Management of Brachial Plexus Birth Injury: The SickKids Experience

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Abstract

This article describes the approach utilized by the multidisciplinary team at Sick Kids Hospital to evaluate and treat patients with brachial plexus birth injury (BPBI). This approach has been informed by more than 30 years of experience treating over 1,800 patients with BPBI and continues to evolve over time. The objective of this article is to provide readers with a practical overview of the Sick Kids approach to the management of infants with BPBI.

Keywords
- brachial plexus
- birth injury
- microsurgery
- glenohumeral dysplasia

In this article, we describe our personal experience in the management of children with brachial plexus birth injury (BPBI). The first formal brachial plexus clinic was established at the Sick Kids Hospital in the 1980s, under the direction of Dr. Howard Clarke and Dr. Ronald Zuker. At the outset, it was established as a multidisciplinary clinic. Soon after its creation, Dr. Howard Clarke assumed the main leadership of this clinic and has gone on to train several generations of pediatric plastic surgeons within Canada and internationally. Our current team of brachial plexus surgeons consists of Dr. Clarke (plastic surgery), Dr. Davidge (plastic surgery), and Dr. Hopyan (orthopaedic surgery).

The global literature on BPBI is very rich, displaying varied but overlapping approaches to the primary and secondary management of infants and children with this condition. We reference key articles in this manuscript, but also focus on certain aspects of the clinic setup and our personal decision-making that are not readily found in other published works. The experience we present here is based on the care of over 1,800 patients with BPBI over the last three decades. Our practice continues to evolve as our understanding of BPBI and its treatment outcomes has matured.

Clinic Structure and Design

This is an often overlooked aspect of BPBI management. Many of our fellows graduate and go on to establish the first brachial plexus clinic at their hospital. The importance of the setup and design of the clinic cannot be overemphasized to ensure long-lasting success.

Personnel and Culture

A multidisciplinary team comprised of properly trained and experienced staff is the foundation of a well-functioning brachial plexus clinic. Our clinic team is comprised of plastic surgeons, orthopaedic surgeons, physiotherapists, occupational therapists, nurses, social workers, psychologists, research support staff, and trainees. We place a high importance on fostering a collegial and supportive culture so as to optimize team dynamics, communication, and ultimately patient care. Ideally, all members of the team are enthusiastic, committed, and engaged in working with children suffering from brachial plexus injuries. Long-term stability of the team is especially beneficial considering that most patients are followed for many years.

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Physical Structure and Layout

Our primary clinical assessments are completed in a large clinic room with sufficient space for families, patient assessment, and involvement of multiple team members and learners. Educational and visual aid materials are available in the clinic room, which are important to facilitate discussions with patients and families around diagnosis and possible interventions.

Clinic Database

Development of a robust and well-curated clinic database is extremely valuable and should be done from the outset. Although implementing and maintaining an accurate database requires time and effort, its impact on the clinic can be significant. Our clinic database has enabled the development of new and innovative tools for patient assessment and evaluation (i.e., Active Movement Scale [AMS], Brachial Plexus Outcome Measurement [BPOM]). Interrogation of the database further drives the evaluation and modification of these tools.

Clinic Visibility/Awareness

To facilitate early patient referrals, general practitioners in the region must be aware of the existence of the clinic. Beyond the pediatric hospital in which the clinic exists, there must be an awareness among referring physicians, nurse practitioners, midwives, and any other health care providers that are seeing infants. An easily accessible central triage and referral process is important to ensure that all patients requiring evaluation are seen in the clinic as early as possible postnatally. Perhaps most importantly, giving the clinic the formal name of "Brachial Plexus Clinic" within the hospital will facilitate this triage process.

Assessment of Infants with BPBI: Indications and Timing of Primary Nerve Surgery

Prior work by our group has demonstrated that a single age or criterion for primary nerve surgery is insufficient to accurately predict who requires operative management.[1,2] Consequently, there are several time points within the first year of life where we may recommend a primary nerve operation. Our algorithm has evolved over time,[3,4] with the most recent iteration demonstrated in ►**Fig. 1**.

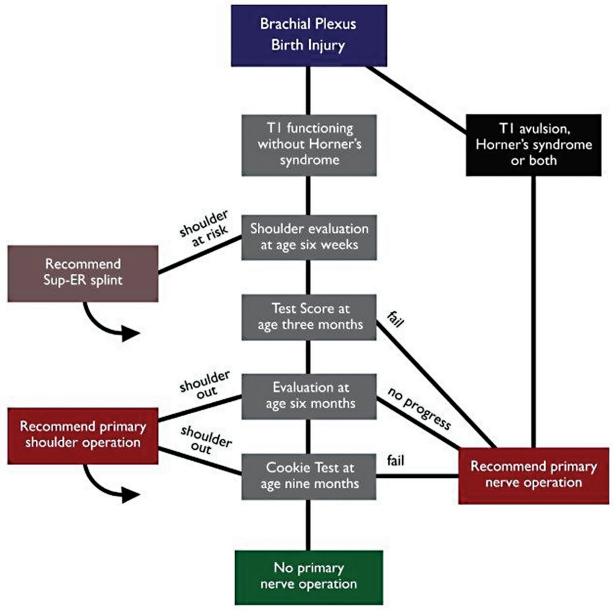

Fig. 1 Sick Kids algorithm for the management of infants with BPBI. This most recent modification includes primary management of the shoulder in infants with glenohumeral dysplasia. (Reproduced with permission from Zuo et al.[32])

Initial Intake Assessment (Prior to 3 Months of Age)

We are fortunate to have a robust referral system, whereby the majority of infants with suspected BPBI are referred within the first week of life. These are triaged to be seen first by the clinic physiotherapist for an initial evaluation at 2 to 3 weeks of age. The physiotherapist follows the infant serially until the first multidisciplinary clinic with the surgeon at 3 months of age.

