Update on Management of Pediatric Brachial Plexus Palsy

Peter M. Waters, MD

Abstract: This manuscript will review the literature and focus on the present controversies regarding the natural history, microsurgical treatment, and secondary shoulder reconstructive surgery in infants with brachial plexus birth palsies. Surgical indications, expected outcomes, and complications will be addressed. The controversy regarding the timing of microsurgery in extrafacial ruptures will be addressed in detail. The developments in assessment and care of glenohumeral deformity with magnetic resonance imaging scans, arthroscopic and open reductions, and tendon transfers about the shoulder will be discussed. Recommendations for microsurgery and shoulder reconstruction will be based on the present evidence from the literature.

Key Words: brachial plexus, pediatric, management, surgery


INFANT EVALUATION

Brachial plexus birth palsy has an incidence of 0.38 to 1.56 per 1,000 live births.5,14 The difference in incidence may depend on the type of obstetric care and the average birthweight of infants in different geographic regions.13,44 Perinatal risk factors for brachial plexus palsy include large-for-gestational age infants (macrosomia),25 multiparous pregnancies, previous deliveries resulting in brachial plexus birth palsy,4 prolonged labor, breech delivery,29 and assisted (vacuum or forceps) and difficult deliveries.44 Delivery by cesarean section does not exclude the possibility of a birth palsy.7 Fetal distress may contribute to muscle hypotonia and provide less protection of the plexus from stretch injury during delivery. Mechanically, shoulder dystocia12 in vertex deliveries and difficult arm or head extraction in breech deliveries increase the risk of neural injury.29 Most commonly, a brachial plexus birth palsy involves the upper trunk (C5–C6), potentially in combination with an injury to C7; less often, the entire plexus (C5–T1) is injured. On extremely rare occasions, the lower trunk can be most significantly involved (C8–T1). Injuries are described classically as neurapraxia (Sunderland I), axonotmesis (Sunderland II–IV), neurotmesis (Sunderland V), or avulsion.78 Mechanically, lesions have been described as stretch (Sunderland I), varying degrees of rupture (Sunderland II–V), and avulsions (Fig. 1). Upper trunk extrafacial ruptures are more common with vertex delivery and shoulder dystocia. The right upper limb is more often involved because of the more frequent left occipital anterior vertex presentation. C5–C6 root avulsions are particularly frequent with breech presentation and can be bilateral.29 Entire plexus involvement can be a combination of stretch, rupture, and avulsion injuries and generally involves a more severe injury.30 Extensive work from Smellie’s28 and Duchenne’s24 original descriptions of brachial plexopathy with delivery through the anatomic studies of Stevens27 and Metaizeau define a mechanical basis for the infantile paralysis. This is the predominant theory regarding the etiology of brachial plexus birth palsies.

Narakas41 and others72 have attempted to categorize this clinical continuum into four categorical groups. The mildest clinical group (group 1) represents a classic Erb’s (C5–C6) palsy29 with initial absence of shoulder abduction and external rotation, elbow flexion, and forearm supination. Wrist and digital flexion and extension are intact. The successful spontaneous recovery rate is reported to be as high as 90% in this group. Group 2 includes involvement of C7 with the additional absence of wrist and digital extension along with C5–C6 impairment. These infants have the classic “waiter’s tip” posture of their hand and wrist. Prognosis is poorer with C5–C6–C7 involvement. Group 3 is a flail extremity but without a Horner’s syndrome. The most severe involvement (group 4) is a flail extremity and a Horner’s syndrome (ptosis, myosis, enophthalmos, anhydrosis). These infants may have an associated phrenic nerve palsy with an elevated hemidiaphragm.60 This can be assessed by observing the abdominal wall for symmetric diaphragmatic movement during respiration or by an expiratory chest radiograph. Phrenic nerve involvement increases the likelihood of an avulsion injury and limited spontaneous recovery.

For prognostic reasons, it is important to determine whether the level of injury is preganglionic or postganglionic.4,26 Due to the proximity of the ganglion to the spinal cord and the fact that the motor cell body is in the spinal cord, preganglionic lesions are avulsions from the cord that will not spontaneously recover motor function. By assessing the function of several nerves that arise close to the ganglion, one can often determine by physical examination the level of the lesion. Specifically, the presence of a Horner’s syndrome (sympathetic chain), an elevated hemidiaphragm (phrenic nerve), winged scapula (long thoracic nerve), and the absence of rhomboid (dorsal scapular nerve), rotator cuff (suprascapular nerve), and latissimus dorsi (thoracodorsal nerve) function all raise significant concern about a preganglionic lesion. Preganglionic lesions can only be reconstructed microsurgically by nerve transfers.62

From Harvard Medical School and Children’s Hospital Boston, Boston, Massachusetts.
Reprints: Peter M. Waters, MD, Children’s Hospital, 300 Longwood Avenue, Boston, MA 02115 (e-mail: peter.waters@tch.harvard.edu).
Copyright © 2004 by Lippincott Williams & Wilkins

J Pediatr Orthop • Volume 25, Number 1, January/February 2005
FIGURE 1. A severe traction injury to the brachial plexus may cause nerve injuries of varying severity in the same plexus. These include avulsion of the nerve root from the spinal cord (not repairable), extraf oralinal rupture of the root or trunk (surgically repairable), and intraneural ruptures of fascicles (some spontaneous recovery possible). (Reprinted from Green’s Operative Hand Surgery, 4th ed., edited by David P. Green, Robert N. Hotchkiss, and William C. Pederson, Fig. 39-1, p. 1273, copyright 1998, with permission from Elsevier.)

most commonly with thoracic intercostals, a branch of the spinal accessory nerve, or contralateral plexus. Postganglionic ruptures have reconstructible proximal and distal nerve beyond the zone of neural injury; thus, a postganglionic injury is a complex peripheral nerve lesion that can be reconstructed with nerve grafts if necessary.

