Use of the Active Movement Scale in Outcome Prediction in Birth Brachial Plexus Injuries: Early Results

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Part 1: Foundations of this project

Personal experience in practicing a pediatric surgical subspecialty has led the author to explore models of care delivery for chronic disorders in children and young adults. These disorders may include genetic entities (eg. cystic fibrosis), acquired disorders (eg. malignancies), birth defects (eg. spina bifida and related disorders) and forms of trauma (eg. birth-related brachial plexus injuries), among others. Given the complex natures of these problems, the care needed by these patients can be quite complex, involving multiple visits to providers over many years or even the entire lifespan. This care can be delivered in a fragmented fashion, by one or more specialists, or could be delivered in a multidisciplinary setting.

It seems intuitive to say that the results of care improve with the degree of coordination of care, with a single-site, multidisciplinary clinic (MDC) being the acme of such approaches. Nevertheless, it is difficult to show evidence in support of this statement in all such diagnostic groups. In the example of care of patients with spina bifida, a well-known attempt at demonstrating this was the report of Kaufman et al. who reported on a group of patients who had been followed in a multidisciplinary clinic that closed. These patients were subsequently contacted 3 years after the clinic was disbanded and their health status and current follow-up systems were identified. They were also compared to a matched cohort of patients followed in an MDC in a neighboring state. The key findings were that, despite having been referred to specialty providers, a majority had not had any follow-up until serious morbidity developed. The authors noted a higher incidence of amputation and nephrectomy in the group with no follow-up. It was concluded

that regular attendance at an MDC was associated with a lower frequency of "preventable" morbidities. (Kaufman 1994)

In the case of oncology care, it has been easier to demonstrate that care through an MDC is linked to improved patient outcomes, both in pediatric and adult settings. In a recent review by Tyler et al., the value of an MDC was taken as an *a priori* in the development of a clinic for care of patients with cutaneous lymphomas, a rare disorder but clearly one with potential involvement of multiple specialty providers. (Tyler 2015). A similar report by Dundee et al. concerning the establishment of MDC centers for prostate cancer management in Australia makes the statement that "It is well recognized that multidisciplinary care (MDC), an integrated team-based approach to cancer care, can lead to improved decision-making and better survival outcomes." However, in the same report, the authors note that "Without specifically designed, prospective trials evaluating the impact of MDC, it is difficult to assess the extent of oncologic benefit from this type of clinic. Such a trial would require the recruitment of thousands of patients over many years and would incur significant cost." (Dundee 2015) In an editorial note, Gupta noted that there were still "pitfalls" even in a well-functioning MDC, giving the example of privately insured patients being held out of such clinics, balanced by the observation that there are clearly cases of exceptional care administered without the benefit of an MDC. (Gupta 2007). Hence it seems still to be the case that the apparent superiority of care delivery in an MDC has not been completely confirmed in oncology care.

One true advantage of this care approach in oncology is that, given a large enough number of patients, it is possible to define relatively homogeneous groups for appropriate study. In addition to being able to group and stratify patients by demographic features, clinical presentation, extent of tumor at diagnosis, the ever-expanding range of molecular descriptions of individual tumors further ensures that treatments and outcomes can be evaluated with

accuracy. For many other disorders, this feature is strongly lacking. Using the techniques of comparative effectiveness research to assess the treatment and outcomes of children with cerebral palsy (CP) becomes very difficult when the treatment groups are so heterogeneous. Children with CP will vary by birth history, type of CP, cause of CP, types of interventions etc. The same is true of children with spina bifida, who have a wide range of clinical presentations, functional abilities, comorbidities, etc.

As a result, the concept of using birth-related brachial plexus injury (BBPI) is appealing because of the feature that this relatively frequent birth injury, which occurs globally, has a very narrow band of presenting features. Obviously, all patients with BBPI suffer the injury at the same age. They are usually larger than average babies, with a limited number of subtypes of injury. They typically are otherwise healthy, and these injuries occur in isolation. They may go on to receive care for this injury at an MDC or other setting, from a variety of specialists. The homogeneous nature of this group at baseline more readily lends itself to comparisons between treatment groups, or between clinics.

