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Outcomes at 2-Year Minimum Follow Up of Shoulder, Elbow and Wrist Surgery in Individuals with Arthrogryposis Multiplex Congenita

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Abstract

Background: Arthrogryposis Multiplex Congenita (AMC) is a rare condition that has not been extensively studied, and the need for further research has been strongly emphasized. Despite substantial functional impact resulting from AMC, minimal research has been completed regarding mid- to long-term outcomes in this population. The objective of this systematic review was to identify existing knowledge concerning outcomes of shoulder, elbow and wrist surgery with a minimum two year follow-up in individuals with AMC.

Methods and findings: A search was conducted in Medline, Embase and Web of Science databases and a hand search of the bibliographies of articles relevant to the research question was completed. Inclusion criteria were English articles with AMC participants only (excluding syndrome-related AMC) who underwent shoulder, elbow or wrist surgeries with a minimum follow-up of two years. 1289 articles were identified through electronic database and hand searches; after review, 12 articles met all inclusion criteria. No articles were identified that described shoulder surgery outcomes in those with AMC. Elbow surgeries discussed were posterior capsulotomy and triceps lengthening, pectoralis major muscle transfer, gracilis muscle transfer, Steindler flexorplasty, and brachial nerve exploration with or without nerve transfer. Wrist surgeries included proximal row carpectomy, osteotomy of radius and ulna, and dorsal carpal wedge osteotomy.

Conclusions: This systematic review identified those procedures with lasting benefit, those that resulted in additional complications or surgeries, and those which are currently recommended as treatment in patients with AMC. These conclusions were based on a small number of articles with low levels of evidence due to the obstacles of performing higher-level studies in this rare, heterogeneous population. To facilitate treatment and to prevent adverse effects on patient quality of life and strain on healthcare and social system resources, additional and continued research is necessary in this population. AMC is a rare congenital condition regarding which further research is required to ensure appropriate and successful treatment as well as expansion of the existing knowledge base.

Keywords: Amyoplasia; Arthrogryposis; Arthrogryposis Multiplex congenita; Upper limb surgery

Introduction

Arthrogryposis multiplex congenita (AMC) or arthrogryposis is a rare, congenital, non-progressive condition presenting with soft tissue contractures affecting two or more joints [1]. AMC occurs between 1:3000 and 1:5000 live births [2], with approximately equal gender ratio [3]. Nearly 400 different conditions are classified within AMC [4]. The upper extremities are involved in 70% of patients [5], and the most common pattern presents with shoulder adduction and internal rotation, elbow extension contractures, forearm pronation, wrist flexion and ulnar deviation, and rigid fingers with a thumb-in-palm deformity [5-7]. The less common presentation has similar deformities of the shoulder and wrist but fixed elbow flexion [5,6].

The elbow joint is thought to be most critical for essential function of the upper limb, allowing self-feeding, self-care of the face and hair, and independent toileting [5,7]. Joint contractures resulting from AMC can often affect these essential functions [8]. Early splinting and range of motion treatment has been shown to be effective management and the majority of children do not require surgical treatment [9]. Surgical procedures are only indicated if these measures fail and basic activities of daily living (ADLs) require assistance [6]. Surgical goals are to improve quality of life and functional independence, accomplished by repositioning the upper limb in neutral alignment while maintaining any joint motion already present [10].

The lack, and subsequent need, of research regarding AMC has been emphasized frequently by those prominent in the research community. At the 2nd International Symposium on Arthrogryposis, there was an emphasis on research specific to long-term outcomes in adults with arthrogryposis [11]. A recent

international study of 177 adults with AMC-the largest AMC-focused study to date-found that patients had an average of nine surgeries throughout their lives, and 29% of the surgeries were specific to the upper extremity [12].

Due to the nature of the study, it was not possible to determine outcomes of specific surgeries. Currently, analysis of surgical management results in this population still relies on pre-post studies using upper extremity outcome measures. A systematic review is needed to know which surgeries are worth targeting in this population, though the authors recognize a priori that only small sample sizes are likely. For this systematic review, the primary research question was what is the functional outcome, a minimum of two years later, following shoulder, elbow or wrist surgery in those with AMC?

Materials and Methods

The Preferred Reporting Items for Systematic Reviews and Meta-Analysis (PRISMA) checklist and guideline were used to develop this review [13]. Medline, Embase and Web of Science databases were searched from January 1970 to June 8, 2016. Peer-reviewed articles were identified using key words arthrogryposis multiplex congenita, arthrogryposis, amyoplasia, surger*, and surgical.

