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Hand

Contemporary Management of the Upper Limb in Apert Syndrome: A Review

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Background: Apert syndrome is a relatively rare genetic disorder with a constellation of distinct craniofacial deformities and bilateral syndactyly of the hands and feet. Although the literature contains ample evidence for the need to treat cranial, midfacial, and hand abnormalities, there are severe shortcomings in the literature when attempting to describe the pathology and management of the entire upper limb in patients with Apert syndrome.
Methods: A thorough literature search was performed using PubMed, Scopus, Web of Science, and Google Scholar, on the management of the upper extremity in Apert syndrome, including the shoulder, elbow, and hand.
Results: Our findings of the literature discuss the clinical presentation and management trends of the upper extremity in patients with Apert syndrome. Through multicenter collaboration, discussion among experts in the field, and evidence gathered from the literature, we propose treatment algorithms to treat deformities of the hand, shoulder, and elbow in patients with Apert syndrome.
Conclusions: This review identifies that even if hand pathologies have been correctly treated, shoulder and elbow abnormalities in patients with Apert syndrome are largely ignored. To optimize outcomes, added cognizance of additional upper limb congenital differences and their management should be highly advocated in this patient population. *(Plast Reconstr Surg Glob Open 2024; 12:e6067; doi: 10.1097/GOX.0000000000006067; Published online 15 August 2024.)*

INTRODUCTION

Apert syndrome (AS) was first described by French physician Eugene Apert in 1906.[1] In his renowned publication, "De l'acrocephalosyndactylie," Apert meticulously describes the complex deformities he observed in similar patients, coining the term acrocephalosyndactyly.[2] Now known as AS, or acrocephalosyndactyly type I, this syndrome is a rare genetic disorder with an estimated prevalence between one in 80,000–200,000 live births.[3,4] The literature contains ample evidence for the need to treat cranial, midfacial, and hand abnormalities. However, there is a paucity of information that describes the pathology, anatomy, and treatment of the entirety of the upper limb, especially as it pertains to the elbow and shoulder. Therefore, our objective is to provide a comprehensive overview of the clinical presentation and management strategies pertaining to the entire upper limb in AS.

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DIAGNOSIS

Diagnosis relies on a combination of clinical, radiological, and genetic findings. The most frequently observed clinical finding includes an atypical head shape, resulting from the premature closure of one or both coronal sutures. This can lead to other conditions such as brachycephaly or plagiocephaly. Other brain anomalies such as hydrocephalus, agenesis, or dysgenesis of the corpus callosum is also common.[5–7] Moreover, complex syndactyly, thumb abnormalities, and symphalangism are characteristic hand findings. Prenatal ultrasounds combined with fetal magnetic resonance imaging can identify fetal morphological abnormalities in utero.[8,9] Anomalies related to cranial development have been observed in utero as early as the 19th week of gestation.[10] Additionally, recent advancements in three-dimensional prenatal ultrasound have been shown to successfully demonstrate characteristic features during the third trimester of pregnancy.[11,12]

Disclosure statements are at the end of this article, following the correspondence information.

Genetic testing for a missense mutation on chromosome 10q confirms the diagnosis.[13]

CLINICAL FEATURES

The most prominent clinical features are a combination of craniosynostosis; midface hypoplasia; and symmetric, complex syndactyly with symphalangism of the hands and feet.[14,15] Symptoms related to the skin, central nervous, cardiac, gastrointestinal, and urogenital system, and vertebral anomalies have rarely been reported.[16] Further clinical features of the upper limb, including the hand, elbow, and shoulder, are detailed as follows.

Takeaways

Question: What is our understanding of the pathology, management, and treatment of upper extremity deformities in children with Apert syndrome?

Findings: This article provides a comprehensive overview of the clinical presentation and management strategies pertaining to the entirety of the upper limb in Apert syndrome. Due to the scarcity of management protocols, we propose treatment algorithms to help guide providers in treating deformities of the hand, shoulder, and elbow.

Meaning: To optimize outcomes, added cognizance of upper limb congenital differences and their management should be highly advocated for in patients with Apert syndrome.

Hand

One of the hallmark characteristics of AS is complex hand syndactyly. Symphalangism and clinodactyly are also frequently observed. The degree of thumb involvement in hand syndactyly provides the overall basis for the Upton classification.[17] In Upton type I (referred to as spade hand), the thumb remains free, whereas the central three digits are fused together, with the thumb appearing flat and separate. There is a variable amount of syndactyly of the fourth webspace in this type (Fig. 1). In Upton type II (known as mitten hand), both the thumb and central three digits are fused, creating a concave palm (Fig. 2). In Upton type III (referred to as rosebud hand), all digits of

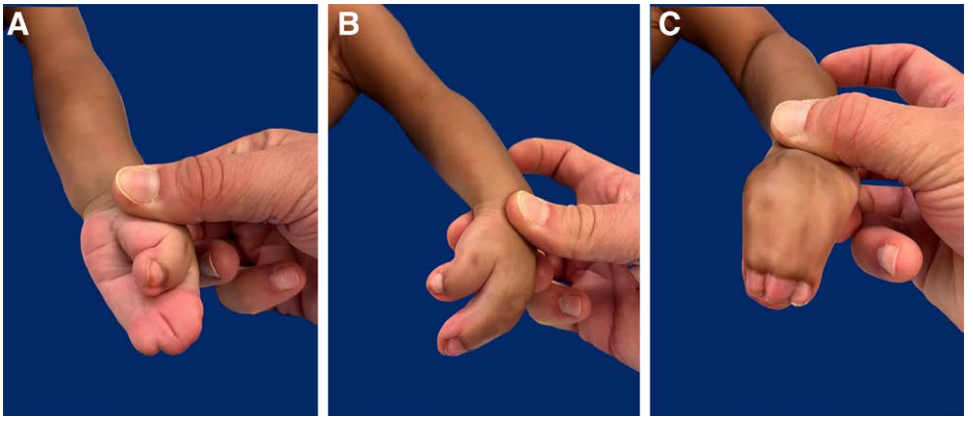

Fig. 1. Upton type I hand in a patient with Apert syndrome. A–C, Preoperative views.

