Amniotic Band Syndrome of the Upper Extremity: Diagnosis and Management

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Abstract

Amniotic band syndrome, a condition involving fetal entrapment in strands of amniotic tissue, causes an array of deletions and deformations. Band formation most frequently affects the distal segments, including the hand. Because of the heterogeneous nature of expression of this disease, treatment is individualized. Timing of repair and surgical planning are important in improving functional outcome. In the patient with distal edema and acrosyndactyly, early repair portends better prognosis. Improvements in prenatal diagnosis and fetoscopic surgical technique may eventually allow treatment of amniotic band syndrome in utero.

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Amniotic band syndrome (ABS) is a rare condition in which fetal parts become entangled in amniotic membrane, leading to constriction, deformation, and deletion. Resulting defects from constricting bands range in severity from mild constriction to complete limb deletion, anencephaly, and fetal demise. Amniotic bands most commonly affect the extremities, with a predilection for the distal segments (Figure 1). Deformations affecting the upper extremity can be disabling and, because of the unique presentation in each individual, pose a treatment challenge.

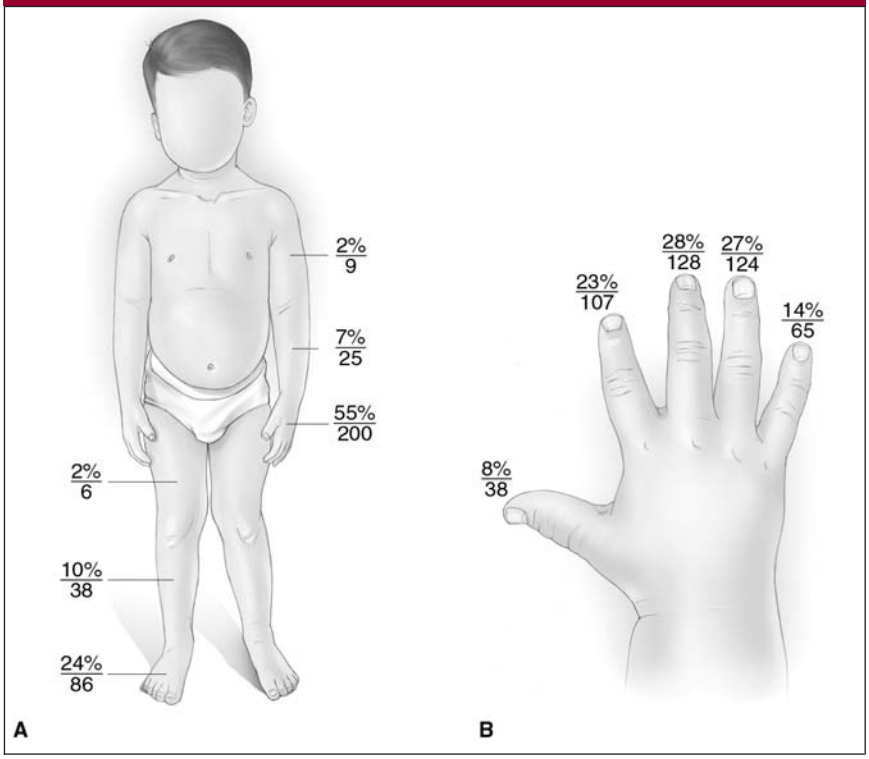
Etiology, Embryology, and Demographics

The earliest recorded account of ABS dates to Hippocrates, who made reference to a syndrome of amputation or band formation caused by fetal membranes encircling the extremities or digits.[1] The number of synonyms associated with ABS (annular bands; amniotic disruption sequence; amniotic deformity, adhesions, and mutations complex; annular defects; Simonart's band; congenital ring-constriction; limb-body wall malformation complex; early amnion rupture sequence; intrauterine or fetal amputation; constriction band syndrome; Streeter's dysplasia[2]) attests to the difficulty physicians have had in determining its etiology.

ABS was first defined based on the presence of constrictive tissue bands, amniotic adhesions, and more complex patterns of anomalies, designated as the limb-body wall complex.[3-5] Streeter suggested that an intrinsic defect in the subcutaneous germplasm resulted in focal mesenchymal hypoplasia, tissue loss, and scarring.[1] The proposed mechanism was thought to be similar to that involved in the development of normal skin folds, which appear very similar histologically.