This project was designed to explore a population of infants seen in a single BBPI MDC and assess if existing clinical assessment tools might lend themselves to measuring or predicting outcomes of these patients. This is intended as an initial exploration in this area.

Part 2: Use of the Active Movement Scale in Outcome Prediction in Birth Brachial Plexus Injuries: Early Results

Abstract

Purpose

Injury to the brachial plexus at birth has an unpredictable and poorly understood natural history. While many infants will have spontaneous recovery of function, others are at risk for permanent disability. The purpose of this study is to explore if the early clinical examination using the Active Movement Scale (AMS), in combination with accurate classification of the extent of injury, can be used to predict the future need for an intervention to improve natural history.

Methods

Single center retrospective review of a cohort of infants seen in a multidisciplinary clinic between 1997 and 2014, with minimum of 18 months follow up. All infants were included who were seen before the age of 6 months and were assessed using the AMS tool. Demographic data and subsequent interventions were recorded. AMS scores were recorded in intervals of 3 months, and the best possible elbow flexion score was identified in each interval. Data was collected using REDCap and analysis of contingency tables was done using commercially available software.

Results

191 infants were included in this study (70M: 121F). Injuries were classified as Narakas type 1 in 102 (53.4%), type 2 in 65 infants (34.0%), type 3 in 16 (8.4%) and type 4 in 7 (3.4%). Spontaneous recovery occurred in 95 infants (49.7% of total). When analyzed by Narakas type, recovery occurred in 75.5% of type 1

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injuries and 24.6% of type 2 injuries. Type 3 and 4 injuries (combined) were significantly less likely to exhibit spontaneous recovery than type 1 and 2 injuries (p<0.0001). Recovery was seen in 12.5% of type 3 injuries and none of the type 4 injuries.

Recovery of normal elbow flexion scores for both type 1 and type 2 injuries was associated with a significant decrease in the risk of needing a subsequent therapeutic intervention. In type 1 injuries, attaining either a full score or functional equivalent, at any age before 9 months, was associated with a significant reduction in the risk of needing subsequent intervention. Similar results were found with type 2 injuries. The frequency of recovery in type 3 or 4 injuries was so low that no correlation between elbow flexion and avoidance of intervention was identified.

Conclusions

Infants with Narakas type 3 or 4 injuries are unlikely to avoid needing an intervention for their injuries. Infants with type 1 or 2 injuries are significantly more likely to have spontaneous recovery, and early return of high elbow flexion scores on AMS testing is associated with further significant reduction in the risk of needing an intervention.

Introduction and Background

Anatomy and mechanism of injury

The brachial plexus is a region of the peripheral nervous system that is vulnerable to injury, in part as a function of its relatively superficial location. It consists of contributions from five spinal segments (C5 through T1), which then exit the posterior cervical triangle, merge and rearrange into three trunks. These

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trunks, in turn, divide and recombine to become the peripheral nerves of the arm. The two highest roots (C5 and C6) become the upper trunk, which then divides further, with the principal terminal branches including the axillary, suprascapular and musculocutaneous nerves, among others. The C7 root forms the middle trunk, which becomes a principal contributor to the radial nerve. The C8 and T1 roots form the lower trunk, whose terminal branches include the ulnar nerve and the majority of the supply of motor function of the hand. Since the bulk of these structures lie in the supraclavicular fossa, they are particularly susceptible to stretch mechanisms of injury, especially those involving forced depression of the shoulder associated with lateral flexion of the head in the contralateral direction. These mechanisms involve the usual sources of high-energy trauma, such as vehicular injury and falls from a height, but these injuries also commonly occur in the context of the birth process.

