A 9-year-old boy was evaluated for a malformation of his right forearm present from birth (Figure 1). Type 1 neurofibromatosis was confirmed by genetic testing. The mother and maternal grandmother were also affected by neurofibromatosis with only skin involvement. Initially, the forearm malformation was attributed to hyperlaxity but a radiograph showed evidence of dysmorphism of the ulna, with bone deficiency along the stem (Figure 2).

In patients with congenital nonunion of long bones, 50%-80% have an underlying diagnosis of type 1 neurofibromatosis. The prevalence of nonunion in patients with type 1 neurofibromatosis is 5%.1 The formation of pseudarthrosis is a result of the nonunion between bone fragments of a long bone fracture and the formation of a false joint; the site most commonly affected is the tibia. This mechanism can occur in type 1 neurofibromatosis following bone dysplasia that can cause alterations in the repair processes.2 Pseudarthrosis of the forearm is rare and usually involves the ulna. Radiologic findings may include cystic lesions, bone deformity, and cortical thinning; anecdotal cases of osteolytic reaction are described. Surgical treatment is recommended in the event of a fracture to prevent recurrence and includes bone fixation and/or debridement of the nonunion fibrous tissue between the bone segments.3

Alessandro Agostino Occhipinti, MD
Institute for Maternal and Child Health IRCCS Burlo Garofolo
Trieste, TS, Italy

Prisca Da Lozzo, MD
Elena Favaretto, MD
Università degli Studi di Trieste
Dipartimento di Scienze Mediche Chirurgiche e della Salute
Trieste, TS, Italy

Andrea Magnolato, MD
Irene Bruno, MD
Egidio Barbi, MD
Institute for Maternal and Child Health IRCCS Burlo Garofolo
Trieste, TS, Italy

Figure 1. Right forearm with evidence of malformation and varus deviation of the right upper limb.

Figure 2. Dysmorphism of the ulna, with bone deficiency along the stem, fragmented in the middle third shaft with loss of distal trophism.

The authors declare no conflicts of interest.
A 1-year-old boy was transferred to our hospital due to severe septic shock that led to cardiac arrest. The patient reportedly had fluid-refractory septic shock that required catecholamine administration. On admission, a characteristic skin rash covered the patient’s entire body (Figure 1). His right buttock and thigh were reddish-purple and swollen (Figure 2). An incision was made in the right thigh, and the patient was diagnosed with necrotizing fasciitis. Surgical debridement was performed within the first 5 hours following his arrival. Wound and blood cultures revealed a *Pseudomonas aeruginosa* infection. The patient underwent continuous renal-replacement therapy with an endotoxin adsorption filter, empiric antimicrobial therapy (meropenem and ciprofloxacin), and necrotic tissue excision. Based on the drug susceptibility test results, the patient’s treatment was de-escalated to piperacillin monotherapy. On the 31st hospitalization day, the infant died of intra-abdominal hemorrhage due to liver failure-associated coagulopathy. Rapid screening via flow cytometric analysis of monocytic intracellular tumor necrosis factor-α production in response to lipopolysaccharide suggested a Toll-like receptor signaling pathway deficiency. The postmortem sequence analysis of the *IRAK4* gene revealed a homozygous frameshift mutation (c.167_172insA).

The skin rash covering the entire body was ecthyma gangrenosum (Figure 1), caused by *P aeruginosa* bacteremia. Ecthyma gangrenosum is characterized by a central necrotic ulcer, sharp edges, and an erythematous halo. First described in 1897, ecthyma gangrenosum is a distinct

---

**Figure 1.** A, Ecthyma gangrenosum in the back, measuring 10 mm in diameter. B, Ecthyma gangrenosum in the left foot.