In contrast, the extrinsic theory proposed that the entanglement of fetal parts by bands of amniotic tissue caused deformation. Histologic evidence of amniotic tissue within constriction grooves provided support for

Figure 1



Frequency of involvement of the extremities (n = 364) **(A)** and digits (n = 462) **(B)** in amniotic band syndrome.[6] (Reproduced with permission from the Mayo Foundation.)

this idea. Case reports of formed, amputated extremities within the amniotic cavity suggested that the syndrome was a deformation rather than a true malformation.[1,6] Houben[7] and Kino[8] duplicated similar limb deletions and syndactyly by performing early amniocentesis on rat embryos. These authors suggested that extrinsic forces on the limb buds led to hemorrhage of the distal marginal blood sinuses, which in turn led to hematoma formation, scarring, deletion, and acrosyndactyly. Medical imaging technology has confirmed the extrinsic theory by demonstrating the presence of constricting bands of fetal membranes that lead to deformation.[9-11]

Fetal membranes are composed of two distinct layers: the amnion and the chorion. The inner amnion, which provides the majority of tensile strength, is composed of the inner, mesenchymal, and outer layers. The inner and mesenchymal layers play an important role in secreting collagen, fibronectin, and lamin to provide a strong elastic and tensile layer. The outer layer, also known as the spongy layer, is adjacent to the chorion; it can swell to accommodate sliding of the amnion across the chorion. The membranes are seen on ultrasound as two separate and distinct entities until approximately 3 months' gestation, when the chorion and amnion fuse. Defects or tears of the inner membrane can lead to fetal entrapment. Transient oligohydramnios caused by the loss of amniotic fluid also may contribute to deformation of early limb buds, including the occurrence of clubfoot deformity.[8]

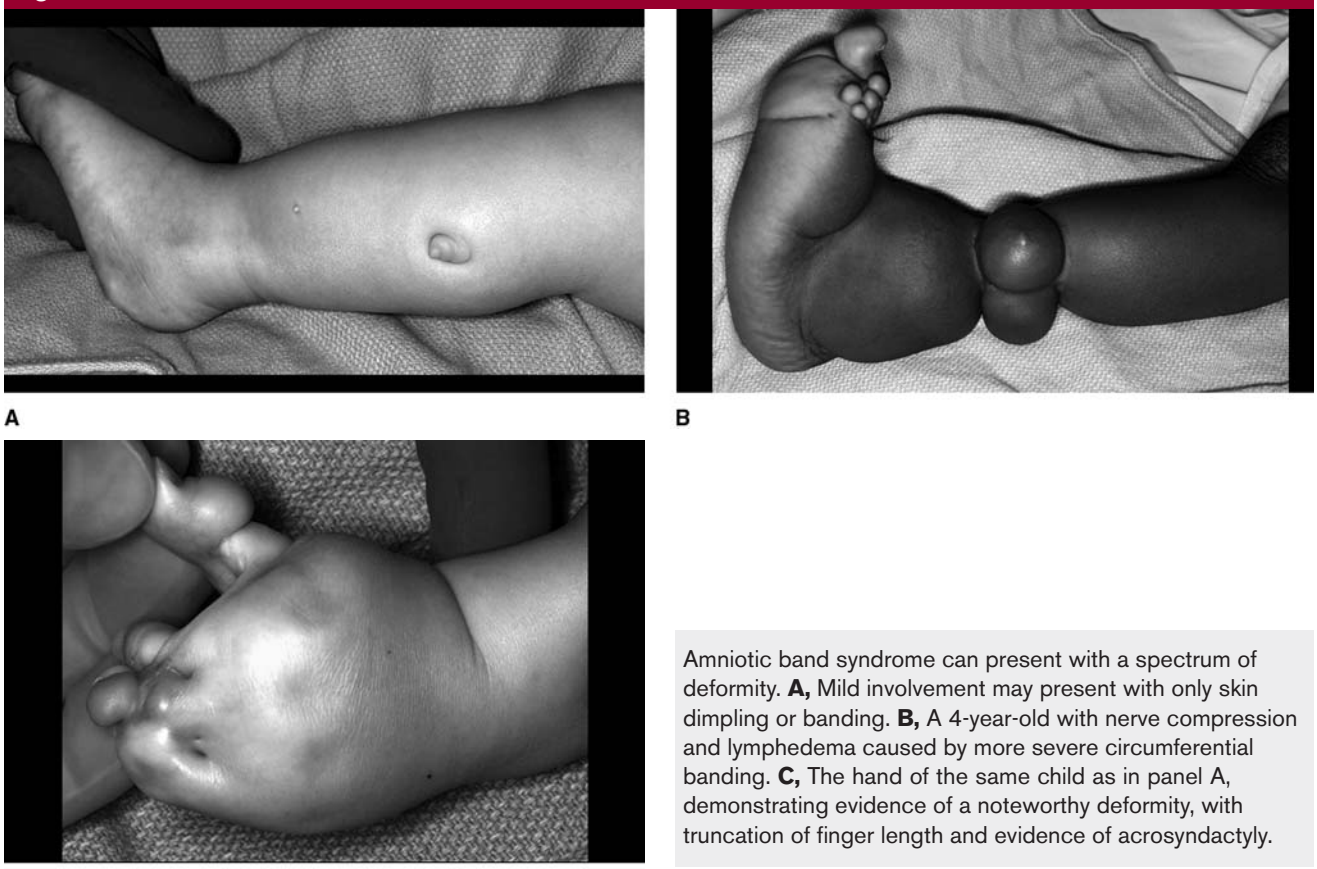
The stage of limb development at time of entrapment determines the physical appearance of the pathogenic process. The upper limb buds appear as small bulges at 24 days, with proliferation of an ectodermal ridge overlying mesodermal tissue. Upper limb morphogenesis occurs in weeks 4 through 8; lower extremity development occurs in weeks 5 through 9. By day 33, the hand plate is visible at the end of the limb bud. Digital rays appear during week 6 on the upper extremity and week 7 on the lower extremity. A process of programmed cell death sculpts these rays into fingers and toes. This process is complete by the end of week 8.