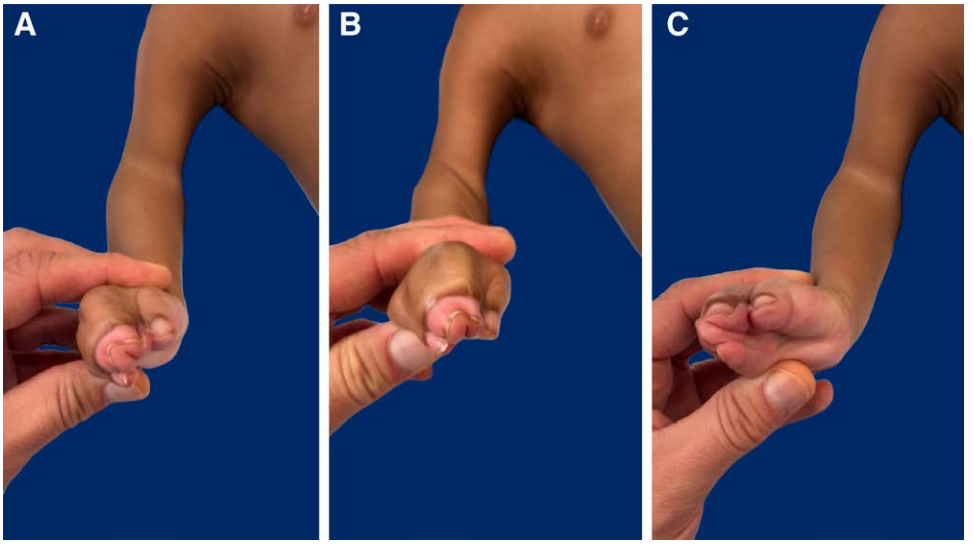

Fig. 2. Upton type II hand in a patient with Apert syndrome. A–C, Preoperative views.

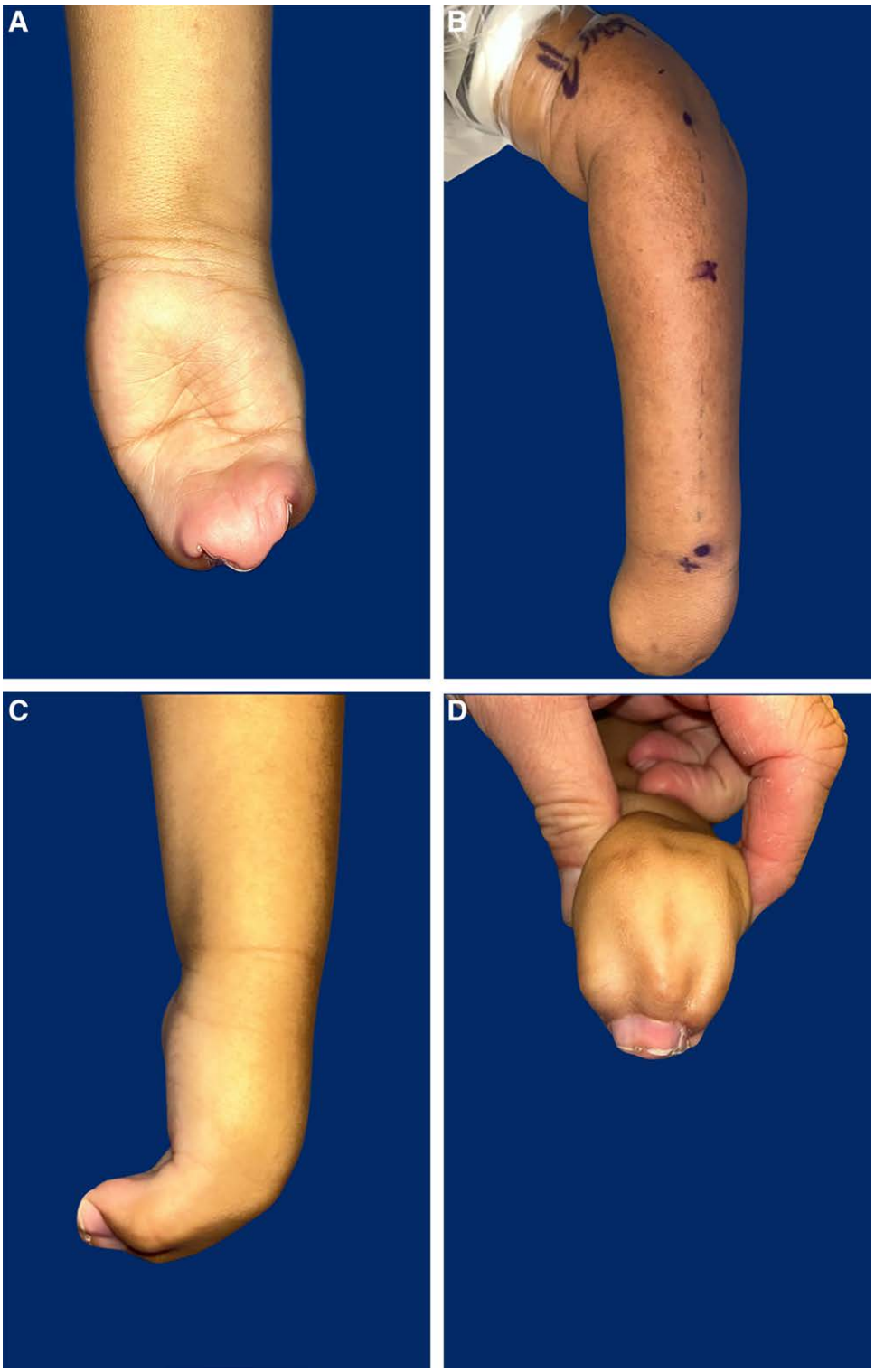

Fig. 3. Upton type III hand in a patient with Apert syndrome. A–D, Preoperative views.

