Current Practice in Neurosciences

Current Practices in Birth Brachial Plexus Palsy

MAY 2024

VOLUME 6, ISSUE 3

Sumit Sinha[1], Vigneswaran V[2], Praveen Bhardwaj[3]

[1]Principal Director, Robotic Neurosurgery and Spine Surgery, Max Hospital Dwarka, New Delhi, [2]Senior Consultant, Department of Plastic Surgery, Hand and Reconstructive Microsurgery, Ganga Medical Center and Hospitals Pvt. Ltd, Coimbatore, [3]Associate Consultant, Department of Plastic Surgery, Hand and Reconstructive Microsurgery, Ganga Medical Center and Hospitals Pvt. Ltd, Coimbatore, India

Introduction

Injury to the brachial plexus at birth can be a great concern to the obstetrician and the parents and was initially termed as 'obstetric brachial plexus palsy'. With evolution in the nomenclature based on the observations by multiple surgeons, it has been called at present as Birth Brachial Plexus Palsy (BBPP). Chuang (1) has classified these injuries as infantile or early BBPP, those seen up to one year of life and late BBPP which are sequelae of secondary contractures and deformities. Growth of the patients and consequent deformities are major determinants of the outcomes and quality of life in these patients. These patients are managed with primary nerve surgeries involving nerve repair/grafting and secondary surgeries which are not performed on the nerves but to correct deformities that are seen in patients with spontaneous recovery of birth brachial plexus palsy (BBPP). These surgeries, which easily outnumber those performed primarily (nerve repair/grafting), include muscle release, muscle/tendon transfer, osteotomies, joint release, and free functioning muscle transfers. Paralysis of the muscles supplied by the C5-T1 spinal nerves results in the varied clinical presentations of BBPP whose incidence ranges between 0.4-5% as per literature and about 5% may be bilateral, with an increased risk with breech presentation. However, there have been various reports of decreased incidence of these injuries in the recent past, probably due to better training of obstetricians to manage shoulder dystocia. (2-6). A majority of them do improve spontaneously, however about 25-30% still require some surgery.

Mechanism of Injury

One of the commonest cited causes for BBPP is the excessive traction applied while attempting natural delivery in a baby who weighs more than 4 kgs. (7,8). Over 50% of these children don't have any risk factors (2). The risk factors are as follows in Table 1. (7-12)

Table 1: Risk factors for BBPP

Maternal	Labour	Newborn
• Age >30	• Shoulder dystocia	• Macrosomia- birth weight >4 kg
• Higher Body Mass Index	• Breech presentation	• Lower head-to-thoracic circumference
• Higher parity	• Protracted active phase	• Higher difference between abdominal and biparietal diameters
• Gestational diabetes	• Induced labour	• Hypotonia
• Previous history of shoulder dystocia	• Epidural anesthesia	• Hypoxia
• Black race	• Operative vaginal delivery	
	• intrauterine maladaptation, excessive fundal pressure which causes the anterior shoulder to impact behind the pubis and bicornuate uterus (11).	

It should also be noted that all the patients with BBPP may not have encountered any trauma at birth. A major determinant, to be considered as pathognomonic in BBPP is fetal distress. When labor gets prolonged, even if a cesarean delivery is performed, the baby in distress develops muscular hypotonia resulting in a flail limb. Even with minimal or a little excessive traction, they can end up with bilateral severe palsy. The Apgar score is extremely important to be recorded in the immediate postdelivery stage to assess fetal distress, especially birth asphyxia. Any deviation from the natural course after the first stage labour should be considered as a warning and the treating obstetrician should consider converting the delivery into a controlled cesarean section. However, cesarean section should not be considered protective against BBPP. It does reduce the incidence but does not eliminate the risk of BBPP.

Classification of BPBP

Al Qattan proposed group 2a- those patients in whom wrist drop recovers within 2 months and 2b- in whom it doesn't recover within 2 months and carries a poor prognosis (14).

Table 2: Narakas classification (13)

Group	Name	Roots injured	Weakness	Outcome
1	Upper Erb's	C5,6	Shoulder abduction, external rotation and elbow flexion	Good spontaneous recovery in 80%
2	Extended Erb's	C5,6,7	Above with wrist drop	Good spontaneous recovery in 60%
3	Total palsy without Horners syndrome	C5-T1	Complete flaccid palsy	Good spontaneous recovery of shoulder and elbow in 50% and a functional hand in many
4	Total palsy with Horners	C5-T1	Complete flaccid paralysis with Horners	Worse outcome in absence of surgery

Morphological classification

Table 3: Seddon, Sunderland and Mackinnon* classification (15)

	Seddon	Sunderland	Description	Recovery
1	Neuropraxia	Type 1	Temporary conduction loss	Full recovery 6-12 wks
2	Axonotemesis	Type 2	Division of axons only	Partial recovery
3	"	Type 3	Division of axons with endoneurium	Partial recovery
4	"	Type 4	Division of axons with endo and perineurium	Partial recovery
5	Neurotemesis	Type 5	Complete division of all elements with epineurium	Needs surgery
6*	Mixed	Type 6	Type 2-4	Mixed recovery

Clinical Presentation and Assessment

This section will include the initial examination of patients with BBPP, assessment of the motor and sensory functions and outline of the primary management of these patients.

At the first presentation, the assessment starts with a detailed birth history and perinatal events should be recorded. Birth weight, mode of delivery, order of pregnancy and history of fetal distress if any should be noted. This is followed by a thorough physical examination of the child in a quiet environment. When the child is seen in the first few weeks of birth, keen observation of all the movements happening in the limb remains the examination. The presence of Horner's syndrome can be identified by observing the face. A detailed history and examination findings are documented, and the visit should not be concluded without explaining the chances of spontaneous recovery in these injuries. The mother is taught about the exercises to be performed at each breastfeeding episode.

The authors follow an algorithm (Figure 1) while evaluating and managing these patients who present early. The treating surgeon should be able to address the following queries by following the algorithm-

- How to confirm the diagnosis?
- Where exactly is the level of injury?
- How long to wait? If and when to explore?
- Nerve grafting or nerve transfer?
- Whether to suture or to use tissue glue?
- How long to wait for the full recovery after surgery?
- When to perform secondary procedures?