During week 5, skeletal elements develop from mesodermal condensations that appear along the long axis of the limb bud. Higginbottom et al[12] showed that fetal development and timing of amniotic entrapment greatly altered the expression of ABS. Rupture before 45 days' gestation led to a higher rate of central abnormalities, including severe central nervous system malformations, truncal defects, and abortion. Later rupture more frequently entrapped the limbs and spared the central structures.[12]

Protruding fetal structures are more vulnerable and much more likely to be entrapped than are other anatomic structures. Numerous observational studies have indicated that involvement is more common in the upper extremities, with a predilection for the distal segments of the hand.[1,8,13] Middle digits are most commonly involved, with relative sparing of the thumb. This pattern correlates with fetal positioning, with either outstretched fingers or a clenched thumb in the palm, protected by the other digits.[1,8,13]

No two cases of constriction bands are identical. Fetal banding gives rise to varying degrees of defects, ranging from skin dimpling and soft-tissue constriction rings to complete deletion (Figure 2). Soft-tissue strictures can disrupt neurovascular bundles, resulting in distal anesthesia, peripheral nerve palsy, vascu-

Figure 2



A B C

Amniotic band syndrome can present with a spectrum of deformity. **A,** Mild involvement may present with only skin dimpling or banding. **B,** A 4-year-old with nerve compression and lymphedema caused by more severe circumferential banding. **C,** The hand of the same child as in panel A, demonstrating evidence of a noteworthy deformity, with truncation of finger length and evidence of acrosyndactyly.

lar insufficiency, venous congestion, and lymphedema (Figure 2,B).[14]

