

Purpura fulminans

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This is a relatively rare condition in which intravascular thrombosis produces rapid skin necrosis and haemorrhagic infarction, which progresses rapidly to septic shock and disseminated intravascular coagulation. Usually seen in children, it can occur in adults and may be subdivided into three types based on the aetiological mechanism: 'acute infectious', 'neonatal' and 'idiopathic' purpura fulminans. - Acute infectious is the commonest form. It is associated with multiorgan with a mortality rate of 40–50%, usually from failure, and is secondary to either an acute bacterial (*Neisseria meningitidis*) or viral infection (varicella). It is most common in children under 7 years, following an upper respiratory tract infection or in asplenia. Endotoxins produce an imbalance in procoagulant and anticoagulant endothelial activity, producing protein C deficiency; this gives the clinical picture of an initial petechial rash developing into confluent ecchymoses and haemorrhagic bullae, which necrose to form well-demarcated lesions that form hard eschars. Extensive tissue loss is common, which often culminates in limb amputation (Figure 45.10

Figure 45.9 Necrotising fasciitis affecting the left orbit and facial skin (courtesy of St John's Institute for Dermatology, London, UK).

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