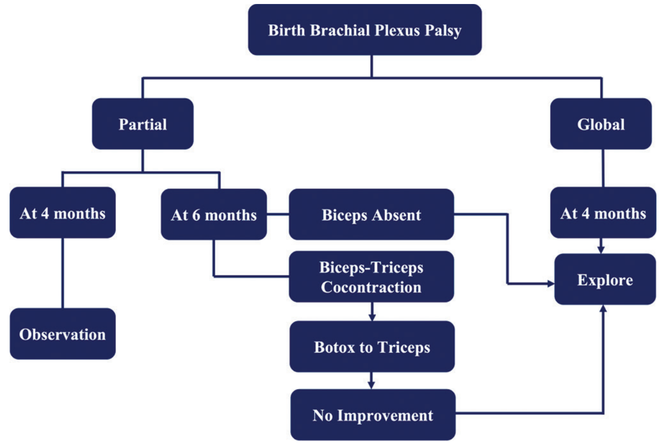

Figure 1: Algorithm for BBPP management

Clinical Examination

Sensory and motor examination is performed. The sensory examination is done by giving mild nociceptive stimuli in specified dermatomes and recording the child's response. Sensory disturbances may be identified by the presence of wounds or ulcers in the fingertips as absolute sensory assessment may pose a challenge in a less co-operative child. These ulcers may result from either lack of sensation or the de-afferentiation pain. A quick guide for sensory examination for distinct dermatomes- : C5- skin over deltoid, C6- Thumb and index fingers, C7- middle finger, C8- Little finger and T1- medial forearm.

Motor examination is performed by examining movements at shoulder, elbow and wrist. When seen at three months, the examination should walk through the plexus as below-

- C5—Shoulder abduction and external rotation (suprascapular and axillary nerves)
- C6—Elbow flexion—biceps (musculocutaneous nerve [MCN]) and brachialis
- C7—Elbow and wrist extension
- C8—Finger and thumb flexion (median nerve)
- T1—Intrinsic muscle activity. Dimpling of dorsal skin at the level of the metacarpophalangeal joints indirectly shows clawing of the fingers (ulnar nerve).

The classical presentation of child with upper plexal injury is the classic waiters tip position, with internal rotation of shoulder, pronation of forearm and flexion of wrist. The pan plexal injuries present with more extensive paralysis of the upper limb depending on the roots and nerves involved.

It is of utmost importance to distinguish between root avulsion and rupture and this has prognostic and therapeutic implications as discussed later. The signs of root avulsion are-

- Horner's syndrome- ptosis, miosis, anhidrosis
- Diaphgramatic injury
- Winging of scapula.

Cookie test- The Cookie test described by Clarke and Curtis (16) can be helpful to identify the presence of elbow flexion recovery in these patients. The test can be conducted at 9 months of age where the baby is offered a cookie and if the baby is able to get it to the mouth, then further waiting is recommended. If the baby fails the test, then exploration is warranted. All authors are unanimous on early exploration in the presence of global palsy as demonstrated by an unrecovered flail hand at 4- 6 months of age. The present authors also recommend exploration of the plexus to a child with flail hand by 6 months and exploration by 9 months in patients with good hand function but absent elbow flexion.

Towel test- Eye cover test or hand to face test- covering the face of child by towel and assessing his ability to reach out to the towel. If he is able to do so, than surgical intervention is not required. (17)

Putti Sign

The elevation of superomedial angle of the scapula on brachio-thoracic abduction is called as putti sign. This is suggestive of abduction contracture of glenohumeral joint.

An assessment of motor and sensory functions can be performed by various methods, the commonly employed being the Active Movement Scale (Table 4). In older children (> 4 years), Mallet score (Figure 2) is useful particularly in assessing the shoulder function.

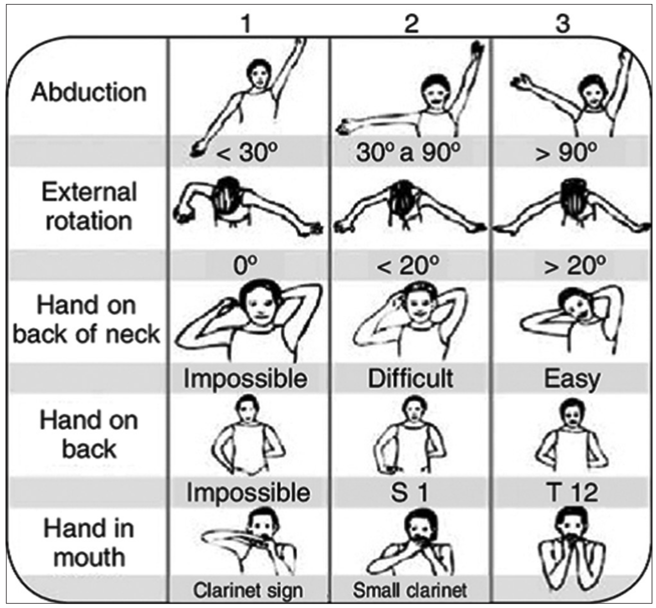

Figure 2: 5-point Mallet Score

1. Gilbert and Tassin- BMRC scale – this scale has limitations that it can't measure partial recovery as it has only one grade. Gilbert suggests early neuroma exploration and plexal reconstruction if there is no elbow flexion by 3 months of age (18,19)
2. Clark and Curtis- developed Hospital for sick children Active movement scale (HSC AMS)- (20) (Table 4)

Table 4: Active movement scale

Observation	Muscle grade
Gravity eliminated	
No Contraction	0
Contraction, no movements	1
Movement <1/2 of range	2
Movement >1/2 of range	3
Full movement	4
Against gravity	
Movement <1/2 of range	5
Movement >1/2 of range	6
Full movement	7

3. Toronto score- derived form AMS and used to predict recovery in BBPP. It evaluated elbow flexion and extension, wrist, finger and thumb extension as per the AMS. Each AMS grade is than converted into a numerical score ranging from 0 (no motion) to 10 (full motion). A score of <3.5 at 3 months of age suggests poor recovery and >3.5 suggests fair recovery (21) (Table 5)

Table 5: Toronto Score

Observation	Muscle grade	Score
Gravity eliminated		
No Contraction	0	0
Contraction, no movements	1	0.3
Movement <1/2 of range	2	0.3
Movement >1/2 of range	3	0.6
Full movement	4	0.6
Against gravity		
Movement <1/2 of range	5	0.6
Movement >1/2 of range	6	1.3
Full movement	7	2

The need for surgical intervention is also suggested by the absence of any improvement in HSC AMS score or Toronto score.

4. Mallet score- This score is used for assessing global shoulder function in children older than 2 years, with grade 0 being no movement in the desired plane and grade 5 being full movement. (22) (Figure 2)

Why Biceps is important landmark in BBPP?

The child with global palsy usually requires surgery with the available roots. The dilemma in kids with Narakas Grade I (Erb's Palsy), because of good chances of spontaneous recovery in this subset of patients. In these patients, the recovery of elbow flexion is considered as the landmark to decide on the need and timing of surgical management. A positive elbow flexion means the entire neural pathway up to the entry of the Musculocutaneous nerve (MCN) into the biceps is intact, which logically means that the other pathways are intact too and therefore spontaneous recovery is likely.