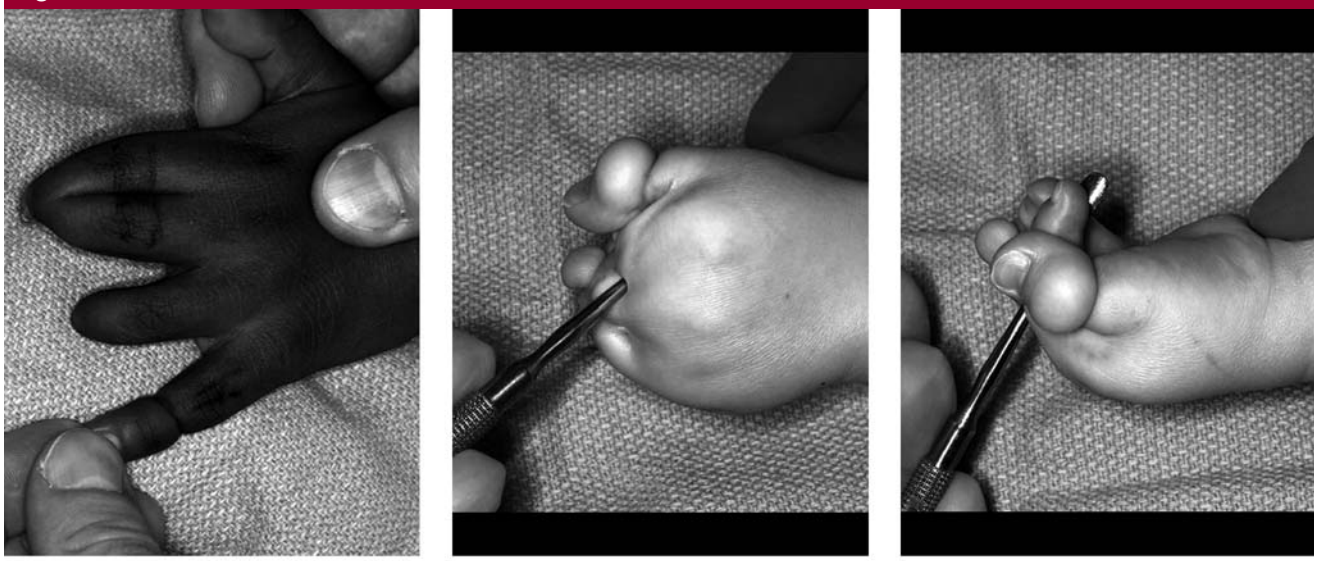
Finger involvement may result in acrosyndactyly, which is characterized by a constriction ring around adjacent digits, with distal soft-tissue webbing and epithelial sinus tracts at the base of the proximal phalanx (Figure 3). Complete digit separation proximal to the band is common[15] (Figure 3, A). The mechanism of acrosyndactyly is not fully understood, but several theories have been postulated. Some investigators have suggested that early bands block programmed cell death and separation of the distal digital rays.[8,11,12] Alternatively, constriction rings may cause distal hemorrhage of fragile vessels, leading to hematoma formation, scarring, and fusion of distal digits.[8] Patterson[16] argued that constriction bands around formed fingers may cause ulceration, with scar formation and ingrowth of ectoderm. The mechanism may, in fact, represent a composite of these theories.

The incidence of ABS has been reported at 1:1,200 births, with equal expression in males and females.[5] No autosomal inheritance pattern has been identified, nor has a connection been made to any infectious agent.[1,16] Some studies have shown a racial predisposition, but these findings may be influenced by selection or referral bias rather than evidence of a true predisposition.[17-19] An association with oligohydramnios may explain the increased frequency of clubfoot deformity; however, this finding is not ubiquitous. Researchers have postulated that early, transient oligohydramnios at the time of amniotic disruption is most likely causative.[7] Other anomalies that may be associated with oligohydramnios include calcaneovalgus, pes valgo planus, congenital hip dislocation, and congenital vertical talus.[20,21] Clubfoot deformity associated with ABS is often more challenging to treat than its idiopathic counterpart because of rigid equinovarus deformity and frequent deep distal circumferential banding of the skin and soft tissue.[22] Other anomalies, such as cleft lip, cleft palate, hemangioma, meningoceles, abdominal wall defects, and skin tags have been noted to occur in association with ABS.[13] Associated hand anomalies include syndactyly, acrosyndactyly, digital hypoplasia, symbrachydactyly, and campodactyly; pseudosyndactyly is the most common.[23]

Differential Diagnosis

The differential diagnosis of ABS includes vasculocutaneous catastro-

Figure 3



A B C

A, Acrosyndactyly of the index and long finger. Note the amputation of the distal phalanx of the ring finger and banding over the small finger. **B** and **C,** Severe acrosyndactyly with a probe in the sinus tract present at the base of the fingers.

