

RESEARCH ARTICLE

CASE SERIES OF COMPLICATIONS ASSOCIATED WITH WRIST CENTRALIZATION IN RADIAL CLUBHAND.

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| Manuscript Info | Abstract |
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Introduction:-

Congenital longitudinal deficiency is divided into radial and ulnar deformities. Radial is more common than ulnar. Radial club hand is a congenital deformity of partial or complete absence of radius due to malformation of preaxial border of the upper limb. It is characterized by deviation of radius and short forearm. More than half of radial clubhand occur bilaterally, and the incidence is more in males.

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Case 1:-



This is a case of 9 year old primii child who was born with bilateral forearm deformity and absence of thumbs. He is a product of uncomplicated spontaneous vaginal delivery with no evidence of prenatal screening.

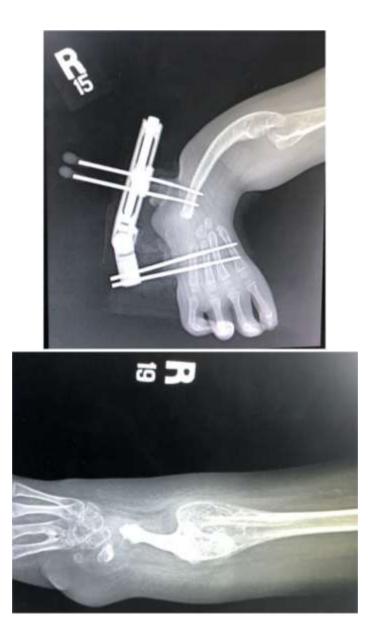
The patient had been thoroughly investigated after birth, for which he had ultrasound of abdomen, he had normal size left ectopic kidney. Also he had echocardiogram that showed small patent ductusarteriosus, and abnormal vertebra at L3. He was 8 month old in his first visit to orthopedic surgery in October/2009.

On examination: No obvious dysmorphic features. He had normal looking face and cervical spine with no abnormal or hair dimples or any clinical scoliosis. His hips were both stable with full range of motion, normal knees, legs and

feet. Local examination of the upper limbs revealed he had bilateral good shoulder and normal contour and movement, but there was obvious bilateral shortening of the forearm and radial deviation of both wrists and bowing of ulna on both sides with complete absence of the thumb on both sides. Forearm x-ray showed revealed he had bilateral complete absence of radius and also bilateral complete absence of the thumb.

Patient was diagnosed with grade IV radial clubhand. The managment began immediately with stretching exercises and brace supplementation then centralisation of wrist after 1 year of age. 18 months later, centeralisation of the right ulnar bone with the right was done, and the patient tolerated the procedure without complication then he was put on slab postoperatively. Follow-up was 2 months after the date of discharge, and there was migration of K-wire that protroded from the distal part of the wrist. It came from metacarpal bone for which the K-wire was removed under aspetic technique in the clinic. Meanwhile, he was put in splinting because there was 30% of callus formation around the osteotomy part which was done during wrist centralisation. He was seen again in clinic after 8 months, and the follow-up x-ray revealed completely healed bone but the deformity recurred for which the parents were offerred to redo centralisation of the right wrist. Surgery was redone in October/2012, and he was put in above right elbow cast and bivalved. Patient was admitted on October/2017 for removal of k-wire of right hand and forearm due to the breakage of k-wire.

Case 2:-



15-year-old Saudi female a product of spontaneous delivery, full term birth weight was 2.5 kg. Patient was referred to king Faisal specialist research center from Asir central hospital she was diagnosed at birth with syndromic bilateral club hand. She had type 4 isolated radial club hand. Patient had no family history of same condition. All of her siblings were doing well. Past surgical history tracheoesophageal fistula repair. Patient was born with one kidney On examination generally she looked good, right upper limb showed shoulder muscle atrophy, but normal joint. Elbow range of motion between 10 to 110 degrees. Supination and pronation 0 and bowing of ulna and shortening. The wrist is subtle and the right thumb is absent.

This patient underwent external fixator application for traction at the right forearm on June 2006. She was discharged. After that patient underwent removal of external fixators and centralization of the right wrist on September/2007. Postoperatively patient was doing well no complications. She had a follow up after 6 weeks This procedure failed to reach full correction due to distal circulation compromise.

Patient had recurrent deformity because the k-wire was accidentally removed and the progress was lost. Patient was admitted on October/2007 for a redo of her right radial clubhand. She was doing well postoperatively Patient was put on full cast above the elbow at 90 degrees. Patient was admitted electively for k-wire removal of right hand on March/2010. Patient developed maluinon of the ulna bone and no further treatment was done for her.