Those infants who recover partial antigravity upper trunk muscle strength in the first 2 months of life should have a full and complete recovery over the first 1 to 2 years of life. Infants who do not recover antigravity biceps strength by 5-6 months of life should have microsurgical reconstruction of the brachial plexus, as successful surgery will result in a better outcome than natural history alone. Infants with partial recovery of C5-C6-C7 antigravity strength during months 3 through 6 of life will have permanent, progressive limitations of motion and strength; they also are at risk for the development of joint contractures in the affected limb. (23,24)

While three months was the 'cut-off' period for intervention in earlier studies, multiple evidence have prolonged this waiting period up to 11 months by which, surgical intervention is warranted if the elbow flexion has not recovered (25).

Investigations in BBPP

The following investigations help in diagnosing the type, location and severity of the injury-

- *Xray of chest to see clavicle*
- *Electrodiagnostic study of upper limb-* provides the following useful information-status of each root commenting on pre or post ganglionic injury, re-innervation and its progress, status of important individual muscles, Compound motor action potential (CMAP) of recipient and donor nerves in cases involving distal nerve transfers and documenting co-contractions.
- *MR Brachial plexus with MR Neurography (3.0 Tesla)-* Magnetic resonance imaging may be helpful at the time of surgery by providing the site of injury- avulsion or rupture of roots. However, it requires the child to be anesthetized and expertise to interpret the findings, the same limitation as with conducting electrodiagnostic studies in these children.

Surgery

There are two types of surgery for BBPP- primary nerve repair and secondary surgery for deformities. The primary nerve repair can be intraplexal repair or distal nerve transfer.

The intraplexal repair is performed between the available proximal and distal stumps by means of intervening nerve grafts harvested from the Sural, Medial Cutaneous Nerve of Forearm and arm (MCNF/A), and the superficial radial nerve (in cases with 5 root rupture), when bilateral Sural plus MCNF/A is insufficient to satisfy the available nerve ends. The distal nerve transfers are used in special circumstances.

Role of distal nerve transfers in BPBP

In BPBP, the surgeon has to choose between the neuroma excision and primary plexal reconstruction or performing the distal nerve transfers initially as the first surgical procedure of choice. In our country, it is very difficult for the patient's family to give consent for neuroma excision and entire plexal reconstruction as the primary procedure, especially in the face of some poor quality shoulder motion or triceps function, for the fear of temporary visible loss of this useless motion. The role of distal nerve transfers is otherwise reserved either as a i) fall back procedure after failed primary intraplexal repair, ii) in patients with late presentation, iii) in cases with dissociated recovery with shoulder recovery without elbow flexion recovery or iv) in avulsion injuries where intraplexal repair is not possible. Typical distal nerve transfers performed are:

1. Oberlin's transfer ulnar to Biceps (26)
2. XIth nerve to Suprascapular Nerve (SSN)
3. Nerve to triceps long head to Axillary Nerve (AXN) described by Somsak from Bangkok (27)

Reconstruction strategies in BPPP

There can be two different kinds of BPBP- i) Ruptures or extra foraminal injuries- which can be repaired by resection and grafting assuming both ends are suitable. ii) Avulsion or intraforaminal injuries- which are truly spinal cord injuries and cannot be repaired. In these cases, reconstruction is accomplished by neurotization wherein nerve fibers from another source are directed into the injured distal plexus. There can be two major varieties of injuries depending on the roots involved- upper plexus injury (C5,6 or C5,6,7) and total palsy. The upper plexal injuries are commonly extra-foraminal and hence reparable mostly. Avulsions arec more commonly seen with central roots- C6,7,8. C8, T1 lesions are the least common in BPBP.

The plexus is exposed under the anterior scalene muscle through a supraclavicular approach under general anesthesia. The nerve trunks and roots are isolated and stimulated with electric current, and neuromatous tissue is excised. The plexus is repaired by intraplexal bridging and/or by extraplexal nerve transfers involving portions of the accessory nerve, intercostal nerves, or uninjured forearm nerves from which functionally expendable motor fascicles can be derived (26). The sural nerves and sometimes the medial cutaneous nerve of forearm, if required, are used as donor sites for nerve transplants.

Recognizing the hand as most important for a useful limb, the order of preference during reconstruction should be (1) hand, (2) elbow flexion, and (3) shoulder.

The Guidelines for repair of specific injuries are included and are as follows. (28,29)

(a) ***C5, C6 injury: ruptures-*** (Figure 3 a) Both proximal root stumps are available for grafting. The strategy is to dissect and transect both the roots as far as possible proximally and distally, achieving a neuroma free distal stump. The distal upper trunk is transected more distally if it is found to have fibrosis and coaptation done between healthy fascicles with food functional organisation. The gap between the healthy roots and the distal normal stump requires 7 supraclavicular grafts, 2–3 cm in length. Typically, C6 to anterior division of upper trunk and C5 to posterior division of upper trunk, is bridged with cable grafts. This repair has an excellent recovery with near normal functional results.

(b) C5, C6 injury, one root ruptured, the other one avulsed- (Figure 3 b, Figure 4a and b) One root is available in this scenario. C5 contains less fibres than C6. Theoretically, prognosis is best if the smaller C5 root is avulsed and if C6 root stump is healthy and normal. C6 is considered to contribute more to biceps than to deltoid and its contrary for C5. The reconstruction involves direct neurotization of distal spinal accessory to suprascapular along with 4 nerve grafts from ruptured proximal root to anterior and posterior divisions of upper trunk. Intercostal nerves may also be used for additional

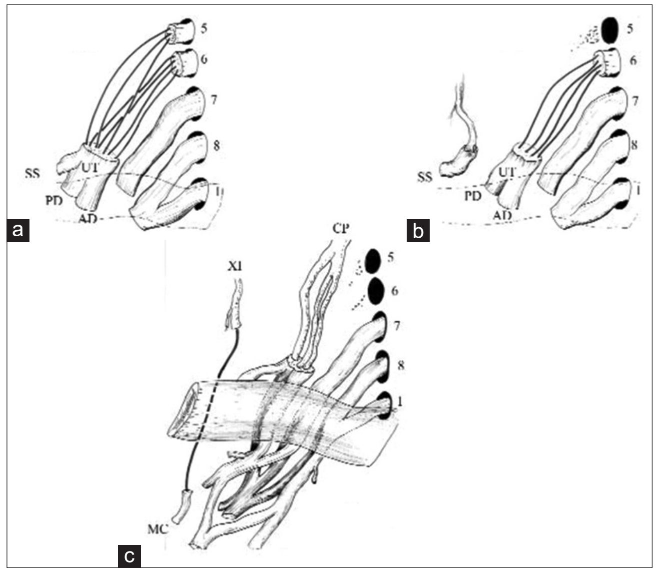

Figure 3: (a) C5, C6 injury: ruptures, b)) C5, C6 injury, one root ruptured, the other one avulsed c) C5, C6 Injury, both roots avulsed d) C5, C6, C7 ruptured e) C5, C6 C7 injury: two roots avulsed, one root ruptured (f) Total plexus injury: C5, C6 rupture, C7, C8, T1 avulsion. SS, suprascapular; PD, posterior division upper trunk; AD, anterior division upper trunk; UT, upper trunk. XI, Spinal accessory; CP, cervical plexus; MC, musculocutaneous nerve

Figure 4: (a) Preoperative, Intraoperative (b) Postoperative photographs of a child with lack of shoulder abduction and elbow flexion at 14 months of age who underwent excision of the neuroma and nerve grafting from C5 root to anterior and posterior divisions of upper trunk with transfer of Spinal accessory nerve to Suprascapular nerve showing the improvement in shoulder abduction and elbow flexion

axillary nerve neurotisation for achieving good shoulder function. This situation expects good results.