Figure 4

A B

Neonatal gangrene in the upper **(A)** and lower **(B)** extremities. Telltale findings include the presence of skin slough and ischemic skin changes throughout the forearm.

phe of the newborn, brachysyndactyly, and transverse growth arrest. Several features can help to distinguish ABS, including the presence of constriction bands in alternative areas and the development of normal structures proximal to the band. In utero, amniotic bands should be distinguished from amniotic sheets and uterine synechia, which are adhesions caused by prior instrumentation, such as curettage, Caesarean section, and myomectomy. These bands protrude into the lumen of the amniotic cavity but do not adhere to the fetus and do not cause ABS. Patient history, ultrasound, and magnetic resonance imaging are useful for diagnosis.[24]

Vasculocutaneous catastrophe of the newborn also has been referred to as neonatal gangrene and neonatal Volkmann contracture. The etiology is not completely understood, but the condition seems to be related to ischemia from a vascular insult in the perinatal period. An altered fibrinolytic system, high plasminogen levels, and antiplasmin inhibitors have been implicated.[17,25] The patient with vasculocutaneous catastrophe may present with a swollen forearm, contracted finger, or digital necrosis (Figure 4). Skin lesions are frequently present on the day of delivery. These lesions are described as bullous or ulcerative and are localized to the dorsum of the forearm, wrist, and hand, with associated dis-

tal edema. Early fasciotomy for associated compartment syndrome can minimize adverse sequellae.[26]

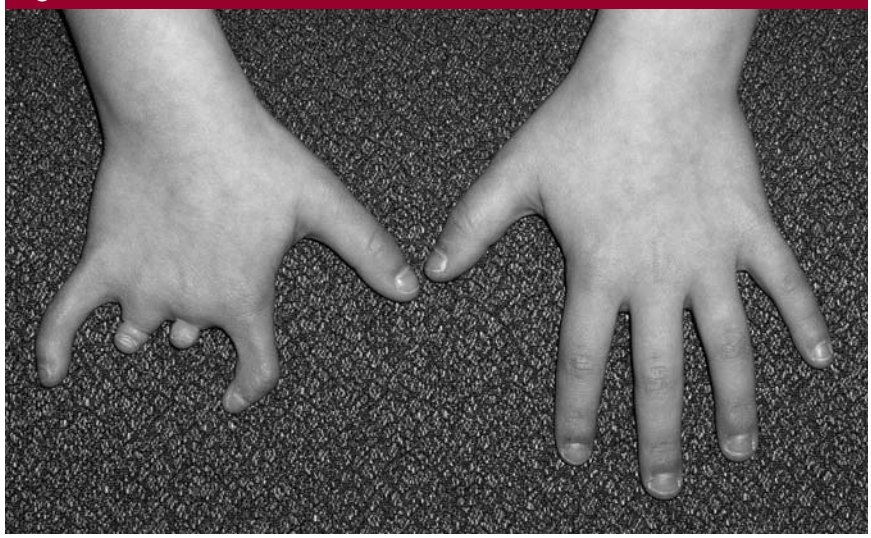
Symbrachydactyly usually affects the entire upper limb; the patient typically has a small hand with simple syndactyly (Figure 5). Bilateral cases are rare, with reported incidence ranging from 1.6% to 10%.[1,20,27] The presence of distal phalanges with fingernails represents ectodermal tissue not destroyed by the mesodermal event. Symbrachydactyly, which may be familial, has been associated with Poland's syndrome.

Transverse growth arrest, which constitutes a failure of formation, is occasionally seen in association with ABS.[28] The deformities in this clinical scenario are usually bilateral and may have an autosomal recessive inheritance pattern with variable expression.[23] Overgrowth and symptomatic diaphyseal amputation is infrequently seen.[27,29] The most common site of amputation is at the proximal third of the forearm as the result of disruption of the apical ectodermal ridge.

