A systematic review and meta-analysis of the outcome of the surgical correction of Congenital Radioulnar Synostosis

Submitted for Partial Fulfillment of the Requirements for Master Degree in ***Orthopedic Surgery***

By

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LIST OF ABBREVIATIONS

Abbreviation	Full Term
ASSH	American Society for Surgery of the Hand
CENTRAL	Cochrane Central Register of Controlled Trials
CI	Confidence Interval
CRUS	Congenital radioulnar synostosis
ICJME	International committee of medical journal association
MD	Mean difference
MOOSE	Meta-analysis Of Observational Studies in Epidemiology
PRISMA	Preferred Reporting Items for Systematic Reviews and Meta-analysis
RCT	Randomized Control Trials
ROM	Range of Motion
RR	Relative risk
SMD	Standardized mean difference

INTRODUCTION

Congenital radioulnar synostosis refers to an abnormal connection between the radius and ulna due to embryological failure of separation. De-rotational osteotomy has been advocated for children with functional limitations, although historically this procedure has been associated with a 36% complication rate including compartment syndrome and loss of correction. (1)

Presentation of radioulnar synostosis varies from cosmetic concerns with no functional limitations to significant pronation deformity which hampers activities of daily living. Surgical management must be considered based on the position of the forearm and functional limitations. (2)

A retrospective evaluation of consecutive patients who underwent de-rotational osteotomy for CRUS was performed. Children with functional limitations secondary to excessive pronation were indicated for surgery with a goal of correction to 10 to 20 degrees of pronation. All patients were treated with a standardized surgical technique including careful sub periosteal elevation, rotational osteotomy at the level of the synostosis, control of the osteotomy fragments, appropriate pinning techniques, and prophylactic forearm fasciotomies. Electronic medical records, preoperative radiographs, and postoperative

radiographs were reviewed. All patients successfully achieved union by 8 weeks postoperatively. There were no cases of compartment syndrome, vascular compromise, or loss of fixation. The overall complication rate was transient anterior interosseous nerve palsies, transient radial nerve palsy, symptomatic muscle herniation. Transient anterior interosseous nerve palsies occurred in patients with rotational corrections exceeding 80 degrees. De-rotational osteotomy can be safely and effectively performed in children with CRUS. Meticulous surgical technique, including control of the osteotomy, judicious pin fixation, and prophylactic fasciotomies, may diminish the risk of neurovascular compromise and loss of correction. [(1)]

Analyses of correlations between the deformities of radius and ulna and degrees of fixed pronation of these forearms through a three-dimensional image of radius and ulna with congenital proximal radioulnar synostosis from their computed tomographic studies. The average ulnar deviation, flexion and internal rotation deformities of the radius were 6°, 3° and 18°, respectively. The average radial deviation, extension and internal rotation deformities of the ulna were 3°, 4° and 30° respectively. The flexion deformity of the radius and the internal rotation deformity of the radius and ulna were correlated significantly with degree of fixed pronation. As a conclusion, the patients with congenital proximal radioulnar synostosis have remarkable flexion deformity

of the radius and internal rotation deformity of the radius and ulna, which might impede forearm rotation after corrective surgery in the proximal part of the forearm. [3]

Proximal radioulnar synostosis is a rare but highly disabling condition. Surgical treatment is an option for functionally limiting proximal radioulnar synostosis; however, the approach can endanger local neurovascular structures, especially if the synostosis affects the level of the bicipital tuberosity. Cases were reported of proximal radioulnar synostosis with a preoperative prono-supination range of motion of 0° and 15° treated by a reverse Sauvé-Kapandji procedure resecting a 1-cm section of the radial shaft distal to the bicipital tuberosity and leaving the synostosis in place. An improvement in prono-supination arc of motion of 82.5° was achieved at 2 years of follow-up with no complications associated with the technique. The reverse Sauvé-Kapandji procedure could be an option in the treatment of proximal radioulnar synostosis in those cases. [4]

Acute elbow extension deficit is an unusual phenomenon that has been observed in patients with congenital radioulnar synostosis. A case of an 11-year-old girl with congenital radioulnar synostosis who developed acute extension deficit of the right elbow and whose elbow range of motion was restored following lateral capsular release was described by professor Cheng PG. [5]

AIM OF THE STUDY

The aim of the study is to assemble, describe and analyze the results of surgical correction for congenital radioulnar synostosis through a systematic review and meta-analysis.

REVIEW OF LITERATURE

Etiology:

Congenital radioulnar synostosis is a rare anomaly in which proximal portions of the radius and ulna fail to separate and restrict rotation of the forearm. Congenital Radioulnar Synostosis has multifactorial etiology including both sporadic, mutation and undefined genetic patterns. [5, 8]

Radioulnar synostosis occurs due to the persistence of the interzonal mesenchyme which undergoes chondrification, ossification, and eventually synostosis. The proximal one-third of the forearm is the most common site of involvement. About 40 % of the cases are unilateral and 60 % bilateral. [8, 10] Males are affected more commonly than females. Approximately one-third of patients with radio-ulnar synostosis have associated anomalies involving the cardiovascular, genitourinary, gastrointestinal, central nervous and musculoskeletal systems. [8]