Early and serial assessments by physiotherapy serve several valuable purposes. First, it enables an experienced team member to screen referrals and conduct an early assessment of function. At the appointment, a full history and comprehensive physical examination are completed. The examination involves assessment of passive range of motion (ROM) and evaluation using the AMS.[5,6] Fracture callus can also be readily palpated at this age. This provides baseline data and information, which is helpful for instructing future clinic appointments.

Based on their assessment, the physiotherapist will independently make three important decisions: (1) whether the child requires dynamic ultrasound imaging of their shoulders at the time of their first full clinic visit; (2) whether the child might benefit from early Sup-ER (supination-external rotation) splinting for their shoulder[7]; and (3) whether the child requires evaluation by the full team prior to 3 months of age—in the case of a completely flail upper extremity or presence of Horner's syndrome.[8]

Additionally, at this first meeting, stretching and ROM exercises are instituted to maintain joint motion. Families are specifically instructed on how to complete the exercises at home. For upper plexus injuries, exercises focus on shoulder, elbow, and forearm movement. For total plexus injuries, the wrist and hand are also included. The exercises are reviewed at subsequent visits with physiotherapy prior to the first full clinic visit.

Perhaps, equally important is the opportunity for the family to meet a member of the team in a nonthreatening environment and have their questions and concerns addressed. An early point of contact is extremely helpful to provide education to the families and explain the process of evaluation. Consequently, when the family comes for their first full clinic visit, they have a basic understanding of BPBI and the typical protocols. Being well informed helps to alleviate stress and anxiety surrounding their child's care. If there are concerns regarding social support, financial stress, and/or family dynamics, the clinic social worker can be engaged at this time as well.

First Multidisciplinary Clinic Appointment (3 Months of Age)

Most new infant referrals are seen for their first full multidisciplinary clinic appointment at 3 months of age. Patients with severely affected extremities (flail limb, Horner's syndrome) are a unique group and are seen as early as possible to discuss surgical intervention.

History and Physical Examination

A standardized form is used to complete the history and ensure that all pertinent features are recorded (►**Fig. 2**). The physical examination is completed by an experienced physiotherapist and focuses on determining AMS scores and assessing passive ROM. AMS scores are obtained for 14 domains of upper extremity movement: shoulder abduction/flexion/internal rotation/external rotation, elbow flexion/extension, forearm supination/pronation, wrist flexion/extension, finger flexion/extension, and thumb flexion/extension. Each movement is assigned a score from 0 to 7 based on the AMS system[5,6] (►**Table 1**). The surgeon evaluates the glenohumeral joint for any evidence of posterior subluxation.

The physical examination also includes screening for coexisting torticollis, positional plagiocephaly, Horner's syndrome, general muscle tone (increased or decreased), and general development.

Indications for Primary Nerve Surgery at or before 3 Months of Age

Failed Test Score

The test score is calculated by converting the AMS scores at 3 months of age for elbow flexion, elbow extension, wrist extension, finger extension, and thumb extension (►**Table 2**). The converted scores are combined to obtain the final test score, out of 10.[3,5] Test scores less than 3.5 are an indication to recommend a primary nerve operation at 3 months of age. If the test score is greater than 3.5, the decision for operative intervention is deferred until the next clinic visit.

Flail Limb

Infants found to have a completely flail extremity and/or Horner's syndrome at the initial intake assessment will be triaged to be seen earlier than 3 months of age, ideally at 4 to 6 weeks of age. A discussion about early operative intervention is held, and we initiate the surgical work-up. If there is no evidence of root avulsion on imaging, we will wait until 8 weeks of age to perform the primary nerve operation to ensure there is no evidence of rapid recovery of hand function indicating a neurapraxic injury. If rapid return of hand function is seen, we will defer the decision to operate until 3 months of age.

Complete Nerve Recovery by 3 Months of Age

Infants that have achieved full spontaneous recovery of their BPBI prior to 1 month of age are considered to have a true neurapraxic injury and are expected to have no long-term sequelae of their injury. These patients are discharged from our care following the 3-month clinic appointment.

Infants that demonstrate complete recovery of upper extremity function between 1 and 3 months of age may, or may not, have subtle sequelae of their injury. These patients may demonstrate differences in the active or passive ROM between the affected and unaffected upper limbs, or differences in complex patterning of the upper limb. For example, the affected limb may be tucked, or held at the side, when running. Consequently, these patients are seen at 2 to 3 years of age to evaluate for any subtle differences and provide appropriate education to the family where indicated.

Conditions Mimicking BPBI

Approximately 6% of patients seen in our clinic are ultimately found to have other conditions that can be confused with a BPBI. These conditions include: pseudoparalysis (secondary to humeral, clavicle, or rib fracture), cerebral palsy, spinal cord lesions, and isolated radial nerve injury. These patients are then provided referrals to appropriate care providers and discharged from our clinic.

Second Multidisciplinary Clinic Appointment (6 Months of Age)

The 6-month clinic appointment is specifically targeted at patients where there are concerns regarding either (1) poor neurologic recovery or (2) shoulder dysfunction. Prior to the 6-month visit, the patients are seen by our physiotherapist in a dedicated visit to reevaluate nerve recovery and ROM, reinforce stretching exercises, and address any new concerns. AMS scores and ROM values are recorded and compared with previous scores to assess for nerve recovery.

Indications for Primary Nerve Surgery at 6 Months of Age

Patients that do not demonstrate significant improvement in AMS scores are considered for primary nerve surgery. Specifically, if elbow flexion AMS scores remain less than 3, it is unlikely that the child will develop sufficient antigravity function by 9 months of age. The families of these patients are offered a primary nerve operation at this visit. Patients with improvement in AMS scores return for follow-up at 9 months of age.

Third Multidisciplinary Clinic Appointment (9 Months of Age)

The infant is reexamined and a final decision regarding a primary nerve operation is made at 9 months of age. Specifically, we perform the "cookie test" (►**Fig. 3**). This test requires the infant to be able to bring their hand to their mouth with the shoulder held in adduction and less than 45 degrees of neck flexion.[3,5] A single demonstration of this ability is sufficient to consider the infant as having completed the test successfully. If the patient clearly demonstrates the ability to pass the test, then they are not offered an operation. Stretching and ROM exercises are reviewed by the physiotherapist at this stage. Arrangements are made for follow-up in clinic at 2 years of age, or earlier if there are ongoing concerns regarding shoulder dysfunction or elbow contracture.