The majority of obstetric plexus injuries involve the upper trunk. The classic Erb’s palsy involves only C5-C6 (46%), while it is also common to have C5-C6-C7 involvement (29%). The level of injury is usually postganglionic. When the lower plexus is involved, it is more common to have a preganglionic avulsion of C8-T1. The exception to this situation is an upper trunk lesion seen with a breech delivery. These neural injuries tend to be preganglionic C5-C6 avulsions from the spinal cord. A true Klumpke’s paralysis (isolated C8-T1) is very rare.

Most authors simplify the clinical continuum of recovery of a brachial plexus birth palsy into timing of recovery of specific motor function and use the absence of motor recovery as an indication for surgical intervention. Wyeth and Sharpe in 1917 advised surgical intervention if there was absence of recovery by 3 months of life. Gilbert and Tassin’s classic study concurred with the 3-month time interval and used the recovery of biceps function as the key indicator of brachial plexus spontaneous recovery. Waters similarly found biceps recovery statistically reliable. Laurent advised monitoring biceps, triceps, and deltoid function. Clarke’s group in Toronto described the timing of return of elbow flexion, with elbow, wrist, finger, and thumb extension as discriminators of outcome. Ultimately, though, in difficult cases of poorly recovering rupture, Clarke advised isolated elbow flexion recovery at 9 months by his “cookie test” to predict outcome and determine microsurgical intervention.

At this stage, most surgeons differentiate avulsion injuries from ruptures in their microsurgical recommendations. Avulsion injuries will not recover spontaneously, and therefore microsurgery is recommended before 3 months of age. Ruptures have varying degrees of recovery and are the source of controversy regarding the indications for and timing of surgery.

Invasive radiographic studies with myelography, combined myelography and computed tomography (CT) scans, and magnetic resonance imaging (MRI) scans have been used in an attempt to distinguish between avulsion and extraf oralinal ruptures. Kawai compared all three techniques with operative findings in infants. Myelography had an 84% true-positive rate with 4% false-positive and 12% false-negative rates. The addition of CT scans with myelography increased the true-positive rate to 94%. The presence of small diverticula was only 60% accurate for an avulsion. However, the presence of large diverticula or frank meningoceles was universally diagnostic. MRI had a true-positive rate similar to that of myelo-CT studies but also allowed extraf oralinal evaluation of the plexus. This permitted evaluation of possible double-crush injuries. High spin-echo MRI, MR myelography, and MR neurography may improve the resolution of MR analysis. MRI has the potential value of sedation only, as myelography requires general anesthesia in an infant. These radiographic studies may improve the quality of preoperative planning, but the final decision regarding the presence or absence of an avulsion injury is still made during surgery.

Electrodiagnostic studies with electromyography (EMG) and nerve conduction velocities (NCV) have also been used in an attempt to improve the diagnostic accuracy of the severity of the neural lesion. The presence of normal sensory nerve conduction in the absence of motor nerve conduction is diagnostic of a root avulsion. The absence of reinnervation at 3 months is indicative of an avulsion. Unfortunately, the presence of motor activity in a muscle has not been accurate in predicting an acceptable level of motor recovery in that muscle. In an EMG, the presence of reinnervation sometimes confuses the clinical picture. It has been documented that a near-normal EMG can be found in infants with a severe lesion or even root avulsion. There are often significant discrepancies between preoperative EMG, SNAP (sensory nerve action potential), and SSEP (somatosensory evoked potentials) testing and intraoperative surgical findings. A potential source for this is the plasticity of the infantile nervous system. For example, Slooff documented innervation of the
deltoid and biceps from C7 in the presence of avulsions of C5 and C6.\(^\text{82}\) At this stage, it is clear that neurophysiologic studies may underestimate the severity of injury and falsely provide optimism regarding recovery.\(^\text{81}\) At present, most centers and brachial plexus microsurgeons still ultimately rely on physical examination to assess recovery and decide on surgical interventions.

