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# Split Forearm and Hand- a case report

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***Abstract:*** *A male of around 24years of age presented with splitting of left forearm and hand. Radial side of forearm continued hand with index finger only and ulnar side continued hand with ring and little finger. Thumb and middle finger were absent. Length of the forearm was comparatively shorter. No relevant family history was found. This was a case of congenital longitudinal growth defect of upper limb.*

***Case Report:*** *A male of around 24 years of age from Bihar presented a congenital defect of splitting of forearm and hand on left side. Splitting of forearm was extended up to just distal to elbow joint. Radial side of forearm was continued hand with index finger only. Thumb and middle fingers were absent. Unlar side of forearm was continued hand with ring finger and little finger. Length of the forearm was comparatively shorter [Fig 1&2]. Extension and flexion movements of the fingers were present. Left arm and right upper limb were normal. No limb defect was present in the lower limb. No relevant family history was found. This was a case of congenital longitudinal growth defect of upper limb. X-ray of the limb was not taken.*

***Key words:*** *split, forearm, hand and congenital.*

## I. Introduction

The upper extremity is developed on the ventrolateral wall of the embryo on day 26 (4-mm crown-to-rump length) (see image below). The limb bud develops from the Wolff crest and lies opposite the 5 lower cervical and 2 upper thoracic segments. At this stage, the limb bud appears as little more than a mesenchymal core covered by a thin layer of epithelium.

A thickening of 2-5 cell thickness, termed as the apical ectodermic ridge (AER) develops along the ventromedial border of the limb bud, which is vital in axis orientation, outgrowth, and digitations. It covered a layer of 5-15 cells thick, undifferentiated, proliferating mesenchymal cells. This underlying mesenchymal layer is known as the progress zone (PZ). PZ is also essential in limb outgrowth. The AER and PZ are closely associated with a large marginal blood vessel, the marginal sinus. These vessels branches at the base of the limb bud and it continued to join the cardinal vein. The AER and PZ are responsible for the outgrowth of the limb along the proximodistal axis and the marginal blood vessel may convey messenger proteins that integrate this process.

In the seventh week, the upper extremity continues outgrowth and rotates 90° so that the elbows project posteriorly and the developing hands lie on the anterior thorax. At this time, the cartilaginous models of the proximal bones are undergoing ossification at ossification centers within the diaphysis of each model. Mesenchymal cells derived from the dermomyotome condense within connective tissue (mesenchymal) scaffolds to form 2 common muscle masses. They are immediately penetrated by the spinal nerves. These dorsal and ventral common muscle masses split later to form the muscles of the extensor and flexor compartments, respectively.

The process of developing digits continues as the AER begins to fragment and notched digital rays are, thus, formed by day 46. At 50 days, the digits are webbed, and at 52 days (week 8, 23-mm crown-to-rump length), they have become separate digits. The process of forming separate digits involves the fragmentation of the AER and subsequent apoptosis, which involves programmed cell death (apoptosis) within the digital interspaces.

Summary for development of upper limb as follows [1]

- Onset of development of arm bud - 27 days
- Well-developed arm bud - 28-30 days
- Elongation of arm bud - 34-36 days
- Formation of hand paddle - 34-38 days
- Onset of finger separation - 38-40 days
- Full separation of fingers - 50-52 days

## II. Discussion:

Studies on these types of defects are important for corrective surgery and prosthetic management. According to different study, the incidence of upper limb defect are 22.5 out of 10000 (in Edinburgh) , 19.76 out of 10000 (in western Australia) and 21.5 out 10000 (in Sweden) life birth. Many authors have described about the different types of upper limb defects.[2]

The international classification for congenital hand anomalies based on an extension of an earlier classification system.[3]

**Groups**

I. Failure of formation; transverse (A), or longitudinal (B) (radial and ulnar deficiencies, (symbrachydactyly)
II. Failure of differentiation
III. Polydactyly (513 anomalies, Madelung deformity, the Kirner deformity and congenital trigger fingers and trigger thumbs, Triphalangeal thumbs)
IV. Overgrowth
V. Undergrowth
VI. Amniotic band syndrome (amniotic bands)
VII. Generalized skeletal syndromes.
viii. Unclassifiable

