

## **A Modified Sayre's Technique, for Correction of Congenital Radial Club Hand Deformity**

**Abdulrahim Aljayar\*, Monem Shakeer and Moattaz Aljayar**

*Hand Surgery Unit, Orthopedics Department, Aljala Trauma Hospital, Benghazi, Libya*

**\*Corresponding Author:** Abdulrahim Aljayar, Hand Surgery Unit, Orthopedics Department, Aljala Trauma Hospital, Benghazi, Libya.

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### **Abstract**

Congenital radial club hand deformity is a spectrum of complex pre-axial upper limb defect. Characterized by radial deviation and flexion of the hand, hypoplasia, or aplasia of the radius and thumb ray, shortened forearm and generalized underdevelopment of the involved extremity. That was and still wondering therapists since first described by (Petit) in 1733. However a huge variety of techniques, ranging from conservative stretching, to the complex microsurgical interventions, have been suggested for treatment, the centralization procedure of (Sayre) remains a core idea for all thereafter attempts, despite a recognized relapse rate of relapse, Such diversity would reflects the great challenge in management. Here is a prospective assessment of promising results, in 15 hands, of 12 patients, with congenital radial club hand deficiency, using a modified technique of (Sayre).

**Keywords:** *Congenital Radial Club Hand Deformity; Sayre Procedure; Sayre Procedure; Modified Sayre Technique*

### **Introduction**

Congenital radial club hand deformity, also known as the radial dysplasia, radial ray deficiency, preaxial deficiency, or the congenital longitudinal radial deficiency. Is a cosmetically disfiguring and functionally disabling socioeconomic tragedy, of a complex preaxial deficiency involving the whole upper limbs from shoulder to phalanges, That may include, a small sized scapula, shorted clavicle, humerus and short curved forearm [1] with hypoplasia, partially, or completely absent radius, scaphoid, trapezium and trapezoid bones as well as the thumb ray. Inexistence of radio-carpal joint and the triangular fibrocartilage complex [1].

An abnormal deltoid, pectoralis major, biceps and brachialis [1] brachioradiales, palmaris longus, flexor carpiradiales and the supinator-pronator muscles, as well as the thumb intrinsic and extrinsic musculature [1].

Also the absence or hypoplasia of the radial artery.

However the brachial plexus is normal, the radial nerve usually terminates at elbow level, where its functions are compensated through the median nerve [1].

Movements are restricted in elbow [1] wrist, metacarpo-phalangeal and the inter-phalangeal joints, with absent forearm rotation in types 3 and 4 (Figure 1).



**Figure 1:** Congenital radial club hand deformity.

A condition that was and still confusing therapist, since first described by petty in 1733 [2].



*Jean-Louis Petit (1673 – 1750).*

**Figure 2**

The source of confusion is obvious when we consider; The variety of classifications brought in for typing the deformity [3] and that in itself adds more confusion.

The associated systemic anomalies, including: the Holt-Oram syndrome (heart-hand syndrome), [4] TAR syndrome (thrombocytopenia-absent radius) [5] Fanconi anaemia (a life-threatening condition) [6,7]. VACTERL syndrome, [8] as some of these associations may

prevent or change the way of treatment; and The age at presentation as well as surgery. Until a hundred sixty years later, when (Sayre), the first American Orthopedic Professor, introduced his centralization procedure in 1893 [9,10,11].



Lewis Albert Sayre (1820 - 1900).

**Figure 3**

In his original technique the distal ulna tapered and shape to fit a centrally created carpal notch, for correction of the hand-forearm alignment. An idea that was disputed by some of his colleagues that time, stressing that even this can correct the deformity, that will be at expense of the growing epiphyses of the distal ulna particularly its peripheral active zone, which is already short [12,13].

Since that many surgical and nonsurgical ideas and several modifications and procedures have been suggested to solve the problem, including that of (Lidge) [14]. Who preserved the ulna intact, by widening the carpal notch to accommodate the whole circumference of the distal ulna.

While (Gramcko), was against disturbing the carpal bones, through his radicalization technique, [15] centralizing the carpals through the scaphoid, instead of the lunate.

The ulnarization procedure of (Paley), [16] who translated the carpals and the whole hand media to the distal ulna.