BRACHIAL PLEXUS BIRTH INJURY INITIAL HISTORY FORM

Referring Physician:
Age:

Affected Side: **Right** **Left**

Maternal History	
Diabetes	Y/N
Preeclampsia	Y/N
Duration of Labour in hours	
Gravida, Parity, Abortion	G _____ P _____ A _____
Child 1 (age, sex, weight)	
Child 2	
Child 3	
Delivery Problems with Earlier Pregnancy	Y/N, (details)

Birth History	
Birth Centre (Details)	
Delivery Position	Vertex/Breech/Other
Delivery	uncomplicated/difficult
Delivery	normal/forceps/vacuum/c-section
Dystocia Head	Y/N
Dystocia Shoulder	R/L/both (details)

Child Factors	
Large Baby	Y/N
Birth Weight in kg	
Gestation in weeks	
Asphyxia	Y/N
APGAR	1 min _____ 5 min _____ 10 min _____
Respiratory Complications	Y/N
Clavicle Fracture	Y/N
Rib Fracture	Y/N
Humerus Fracture	Y/N
Horner's Syndrome	Y/N
Hemi-Diaphragm Paralysis	Y/N
Shoulder Subluxation	Y/N
Torticollis	Y/N
Torticollis true mass, positional	Mild/moderate/severe

Fig. 2 Initial intake assessment form. The therapist obtains this information from the family and referring physician at the initial visit. It is subsequently reviewed by the surgeon at the 3-month multidisciplinary clinic visit. This information is also included in our research database after obtaining guardian consent.

Table 1 Active Movement Scale (AMS) scoring system[5]

Observation	AMS score
Gravity eliminated	
No contraction	0
Contraction, no movement	1
Motion < ½ range	2
Motion > ½ range	3
Full motion	4
Against gravity	
Motion < ½ range	5
Motion > ½ range	6
Full motion	7

Abbreviation: AMS, Active Movement Scale.

Table 2 Conversion score corresponding to AMS scores for the calculation of the test score

AMS score	Conversion score
0	0
1	0.3
2	0.3
3	0.6
4	0.6
5	0.6
6	1.3
7	2.0

Abbreviation: AMS, Active Movement Scale.
Note: The test score is calculated by adding the conversions scores for elbow flexion, elbow extension, wrist extension, finger extension, and thumb extension, which generates a score out of 10. The test score is only calculated when the infant is 3 months of age; it is not validated for other time points.[3,5]

Indications for Primary Nerve Surgery at 9 Months of Age

Persistently weak elbow flexion leading to an inability to pass the cookie test is an indication for a primary nerve operation at 9 months of age. In rare circumstances, elbow flexion has recovered and the cookie test is passed, but shoulder abduction, flexion, and external rotation remain poor. We have recommended a primary nerve operation in this setting as well.

Special Considerations: Infants with Poor Neurologic Recovery and Glenohumeral Dysplasia

In upper plexus palsies, every effort is made to manage the glenohumeral joint nonoperatively, using the Sup-ER splint, botulinum toxin, and casting when needed, while we await nerve recovery. In patients with both poor recovery of active external rotation and persistent dysplasia/posterior subluxation of the glenohumeral joint, the resting position of the shoulder (in internal rotation and adduction) can physically limit the ability to fully flex the elbow. In these situations, it may be challenging to accurately assess neurologic recovery of elbow flexion in infancy. We do not perform the shoulder surgery and the nerve operation in the same setting, as we feel it is important to evaluate the impact of one operation prior to proceeding with the other.

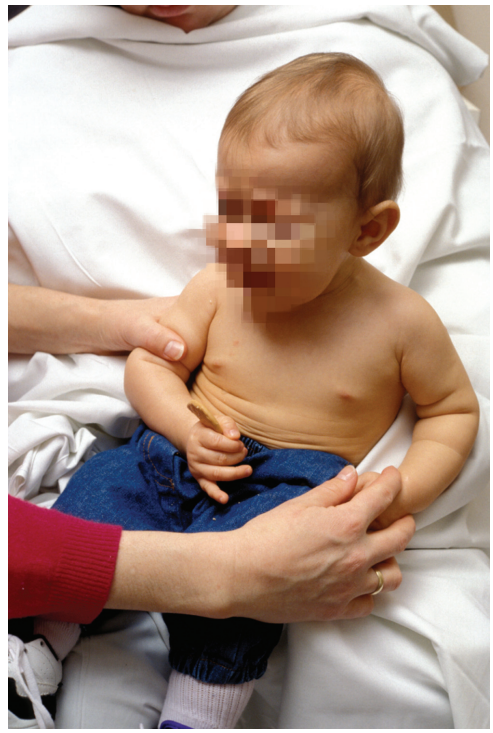

Fig. 3 Failed cookie test at 9 months of age. The infant is unable to sufficiently flex their elbow to bring a cookie to their mouth with the shoulder adducted. (Reproduced with permission from Swan and Clarke.[20])

When nerve recovery is globally poor at 9 months of age (i.e., AMS less than or equal to 5 for shoulder abduction/flexion and elbow flexion), a primary nerve operation will be recommended as the first priority. Conservative management of the glenohumeral dysplasia continues during this time; depending on nerve and functional recovery postoperatively, surgical management of the shoulder joint may be recommended as a second stage.

In a highly select subset of infants with overall good nerve recovery (AMS greater or equal to 6 for shoulder abduction and elbow flexion), who marginally fail the cookie test and have poor external rotation and a subluxed shoulder, the glenohumeral joint is surgically addressed first and upper limb movement reassessed 2 months later. If the patient now passes the cookie test, no nerve operation is recommended. If the patient is still unable to pass the cookie test, we offer a nerve transfer to augment elbow flexion (►**Fig. 1**).

