



قَالَ سُبْحَانُكَ لَا إِلَهَ إِلَّا مَا عِلْمُنَا
وَلَا نَعْلَمُكَ إِلَّا مَا عِلْمُنَا

إِنَّمَا أَنْتَ الْعَلِيمُ الْحَكِيمُ

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Management of Radial Club Hand

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Aim of Essay

The aim of this essay is to review the literature as regard the diagnosis and the new advances in the treatment of radial club hand, including: indications , techniques , results and possible complications.

Abbreviations

AER Apical ectodermal ridge

AP Antroposterior

CRHD Congenital radial head dislocation

CBC complete blood count

ECU extensor carpi ulnaris

EME Ectoderm Mesoderm Endoderm

FCU flexor carpi ulnaris

HFA hand–forearm angle

HFP hand–forearm position

HFM Hemifacial Microsomia

MTP metatarsophalangeal

PIP proximal interphalangeal joint

PRUS Proximal radioulnar synostosis

RCH Radial club hand

RLD Radial longitudinal deficiency

TAR Thrombocytopenia–absent radius syndrome

UB the ulnar bow

ULD Ulnar longitudinal deficiency

VATER/VACTERL V = vertebral, A = anal atresia, C = cardiac, TE = tracheoesophageal fistula, R = renal, radial, L = limb

ZPA zone of polarizing activity

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Introduction

A child with a congenital anomaly can cause stress for the parents.⁽¹⁾ Radial club hand is one of the dysplastic manifestation of radial-side deficiency of the upper limb. This defect describes a spectrum of osseous, muscular, cutaneous tissues and nerves tissues dysplasia of the radial border of the upper limb. The dysplastic manifestations of radial deficiency range from the minimum finding of hypoplasia of the thenar muscles or an absent thumb to the more severe anatomical and functional abnormalities of radial club hand.⁽²⁾

The rate of outspread insufficiency has been evaluated to be 1 in 30,000 to 1 in 100,000 births.⁽³⁾ Radial insufficiency can show as a spectrum of distortion running from negligible wrist weakness to wrist, thumb what's more, elbow impairment.⁽³⁾ Associated oddities and skeletal disorders are to a great degree regular with spiral hypoplasia and must be prohibited preceding any sort of surgical intervention.⁽⁴⁾ At slightest 40% of kids with outspread club hand have some related restorative issue. Generally essential is to recognize any related hematologic or cardiovascular variation from the norm on the grounds that these are regularly life-undermining.⁽⁵⁾

Radial aplasia is divided into 4 types based on severity and range from shortened radius to complete absence of the radius.⁽⁶⁾ Radial club hand can be detected by clinically, radiographs, laboratory to screen associated conditions.⁽⁷⁾

Extending and orthotic mediation for outspread deficiencies can begin soon after birth. The objective is to diminish the hand/carpus onto the distal ulna and anticipate contracture of the wrist in spiral deviation.

The extending convention comprises of dynamic longitudinal diversion, ulnar deviation, and expansion with adjustment of the

ulnocarpal joint. Any change in ROM accomplished through activities can make future surgical correction less complex.⁽⁸⁾ Notwithstanding extending , outspread canal orthosis which leaves the thumb and fingers free is prescribed amid the day to keep up the remedy got from latent control. Patients with an abbreviated lower arm and those with elbow expansion restrictions may require an orthosis over the elbow.⁽⁹⁾

Centralization and radialization have become accepted treatment options for children with complete absence. The objective is to give wrist dependability while expanding the useful length of lower arm by putting the wrist on the distal end of the ulna. These methodology likewise enhance the arrangement of the flexor tendons.⁽⁹⁾ A temporary pin is placed across the ulnocarpal joint to stabilize the hand and wrist on the ulna.

Amid these systems the staying spiral tendons are regularly exchanged to the ulnar part of the carpus trying to rebalance the wrist and avoid repetitive contractures. Reestablishing muscle parity is imperative for forestalling repeat of distortion.⁽¹⁰⁾

Repeat of spiral deviation of the wrist as the youngster develops has been connected with the measure of remedy acquired at the underlying surgery and the age at the season of beginning surgery.⁽¹¹⁾ A provisional procedure using an external fixator from the ulna to the small and ring metacarpals is often required.⁽¹²⁾ The Ilizarov method is recently developed and applied three dimensional distraction-correctional device that has great value in upper extremity.⁽¹³⁾ The surgical treatment of radial club hand would be complete with comment on role of ulnarization of wrist followed by pollicization of index finger.⁽¹⁴⁾

EMBRYOLOGY OF LIMB DEVELOPMENT

Normal development of the limbs begins at the end of the fourth week after fertilization (table 1)⁽¹⁵⁾, with limb buds forming in the mesoderm along the flank of the embryo (Fig. 1–1).⁽¹⁶⁾ The limb bud is divided into three major regions. The apical ectodermal ridge (AER), in which several fibroblast growth factors are expressed, keeps the adjacent mesenchymal cells in an undifferentiated, rapidly proliferating state. This mesenchyma is known as the progress zone. The third zone is the zone of polarizing activity (ZPA). This region is responsible for anteroposterior polarization as the limb develops. The cells that remain in this region the longest populate the distal portion of the extremity.⁽¹⁷⁾ The limbs develop in a proximodistal direction from the limb girdle to the digits. The proximal bones of the limb girdle and the humerus or femur form before the differentiation of ridge ectoderm, whereas development of the remaining bones and digits depends on the AER.⁽¹⁸⁾⁽¹⁹⁾