the hand are fused together, with a complex bony coalition (Fig. 3).[18]

Shoulder

Less commonly, affected individuals present with a range of developmental abnormalities of the shoulder girdle, leading to limited shoulder mobility. Cuthbert[19] first drew attention to the prominence of the acromial process with associated deformity of the humeral head and glenoid. Glenoid dysplasia emerged as the most prevalent abnormality in the cohort described by Kasser and Upton.[20] Murnaghan et al. corroborated many of

these physical and pathological findings, with the additional observation of medial humeral head hypoplasia.[21] When reviewing the magnetic resonance imaging findings in a 10-month-old with AS, the results revealed glenoid hypoplasia and growth arrest of the medial aspect of the humeral head.[22] Complete glenohumeral synostosis has also been documented as a rare finding.[23]

Progressively worsening with age, the glenohumeral joint can result in restrictions in abduction, forward flexion, and internal and external rotation.[20,24] Kasser and Upton[20] observed that the motion of the glenohumeral joint was consistently impaired and decreased as the child matured. Wood et al[25] observed shoulder abduction limitations, suggesting anterior subluxation, with four of nine patients demonstrating shoulder incongruity.

Elbow

Park and Powers[26] were the first to allude to a congenital abnormality of the elbow with an illustration of a patient with significant bilateral flexion contractures. In a prospective radiographic study, evaluating the incidence of elbow anomalies in patients with syndromes associated with craniosynostosis, it found that anomalies of the elbow were very common in AS, with the most common being complete or proximal radiolunar synostosis.[27,28] Proximal radioulnar synostosis occurs due to the incomplete prenatal segmentation of the forearm bones, leading to limited rotational movement of forearm.[29]

Beligere et al[24] recorded limited extension of the elbow joint in seven of 10 children with AS. Upon retrospectively analyzing radiographic and clinical findings at the elbow joint, the authors discovered that of 19 patients, seven experienced reduced elbow extension, another seven demonstrated a lack of elbow flexion, two had fused elbow joints, and the remaining had segmented joints.[20] Furthermore, Wood et al[25] noted flattening and subluxation of the radial head in radiographs with a positive click on physical examination in most patients with AS.

MANAGEMENT

Shortly after birth, children are first referred to craniofacial centers for treatment of calvarial and midfacial anomalies.[30] In cases in which children are born with an obstructed airway, immediate attention is required. Next, typically between 2 and 4 months old, release of the fused cranial sutures can be performed. If this is not indicated, then posterior cranial vault distraction, calvarial vault remodeling, or a fronto-orbital advancement is typically performed between 9 and 11 months. It is typically between these two critical surgical events that care of the upper limbs is coordinated with the orthopedic or plastic hand surgeon specializing in congenital hand abnormalities. In the following section, we discuss past literature recommendations and our current model for management of the upper limbs in AS.

Hand

The management of hand phenotypes in AS demands a multifaceted and individualized approach, considering the wide variability in anatomical presentation. There are many staging algorithms and technical variations that have been proposed for syndactyly release and reconstruction of metacarpal structures.[3,31] After determining the extent of hand malformation, a surgical correction strategy can be coordinated, typically spanning over the course of several years. Overall, the goal is to minimize the number of operations by addressing as many digits or hand components as possible, performing bilateral procedures, combining procedures with different anatomical regions (such as the feet), and achieving a functional and aesthetic hand with an opposable thumb with three to four fingers, depending on individual anatomy.[17]

The optimal timing for initiating surgical correction of the hand is generally recommended around 6 months of age.[3,32] This recommendation is based on the understanding that operating on a hand younger than this age presents with significant surgical challenges. These include dealing with smaller structures and encountering a greater amount of subcutaneous fat which complicates the mobilization of skin flaps, and, because of the little hand growth which has occurred, the risk of significant web creep greatly increases (the hand will roughly double in size in the first 3 years of life). Additionally, because lateralized manual hand preference manifests at 21 months of age, but fine reach-to-grasp does not manifest until 3 years old, the goal of early surgery is to complete prehensile hand reconstruction by the age of 3. Exceptions for this timeline, however, do exist; for instance, early correction of digit abnormalities can commence as early as 3 months of age, offering benefits in preventing growth discrepancies, especially in cases of syndactyly involving border digits and with significant contracture. Conversely, delaying surgical correction presents with advantages such as enhanced growth potential, enabling operations on a more developed structure, and decreasing the risk of postoperative complications.

Surgical planning first is necessary to determine the degree of hand malformation and the steps needed for reconstruction. Radiographs combined with a detailed physical examination provide the basis for a comprehensive surgical plan, that is, by necessity, staged over many years. Almost universally, consensus dictates addressing the first webspace first; a mobile and stable thumb with a broad first web space is essential to good hand function. In type I hands with incomplete syndactyly (and a minority of type II hands) leading to a shallow first webspace, the most common approach is a 4- or 5-flap Z-plasty to deepen the web space at the same time of finger separation (Figs. 4 and 5).[33] In complete first webspace syndactyly as in type II or type III hands, a large dorsal rotational flap (Ghani modification of the Buck-Gramcko flap) or first dorsal metacarpal artery flap[3,31,32] is required to provide adequate soft tissue for the webspace. More avant-garde approaches, such as a reversed posterior interosseous artery flap have also been described (Fig. 6).[34] Release of tight adductor muscles is often required. In selected cases, especially severe type III hands with a short thumb, index amputation has been advocated to provide relative widening of the first webspace and gain additional dorsal tissue.[17] The

