

Ulnar Longitudinal Deficiency (Ulnar Club Hand)



What is ulnar longitudinal deficiency?

Ulnar longitudinal deficiency, previously known as ulnar club hand, is a condition in which your child's wrist is in a fixed and bent position toward the side of the hand with the little finger. This condition is also sometimes called ulnar dysplasia. It happens when 1 of the long bones of the forearm, the ulna, and other soft tissues of the hand, develop abnormally in the womb. Your child's fingers and thumb may also be affected. Other muscles and nerves in the hand may be unbalanced or missing.

Half of the time, children also have other malformations in the musculoskeletal system, such as scoliosis. The most severe cases lead to significant problems in the function of the hand, fingers and elbow.

What causes ulnar longitudinal deficiency?

Most of the time, the cause of ulnar longitudinal deficiency is unknown. It may sometimes run in families, particularly as part of inherited syndromes, like ulnar mammary syndrome and Klippel-Feil syndrome. It is also associated with syndromes that are not inherited, like Cornelia de Lange syndrome.

How common is ulnar longitudinal deficiency?

Ulnar longitudinal deficiency affects about 1 in 100,000 babies. It is much less common than failure of the radius to develop, called radial longitudinal deficiency.

How is ulnar longitudinal deficiency diagnosed?

Your child will have a complete physical exam to evaluate the arm difference, but also to check for other orthopaedic differences, such as scoliosis. Your child will likely have an X-ray to evaluate the development of the bones in the forearm and hand. Your child may have other tests, depending on whether the doctor suspects any associated conditions.

Ulnar longitudinal deficiency is classified into the following 4 types:

Type 1

This is the mildest form. The ulna is slightly smaller than normal. Sometimes there are other malformations of the hand. This type will not cause many problems in your child's development.

Type 2

This is the most common form. It involves a partially missing ulna. There is an abnormal bar of fibrous tissue (anlage) extending from the distal (wrist side) end of the ulna to the wrist. The elbow is functional and stable. There may be some differences in the fingers or thumb.

Type 3

This type means there is a completely missing ulna. The elbow is unstable. Hand and wrist malformations are common.

Type 4

This type involves an abnormal connection between the humerus and radius, called a humeroradial synostosis, so that the elbow is fixed in one position. There is considerable bowing of the radius and usually malformation of the hand as well.



