Centralization of Radial Club Hand

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Abstract
Radial club hand, often referred to as radial longitudinal deficiency (RLD), is a rare congenital condition characterized by an underdevelopment and malformation of the radial side structures of the forearm and hand. The anomaly, first described by Petit in 1733, is characterized by a significant shortening of the forearm, radial angulation of the wrist, impaired range of digital motion and limited strength in pinch and grip. In addition, the thumb is always affected with a varying degree of hypoplasia or aplasia. It, encompasses a collection of congenital deformities characterized by hypoplasia of bones, joints, muscles, tendons, ligaments, nerves, and vasculature on the radial aspect of the forearm [1,2]. Radial club hand is a rare condition, occurring in approximately 1 in every 30,000 live births, with a slightly increased incidence in males. The anomaly occurs bilaterally 50% of the time, and is frequently asymmetrical [3].

Keywords: Hypoplasia; Thrombocytopenia Absent Radius

Abbreviations: TAR: Thrombocytopenia Absent Radius; RLD: Radial Longitudinal Deficiency; PIP joint: Proximal Interphalangeal Joint; MCP joint: Metacarpophalangeal Joint; DIP joint: Distal Interphalangeal Joint; AP: Anteroposterior; SPSS: Statistical Package for Social Sciences

Introduction
Most cases of radial deficiency are sporadic, and they are commonly associated with a number of congenital syndromes: thrombocytopenia absent radius (TAR) syndrome; vertebral defects, anal atresia, cardiac malformation, tracheoesophageal fistula, esophageal atresia, renal anomalies, and limb anomalies (VACTERL); Fanconi’s anemia; Holt-Oram syndrome; and trisomies 13 and 18 [1-4].

Since the 19th century there has been an evolution of surgical methods to correct the radial angulation of the wrist with the aim of improving function and appearance. In spite of this, the long-term results of surgery are discouraging with a high rate of late deformity recurrence and impaired ulnar growth [5,6]. Functional improvements have been difficult to verify and the routine use of surgical correction of the radially deviated wrist in individuals with RLD is currently debated [7,8].

Current knowledge about the relationships between, on the one hand, different physical aspects of the deformity in RLD (body function and structure) and, on the other, activity and participation among individuals with RLD, is sparse. The modalities of treatment have traditionally varied from stretching casts with soft-tissue procedures to the use of multiple corrective osteotomies. These osteotomies can be stabilized by a variety of methods, including external fixators that allow the possibility of gradual distraction with neohistiogenesis [9].

Clinically, the radial club hand spectrum is classifiable by severity into one of four types based on the degree of radius present (as described by Bayne and Klug) or by a modified scheme that grades thumb, carpal, and radius deficiencies in summation [2,6].

Treatment of radial club hand has progressed over the years from no treatment to aggressive surgical correction. In an untreated radial club hand, although the deformity did not change much, the hand’s prehensile function was never able to develop. Various surgical methods of correction have been described, such as soft tissue releases with or without ulnar osteotomy, proximal fibular transplants, arthrodesis, centralization, correction by distraction in an external fixator, radialization, and Ilizarov’s methods. Centralization of the hand over the distal ulna is still the basis for the modern-day treatment of this condition [10].
Aim of Work

This study is designed to study the results of Centralization for children with congenital radial club hand presented to Hand and Upper Limb Unit in Mansoura University Hospital in Egypt.

Patients

Study design

A study for cases with congenital radial club hands presented to the Hand and Upper limb Surgery unit; managed by Centralization of the hand over the distal ulna.

Inclusion criteria

Children who have radial club hands of Heikel’s Grade III and IV: 2- Unilateral and bilateral cases of radial club hand.

Exclusion criteria

a. Children who have radial club hands of Heikel’s Grade I and II. 2- Older children who have radial club hands with good function.
b. Children who have other causes of radial club hand e.g.: (arthrogryposis, post traumatic radial club hand).
c. Children who have radial club hands with elbow extension contracture who rely on radial deviation.
d. Children with multiple congenital anomalies not compatible with life.