Classification

There are several proposed classification systems for ABS, based on location and severity of disease. The Ossipoff and Hall classification is based on involvement of three major anatomic areas: limb, visceral, and craniofacial. Swanson[28] developed a subclassification of upper extremity ABS. The most widely used classification system, described by Patterson, is based on severity of the syndrome, ranging from simple constriction rings, to constriction rings associated with deformity of the distal part, with or without lymphedema (Figure 2, B), to constriction rings associated with acrosyndactyly (Figure 3), to intrauterine amputation.[16] Patterson divided constriction rings associated with acrosyndactyly into three types: type I, conjoined fingertips with well-formed webs of the proper depth; type II, the tips of the digits are joined, but web formation is not complete; and type III, joined tips, sinus tracts between digits, and absent webs.[29]

Figure 5



A patient with symbrachydactyly who has characteristic preservation of nails without evidence of acrosyndactyly.

Clinical Presentation in the Hand

In the hand, clinical presentation of ABS can vary from slight indentations on the affected part to distal atrophy, lymphedema, acrosyndactyly, and amputation.[1] Histologically, mild constriction bands resemble naturally occurring skin creases. The dermis is thinned with atrophy of the underlying subcutaneous tissue. More severe constriction bands cause fibrous scar formation that can lead to severe vascular, lymphatic, and neural damage. Symptoms may progress as the child grows. Some patients present with ulceration of the base of the constriction ring or with firm skin protuberances on the dorsum of the fingers.[15,30] Neurovascular compromise leads to ischemia, nerve injury, and distal atrophy. Nail bed changes are common, with frequent absence of nail growth.[14] Temperature gradients have been measured across constricting rings.[6] Forearm constriction may cause palsy of the median and ulnar nerves, with resulting sensory defects and, possibly, permanent nerve damage.[14,18,31] Distal lymphedema is common, with histologically dilated lymphatics and wide open tissue spaces filled with excess fluid. Acrosyndactyly (ie, pseudosyndactyly) is common in affected digits. Although it may affect all fingers, the index, middle, and ring fingers are most frequently involved.[17]

Management

Management, which must be individualized, ranges from cosmetic repair to emergent limb-sparing band release. Z-plasty, W-plasty, and V-Y flaps are the mainstay for release of superficial and deep rings in the presence of good distal function. Deep rings with neurovascular compromise may require special attention to neurovascular reconstruction. Ischemia, which may lead to osteomyelitis, may necessitate amputation.[21] Finger and toe transfer, augmentation, lengthening procedures, bone

grafting, and composite toe transfer have been used to restore function in patients with amputation.[32,33] In the patient with acrosyndactyly, syndactylized digits are separated and web spaces deepened to enhance independent finger function.

Timing of Repair

Timing of repair is dictated by disease severity and predicted skeletal growth. Cosmetic repair of shallow constriction rings that present without distal edema may be done electively.[1] Release of bands associated with severe distal edema should be performed within the first few days after birth.[30,34] Left untreated, chronic stretching and fibrosis of the tissue develops, resulting in poor resolution of edema. In the patient with acrosyndactyly, repair in the first 3 to 6 months allows the best chance for proper longitudinal bone growth. This is particularly important when the joined fingers are of different lengths. Cerebral pattern hand use after separation of digits is optimized when repair is performed before age 1 year.[30]

Single Versus Staged Release

No clear guidelines have been established regarding the amount of tissue to release and reconstruct at one time. For digits that are circumferentially involved, release of the entire digit may be done in a single operation.[1,31,35-37] Digital endosteal blood supply and myocutaneous arteries are adequate for tissue viability distal to the constricting band.[1,31,35-37] Venocongestion that occurs after single-stage release, which is most problematic with deep bands, is treated with leech therapy or suture release. The rationale for the staged procedure is that once cutaneous circulation has been reestablished across the scar, the remaining 50% of the band can be removed safely. Six to 12 weeks between procedures is advised.[1] Most experts recommend single release for superficial bands and staged release for deep bands.