Discussion:-

Radial clubhand is a congenital deformity of partial or complete absence of radius due to malformation of preaxial border of the upper the limb, and characterized by deviation of radius and short upperlimbs¹². Half of radial clubhand occur bilaterally³. Radial clubhand is considered as the most common longitudinal malformation, although its is rare condition. It occures once in every 20,000 livebirths, and it is slightly more common in males⁴. The deformities can be either associated with multiple congenital defects or syndromes such as VACTREL, or sporadic and less commonly inherited⁵.

Radial clubhand deformity was classified by Bayne into four grades according to the radiological findings: grade I: short radius with the presence of the distal growth plate; grade II: hypoplastic radius with absence of the distal growth plate; grade III: partial absence of the radius, and the most severe grade IV: complete absence of the radius with abnormal ulnar curvature⁶.

Selection of treatment modality of radial clubhand is based on the severity of the deformity, and the age. For type I and II, which considered mild types, treatment modality bone lengthing with temporary external fixation of the wrist, or conservatively by splinting⁷.

Surgical correction by hand centralisation is the treatment of choice for radial clubhand in type III and IV^8 . It is usually done after 1 year of age, and delaying the intervention will make it more complex and challenging to correct⁹. Piror to surgery, muscle stretching is beneficial in correction of ulnar malalignment for successful centralisation, starting immediately after birth then splinting after maturity of the forearm, but conservative treatment is not helpful in late presentation or after two to three years of age¹⁰. Unfortunately, more than 45% of

¹James MA, Green HD, McCarroll HR, Jr, Manske PR. The association of radial deficiencywiththumbhypoplasia. J Bone Joint Surg Am. 2004;86(10):2196–2205.

²Walia JPS, Singh R, Sareen S et al. Radial club hand: a case report. Indian J Orthop 2006;40:267–8 doi:10.4103/0019-5413.34511

³Louis S, David W, Selvadurai N. Systems of orthopedics and fractures. 9th ednHodderAnorld, Hatchet UK Company, 2010:386–8.

⁴Ekblom AG, Laurell T, Arner M. Epidemiology of congenitalupperlimb anomalies in 562 childrenbornin 1997 to 2007: a total population studyfrom Stockholm, Sweden. J Hand Surg Am. 2010;35(11):1742–1754.

⁵Radial ClubhandTreatment& Management. **Medscape**, 2015. http://emedicine.medscape.com/article/1243998-treatment (accessed 30 Jan 2018). ⁶Bayne LG, Klug MS. Long-termreview of the surgicaltreatment of radial deficiencies. J Hand Surg Am. 1987;12(2):169–179.

⁷Lamb DW. Radial club hand: acontinuingstudy of sixty-eight patients with one hundred and seventeen club hands. J Bone Joint Surg Am. 1977;59(1):1–13.

⁸Manske PR, McCarroll HR, Jr, Swanson K. Centralization of the radial club hand: an ulnar surgical approach. J Hand Surg Am. 1981;6(5):423–433.

⁹Golfarb CA, Manskee PR, Busa R et al. Upperextremityphocomeliareexamined: a longitudinal dysplasia. J Bone Joint Surg Am 2005;87:2639–48 <u>doi:10.2106/JBJS.D.02011</u>

cases are prone to recurrence after surgical correction¹¹. In addition, centralisation procedures result in physeal injuries that can worsen it is proved that patients who underwent centralisation grew almost half of the normal ulnar length, while 64% of normal ulnar length¹²¹³. With these challenges and difficulties in treatment of radial clubhand, seeking other alternative treatment choices with less complications is necessary today.

¹⁰Kotwal PP, Varshney MK, Soral A. Comparison of surgical option and non operative management for radial longitudinal deficiency. J of Hand SurgEur Vol2012;37:161–9 doi:10.1177/1753193411413070

¹¹Shariatzadeh H, Jafari D, Taheri H, Mazhar FN. Recurrence rate after radial club hand surgery in long termfollow up. J Res Med Sci. 2009;14(3):179–186.

¹²Heikel HV. Aplasia and hypoplasa of the radius: studies on 64 cases and on epiphyseal transplantation in rabbits with the imitated defect. Acta OrthopScand Suppl. 1959;39:1–155.

¹³ Sestero AM, Van Heest A, Agel J. Ulnargrowth patterns in radial longitudinal deficiency. J Hand Surg Am. 2006;31(6):960–967.