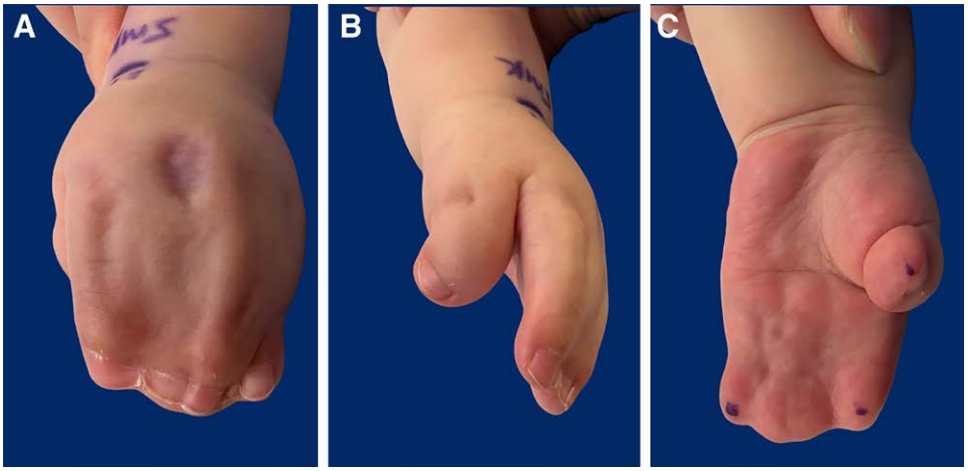

Fig. 4. Upton type I hand with multiple views (A–C).

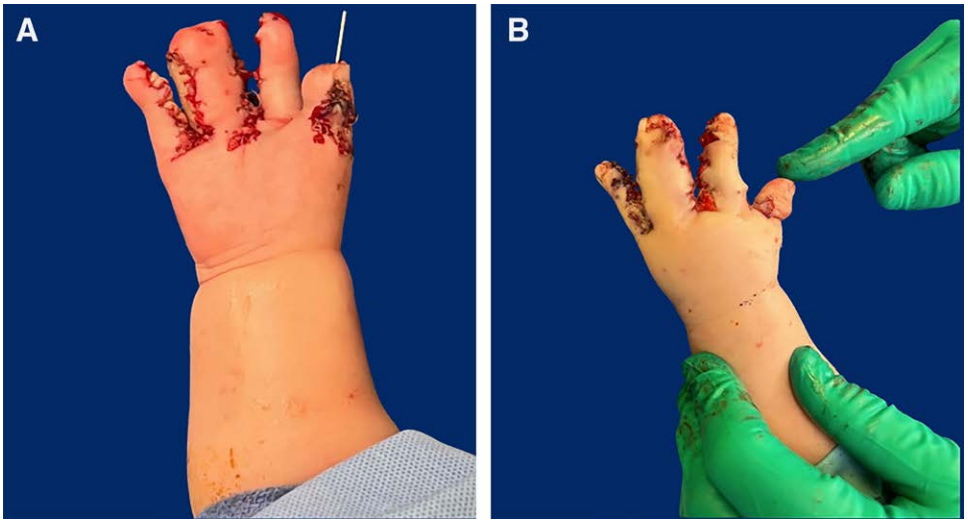

Fig. 5. Upton hand type I undergoing deepening of the first webspace and separation of the second and fourth webspaces. A–B, Intraoperative images.

non-Z-plasty solutions avoid interdigitating flaps in the webspace and allow for subsequent readvancement of tissue during subsequent surgery to deepen the webspace.

Release of complex syndactyly follows the general surgical principle of ensuring adequate soft-tissue coverage after release by using local flaps or skin graft and avoidance of releasing both sides of the same digit in one session to ensure vascularity, which is of particular importance in given common neurovascular anomalies in affected children.[33,35] Although the order of separation is contentious (we advocate for border digit separation first, along with the first webspace), prioritizing the release of the border digits, by necessity, will result in three operations to create a five-digit hand, rather than a complete release in two stages as advocated by Fearon and others.[3,31,36]

Separation of the fingers can be achieved by creating the webs using dorsal metacarpal flaps[37] and a zigzag (Fig. 7)[31,32] or straight-line[3] incisions distally. Cordray et al[38] advocate for the use of straight-line incisions due to decreased complication rates and less visible scars. Remaining skin defects can be addressed by applying full-thickness skin grafts harvested from the lower abdomen.[3,31,39] Groin flaps may be used to provide coverage to the central digit.[32] To address nail deformity commonly associated with type III hands, Buck-Gramcko flaps are utilized to create a paronychium by utilizing adjacent pulp tissue.[40] Although there are descriptions of utilizing external distraction to progressively recruit soft tissue and nail matrix before separating complex syndactyly, this approach is less commonly used due to its demanding nature.[41] Revision procedures may be necessary due to web creep and to further deepen the commissures.

For synostosis of the proximal fourth and fifth metacarpals, early separation during syndactyly release is crucial to enhance grip strength and prevent stiffness.[3] This must also be monitored, as it has been reported to occur later in growth. As such, it is sometimes performed in concert with thumb osteotomy corrections. Border fingers