(c) C5, C6 Injury, both roots avulsed- (Figure 3c) There are no proximal roots available for plexal reconstruction and neurotization is the only solution. Assuming C7 is perfect and hand is normal, priorities should be elbow flexion and shoulder function in that order. The elbow flexion can be achieved with Oberlin's procedure, which involves ulnar motor fascicle as a donor and biceps branch of the musculocutaneous nerve as the recipient. The shoulder function can be restored by neurotization of spinal accessory to axillary nerve (with a graft), or cervical plexus to upper trunk. Poor shoulder function is expected in this scenario.

(d) C5, C6, C7 ruptured- (Figure 5a) In this situation, as in all upper plexus lesions, hand function is good and the priority is elbow flexion, shoulder function and wrist extension. All the proximal root stumps are available, and distal upper and middle trunks are present with little or no fibrosis after transecting them to appropriate clean levels distally. Multiple grafts, approximately 10 grafts (2–3 cm each) can be used for plexus to plexus repair. The problem is not having enough grafts, but good results are expected if all proximal stumps are healthy and suitable and distal upper and middle trunk show minimal fibrosis supraclavicularly.

(e) ***C5, C6 C7 injury: two roots avulsed, one root ruptured-*** (Figure 5b) This situation demands the neurotization of spinal accessory to suprascapular nerve (extraplexal), and grafts from available root to divisions of upper trunk and to middle trunk. There is a significant dilution of proximal nerve fibers. Distal upper or middle trunk fibrosis calls for a more distal transection, thus longer grafts to anterior and posterior division of upper trunk and to distal middle trunk will be needed. The problem is not having enough proximal fibers. Since elbow flexion is a priority, hence grafts are often directed to the anterior division.

(f) ***Total plexus injury:***

- *4-5 roots available-* this is a very rare situation and simple reconstruction of plexus with intervening cable grafts is all that is required.
- *3 roots available-* one each root is given to medial, lateral and posterior cords. Spinal accessory to suprascapular nerve may be used if required. The best root should be given to the lower trunk or medial cord.
- *2 roots available* - (Figure 6) Root 1 to Lower trunk /medial cord, Root 2 to Lateral cord (or shared between lateral and posterior cord) and XIth to SSN. Some authors

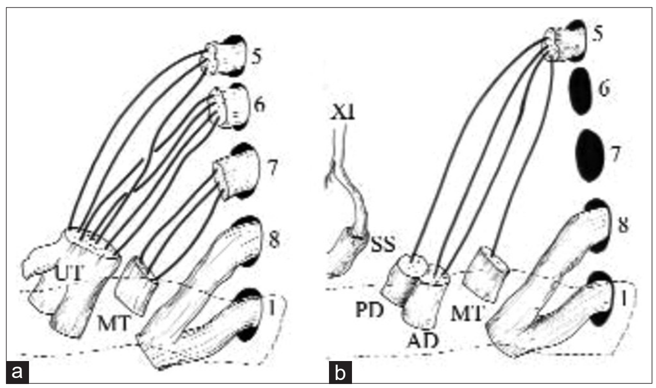

Figure 5: (a) C5, C6, C7 ruptured, all proximal stumps suitable, distal upper and middle trunks with little fibrosis. Multiple grafts used for plexus to plexus repair. (b) C5, C6 C7 injury: two roots avulsed, one root ruptured. Neurotization of spinal accessory to suprascapular (extraplexal), grafts from available root to divisions of upper trunk and to middle trunk- two grafts are directed to the anterior division to illustrate priority for elbow flexion. UT, upper trunk; MT, middle trunk. XI, spinal accessory; SS, suprascapular; PD, posterior division upper trunk; AD, anterior division upper trunk; MT, middle trunk

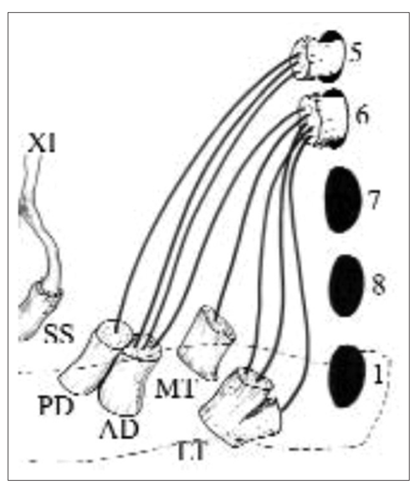

Figure 6: Total plexus injury: C5, C6 rupture, C7, C8, T1 avulsion. In this example two proximal roots allowing 8 grafts are available for reconstructing the entire plexus. Priorities should be hand, elbow flexion and shoulder function. Best proximal sources provide grafts to lower trunk and to anterior division of upper trunk. Neurotization of spinal accessory to suprascapular. XI, Spinal accessory; SS, suprascapular; PD, posterior division, upper trunk; AD, anterior division, upper trunk; MT, middle trunk; LT, lower trunk

would prefer Root 2 to posterior division of upper trunk/posterior cord and 2 or 3 Intercostal nerves (ICN's) to biceps. Their point is getting a strong triceps and a stable shoulder with deltoid and triceps as well as the rotator cuff is important to balance a well- recovered biceps

- *1 root available-* root to lower trunk/ medial cord along with XIth to SSN and 3 ICN to biceps. Secondary transfers will be required for shoulder in future.
- *no root available-* total avulsion is rare. In total plexus injuries, as in all plexus injuries, reconstruction strategy is variable depending on what is available. Graft shortage and not enough proximal source of fibers are serious problems. Results will vary and major deficit will remain. Still these patients are better off than if not treated (spontaneous recovery).Opp. C7 root(Full or Posterior division) to lower trunk/Medial cord and XIth to MCN or XIth to SSN and ICN's to MCN plus one ICN to Long Head of triceps.