Surgical Considerations

Preoperative radiographs, nerve velocity studies, and vascular mapping may aid in surgical planning and allow for a more reliable prediction of postoperative outcome. Nerve velocity studies may be indicated for the patient who presents with distal neuralgia, paresthesia, or paralysis. For the patient with irreversible nerve damage, a thorough discussion of treatment and expected functional outcomes with the parents is indicated.[14]

Simple Constriction Rings

Treatment may not be necessary for superficial constriction rings because they may become less apparent as the infant fat is absorbed from the hands during growth. Grooves can be excised with mobilization of the adipose tissue and reapproximation of skin via Z-plasty. Simple excision without Z-plasty leaves a circular scar that may contract, producing a more noticeable defect than was present before repair.[6]

We prefer to use the Upton technique when repairing moderate to severe rings presenting with or without edema.[38] Before incision, one sidewall of the band is marked with ink and pressed against its partner to create a superimposed image. These markings help guide skin resection. All ring tissue is considered scar and should be removed. Skin flaps are then mobilized proximally and distally. Excess skin is left proximally until skin closure. The adipose tissue is then mobilized below the dermis and above the muscles or tendons. The proximal and distal adipose tissue is mobilized and approximated over the site of the annular groove to correct the contour deformity. During dissection, at least one or two large subcutaneous veins are preserved, along with the neurovascular bundle, thus helping to prevent postoperative venous congestion. In the case of deep dorsal grooves, there is often a paucity of dorsal veins, and a staged release should be performed. The skin is closed after the subcutaneous fat is approximated with fine absorbable suture. The skin suture line should be staggered from the adipose suture line (Figure 6, A-E).

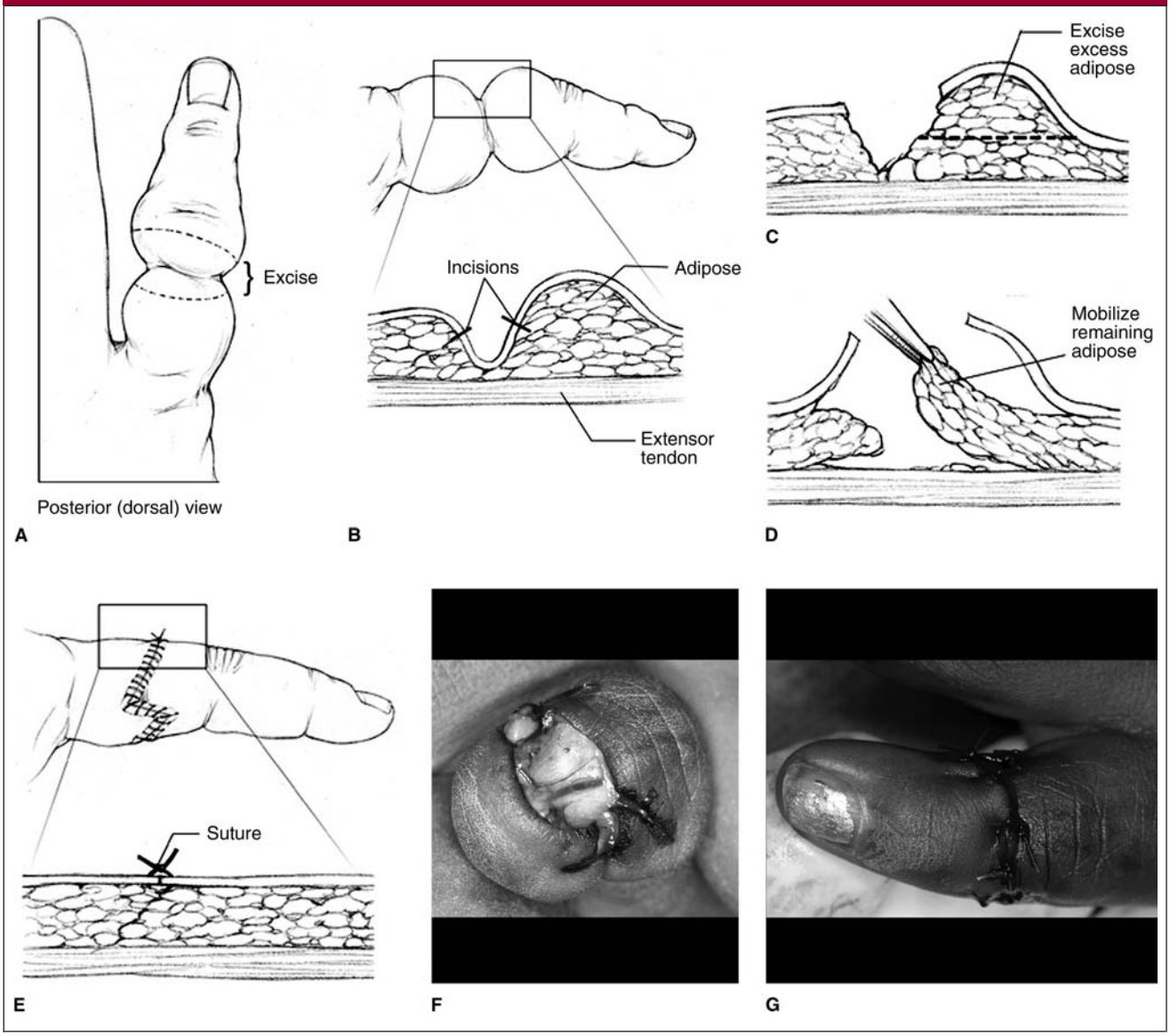
Skin approximation on the palmar surface usually does not require substantial skin advancement and may be closed with simple reapproximation. Dorsal digital deformities frequently require Z-plasty, which is best placed on the side of the finger in the midlateral line to minimize visible scarring. After mobilizing the subcutaneous tissue, the excess skin on the proximal flap is advanced distally and palmarly, starting at the dorsal midline. Occasionally, a broad constriction band necessitates a cross-finger flap to replace the deficient area. Bands in series are repaired in stages; the more distal band is corrected first.[6,38]