Some of these abnormalities can be initially detected prenatally by ultrasound and may be associated with other syndromes or karyotype anomalies such as Split hand-foot malformation1- 7q21.3-q22.1 , Split hand-foot malformation 2- Xq26, Split hand-foot malformation 3-10q24-35, Split hand-foot malformation 4-3q27 and Split hand-foot malformation 5-2q31. .[2]

The development of hand is a complicated interaction. During the development of hand during fifth week, homeobox transcription factors (HOX) and sony hedge hog (SHH) gene together determine digit number and identity. SHH induces an ulnar-to-radial gradient of bone morphogenic proteins (BMPs) that induces programmed cell death (apoptosis) in the interdigital space by suppressing the FGF expression in the overlying AER. BMP also helps establish digital identity by maintaining FGF in the AER overlying digits and inducing sex-determining region Y-related, high-mobility group box 9 (SOX9) there. SOX9 regulates chondrogenesis in the phalanx-forming (PFR) region of the developing hand. It is not clear how the family of BMPs interacts to produce the pattern of alternating digit and webspace recession. [2]

Disturbance of AER by amniotic band or other causes lead to abnormal digit formation. It has been mentioned that if the length of AER is reduced, then fewer digit will form, whereas if the AER is not reduced and becomes longer, then more digits will form.[4]

Defects present in the present case i.e. split of forearm and hand which is not found in the available literatures, may be postulate due to 1) prolongation of digital rays up to forearm, but absence of some of the fingers can't be explained, 2) amniotic band defect separating hand and forearm, and cutting of other fingers, 3) a unknown genitical cause or 4) combination of amniotic band defect and a genetical cause.

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Fig 1

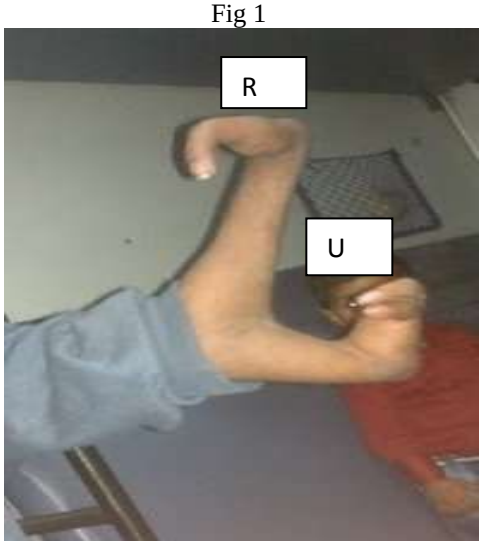
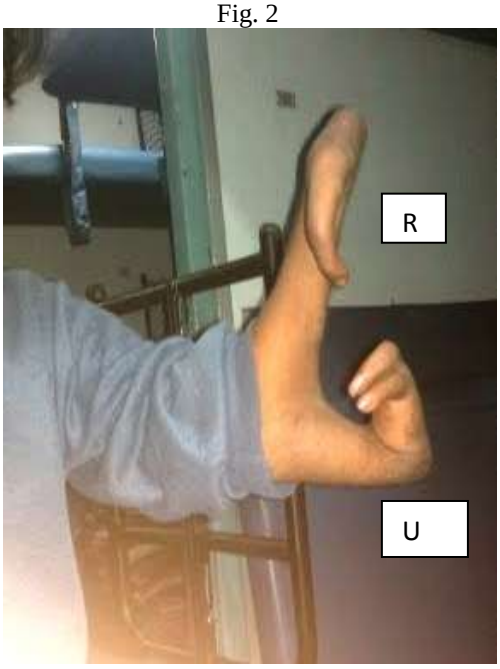



Fig. 2




Showing Radial side(R) with index finger Ulnar side (U) with ring and little fingers