Preoperative Planning

Selection of Operative Procedure

Neuroma Excision with Interpositional Nerve Grafting

Patients that meet criteria for a primary nerve operation are offered formal brachial plexus exploration, neuroma excision, and reconstruction with sural nerve grafting and

possible extraplexal nerve transfers. Extraplexal nerve transfers are used in situations where there are insufficient donor roots or insufficient graft material. The most commonly utilized extraplexal nerve transfers include the spinal accessory nerve (SAN) to suprascapular nerve (SSN) transfer via an anterior approach, and the intercostal to musculocutaneous nerve transfer in total plexus injuries.

Role of Neurolysis

We have previously demonstrated that there is no role for neurolysis alone in the management of these injuries.[9] The role of external neurolysis is limited to situations where a nerve is otherwise normal but is surrounded by scar from a neighboring, more proximal nerve injury.

Role of Distal Nerve Transfers

In our practice, distal nerve transfers are utilized in select circumstances, including late presentation after 12 months of age, lack of available donor roots (i.e., C5–C6 injury where both roots are avulsed), and in cases of otherwise good spontaneous recovery when a specific movement fails to recover.[10] For example, patients without glenohumeral dysplasia but who fail to recover active external rotation may benefit from the SAN to SSN transfer. Or, in patients who underwent primary shoulder stabilization surgery and demonstrate persistently weak elbow flexion, a single or double fascicular nerve transfer from redundant median and/or ulnar fascicles is performed.

Nerve transfers as a complete, primary approach to BPBI are gaining popularity for selected patients with upper trunk injuries, and thus far appear to have equivalent postoperative outcomes.[11,12] However, further long-term data are required given that results of nerve grafting have been shown to improve up to 4 years postoperatively.[13] As well, sensory outcomes following primary nerve transfers as a complete approach are lacking. Consequently, our preferred approach at this time remains neuroma excision and interpositional nerve grafting.

For patients with 4- or 5-root avulsion injuries, we consider transfer of the contralateral C7 root to the lower trunk via a retropharyngeal approach. Our otolaryngology colleagues assist us with the retropharyngeal dissection. There are limited data on the outcomes following this transfer in infants with BPBI, but our personal experience and the existing data[14] suggest that this approach is promising in these devastating injuries.[14]

Discussion and Consent Process

The preoperative discussion begins with a detailed description of the nature of BPBI and the rationale for treating the patient surgically. The risks of the operation and the expected outcomes are explained. Care is taken to set expectations very clearly with the family regarding the postoperative long-term prognosis. Based on our center-specific data, 85% of children have improved function, 10% of children have similar function, and 5% of children have reduced function. The families are informed that it will take 3 to 6 months before the child has returned to their preoperative baseline function. We explain that we will not see the final functional outcome from the surgery for up to 4 years postoperatively.

We specifically review the risks of pneumothorax, bleeding, phrenic nerve injury, infection, and wound-healing issues, as well as the use of fibrin glue.[15]

Investigations

Imaging

All patients undergoing primary nerve surgery at Sick Kids undergo two preoperative imaging studies: (1) diaphragmatic ultrasound and (2) computed tomography (CT) myelogram. The ultrasound is completed to evaluate and document the function of the phrenic nerves. A CT myelogram provides information regarding the presence or absence of pseudomeningoceles and whether rootlets are seen crossing these pseudomeningoceles. A prior study at our center demonstrated that a pseudomeningocele with absent rootlets on CT myelogram had a specificity of 0.98 in correctly identifying a root avulsion.[16] However, its sensitivity was lower and thus CT myelogram alone was insufficient to rule out root avulsion.[16] Nevertheless, the findings on CT myelogram provide good baseline information for preoperative planning.

Electrodiagnostic Testing

We do not perform electrodiagnostic studies (electromyography and nerve conduction studies) in the clinical management of our patients. We feel that these studies are unreliable in this patient population and generally result in overly optimistic assessment of neurologic recovery.[17,18] Consequently, we rely on clinical recovery to guide decisions on treatment.

Operative Protocol

A detailed description of our technical approach to microsurgical reconstruction of the brachial plexus has previously been published[3,19,20] (►**Table 3**). One can refer to these reports for details regarding sural nerve harvesting and brachial plexus exposure/dissection. Briefly, we conduct sural nerve harvest first in a prone position through three 2-cm transverse incisions, followed by a supraclavicular approach to the plexus through a V-shaped incision in the supine position (►**Fig. 4**).

Each nerve root and distal nerve target are carefully dissected and identified. Roots are dissected proximally to the vertebral foramina to identify evidence of root avulsion (visible rootlets and/or dorsal root ganglia identified in the operative field, an "empty" foramen). The nerves are visually inspected and gently palpated to identify the transition between firm neuroma tissue, and soft, pliable healthy neural tissue. Distally, we dissect until healthy nerve is seen. The clavicle can be elevated to facilitate better visualization. In rare circumstances for very distal lesions, we have dissected on the caudal aspect of the clavicle, but do not perform a clavicular osteotomy. The incision can also be extended along the deltopectoral groove to allow for a

Table 3 A step-by-step approach to exploration of the brachial plexus[20]