Management of Secondary Deformities

These deformities are exclusively associated with BBPP which are low velocity injuries rather than the higher velocity avulsion injuries seen in adults. These injuries are mostly partial including ruptures and in continuity lesions. These injured segments lie in close proximity to each other facilitating spontaneous reinnervation. In such instance, there could be cross innervation of fibres resulting in co-contractions and partial recovery. There exists an imbalance between the recovery of the agonists and the antagonists, which when combined with co-contractions and growth-related disparity result in inadequate functional range of movement. At initial stages, these deformities are passively correctable but have the potential to become fixed contractures later. However, these contractures can be prevented with early institution of physiotherapy associated with high compliance rate which otherwise can lead to deranged functionality and aesthetics. While all the joints can be affected, the deformities most commonly involve the shoulder joint (30,31).

Secondary surgeries in BBPP are indicated in four situations:

- To restore function in situations when the child presents beyond the time at which the primary surgery, that is, nerve repair/grafting or nerve transfer surgery can be offered.
- To augment the function attained by incomplete spontaneous or post-surgery recovery.
- To correct the deformities occurring during the process of spontaneous recovery or after surgery (secondary deformities).
- To restore function after failed nerve surgery.

Deformities of the shoulder

Limitation in abduction and external rotation is the commonest deformity of the shoulder joint in patients with BBPP. The functional disability noted in these patients is directly proportional to the limitation in the working space of the hand. With good hand function, these patients are able to place their hands in various positions in the space with the improvement in the range of shoulder abduction obtained with surgeries. The secondary deformities of the shoulder joint can be classified into three categories for appropriate management as follows:

1. Limited abduction with good external rotation:
   A. Recovered abductors with adductor–abductor co-contractions.
   B. Paralyzed abductors
2. Limited external rotation with good abduction
   A. Limited active external rotation with preserved passive external rotation
   B. Limited active and passive external rotation
3. Limited abduction and external rotation

   These categorical management practices should not be considered as strict guidelines as the presentation can be highly variable with patients not fitting into any of these categories. However, the principles stated are time tested and should be helpful to make a treatment decision.

I- Limited abduction with good external rotation

A: Limited abduction with recovered abductors

These patients have their shoulder abductors recovered but the abduction remain limited secondary to the co-contractions of the shoulder adductors. The limitation therefore could be corrected by releasing the co-contracting adductors, a procedure popularized as the "Modified Quad" procedure by Nath and Paizi (32). The original procedure has undergone various modifications, and the authors perform by releasing the pectoralis major, teres major, latissimus dorsi and decompression of the axillary nerve in the quadrangular space. The released tendon of latissimus dorsi is then sutured to the teres minor in low tension. Shoulder abduction exercises started two weeks later could provide good results and can be maintained over long term (Figure 7).

B: Limited abduction with paralyzed abductors

The deltoid is either poorly recovered or paralyzed in these patients resulting in loss of antigravity shoulder abduction. The adductors may have co-contraction as well. These patients require augmentation of their abduction power with muscle transfers to the deltoid, the donor, most commonly, is the Trapezius. The adductors are released simultaneously as and when required.

II- Limited external rotation with good abduction

A: Limited active external rotation with good passive external rotation

With weak external rotators and persistent internal rotated posture resulting in capsular contracture and restricted passive external rotation, this presentation is uncommon. Preoperatively, external rotation must be assessed by stabilizing the scapula to avoid faulty measurements which is possible because of hypermobile scapula. True external rotation of the glenohumeral joint should end once the scapula moves. Patients with at least 45° passive external rotation can be treated under this category with transfer of the latissimus dorsi to the infraspinatus. Lower trapezius can also be considered as a donor to restore external rotation (33).

B: Limited active and passive external rotation

In this category, the external rotation rarely reaches to the neutral position. As discussed earlier, persistent internal rotation posture and capsular contractures, can result in posterior subluxation of the humeral head, which over time, can cause permanent deformation of the glenoid and the humeral head (34). Aggressive passive external rotation exercises can be instituted in patients presenting early to prevent the secondary changes. However, if the passive external rotation is noted to be lost in the follow-up, it is considered as a red flag and warrants intervention (35,36). These patients are managed according to their age at presentation. At one year of age, these patients can be managed with Botulinum toxin A injection into the shoulder internal rotators. The patients are then immobilized in a cast with shoulder in external rotation for a month followed by passive external rotation exercises. The injection can also be used for the

Figure 7: (a) Preoperative and (b) Postoperative photographs showing the improvement in shoulder abduction in a patient with Category I-A of shoulder deformity. Patient has undergone soft tissue release- the 'Mod Quad' procedure to improve abduction

shoulder adductors simultaneously, if found to be co-contracting. With increasing age, the contracture becomes less amenable for manipulation, therefore requiring surgical intervention. In patients with severe internal rotation contracture, anterior capsular release and subscapularis slide can be helpful (Figure 8). The latter alone can prove to be sufficient if the contracture is mild. In severe cases and when the children are younger (3-5 years) Z-lengthening of the subscapularis is an effective operation. However, one must assure that the muscle in not overly lengthened and just sufficient to maintain the reduction of the shoulder joint lest there is gross limitation of internal rotation at the shoulder. Patients beyond six years of age, where the deformation of the shoulder is severe, classically presents with the 'trumpet' sign. This is characterized by the abducted posture of the shoulder on attempting hand to mouth movement (more annoying if the patient uses the involved hand for eating) and is secondary to lack of external rotation coupled with co-contraction of the shoulder abductors and the elbow flexors (37) (Figure 9). External rotation osteotomy of the humerus can be helpful in managing these patients. Though the procedure does not address the pathology, it does position the limb in a more useful externally rotated position and abolishes the 'trumpet' sign completely (Figure 10).The osteotomy can be carried out at proximal or distal metaphysis

Figure 8: (a) Preoperative and (b) Postoperative photographs showing improvement in shoulder external rotation in a patient of four years of age with Category II-B of shoulder deformity. Patient has undergone anterior shoulder release to improve external rotation

Figure 9: Preoperative photographs of a patient of 12 years of age with limited active and passive external rotation of the shoulder- Category II B. Note the 'trumpet' sign on attempt to reach the mouth

Figure 10: Postoperative photographs of the same patient after external rotation osteotomy of the humerus. Note the complete disappearance of the 'trumpet' sign

humerus which offers better healing potential. The authors recommend performing the osteotomy proximal to the deltoid insertion, thereby creating a lateral abductor moment by the deltoid rather than a forward flexor moment following the osteotomy which transfers the deltoid laterally. The amount of correction required should be assessed preoperatively, ensuring that the patient should be able to reach the midline for buttoning activities, to the mouth and the head. An approximate of 15° external rotation beyond neutral should be an ideal position to be achieved. If preoperative examination shows abduction contracture and elevation of the scapula (as judged by the prominence of the medial border of the scapula on adducting the shoulder—Putti sign)), a 15°–20° varus angulation can be added at the osteotomy site which tends to correct the scapula elevation.