Birth brachial plexus injury

Birth brachial plexus injury (BBPI) most often occurs during vaginal, vertex deliveries, although it has been observed in all obstetrical presentations, including operative delivery. It occurs in 2-4 per 1000 live births and is most often associated with shoulder dystocia. It is believed that the stretch injury occurs while the shoulder is still in the pelvis, as traction is applied to the infant's head. The nature and extent of the resultant nerve injury is highly variable, with involvement of the upper trunk alone being the most common location of injury (although the entire plexus can be in injured) and a transient, spontaneously recovering neuropraxic type of injury being commonly observed. However, while the infant's function may seem quite severely impacted at initial assessment, varying degrees of recovery can and do occur. In the mildest injury, nerve function can return within a few weeks. Nerve injury with neuroma formation is the next injury by severity and can recover over a period of weeks to months. Nerves which are either completely disrupted or develop non-functioning

neuromas tend to reach a plateau in their clinical recovery. Lastly, nerve injuries that involve nerve root avulsion from the spinal cord are non-recoverable, and are considered the most severe form of injury within the brachial plexus. Currently, no feature on initial exam is known to predict the individual infant's outcome in BBPI, nor is there any currently available imaging or neurophysiologic technique which distinguishes between those infants likely to have a good, functional spontaneous recovery and those who will not. (Andersen 2006)

Classification of BBPI

Early descriptions of BBPI identified patterns of involvement and gave rise to the earliest, eponymic classifications of these injuries. Duchenne had identified the occurrence of birth injuries to the brachial plexus as early as 1861, which subsequently was refined by Erb in 1875 when an injury involving the muscles innervated by the upper trunk was described. This was the origin of the eponym Erb-Duchenne, or simply Erb palsy, describing the isolated upper trunk injury. Also in 1875, Klumpke described the injury involving the lower brachial plexus in isolation. This lesion is rare in BBPI and is more often seen in non-birth related injuries, particularly in association with pathological processes of the upper chest (eg. apical tumors of the lung).

Narakas proposed a classification scheme based on the anatomical extent of involvement. In this system, involvement of the upper trunk alone (ie. C5-6 roots) is termed Type 1, while the Type 2 injury adds middle trunk involvement to the Type 1 injury (ie. C5-7 roots). The Type 3 injury involves all levels of the plexus, while the Type 4 injury is also a pan-plexus injury but with the addition of Horner syndrome. The presence of Horner syndrome implies a proximal injury to the T1 root. This scheme is summarized in Table 1. This classification is easily applied and is in extensive use, as will be seen below. (Narakas 1986)

Natural history

The natural history of BBPI varies greatly by individual. As stated above, injuries of lower severity are likely to recover spontaneously, while the most severe do not. At initial exam, particularly in the first few weeks of life, there is no feature of the clinical exam that enables one to distinguish the infant who is going to require an intervention (because of failure to recover) from one that will not. This is a result of a combination of two phenomena. The first of these is the fact that at first all motor deficits are similar, and the underlying injury type only becomes recognizable by observing recovery. The other is that the child with multiple root level involvement can have differing degrees of severity at different levels, eg. an infant who appears to have injury to the entire plexus at first exam may subsequently have quick recovery of function in the lower trunk muscles but turn out to also have root avulsions involving the upper level muscles. Over time, clinical observation has shown that a significant number of infants with BBPI will go on to have excellent functional recoveries without the need for intervention. The frequency of spontaneous recovery is unknown, but is variously estimated to range from 52 to 90%. (Andersen 2006, Clarke 1995, Foad 2009, Ali 2014)

Specific aim: Outcome prediction

Given that spontaneous recovery is common, but difficult to predict at initial clinical exam, the goal in this paper is to explore whether the use of a standardized clinical assessment tool, specifically derived for BBPI, can be used to identify those infants who will need a therapeutic intervention because of lack of recovery and distinguish them from those who will not. The benefits of early identification would, most importantly, be to reassure parents early on in their child's course if they will not need intervention. This would also allow for appropriate conservation of resources, if the need for clinical follow-up assessments could be tailored to the specific needs of the child and family. Our

clinical experience has been based on the use of the standardized tool, the Active Movement Scale (AMS), which will be described below.

