



Orthopedic Care Center Hand and Orthopedic Upper Extremity Program Ulnar Club Hand

Whether your child or loved one suffers a broken arm, a sports-related injury or the most complex spine condition, The **Orthopedic Care Center** at Children's Hospital Boston is committed to providing comprehensive and compassionate care. Established in 1903, we are among the world's most experienced pediatric orthopedic programs, treating a high volume of some of the most complex orthopedic conditions. And with 10 specialty clinics, we are the largest in the country. We are also one of the busiest. Each year, our staff attends to about 80,000 patient visits and conducts about 5,000 surgeries. In 2010, we were ranked #1 in pediatric orthopedics by *U.S. News & World Report*.

The **Hand and Orthopedic Upper Extremity Program** provides comprehensive care for infants, children and adolescents with a wide range of complex upper limb conditions. Multidisciplinary care involving occupational and physical therapy, splinting, casting and reconstructive surgeries is provided for congenital, neuromuscular, sports-related oncologic, traumatic or post-traumatic conditions.

What is Ulnar Club Hand?

Ulnar club hand refers to a congenital difference in which one of the long bones of the forearm, the ulna, and other soft tissue structures on the ulnar side of the hand develop abnormally. The improperly formed ulna may cause the wrist to deviate toward the little finger. This may cause the radius to bow. There may be deformities in the fingers and thumbs as well. In most forms of ulnar club hand - which can present itself in a wide range of severity - not only is the bone abnormal but the soft tissues and flesh of the forearm are also abnormal. The arrangement of muscles and nerves may be unbalanced and some muscles and nerves may even be absent. The condition is sometimes called "ulnar dysplasia" which simply means abnormal development of the ulna. As with the majority of birth defects, scientists do not know why ulnar club hand occurs, but information that is known about the condition indicates that it does not result from the mother's lifestyle or anything the mother did during her pregnancy.

What causes ulnar club hand?

In most cases, the cause of ulnar club hand is unknown. There have been reports of ulnar club hand occurring in families, particularly in associated inherited syndromes. These syndromes include ulnar mammary syndrome and Klippel-Feil Syndrome. It is also associated with syndromes without a familial inheritance pattern such as Cornelia de Lange syndrome. Half of ulnar club hand cases occur along with other malformations in the musculoskeletal system.

How common is ulnar club hand?

Ulnar club hand is much less common than a similar condition known as radial club hand, occurring in 1 in 100,000 live births.

How is ulnar club hand diagnosed?

Ulnar club hand is diagnosed after a thorough history and physical examination. X-rays will often aid in the diagnosis. Your child's doctor will want to check for other congenital deformities or syndromes that are associated with this condition. Further tests may depend on whether the doctor suspects any of these associated problems. In diagnosing the condition, the doctor will want to note the severity and will likely classify it as one of the following four types:

Type 1: This is the mildest form of ulnar club hand. Although the ulna is underdeveloped, both ends of the bone (epiphyses) are present. The growth plates at both the distal (hand side) and proximal (elbow side) are in tact, but growth is slowed. Deviation of the wrist and bowing of the radius is minimal. Sometimes there are other malformations of the hand.

Type 2: This is the most common form of ulnar club hand. It involves a partial absence of the ulna and the hand appears deviated toward the ulnar side. There is an abnormal bar of fibrous tissue, known as anlage, extending from the distal (wrist side) end of the partial ulna to the wrist. Bowing of the radius may also occur. The presence of the ulna in Type 2 cases is usually sufficient enough that the elbow is functional and stable. There may be some degree of deformity in the fingers or thumb.

Type 3: This type involves complete absence of the ulna. The elbow is unstable. Hand and wrist malformations are common.

Type 4: This type involves an abnormal connection between the humerus and radius (synostosis). There is an ulnar anlage present from the distal (hand side) humerus to the wrist, with considerable bowing of the radius and ulnar deviation of the hand. Hand anomalies are common.

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Treatment

Specific treatment for ulnar club hand will vary from child to child and will be determined by your child's physician based on:

- your child's age, overall health, and medical history
- the severity of the condition
- any other deformities or syndromes associated with the problem
- your child's tolerance for specific medications, procedures, or therapies
- your opinion or preference

Exercises and splinting

During infancy, the first goal of treatment is to achieve passive extension of the wrist and elbow into a normal position. Your child's doctor will guide you in performing gentle but firm and frequently repeated passive stretching exercises for the wrist and elbow to help accomplish this goal. Corrective casting and splinting may also be used.

In more severe cases, surgery is necessary. If your child's doctor recommends surgery, keep in mind that range of motion exercises are still extremely important. Even if wrist therapy alone doesn't resolve problems with the wrist, you should not be discouraged from continuing the exercise regimen. Remember that any improvement in range of motion achieved through exercise will make subsequent surgery more effective and perhaps less complex.

Surgery

Operations involved with correcting ulnar club hand include:

- Surgical excision of anlage to reduce or eliminate deviation of the hand.
- A procedure to correct the bowing radius called osteotomy, in which a surgeon cuts the radius into wedges and refits them into a straight position.
- Osteotomy of the humerus (upper arm bone) to better position the wrist and hand
- Repair of digital and thumb deformities.

What is the long-term outlook for a child with ulnar club hand?

Most children with ulnar club hand will have some shortening of the forearm when fully grown. There may be some limitation of motion, function, and strength, but overall the limb and hand is very functional.



X-ray of an ulnar club hand.

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