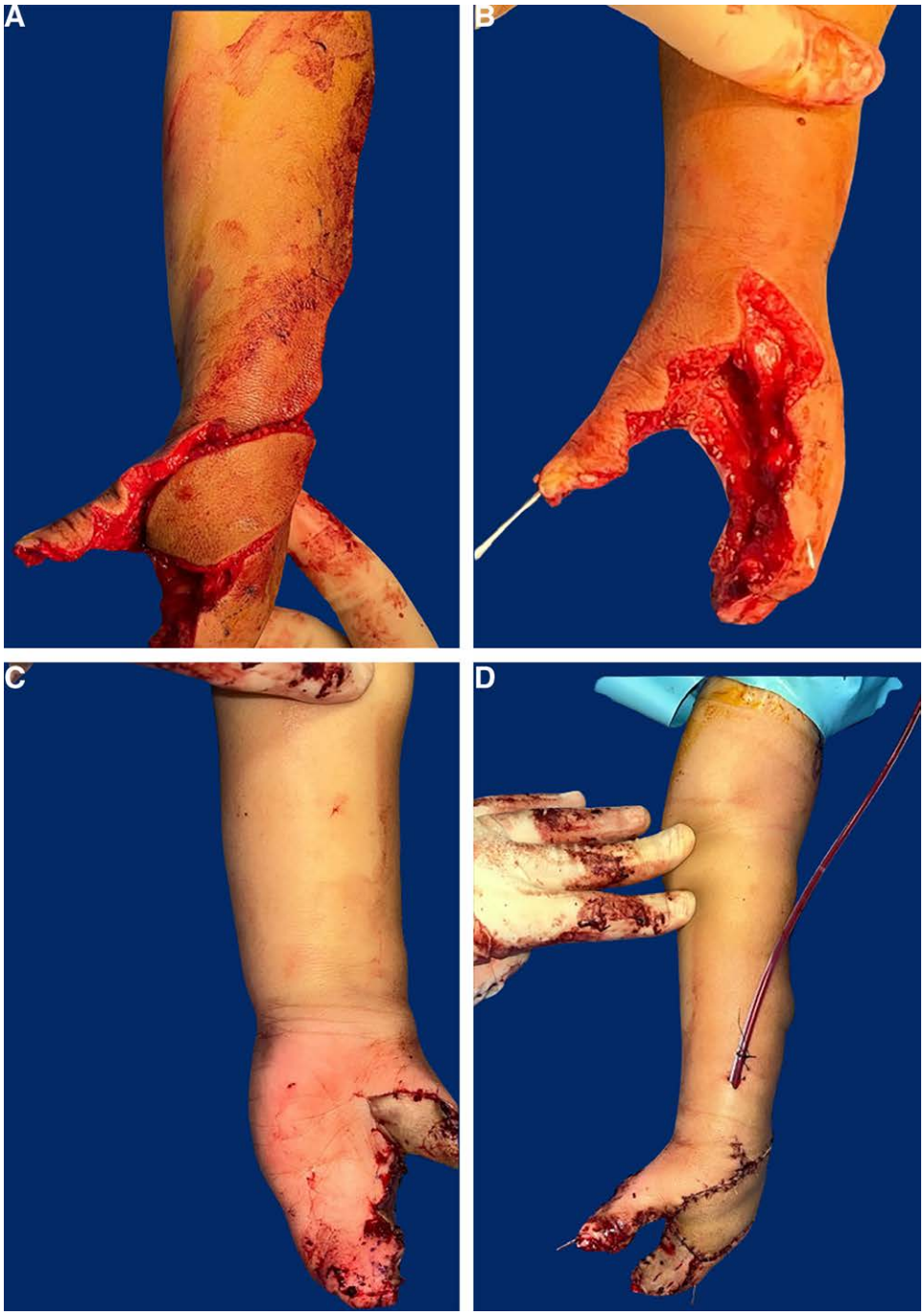

Fig. 6. Separation of the first webspace using a pedicled reverse posterior interosseous artery in Upton hand type III. A–D, Intraopertiave images.

that tend to laterally deviate or develop clinodactyly may require corrective osteotomy at a later stage.

Clinodactyly of the thumb is commonly observed in type I and type II hands. Although correction of metacarpal synostosis is typically combined during syndactyly release, correction of thumb clinodactyly is typically delayed. The correction of thumb abnormalities may be delayed to a later time, as suggested by Upton and Fearon[3,17] or performed concurrently with the second stage of finger separation, as advocated by Chang et al.[39] The goal of correcting this deformity is to re-establish the pinch mechanism by decreasing the radial deviation.[31] Radial deviation at the metacarpophalangeal joint is commonly recognized due to the abnormal insertion of the abductor pollicis brevis.[3,17,31] To correct thumb deformity without osteotomy, some advocate for soft-tissue correction along with abductor pollicis brevis release and capsulotomy.[42] However, most typically address thumb clinodactyly between 2 and

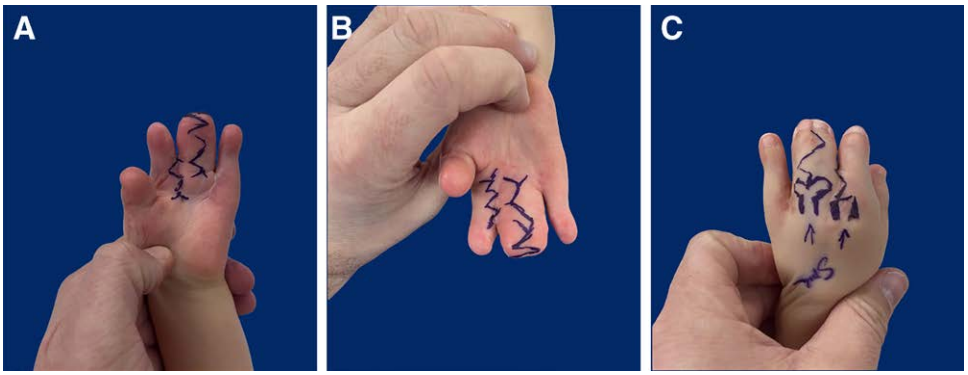

Fig. 7. Separation of the second, third, and forth webspaces using zig-zag incisions distally to address symphalangism. A–C, Intraoperative images.

4 years of age through an open or reverse wedge osteotomy to correct the delta phalanx[31] with a Z-plasty or local flap of the contracted skin at the interphalangeal level. Additionally, correcting symphalangism with angular osteotomies between 5 and 8 years of age has shown promising outcomes.[3]

Shoulder

Shoulder abnormalities can include malformations of the shoulder joint and muscle imbalances leading to progressive dysplastic changes that result in restriction of shoulder mobility.[18] Patient-reported outcome scores have revealed significant functional impairment associated with shoulder deformity, comparable to the disability reported for their hand deformity.[21] Despite this, surgical correction of the shoulder joint has not been well defined or discussed, as symptomatic presentation varies widely in severity and timing.[43] Currently, there is no evidence in the literature to support early or prophylactic correction.