**NATURAL HISTORY**

Despite flaws in methodology or loss of patient enrollment in many of the natural history studies, there is sufficient evidence from several studies\(^\text{3,31,33,63,82,84}\) to draw significant conclusions. Most brachial birth palsies are transient. 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 to 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. At some point these limitations cross the line from clinical observations to functional impairment with permanent consequences. This is most evident about the shoulder, where children with incomplete recovery almost universally develop an internally rotated, adducted shoulder. With the development of limited glenohumeral motion, there is universal increased compensatory scapulothoracic motion. These patients can have functional limitations for above-shoulder, facial, and occipital region activities. The muscle imbalance of external rotation and abduction weakness and relatively normal internal rotation and adduction strength leads to glenohumeral joint deformity.\(^\text{66,84}\) This was described in the early 20th century, \(^\text{73}\) but more recent studies have defined the risk factors and progressive deformities in more detail with the use of arthrograms\(^\text{66}\) and CT and MRI scans.\(^\text{86,88}\) The glenohumeral deformity is progressive with age and appears very early in infancy. The development of glenohumeral deformity in these children follows a basic pediatric orthopaedic principle that muscle imbalance in a growing child will lead to bone and joint deformity. Adapting Severin’s classification of hip dysplasia, Waters described various grades of deformity\(^\text{88}\) (Fig. 2). The deformity evolves sequentially by grade from normal (I) to increased glenoid retroversion (II); posterior glenohumeral subluxation with posterior glenoid dysplasia (III); development of a false glenoid (class IV); to flattening of the humeral head and glenoid (class V). In addition, on

**FIGURE 2.** Glenohumeral deformity classification. Radiographic types as determined with use of CT or MRI scans. A, MRI scan of a type I (normal) glenohumeral joint (less than a 5-degree difference in glenoid version compared with that on the normal, contralateral side). The bisecting line extending from the spine of the scapula through the humeral head is outlined. The angle in the posterior medial quadrant is indicated by the arrow. Ninety degrees is subtracted from this measurement to determine the glenoid version. B, CT scan of a glenohumeral joint with type II deformity (on the right). The deformity is minimum (more than a 5-degree difference in glenoid version compared with that on the normal, contralateral side and no evidence of posterior subluxation of the humeral head. C, MRI scan of a glenohumeral joint with type III deformity. There is moderate deformity of the glenoid with posterior subluxation of the humeral head (less than 35 percent of the head is anterior to the scapular line). The scapular line and the tangential line indicating the anterior and posterior cartilaginous margins of the glenoid are shown. The angle in the posterior medial quadrant is indicated by the arrow. D, MRI scan of a glenohumeral joint with type IV deformity. There is progressive deformity (a false glenoid) and subluxation. (Reprinted with permission from *The Journal of Bone and Joint Surgery*, volume 80-A, no. 5, Peter M. Waters, Garth R. Smith, Diego Jaramillo, “Glenohumeral Deformity Secondary to Brachial Plexus Birth Palsy,” Fig. 2A-D, pp. 670–671, copyright 1998.)
At present, too few microsurgeons will participate in patients with incomplete recovery. The goals are to prevent joint contractures due to muscle imbalance, strengthen recovering muscles, and achieve developmental milestones in the presence of incomplete neural and muscular function. A multidisciplinary approach in the clinical setting is important to monitor recovery and guide decisions regarding nonoperative and surgical care.

At present, the major clinical dilemma early in life is to determine whether infants without antigravity return of C5-C6-C7 function at 3, 4, 5, or 6 months of age warrant surgical exploration and neural reconstruction. These infants have varying degrees of neurologic injury (Sunderland II–IV) and their ultimate neuromuscular recovery from spontaneous recovery and subsequent tendon transfers has not still been compared with similar group of infants with microsurgery and tendon transfers. Parents, primary care physicians, and therapists are under significant emotional pressure to do what they believe is best for the affected infant. Unfortunately, despite strong opinions and at times solicitous pressure from specific medical centers, there are still insufficient data to answer this question. Ideally this would be addressed with a prospective randomized clinical trial with sufficient enrollment. At present, too few microsurgeons will participate in a randomized trial due to clinical equipoise. The American Society for Surgery of the Hand and the Pediatric Orthopedic Society of North America have sponsored a center randomized trial that is underway; we hope it will further our knowledge base regarding this very important issue.

MICROSURGERY

Without question, the role and timing of microsurgery are the most controversial issues in the care of these infants. The original surgical interventions of the brachial plexus were established at the turn of the 20th century with resection of the neuroma and direct repair. Kennedy initially described three cases in 1903, with subsequent reports by Wyeth and Sharpe in 1917 and Taylor in 1920. However, in a report on 1,100 infantile brachial plexus patients in 1925, Sever was uncertain of the benefit of surgical intervention. By the 1930s, brachial plexus nerve surgery had fallen out of favor. It was not until the advent of microsurgical advances and the extensive work in the 1970s and 1980s by Narakas, Millesi, Gilbert and others in Europe, and comparable work by Kawabata and others in Asia, that brachial plexus microsurgical reconstruction became common. At this stage, it is nearly universally performed by major medical centers throughout the world to have plastic surgery, neurosurgery, and/or orthopaedic surgery sub specialized in microsurgical reconstruction.

