EDUCATION AND TAKE-HOME INSTRUCTION

Radial Longitudinal Deficiency (Radial Club Hand)

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What is radial longitudinal deficiency?

Radial longitudinal deficiency, previously known as radial club hand, is a non-painful, rare, congenital (meaning your baby was born with it) difference in which the radius bone of the forearm does not form properly.

The radius bone is the lateral (on side of thumb) bone in the forearm. The condition causes the wrist to be in a bent position toward the thumb-side of the forearm. It is also called radial dysplasia or radius deficiency.

What problems does it cause?

Radial longitudinal deficiency affects more than the bone. It also affects the soft tissues and flesh of the forearm. The arrangement of muscles and nerves may be unbalanced and some muscles and nerves may be missing. If your child has a mild form, it generally does not cause many problems with your child's development or hand movement.

If your child has a severe form, they may have significant problems moving their hand, fingers and elbow. Their entire arm will be shorter, with a curved forearm and stiff elbow and fingers. Your child may also have a missing or small thumb.

What causes radial longitudinal deficiency?

It usually occurs by chance. Doctors and scientists do not know why it affects certain children. Several theories have been raised, including compression of the uterus and blood vessel injury. But none of these have been proven. Experts do not think that it happens because of the mother's lifestyle or anything the mother did during pregnancy.

Radial longitudinal deficiency is associated with several congenital syndromes, including those affecting the heart, digestive system and kidneys. It has also been linked to some chromosomal abnormalities, including Trisomy 13, 18 and 21.

How is radial longitudinal deficiency diagnosed?

It is sometimes picked up on a prenatal ultrasound but cannot be treated until after the baby is born.

If your child's condition is seen after birth, a doctor will examine their forearm and take an X-ray. Your child's doctor will check for other differences or syndromes that are associated with radius deficiency. Additional tests could include blood tests and ultrasounds of the heart and/or kidneys.

Radial longitudinal deficiency is classified into the following 4 types:

Type 1

This is the mildest form. Your child will be able to move their hand normally. They will only need surgery if it is necessary to correct an underdeveloped thumb.

Type 2

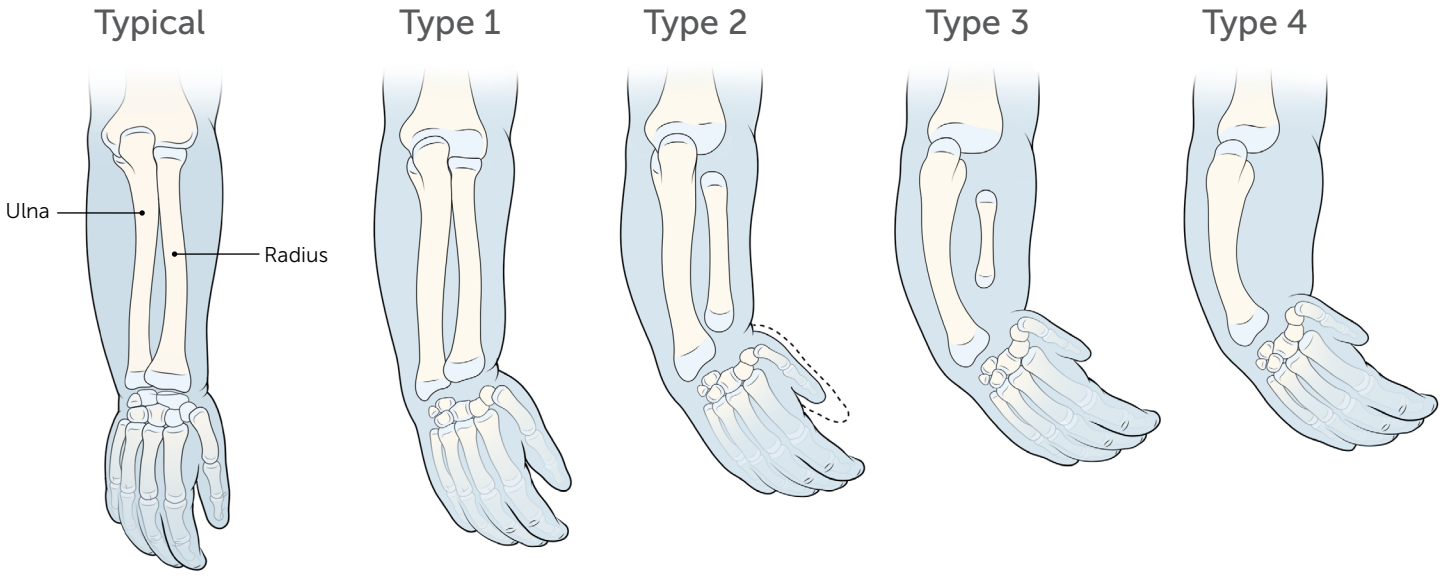
This involves limited growth of the radius on both sides. The wrist is more bent toward the radius, and the ulna bows out. This type is commonly associated with an underdeveloped thumb.