Surgical steps
Tattoo preoperative markings to facilitate subsequent wound closure
Elevate skin flap in subplatysmal plane and reflect superolaterally
Divide clavicular head of the sternocleidomastoid and external jugular vein
Identify the cervical plexus and C4 nerve root; divide supraclavicular branches
Divide omohyoid muscle and reflect Brown's fat pad off the clavicle
Divide the transverse cervical and suprascapular artery and vein
Identify neuroma/plexus between anterior and middle scalene muscles
Identify phrenic nerve and perform neurolysis if required
Using the C4 marker, identify the C5 root and dissect proximally to the foramen to exclude a preganglionic avulsion
Sequentially identify and dissect the C6 root and foramina, assess for a preganglionic avulsion
Dissect antegradely along the lateral border of the brachial plexus to identify the suprascapular nerve and the upper and middle trunks
Identify the remaining nerve roots and foramina, taking care to protect the subclavian artery and parietal pleura, assess for a preganglionic avulsion
Identify the lower trunk of the brachial plexus and its branches distally
Review the operative findings:
1. Which roots appear intact?
2. The position and length of any neuromata that require excision
3. The position of distal plexus targets requiring nerve grafting
4. Length of sural nerve (±supraclavicular nerve) graft available
Perform intraoperative nerve stimulation to help differentiate between an intraforaminal root avulsion and an intact nerve root if required
Prepare for neuroma excision and stump sampling for frozen section
Calculate amount of sural nerve graft required to reconstruct resultant defect and consider options for intraplexal or extraplexal neurotization
Cut grafts to length for proposed plexus reconstruction in a tension-free manner; prepare fibrin sealant
Ensure meticulous hemostasis within the wound bed; no further irrigation during gluing of grafts
Glue grafts in situ using a fibrin sealant; ensure optimal orientation using an operating microscope
Wound closure to include redraping of Brown's fat pad, repair of omohyoid muscle, and reattachment of sternocleidomastoid muscle
No surgical drain required
Skin closure in layers using absorbable sutures; infiltrate local anesthesia
Simple wound dressing; application of Velpeau sling, or external rotation splint/cast

Source: Reproduced with permission from Swan and Clarke.[20]

complete infraclavicular exposure of the plexus as necessary. We rarely find that this is necessary.

The nerve roots are stimulated and the response to stimulation recorded. Then, the neuroma and scarred segments of the brachial plexus are excised, and the proximal and distal nerve ends cut back to healthy-looking nerve. A fresh 15-blade scalpel is utilized for each cut. The transected proximal and distal nerve ends are passed off for pathology and carefully labeled.

Role of Neuropathology (Frozen Section)

It is critically important to cut back to healthy nerve ends to optimize nerve regeneration and functional outcomes. When we reviewed how well visual inspection of the nerve roots faired in comparison to frozen-section analysis, we found that frozen-section analysis prompted reresection of at least one proximal nerve root in 25% of patients.[21] With increasing experience, this percentage likely decreases with time, yet frozen section analysis remains a key component to our intraoperative protocol. Our surgical team physically delivers the fresh specimens to the Pathology department. We participate in the slide preparation process and ensure that all specimens are carefully labeled for analysis. Once the neuropathologist has completed their initial evaluation of the slides, the surgical team joins for a group review. Any concerns are addressed directly at this time (i.e., equivocal/unusual

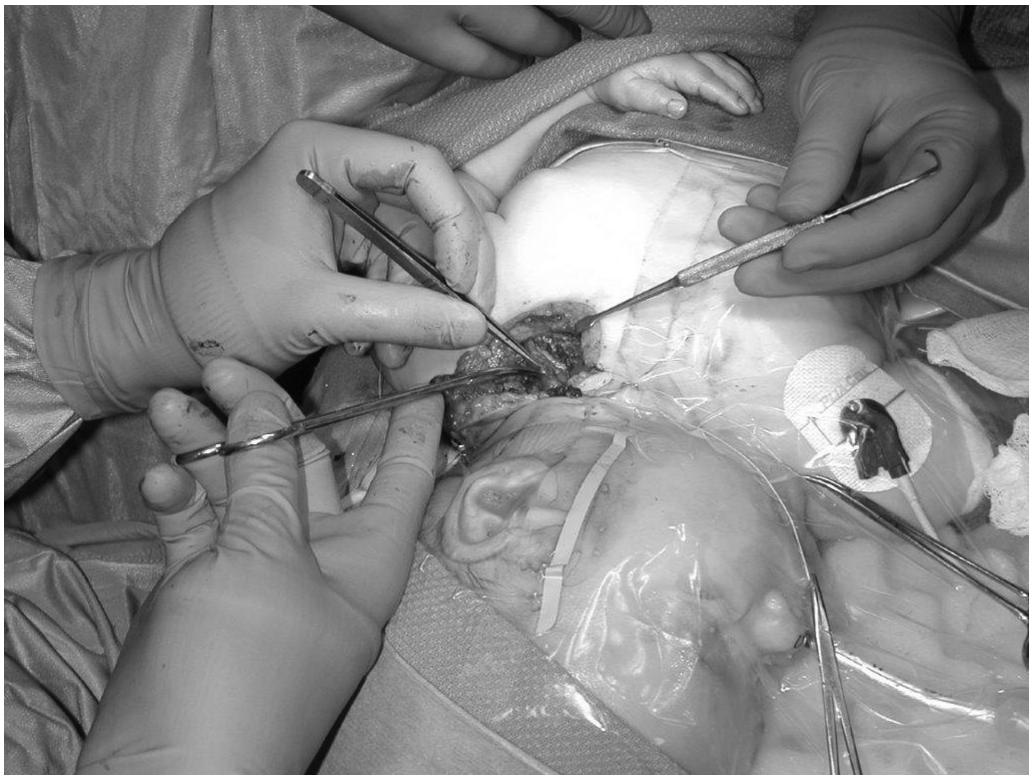

Fig. 4 A V-shaped flap is created and reflected posterolaterally by an incision along the posterior border of the sternocleidomastoid and the clavicle. Through this exposure, the entire posterior triangle is visible. For longer lesions, the incision may be extended along the deltopectoral groove, if necessary. The infant is positioned in such a way that the surgeon may work above, beside, and below the field. The neck is slightly extended, and the head faces away from the field. The hand and arm are accessible for observation of stimulated movements. A clear plastic head drape is used so that the patient's face and the nasotracheal tube are always visible to the anesthetist and the other members of the surgical team. (Reproduced with permission from Davidge et al.[19])

pathological findings, note of particularly challenging specimens to obtain, any relevant intraoperative findings). Once all slides have been reviewed and the findings discussed, the surgical team returns to the operating room. Recuts of any proximal or distal nerve ends are performed as necessary.