As such, surgical management has focused on addressing late-stage and severe arthritis secondary to

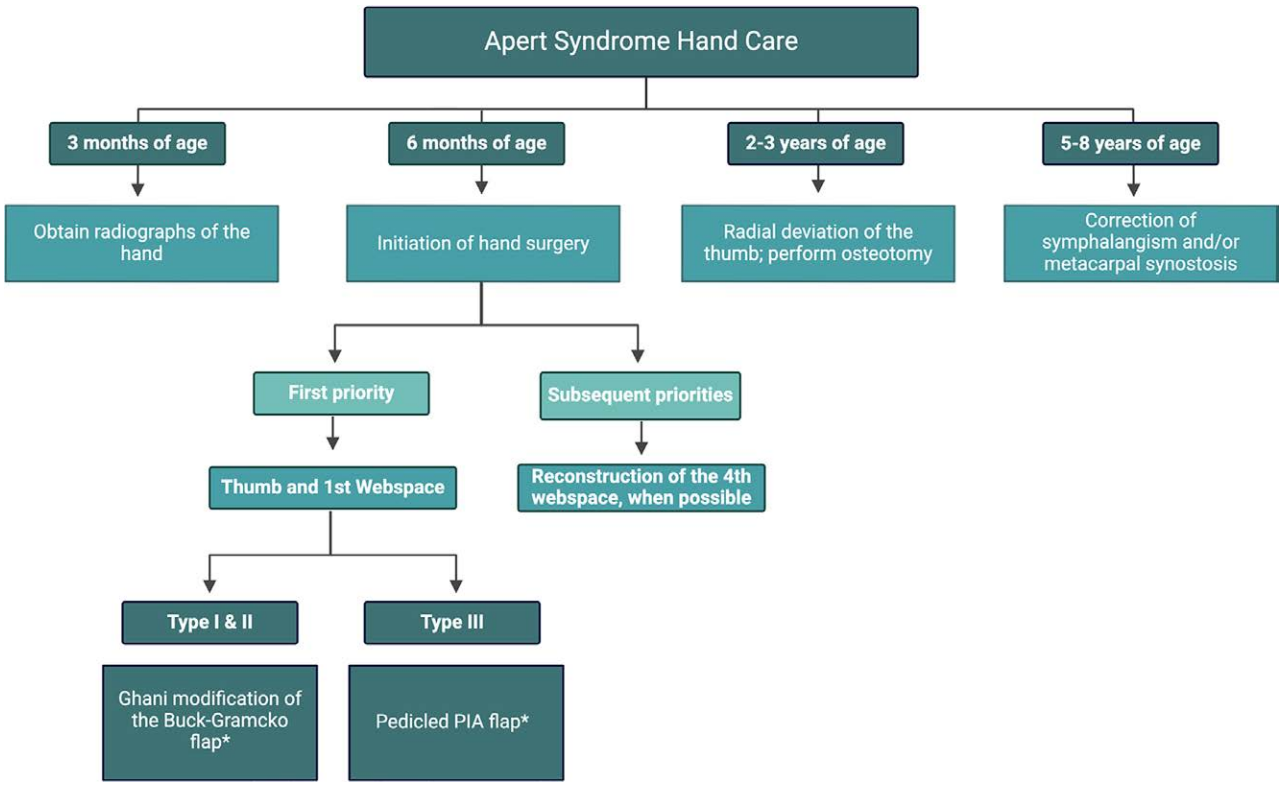

Fig. 8. Recommended algorithm for treatment and management of the hand in patients with AS. *Important to note that due to significant hyperhidrosis in these patients, which can result in compromised skin grafts, we suggest utilizing Unna dressing and often to K-wire the fingers apart to keep the commissures open during warmer season reconstructions.

