Symposium

Elbow, Forearm and Wrist Issues in Brachial Plexus Birth Palsy: Current Concepts

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Abstract
The variable presentation of the sequelae of brachial plexus birth palsy (BPBP) at the elbow, forearm and wrist and their association with much extensive brachial plexus involvement makes reconstruction at these levels demanding. Functional limitation and cosmetic concern are common indications for surgical intervention. This article presents a synopsis of the incidence, pathogenesis, clinical presentation and parental concerns related to these deformities, decision-making considerations, management strategies and expected outcome for correction of these deformities. Deformities at the forearm and wrist can be often corrected simultaneously as they could be interrelated. The pattern of deformities, their severity and their impact on the overall function of the limb and parental concern differ. Each child needs a tailor-made management plan, weighing the expected outcome against parental expectation.

Keywords: Brachial plexus birth palsy; Forearm deformity; Supination deformity; Elbow flexion deformity; Pronation deformity; Ulnar deviation deformity.

Introduction
The elbow, forearm and wrist issues are much less discussed compared to the indications for nerve surgery and shoulder deformities. However, their treatment is more challenging because they tend to occur with extensive brachial plexus involvement characterised by widespread weakness, multiple level deformities and lesser function of limb [1, 2].
The deformities at these levels are generally tackled once the shoulder deformities have been corrected, as the proximal deformity can have an

influence on the distal ones [1, 3].

1. Elbow Issues in BPBP:

At the elbow, flexion deformity or absence of flexion can occur.

a. Flexion deformity at the Elbow:

i. Incidence, Clinical presentation, and Parental Concern:

Flexion deformity of >10 degree was noted in 90 of the 113 consecutive children with BPBP visiting the outpatient department of one of the authors (PB) institute. In 48 children, the deformity was >30 degrees and of concern to parents [4].

ii. Pathogenesis:

The factors causing the deformity are muscle imbalance i.e., strong elbow flexors vs weak extensors, contracture of the biceps due to partial re-innervation or a combination of both [5-8]. It can occur as an adaptation to an abduction contracture at the shoulder or in an attempt to stabilise the shoulder by activating the long head of biceps [9, 10].

Elbow flexion deformity is graded as mild: less than 30 degrees, moderate: 30 to 60 degrees and severe: greater than 60 degrees. The median age of onset of deformity was 5.1 years and prevalence of contracture increased with age. An associated radial head dislocation was present in 6% of cases [8]. No significant association was found with the extent of brachial plexus involvement, but others have noted increased prevalence and severity of elbow flexion deformity in Narakas groups II and III as compared to group I [7].

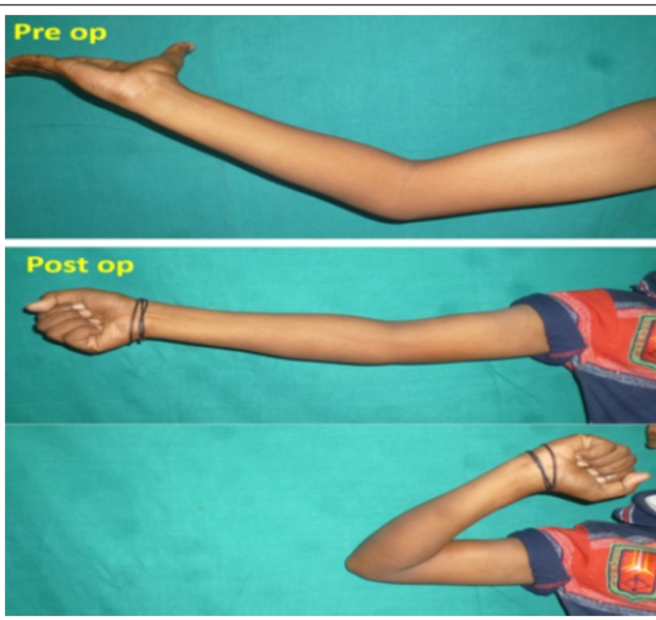

Figure 1: A child with 45 degrees of elbow flexion deformity was managed by three session of stretching and serial casting at weekly interval to attain full correction. He maintained full flexion at the elbow. Through stretching exercises and night splinting, the correction was maintained at two-year follow-up.

iii. Management:

Mild deformities of less than 30 degrees may not hinder activities of daily living, however moderate to severe deformity warrant treatment. Correction of internal rotation deformity in the proximal shoulder joint may improve the elbow flexion deformity [9].