Radioulnar synostosis can be isolated or associated with a radial head dislocation. Radioulnar synostosis can be one of the physical findings a variety of syndromes, including Apert syndrome (acrocephalosyndactyly), Carpenter's syndrome (acropolysyndactlyly), Arthrogryposis, Mandibulofacial dysostosis, Klinefelter's syndrome (XXY) and other sex chromosome

abnormalities. These syndromes present numerous problems that are more compelling than the absence of forearm rotation, and this is usually the reason for a delayed diagnosis of radioulnar synostosis. In addition, shoulder and wrist motion can compensate for the lack of forearm rotation during many childhood daily activities. Even in a healthy child, a delay in presentation is common until the child begins engaging in more complex daily activities, such as catching a ball or eating. A careful examination is necessary to identify a lack of forearm rotation, particularly in the presence of compensatory inter-carpal rotation. [7]

Failure of longitudinal segmentation and the persistence of the cartilaginous anlage between the radius and ulna during the seventh week of intrauterine development is thought to be the major cause of congenital radioulnar synostosis. The fixed position of the forearm in pronation reflects the fetal position at this specific embryonic period. [8, 15, 16]

Patho-anatomy:

Synostosis may be longitudinal or transverse. Radio-ulnar synostosis is the most common form, resulting from the failure of completion of the distal-to-proximal separation of the forearm mesenchyme. It may be unilateral or bilateral and is usually at

the proximal end of the forearm. Some compensatory rotation occurs at carpal level. (9)

Disability depends upon the rotational alignment of the forearm. Shoulder motion compensates less readily for fixed pronation. Unilateral synostosis causes little disability and seldom requires treatment. Bilateral synostoses are often fixed in marked pronation. (9)

Clinical evaluation:

Most cases are asymptomatic, noticed by parents and teachers especially with specific tasks such as keyboard, tabletop activities that indicate deficient pronation, or eating, washing face, or catching a ball that indicates deficit supination. (11)

Patients with congenital radioulnar synostosis can go unnoticed until early adolescence, especially in unilateral cases, most commonly discovered at 6 years old. Most cases can do compensatory motion such as; shoulder abduction to compensate for loss of active pronation. (11)

The axis of rotation of forearm runs through radial head (proximal) and ulna fovea (distal); distal radius effectively rotates around the distal ulna in prono-supination, (***Figure 1***). In patients with congenital radioulnar synostosis, a prono-supination degree of the palm is possible by the means of compensatory rotation

motion in the wrist and the carpometacarpal joints. Therefore, the range of prono-supination of the palm was defined by the angle between the longitudinal axis of the humerus and the axis of metacarpophalangeal joints from the index to the little finger in the palm (apparent rotation). [11]

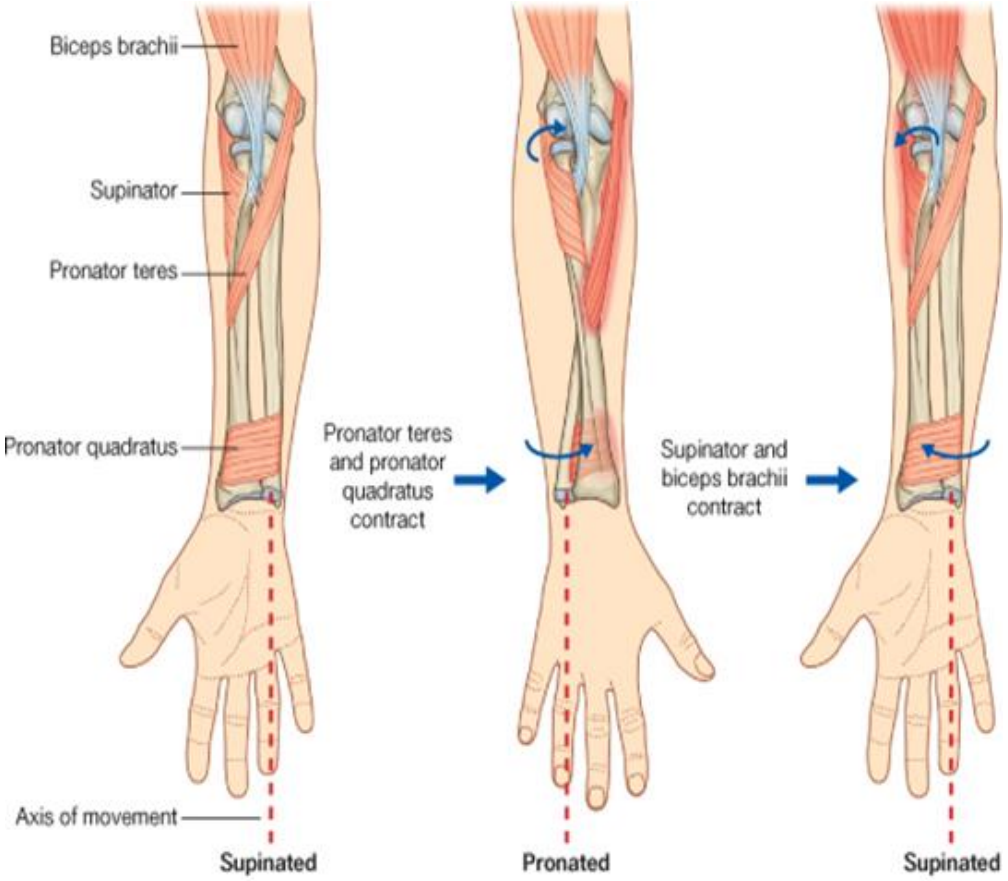

Figure 1: *The axis of rotation of forearm runs through radial head (proximal) and ulna fovea (distal); distal radius effectively rotates around the distal ulna in prono-supination.* [22]