III- Limited abduction and external rotation

Though multiple muscle transfers have been described to address this category with variable outcomes (38,39), the authors propose and use a two-stage procedure to manage the limitation in both abduction and external rotation. In the first stage, the patients are managed as per the first category mentioned previously and at the second stage humerus external rotation osteotomy is done after an interval of one year. External rotation osteotomy provides the advantage of tailoring the expected reduction in internal rotation to avoid any functional limitation.

Deformities of the elbow

The commonest presentation is a fixed flexion deformity of the elbow (40). Though parental concerns may be high to correct the deformity, the functional importance of elbow flexion should be highlighted, and treatment should not result in loss of elbow flexion with multiple elbow release procedures and osteotomies described. Flexion deformity of the elbow in the presence of weak elbow flexors may be amusing but the cause for the deformity is multifactorial as listed below:

1. The spontaneously recovering elbow flexors with their contractile properties deranged could not grow as much as the skeleton or the triceps, resulting in relative shortening (41).
2. These patients with internal rotation contracture at the shoulder tend to keep the elbow flexed to bring it out for functional activities.
3. Presence of co-contractions between the shoulder abductors and the elbow flexors which over time results in the development of flexion contracture of the elbow.

Elbow stretching exercises and elbow extension splint are invaluable to prevent the deformity and should be advocated as early as the patient recovers antigravity elbow flexion power. The splinting should continue as a night splint till the growth is complete to maintain the correction as the altered muscle growth is a causative factor. Patients with deformity greater than 30° are candidates for correction using serial manipulation and casting the elbow in maximally extended position. A minimal of three to four sessions separated by a week is usually required to correct the deformity. The correction achieved should be maintained with an elbow extension splint to avoid recurrence.

Deformities of the forearm

Though a very common occurrence in patients with BBPP, forearm deformities and their management have not gained importance as much as the shoulder deformities (42). The patient can develop both supination and pronation deformity and it is the former which pushes the parents to seek treatment as the posture is unacceptable for them which has been described as the “beggar’s hand” or the “unshakeable hand” (43). Preoperative discussion with the parents is important before correcting these deformities as these are commonly fixed, therefore, improvement in one direction would decrease the opposite motion. Pronated posture of the forearm is more functional for bimanual and table-top activities whereas the supinated posture will be useful for receiving offerings and eating if the patient uses the hand for eating. The supination deformity puts the thumb in an unfavorable position resulting in adduction contracture of the first web, compromising the hand function (44). These functional and aesthetic considerations tilt the balance towards restoring the pronated posture of the forearm.

Restoration of pronation

Presence of active wrist extension is mandatory as once the supination deformity is corrected, the patient with absent wrist extension will develop a wrist drop. The authors use and recommend a protocol (Figure 10) for managing the supination deformity.

Supination deformity- fixed deformity/absence of donors- pronation osteotomy of radius.

Supination deformity- passively correctable- triceps functioning- Biceps rerouting; triceps nonfunctioning- Brachioradialis rerouting.

Triceps' power of more than grade 3 is the prerequisite for doing a biceps' rerouting procedure (45). Patients with weak triceps tend to develop elbow flexion deformity after the surgery and checking the power of the triceps is very important during evaluation.

While performing a pronation osteotomy, the aim should be to achieve 30° of pronation. With radius osteotomy alone, approximately 100° of rotation could be achieved and when the requirement is more, interosseous membrane release and additional ulna osteotomy may be required.

Restoration of supination

Surgeons must be aware to retain or should not decrease the range of pronation present while restoring supination. Though fixed supination deformity is well tolerated in other conditions, patients with BBPP fare poorly because they could not compensate the deformity with shoulder movements which itself is abnormal. When the deformity is passively correctable, the options available are pronator teres rerouting (46) or transfer of flexor carpi ulnaris (FCU) to split Brachioradialis (47). The latter requires a normal flexor carpi radialis for the FCU to be harvested as a donor. It should be ensured that the future restoration of more useful function like wrist or finger extension does not require the wrist flexor as donor. In fixed deformities, supination osteotomy of the radius could be performed but care should be taken to maintain at least 45° of pronation intact to allow tabletop and manual activities.

Deformities of the wrist and hand

The common presentations include ulnar deviation deformity of the wrist, weak or absent wrist and finger extension, lack of wrist flexion, adducted and weak thumb, or poor intrinsic function (48,49). Managing these, require detailed assessment and tailored approach for each individual patient based on their requirements. A major constraint faced in these patients is the paucity of donors as these muscles are also recovered muscles or partly affected, therefore lacking normal function (50,51). However, the wrist and hand function can be improved with tendon transfers with proper patient selection (Figure 11).

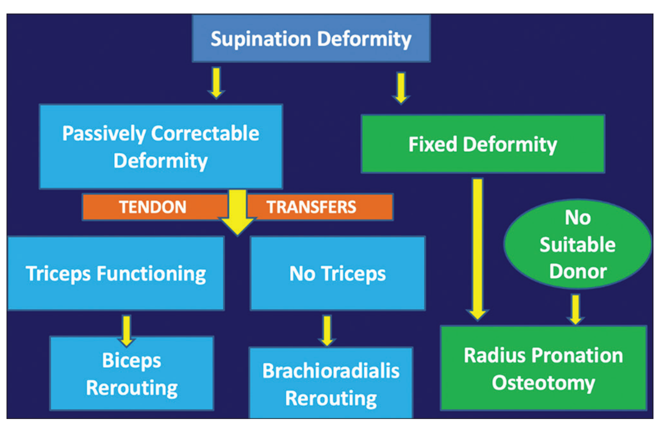

Figure 11: Algorithm for management of supination deformity of the forearm

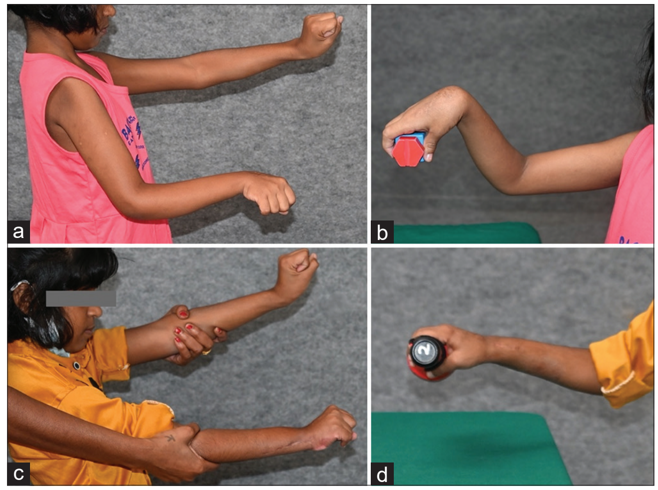

Figure 12: (a and b) Preoperative photographs of a patient with ulnar deviated wrist with weak wrist extension (c and d). The patient underwent Extensor carpi ulnaris to Extensor carpi radialis brevis transfer resulting in aesthetic and functional improvement