Mild and moderate deformities (Fig. 1) can be treated by serial casting followed by night splinting and respond well [11].

A severe deformity may be treated initially with serial casting and night splinting with surgical release indicated if not responsive to the same. In resistant cases, open release of the joint and biceps tendon lengthening is an option [12] but should be balanced against the risk of weakening elbow flexion. Arthrodiastasis by gradual distraction with hinged external fixator has been described in deformities greater than 40 degrees [13].

b. Absent Elbow Flexion:

Recovery of elbow flexion does occur even with global plexus involvement. If recovery does not occur spontaneously, restoration of elbow flexion is a priority. Nerve surgery in the form of grafting or nerve transfer is most effective for restoring elbow flexion up to 2 years of age. In older children, free functioning muscle transfer, latissimus dorsi bipolar transfer, Steindler's flexorplasty, pectoralis major transfer and triceps to biceps transfer can be considered [1, 2] based on availability of a strong donor, existing function of the hand and availability of the microsurgical expertise.

2. Forearm Deformities in BPBP:

a. Supination Deformity:

i. Incidence, Clinical presentation, and Parental Concern:

Incidence of supination deformity increases with Narakas group. In a series of 750 operated BPBP patients, secondary surgical intervention was required to correct supination

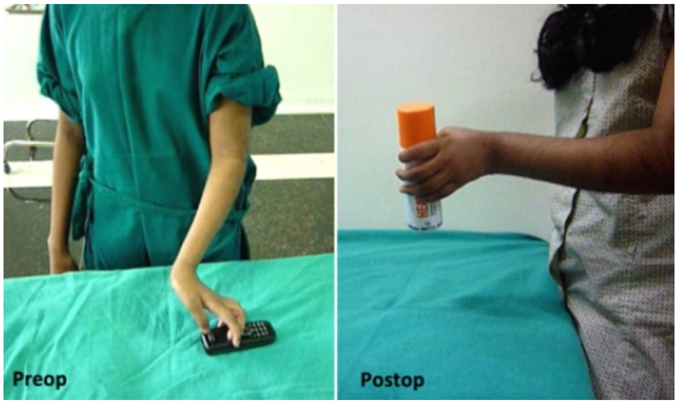

Figure 2: A child with forearm supination deformity has difficulty in tabletop activities as the hand cannot approach on an object well enough to grasp it, as seen in this 14-year-old girl. Also, the normal compensation for the lack of pronation by the shoulder is lacking because of the co-existing shoulder deformity. Mere placement of the hand in pronated position by derotation osteotomy of radius allowed her to use the existing power in the hand much more effectively.

Figure 3: A severe supination deformity worsens the hand function just by virtue of the position of the thumb in adduction (open arrow) and wrist in hyperextension (arrow). Even a reasonably good hand is rendered useless by this combination of postures.

deformity in about 8.8% patients [15]. In the series of Yam et al, it was absent in Narakas group I, it occurred in 5.7% of group II, 9.6 % of group III and 23.4% of group IV [16]. Zancoli and colleagues observed supination deformity in 69% patients with distal paralysis [17].