No key words were used regarding shoulder, elbow or wrist to ensure the largest quantity of articles identified. References of relevant studies were also searched. Inclusion criteria for this review were: English only; participants with AMC (excluding syndrome-related AMC); reported shoulder, elbow or wrist surgery, and; reported the functional outcome of the described surgery a minimum of two years later. No age limit was defined.

1289 articles were found by the primary electronic and hand searches (Figure 1). After duplicate removal, 904 articles remained. The titles and abstracts of the 904 articles were reviewed and 738 articles were excluded. A full-text review of the remaining 166 articles was completed and the authors determined article eligibility.

From these, 12 articles met the inclusion criteria while 154 articles were excluded. 11 articles were identified by the electronic search, [1,5-8,10,14-18] while one study was found by hand search (Figure 1) [19].

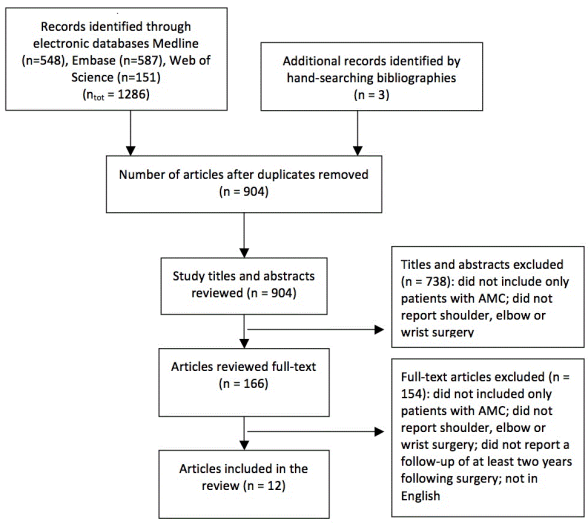

Figure 1: Flow chart of literature search and study selection.

Information related to the research question was extracted from the reviewed articles by the authors independently; consensus was achieved following. The extracted data included number of participants, number of procedures, type of AMC, mean age at surgery, mean follow-up time, surgery performed, surgery outcomes a minimum of two years later, if the measured outcomes were standardized, and any complications.

The level of evidence guidelines from the Centre for Evidence-Based Medicine (CEBM) were used to determine the level of evidence for the reviewed articles [20].

Results

Twelve articles that met all inclusion and exclusion criteria were included, [1,5-8,10,14-19] and all included articles were published as Level 4 evidence.

Demographics

Altogether, the studies included 159 individuals with AMC. Seven studies involved 88 patients diagnosed with AMC but no further classification [6,7,14,15,18,19]. Five studies included 71 patients with the amyoplasia form of AMC [1,5,8,10,16]. The mean age at surgery of individuals in all studies was 5.1 years and mean follow-up was 6.8 years (Table 1).

Table 1: Patient population of the studies reviewed.

Author	Year	N	Location of surgery (elbow, wrist)	Number of surgeries	Type of AMC	Mean Age at Surgery (years)	Mean Follow-Up Duration (years)
Axt et al. [14]	1997	16	Elbow	22	AMC	4.4	12.3
Bahm[15]	2013	2	Elbow	2	AMC	1.0	4.9
Burgess and Robbe [1]	2012	6	Wrist	9	Amyoplasia	Not recorded	5.8

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Chomiak et al. [5]	2014	5	Elbow	9	amyoplasia	6.3	15.1
Doi et al. [16]	2011	2	Elbow	4	amyoplasia	2.2	5.3
Foy et al. [9]	2013	46	Wrist	75	amyoplasia	4.3	5.7
Goldfarb et al. [17]	2004	10	Elbow	17	AMC	7	5
Lahoti and Bell [6]	2005	7	Elbow	10	AMC	5.2	11.4
Mennen [19]	1993	25	Elbow & Wrist	47	AMC	1.5	7.5
Van Heest and Rodriguez [8]	2013	12	Wrist	20	Amyoplasia	8	3.8
Van Heest et al. [7]	2008	23	Elbow	29	AMC	2.9	5.4
Wenner and Saperia [18]	1987	5	Wrist	5	AMC	11	4.6
Totals		159		249		5.1	6.8

Shoulder

There were no studies identified which described functional outcomes after shoulder surgeries with follow-up at least two years later.