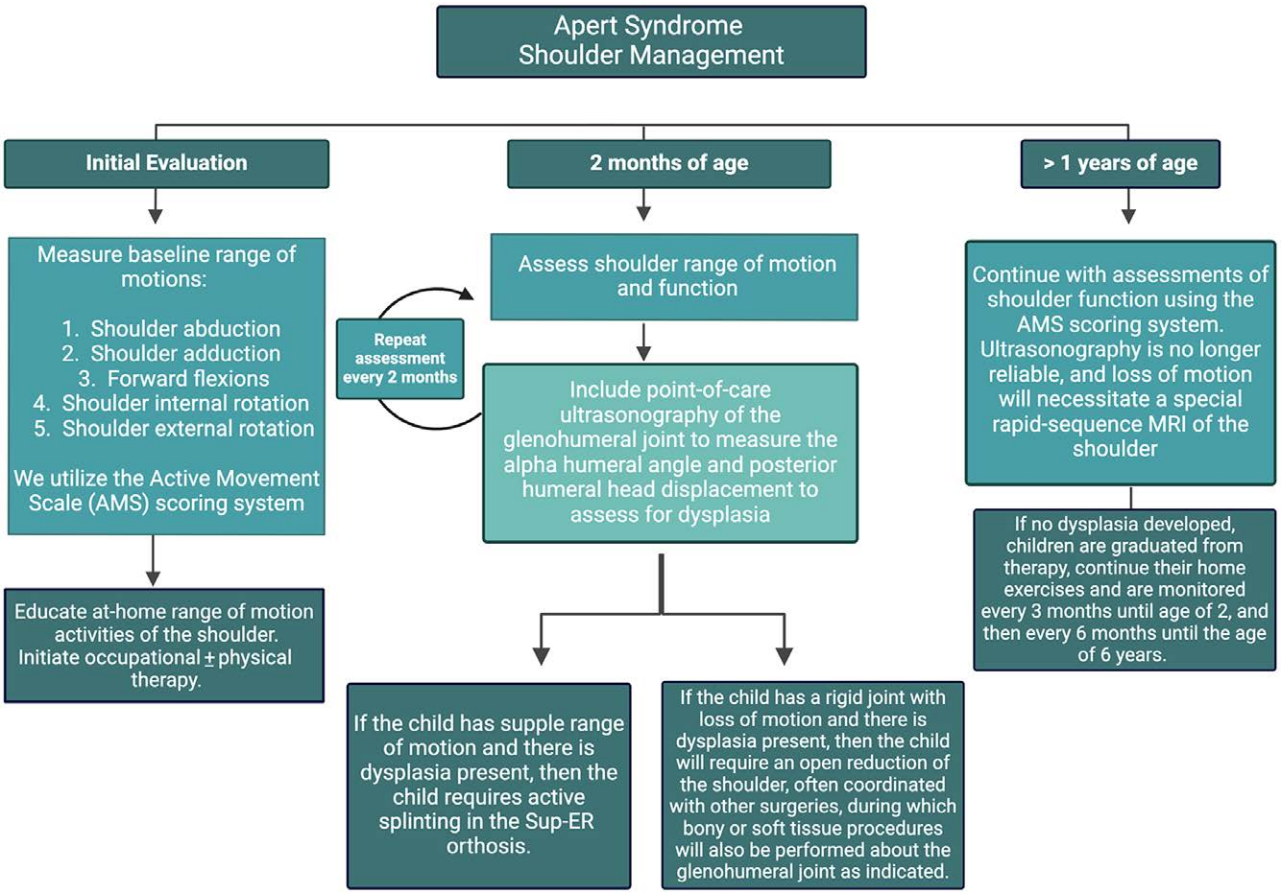

Fig. 9. Recommended algorithm for treatment and management of the shoulder in patients with AS.

glenoid dysplasia, with mixed results. Humeral head resurfacing and hemiarthroplasty do not consistently provide pain relief and cannot correct persistent glenohumeral subluxation.[44] Total shoulder arthroplasty can be performed with satisfactory pain relief; however, underlying glenoid hypoplasia often results in subluxation of the prosthetic glenoid component and requires revision or results in abject failure.[44] A case report on the use of reverse arthroplasty suggested promising results with reliable fixation.[45] Soft-tissue modifications such as capsular release, capsulorrhaphy, muscle lengthening, and tendon transfers are techniques that are well described in brachial plexus birth injury related glenohumeral deformity but have not been reported in the treatment of AS. Similarly, osseous procedures such as coracoidectomies or those aiming at glenoid reorientation using osteotomy or glenoid augmentation have been described as reconstructive options.[46] These options can be considered on an individual basis in patients with AS, considering anatomy, presentation, age at presentation, and surgeon familiarity and comfort. We currently screen children for the development of glenohumeral dysplasia and aggressively intervene at the onset of presentation with many of the above noted interventions on an individualistic basis outside the scope of this article.

Elbow

Surgical management of congenital deformities of the elbow joint has rarely been described in AS. It has been postulated that this is likely due to the fact that these elbow deformities are well compensated.[21] However, this is rarely the case. It is well established in reconstructive upper limb surgery that the elbow is a priority joint and the conduit for the hand to reach the mouth and face. With a poorly functioning elbow, significant upper limb disability results. What little has been reported primarily addresses bony deformities and instability through osteotomies and ligamentous procedures. The optimal timing of surgical management has not been determined, although as these children grow, the bony deformity and soft-tissue contractures can worsen and increase symptoms and surgical complications.[47] This is also likely why pathology is missed by surgeons—most hand surgeons complete surgical reconstructions by 4–6 years of age, and elbow pathology may manifest at a later time. Ultimately, optimal elbow outcomes may come to rely on a comprehensive approach with a strong emphasis on early range of motion, surgery, and life-long physical therapy. In our practice, elbow pathology is commonly screened for and treated. We frequently encounter various synostoses about the elbow, mostly involving the radial head; thus, radial head resection can provide a straightforward solution to

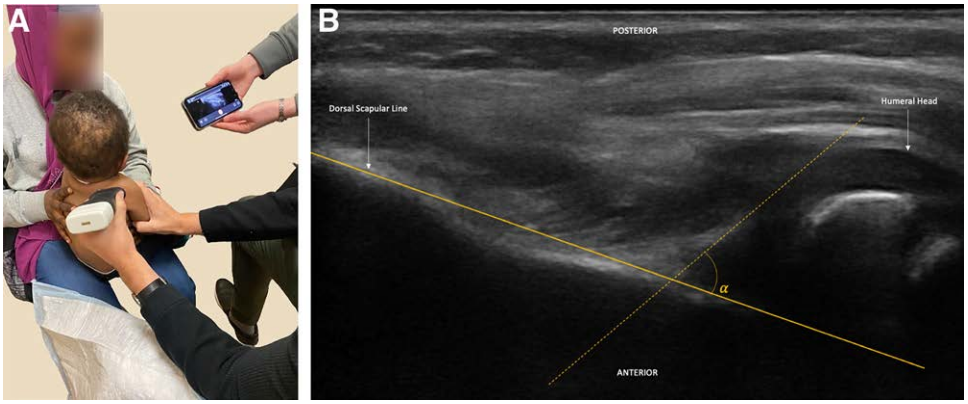

Fig. 10. Ultrasound imaging of the shoulder in a patient with AS. A, Preparing the patient. B, Image depicting calculation of the alpha angle from transverse ultrasound images of the shoulder adducted and internally rotated. The alpha angle is calculated by the intersection of the dorsal scapular line and the line tangential to the humeral head at the glenoid (B).

Fig. 11. Posterior (A) and anterior views (B) of an orthosis fabricated bilaterally in a patient with AS.

preserving elbow motion in the Apert population, in our experience.

DISCUSSION

AS represents a spectrum of congenital differences, necessitating individualized consideration and prioritization by a multidisciplinary surgical team. The entirety of the upper limb should remain paramount on this list of conditions to be fully addressed.