Prior attempts at outcome prediction most famously included the "cookie test," in which a child of developmentally appropriate age (about 9 months) was given a cookie in their hand on the affected side and observed to see if they would bring it to their mouth. The baby was assumed to be sitting upright, with no more than 45 degrees of forward flexion of the neck. A baby who passed this test was thought to be likely to avoid any need for further intervention. It was later reported by Michelow et al. that achieving good elbow flexion by the cookie test was actually incorrect in a number of instances, measuring 12.8% in their series (combining both false positive and false negative results). These authors subsequently developed a scale in which points were assigned for degrees of movement in a number of muscle groups in the arm, and found that with this modification of the cookie test they were able to correctly predict avoidance of interventions more consistently, with only 5.2% of infants with a good score going on to need an intervention. (Michelow 1994) This numerical scale for describing function was the forerunner of the Active Movement Scale, described by members of the same group of authors.

Active Movement Scale
The Active Movement Scale (AMS) was developed at the Hospital for Sick Children in Toronto and was first reported by Clarke and Curtis in 1995. It is designed specifically for use in birth brachial plexus injuries. All previously published reports used adult scales (such as the 0-5 muscle power scale), none of which had been validated in BBPI and were never intended for use in infants. The principle of the AMS is that it assigns a numeric score to each muscle group, based on the observed range of active motion. Each muscle group is observed

while moving in both the gravity-eliminated plane and then in the antigravity plane. A score from 0-7 is assigned based on the observed range. In its original form, the muscle group must demonstrate active movement through its full range in the gravity-eliminated position before it can be given a higher grade. The scale is outlined in Table 2. In their original report, it was presumed that achieving a score of 6 or 7 represented good clinical function. The AMS has subsequently been validated independently, has a high inter-observer reliability and has been correlated with outcome in BBPI (see Chang for review). (Clarke 1995, Chang 2013)

The AMS assessment is performed in our institution by an occupational therapist with specific training in the test. Individual scores are reported for each of fifteen muscle groups (or functions) of the upper extremity. These groups are summarized in Table 3. The AMS is repeated at each clinic visit, but is discontinued once the child reaches school age.

Methods

This is a single center, retrospective study, carried out with IRB approval. All patients were seen in a multidisciplinary Brachial Plexus Clinic, and were assessed by an occupational therapist trained in the administration of the AMS and a team of three physicians (pediatric physiatrist, upper extremity orthopedist and pediatric neurosurgeon). The composition of this team remained unchanged over the course of this study. Intake history and exam was done by each of the physicians, and the AMS assessments were reviewed. A consensus was reached on the management of each infant, with serial visits to the same clinic. If the child showed no evidence of improvement, or reached a plateau in recovery with significant residual deficits, a decision was made to consider primary nerve

surgery. Most of these infants would undergo electromyography prior to surgery. MRI of the plexus was only obtained in those infants with complete plexus injuries without return of function. These were performed to evaluate for root avulsions: if all roots were avulsed, they would not be considered candidates for primary exploration, but still would be followed in the clinic and often required secondary procedures to assist function.

If any child developed evidence of shoulder dysplasia (pain or restriction of movement during passive range of motion of the shoulder), an assessment of the shoulder joint would be added. This most often used ultrasound of the shoulder joint to assess for subluxation, although ossified joints are better evaluated using CT. If evidence for early subluxation was identified, initial intervention would most often be botulinum toxin injection into the internal rotators of the shoulder (pectoralis major and subscapularis) with increased therapy and range of motion exercises. Frank dislocation would be treated with closed reduction and casting, open reduction with soft tissue releases, or open reduction with tendon transfers (latissimus dorsi and teres minor). Follow-up is continued until the child has reached skeletal maturity.

For the purposes of this study, the group of children who required an intervention is composed of those children who underwent primary or secondary nerve surgery, injection therapies, constraint therapy, closed or open reduction of subluxated joints or tendon transfers. All other children, who had good function at last follow-up and did not require intervention, are in the non-intervention group.