Supination deformity places the hand in a poor functional position described as the "beggar's hand" or "unshakeable hand" in the literature [18]. A supinated posture precludes effective use of the hand for table-top activities particularly in patients who are unable to use their shoulder to compensate for lack of pronation (Fig 2). Supination deformity makes the simple activities like, dressing, writing, eating, and holding a bicycle handle laborious. Pronation of -10° to +40° is required to carry out common self-care tasks. Extreme supination associated with wrist hyperextension leads to a greater functional loss than what the existing power in the hand would dictate (Fig 3). Hence, mere placement of the hand in a better position by surgery improves function.

ii. Pathogenesis:

Imbalance between a stronger supinator-biceps group and weak flexor-pronator group leads to supination deformity. A fixed deformity results from a secondary contracture of the interosseous membrane [17]. In severe cases, it produces curvature in the radius and volar subluxation of the distal ulna or volar dislocation of the radial head. There are two subsets of patients with BPBP who develop supination deformity [19]. Extended Erb's palsy with forearm muscular imbalance. And more frequently, global palsy patients, who also have wrist hyperextension and ulnar deviation. This is due to weak volar flexors and partial activity of extensor carpi ulnaris.

iii. Management:

Preventive Measures: The deformity usually becomes fixed by 2 years of age. Passive stretching and manipulation cannot prevent fixed deformity. Early identification and surgical treatment prevents bony changes and improves digital function [17, 19]. In patients undergoing shoulder rebalancing surgery with weak forearm pronation (C5-7 cohort), shoulder spica should be applied with the forearm in mid-prone position.

Surgical management depends on multiple factors such as type of deformity (flexible or fixed), triceps strength and congruency of the radio-ulnar joints [1]. Severe shoulder deformities should be corrected first. This unmasks the true supination deformity in the subset of patients with co-existing arm internal rotation and forearm supination (ARMS variant) [3]. Mild-to-moderate elbow flexion deformity is not a contraindication for forearm surgery. However, presence of active wrist extension power of grade 3 or more is a must when considering correction of supination deformity [1]. Weakness of the wrist

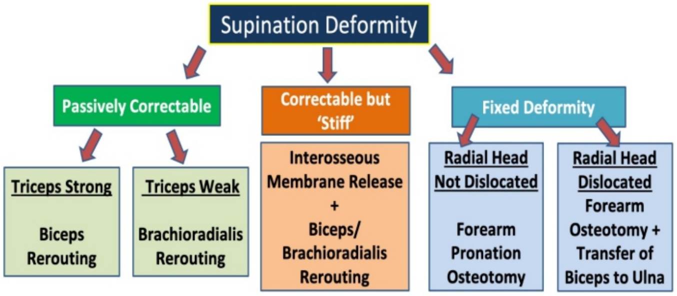

Figure 4: Authors' preferred management algorithm of the management of supination deformity in children with BPBP.