The spectrum of nerve surgery includes neurolysis, neuroma resection and nerve grafting, and nerve transfers. Direct repair is rarely performed due to the extensive nature of the lesion and inability to obtain a tension-free repair without grafting. Although neurolysis has been performed extensively, most centers have abandoned its independent use. Clearly there is no role for neurolysis in the presence of an avulsion injury. It has been shown to be no different than natural history in total plexopathy, and the evidence is similar, although less conclusive, in upper trunk rupture situations. Gilbert strongly stated that he sees no role for neurolysis. Laurent advocated its use in conjunction with intraoperative electrophysiologic studies. If there is maintenance of more than 50% of a muscle action potential across the lesion and inability to obtain a tension-free repair without grafting, then Laurent indicated that neurolysis should be performed. Otherwise, the neuroma is resected and grafted. However, the recovery of muscle strength results with nerve grafting was more dramatic than neurolysis despite the fact that the preoperative status of the neurolysis patients was better than the grafting patients. Capek and Clarke described better long-term results after resection and grafting of both conducting and nonconducting neuromas than with neurolysis, despite an initial worsening of the situation with resection. Based on the available information at this time, neurolysis alone should be viewed as having little therapeutic benefit.

The present microsurgical standard of care is transection of the neuroma and sural nerve grafting in extraforaminal ruptures. In the usual upper trunk rupture, sural nerve grafts are performed from the C5 and C6 roots to the most proximal healthy nerve tissue of (1) the upper trunk anterior division/lateral cord/musculocutaneous nerve, (2) suprascapular nerve, and (3) upper trunk posterior division/posterior cord/axillary-radial nerves (Figs. 3, 4). In the case of segmental avulsions, nerve transfers in conjunction with nerve grafting are performed using the thoracic intercostals (T2-T4) and/or a branch of the spinal accessory nerve (CN XI) after it innervates the trapezius. In total plexal avulsions, nerve transfers are the only nerve reconstructive option and may include intercostals, spinal accessory, phrenic, cervical plexus, contralateral C7, and even the hypoglossal nerve. With biceps presentation avulsions of C5-C6, transfer of a part of the ulnar nerve to the motor branch of the biceps is useful. Carlstedt has done experimental and limited clinical work on direct reimplantation or grafting into the spinal cord, but at present the risk of cervical instability from the laminotomy and fasciectomy or injury to the nerves to uninvolved limbs does not warrant its use. Gilbert and Slooff argued that priority be given to microsurgical reconstructions of the hand in infants with extensive avulsions and limited nerve options. Unlike adults, infants with brachial plexopathy may have the potential to regain hand function after nerve grafting or nerve transfers. In each microsurgical case, the plan is individualized depending on the extent of injury and available reconstructive options.

Although there is ongoing debate about the timing of microsurgical intervention, the most common criterion used in clinical practice as an indication for microsurgery is the absence of return of biceps muscle function associated with total plexopathy and a Horner’s syndrome, or an upper trunk...
lesion. Avulsion surgical reconstruction is advocated at 3 months or less of life to limit motor endplate loss and maximize recovery. Reconstruction of extraforaminal ruptures is performed at between 3 and 9 months of age, depending on the center; the range of repairs cited in the literature extends from 1 to 24 months.

The quality of muscle recovery is open to debate and observer error, making comparative analysis difficult. Gilbert and others advocate for microsurgery when there is absence of antigravity biceps recovery by 3 months of age. The reasons for early intervention include less risk of irreversible loss of motor endplates with prolonged denervation and better parental acceptance of surgical intervention if performed while the limb is still flail or with minimal motion. However, prospective studies by Al-Qattan and Waters indicate that recovery of antigravity biceps function by 4 and 5 months of age, respectively, results in natural history outcomes that are equivalent to microsurgery, especially when combined with secondary tendon transfers to improve shoulder external rotation and abduction. Microsurgical results in both these papers resulted in improved function when performed later than 3 months of age. Finally, Clarke advocated microsurgery as late as 9 months in infants who fail the “cookie test” and thus have less than a grade 6 strength of biceps on the Hospital for Sick Children muscle scale. Microsurgical reconstruction at that relatively late time was of benefit. However, there is an initial worsening of the child's condition with microsurgery performed later in infancy. On average, recovery extends to 18 to 24 months after surgery, and final outcome improvement warrants microsurgery. Therefore, the motor endplates in infants may be more resilient than adults. Ultimately, the best time for microsurgical intervention remains unknown in the situation of an extraforaminal rupture. However, it is clear that there is a marked difference in number of procedures performed depending on the individual surgeon's bias toward timing.

