravascular coagulation (3). In this baby the protein C level was low and there was marital consanguinity but no family history of the disease. Highly

The lesions spread over the next few days to involve the scrotum, abdomen and upper limbs, and the baby became icteric. The indirect serum bilirubin level was 280 $\mu\text{mol/l}$. The haemoglobin level was 9 g/dl. The white blood count, platelet count and SGPT were normal, but the prothrombin time was prolonged. The protein C level was 0.37 mg/l (normal 1.77 to 3.11).

purified protein C concentrates have been used successfully in the treatment of neonatal purpura fulminans (4). Unfortunately, protein C concentrates are not available in Sri Lanka.

References

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G N Lucas, Paediatrician, and **T G Y R Gunapala**, Registrar, Lady Ridgeway Hospital, Colombo.