Overall, the reconstructive goals of the hand highlighted are synonymous in the literature, despite there being differences on how to achieve those goals. This is unsurprising, as the hand is the dominant published aspect of the upper limb differences in AS. We too, ascribe to many of the same tenets as our colleagues around the globe for our hand care (Fig. 8).

However, the shoulder is unique from the hand in many respects; there is little literature regarding the shoulder, yet shoulder motion is almost never normal and may be quite restricted. Despite this understanding, minimal care has historically been provided. Fundamentally, this stems from the absence of evidence-based literature or a consensus in the literature regarding shoulder management in these children. In our shoulder protocol (Fig. 9), we utilize the Active Movement Scale scoring system to assess for shoulder function,[48] and ultrasonography measurements, such as measurement of the alpha angle,[49] to assess for glenohumeral dysplasia (Fig. 10). As described in our protocol, treatment for glenohumeral joint, as evidenced in these

Fig. 12. Clinical examination findings of external rotation after treatment of glenohumeral dysplasia in a child with AS (A–B).

infants, can include home exercises, occupational and/or physical therapy, fabrication with Sup-ER orthosis (Figs. 11 and 12),[50] and open reduction of the shoulder.

The elbow is limited in a much smaller percentage of patients and therefore is even less addressed in the literature. It is often thought to be clinically insignificant unless an elbow fusion has occurred. Therefore, it is of the utmost importance to have a high index of awareness during continuous follow-ups. Because there is truly no guidance in the literature regarding the elbow in AS, we have developed our own elbow protocol (Fig. 13).

Unfortunately, the functional capacity in patients with AS tends to be limited due to the severity of the deformities involved. Although the literature is limited in the long term, one study revealed that average total times on the Jebsen Hand Function Test were notably slower than population norms, and measures of total active motion and pinch strength were considerably lower in adults with AS.[18] Despite evident functional deficits, individuals with AS often lead remarkable lives, as those born with these conditions tend to demonstrate great adaptability. This adaptability, along with early surgical intervention, contributes to the ability to live a fulfilling life.[51] This can only be further improved with the added cognizance of additional upper limb congenital differences and their management.

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DISCLOSURES

Dr. Koehler is a committee member of the American Society for Surgery of the Hand, and a stockholder and member of the medical advisory board for Reactiv, Inc. Dr. Mendenhall is an educational consultant for PolyNovo. The other authors have no financial interest to declare in relation to the content of this article.

PATIENT CONSENT

Patients or guardians provided written consent for the use of their images.

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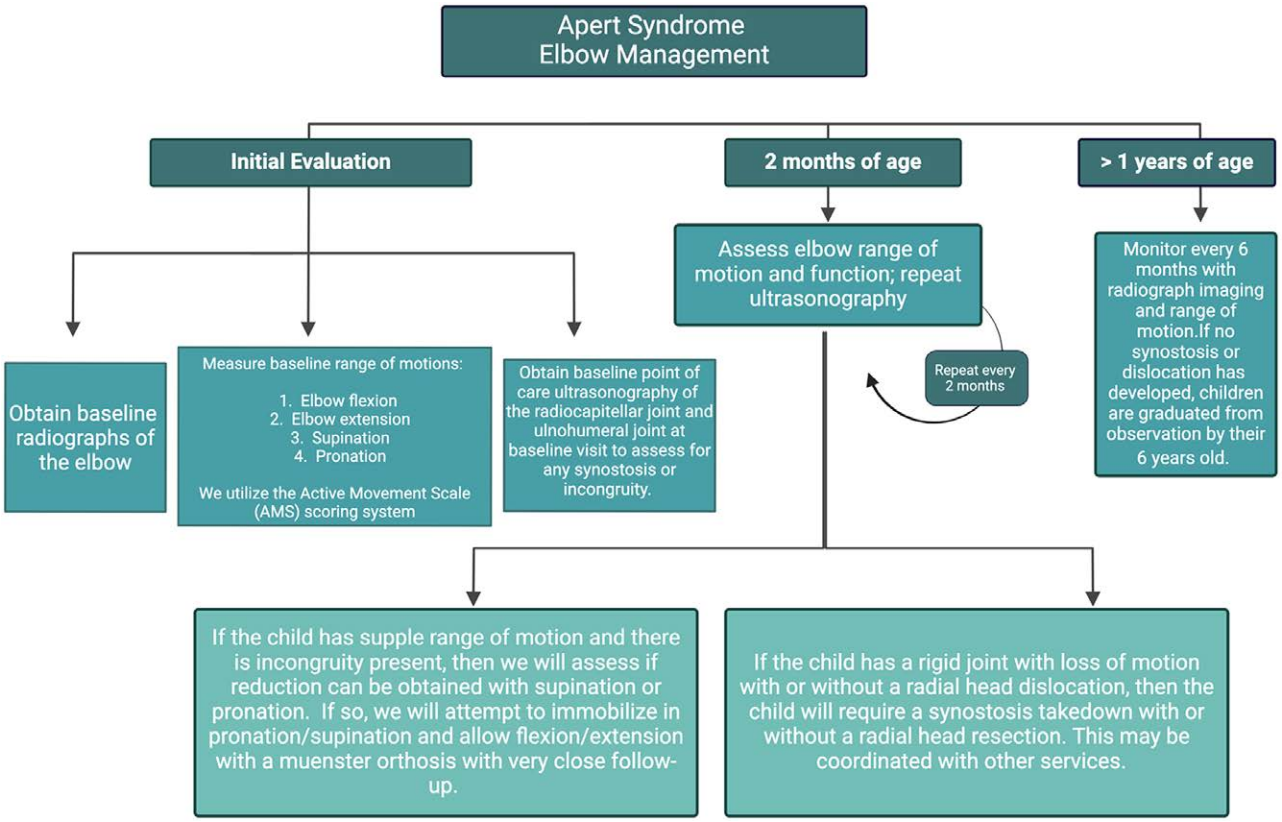

Fig. 13. Recommended algorithm for treatment and management of the elbow in patients with AS.

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