Elbow

Three studies were completed studies investigating posterior capsulotomy and triceps lengthening procedures performed to increase passive extension of the elbow [7,14,21]. In all studies, the procedures improved passive elbow flexion and arc of motion, resulting in functional improvement. The more recent study by Van Heest et al. found that 23 of 23 patients were able to reach their mouth passively following surgery and 22 patients could actively flex their elbow for self-feeding [7]. These results were better than the previous study by Axt et al. which showed 17 patients with improved passive flexion, five patients with either loss or no functional gain, and only three patients with active flexion for self-feeding [14]. The differences in results were suggested to be due to longer follow-up time in the Van Heest et al. study [7]. Despite promising outcomes, the results of Van Heest et al. showed that mean extension decreased from -1° to 34° flexion [7], and there were five patients in the Axt. et al. study with poor results but with no functional loss [14]. Regardless, all studies demonstrated an increase in elbow range of motion following surgery and concluded that posterior capsulotomy and triceps lengthening were reliable in successfully increasing both passive elbow flexion and arc of motion (Table 2) [7,14,21].

Table 2: Characteristics and results of studies addressing elbow surgeries.

Author	Surgical Procedure	Standardized Outcome Measure	Outcome Measure	Results Following Surgery	Complications
Axt et al. [14]	a) Posterior capsulotomy and triceps lengthening. b) Additional triceps transfer. c) Only triceps transfer.	No	ROM and if patient could actively or passively reach mouth.	a) Elbow ROM doubled from 20° to 29°. Extension reduced by 39°. Functional position improved. 17 good results: 15 patients able to passively reach mouth, two patients able to actively reach mouth. Five poor results, but no functional loss. b) Mean elbow ROM increased from 60° to 83° and shifted to improved flexion position. c) One decreased ROM, one increased ROM.	None described.
Bahm [15]	a) Exploration to identify functional brachial plexus nerves. b) Above procedure as well as nerve transfer from median nerve to biceps motor branch.	No	Elbow flexion.	a) No improvement in case 1; developed as a typical AMC patient. b) Active elbow flexion was seen.	None described.

Chomiak et al. [5]	Unipolar transfer of three-fifths of pectoralis major muscle.	Modified evaluation according to Atkins:23 grade 1 (excellent), grade 2 (very good), grade 3 (good), grade 4 (satisfactory), grade 5 (unsatisfactory).	ROM, elbow flexion and extension; Medical Research Council muscle strength.	Improved flexion in five elbows. Global improvement of function in five elbows. Flexion deformity developed in four elbows. One grade 2 elbows with 90° flexion and 35° decrease in extension. Two grade 3 results with flexion of 92° and 100°, and lack of extension of 42° and 45°. Two grade 4 results with limited arc of motion between 20° and 45°. Four grade 5 results with arc of motion between 5° and 15°.	No intra- or perioperative complications. Subsequent procedures required in seven extremities: Five extremities required the transferred muscle to be fixed to the alternative tendon of the forearm 10-26 months after the primary procedure. In one patient, the hypertrophic scar was corrected one year after surgery, the tendon of the transferred muscle was lengthened, and the ventral elbow capsule was released 11 years after primary surgery. In one extremity, the tendon pulley was reconstructed for a large cutaneous fold on ventral side of elbow.
Doi et al. [16]	Functioning free muscle transfer with the use of the gracilis muscle.	No	Elbow active flexion.	Case 1: Active elbow flexion and strength increased over time. At 11 years, active elbow flexion 50°-130° on right, 60°-130° on left. Case 2: 21 months following second transfer, able to actively flex elbows < 70° due to elbow contracture. Three years after procedure, active elbow flexion 40°-105° on right, 40°-100° on left. Residual shoulder contracture persisted.	Case 1: Flexion contracture developed due to triceps brachii weakness; total arc of active elbow motion <60°. Case 2: Posterior capsulotomy of elbow with triceps lengthening performed to improve flexion.
Goldfarb et al. [17]	Steindler flexorplasty.	No	Individual skill performance post-operation.	Mean elbow flexion improved from not possible to 85°. Average 27° of elbow extension was lost, but without negative affect on ADL performance. 9/10 patients satisfied with procedure.	In one elbow, the screw attaching flexor pronator mass to anterior humerus dislodged; resolved with revision surgery with a longer immobilization period. In one patient, ulnar nerve paresthesias developed immediately after surgery and resolved by first postsurgical day. Surgery on the contralateral elbow resulted in development of more considerable paresthesia that did not resolve spontaneously; ulnar nerve transposition was required to resolve symptoms.
Lahoti and Bell [6]	Modified pectoralis major muscle transfer.	No	Self-feeding.	Initially, 6/7 patients retained useful power. Gradual and progressive increase in flexion deformity, > 90° in all cases. 8/10 elbows also had decrease in arc of flexion.	None described.
Van Heest et al. [7]	Posterior capsulotomy and triceps lengthening.	No	Flexion and extension, arc of motion.	Mean extension improved from -1° to 34°; mean flexion improved from 38° to 100°. All patients achieved an arc of motion of at least 90° (mean increase 33°). All patients could reach mouth passively; 22/23 patients could independently feed themselves. No additional tendon transfers were required to further improve flexion.	None described.

