Long-Term Outcome Following Carpal Wedge Osteotomy in the Arthrogrypotic Patient

Christian A. Foy, MD, Janith Mills, MPAS, Lesley Wheeler, BA, Marybeth Ezaki, MD, and Scott N. Oishi, MD

Investigation performed at Texas Scottish Rite Hospital for Children, Dallas, Texas

Background: Wrist flexion and ulnar deviation deformity is a common presentation in children with amyoplasia congenita. Multiple surgical procedures have been reported to correct the deformity to enhance functional independence and improve quality of life. We performed a retrospective review to detail our long-term results with carpal wedge osteotomy in these patients.

Methods: Medical records of all patients with the amyoplasia form of arthrogryposis who underwent carpal wedge osteotomy between 1994 and 2008 were reviewed. Patients with a follow-up of two years or less were excluded. Preoperative and postoperative resting position and range of motion of the wrist were recorded. Interviews and questionnaires were completed to assess the mean overall satisfaction level of the parent or guardian with the outcome of surgery, function, and task completion with use of parent-guardian surveys, the Manual Ability Classification System, and the ABILHAND-Kids measure of manual ability.

Results: Seventy-five wrists in forty-six patients who met the inclusion criteria were reviewed. The average age of the patients at the time of surgery was 4.3 years (range, nine months to eighteen years; median, 2.7 years). The average duration of follow-up was 5.7 years (range, two to 10.3 years; median, 5.3 years). The average resting position of the wrist postoperatively (11° of flexion) was significantly different from that measured preoperatively (55° of flexion) (p < 0.001). The arc of wrist motion measured preoperatively (32°) did not differ significantly from that measured postoperatively (22°) (p = 0.4903). The location of the motion arc was significantly improved to a more functional position. The average active extension of the wrist changed from –37° preoperatively to –11° postoperatively (p < 0.001). Active wrist flexion also significantly changed from 69° preoperatively to 33° postoperatively (p < 0.001). Parent-guardian surveys indicated that the mean overall satisfaction score after surgery was 9.1 of 10 possible points and that the mean ranking for task completion in activities of daily living was 4 (easier following surgery).

Conclusions: Long-term outcomes reveal that surgical correction of wrist flexion posture in children with amyoplasia congenita results in improvement that is sustained over time. The surveys and questionnaires completed by parents or guardians indicated that they were satisfied with the results of the operation.

Level of Evidence: Therapeutic Level IV. See Instructions for Authors for a complete description of levels of evidence.

Arthrogryposis is a term that is used to describe a group of unrelated diseases with the common phenotypic characteristic of nonprogressive multiple congenital joint contractures. The group of conditions together is called arthrogryposis multiplex congenita. The most well-recognized or “classic form” is termed amyoplasia congenita. Patients with amyoplasia congenita have a congenital syndrome that is characterized by stiffness of multiple joints, with the joints fixed in various positions. The muscles are smaller and reduced in number and are replaced by fibrous tissue. The characteristic positioning of the upper extremity in patients with amyoplasia includes internal rotation of the shoulders, extension of the elbows, flexion and ulnar deviation of the wrist, and stiff fingers with a thumb-in-palm deformity. Performing activities of daily

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living can be difficult, and the child uses bimanual functioning and substitution maneuvers to accomplish necessary tasks.

The goal of upper limb surgical procedures is to enhance functional independence and improve quality of life. This can be accomplished by repositioning the upper extremity in neutral alignment while ideally preserving the limited joint motion. In recent years, different techniques for correction of upper limb deficiencies in patients with amyoplasia have been reported in the literature. A tricepsplasty can be performed to correct fixed elbow extension contracture, a transfer involving the long head of the triceps can provide active elbow flexion, a thenar muscle release or “slide” can bring the thumb out of the palm, local rotation flaps and soft-tissue reconstruction can provide better digital extension, and tendon transfers and/or osteotomies can improve the flexion deformity of the wrist.

The correction of wrist flexion deformities with use of carpectomy has been described since the 1940s. Early reports of wrist flexion deformity correction in patients with amyoplasia recommended proximal row carpectomy or distal radial exten-

sion osteotomy. Both proximal row carpectomy and distal radial extension osteotomy further limit or alter motion but do not correct the wrist flexion deformity. The technique of carpal wedge osteotomy has been described more recently. The procedure is based on the presumption that, in the untreated wrist in the child with amyoplasia, multiple carpal coalitions obliterate the midcarpal joint, resulting in most of the wrist motion occurring through the radiocarpal joint (Figs. 1-A and 1-B). With the performance of a biplanar wedge resection of the impending carpal coalition, the wrist flexion deformity is corrected to a position of neutral alignment while still allowing for motion at the intact radiocarpal articulation (Figs. 2-A and 2-B). Recent literature reports have indicated that dorsal carpal wedge osteotomy can significantly improve wrist extension while preserving wrist motion.