Until recently, we have reached the more complicated microsurgical interventions, through the free vascularised epiphyseal plate transfer from the proximal fibula for example and the metatarso-phalangeal joint transfer of (Vilkki) [17-19].

Of course, passing through some historical procedures like, the sagittal splitting of the distal ulna to accommodate the carpals and the free bone graft procedure of (Albee) [20] providing proximal-radial bony support of the carpal bones.

Unfortunately none proved to be effective yet, because of a recognized tirade of relapse [21-29], That was attributed to; the inadequate intraoperative correction of the deformity; the musculotendinous tension imbalance across the created ulno-carpal joint; the lack of proximal-lateral skeletal support of the carpal bones; the premature removal of internally splinting Kirschner wires; the deficient follow up; and lack of patient and family compliance. So that, the original (Sayre) centralization procedure remains a reference for all attempts thereafter.

Here we are trying to contribute humbly, avoiding these and other negative remarks, introducing technically easier reduction and expectedly sustained correction of the deformity.

Through the creation of medial slope or tilt at the distal ulna of about 10 degrees resembling the natural slope of normal radius.

**Methods**

**Patients:** Out of total 102 upper limb congenital anomaly cases, referred to our unit, between 2012 and 2017. The results of centralization procedure using a modified technique of (Sayre) were collected, for 15 hands, of 12 patients, less than 2 years of age. Who were diagnosed as type 3 and 4 Congenital Radial Club Hand Deformity, according to the most frequently used (Bayne and Klug) classification [30,31].

Type I - Radius apparently normal, but shorter, since the distal epiphysis takes longer to appear and has lower potential for growth.

Type II - Hypoplastic radius. Abnormalities in the proximal and distal epiphyses, resulting in a miniature radius.

Type III - Absence of the proximal or distal third. Absence of the distal third is more common. The ulna is hypertrophied and angled radially.

Type IV - Complete absence of the radius. This is the commonest and severest form (Table 1 and Figure 4)

Type	Criteria
I	Radial shortening
II	Radial shortening
III	Partial aplasia
IV	Total aplasia

**Table 1:** Bayne and Klug classification of Congenital radial club hand deformity.



**Figure 4:** Bayne and Klug classification Congenital radial club hand deformity A. type 1, B. type 2, C. type 3, D. type 4.

Excluded are; the types 1 and 2 Congenital Radial Club Hand Deformity cases; the stiffed elbow joints in extension; patients of more than 2 years of age, at presentation; and those were unfit for general anesthesia.

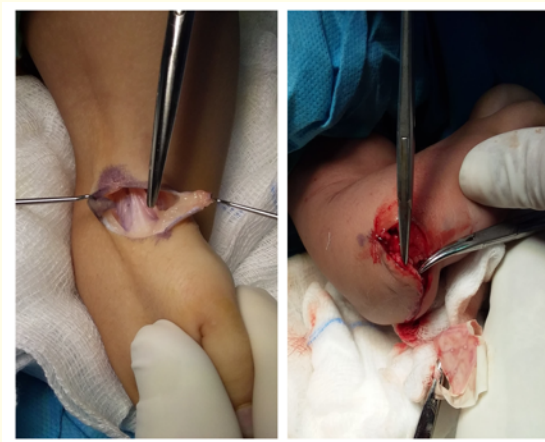
**Procedure**

Intervening as settled under general anesthesia, tourniquet control and TV monitoring. Through double approach, a lateral Z and medial curved incision, curved smoothly distal up to the proximal hamate level, creating a proximally based skin flap (Figure 5).



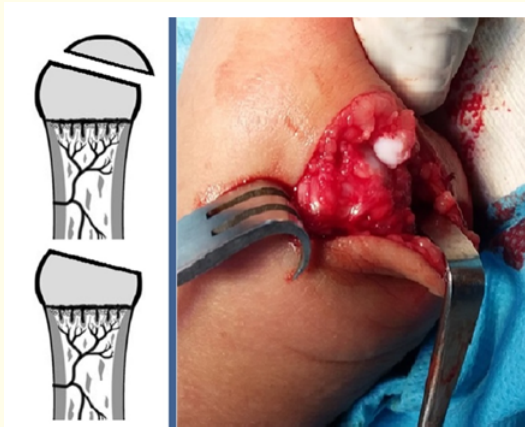
**Figure 5:** The lateral Z, and medial curved approaches.