Data collection was done by chart abstraction, and the demographics, Narakas type, AMS exams, other test results and therapeutic interventions were collected and managed using REDCap electronic data capture tools hosted at Gillette Children's Specialty Healthcare. REDCap (Research Electronic Data Capture) is

a secure, web-based application designed to support data capture for research studies, providing 1) an intuitive interface for validated data entry; 2) audit trails for tracking data manipulation and export procedures; 3) automated export procedures for seamless data downloads to common statistical packages; and 4) procedures for importing data from external sources. (Harris 2009) The frequencies of interventions were sorted and analyzed by gender, as well as by Narakas type. AMS data were analyzed with a specific goal of correlating return of elbow flexion (ie. achieving a score of 6 or 7 at specific ages) with long–term outcome. Statistical analysis was done using commercially available software (Graphpad software, see graphpad.com/quickcalcs). Contingency tables were analyzed using Fischer's exact test and Peto method for odds ratio and confidence intervals.

Results

Patient population

Between 1997 and 2014, a total of 561 patients were referred for initial evaluation for possible BBPI. A total of 370 patients were excluded, as detailed in Table 4. The remaining 191 patients were all seen for the initial examination between birth and 6 months of age, were assessed in the clinic and had AMS scores recorded, and had at least one year of follow-up exams in the clinic.

Gender distribution

Gender distribution was found to be 70 males, 121 females (36.6% males). Of those children who subsequently required any intervention, the proportion of males was similar: 33 males and 63 females required interventions (ie. 33/96

(34.4%) of treated patients were males, which was not significantly different from the baseline gender distribution (Fisher's exact test, p= 0.794).

Distribution and frequency of interventions by Narakas type

Table 5 summarizes the distribution of the study population by Narakas type. One patient does not fit this classification as the infant had an isolated lower trunk BBPI, which is a rare lesion. As expected, Type 1 injuries are the most common, with decreasing frequency in successive grades. The rates of spontaneous recovery were also established: 75.5% of Type 1 injuries recovered without intervention, as compared with 24.6% of Type 2 injuries (p<0.0001). When Type 1 and 2 patients were combined and compared with the combined Type 3 and 4 patients, the frequencies of spontaneous recoveries were 93/167 (55.7% of Type 1 and 2 injuries) versus 2/23 (8.7% of Type 3 and 4 injuries), which was significantly different (p<0.0001, Fischer's exact). For the overall study population, spontaneous functional recovery (defined as having no intervention during the observed follow-up period) occurred in 95 of 191 patients, or 49.7%.

When the surgical findings on primary nerve surgery were assessed as a function of increasing Narakas type, no significant findings were identified. For example, since the C5, C6 and upper trunk is the anatomy common to all four types within this classification, the frequency of severe, non-conducting nerve injuries (which will require repair or grafting) was examined. Although the frequency of such injuries increased with Narakas type, these were not significantly different: non-conducting lesions were found in the C5/C6 roots or upper trunk in 22.2% of Type 1 lesions, 24.6% of Type 2 lesions, 62.5% of Type 3 lesions and 66.7% of Type 4 lesions (no pair was significantly different when tested).

Active Movement Scale scores and spontaneous return of function

Because of the small numbers of patients with more extensive injuries, this portion of the analysis was restricted to those patients with Narakas Type 1 or 2 injuries only. For the remaining patients, the best score obtained for elbow flexion was assessed for each subject in three distinct age groups: 0-3 months, 3-6 months and 6-9 months. As a function of when some of these infants returned for clinical assessment, the number of assessments per child in each age range varied, and some infants were assessed more than once (ie. sampling was with replacement). In each group, the achievement of a normal score for elbow flexion (EF = 7) or a functional score (EF = 6 or 7) was then correlated with the need for subsequent intervention vs. spontaneous recovery.

Results for the Narakas Type 1 injuries are summarized in Table 6. In any of the age groups tested, the achievement of either a full score for elbow flexion or a functional score for elbow flexion is associated with a significantly reduced risk of the need for a subsequent intervention of any type. For those infants achieving a full elbow flexion score, the odds ratio of requiring a subsequent intervention ranged from 0.196 for the infants aged 0-3 months to 0.118 for those infants assessed between 3 and 6 months. For those infants with spontaneous functional recovery (elbow score of 6 or 7), the odds ratio is also significantly reduced when compared with infants with lower scores for elbow flexion, ranging from 0.138 for the infants aged 0-3 months to 0.0435 for those assessed between 6 and 9 months.