Active participation of the surgical team in this process recognizes the vital contribution of the neuropathology team. We feel this empowers the pathology team to take ownership of the case and helps provides direct clinical continuity. It also provides valuable learning opportunities for the surgical team and trainees.

Final Evaluation of Donor Roots

The definitive decision regarding which nerve roots are viable donors is made by combining preoperative and intraoperative information. Preoperatively, evidence of root avulsion on CT myelography is combined with preoperative physical examination AMS scores to determine presurgical probability of root avulsion. Intraoperatively, absence of a response to stimulation, presence of visible dorsal root ganglia or rootlets in the operative field, ganglion cells on frozen section, and an empty foramen are all further evidence of a root avulsion injury. We abandoned the use of intraoperative electrophysiology over 20 years ago, as we found it unreliable as a predictor of root avulsion.[22]

The quality of the nonavulsed proximal roots on frozen section is also important to our decision-making as to what roots to graft from. For example, if a proximal nerve root is cut back as proximally as possible but still shows evidence of scarring and substantially fewer viable axons, we would not count on this root to provide useful recovery. The reconstruction would be planned with the best quality roots and, if residual nerve graft material was still available, we would graft from the poor-quality root secondarily. Or, the poor-quality nerve root might not be used at all. Importantly, an intraforaminally avulsed root can look completely normal on histopathology.

Reconstruction Design

Once the viable donor roots have been identified, the reconstructive plan can be created. The gaps between each donor root and the distal targets are measured and recorded. These measurements are typically transcribed graphically on a white board. The total length of available nerve graft (sural and/or cervical plexus) is recorded. A proposed reconstruction is designed and drawn schematically, taking into consideration the gap distances and the length of nerve graft available.

Anatomic reconstruction is preferred if appropriate nerve root donors are available (►**Fig. 5**). However, when root avulsions are present, nonanatomic reconstructions must be employed. In all cases where the lower trunk is involved,

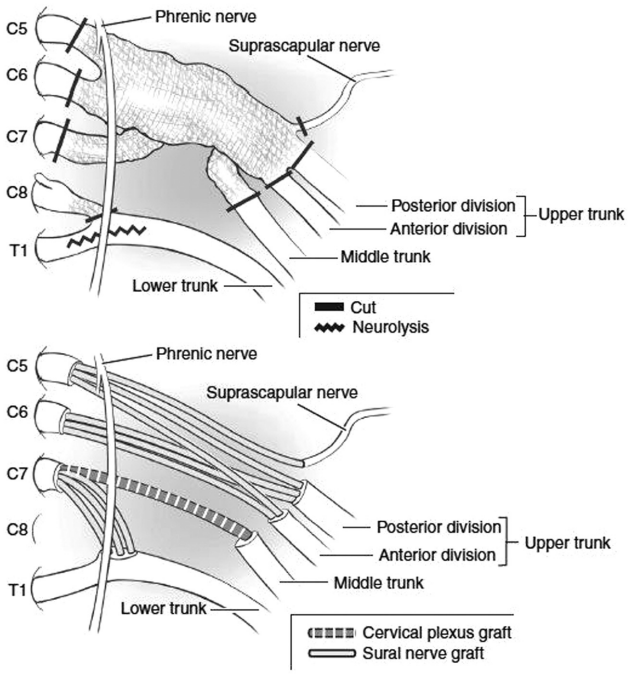

Fig. 5 Intraoperative case example of neuroma resection and interpositional nerve grafting in an infant with BPBI. The upper trunk (C5, C6) demonstrated a neuroma-in-continuity. The middle trunk (C7) was ruptured and the proximal and distal ends scarred to the upper trunk neuroma. Avulsion of C8 was confirmed unequivocally by the presence of a dorsal root ganglion in the surgical field. T1 was found to have a normal fascicular pattern with minimal involvement in the neuroma; external neurolysis was completed. The C5, C6, and C7 roots were all cut back to healthy nerve. Anatomic grafting was performed from C5 and C6 to the suprascapular nerve, anterior and posterior divisions of the upper trunk. Intraplexal neurotization to distal C8 was performed from C7. The lower quality graft material (cervical plexus graft) was utilized to graft from C7 to middle trunk. The phrenic nerve is shown traversing longitudinally over the plexus. (Reproduced with permission from Davidge et al.[19])

hand function is prioritized. The most important consideration is to robustly reconstruct the lower trunk to provide optimal hand function. Multiple cable grafts are directed from viable nerve roots to the lower trunk. The "best" root will be used for hand reconstruction, even if this results in a nonanatomic reconstruction.

When anatomic reconstruction of upper trunk is possible, equal priority is given to the anterior and posterior divisions. Ideally, there should be contributions from both C5 and C6 to each division (grafts running from both roots to both divisions). If only one of C5 or C6 is available for upper trunk reconstruction, then the single available donor root is used to graft to both posterior and anterior divisions of the upper trunk.[23] Typically, the posterior division of the upper trunk is larger and can accommodate up to three grafts, whereas the anterior division is smaller and can accommodate up to two grafts on average. Reconstruction of the middle trunk is given the lowest priority overall.

As noted earlier, the SAN to SSN transfer may be used in cases where there is either insufficient graft material or donor nerve roots. This transfer is robust and synergistic in function. We and others have demonstrated that outcomes of grafting from C5 versus nerve transfer from SAN are equivalent.[24] Direct transfer of three intercostal nerves to the musculocutaneous transfer is considered in cases of total plexus injury with limited donor roots.

Reconstruction Method

All nerve coaptations are performed with fibrin glue under the operating microscope.

Postoperative Care

Immobilization and Dressings

Choice of postoperative immobilization is based on the presence or absence of glenohumeral dysplasia. If there are concerns regarding glenohumeral dysplasia, we will immobilize the patient in an external rotation cast or splint. Otherwise, a Velpeau-style sling is applied[25] (►**Fig. 6**). This sling is applied using stockinette and safety pins, and is inexpensive, comfortable, and easy to use. Either way, dressings are maintained for 3 weeks. The neck is not immobilized in any of our patients.

