



Orthopedic Care Center Hand and Orthopedic Upper Extremity Program Radial 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 radial club hand?

The normal hand connects with both bones of the forearm, the radius and the ulna, at the wrist. In radial club hand, a baby's radius was not formed properly in the womb, which in turn causes the wrist to be in a fixed bent position toward the thumb side of the hand. There may be a deformity or absence of the thumb as well. The condition is sometimes called "radial dysplasia" which simply means abnormal development of the radius. In most forms of radial club hand, 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 most severe cases lead to significant problems in the function of the hand, fingers and elbow. The entire arm will be shorter, with marked curving of the forearm, stiffness of the elbow and fingers. In these situations, the thumb will either be very small or absent.

Radial club hand is a birth defect. As with the majority of birth defects, scientists do not know why it occurs, but information that is known about radial club hand indicates that it does not result from the mother's lifestyle or anything the mother did during her pregnancy.

Radial club hand develops early in pregnancy, sometime between the 28th and 56th day of gestation, when the bones of the hand and forearm are being formed. It is sometimes, but not always, picked up on a prenatal ultrasound. Even if it is detected prenatally, the condition cannot be treated until after the baby is born.

The hand is comprised of many small bones called carpals, (at the base of the hand) metacarpals (in the palm area) and phalanges (in the fingers) The two bones of the forearm - the radius and the ulna - meet at the hand to form the wrist. The radius provides more of the support for the hand than does the ulna.

When describing a particular side of the arm, you may hear your child's doctor refer to the "radial side" which indicates the side of the arm on

which the thumb lies, or the "ulnar side" which describes the side on which the little finger lies. These terms are used rather than "inner" or "outer" arm because the palm of the hand can face either forward or backward. As the hand rotates, the radius swings around the ulna, which is hinged to the humerus (upper arm bone) at the elbow joint.



X-ray of a radial club hand.

How much does radial club hand interfere with function?

There are varying degrees of malformation of the radius and therefore varying degrees on how much a child's radial club hand will affect function. In the most severe cases, the radius is completely absent, leading to very limited range of motion at the wrist. The elbow joint may also be disturbed or even fused with no motion. There may also be underdevelopment or absence of the thumb, which interferes with hand function.

continued on page 2.

Clinical Team

Peter M. Waters, MD
John E. Hall Professor of Orthopedic Surgery, Harvard Medical School
Clinical Chief, Orthopedic Surgery, Children's Hospital Boston

Donald S. Bae, MD
Assistant Professor of Orthopedic Surgery, Harvard Medical School

Apurva S. Shah, MD
Clinical Fellow in Orthopedic Surgery

Locations

Children's Hospital Boston
300 Longwood Avenue
Fegan 2
Boston, MA 02115

Children's Hospital Boston
at Waltham
9 Hope Avenue
Waltham, MA 02453

Children's Hospital Boston
at Lexington
482 Bedford Street
Lexington, MA 02420

Boston Children's North
10 Centennial Drive
Peabody, MA 01960

In the mildest cases, the radius is merely slightly smaller than the ulna and there is minimal deviation at the wrist.

The severity of radial club hand is also affected by the presence of an abnormal bar of fibrous tissue that may appear in any case where the distal part (hand end) of the radius is absent. This fibrous tissue which connects the remnants of the radius to the hand, is known by the German word anlage and has a very limited ability to grow. It is attached to the radial side of the hand and wrist. As the ulna grows in the mother's womb, the lack of growth in the radial anlage draws the hand into a deviated "club" position. This may also cause the ulna to bow.

The degree to which the hand and wrist deviate at birth, depends on two factors: the degree to which the support of the radius is lacking and the degree to which the anlage tethers the wrist, allowing the ulna to push the wrist further and further into deviation as it grows.

What causes radial club hand and who is affected?

Radial club hand occurs sporadically with no known cause. It occurs between 1 in 30,000 and 1 in 100,000 live births. Several theories have been raised, such as maternal drug exposure, compression of the uterus, and vascular injury, but none of these have been proven. There is no known genetic link to radial club hand except when the problem is associated with other congenital anomalies or syndromes that do have a known genetic component. It has been associated with many congenital syndromes including those affecting cardiac, gastrointestinal and renal systems. It has also been linked to some chromosomal abnormalities including Trisomy 13, 18 and 21.

How is radial club hand diagnosed?

Radial club hand is diagnosed after inspection of the forearm and an X-ray. Your child's doctor will want to check for other congenital deformities or syndromes that are associated with radial club hand. Further tests may depend on whether the doctor suspects any of these associated problems, including concern regarding the heart, kidneys, vertebral column, blood cells, and digestive system. 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 radial club hand, involving mild deviation of the wrist throughout life. Problems that can result from more severe forms, such as loss of motion, usually do not occur. Type 1 involves a defective growth plate (physis) on the distal end (hand side) of the radius. This defect leads to a minor foreshortening of the radius and a prominent distal ulna. Surgical intervention is usually not required. However, some surgery may be required to correct underdevelopment (hypoplasia) of the thumb, an associated problem that sometimes occurs in Type 1 cases.

Type 2: This involves limited growth of the radius on both its distal and proximal sides. The wrist is therefore more deviated toward the radius,

and the ulna bows out. Underdevelopment of the thumb is usually more significant with more deficiency in the bones of the wrist.

Type 3: This involves the absence of two thirds of the radius on the hand side. The wrist is more severely deviated and the hand has limited mechanical support. The ulna is thickened and bowed. Associated problems with the thumb and fingers, such as underdevelopment or camptodactyly, a deformity in the finger joints that leads to a flexed finger or fingers.

