

# WOUNDS Congenital

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Cutis aplasia congenita This is a rare condition characterised by the congenital absence of epidermis, dermis and, in some cases, subcutaneous tissues, with underlying bony defects in 20%. Treatment depends on the severity of the presentation, but usually involves plastic surgery .

Parry-Romberg disease (linear morphoea) A rare variant of scleroderma affecting up to 3:100 /uni00A0 000 children. It is linked to certain HLA subtypes and to a family history of autoimmune disease. It is three times more common in females and often develops after an external physical trigger, such as friction or trauma. Moritz Kaposi , 1837–1902, Austrian dermatologist, described xeroderma pigmentosum in 1874. He also described a rare cutaneous sarcoma in Ashkenazi Jews, now more often an AIDS-defining condition. Caleb Hillier Parry , 1755–1822, physician, The General Hospital, Bath, UK. Moritz Heinrich Romberg , 1795–1873, neurologist, Director of the University Hospital, Berlin, Germany . Parry-Romberg disease is a progressive, hemifacial atrophy of skin, soft tissue and bone. The disease commonly starts in the late twenties but can present in childhood, when the resulting deformity is worse because it is magnified by differential growth elsewhere. The condition is self-limiting, usually by 5–10 years after onset. Once stable, plastic surgery techniques can be employed alone or in combination to reconstruct an aesthetic contour. Coup de sabre is a variant that affects the cranium and scalp and resembles a 'blow from a sabre'. Spina bifida Failure of closure of the caudal neuropore during the fourth week in utero results in incomplete development of some or all of the structural elements posterior to the spinal cord. This can occur anywhere, but is commonest in lumbar vertebrae and presents as gross variants: spina bifida occulta, in which there is a bony defect without neural protrusion, and spina bifida cystica, in which there is herniation of the meninges (meningocele), spinal cord (myelocele) or, most commonly ,

Figure 45.49 Angiosarcoma (courtesy of St John's Institute for Dermatology, London, UK). Figure 45.50 Kaposi's sarcoma (courtesy of St John's Institute for Dermatology, London, UK).

multidisciplinary approach and is directed towards protecting the spinal cord and preventing cerebrospinal fluid contamination, secondary hydrocephalus and meningitis. WOUNDS Congenital

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