Identifying and isolating the subcutaneous dorsal branch of median nerve, the extensor and flexor carpi-radialis tendons and dissecting carefully adequate the contracted soft tissues to free the carpal bones through the first and exposing the distal ulna, through the second incision (Figure 6).



**Figure 6:** The identification and protection of subcutaneous dorsal branch of median nerve, isolation of the radial wrist extensor and flexor tendons laterally, and the exposure of the ulna head medially.

Then under TV. control, a medially based triangular piece is sharply excised from the radiologically nonvisible head of the ulna, between the tip laterally and at least 3 mm. distal to the epiphyseal line medially (Figure 7).



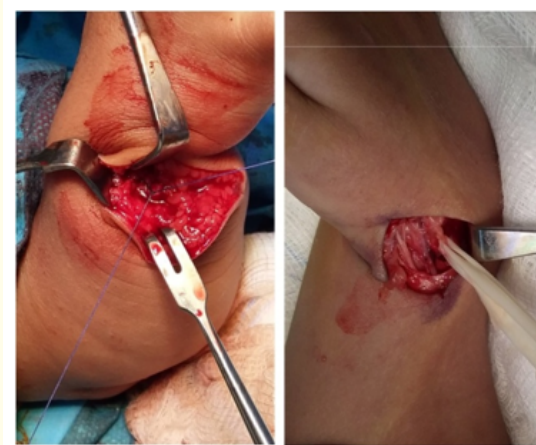
**Figure 7:** The excision of medially based triangular piece, from the radiologically nonvisible head of the ulna.

A suitable sized kirschner wire usually 1.5 or 2 mm, is then inserted longitudinally through the head of the third metacarpal, till appears about 1 cm. through the lunate proximally, when the drill hand piece detached and the inserted wire tip adjusted under vision towards the central portion of the distal ulna, to be used as a fulcrum when the hand is deviated medially in extension, to be maintained in about 10 degrees over correction, helped by the created tilt, through proximal advancement of the wire to penetrates the lateral-volar cortex of the ulna (Figure 8).



**Figure 8:** The centralization, of the hand in about 10 degrees over correction, maintained by advancing the K. wire to penetrate the lateral volar cortex.

The wire is then cut off distally and pended out of the skin. The redundant medial capsule tightened by augmentation or excision of excess tissue and repair. The Extensor Carpi-Ulnares tendon tensed up as needed and the Extensor Carpi-Radiales tendon is tunneled medially to be fixed to the Extensor Carpi-Ulnares tendon, while the Flexor Carpi-Radiales tendon if present, is tunneled dorso-medially to be fixed to and along the new axis of the Extensor Carpi-Radiales tendon (Figure 9).



**Figure 9:** Augmentation of the redundant capsule, tightness of the lax extensor carpi-ulnares and transfer of the available radial extensor and flexor tendons.

Lastly, the tight lateral skin lengthened by Z-plasty repair and the proximally based medial skin flap is harvested to close the medial wound. Finally, the limb immobilized in a long arm slab.

Postoperatively, the sutures removed in 2 weeks time, the wire extracted at 12 weeks, while splinting is continued in long arm splint for another 8 weeks (Figure 10).



**Figure 10:** Closure of the wounds and immobilization of the limb in long arm slab.

Then as night bracing for 76 weeks more (Figure 11).



Figure 11: Short arm night splint.

**Results**

Using this simple modified technique in 15 cases, of 12 patients, categorised as type 3 and 4 Congenital Radial Club Hand Deformity, 7 Males and 5 Females. 3 were bilateral involved, In 4, thumbs were totally absent, hypoplastic in 7 and normal in 4.

An associated ASD, detected in one bilaterally involved female, bilateral DDH, in a bilaterally involved male and an epsilateral Torticoles, in a unilaterally involved male patient. Their mean age at presentation was 2, while at surgery was 10 months (Table 2).