The purpose of this study was to analyze the long-term outcome of dorsal carpal wedge osteotomy for the treatment of wrist flexion deformities in children with the amyoplasia form of arthrogryposis multiplex congenita. We reviewed the efficacy of dorsal carpal wedge osteotomy in our patients by determining the ability of the procedure to initially correct the flexion deformity, maintain wrist motion, and maintain the correction over time. We also assessed the functional outcome of patients with use of objective as well as subjective methods.

**Materials and Methods**

The medical records of all children with the amyoplasia form of arthrogryposis who presented to Texas Scottish Rite Hospital for Children in Dallas, Texas, and were treated with a dorsal carpal wedge osteotomy (as described by Ezaki and Carter) between the dates of January 1, 1994, and December 31, 2008, were retrospectively reviewed. The procedure as described involves a biplanar wedge resection of the carpus, wider radially than ulnarly, that will preserve the intact radiocarpal joint while reducing the resting ulnar deviation position of the wrist (Fig. 3). Tendon transfers of the extensor carpi ulnaris to the extensor carpi radialis brevis aid in providing more extension power and eliminating some of the ulnar-deviated force. Volar procedures in the distal part of the forearm include evaluation and release of fascia as well as fibrotic wrist flexors to reduce the flexion deformity of the wrist, as well as decompression of fascial compartments to reduce postoperative intracompartmental pressures.

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**Fig. 1-A** Posteroanterior (Fig. 1-A) and lateral (Fig. 1-B) radiographs, made when the patient had reached skeletal maturity, showing an untreated arthrogrypotic wrist. Note the pancarpal coalition that is present.

**Fig. 2-A** Posteroanterior (Fig. 2-A) and lateral (Fig. 2-B) radiographs, made when the patient had reached skeletal maturity, showing an arthrogrypotic wrist that had been treated with carpal wedge osteotomy.
Patients with rigid wrist flexion deformity not responsive to stretching or splinting were included in the study. These patients were managed postoperatively with casting for six weeks and splinting for a minimum of six months. Patients who underwent dorsal carpal wedge osteotomy for treatment of a diagnosed disease other than amyoplasia and patients with a follow-up of less than twenty-four months were excluded from the study. Concomitant procedures included thenar muscle release, elbow releases, and tricepsplasties, when indicated. The surgical procedures were all performed by one of three experienced staff hand surgeons. With use of a goniometer, preoperative and final postoperative wrist resting position and wrist flexion and extension were recorded by the hand fellow or physician assistant and verified by one of the staff hand surgeons. These results were then reviewed with use of a paired t test.

A parent or the guardian of each child who participated in the study group completed a follow-up parent-guardian survey by telephone. Parent-guardian survey scores were assigned on the basis of the overall satisfaction of the parent or guardian with the surgical outcome (on a scale of 0 [not satisfied] to 10 [very satisfied]), on the overall appearance of the wrist (on a scale of 0 [no improvement] to 10 [great improvement]), and on the level of function of the wrist (on a scale of 0 [no improvement] to 10 [great improvement]). Other questions targeted manual ability as well as the ability to perform specific tasks postoperatively. For the specific task assessment, the parents were asked to evaluate their child’s ability to perform activities of daily living, such as eating, keyboarding, working at a desktop, tasks of personal hygiene, and ambulation, on a scale of 1 (difficult) to 5 (easy), depending on their perception of how difficult or easy it was for their child to perform the activity following surgery.

Parents’ questionnaires included two different scoring systems: the Manual Ability Classification System (MACS)\(^\text{13}\) and the ABILHAND-Kids\(^\text{14-16}\). The MACS is a validated scoring system for cerebral palsy that can be useful in assessing other neuromuscular disorders, such as arthrogryposis. The purpose of the MACS is to provide a systematic method to classify how children with cerebral palsy use their hands when handling objects in daily activities. It is based on self-initiated manual ability in an individual’s personal space. The focus is to determine which level best represents the child’s usual performance in home, school, and community settings. The level is determined by someone who knows the child well and not by conducting a specific assessment. The classification system describes five levels of ability, with level I indicating that the child can handle most kinds of objects easily and successfully and level V indicating that the patient cannot handle objects and has a severely limited ability to perform even simple actions\(^\text{13}\).