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Two papers reported pectoralis major muscle transfers to improve active elbow flexion. Both studies found that results declined over time due to persistent flexion deformity development [5,6]. Chomiak et al. showed improvement in 55% of operated elbows. However, five elbows required additional surgeries to adjust the transferred muscle tendon, one tendon required lengthening in addition to posterior capsulotomy, and the tendon pulley was reconstructed in one elbow due to development of a large cutaneous fold on the ventral side of the elbow [5]. Lahoti and Bell showed that although six of seven patients initially retained useful power, progressive flexion deformity developed with decreased arc of flexion in eight of ten elbows [6]. Chomiak et al. recommended bilateral transfer in patients with existing passive elbow extension greater than 90°, [5] and both studies proposed the procedure be performed unilaterally in all other patients to prevent loss of extension in both elbows. Despite a flexion deformity, pectoralis major transfer was shown to be effective in improving active flexion in the majority of patients in both studies [5,6].

One case study addressed results of free muscle transfer using the gracilis muscle. The first patient showed increased active elbow flexion and power over time, but also development of a flexion contracture causing the arc of active elbow motion to be <60°. The second patient showed a more severe flexion contracture and required a posterior capsulotomy and triceps lengthening to restore extension. The study concluded that despite flexion contracture development, when compared to other procedures free muscle transfer achieved sufficient function without suitable donor muscles for pedicle transfer [16].

One study analysed results of Steindler flexorplasty to increase active elbow flexion. Mean elbow flexion improved while elbow extension was lost, but without a negative effect on ADL performance. In one elbow, a screw attaching the flexor pronator mass became dislodged but was resolved with revision surgery. In another patient, ulnar nerve paresthesia developed after surgery on each elbow, with spontaneous resolution in one elbow but requiring ulnar nerve transposition in the second. The results obtained were similar to that of other procedures, and was noted by the authors to be less surgically extensive than other muscle transfers [17].

One case study addressed results of brachial nerve plexus exploration and/or nerve transfer. In the first patient, no procedure was performed following nerve exploration and the patient developed with “typical unilateral upper limb [presentation] consistent with AMC”. In the second patient, a nerve transfer from the median nerve to biceps motor branch was performed following nerve exploration. Active elbow flexion was achieved after nerve transfer. The study argued for nerve transfers prior to muscle transfers in selected children, suggesting that targeted muscles could be salvaged and functionally upgraded before a muscle transfer [15].

Table 3: Characteristics and results of studies addressing wrist surgeries.

Author	Surgical Procedure	Standardized Outcome Measure	Outcome Measure	Results Following Surgery	Complication
Burgess and Robbe [1]	Shortening osteotomy of radius and ulnar with volar wrist capsulectomy.	No	Wrist flexion and extension.	Initial improvement in wrist not maintained long-term.	Three wrists required salvage procedures for recurrent flexion deformity. Two wrists had salvage procedures recommended. One wrist had distal forearm fracture that compensated for wrist contracture. Three remaining wrists had wrist flexion contractures recur by three-year follow-up.
Foy et al. [10]	Carpal wedge osteotomy.	Manual Ability Classification System, ABILHAND-Kids.	Flexion and extension, ROM.	Mean resting wrist flexion posture improved from 55° to 11°. Arc of motion unchanged, but location improved to more functional position. Mean active wrist extension improved from -37° to -11°. Mean satisfaction after surgery 9.1/10 and ADL task completion was easier.	13 patients had additional surgeries: thenar slides, tricepsplasty, elbow releases.
Van Heest and Rodriguez [8]	Dorsal carpal wedge osteotomy.	No	Flexion and extension, arc of motion.	Mean wrist extension improved by 44°, mean flexion decreased by 35°. Greater improvement in wrist extension in children <7 years old at time of surgery compared to those >7 years old. No change in arc of motion. Parents of patients reported subjective improvement in position and appearance, and increased ADL task completion ability.	One wound infection: treated with irrigation, debridement and antibiotics. Healed without additional complications.