The sural nerve harvest sites are dressed with Friar's balsam, Steri-Strip, gauze, Sof-Rol, and Kling wrap. The Sof-Rol and Kling wraps extend from the toes to above the knees. These outer dressing are allowed to fall off in the first 1 to 2 days postoperatively as the patient moves around.

Hospital Course

Patients are admitted to hospital postoperatively to a constant observation setting on our Plastic Surgery inpatient unit. Patients are monitored for complications, specifically any respiratory insufficiency, and provided analgesia in the form of acetaminophen, ibuprofen, and oral morphine.

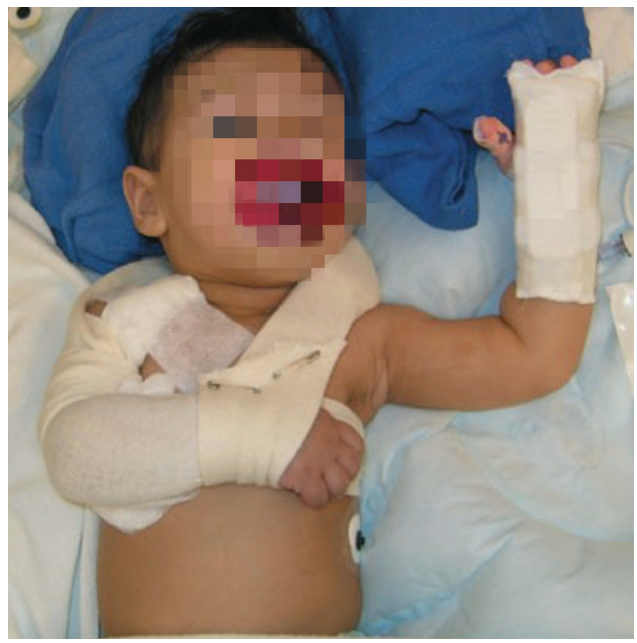

Fig. 6 Velpeau sling. The Velpeau sling is inexpensive and securely maintains the reconstructed extremity in full internal rotation with the elbow fixed at 90 degrees of flexion. (Reproduced with permission from Borschel and Clarke.[3])

Discharge Home and Follow-up

Patients are typically discharged on the second postoperative day. Patients are seen for the first follow-up in clinic at 3 weeks postoperatively. The immobilization device is removed at this visit and the wounds are inspected. The physiotherapy team begins stretching and mobility exercises at 5 weeks postoperatively. The parents are provided with ongoing education regarding expected timelines for recovery and improvement.

Management of Glenohumeral Dysplasia in BPBI

Glenohumeral dysplasia is common in infants and children with BPBI, particularly those with upper plexus injuries. Strong internal rotator muscles often overpower the weaker external rotators, leading to abnormal positioning of the shoulder in internal rotation.[26,27] This can result in remodeling of the glenohumeral joint, eventually leading to glenohumeral dysplasia and posterior humeral head subluxation. The impact of shoulder dysfunction is increasingly being recognized, and our clinic has adjusted our practice to address this issue. There is currently no consensus on the optimal management of the shoulder in BPBI.

Management of the Shoulder in Infancy

Initial Intake Assessment by Physiotherapy

It is imperative to identify patients at risk of shoulder dysfunction as early as possible. Early identification enables the team to institute treatment quickly, potentially preventing the longer-term sequelae associated with abnormal shoulder positioning. The physiotherapist initiates passive ROM exercises for the shoulder as of the initial visit.

Clinical Examination

During each clinical visit, the glenohumeral joint is evaluated for any evidence of posterior subluxation of the humeral head. The examiner places one hand on the infant's shoulder with their thumb on the humeral head. The other hand is placed on the distal humerus at the level of the epicondyles. The humerus is moved through internal and external rotation, while the humeral head is palpated for evidence of posterior subluxation. Both shoulders are examined to enable comparison to the contralateral side.

Dynamic Ultrasound Imaging

Indications for scheduling a dynamic ultrasound of bilateral shoulders at 3 months of age include any tightness into passive external rotation and/or external rotation AMS score less than or equal to 2 for at the initial visit with the physiotherapist. During the test, each shoulder is examined in internal and external rotation to assess the position of the humeral head in relation to the glenoid fossa. The affected limb is compared with the contralateral side for reference. An absolute α angle of >30 degrees is considered abnormal.[28,29] However, the α angle itself does not dictate our level of concern or treatment. Rather, it is the ultrasound findings in correlation to the clinical examination that guides management.

Sup-ER Splinting and Botulinum Toxin

Conservative management of the shoulder joint aims to improve passive ROM, maintain congruity of the glenohumeral joint, and improve muscle balance while the nerves are recovering.

The Sup-ER splint was pioneered by the team at the British Columbia Children's Hospital to treat infants with glenohumeral instability. The splint functions to keep the infant's arm in a supinated and externally rotated position to decrease the risk of developing glenohumeral dysplasia[7] (►**Fig. 7**).

If there is clinical and/or radiologic evidence of glenohumeral instability, we recommend treatment with Sup-ER splinting. This decision may be made prior to the 3-month assessment if deemed prudent by the physiotherapist at the early evaluations. The Sup-ER splint is fabricated on the same day by our occupational therapy team and the parents are instructed to apply the splint at night and during naps. If a Sup-ER splint is initiated, the patient will be seen in our occupational therapy department in 2 to 3 weeks to ensure a proper fit and compliance with the splinting protocol.