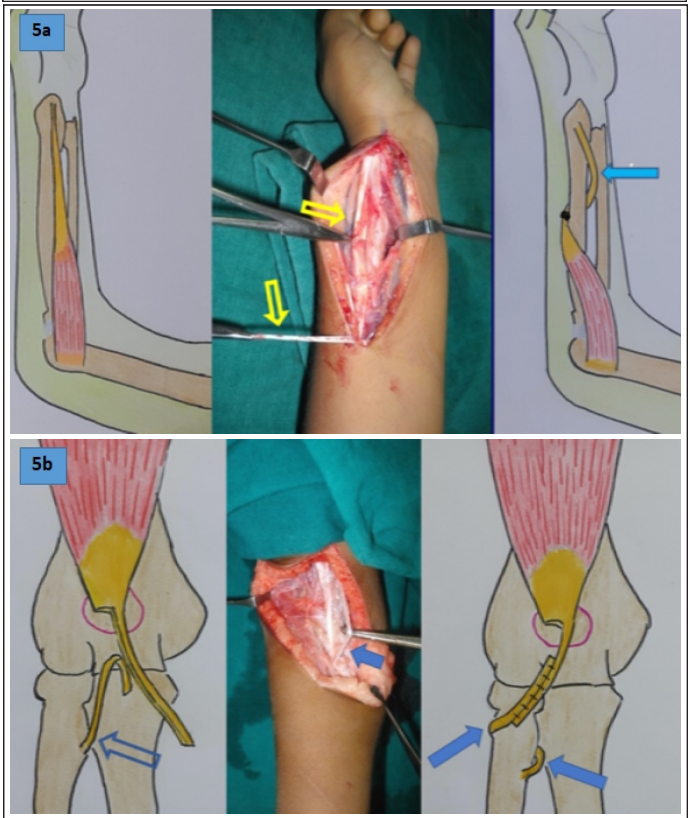

Figure 5: Tendon transfer options for restoration of forearm pronation: (A) Brachioradialis Rerouting: The BR tendon is divided in a Z-manner over a length of 6-8 cm to get two long slips (open arrows). The distal slip attached to the original insertion site is rerouted around the ulnar side of the radius through the interosseous membrane to the dorsum (arrow) and then brought volar and sutured to its proximal Z limb while keeping the forearm in full pronation. By this route now BR provides a pronation torque. (B) Biceps Rerouting: The biceps tendon is divided in a Z-fashion (open arrow and small arrow). The slip attached to the radial tuberosity is rerouted around the radius from anterior to posterior to bring out on the radial aspect of the radius (arrows) and sutured to it proximal z-limb while keeping the forearm in full pronation and elbow 90 degree flexed. This route converts biceps from a supinator to a pronator.

dorsiflexion should be addressed in the same sitting or as the staged procedure [18]. If wrist extension power cannot be restored due to paucity of donors, pronation osteotomy can be combined with wrist fusion. In a child with insensate hand, it is better to leave the supination deformity untreated [20]. An insensate palm is better helped by visual gnosis which is hampered if the palm is placed in pronation. Figure 4 outlines the authors' preferred treatment plan for management of supination deformity. Figure 5 details the commonly employed tendon transfers for supination deformity of the forearm.

Biceps Re-routing:

Biceps re-routing is indicated in flexible supination deformity with active triceps. When triceps is weak, biceps re-routing can increase the elbow flexion deformity. Biceps strength should be grade 4 or greater to provide active pronation without jeopardizing elbow flexion. A Z-incision is placed in the front of elbow with horizontal limb along the elbow crease. Biceps tendon is exposed through its full course. Distal limb of Z-plasty is kept attached at insertion and rerouted around radius through the interosseous membrane to give pronation force. It is then sutured with the proximal limb (Fig 5 A). Mid to long term outcome studies have shown maintenance of active pronation with improved wrist and hand function due to better positioning [15]. Recurrence after biceps rerouting is uncommon as it corrects the primary muscle imbalance. Some degree of elbow extension loss is expected from progressive biceps contracture and not due to the procedure.

Inter-osseous membrane release:

Zancolli & colleagues considered inter-osseous membrane retraction as one of the important factors in developing supination deformity. For a partially fixed deformity, they recommend release of interosseous membrane through a dorsal approach followed by biceps rerouting [17, 19].

Forearm Osteotomy:

Forearm osteotomies are indicated for fixed supination deformities. Given the paucity of donors for restoration of active pronation in these children and the deformity being stiff at presentation, derotation osteotomy is the commonest operation performed for these children (Fig. 6). Isolated radial osteotomy is adequate for moderate supination deformity but both radius and ulna osteotomy

Figure 6: Forearm osteotomy for correction of supination deformity: (A) Supination deformity in this 8-years girl was corrected by pronation osteotomy of the radius to get a much better aesthetic posture to the limb. (B) The improved position of the hand also dramatically improved her function.

Figure 7: Picture showing the disability with the pronation deformity of the forearm in this six-year boy who in an attempt to reach the mouth as for eating has to bend the trunk to compensate but in vain. Shoulder is unable to compensate for even the terminal half of the supination because of the obstruction by the thorax.

is needed for severe deformities [15, 21]. The forearm is stabilised in 30 degrees of pronation. In younger children, this angle of pronation can be slightly higher as remodelling occurs over time. Gilbert et. al. found recurrence of deformity in almost 20% patients after isolated radial osteotomy at an average of 64 months follow up [15]. Manske et. al. combined biceps re-routing with forearm osteotomy and found no recurrence of supination at longer follow up [22]. Lipskier and Weizenbluth preferred double osteotomy over single bone osteotomy to avoid recurrence [23].