The problem with reviewing the results of microsurgery is that few of these patients have both long-term follow-up and microsurgery alone. Gilbert and Tassin's original study compared microsurgery with spontaneous recovery. For C5-C6 lesions, 100% had class III recovery spontaneously, while 37% were class III and 63% were class IV with microsurgery. With C5-C6-C7 lesions, 30% were class II and 70% class III with observation. With microsurgery, 35% were class II, 42% class III, and 22% class IV. Later, Gilbert and Whittaker cited results of Mallet scores of III, IV, or V for abduction at 81% in C5-C6 reconstructions and III or IV at 64% with total plexus reconstruction at greater than 2 years of follow-up. In combination with secondary shoulder reconstructions, at 5 years these results increased to 70% Mallet class IV or V abduction for C5-C6 lesions. Similarly, with total plexopathy reconstructions prioritizing the hand, at 2 years of follow-up there were only 25% grade III or IV shoulder function; 70% had grade III, IV, or V elbow function; and 35% had grade III or IV hand function by Gilbert's new classification scheme. With the addition of secondary shoulder and hand procedures, this increased to 77% in the shoulder and 75% in the hand at 6 years of follow-up. Gilbert maintained that microsurgery not only improves function over natural history in selected patients but also increases the possibilities for secondary tendon transfers that will then further improve the clinical situation. These results, however, are comparable with the limited natural history data available. Benson et al reviewed 142 patients with follow-up to assess the natural history of brachial...
plexopathy in regard to the timing of biceps recovery. Seventy-one patients had full recovery by 6 weeks; the other 71 had recovery of biceps at greater than 6 weeks of age. At final follow-up of the latter group, 67% had excellent, 12% good, 5% fair, and 10% poor results by their assessment of shoulder function. Zancolli et al. found that 82% of the affected infants followed from birth had recovery of biceps, 75% of whom began recovery between months 4 and 5 of life. Waters addressed the same issue and found prospectively that of the 49 infants with no biceps recovery at 3 months, 42 recovered antigravity biceps function by 6 months. At follow-up greater than 2 years of age, infants who recovered biceps function between 3 and 6 months had a progressive decrease in Mallet grades for abduction, external rotation, hand to mouth, and hand to neck activities depending on the month of biceps recovery. In infants with biceps recovery between 3 and 6 months of age, the recovery of function by Mallet class was as follows: global abduction II (3%), III (52%), IV (46%); global external rotation II (54%), III (31%), IV (15%); hand to neck II (39%), III (33%), IV (28%); and hand to mouth II (33%), III (24%), IV (43%). These results are similar to Gilbert’s published microsurgical results.

In addition, in both natural history and microsurgery patients, secondary shoulder transfer and osteotomies significantly improved function. In the subgroup of 20 patients with recovery of biceps function between 3 and 6 months of life with shoulder reconstruction, there was an improvement to an average grade IV for all Mallet classes. Therein lies one of the present controversies: How different are patients with microsurgery at 3 months from those who recover biceps function between 3 and 6 months and have secondary reconstructions? It is critical to know the answer to this question to resolve whether unnecessary surgery is being performed and whether some centers are failing to adequately treat these infants by not being aggressive enough with microsurgery at 3 months of life. Further, since the published microsurgery results include secondary procedures, this controversy remains unresolved because of lack of comparable data. Although there are many believers of the importance of microsurgical intervention at 3 months, at present there are no studies that fully answer these questions. As mentioned above, there is a multicenter international study underway to address these issues.

The microsurgical treatment and long-term expected outcomes of infants and children with brachial plexus birth palsies remains controversial. Since the type and severity of neural injury vary per infant afflicted, the treatment and outcome are not expected to be the same. For the mild neuropritic lesion, spontaneous recovery is to be expected over the first several months of life, with complete recovery evident by the first year of life. This is in contrast to the infant with a severe avulsion injury, who will probably have a lifetime of disability despite extensive physical therapy and surgical management. Unfortunately at the time of this writing, a severe brachial plexus birth palsy is not a solved problem. Although there have been many advances since Duchennes’s (1872) and Erb’s (1874) classic descriptions of infantile paralysis, many difficult challenges remain. These include neural injuries that are more severe than can recover spontaneously; more complex and extensive neural injuries than available donor nerve grafts or transfers can repair; and avulsion injuries from the spinal cord that cannot be repaired directly or replaced adequately with nerve transfers. In these circumstances, therapeutic intervention with microsurgical reconstruction of the plexus in the first 3 to 9 months of life will improve the situation (one hopes) but not normalize it. Further advances in neural repair, regeneration, and growth are necessary to solve this problem. Complications of infection, systemic deterioration, bleeding, or worsening neural condition are rare. At times, a phrenic nerve palsy can recur with a postoperative elevated hemidiaphragm. If this persists and affects respiratory function, a late diaphragmatic plication is appropriate. The major complication of microsurgery is the failure to achieve the desired outcome. It is important for parents to understand that perfect range of motion, strength, posture, and function are rarely achieved with microsurgery. However, improvement from the natural history in all those areas is desired and possible. Still, 10% to 20% of ruptured and avulsed patients will not have an optimal result. In addition, even with a positive outcome for arm function and strength, most of these patients will require secondary tendon transfers to further improve their situation. Finally, the life-long implications for self-esteem, psychosocial well-being, and function in these children remain unclear.

**SHOULDER PROBLEMS**

There is frequent shoulder weakness, contracture, and joint deformity in infants and children with brachial plexus birth palsies. Even children with the mildest chronic plexopathy will have some limitation of glenohumeral motion with increased scapular winging. Only infants who recover antigravity biceps strength in the first 4 to 6 weeks of life will have no asymmetry of their shoulder girdle on long-term examination. The initial trauma of the birthing process may cause muscular injury that leads to myostatic contracture. Periarticular injury can be the result of the same process with resultant glenohumeral capsular and ligamentous tightness. On rare occasions, an infantile glenohumeral dislocation can occur due to the birth trauma. More commonly, abduction and external rotation weakness from a failure of neuromuscular recovery, in conjunction the muscular and periarticular tightness, leads to an internal rotation and adduction contracture. If the soft tissue contracture and muscle imbalance are allowed to persist, progressive glenohumeral joint deformity is universal. This only worsens the clinical situation of limited motion, strength, and function. The infantile treatment options of physical therapy, splinting, Botox injections, and surgical interventions are all designed to reverse this process.