Results for the Narakas Type 2 injuries are summarized in Table 7. For infants achieving a full elbow flexion score (EF =7) before the age of 3 months, the odds ratio was not significantly lowered (OR =0.188 but p= 0.235). For the remaining

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infants with full or functional elbow flexion scores, the risk reductions were highly significant, with the OR ranging from 0.025 to 0.058, with p values between <0.0001 and 0.023. This reflects the fact that only 47 of infants with Type 2 injuries were assessed between birth and 3 months. By the age of 6 months or 9 months a larger number of infants had been assessed (60 and 54, respectively) and a significant difference was detectable.

Thus in either Type 1 or Type 2 injuries, the early return of full elbow flexion or functional elbow flexion was significantly correlated with avoidance of any intervention during the follow-up period.

Discussion

Population and Gender Distribution

The study population was derived from the entire known patient population seen in this clinic. The majority of exclusions were because the initial assessment did not include an AMS, either because the exam was not in use at that time or because the infant was initially seen at an age that was too late to include early AMS. Of those infants excluded because of a different final diagnosis, this was most commonly one of the subtypes of cerebral palsy.

Gender distribution

A minority of the infants were boys, but this ratio was also observed among those infants requiring interventions. Interestingly, in a number of articles concerning BBPI, sex distribution was usually either even or with a slight preponderance of males. (Michelow 1994, Capek 1998, Gosk 2014, Hulleberg 2014). None of these reports describe large populations, but in none was a significant excess of

females noted like that observed in our series. Any attempt to explain this phenomenon would be purely speculative.

Distribution and interventions by Narakas type

The frequency of spontaneous recovery observed across the study population is similar to that reported elsewhere, but remains at the low end of that range (49.7%, as compared with 52-90% in the previously cited studies). (Andersen 2006, Clarke 1995, Foad 2009, Ali 2014) One possible reason for this low rate of recovery might be reflected in the aggressive nature of management in our series. Given the availability of the entire range of possible interventions in this clinic, it is possible that fewer patients were considered to have spontaneous recovery when we were including some later interventions not routinely available. For example, any child requiring later non-surgical treatments for elbow contracture (such as serial casting, splinting or Botox injection) would be considered a failure of spontaneous recovery. Another issue is the potential introduction of bias by the clinician's decision-making. Although the general sense is that clinical evidence of return of function, combined with AMS scores in the "functional" range of 6-7 on exam, is considered "spontaneous recovery," the providers in clinic ultimately defined spontaneous recovery as being those children for whom no intervention was needed by their own judgement.

When analyzed by Narakas subtype, the expected finding was confirmed, with a greater frequency of spontaneous recovery in patients with the lower severity injuries. This was found to be significant in comparing the Narakas Type 1 vs Type 2 groups, but the small size of the Type 3 and Type 4 groups limits the value of comparing the individual groups. Nevertheless, the frequency of spontaneous recovery in Type 1 and 2 combined was significantly better than that of Type 3 and 4 combined (55.7% vs 8.7%, p<0.0001). The surgical findings

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suggest a trend towards more severe injury of the common anatomical elements (ie. the C5 root, the C6 root and the upper trunk), but not in a significant fashion. These findings are very similar to those identified in the meta-review by Foad et al, who also found that the rate of spontaneous recovery was significantly greater in Narakas Type 1 and 2 patients than in the combined Type 3 and 4 patients. (Foad 2009)

Active Movement Scale scores and spontaneous return of function

The fundamental finding in this portion of this study is that early return of elbow flexion, whether full scale or the functional equivalent, is indeed strongly associated with subsequent spontaneous recovery and the ability to avoid other interventions. This seems to be an even stronger association than that observed with the "Cookie test," whose limitations were previously noted by Michelow et al. (Michelow 1994). One possible reason for this is that to obtain a full score on the AMS for elbow flexion, or even a functional score, the shoulder needs to have an underlying degree of external rotation function present. Specifically, if the shoulder is fully internally rotated and external rotation is absent, the full range of elbow flexion cannot be obtained, as the flexing arm cannot clear the infant's abdomen and hence a full range movement is not achieved. Thus the normal range of elbow flexion predicts that the shoulder can be externally rotated, and thus good elbow flexion is a surrogate marker for external rotation. The overall finding that spontaneous recovery of elbow function is correlated with avoidance of later interventions is also supported by the observations of Foad et al. (Foad 2009)