Other Uncommon Procedures:
Ozkan reported good results of brachioradialis rerouting with interosseous membrane release ('Pronationoplasty') in four patients with flexible supination deformity and weak biceps (Fig. 5B) [24]. Zancolli preferred resection of distal ulna and distal metaphyseal radio-ulnar fusion in patients with fixed deformities with volar dislocation of distal ulna [17, 19].

Anterior Radial Head dislocation:
When the radial head is reducible, capsuloplasty is recommended but redislocation due to anterior pull of biceps tendon is common. With a dislocated radial head, derotation osteotomy can be combined with transfer of biceps tendon to the ulna or to the brachialis tendon [15, 17, 19].

b. Pronation Deformity:

i. Incidence, Clinical Presentation and Parental Concern:
Pronation contracture or limited supination accounts for 28% of cases with forearm sequelae in BPBP [25]. Self-care, keyboard and tabletop activities require the hand in pronation and since the deformity is rarely extreme due to, the available supination range is sufficient to maintain satisfactory overall function. However, activities needing full supination such as eating with the right hand and accepting offerings in the temple offerings are difficult and result in an awkward posture as the compensation for terminal supination by the shoulder is limited by the thorax (Fig. 7). Most of these children if presenting late have already cortically adapted the dominant use of the opposite upper limb in unilateral cases. Surgeries aimed at improving supination must preserve at least 40 degrees of pronation.

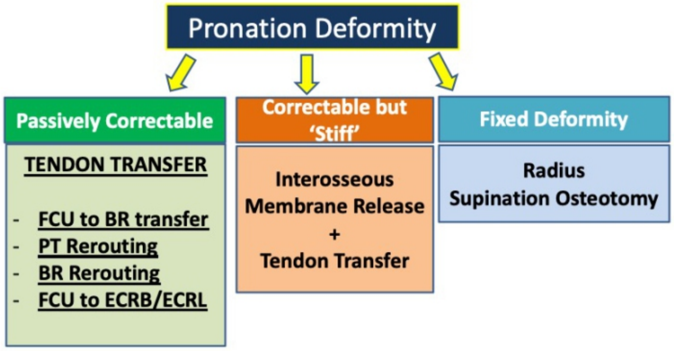

Figure 8: Authors' preferred management algorithm of the management of pronation deformity in children with BPBP.

