

fontanelle and mineralization of sutures. Wormian bones are present. The mandible is prognathic as a result of hypoplasia of the maxilla and other facial bones. Skeletal defects in the hands and pelvic bones have also been described. The permanent teeth erupt late and are often abnormal with numerous supernumerary teeth. The patients may also have conductive deafness. The

course is usually uncomplicated except for dislocations, especially of the shoulders, and dental anomalies that require therapy.

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Necrotizing Fasciitis

A 6-year-old, male child was admitted with history of minor penetrating injury with nail on anteromedial aspect of left thigh 7 days prior to admission. Three days later, he developed fever, discoloration of skin and swelling of left thigh. On examination child was toxic, drowsy, febrile with poor respiratory effort and shock. Local examination of left thigh revealed 8×10 cm necrotic area with blue-black discoloration with marked swelling of thigh and leg with no palpable crepitus (*Fig. 1*). Investigation revealed multi-organ dysfunction. A diagnosis of necrotizing fasciitis was made. The child was provided intensive treatment with inotropes, antibiotics and mechanical ventilation and extensive surgical debridement. Child deteriorated over next 5 days and expired.

Necrotizing fasciitis is characterized by involvement of soft tissue of deep dermis, adipose tissue and subcutaneous fascia. Hallmark of the disease is extensive areas of both necrosis and cellulitis. Portal of entry of organisms is usually through a cut or surgical wound. Common organisms implicated are β -hemolytic group A streptococci, *Pseudomonas* and anaerobes. The disease has a varied



Fig. 1. Necrotizing fasciitis of left thigh.

spectrum ranging from simple cellulitis to septic shock and multiorgan dysfunction. Over 50% mortality has been reported. Management of necrotizing fasciitis includes early surgical debridement. Antibiotics alone do not improve outcome. Amputation can be done in difficult cases. Role of hyperbaric oxygen is doubtful.

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