**PATIENT EVALUATION**

Recovery of active muscle strength in infants is graded by Medical Research Council grading, Mallet classification, or Hospital for Sick Children scores. Assessing infants for shoulder function involves observation of spontaneous activity, neonatal reflex activity (Moro, asymmetric tonic neck, symmetric
tonic neck), and stimulated activity with and without gravity assistance. Passive glenohumeral motion is assessed with scapular stabilization. Internal and external rotation is performed both in adduction and 90 degrees of abduction while stabilizing the scapula against the thorax. The degree of scapular winging posteriorly with internal rotation, superiorly with full adduction, and into the axilla with full abduction and forward flexion is recorded. It is important to palpate for posterior glenohumeral joint dislocation in the posterior soft spot because it is critical not to miss the rare infantile dislocation. Dynamic instability is assessed by palpation posteriorly while monitoring passive and active range of motion, especially with adduction and internal rotation. Subscapularis tightness is evaluated by measuring the scapular-humeral angle in abduction, again with scapular stabilization. Pectoralis major, latissimus dorsi, and teres major tightness is assessed by palpation of the muscles with abduction and external rotation.

Infantile treatment of the shoulder focuses on maintaining a full passive range of motion. It is unclear how long after birth the nerves should be protected with immobilization to lessen recurrent traumatic stretch. However, early motion is important to prevent myostatic and periarticular contracture. Initially a gentle home program is begun at 7 to 10 days of life, followed a formal physical therapy program with home supervision in infants who do not recover rapidly in the first month of life. Full glenohumeral range with scapular stabilization is the goal. Abduction and external rotation splints have been used to improve or maintain range of motion. Compliance can be poor, and Zancolli expressed concern that these splints may increase the risk of injury to the physis and developing joint.92,94 Botox injections have been used to lessen the contracting muscle forces.95,96 Post-Botox splints, spica casts, electrical stimulation, and intensified therapy have all been advised. However, at present there are no data to guide the clinician on the indications and expected outcomes from these various treatments. It is clear, though, that failure to maintain full passive range of motion of the joint puts the child at risk for the development of glenohumeral deformity. Monitoring the patient’s passive external rotation in adduction limitations correlates with the degree of glenohumeral deformity and is a useful guide in terms of failures of physiotherapy and indications for MRI scans and surgical intervention.

If the child fails to recover external rotation strength and motion, there will be significant functional consequences.46 External rotation of the shoulder is necessary to achieve above- horizontal-shoulder activity (ie, reach the hand to the occupant, forehead). Scapulothoracic winging can compensate for limited glenohumeral motion in all planes except external rotation. In this circumstance, the scapula abuts the posterior thorax. This leads to marked limitation of function due to the inability to place the hand appropriately in space for many activities. In addition, weakness about the shoulder further limits hand use away from the body. The child has difficulty placing and maintaining the hand at a desired location in space due to fatigue. The affected limb will therefore most often be used at the child’s side or with support from furniture or the ipsilateral leg.

Arthrograms have been used to assess joints and bony development at a young age in infants and children with birth palsies.97 However, most institutions use MRI scans to assess the cartilaginous surfaces of the glenohumeral joint in young children and CT or MRI scans to evaluate the joint and bony structures in older children. In infants and young children, MRI scans require sedation, while arthrograms usually have been performed under anesthesia.98 The glenohumeral deformity progresses from normal (I) to posterior glenoid deformity (II), to humeral head subluxation and further glenoid dysplasia (III), to the development of a false glenoid (IV), to flattening of both the glenoid and the humeral head (V) (see Fig. 2). The degree of joint deformity can guide the surgeon in selection of surgical procedure. It is clear that with minimal joint deformity (grades I and II), releases or lengthenings of contracted muscles and tendon transfers for improved external rotation and abduction have a very favorable outcome.92,96 With more severe deformity of the joint (grade V and advanced grade IV), a humeral derotational osteotomy improves function significantly.96 The roles of arthroscopic debridement and reduction, open reduction and capsulorrhaphy, and glenoid osteotomy are still not fully defined.96 Our understanding of the degree of glenoid remodeling possible with a soft tissue procedure is still incomplete, but age, degree of preoperative deformity, and surgical procedures performed must all be factors. In patients with more advanced deformities (grades III and IV), these issues become important in the decision regarding the timing and choice of surgical procedures.

### SURGICAL RECONSTRUCTION

Indications for surgical intervention about the shoulder include infantile dislocation; persistent internal rotation contracture despite aggressive nonoperative management; limitation of abduction and external rotation function with plateauing of neural recovery; and progressive glenohumeral deformity. The age at intervention depends on the problem and its severity.