ABILHAND-Kids was developed as a measure of manual ability in a sample of children with cerebral palsy. It explores the most representative inventory of manual activities and is based on the perceptions of the parent or guardian. ABILHAND-Kids was originally developed with use of the Rasch measurement model. It allows the conversion of ordinal scores into linear measures located on a unidimensional scale. Parents are asked to fill out the questionnaire by estimating their child’s ease or difficulty in performing each activity when the activities are performed without technical or human help, irrespective of which upper limb is used or the type of strategy that is used.

![Graph showing the mean rating on activities of daily living on the specific task assessment. 1 = much more difficult following surgery, 2 = more difficult following surgery, 3 = no change, 4 = easier following surgery, and 5 = much easier following surgery.](http://jbjs.org)
Fig. 5
Graph showing the mean ranking for the level of performance at home, at school, and in community settings, with use of the Manual Ability Classification System (MACS). I = Handles objects easily and successfully; II = Handles objects, but with somewhat reduced quality and/or speed of achievement; III = Handles objects with difficulty—needs help to prepare and/or modify activities; IV = Handles a limited selection of easily managed objects in adapted situations; and V = Does not handle objects and has a severely limited ability to perform even simple actions.

Fig. 6
Graph showing the results of the ABILHAND-Kids measure of manual ability.
They must rank the child’s ease or difficulty according to a three-level scale in which the choices are “impossible,” “difficult,” or “easy.” “Impossible” describes a child who is unable to perform the activity without any other help, “difficult” describes a child who is able to perform the activity but with some difficulty, and “easy” describes a child who is able to perform the activity without difficulty.13

Source of Funding
No external funding source was used for this study.

Results
Carpal wedge osteotomies were performed in ninety-eight wrists (fifty-eight patients) during the study inclusion period. Seventy-five wrists (forty-six patients) had a follow-up of greater than two years. Of these, twenty-nine patients underwent bilateral procedures. The average age at time of surgery was 4.3 years (range, nine months to eighteen years; median, 2.7 years). Eighteen wrists were in patients who were younger than two years, forty-one wrists were in patients whose age fell within the range of two to five years, fourteen wrists were in patients whose age fell within the range of six to twelve years, and two wrists were in patients whose age fell within the range of thirteen to eighteen years. The average follow-up was 5.7 years (range, two to 10.3 years; median, 5.3 years). Seventeen patients were followed to skeletal maturity. Thirteen patients had additional surgery, mostly including thenar slides, tricepsplasties, and elbow releases. When comparing the resting position of the wrist, the mean final postoperative resting position of the wrist (11° of flexion) was significantly different from the mean preoperative resting position of the wrist (55° of flexion) (p < 0.001). The arc of wrist motion measured preoperatively (32°) did not differ significantly from that measured postoperatively (22°) (p = 0.4903). The location of the motion arc was improved to a more functional position. The average preoperative range of motion of 69° to 37° of flexion changed to a range of motion of 33° to 11° of flexion postoperatively (p < 0.001).

Data obtained from the twenty-one different tasks assessed in the ABILHAND-Kids measure of manual ability revealed that their long-term results with carpectomy had generally been unsatisfactory despite excellent intraoperative correction and prolonged postoperative immobilization.

Discussion
The initial goal in the treatment of infants who have the amyoplasia form of arthrogryposis is to enhance functional independence and improve quality of life. For stiff upper limbs, the objective is to passively flex the elbows and extend the wrists. Treatment of flexion contracture of the wrist usually begins with stretching, followed by a period of splinting. The lower extremities are treated concomitantly due to the importance of a supportive structure by the age of eighteen months. While treatment of the lower extremities is underway, passive stretching and splinting of the elbows and wrist can provide motion to a stiff joint. If sufficient motion is obtained, and the patient’s wrist can be brought to a neutral position, functional bracing can then be instituted to develop normal use patterns. Bracing can then be appropriately replaced by tendon transfers in the future. Dorsal carpal wedge osteotomy is reserved for patients with rigid flexed wrist joints that are unresponsive to passive stretching and splinting. Due to the nature of the condition, many of the joint contractures are resistant to cast correction and soft-tissue surgery.