Future considerations

If spontaneous recovery of the infant with a BBPI can be made at an early clinical assessment, clinicians should then be able to determine which infant requires close follow up and which can be safely discharged from clinic. From this study, it is confirmed that the combination of both the Narakas type and assessment of early recovery of elbow flexion can contribute to this decision. For example, the Narakas type 1 patient who achieves an elbow flexion score of 6 or 7 by nine months of age is not likely to require an intervention. Subsequent follow-up could then be either deferred to the primary provider or the child can be seen at greater intervals in a specialty clinic. Conversely, the early identification of a significant deficit in a child with a Narakas type 3 or 4 should be recognized as indicating a need for close follow-up, and the family can be warned of the greater likelihood of needing an intervention.

Before fully implementing an algorithm based on these findings, it would be appropriate to validate the findings further. One proposal for a future study would be to sample patients from another clinic that uses the AMS, and then attempt to predict if the infant did or did not subsequently require an intervention. We are proposing to do this by using records from patients at an outside institution that uses the AMS tool and a similar clinical paradigm. Another validation tool, which has been proposed, is to contact patients who were believed to have made full recoveries and were subsequently discharged from clinic. Patients (or parents) will then be then asked to complete the Pediatric Outcomes Data Collection Instrument (PODCI), which has been validated for use in the assessment of BBPI (see Chang et al). (Chang 2013) The purpose of this would be to confirm that patients who were discharged from follow-up early on had long-lasting results without functional compromise. If these can help to confirm and validate our ability to predict a child's outcome, then further work can be done to develop a specific outcome prediction instrument.

Table 1: Narakas classification of brachial plexus injuries

Type	Roots involved	Trunks involved
1	C5, 6	Upper
2	C5, 6, 7	Upper + middle
3	C5, 6, 7, 8, T1	All
4	C5, 6, 7, 8, T1	All plus Horner syndrome

Table 2: Active Movement Scale

Gravity-eliminated:

Score	Observed movement
0	No response
1	Twitch, without movement
2	< Half range
3	>Half range
4	Full range

Antigravity:

Score	Observed movement
5	<Half range
6	>Half range
7	Full range

(Clarke 1995)

Table 3: Muscle functions tested using the AMS assessment

Upper trunk:
   Shoulder flexion
   Shoulder internal rotation
   Shoulder external rotation
   Shoulder abduction
   Shoulder adduction
   Elbow flexion
   Supination

Middle trunk:
   Elbow extension
   Wrist extension

Lower trunk:
   Pronation
   Wrist flexion
   Finger extension
   Finger flexion
   Thumb extension
   Thumb flexion

Table 4: List of exclusion criteria

Reason for exclusion	Number
Initial visit after age 6 months	166
Initial visit prior to introduction of AMS	82
Lost to follow-up	77
Short follow-up (< 1 year)	24
Final diagnosis not BBPI	21
Total	370

Table 5: Distribution of patients and rate of spontaneous recovery by Narakas type

Narakas type	Number (%)	Spontaneous recovery (%)**
1	102　(53.4)	77/102　(75.5)
2	65　(34.0)	16/65　(24.6)
3	16　(8.4)	2/16　(12.5)
4	7　(3.7)	0/7　(0.0)
Other*	1　(0.5)	0/1　(0.0)
Total	191	95/191　(49.7)

*Patient with isolated lower trunk BBPI

**Rate of spontaneous recovery significantly higher for Type 1 vs Type 2 (p<0.0001) but not Type 2 vs Type 3 (p=0.503)

Table 6: Does return of spontaneous elbow flexion predict decreased risk of
intervention in Narakas type 1 patients?

