Systematic review on Ilizarov technique in the management of congenital radial club hand

An Essay

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List of Abbreviations

A.P.	:	Antro-posterior
CORA	:	Centre of rotation of angulation
F	:	Female
M	:	Male
MTP	:	Metatarso-phalangeal Joint
R TA	:	Resolved total angle
RDL	:	Radial longitudinal deficiency
TA	:	Total angle
UB	:	Ulnar bow
UE	:	Ulnar epiphysis
WA	:	Wrist angle
3-D	:	3-dimentional

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Introduction

Radial club hand is a term first used in 1733 by Petit in an autopsy of a neonate to describe a congenital deformity of the upper extremity. [1]

It presents with a spectrum of hand and forearm anomalies ranging from mild radial hypoplasia to complete absence of the radius. Severe radial hypoplasia is clinically characterized by shortening of the forearm and radial deviation of the wrist, the thumb may be absent or hypoplasic. [2,3]

Paul and Manske 2009 reported that the incidence of radial club hand ranges from 1 in 30.000 to 1 in 100.000 ,slightly more common in male than female. Bilateral in 38 to 50 percent of cases. [4]

Many factors were claimed to cause congenital radial club hand, mostly environmental factors as compression, inflammatory processes, nutritional deficiency, irradation, infection and medication especially thaliodomide.[5]

Radial club hand was classified the into four types. Type1: mildest form with defective distal radial epiphysis, minor radial deviation of the hand is apparent, thumb hypoplasia may be present.

Type 2: limited growth of the radius on both its distal and proximal sides, characterized by a miniature radius and moderate radial deviation of the wrist.

Type 3: absence of two thirds of the radius, most commonly the distal side and severe radial deviation of the wrist.Type 4: is the most common and the most severe, it involves complete absence of the radius, the hand tends to develop a perpendicular relationship to the forearm. [6]

James et al has modified the classification scheme by adding type N and type 0. Type N: has a normal length radius and a normal carpus with thumb hypoplasia. Type 0: has a normal length radius and radial side carpal abnormalities.[7]

Flatt in 1994 described the condition as"an abnormal hand joined to a poor limb by a bad wrist".[8]

Treatment goals of radial club hand focus on the creation of a stable centralized and functionally hand ,maintenance of a mobile and stable wrist and preservation of longitudinal forearm growth. [9]

Correction of radial club hand requires a combination of operative and non operative management.[10]

The non- operative management comprises of stretching of the taut radial structures by physiotherapy and splintage.[11]

Dobyns et al., 1993 stated that in type I radial club-hand with mild radial deviation of the wrist, serial manipulation and casting in early infancy is effective in correcting deformity.[12]

The operative treatment aims at centralization of the carpus on the distal end of the ulna in conjunction with soft tissue release.[3]

Radialization was introduced more recently to overcome the short comings of centralization ,it entails release of contracted soft tissue on the medial side of the wrist and forearm,osteotomy of the ulna ,transfer of flexor and extensor carpi radialis tendons to extensor carpi ulnaris and realignment of the carpus on the ulna .It avoids excision of any carbal bones.[13]

Previous methods of treatment were concerned largly with correction of radial deviation of the wrist and pay little attention to the ulnar hypoplasia,and so recurrence of radial deviation had been commonly reported leading to unsatisfactory functional and cosmetic results.[14]

The introduction of Ilizarov method has dramatically changed the possibilities for treatment of many congenital and acquired conditions. This procedure had been used to correct severe residual shortness and bowing of the ulna and recurrent radial deviation at wrist in congenital radial club hand in which a previous wrist operation had been performed.[15]

The Ilizarov technique applied three dimensional distraction osteogenesis device that has a great value in the upper extremity. It has been used to lengthen the radius alone(type 1or2 radial dysplasia),lengthen ulna for late radial aplasia (type 3or4). [16]

Pickford, Scheker believe that the ilizarov device offers certain advantages over other external fixators for pre-centralization soft tissues distraction, lengthening of ulna and Multi-planer deformity correction. [17]

Children with radial longitudinal deficiency have very short forearm. The ilizarov method of distraction osteogenesis has been used to increase the extremity length but results differ with the underlying condition. The process of lengthening is prolonged with frequent complications. In successful cases, patients are pleased with the function and appearance of the limb.[18]

Aim of work

The aim of this work is to systematically review the literature as regarding the the management of congenital radial club hand by Ilizarov technique including:

Indications, technique, results and possible complications.

Embryology of the upper extremity

Embryogenesis

Limb development begins during embryogenesis with events that affect the position, number, and orientation of the limb as follow:

- At 26 days after fertilization, the limb bud starts to develop & continue developing rapidly through 47 days.
- Fifty-two to fifty-three days after gestation, the fingers are entirely separate. Eight weeks after fertilization, embryogenesis is complete and all limb structures are present.
- At this point, the joints develop by condensation of chondrogen to form dense plates between future bones. Joint cavitation further forms the articulation, although proper joint development requires motion for modeling of the ultimate joint surface.

> The majority of upper extremity congenital anomalies occur during this 4- to 8-week period of rapid and fragile limb development.

- After 8 weeks' gestation, the fetal period commences with differentiation, maturation, and enlargement of existing structures. ***(19)***

The limb bud represents an outgrowth of the mesoderm into the overlying ectoderm.

Two sources of cells migrate from their origins into the limb bud:

- The cells from the lateral plate mesoderm become bone, cartilage, and tendon.
- The cells from the somatic mesoderm form the muscle, nerve, and vascular elements of the limb bud **(Fig . 1)**.

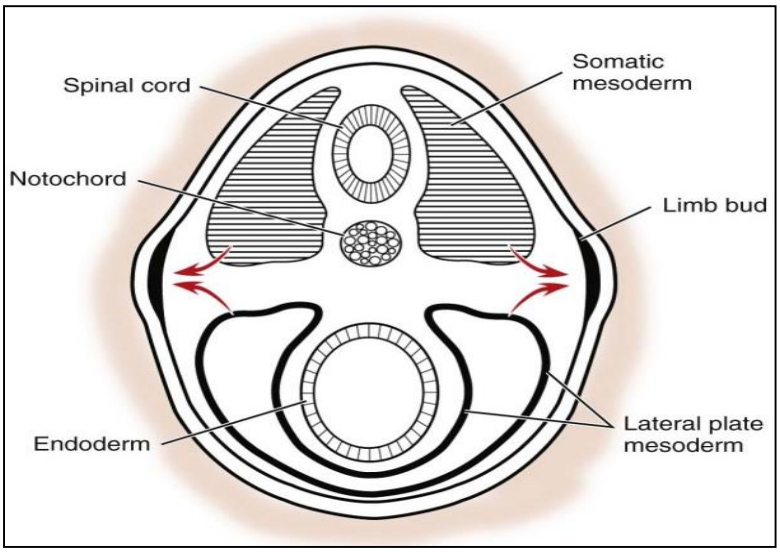

Fig.(1): Axial view of an embryo. The lateral plate mesoderm forms the bone, cartilage, and tendon, and the somatic mesoderm forms the muscle, nerve, and vascular elements of the limb.*(20)*

The signaling centers that control the three spatial axes of limb development: proximal-distal, anterior-posterior, and dorsal-ventral are the apical ectodermal ridge (AER),