Prevention of secondary deformities

All the deformities discussed above can potentially be prevented with regular and proper physiotherapy. The parents should be taught and be provided with a printed exercise protocols or a video recording of the exercises to be carried out. Regular follow-ups with the physiotherapists and the treating surgeons cannot be overemphasized. The surgeons should ensure that the prescribed exercises are carried out properly. It is common to note that the shoulder external rotation exercises being performed in inappropriate method. The correct way to perform abduction exercises is by stabilizing the scapula and then abducting the arm to go and touch the ear. The external rotation exercises are done by flexing the elbow to 90 degrees, tucking the arm to the chest, and then performing external rotation while maintaining the adduction at the shoulder (arm remains touching the lateral chest wall). Normally, there is no scapular movement while performing external rotation, but in these children, there is hypermobility and superior migration of the scapula while doing external rotation. Hence, while doing the external rotation exercise, one hand of the therapist or mother should stabilize the scapula. Once the scapular movement is prevented, true glenohumeral external rotation stretching is possible. Use of night splinting for correction of elbow flexion deformity and continuous use to maintain the correction achieved must also be stressed to the parents.

Summary

Birth brachial plexus injury (BBPP), is unfortunately a rather common injury in newborn. The incidence of BBPP varies between 0.15-3 per 1000 live births in various series in literature. A majority of these patients improve spontaneously, however, a large subset of them don’t recover, requiring either a primary or secondary surgical intervention and up to 35% of children may have some degree of life-long functional impairment of the affected limb. Management of children with BBPP depends upon the degree of nerve injury and involvement. A multidisciplinary team approach incorporating rehabilitation strategies and surgical interventions can help to optimize recovery and prevent long-term impairment. Secondary deformities in BBPP are common and have ability to cause long term disability in such patients (52,53). However, many of these deformities are preventable or at least correctable with timely intervention. Patients and parents must be advocated for regular follow-ups with both the physiotherapists and the treating surgeon. The management protocol should be individualized for each deformity in every patient. Gratifying results with immense satisfaction could be achieved in these patients with appropriate surgical intervention.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Chuang DC, Mardini S, Ma HS. Surgical strategy for infant obstetrical brachial plexus palsy: experiences at Chang Gung Memorial Hospital. Plast Reconstr Surg 2005;116(1):132–142, discussion 143–144.
2. Foad SL, Mehlman CT, Ying J. The epidemiology of neonatal brachial plexus palsy in the United States. J Bone Joint Surg Am. 2008;90(6):1258-1264.
3. Buterbaugh KL, Shah AS. The natural history and management of brachial plexus birth palsy. Curr Rev Musculoskelet Med. 2016;9(4):418-426.
4. Foad SL, Mehlman CT, Foad MB, Lippert WC. Prognosis following neonatal brachial plexus palsy: an evidence-based review. J Child Orthop 2009;3(6):459-463.
5. Hale HB, Bae DS, Waters PM. Current concepts in the management of brachial plexus birth palsy. J Hand Surg Am. 2010;35(2):322-331.
6. Lagerkvist AL, Johansson U, Johansson A, Bager B, Uvebrant P. Obstetric brachial plexus palsy: a prospective, population-based study of incidence, recovery, and residual impairment at 18 months of age. Dev Med Child Neurol. 2010;52(6):529-534.
7. Abzug JM, Mehlman CT, Ying J. Assessment of Current Epidemiology and Risk Factors Surrounding Brachial Plexus Birth Palsy. J Hand Surg Am. 2019;44(6): 515.e1-515.e10.
8. Thatte MR, Mehta R. Obstetric brachial plexus injury. Indian J Plast Surg. 2011;44(3):380-389.
9. Toupchizadeh V, Abdavi Y, Barzegar M, Eftekharsadat B, Tabrizi I. Obstetrical brachial plexus palsy: electrodiagnostical study and functional outcome. Pak J Biol Sci. 2010;13(24):1166-1177.
10. DeFrancesco CJ, Shah DK, Rogers BH, Shah AS. The Epidemiology of Brachial Plexus Birth Palsy in the United States: Declining Incidence and Evolving Risk Factors. J Pediatr Orthop. 2019;39(2): e134-e140.
11. Mollberg M, Hagberg H, Bager B, Lilja H, Ladfors L. Risk factors for obstetric brachial plexus palsy among neonates delivered by vacuum extraction. Obstet Gynecol. 2005;106(5 Pt 1):913-918.
12. Al-Qattan MM, Al-Kharfy TM. Obstetric brachial plexus injury in subsequent deliveries. Ann Plast Surg. 1996;37(5):545-548.
13. Narakas AO (1987) Obstetric brachial plexus injuries. In: Lamb DW (ed) The paralysed hand. Churchill Livingstone, Edinburgh, pp 116–135.
14. Al-Qattan MM, El-Sayed AA, Al-Zahrani AY, *et al*. Narakas classification of obstetric brachial plexus palsy revisited. J Hand Surg Eur Vol. 2009;34(6):788-791.
15. Seddon H.J. A Classification of Nerve Injuries. Br Med J. 1942 Aug 29;2(4260):237-239.
16. Clarke HM, Curtis CG. An approach to obstetrical brachial plexus injuries. Hand Clin. 1995;11(4):563-581.
17. Bertelli JA, Ghizoni MF. The towel test: a useful technique for the clinical and electromyographic evaluation of obstetric brachial plexus palsy. J Hand Surg Br. 2004;29(2):155-158.
18. Gilbert A, Tassin J-L. Obstetrical palsy: a clinical, pathologic, and surgical review. In: Terzis JK, ed. Microreconstruction of nerve injuries. Philadelphia: Saunders, 1987: 529.
19. Gilbert A, Razaboni R, Amar-Khodja S. Indications and results of brachial plexus surgery in obstetrical palsy. Orthop Clin North Am. 1988;19(1):91-105.
20. Curtis C, Stephens D, Clarke HM, Andrews D. The active movement scale: an evaluative tool for infants with obstetrical brachial plexus palsy. J Hand Surg Am. 2002;27(3):470-478.
21. Michelow BJ, Clarke HM, Curtis CG, Zuker RM, Seifu Y, Andrews DF. The natural history of obstetrical brachial plexus palsy. Plast Reconstr.
22. 33. 28.Abzug JM, Kozin SH. Evaluation and management of brachial plexus birth palsy. Orthop Clin North Am. 2014;45(2):225-232.
23. Tassin JL. Paralysies obstetricales du plexus brachial : evolution spontanee, resultats des interventions reparatrices precoces. Thesis. Paris, Universite Paris VII, 1983.
24. Gilbert A, Tassin JL. Reparation chirurgicale du plexus brachial dans la paralysie bstetricale. Chirurgie. 1984;110:70–75.
25. Waters PM, 2005 Update on Management of Pediatric Brachial Plexus Palsy J Pediatr Orthop 2005;25:116– 126.
26. Oberlin C, Beal D, Leechavengvongs S, Salon A, Dauge MC, Sarcy JJ. Nerve transfer to biceps muscle using a part of ulnar nerve for C5/C6 avulsion of the brachial plexus. Anatomical study and report of 4 cases. J Hand Surg. 1994;19:232–237.
27. Leechavengvongs S, Witoonchart K, Uerpairojkit C, Thuvasethakul P. Nerve transfer to deltoid muscle using the nerve to the long head of the triceps, part II: a report of 7 cases. J Hand Surg Am. 2003 Jul;28(4):633- 638.