Normal elbow flexion score on AMS (EF =7) and risk of subsequent intervention,
by age group

Age (months)	Number (n)	OR (95 CI)	P value	Sensitivity (%)	Specificity (%)	Prevalence (%)	PPV	NPV
0-3	84	0.196 (0.056-0.693)	0.0087	31.9	100	17.9	100	24.2
3-6	92	0.118 (0.045-0.314)	<0.0001	51.4	100	23.9	100	39.3
6-9	65	0.121 (0.039-0.377)	<0.0001	50.0	100	26.1	100	41.5

(OR= odds ratio of a child with EF =7 requiring an intervention, compared with
those with lower EF scores in this age interval, PPV = positive predictive value,
NPV = negative predictive value)

Functional elbow flexion score on AMS (EF =6 or 7) and risk of subsequent
intervention, by age group

Age (months)	Number (n)	OR (95 CI)	P value	Sensitivity (%)	Specificity (%)	Prevalence (%)	PPV	NPV
0-3	84	0.138 (0.044-0.430)	0.0003	47.8	100	17.8	100	29.4
3-6	92	0.104 (0.040-	<0.0001	70.0	86.3	23.9	94.2	47.5

		0.271)						
6-9	65	0.0435 (0.014-0.134)	<0.0001	81.2	94.1	26.2	97.5	64.0

Table 7: Does return of spontaneous elbow flexion predict decreased risk of intervention in Narakas Type 2 patients?

Normal elbow flexion score on AMS (EF =7) and risk of subsequent intervention, by age group

Age (months)	Number (n)	OR (95 CI)	P value	Sensitivity (%)	Specificity (%)	Prevalence (%)	PPV	NPV
0-3	47	0.188 (0.016-2.25)	0.235	12.5	100	48.9	100	52.3
3-6	60	0.0267 (0.0022-0.331)	0.023	18.2	100	63.3	100	67.9
6-9	54	0.039 (0.0084-0.180)	0.0002	57.1	100	61.1	100	78.6

(As per notes for Table 6, above)

Functional elbow flexion score on AMS (EF =6 or 7) and risk of subsequent intervention, by age group

Age (months)	Number (n)	OR (95 CI)	P value	Sensitivity (%)	Specificity (%)	Prevalence (%)	PPV	NPV
0-3	47	0.058 (0.011-0.319)	0.0026	29.1	100	48.9	100	57.5
3-6	60	0.025 (0.0052-0.123)	<0.0001	45.4	100	63.3	100	24.5
6-9	54	0.048 (0.012-0.189	<0.0001	76.2	97.5	61.1	94.1	86.5

<u>Part 3: Future directions</u>

This study is limited by its retrospective nature, and by being developed from the experience of a single center. The necessary next step in this would be to validate the findings of this work. Internally, this could be done by identifying the elbow flexion status of infants before 6 months of age and following to see if an intervention is subsequently needed (ie. establish a prospective cohort). However, because the results of earlier AMS assessments would be known to the providers in clinic, this could not be blinded in any way that would limit the introduction of bias in decision-making. Hence, it would be preferable to extend this model to another BBPI MDC, with similar diagnostic and therapeutic capabilities, and see if a sampling of early AMS exams correlates with the need for subsequent interventions.

It would also be strongly worthwhile to establish an outcome measure for use in this clinic population, preferably a patient-reported outcome. As was noted in Chang's review article, both the Pediatric Outcome Data Collection Instrument (PODCI) and the Pediatric Evaluation of Disability Inventory (PEDI) have been cited in the literature on reviews of BBPI management. However, only PODCI has been specifically validated for BBPI. (Chang 2013, Haley 2010) The author is considering studying a sample of children from the data set used in this study, specifically those who were discharged form clinic after having been identified as having had a good spontaneous recovery. The upper extremity portion of PODCI and the daily cares domain of PEDI would be administered to these patients (either by patient report or parent as proxy) and an overall measure of functional outcome could be obtained.

Once an adequate, validated measure of functional outcome is established, in combination with the use of the standardized AMS assessment, it should be possible to compare patient results at different BBPI clinics or practices and work to establish the efficacy of MDCs for this disorder.

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