For patients with ongoing evidence of glenohumeral instability, the Sup-ER splint is continued for as long as the infant can tolerate it. We find that it is often challenging to continue Sup-ER splinting in infants older than 6 months

If the Sup-ER splint alone is insufficient to hold the humeral head in joint, our next step is to utilize botulinum toxin A injections to the strong internal rotators, followed by casting in external rotation. Botulinum toxin is injected into the pectoralis

Fig. 7 The Sup-ER splint has two components: a thermoplastic splint and a diaper-like elastic waistband. The arm splint holds the elbow in extension and supination. Two straps connect the arm splint to the posterior waistband of the diaper to position the shoulder in external rotation and hold the humeral head within the joint. The position of the glenohumeral joint in the splint can be confirmed with ultrasound. (Reproduced with permission from Zuo et al.[32])

major, latissimus dorsi, and subscapularis muscles in the main operating room. Although botulinum toxin can be effective in the short term, its benefit is not sustained over time and a significant proportion of patients with glenohumeral dysplasia will still require secondary shoulder surgery.[30,31]

Primary Shoulder Surgery within the First Year of Life

As outlined previously, a small subset of our patients is recommended for primary shoulder surgery in infancy (►**Fig. 1**). We counsel the families that shoulder surgery changes the location of the internal–external rotation arc but does not increase it. Our goal is to have the child be able to internally rotate to touch their belly and externally rotate to at least neutral. We also counsel families that there is a 5% chance of reoperation due to under- or overcorrection of the internally rotated posture.

Primary shoulder surgery at our center typically consists of teres major and latissimus dorsi tendon transfers, in addition to a subscapularis slide. The tendon transfers reorient teres major and latissimus dorsi into shoulder external rotators, correcting the underlying muscular imbalance. These two tendons are reinserted separately into the greater tuberosity of the humerus, rather than as a conjoined tendon.[32] The subscapularis slide allows partial release of the tight internal rotator, reducing the internal rotation contracture. Following shoulder surgery, the infant is immobilized for 4 weeks in external rotation, following which several weeks of physiotherapy are required to stretch the shoulder back into internal rotation.

Late Management of the Shoulder in BPBI

We continue to monitor the shoulder joint beyond the first year of life. Our team screens for joint contracture and abnormal positioning of the joint. Active and passive ROM is meticulously documented to assess trends and identify concerns as early as possible. Physiotherapy is recommended to improve and maintain passive ROM around the shoulder. Secondary shoulder surgery is indicated in children who lack active external rotation and in those with progressive glenohumeral dysplasia to promote joint remodeling. Preoperative magnetic resonance imaging (MRI) of bilateral shoulder joints is used for operative planning and staging of the dysplasia using the Waters classification.[33] In children younger than 4 years, tendon transfers alone may be sufficient to allow for glenohumeral remodeling. However, older children and adolescents require open joint reduction with glenoid anteversion osteotomy in addition to teres major and latissimus dorsi tendon transfers. Details of the surgical technique at our center have been previously published.[34] Postoperatively, the patients are maintained in an external rotation splint for 4 weeks. At that time, the splint is removed and the physiotherapy team begins a postoperative ROM protocol.

Outcome Assessment

Rigorous evaluation of clinical outcomes following BPBI surgery is vital for continued optimization of care. Our clinic maintains a robust patient database, which includes all patients that are seen by the team (including both operative and nonoperative cases). This facilitates longitudinal assessment of patient outcomes.

Our clinic utilizes multiple tools to track outcomes; the choice of tool depends on the age of the patient.

Birth to 4 Years of Age

Infants and young children are evaluated using the AMS scoring system and documentation of passive ROM values. These children are too young to employ other tools that require patient compliance and the ability to follow specific direction.

Patients Older Than 4 Years of Age

Older children are more reliably able to engage with examiners and complete more complex evaluations. These children undergo several detailed assessments by our physiotherapy and occupational therapy teams:

- Active ROM (all domains of upper extremity movement).
- Mallet score.[35]
- BPOM.[36]
- Sensory and pain evaluation by occupational therapy.

Long-term Follow-up

All patients with BPBI are followed in the clinic until they are 18 years old (regardless of whether they required a primary nerve operation). As children grow, it is common to encounter new functional concerns, and these are addressed at subsequent clinic appointments. Patients are typically followed in the full clinic on a yearly or biyearly basis depending on their concerns and progress.

Conservative Management

Patients with active rehabilitation concerns are followed more closely by the specific teams involved (physiotherapy or occupational therapy).

We know that affected muscles in BPBI do not grow longitudinally to the same extent as nonaffected muscles.[37] Therefore, a common concern during childhood and adolescence is loss of passive and active ROM after periods of growth, requiring bursts of therapy to regain motion. In particular, elbow flexion contractures are common and for the most part are managed nonoperatively, with a combination of stretching, splinting, and/or serial casting.[38] The management of elbow flexion contractures is an area of active research at our center.[39]

For functional concerns, our occupational therapy team provides support, adaptive devices, and education to enable patients to meet specific goals (i.e., sports, writing, grooming/hygiene, dressing, etc.). Management of pain, which is still not well understood in BPBI, consists of multimodal therapy including massage, heat, activity modification, stretching, and specific exercises. When pain is severe, we refer to our multidisciplinary pain clinic.

Other Secondary Surgeries

Decisions around secondary procedures for the elbow, forearm, wrist, and hand are made on an individualized basis and

are carefully considered to ensure that the operation will enhance but not downgrade function. This requires comprehensive functional assessments by our therapists and thorough discussion with patients and their families.

Peer Mentorship Program and Brachial Plexus Family Day

Patients with BPBI often benefit from support from peers, specifically those who may have similar functional concerns or those who previously underwent surgery. Our clinic offers a Peer Mentorship Program, which connects older patients (volunteers) with younger patients to provide support and guidance. Additionally, patients and families are invited to participate in the annual Brachial Plexus Family Day, which provides an opportunity to meet other patients/families/caregivers and provide support for our patients. These events have been very well received by patients and families alike and we consider them vital to the overall Brachial Plexus Clinic program.

Conclusion

Management of patients with BPBI is complex, challenging, and requires longitudinal care by a compassionate and dedicated multidisciplinary team. This article has described the current approach taken by the team at Sick Kids Hospital. Our research database has been central to providing evidence-based care of these children and will continue to allow us to evaluate and modify our management algorithm to optimize patient care. As we look to the future, we are excited to focus more on patient-centered outcomes and to engage in international collaborative research that has even greater potential to drive improvements in care for patients of all ages with BPBI.

Conflict of Interest
None declared.

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