Type 3

This type means that your child is missing most of the radius on the hand side. The wrist is more severely bent and the hand has limited support. The ulna is bowed. Most children with this type have an underdeveloped or missing thumb.

Type 4

This is most common form. It is also the most severe, with ulna bowing that leads to a significant difference in the length of the forearm. Most children with this type have an underdeveloped or missing thumb, and may have differences in the other fingers as well. The elbow may have limited range of motion. Your child may preferentially use the pinky finger for picking up and manipulating objects, called "ulnar prehension."



How is radial longitudinal deficiency treated?

Exercises and splinting

When you child is an infant, the goal of treatment is to allow your child's wrist to extend and their elbow to move into a normal position. Your child's doctor will talk to you about how to perform gentle stretching exercises for their wrist and elbow to help improve your child's range of motion.

If your child's condition is severe, they may wear a cast splint to gradually stretch the tight soft tissues. **Even if your child needs surgery, keep in mind that the range-of-motion exercises are extremely important.** Any improvement in range of motion achieved through exercise will make any surgery needed later on more effective and perhaps less complex.

Your baby will probably need to wear a splint at night throughout infancy and during times when they grow quickly.

Surgery

If your child's case is more severe, they will need surgery after stretching and splinting. Surgery is also an option when problems with the thumb impact your child's ability to use their hand. But doctors must weigh the decision to perform surgery against any other medical conditions your child may have, as well as the overall function of their arms.
Each problem is treated during separate surgeries that may take place over months or years, depending on the problem. Below are some ways we treat different parts of the arm and hand:

Surgery - The wrist

Lengthening This procedure is used before surgery to gently stretch the hand into a straighter position. A frame is surgically placed on the hand and forearm so that pins go through the ulna and hand. The pins are slowly pushed apart.
This may be recommended while your child is an infant if it is hard to stretch their tight wrist. It is also used when a child is older and there is another deformity or significant shortening of the forearm.

Centralization This involves repositioning the wrist so the hand sits straight on the end of the ulna. At the time of surgery, surgeons place a pin through the wrist and the ulna. This may stay in place for a year or more after surgery until the ulna broadens and becomes a more stable platform for the wrist to balance on.

Bilobed flap or soft tissue centralization This involved repositioning the soft tissues (skin and muscles/tendons) to improve the motion of the wrist, without any bony surgery as would be done in a formal centralization.

Splinting This is usually necessary both before and after bony or soft-tissue centralization.

Surgery - The forearm bones

Osteotomy Radial deficiency often leads to bowing of the ulna bone. This can become extreme and may cause the forearm to twist. At the time of the operation for the wrist, the surgeon may straighten the forearm bones by cutting them into wedges and rearranging them into a straighter position. These are held in place by pins which are later removed.

Surgery - Underdeveloped or missing thumb

Many children need surgery to reconstruct their thumb. If the thumb is only mildly underdeveloped, surgery called an opponensplasty can be performed to make the thumb muscles stronger. When the thumb is more significantly underdeveloped or missing, a pollicization may be performed, in which the index finger is repositioned as a thumb.

Surgery - The elbow

Elbow surgery is rarely needed. Some children with difficulty bending the elbow will undergo a procedure to release the tight soft tissue structures at the back of the elbow causing the elbow to stiffen. This can be done separately or in combination with procedures for the wrist and thumb.

What is the long-term outlook?

Your child's long-term outlook depends on the severity of the condition and any associated conditions they have.

If your child has a more severe form, their arm will have limited range of motion, strength and movement.

If your child has a mild case, they will need therapy as they grow in order to keep alignment and strength. They may have small problems moving and using their arm.

Notes

Produced by the Hand and Orthopedic Upper Extremity Program at Boston Children's Hospital.
For more information or to request an appointment visit **BostonChildrens.org/Hand**.

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