The AER is formed by the thickening of lateral plate mesoderm, which signals the overlying ectoderm to thicken and establish a ridge over the tip of the limb bud. The AER regulates the proximodistal growth of the limbs (Fig. 1–2). Although the AER causes outgrowth of the limbs, the mesenchyma determines the type of limb that will develop.⁽²⁰⁾⁽²¹⁾ The bones and connective tissues of the limbs are formed by lateral plate mesoderm, and the muscles originate from myotome regions of the somatic mesoderm.⁽¹⁵⁾ Fore limb and hind limb development occurs via similar mechanisms, with upper limb growth preceding lower limb growth by 1 to 2 days. By 6 weeks, as the buds extend distally, the terminal parts of the limbs flatten to form hand and footplates, complete with distal rays, and cartilage begins to appear in the proximal portions of the limbs.

Table 1: Timeline progression of limb development. ⁽¹⁵⁾

Gestational Day	Embryonic Stage	Developmental Events
21	9	Notochord expresses EME
26	12	Upper limb bud forms
31	14	Limb bud curves
33	15	Formation of hand paddle Subclavian/axillary/brachial arteries form
36	16	Nerve trunks enter upper limb Chondrification of humerus and forearm Glenohumeral cavitation begins
41	17	Digital rays visible Chondrification of rays Ulnar artery forms
44	18	Chondrification of proximal phalanges Radial artery forms Pectoralis muscle splits into two heads
47	19	Chondrification of middle phalanges Initial separation of digits Digital cavitation/joint formation begins
50	20	Chondrification of distal phalanges Digital separation
54	22	Humerus ossifies Digital separation complete
56	23	Ossification of distal phalanges Nutrient vessel forms in humerus

During the seventh week, the limbs begin to rotate, with the forelimb turning 90 degrees laterally (positioning the thumb laterally) and the hind limb turning 90 degrees medially (positioning the big toe medially). Digital rays appear in the hand- and footplates. By the eighth week, the limbs have rotated to their final position, and all segments are complete, including the digits. During this time, ossification starts. By 12 weeks, ossification centers are present in all the long bones.⁽²¹⁾

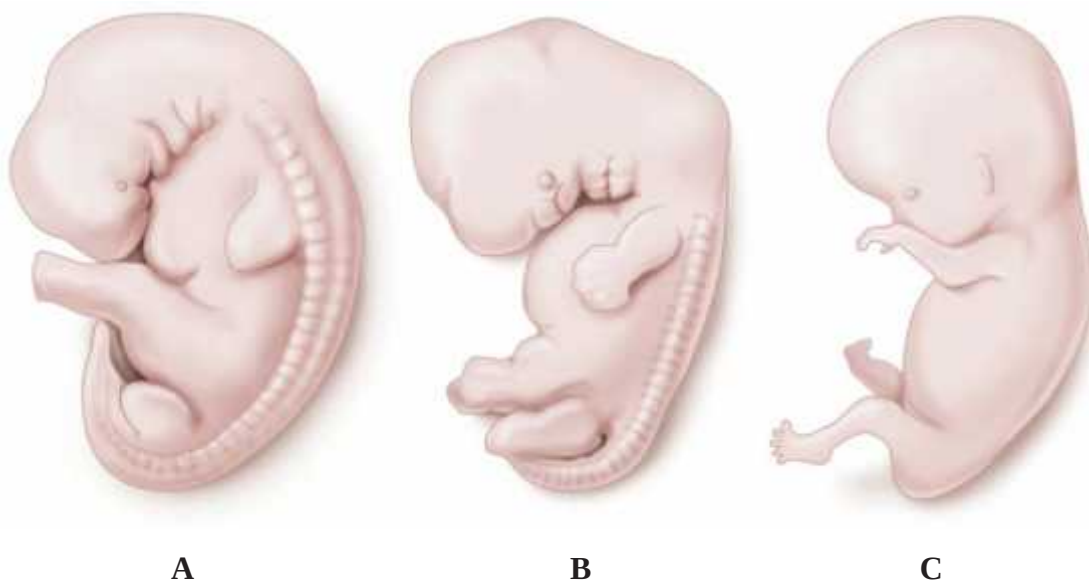


FIGURE 1–1 Limb bud development. **A**, The limb buds appear at the end of the 4th week after fertilization as mesodermal outpouchings on the flank of the embryo. **B**, During the 6th week, the terminal portion of each bud flattens to form the hand- and footplates, complete with digital rays. **C**, By the 12th week, cartilage appears in proximal segments, and ossification centers are present in the long bones.⁽²¹⁾