Radial club hand represents a wide spectrum of congenital anomalies of the pre-axial or radial side of the upper limb ranging from mild hypoplasia of musculature to complete absence of structures. It is also known as radial longitudinal deficiency (RLD) and is a failure of formation of parts — longitudinal arrest type of congenital anomaly of the upper limb.

It is an uncommon entity with an incidence of 1 in 30,000 to 100,000 live births. Males are more frequently affected and it is bilateral in 38 to 50 percent of the cases. If unilateral, the right side is affected twice as much as the left side.

The usual deformity that presents to plastic surgeons is that of a radially deviated hand, prominence at the distal end of the ulna and bowing of ulna. The thumb is absent or severely deficient. The hand is typically small and the metacarpophalangeal (MCP) joints have limited flexion with or without some hyper-extensibility.

Bayne and Klug classified this entity into four types and their classification is based on the amount of radius present. Type I is the mildest type characterised by mild radial shortening. Cases with a hypoplastic (or “miniature”) radius are considered type II, partial absence of the radius constitutes type III, and type IV represents cases where there is a complete absence of the radius.

The goals of treatment are to create a centralised wrist whilst maintaining function and avoiding stiffness and recurrence of the deviation. That, however, is seldom accomplished. The wrist is either stiff and centralised or flexible and deviated, a third combination is rare.3,4

Bayne type I deformities rarely require any treatment, whereas type II deformities are usually managed conservatively with stretching and splinting along with distraction osteogenesis if required. It is the type III and IV which require surgical attention and are treated by various techniques, some of which hold only a historic and academic interest.5

RLD is often associated with syndromes like thrombocytopenia-absent radius (TAR), Holt-Oram syndrome, Vertebral anomalies, Anal atresia, Cardiac defects, Tracheoesophageal fistula and/or Esophageal atresia, Renal and Radial anomalies, and Limb defects (VACTERL) association, Craniofacial microsomia, Fanconi anaemia, etc. The importance of diagnosing these syndromes lies in the fact that these associated defects are particularly common as they are present in 40% of patients with unilateral absence and 77% of patients with bilateral absence.

In our study, we focused on the soft tissue release...
component of the correction and used two techniques and compared them with each other in terms of better longevity of the correction and pliability of tissues transposed in the deficient radial side. The current study was planned to assess if the bilobed flap as soft tissue release component for the radial club hand was superior to the z-plasty technique in type III and IV radial club hands.

**Patients and Methods**

This study was conducted at the Shifa International Hospital, Islamabad, Pakistan, from 2009 to 2015, and comprised patients with radial club hands operated by a single consultant plastic surgeon. In all the cases, paediatric review was done to rule out the presence of associated syndromes before the relevant surgical intervention was carried out. Therapy was started with splinting and stretching and centralisation and soft tissue release was performed at 08 months. Pollicisation in Blauth Type IV and V thumbs was done 3 to 6 months after the previous surgery, but before the age of 2 years. The procedures were done under general anaesthesia. Skeletal stabilisation was achieved with a Kirschnerwire (K-wire). The extremity was then casted for 08 weeks and kept in a long arm cast for 3 to 6 months.

**Results**

There were 9 patients with 12 radial club hands in the study. Z-plasty was used on the radial side to correct the soft tissue deficiency (Figure 1a) in 5(42%) hands, and a bilobed flap in 7(58%) hands for soft tissue rearrangement on the radial side (Figure b). Their treatment showed good to excellent results. The male-to-female ratio was 4:1. Of all, there were 7(58.3%) type IV radial club hands, 4(33.3%) type III and 1(8.3%) type II.

Moreover, 1(8.3%) hand developed partial flap necrosis, 1(8.3%) developed infection and implant (K-wire) removal was also encountered in 1(8.3%). No recurrence was encountered during a follow-up of 2.79±0.32 years.

The mean hand-to-forearm angle on 12-month follow-up was 30 to 35 degrees. The ulnar growth was 60 percent of the contralateral normal limb in cases that had unilateral radial club hands over a 3-year follow-up. Hand function remained independent of the wrist stability since it also depends on stiffness of the fingers prior to surgical intervention and other variables like the presence of the thumb.

**Discussion**

It is important here to go through the history and evolution of various techniques and procedures for correction of the radial club hand. Watson et al. used two z-plasties for soft tissue release in their series. The use of bilobed flap for soft tissue release was described by Evans. Sayre described the use of centralisation for radial club hand in 1893. This was later modified by Lidge in 1969.6-8

For the past several decades, centralisation of the carpus on the distal end of the ulna in conjunction with soft tissue release has become the most commonly performed surgical procedure for the treatment of type III and type IV radial club hand.9

In 1985, Buck-Gramco further modified the centralisation procedure and termed it radialisation.10 This was introduced as a means of correcting the ongoing muscle imbalance across the carpus in these cases. In severe wrist deformities, surgery encompassed extensive dissection, manipulation and bony resection. Failed z-plasties, flap necrosis and repeat surgeries aggravated this and resulted in soft tissue contractures. Therefore, Kessler11 introduced soft tissue
distraction prior to any surgical intervention by using an apparatus originally designed to lengthen fingers and the forearm. These distraction techniques were further refined by Smith and Greene.¹² The excellent results following soft tissue distraction led to its inclusion in the management protocols of radial club hand, especially effective for late presenting cases and severe wrist deformities.¹³⁻¹⁷

However, numerous reports of centralisation at long-term follow-up have shown recurrent radial wrist deviation, stiffness and growth disturbances. This has dissuaded surgeons from conventional centralisation procedures.¹⁸⁻²¹

The conventional approach to radial club hands is to perform a z-plasty on the radial side to account for the deficiency of tissue and this also mandates the removal of some tissues from the ulnar side in order to balance the soft tissue. Thus, soft tissue is wasted. There is also more chances of recurrence of deviation in centralisation and soft tissue deficiency corrected with z-plasty. The repetitive failure and dissatisfaction with centralisation and z-plasty has led us to seek an alternative means of treatment for this problem.²²⁻²⁴

The bilobed flap provides a very good alternative to the conventional approach. The excess tissue on the ulnar side is transposed as a bilobed flap on the deficient radial side which maintains the tissue balance on both sides. There is no need to waste the excess tissue on the ulnar side abiding by the principles of plastic surgery.

Although in our series we encountered no recurrence of the deformity and no stiffness, we encountered one partial flap necrosis in one of the z-plasty flaps, whereas all the bilobed flaps healed uneventfully.

Apart from flap viability, the bilobed flap also offers more advantages over the z-plasty, like pliable and good quality tissue transposed on the radial side which maintains long-term stability with no risk of contractures on the radial side. Post-operative physiotherapy can be carried out easily and early in the presence of good quality tissue without the risk of wound dehiscence or flap tip necrosis.

Conclusion
The bilobed flap was found to be superior to z-plasty since no tissue was wasted and the procedure had better predictability.

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References