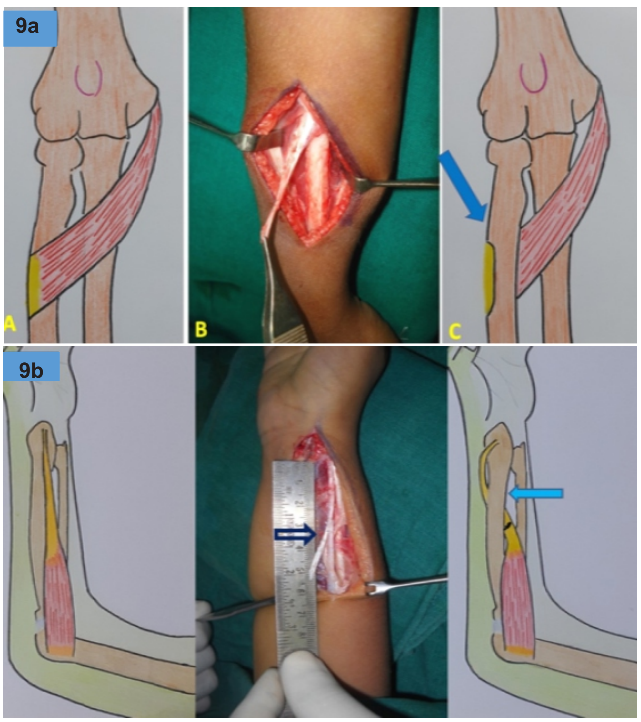

Figure 9: Techniques for achieving supination: (A) Pronator teres Rerouting: Pronator teres is elevated from its insertion with a periosteal extension and is rerouted dorsally along the ulnar border of the radius to reach radially and is inserted over the anterior aspect of the radius at the level of its original insertion site. The PT now works as a supinator instead of a pronator. (B) Brachioradialis Rerouting: The BR tendon is divided in a Z-manner over a length of 6-8 cm to get two long slips (open arrow). The distal slip attached to the original insertion site is rerouted radial to the radius and then around it dorsally and brought through the interosseous membrane volarly (arrow) and sutured to its proximal Z limb while keeping the forearm in full supination. By this route now BR provides a supination torque. Note the route in opposite direction, as in Fig 5A, causes pronation.

ii. Pathogenesis:

Pronation deformity is caused by the weakness of supinator i.e., C7 lesions. Where biceps recovery is partial, the child uses flexor-pronator synergy to assist elbow flexion. This deformity is often accompanied with contracture of the interosseous membrane and radio-ulnar joint dislocation.

iii. Management:

Passive stretching to full supination is the first line treatment but in the setting of muscle imbalance it is rarely effective. Lack of external rotation contributes to limited upturning of the hand; it is imperative to correct the shoulder internal rotation deformity before considering correction of the forearm. Authors' preferred treatment plan for correcting the pronation deformity is presented in figure 8.

Common procedures to restore supination include:

<u>Flexor carpi ulnaris (FCU) to brachioradialis transfer:</u>

The FCU is divided distally and transferred subcutaneously along the ulnar border to reach the dorsum and is attached to a distally-based slip of brachioradialis (BR) in order to make up for the length (Fig. 10 A) [26].

<u>Pronator Teres (PT) Rerouting/Brachioradialis rerouting:</u>

Similar to its use in cerebral palsy, these operations have been extrapolated to birth palsy with satisfactory results provided the donor muscle is of good strength and the deformity is supple [27, 28]. An FCU to extensor carpi radialis brevis transfer may add substantial supination power (Fig. 10A and 10B) [29].

Tendon transfers provide good results in cases with supple and passively correctable deformity. The choice of transfer would largely depend on the status of the donors. If flexor carpi ulnaris and radialis are functioning well and not needed for any other reconstruction, transfer of FCU to BR is our first choice. If FCU is not available for transfer, we consider BR or PT rerouting as per the availability, and in mild cases release interosseous membrane simultaneously with tendon transfer.

A forearm derotation osteotomy is required in fixed deformities. The desired correction depends on the severity of the deformity and the aim of surgery. This procedure can be done even in the presence of a fixed flexion deformity of the elbow and enables the individual to use the extremity as an assistive hand.