The rare infantile dislocation is treated as early as it is recognized, ideally before the glenoid deficiency becomes unreconstructable.79 Ideally this is within the first 3 to 12 months of life. An anterior release of the thickened capsule and middle and inferior glenohumeral ligaments, with debridement of the joint, is necessary for reduction of the humeral head. This can be performed open or arthroscopically. Lengthenings of the subscapularis and pectoralis major muscles are often required. A posterior capsulorrhaphy is performed, usually via a second incision. The deltoid is elevated from the spine of the scapula while protecting the axillary nerve, and the latissimus dorsi and teres major tendons are released from their often conjoint insertion onto the humerus. The infraspinatus–teres minor interval is used to expose the joint. The degree of glenoid dysplasia is assessed. The joint is atraumatically reduced into the deformed glenoid. This may require more extensive releases or lengthenings of additional myostatic contractures, such as the deltoid. Stability of the joint is assessed. On rare occasions, a glenoid osteotomy to elevate the posterior glenoid, with a posterior acromial bone block or allograft, is necessary to maintain joint reduction and congruency. The posterior capsule is reeved, but not excessively so, as this will limit postoperative glenohumeral motion.
The latissimus dorsi and teres major tendons are transferred to the greater tuberosity of the humerus. Postoperative immobilization involves a shoulder spica cast for 4 to 6 weeks.

Carlioz described a subscapularis lengthening for infants who fail to regain passive external rotation in the first 6 to 12 months of life with extensive physical therapy. The subscapularis can be lengthening by an anterior Z-lengthening of the tendon, musculotendinous lengthening via an anterior axillary incision, or elevation of the subscapularis off the anterior scapula as a muscle slide. All can improve passive external rotation and glenoscapular excursion.

Arthroscopic surgery has recently been used in children with brachial plexopathy and glenohumeral problems. Arthroscopy allows for direct visualization of the joint to assess the degree of deformity and soft tissue contractures. Anterior release of the thickened middle and inferior glenohumeral ligaments and the subscapularis can be performed. The joint can be debrided of interposing, intra-articular tissue and synovitis. In an infant, this can performed in a lateral decubitus position with manual traction of the affected limb. The smaller arthroscopic equipment (2.7-mm arthroscope) is used with standard shoulder arthroscopy techniques. An electrocautery device is most useful in performing the anterior release from the posterior portal. Direct visualization of the joint reduction can be performed. The extent of the glenoid deformity and the degree of joint stability after release and reduction can be assessed. Stability is often achieved, with greater than 70 degrees of external rotation in adduction reached passively. Open tendon transfers can be performed in the same surgical setting without difficulty. It is too preliminary at the time of this writing to know the ultimate role of arthroscopic surgery in these children. However, early clinical experience is promising, and arthroscopy may provide a less invasive form of treatment based on the principles Fairbanks outlined in 1913.5,6,7

Infants who present with a brachial plexus birth palsy are examined closely for a dislocation or instability of the glenohumeral joint as well as for contractures about the joint. In the presence of an infantile dislocation, an MRI scan under sedation is obtained to assess the development of the glenoid and the joint alignment. If the patient is young and the glenoid is not severely deformed, either an arthroscopic or open reduction is performed. A posterior capsulorrhaphy and latissimus dorsi and teres major tendon transfers are usually performed at the same surgical setting.

Commonly infants and children with a chronic brachial plexus birth palsy will develop an internal rotation/adduction contracture about the shoulder. Ideally, physical therapy for passive range of motion with scapular stabilization is begun in the first few weeks of life in an attempt to prevent contractures and joint deformity. If an internal rotation contracture develops in the first 1 to 2 years of life and does not resolve with 3 months of supervised therapy, an MRI scan is used to assess the glenohumeral joint. If the joint is normal, then more aggressive therapy is considered. At times, an open or arthroscopic subscapularis release is performed. This situation is rare. Unfortunately, joint deformity occurs commonly early in life in the presence of muscle imbalance and periarticular contractures. In the presence of mild deformity (types II and III), an anterior release of myostatic contractures and posterior latissimus dorsi and teres major tendon transfers are performed (Fig. 5). Results from this surgery have been excellent and have stood the test of time for many patients at multiple centers throughout the world. If the joint deformity is more severe (more advanced type III or type IV), it is difficult to decide whether the joint can be reconstructed. Visualization of the joint, either open or arthroscopically, can aid in the decision. Reduction of the humeral head and stabilization of the joint by an anterior capsule release (arthroscopic or open), posterior capsular reefing, and posterior tendon transfers is preferred. However, to be successful, this requires maintenance of the reduction, acceptable glenohumeral motion, and remodeling of the glenoid. The degree of glenoid remodeling with soft tissue procedures is still unknown. If the joint deformity is too severe at any age (advanced type IV or type V), a humeral external rotation osteotomy is performed. This is done when it is determined that the joint cannot be reconstructed. This is common in adolescent patients with an internal rotation contracture, external rotation limitation, and advanced deformity by CT or MRI scan. Multiple publications over time have indicated excellent outcomes with this operation. A posterior opening wedge glenoid osteotomy with bone graft is used rarely to provide posterior bony stabilization in reconstructible type IV deformities. However, the indications for and outcomes from this procedure remain unclear.