Wenner and Saperia [18]	Proximal row carpectomy.	No	ROM	Improved function subjectively in 4/5 patients, but was state that procedure produces uncertain results; long-term results generally unsatisfactory	Four salvage procedures required: three arthrodesis and one simultaneous flexor carpi ulnaris >extensor radialis carpi brevis.

Wrist

Two studies examined outcomes of proximal row carpectomy [18,19]. Wenner and Saperia showed subjective improved function in four of five patients, but noted their sample was small and long-term results were unsatisfactory. Additionally, four salvage procedures were required and osteotomy of the radius and ulna was stated to be the preferred procedure [18]. Mennen found that wrist and finger range of motion was increased in all joints following surgery and recommended the ideal age for surgery as three to six months (Table 3) (Table 4) [19].

Table 4: Characteristics and results of studies including surgeries on more than one joint.

Author	Surgical Procedure	Standardized Outcome Measure	Outcome Measure	Results Following Surgery	Complications
Mennen [19]	a) Volar radiocarpal capsulotomy, posterior capsulotomy. b) Distal radial and ulnar osteotomy.	No	Flexion, extension	a) Elbow flexion improved 30° to 100° (mean active flexion of 49°). Wrist extension improved 10° to 30° (mean active motion 27°). Finger flexion improved (metacarpophalangeal 20° to 85°, proximal interphalangeal 20° to 80°, distal interphalangeal 15° to 35°). b) Corrected position lost within 2 years due to remodeling.	Loss of correction was seen until one-step carpectomy with tendon transfer was implemented.

In studies addressing the results of distal radius and ulnar osteotomy and volar capsulotomy, it was shown that initial improvement was not sustained and the procedure was not recommended as treatment [1,19].

Two studies addressed dorsal carpal wedge osteotomy as treatment for wrist flexion deformities [8,10]. Both found the procedure resulted in decreased resting flexion position, increased active extension, and promoted a more functionally located arc of motion. It was also shown in the study by Van Heest et al. that completion of the procedure before age seven resulted in greater functional improvement. One wound infection occurred in the study by Van Heest et al., and healed without further complication following irrigation, debridement and antibiotics [8]. In the study by Foy et al., 13 patients required additional surgery (thenar slides, tricepsplasty, elbow release) but no other complications were noted [10]. Both reviews concluded that dorsal wedge osteotomy is capable of correcting wrist flexion deformity and maintained results over time [8,10,22].

Discussion

While the authors acknowledge the importance of physical therapy and splinting as treatments for upper extremity deformities in AMC, the purpose of this systematic review was to explore the current knowledge available regarding mid-term functional outcomes of shoulder, elbow and wrist surgery in individuals with AMC. Little research has currently been performed in this population; however, low level but supportive evidence was obtained in several studies.

While most patients with AMC present with a fixed elbow extension contracture [23], the majority of elbow surgical procedures are aimed at correcting fixed flexion contractures [14,17], with the first goal of treatment to provide a passive arc of elbow motion [7,14,19]. Posterior capsulotomy with triceps lengthening and pectoralis major muscle transfer are two procedures used to improve active elbow flexion. Pectoralis major transfer is more recently described than posterior capsulotomy with triceps lengthening, but despite being a newer procedure the results are not more promising. Posterior capsulotomy and triceps lengthening has been shown to consistently improve passive elbow flexion [7,14,19] and, more recently, active flexion [7]. Although the pectoralis major transfer also improves active flexion, it results in flexion contractures that prevent elbow extension and is only recommended on one elbow in cases of bilateral involvement to prevent the loss of independent perineal care [5,6]. It is important when addressing elbow flexion contractors to identify which patient goal is desired in order to identify the appropriate procedure.

Wrist flexion and ulnar deviation is typically present in patients with AMC [10,23], and dorsal carpal wedge osteotomy is the most recent procedure used to treat wrist flexion. Two articles discussed outcomes and results, and both showed sustained correction of the deformity with minimal or no complications [8,10]. Compared to poor results obtained from other wrist procedures, dorsal wedge osteotomy consistently improved flexion, extension and function of the operated wrists. Additionally, this procedure is advantageous as it preserves wrist motion in the radiocarpal joint while repositioning the hand into a more functional position [8]. Due to strong positive results

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obtained with minimal complications and the consistency of results between studies, the dorsal wedge osteotomy is noted to have the best outcome and is generally successful in treating wrist flexion deformities.