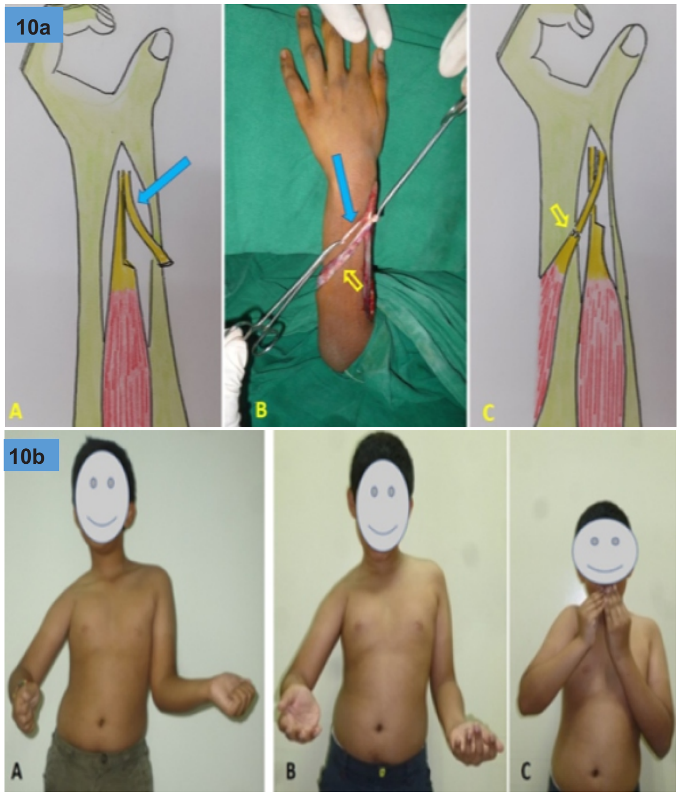

Figure 10: Technique and outcome of FCU to Split BR transfer to achieve supination:
A- Flexor carpi ulnaris is divided distally and mobilized till proximal third of the forearm. It is routed dorsally along the ulnar border of the forearm subcutaneously (open arrow) and sutured to the distally based half-slip of the BR while holding the forearm in full supination.
B- FCU to split BR transfer and interosseous membrane release was performed for this 8-year boy who had active supination possible only till neutral and passive supination of 45 degrees preoperatively (A). Post operatively (B & C) he could achieve 70 degrees of active supination while retaining 60 degrees of active pronation and was comfortably able to reach his mouth; and more interestingly, he could now toss the tennis ball for serving! - one of his desires before surgery.

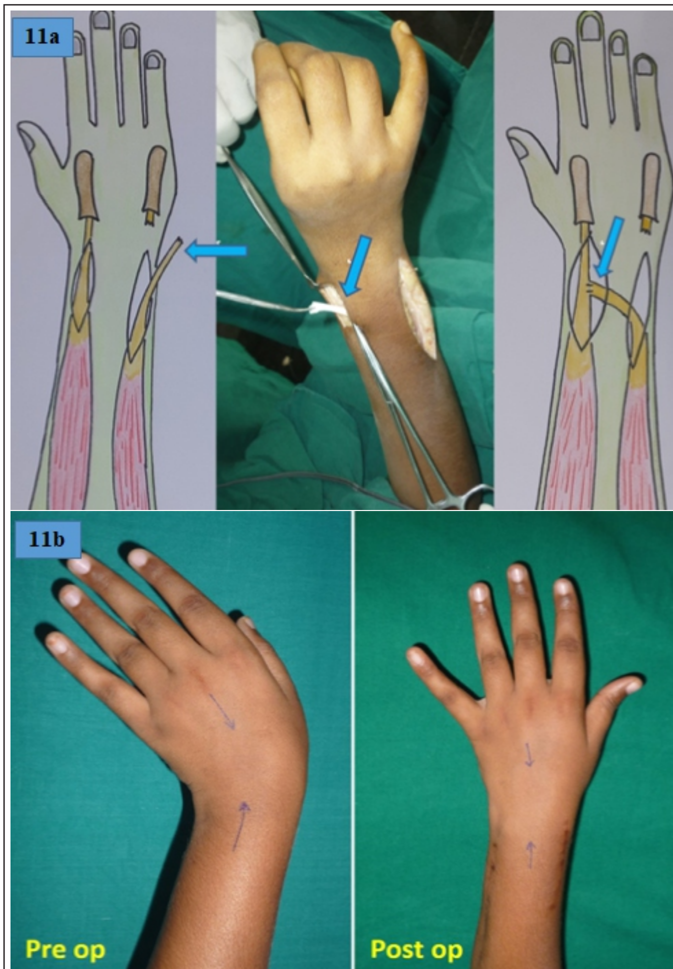

Figure 11: Technique and outcome of ECU to ECRL transfer to correct the ulnar deviation deformity at the wrist:
A- ECU tendon is divided distally and mobilized proximally in order to reach the ECRL in straight line. It is tunneled subcutaneously and sutured to the ECRL while holding the forearm in 20-30 degrees of radial deviation.
B- Comparative pre and post-operative photos showing a 35 degrees of ulnar deviation deformity preoperatively which got effectively corrected by the ECU to ECRL transfer.