Complications are rare in these operations. Even in situations of marked weakness of the affected arm, the latissimus dorsi and teres major transfers usually provide a positive antigravity result.86 Preoperative antigravity deltoid strength is necessary for a successful outcome with a latissimus dorsi nerve repair.
transfer to the rotator cuff. The status of the glenohumeral joint clearly affects the outcome. Preoperative assessment of the joint with an MRI scan or arthrogram will aid in the process of selecting a specific surgical intervention. If the joint is deformed, there may be continued postoperative restriction of motion with a tendon transfer. This is most common with limited external rotation in adduction. The increased scapulothoracic motion cannot compensate for the limited glenohumeral motion in those planes. A secondary humeral osteotomy with external rotation may be necessary to remedy the marked trumpet posture and inability to externally rotate the arm from an adducted position. This situation could be viewed potentially as failure of preoperative selection, as an original joint reduction and stabilization or osteotomy may have been indicated.

Joint remodeling may be limited with extra-articular tendon transfer procedures alone. Closed reductions have been described, but follow-up radiographic studies are limited. Open or arthroscopic reductions may improve the potential for joint stabilization and remodeling. Longer follow-up studies are necessary to answer these questions. The risk of long-term pain and disability from a deformed joint is unknown, but it appears to be much less than a dysplastic hip. This issue is central to many medical-legal disputes regarding the outcome of these children. In the situation of a flail shoulder, arthrodesis may be indicated to improve upper extremity function.  

ELBOW AND FOREARM RECONSTRUCTION

Elbow flexion and forearm supination deformities can occur with a permanent Klumpke (C8-T1) or mixed brachial plexus lesion. Contractures, bony deformity, and joint instability are the result of muscle imbalance in a growing child. In the rare patient with residual C8-T1 neuropathy with recovery of C5-C6 function, the elbow and forearm deformities are secondary to an intact biceps muscle in the presence of weak or absent triceps, pronator teres, and pronator quadratus muscles. Progressively, the biceps creates an elbow flexion and supination deformity from unopposed muscular activity. Soft tissue contractures develop, followed by rotation deformities of the radius and ulna.  

Radial head dislocation may occur. The wrist and hand are often in extreme dorsiflexion because of unopposed wrist dorsiflexors. In the position of forearm supination, gravity exacerbates the dorsiflexion deformity. The patient is left without use of the hand and performs bimanual activities using the volar and ulnar forearm as an assist. Often shoulder abduction and internal rotation are required to improve assistive function. Activities that require simultaneous elbow flexion and forearm pronation, such as dressing, eating, and writing, are significantly limited. In addition, the forearm and hand posture is a major cosmetic concern to both the patient and the family.

The biceps tendon can be treated by Z-lengthening and rerouting around the radius to convert it from a supinator to a pronator. This will improve elbow extension and forearm pronation. Surgically, the biceps tendon is identified as it inserts into the radial tuberosity. By dissecting lateral to the tendon, the brachial artery and median nerve are protected. A long Z-plasty of the tendon is performed from the musculotendinous junction to the insertion site. The distal attachment of the tendon is rerouted posteriorly around the radial neck, from medial to lateral. Care must be taken to stay adjacent to the radial neck to avoid injury to or compression of the radial nerve. The distal tendon is reattached to its proximal counterpart in a lengthened position. This converts the bicaps into a forearm pronator.

In the presence of a supination contracture, the rerouting procedure alone will fail because of recurrence of the deformity. Zancolli suggested performing simultaneous interosseous membrane release, but active pronation was maintained in only 50% of patients who underwent this procedure. Bony correction of the forearm deformity can be performed more predictably. Manske et al proposed staged procedures of tendon rerouting and forearm osteotomy. Waters and Simmons described simultaneous tendon rerouting and osteotomy, using internal fixation to avoid multiple operations and loss of alignment. In both techniques, the forearm is positioned in approximately 20 to 30 degrees of pronation.

These patients clearly have significant improvement in their functional capabilities. Bimanual tasks such as lifting, carrying, and transferring are easier. The affected extremity becomes a better assistive extremity to the unaffected side. The wrist and hand now have greater assisted palmar flexion and resolution of their dorsiflexion deformity. In addition, the patients are usually pleased with their cosmetic results.

WRIST AND HAND RECONSTRUCTION

Children with marked, permanent limitations of hand function are very difficult to successfully reconstruct to a high level of function with secondary procedures. This is due to the limited number of donor muscles with sufficient antigravity strength for transfer. Flail hands and wrists are fortunately rare but are functionally and cosmetically disruptive to the patients and their families. For this reason, microsurgical reconstruction in infancy prioritizes the hand in total, severe plexopathy situations.

Transfer of the flexor carpi ulnaris to restore active wrist extension is performed in the chronic C7-radial neuropathy situation. Wrist fusion, while maintaining distal radial physesal growth, can bring stability to the flail wrist and improve assistive hand function. Surgical procedures about the hand otherwise need to be individualized depending on the number of antigravity, expendable donor muscles available and the functional loss. Alternatives are limited and outcomes marginal in general.

ACKNOWLEDGMENT

The author recognizes the hard work of Ms. Anne Kuo in assisting with the references and illustrations for this manuscript.

REFERENCES


© 2004 Lippincott Williams & Wilkins
Management of Brachial Plexus Birth Palsy