28. MR Thatte, A Hiremath, N Nayak, N Patel. Obstetric brachial plexus birth palsy. Diagnosis and Management. Journal of Peripheral Nerve Surgery (Volume 1, No.1, July 2017) 2-9.
29. Jose L Borrero. Surgical technique. Brachial Plexus Injuries: Published in Association with the Federation Societies for Surgery of the Hand. (2001). United States: CRC Press. Alain Gilbert (ed). 2001 Martin Dunitz Ltd. Pp 189-205.
30. Waters PM. Update on management of pediatric brachial plexus palsy. J Pediatr Orthop 2005;25(1):116–126AQ10 32.
31. Yuceturk A. Palliative surgery: tendon transfer to the shoulder in children. In: Gilbert A, ed. Brachial Plexus Injuries. London, UK: Martin Dunitz; 2001:239–24.
32. Nath RK, Paizi M. Improvement in abduction of the shoulder after reconstructive soft-tissue procedures in obstetric brachial plexus palsy. J Bone Joint Surg Br 2007;89(5):620–626.
33. Elhassan B. Lower trapezius transfer for shoulder external rotation in patients with paralytic shoulder. J Hand Surg Am 2014;39(3):556–562.
34. Waters PM, Smith GR, Jaramillo D. Glenohumeral deformity secondary to brachial plexus birth palsy. J Bone Joint Surg Am 1998;80(5):668–677.
35. Kozin SH. Correlation between external rotation of the glenohumeral joint and deformity after brachial plexus birth palsy. J Pediatr Orthop 2004;24(2):189–193.
36. Bhardwaj P, Burgess T, Sabapathy SR, Venkataramani H, Ilayaraja V. Correlation between clinical findings and CT scan parameters for shoulder deformities in birth brachial plexus palsy. J Hand Surg Am 2013;38(8):1557–1566.
37. Chuang DC, Ma HS, Wei FC. A new strategy of muscle transposition for treatment of shoulder deformity caused by obstetric brachial plexus palsy. Plast Reconstr Surg 1998; 101(3):686–694.
38. Kirkos JM, Kyrkos MJ, Kapetanos GA, Haritidis JH. Brachial plexus palsy secondary to birth injuries. J Bone Joint Surg Br 2005;87(2):231–235.
39. Ozben H, Atalar AC, Bilsel K, Demirhan M. Transfer of latissmus dorsi and teres major tendons without subscapularis release for the treatment of obstetrical brachial plexus palsy sequela. J Shoulder Elbow Surg 2011;20(8):1265–1274.
40. Sheffler LC, Lattanza L, Hagar Y, Bagley A, James MA. The prevalence, rate of progression, and treatment of elbow flexion contracture in children with brachial plexus birth palsy. J Bone Joint Surg Am 2012;94(5):403–409 58.
41. Stefanova-Uzunova M, Stamatova L, Gatev V. Dynamic properties of partially denervated muscle in children with brachial plexus birth palsy. J Neurol Neurosurg Psychiatry 1981;44(6):497–502.
42. Bhardwaj P, Varadharajan V, Salyan S, Venkatramani H, Sabapathy SR. Forearm Deformities in Birth Brachial Plexus Palsy - Patient Profile and Management Algorithm. J Hand Surg Asian Pac Vol. 2023;28(6):624-633.
43. Al-Qattan MM, Al-Khawashki H. The “beggar’s” hand and the “unshakable” hand in children with total obstetric brachial plexus palsy. Plast Reconstr Surg 2002;109(6):1947–1952.
44. Zancolli EA II. Palliative surgery: pronosupination in obstetric palsy. In: Gilbert A, ed. Brachial Plexus Injuries. London, UK: Martin Dunitz; 2001:275–291.
45. Zancolli EA II. Palliative surgery: pronosupination in obstetric palsy. In: Gilbert A, ed. Brachial Plexus Injuries. London, UK: Martin Dunitz; 2001:275–291.
46. Amrani A, Dendane MA, El Alami ZF. Pronator teres transfer to correct pronation deformity of the forearm after an obstetrical brachial plexus injury. J Bone Joint Surg Br 2009;91(5):616618.
47. Anderson GA, Thomas BP, Pallapati SC. Flexor carpi ulnaris tendon transfer to the split brachioradialis tendon to restore supination in paralytic forearms. J Bone Joint Surg Br 2010; 92(2):230–234.
48. Sebastin SJ, Chung KC. Reconstructive strategy for recovery of hand function. In: Chung KC, Yang LJS, McGillicuddy JE, eds. Practical Management of Pediatric and Adult Brachial Plexus Palsies. New York, NY: Elsevier Saunders; 2012:114–142.
49. Bhardwaj P, Parekh H, Venkatramani H, Raja Sabapathy S. Surgical correction of ulnar deviation deformity of the wrist in patients with birth brachial plexus palsy sequelae. Hand Surg 2015;20(1):161–165.
50. Duclos L, Gilbert A. Restoration of wrist extension by tendon transfer in cases of obstetrical brachial plexus palsy. Ann Chir Main Memb Super 1999;18(1):7–12.
51. Ruchelsman DE, Ramos LE, Price AE, Grossman LA, Valencia H, Grossman JA. Outcome after tendon transfers to restore wrist extension in children with brachial plexus birth injuries. J Pediatr Orthop 2011;31(4):455–457.
52. Squitieri L, Larson BP, Chang KW, Yang LJ, Chung KC. Understanding quality of life and patient expectations among adolescents with neonatal brachial plexus palsy: a qualitative and quantitative pilot study. J Hand Surg Am 2013;38(12): 2387–2397.e2.
53. Partridge C, Edwards S. Obstetric brachial plexus palsy: increasing disability and exacerbation of symptoms with age. Physiother Res Int 2004;9(4):157–163.