3.Wrist Issues in BPBP:

Weakness of wrist extension and ulnar deviation deformity require consideration. The former is generally associated with poor hand function and the latter is often associated with forearm deformities. Hence, the treatment plan needs to be individualised. The general principles influencing the decision making are discussed below.

a. Ulnar Deviation Deformity:

I. Clinical Presentation and Pathogenesis:

Ulnar deviation deformity can be functionally hindering and socially awkward2. They are commonly seen in Narakas group 2, 3 & 4 [30, 31]. The deformity is the result of weakness of abductor pollicis longus (APL), extensor carpi radialis longus (ECRL) and brevis, and flexor carpi radialis (FCR) powered by the C7 and C8 against the better recovered extensor carpi ulnaris (ECU) and FCU innervated by C8 and T1. ECU appears to be the main deforming force [32]. Ulnar deviation deformity can be isolated or associated with a supination deformity [31]; the treatment considerations differ in the two scenarios.

ii. Management:

Better outcomes are seen in children younger than 10 years. Careful clinical examination will help differentiate between ECU or FCU as the cause of the ulnar deviation [31, 32]. Bhardwaj et al reported good results with ECU to ECRL transfer in select cases of wrist ulnar deviation deformity (Fig. 11) [32].

When associated with supination deformity, its correction should be combined with the supination deformity correction using the principles outlined above for correction of these deformities. Özkan et al suggested the "switch transfer" to correct supination and ulnar deviation, which consists of ECU to BR and BR to APL transfer. Passive supination is a prerequisite and pre-operative evaluation of BR and ECU is important. A strong BR is useful to correct the ulnar deviation [33]. In fixed supination deformity, the tendon transfer for correction of ulnar deviation can be combined with a forearm pronation osteotomy.

b. Weakness of wrist extension:

Paralysis of wrist extensors form part of the extensive weakness noted in the children with extended upper or global brachial plexus involvement [31, 34]. A paralysed and 'dropped' wrist severely impairs the ability to grasp. Surgery is generally performed after the age of 4-years to enable better compliance with rehabilitation. A wrist splint can be provided in the interim. The donor tendon should be at least of grade 4 power limiting choice of donors. Conventional donor options include PT, BR, FCU, FCR, flexor digitorum superficialis (FDS) [1]. Bertelli described brachialis tendon transfer to ECRL to stabilize the wrist, but the co-contraction between biceps and triceps must be ruled out else there could be loss of elbow flexion [35]. Duclos and Gilbert reported their experience with tendon transfer for wrist extension in 50 children with BPBP and found that in cases with C5, 6, 7 palsy, PT or FCU was effective but in cases of total brachial plexus palsy FCU was a better choice [34]. Tenodesis procedures to stabilize the wrist, in absence of strong motor donors, often lead to recurrence of the deformity or loss of the correction. The wrist can be arthrodesed in the adolescent or adult. The potential disadvantage in children is the growth disturbance due to injury to the distal forearm physes. Chondrodesis of the wrist joint is a useful alternative to fusion in a young child with paralytic flail wrist. To avoid the hardware complications in children, Boulahouache et al described chondrodesis using a double-frame suture of vicryl [36]. In extreme situations, these options provide a reasonable shape to the hand and augment bimanual activities.

Summary

- Sequelae of BPBP at the elbow, forearm and wrist are challenging in many aspects.
- The combination and severity of deformities vary, as do patient and parental expectation. Each case demands a tailor-made management plan.
- Shoulder deformities are addressed first as they influence the elbow and forearm.
- At the forearm, supination deformity is more common and requires surgical intervention more often.
- Tendon transfer for wrist extension requires cautious choice of the donor in view of the widespread weakness of potential donors.
- Early intervention, both preventive and corrective, should be instituted early.
- The operative procedures are not technically complex but decision making and choice of procedure is crucial.

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