Type 4: This type of radial club hand is most common and the most severe, causing profound limitation of hand, wrist and forearm function. It involves the complete absence of the radius along with complete or near complete absence of the thumb. The ulna bowing is also most severe. The index, long and ring fingers may be involved and the elbow may have limited range of motion.

How is radial club hand treated?

Treatment for radial club hand can be very complex. Specific treatment 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.

More severe cases may require casting or splinting to gradually stretch the contracted soft tissues. Still, a rigorous stretching program is an important component of any treatment plan.

Once passive motion is achieved, your baby will likely need to wear a splint during the night throughout infancy and during periods of rapid growth.

More severe cases, in which there is no support for the wrist without the splint, usually require surgery. However, even if your child needs surgery, keep in mind that the range of motion exercises are extremely important. Even if wrist therapy alone doesn't resolve the wrist contracture (shortening) and deviation, this should not discourage you from continuing the exercise regimen. Any improvement in range of motion achieved through exercise will make subsequent surgery more effective and perhaps less complex.

continued on page 3.

Clinical Team

Peter M. Waters, MD
John E. Hall Professor of Orthopedic Surgery, Harvard Medical School
Clinical Chief, Orthopedic Surgery, Children's Hospital Boston

Donald S. Bae, MD
Assistant Professor of Orthopedic Surgery, Harvard Medical School

Apurva S. Shah, MD
Clinical Fellow in Orthopedic Surgery

Locations

Children's Hospital Boston
300 Longwood Avenue
Fegan 2
Boston, MA 02115

Children's Hospital Boston
at Waltham
9 Hope Avenue
Waltham, MA 02453

Children's Hospital Boston
at Lexington
482 Bedford Street
Lexington, MA 02420

Boston Children's North
10 Centennial Drive
Peabody, MA 01960

Surgical treatment

Surgical treatment is usually required to correct deviation of the wrist that persists despite non-surgical means of treatment. It is also an option when the thumb limits function of the hand. However, the decision to perform surgery is weighed against any other problems that the child may be experiencing due to associated medical conditions. Also, sometimes if the child cannot flex the elbow, the presence of a bent wrist and short forearm can be an advantage, enabling a child to, for example, feed himself. Surgery is divided into phases. Each problem is treated in separate surgical procedures that may take place over the course of months or years, depending on the problem. The following is a general description of how the different parts of the arm and hand affected by radial club hand are surgically treated:

The wrist

Any of the following procedures may be used to offset the wrist deviation and contracture:

Lengthening: This procedure is used prior to surgery to gently stretch the hand into a straighter position. A customized frame is surgically placed on the hand and forearm so that pins go through the ulna and hand. The pins in the ulna and hand are then pushed apart gradually, (about 1mm per day). Lengthening can be recommended in infancy in situations where it is very difficult to stretch out the tight wrist. It is also used in adolescence when there is recurrent deformity or marked shortening of the forearm. **Centralization:** This procedure involves removing bones from the wrist so that the hand sits straight on the end of the ulna, which is sometimes placed in a slot within the wrist.

Radialization: This procedure involves moving the hand slightly further across towards the ulnar border of the forearm and reorganizing the tight muscles of the wrist so that the hand is balanced on the end of the ulna. The benefit of this procedure is that it is less likely to interfere with growth because less bone is resected and there is usually more mobility in the wrist because the muscles are restored and the ulna is not placed in a slot within the wrist.

Splinting: This is usually necessary following centralization or radialization procedures. At the time of surgery, surgeons will likely create an internal splint -- a pin that is placed through the wrist and the ulna. This may remain in place for a year or even more after surgery, until the ulna gradually broadens and becomes a more stable platform on which the wrist may balance.

The forearm bones

Osteotomy: Often, radial club hand leads to bowing of the ulna bone. This bowing can become extreme and may even result in a twist in the forearm because of the complex arrangement of the anlage. At the time of the operation for the wrist, your child's surgeon may want to straighten the forearm bones by cutting them into wedges, then rearranging them

into a straighter position. These will be held in place by the same pin that holds the wrist.

Underdeveloped or absent thumb

Problems with the thumb are extremely common in radial club hand, particularly in more severe cases. In many cases, surgery to reconstruct the thumb is recommended. Reconstructive surgery, either through a procedure known as pollicization or by tendon transfers, is usually performed after other procedures described above are completed. For a description of these thumb problems and surgery to correct them, see link below. Common congenital hand conditions and treatments

The elbow

Surgery on the elbow is very uncommon in radial club hand. However, in some cases, your child's doctor may want to perform an operation to release the tight soft tissue structures at the back of the elbow that are causing the elbow to stiffen. This may help your child's arm attain passive range of motion.

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

The long-term outlook is dependent on the severity of the deformity. In mild cases, intermittent therapy throughout growth is required to maintain alignment and strength. These children will have minor limitations of motion, function, and strength. In the more severe cases, there will be marked limitation of motion, strength, and function. In these situations, growth will also be limited in that forearm.

Clinical Team

Peter M. Waters, MD
John E. Hall Professor of Orthopedic Surgery, Harvard Medical School
Clinical Chief, Orthopedic Surgery, Children's Hospital Boston

Donald S. Bae, MD
Assistant Professor of Orthopedic Surgery, Harvard Medical School

Apurva S. Shah, MD
Clinical Fellow in Orthopedic Surgery

Locations

Children's Hospital Boston
300 Longwood Avenue
Fegan 2
Boston, MA 02115

Children's Hospital Boston
at Waltham
9 Hope Avenue
Waltham, MA 02453

Children's Hospital Boston
at Lexington
482 Bedford Street
Lexington, MA 02420

Boston Children's North
10 Centennial Drive
Peabody, MA 01960