No.	Sex	Involvement		Thumb condition	Associated anomalies	Age/M	
		Rt.	Lt.			At presentation	At surgery
1	F	+	+	Absent	ASD	1	8-15
2	M	+		Normal		2	7
3	M	+	+	Hypoplastic	Bilateral CDH	1	7 - 16
4	M	+		Hypoplastic		3	11
5	M		+	Normal		2	9
6	M	+		Absent	Epsilateral torticoles	3	11
7	F		+	Absent		1	8
8	F	+		Hypoplastic		4	10
9	F		+	Normal		1	9
10	M	+	+	Hypoplastic		2	9 - 14
11	M	+		Normal		3	8
12	F	+		Hypoplastic		2	9
						2 / M	10 / M

Table 2



Using this modified technique, we could achieve an averaged postoperative hand-forearm angulation correction of 74.6 degrees, that was 84 degrees preoperatively, so we left with only 9,4 degrees residual angulation, following an average follow up period of 32.7 months (Table 3 and Figure 12).

No.	Angulation correction/ degrees	Preoperative angulation/ degrees	Residual angulation	Age at last follow up	Follow up period/M
1	72 - 90	80 - 100	8 - 10	62	54 - 47
2	54	60	6	56	49
3	80 - 62	90 - 70	10 - 8	50	43 - 36
4	82	90	8	48	36
5	62	70	8	44	34
6	80	90	10	42	31
7	72	80	8	40	31
8	54	60	6	39	28
9	80	90	10	36	28
10	80 - 62	90 - 70	10 - 8	30	21 - 16
11	80	90	10	22	13
12	74	80	6	16	6
	72.2/Degrees	80.6/Degrees	8.4/Degrees		31.7/M

Table 3



Figure 12: The results of hand-forearm angulation correction, using this modified centralization technique of Sayre's procedure.

### Discussion

Congenital Radial Club Hand Deformity is a complex pre-axial deformity affecting all the pre-axial structures of the upper limb [32]. With an incidence of 1:100,000 live births and may reaches 1:30,000 birth, if stillbirths included [32].

Reports were varying regarding sex predominance, it seems that there is an overall increased male predilection, Where had been reported as 3:1 male prevalence by Banskota and 2:1 male predilection by others [32]. In our series there was also a slight male predominance of 1.4:1 and in 20% of cases the involvement bilateral.

The deformity may arise as an isolated alteration, or in relation to the already mentioned medical syndromes and other sporadic anomalies, such as the, cleft pallet, scoliosis, kyphosis, talepes equinovarus, DDH and torticoles.

Where some of these syndromes are associated with short life spans, made correction of the deformity a meaningless attempt, others may prevent or delay the recommended golden early surgical interventions for correction.

In our sample, the problem was mostly isolated variation 66.6% and such associations of other systemic anomalies detected only in 33.3%.

The incidence of associated thumb absence was 26.6%, while the thumb hypoplasia was 46.6%.

Treatment of Congenital Radial Club Hand Deformity, should be started soon after birth, by stretching programs, which is well tolerated from 0 to 6 months of age, or distraction using external fixators, by the age of 8 months [15,16] as preliminary steps for the definitive surgical correction, that is agreed best performed between 6 months, to 2<sup>nd</sup> year of life, before development of persistent soft tissue contractures, secondary configurational bony changes and the exhibition of compensatory mechanisms.

This modified technique, provides a near normal carpal proximal- lateral support simulating the natural radio-carpal articulation, preserving the existing carpals configuration and saving the growing epiphyses of the distal ulna.

Supported by the recognition of an anticipated remodeling of the ulnar head, holding the proximal carpal raw in 5 cases, by the third to fourth postoperative years (Figure 13).



**Figure 13:** The observed remodeling of the ulna head, by the third-fourth postoperative years.

Balancing the deforming tension forces, by transferring the Extensor and Flexor Carpi-Radiales tendons if present, along the natural pulling axis of the tightened ECU tendon [1,32].

The practical delayed extraction of the internally splinting K. wire and enough external splinting period.

### Conclusion

Though, admitting the small sample number and the relatively short follow up period as limitations of our effort, we are introducing the promising result of this modification, with the promise to expose hopefully more satisfying results, in more convenient follow up period.

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