Conversely, articles that examined osteotomy of the radius and ulna with volar wrist capsulotomy to treat wrist flexion deformities reported the poorest outcomes. The study by Burgess and Robbe highlighted this, with the operation in nine extremities resulting in salvage procedures completed or suggested, or recurrent flexion contractures in all nine joints [1]. An older study by Mennen also found the results were not sustained until an additional one-step carpectomy with tendon transfer was included [21]. The suboptimal or lack of improvement indicates that radius and ulna osteotomy with volar wrist capsulotomy does not provide sustained correction of wrist flexion deformity.

This review strongly highlights the lack of standardization in assessing functional outcomes following surgery. While some outcome measures are described, such as those by Atkins et al. [24] and Foy et al. [10], the majority of results were described in terms of degrees of flexion and/or extension gained or lost, arc of motion and its location, range of motion, or general ADL task completion. Vascellari et al. assessed functional versus patient-reported outcomes following total knee arthroplasty and found a non-significant, very weak correlation between range of motion and ADL sub-scores [25]; Gialanella et al. investigated determinants of outcome in hip fractures and showed that basic ADLs had a strong relationship with functional outcomes [26]. Together, these two studies demonstrate that functional outcomes are better assessed using ADL task completion as compared to range of motion. Additionally, Nakahara et al. established that patient satisfaction scores correlate significantly with patient-derived functional scores [27], indicating that patient satisfaction is also an important outcome measure. The lack of consistency between the published articles included in this review therefore suggests the development, description and validation of functional outcome as well as patient satisfaction measures to standardize results of current and ongoing research.

Despite promising outcomes shown to result from certain procedures, it is important to note that the level of evidence of all articles included in this review was level 4. A significant contributing factor to this level of evidence is the difficulty and unfeasibility of completing higher-level studies, such as randomized control trials, in such a small, rare, relatively heterogeneous population. This does not mean that such a review is not warranted. Even with these obstacles, studies such as those by Foy et al. [10], Van Heest et al. [7], and Mennen [19] attempted to create larger sample sizes with longer follow-up to further strengthen the results of their studies.

Arthrogryposis and other rare conditions are difficult to research. One cause of this is that the research community is often distributed nationally or internationally. Additionally, many conditions, arthrogryposis included, require a multidisciplinary research and treatment approach. Resources needed to conduct the necessary research may similarly be spread or absent altogether. A challenge specific to AMC research is the heterogeneity of presentations inherent to the condition and the low number of individuals with each presentation. To reduce this heterogeneity this review did not include syndrome-like AMC, which limited the analysis to articles that largely included patients with amyoplasia, the most common form of AMC. This is important as it suggests that the results shown by the studies included in this review may not be achieved in all patients with AMC, which supports and further necessitates the call for additional research. Additionally, a randomized control trial may not be achievable in this population. Therefore, the idea of creating a centralized patient registry to follow patients longitudinally may provide better insights into surgical treatment outcomes.

Despite the abovementioned barriers to research in the field of AMC, continued research is crucial. The lack of knowledge regarding treatment options and outcomes may negatively affect the quality of life of the patient or delay access to appropriate treatments. Misdiagnosis, delayed diagnosis, or mismanagement often results in wasted resources and increased expenses for healthcare and social systems. Therefore, research related to any rare condition that improves diagnosis or treatment not only improves patient outcomes, but can translate to a cost reduction for healthcare systems. Furthermore, innovative multidisciplinary approaches or new treatment methods are often able to benefit the wider public affected by more common diseases. Thus, research into rare conditions such as AMC would not only benefit those affected, but also the general population.

This unique, novel review of functional outcomes at least two years following shoulder, elbow or wrist surgery in individuals with AMC provides some knowledge as to which procedures have shown sustained benefits, which have not, and which are recommended as treatment. In particular, the dorsal carpal wedge osteotomy is highlighted as being consistently able to functionally correct the wrist and preserve its motion with minimal associated complications. The conclusions made in this review are based on articles with low levels of evidence, which should be considered when making treatment decisions; however, due to the rarity of this condition no single study will be able to meet the requirements of a randomized control trial. This review also clearly reveals the limited knowledge presently available regarding mid- and long-term outcomes in those with AMC. Minimal knowledge and research has the potential to result in deficient and substandard treatment of individuals with AMC, affecting both the patient's quality of life and causing increased strain to healthcare and social systems. Thus, clinicians and researchers are encouraged to begin and continue research related to this population to provide both clinicians and patients with clarity regarding early treatment options and expected results, and to further expand the knowledge base of this rare condition.

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