Methods

Pre-Operative Assessment

Clinical assessment: The deformity will be measured with the help of a goniometer. Radial deviation and volar flexion of wrist at rest will be measured to find out the initial deformity. Movements at elbow and fingers will be noted. The movements of fingers will be taken individually of each finger as total digital motion, i.e., the sum of the movement at metacarpophalangeal joint (MCP joint), proximal interphalangeal joint (PIP joint), and distal interphalangeal joint (DIP joint). The status of thumb will be noted.

Radiological assessment: Radiological assessment with full length anteroposterior and lateral views of hand, forearm, and humerus. The length of ulna and humerus, angle of ulnar bowing and, third metacarpal to ulna angle in both views will be measured. The patients will be also screened radiologically to find whether any evident deformities will be present in other areas like chest, spine, and hip. Using Bayne and Klug Classification and Modified Bayne and Klug Classification.

Hematological investigations: In form of hemoglobin, total leucocyte counts, differential leucocyte counts, and platelet counts will be done to rule out any associated hematological derangements.

Operative Technique

Under general anesthesia, A lazy ‘S’ or ‘C’ dorsal midline incision, or a longitudinal ‘Z’ plasty on radial side with transverse incision on ulnar side, or a bilobed incision will be used depending on the amount of initial correction obtained and the amount of redundant skin left in ulnar side. A slot will be cut in the proximal carpal row, with its depth equal to width of distal ulna. The cartilage of the distal ulna will be shaved with a sharp blade if needed to fit into the carpal notch. A K-wire will be passed through this slot into the second metacarpal and will be retrogradely pushed into the ulna after seating the ulna in the notch. The wrist will be immobilized in an above elbow plaster cast in maximum elbow flexion and mid prone position.

Post-Operative care

The limb will be elevated and observed for any swelling, discoloration of the fingers, and stretch pain. Assisted finger movement will be done by the mothers for first 48 hours. Sutures will be removed on 10th day and the plaster cast will be changed. The K-wire will be left in situ. Plaster cast will be changed at monthly intervals until 3 months when the cast will be removed, and a below elbow polypropylene splint with radial support will be applied and continued for another 6 months, after which this splint will be removed in day time and used only at night. During this period of splinting, the parents will be advised to passively mobilize all the finger joints and elbow and promote the child to use the hand in day-to-day activities so as to develop prehension with the corrected wrist position.

Follow up of cases

Follow-up evaluation will be done at regular intervals of 3 months. The average follow-up period will be 1.5 years (range 8 months to 2 years). At final follow-up, the range of motion of the fingers and elbow will be recorded. Radiological evaluation to find change in length of ulna, change in bowing of ulna, and hand to forearm alignment with third metacarpal to ulna angle in anteroposterior (AP) and lateral views will be recorded. Final data will be compared with the data at their initial presentation to find out the amount of growth the ulna has attained during this period, to find out any increase in ulnar bowing, and to see if the hand forearm alignment is maintained. The final assessment of the result will be made based on criteria of Bora. The results will be classified satisfactory if the third metacarpal is aligned in long axis of the distal part of the ulna in AP and lateral views, no increase in ulnar bowing, and longitudinal measurement of the ulna had increased at least 50% of the projected normal increase during the interval in which patient has been followed.

Finally, counseling of the parents will be undertaken, to explain the procedure, its complications, and prognosis, and written consent will be obtained for participation in study. Rehabilitation program will be discussed with the parents.
Data Analysis

Data will be entered, cleaned to identify inconsistence and statistically analyzed using the statistical package for social sciences (SPSS) version 22. Qualitative data will be described as numbers and percentages. X2 test or Fisher’s exact test will be used for comparison between groups, as appropriate. Quantitative data will be described as means (SD) or medians, as appropriate. They will be tested for normality by Kolmogorov-Smirnov test. In normally distributed variables, t-test and allied will be used while in non-normally distributed variables non-parametric tests will be used for comparison between groups. Odds ratios and their 95% confidence interval will be calculated. “P value ≤ 0.05” will be considered to be statistically significant.

References