A 29-year-old man was born with radial clubhands that required several operations during his early weeks of life. In addition to shortened forearms, the patient was left with severe limitation of wrist motion bilaterally.

Robert P. Blereau, MD of Morgan City, La, noted normal proximal upper extremities. Both forearms were 5.5 in long. The patient had full range of motion of both elbows. He held the right wrist at 20 degrees ulnar deviation and was unable to move it medially or laterally. The right wrist was fixed in a neutral position and could not be flexed or extended. The left wrist, which was held at 45 degrees radial deviation, could be flexed 55 to 90 degrees but had no other motion. Both hands demonstrated equally good strength, dexterity, and grasping ability. The patient had no other associated physical deformities and no family history of limb or other congenital anomalies. His platelet count at birth was unknown.

Roentgenograms revealed short ray deformities of the forearms, with absence of the radius bilaterally. Severe degeneration of the carpal joints and articulation of the distal ulna with the wrist bilaterally also were seen. The humeri at the elbows appeared fairly unremarkable. Congenital absence of the radius can be unilateral or bilateral. The condition is bilateral in thrombocytopenia with absent radii syndrome. A missing radius causes the hand to deviate radially at birth. The thumb may or may not be present; other fingers are usually normal. Associated knee instability may be present.

Radial clubhand is estimated to occur in 1 in 55,000 births, with a male to female ratio of 3:2. The condition is bilateral in 50% of patients. Manifestations can vary from slight hypoplasia of the thumb to complete absence of the radius. Initial treatment is the application of splints to prevent further radial deviation and to help correct the deformity. The goal of surgical correction is to centralize the hand over the ulna. A low platelet count may delay surgical intervention for months to years. Radial ray defects can be associated with other congenital diseases and defects. Among these are Fanconi anemia, Holt-Oram syndrome (heart-hand syndrome), VATER (vertebral defects, imperforate anus, tracheoesophageal fistula, and radial and renal dysplasia) syndrome, Cornelia de Lange syndrome, and trisomy 13 and trisomy 18 syndromes.

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