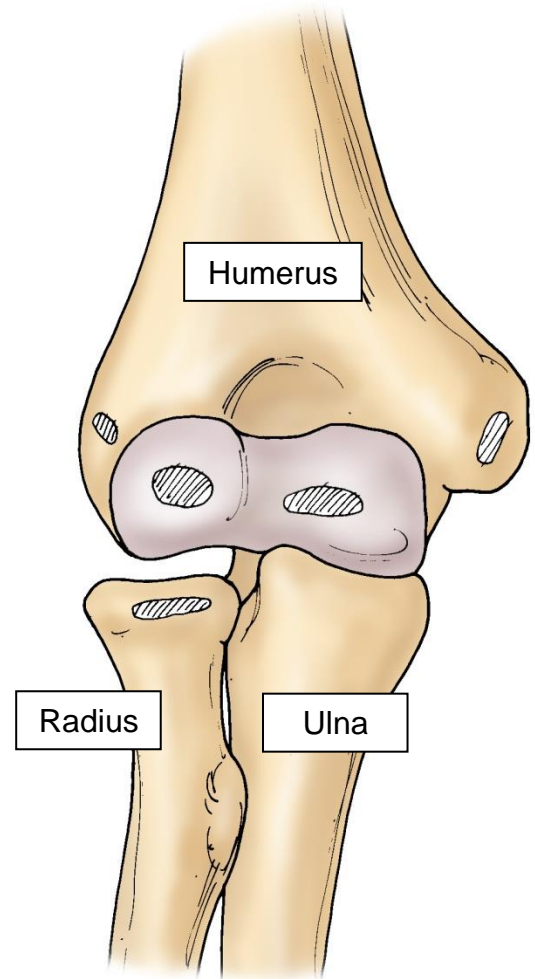


CONGENITAL RADIOULNAR SYNOSTOSIS

The elbow joint is formed by the end of the humerus and the radius and the ulna. These three bones together allow for the arm to be flexed to one's mouth and extended fully, as well as contribute to the ability to place the hand in a palm down appearance and in a palm up appearance. In the first six weeks of life, the embryo creates a projection, which is called the limb bud. The limb bud will separate and form these three bones called the humerus, the radius and the ulna. Very early during this process certain hormonal or timing factors may not signal that the radius and the ulna should separate into two separate and discrete bones.

During this period of intrauterine development this congenital abnormality will be an isolated event.

Approximately boys will be affected by girls in a ratio of 3:2 and 80% of the time of the right and the left arm will be affected. This disorder is associated with certain genetic disorders such as acroplasyndactyly called Carpenter's syndrome, acrocephalosyndactyly called Apert syndrome, arthrogryposis and Klinefelter syndrome.

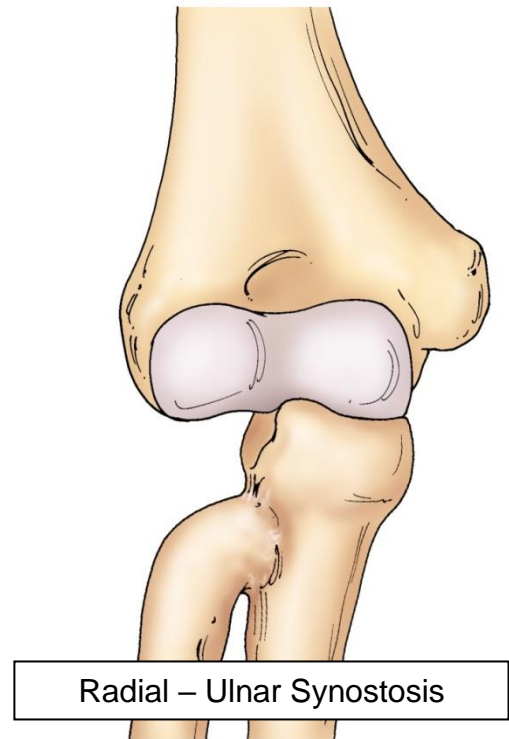


Only a small number of these children are identified at birth. Most children are identified between 18-months and 3-years-of-age when certain activities are identified by the parent or schoolteacher as being awkward compared to their peers. Functional limitations that may prompt the child to be evaluated by a pediatric orthopedist may include difficulty holding small objects such as pencils, crayons or spoons, difficulty closing and opening shirt buttons, coat buttons or belts, the inability to rotate a baseball glove in a position to catch a ball and occasionally with lack of turning the hand in a palm up position to accept certain objects. On closer examination in addition to the complete lack of flexibility or motion into a palm up or palm

down motion there is oftentimes a lack of complete straightening of the elbow, as well as a lack of complete flexion of the elbow. In assessing individuals the position of the hand relative to the elbow is important. The x-rays will demonstrate one of several patterns. Most of them can be demonstrated that the radius and the ulna are fused together and that the radial head does not have a normal appearance or is completely missing due to the fusing of the radius and the ulna together.

TREATMENT OPTIONS

Treatment options are discussed if there are a lack of school tests or recreational tests that are particularly difficult to accomplish. There have not been surgical techniques developed that would restore complete palm up/palm down motion especially if the upper part of the radial bone is absent or completely missing. For a small number of children the hand is stuck in a complete palm down or complete palm up position. If certain activities of daily living are compromised by this extreme position then a recommendation to cut the two bones in the arm and rotate the hand in a more functional position will be discussed. But again, even with this surgical technique the forearm and hand will not be able to rotate.



The natural history still suggests that most children tolerate this birth defect into adulthood without pain or significant functional disability and counseling is required for the family to understand that any operative intervention is unlikely to improve their range-of-motion but only better position the affected hand as a helper hand or an assist hand.