Early proponents of correcting flexion deformity of the wrist in patients with arthrogryposis suggested the use of proximal row carpectomy or distal radial and ulnar extension osteotomy. Mennen reported on twenty-five patients (forty-seven limbs) who had a diagnosis of arthrogryposis multiplex congenita (age range, three months to nine years) and underwent proximal row carpectomy for the treatment of flexion deformity of the wrist. The average follow-up period was 7.5 years. Results from the surgery revealed that the active range of wrist motion was from 10° of flexion to 40° of extension (average active range of motion, 27°). One patient underwent distal radial and ulnar extension osteotomies. The flexion deformity recurred two years after surgery. Mennen did not recommend this procedure as an alternate method for correcting the wrist deformity. He concluded that wrist surgery is better performed at the age of three to six months, which is an age when carpal bones are not ossified and the procedure is easier with less bone resection needed. A prerequisite for successful proximal row carpectomy is an intact capitae head and lunate fossa. In the untreated wrist in the child with amyoplasia, multiple carpal coalitions obliterate the midcarpal joint and can be seen as the corpus ossifies. Because of this, wrist motion only occurs at the radiocarpal joint. Resection of the articular surface of the proximal aspect of the carpus in this case will ultimately result in further stiffening.

Wenner and Saperia reported on five patients with arthrogryposis who underwent proximal row carpectomy for the correction of flexion deformity of the wrist. The average age of the patients at the time of the operation was eleven years (range, eight years and nine months to twelve years). Three of the five patients subsequently underwent arthrodesis for correction of recurrence of deformity. They concluded that, despite the lack of experience with distal radial and ulnar extension osteotomy for the correction of flexion deformity of the wrist, they would prefer this surgery to proximal row carpectomy. They reported that their long-term results with carpectomy had generally been unsatisfactory despite excellent intraoperative correction and prolonged postoperative immobilization.

Van Heest and Rodriguez recently reported their experience with twelve patients (twenty wrists) who had undergone...
dorsal carpal wedge osteotomy and who together had an average follow-up of forty-five months. In their study there is no report of the resting position of the extremity preoperatively or postoperatively, but the patients did obtain an average postoperative arc of motion that was centered over neutral alignment, with a total range of motion that did not differ significantly from its preoperative value. These authors reported better outcomes for patients who underwent surgery after the age of seven years, and they also reported improved extension for patients who underwent extensor carpi ulnaris transfer at the time of surgery.

The technique for carpal wedge osteotomy was initially described by Ezaki and Carter in 2004. In their article, the authors presented the results of forty-two wrists in twenty-four patients who together had an average follow-up of eighteen months. Patients’ average resting position changed from 58.5° of flexion preoperatively to 11.2° of flexion postoperatively. Their arc of passive wrist motion decreased by an average of 10° but remained centered over neutral alignment for bimanual activities. We are now reporting long-term outcome in seventy-five wrists that underwent carpal wedge osteotomy. The mean final postoperative resting position of the wrist was 11° of flexion, with a total arc of motion of 22°, at a mean follow-up time of sixty-eight months. These data are consistent with the previous short-term data reported above and shows that results can be maintained over the long term with use of this procedure. Furthermore, parent-guardian surveys showed that overall satisfaction was 9.1 of 10 possible points, with task completion being facilitated by the procedure. Importantly, parents reported that tasks requiring bimanual positioning in the frontal plane, such as opening a breadbox, filling a glass with water, and unwrapping a chocolate bar, were improved following the procedure. However, because of the nature of the underlying diagnosis, parents reported that children still had a difficult time performing tasks such as jar opening, buttoning shirts and/or trousers, and snapping a belt, which are all tasks that require strength or a higher level of dexterity.

The present study had several limitations. First, it was retrospective in design and patient follow-up for the assessment of satisfaction and outcome measures was done via telephone interviews. Second, preoperative objective measures on manual abilities were not recorded for comparison after surgery. Third, the patient population was not surveyed directly; rather, we relied on the perceptions of a parent or guardian, which may not correlate with the patient’s reality. Fourth, no control group exists to compare carpal wedge osteotomy with other treatment options such as splinting, tendon transfers, osteotomy of the radius, or proximal row carpectomy. Fifth, concomitant procedures, including tenar slides, elbow releases, and tricepsplasies, could potentially influence the overall outcome of patients after surgery. Finally, the results of our study do not allow us to conclude that the age of the patient at the time of surgery affects the surgical outcome.

In conclusion, dorsal carpal wedge osteotomy is an effective method for treating flexion contracture of the wrist in patients with arthrogryposis. Correction of the flexion posture of the wrist was improved, and this improvement was sustained over time. This operation shifted the arc of wrist motion to a more useful position and did not reduce range of motion to enhance functional independence and improve quality of life.

References