Constriction bands with considerable distal lymphedema, cyanosis, and circulatory embarrassment may progress quickly to ulceration or infection. In such patients, urgent release of the ring is required. The surgical principles are the same as above, with staged excision of the band. After the first operation, lymphedema decreases as the lymph channels are restored. The second release can be performed 2 to 3 months after the first. The surgeon must inform the parents that some residual edema may persist.[6] Postoperative extremity wrapping with an elastic circumferential bandage or compressive sleeve facilitates edema resolution.

Separation of Digits and Web Reconstruction

Treatment of acrosyndactyly varies depending on the extent of fusion

Figure 6



Surgical treatment of digital constriction rings. **A,** Dorsal view of a constriction ring demonstrating the area to be excised. The ring is excised in its entirety. **B** and **C,** Surgical correction begins with excision of all skin in the side walls and debulking of dorsal excess adipose tissue. **D,** Flaps are mobilized proximally and distally as needed to correct the contour deformity. **E,** Skin and subcutaneous closures are preferably staggered, and Z-plasties are positioned along the side of the digit, with straight line closure dorsally to minimize visible scarring. **F**, The constriction ring has been excised, and a dorsal bridging vein has been preserved. **G**, The appearance of the finger at time of closure. (Reproduced with permission from the Mayo Foundation.)

and associated band deformities. Narrow skin bridges can be cut or tied off in the neonatal unit. Deep bands with severe syndactyly pose a considerable challenge. Surgical planning should be guided by the dictum that the number of fingers is not as important as their spacing, length, bulk, stability, and control. Surgical approach is guided by severity of disease. Walsh[39] proposed a helpful distinction of groups into mild, moderate, and severe.

The patient with mild acrosyndactyly presents with three phalanges and two interphalangeal joints in the affected digits, typically with long clefts between the fingers. The surgical goal is early release to allow parallel growth of the fingers without articular deformities. Surgery should be performed by age 6 months, or as soon as possible for the patient who presents at an older age. Split-thickness skin grafts are used to cover the interdigital skin defect. To provide increased function, subse-

quent surgery may be performed to deepen the web space.[6]

The patient with moderate acrosyndactyly presents with two phalanges and one interphalangeal joint. Fusion of the digits will cause considerable articular defects, which must be released early to prevent permanent damage. Distortion of the distal fingertips is the greatest challenge. It may be difficult to determine which fingertip goes to which finger. Preservation of the distal tips is preferred over amputation because the tips may contain phalangeal buds that can be associated with articular spaces. In many patients, increased function can be gained by deepening the commissures.

The patient with severe acrosyndactyly has only stubby fingers, with one phalanx and no interphalangeal joints. The heterogeneity of the disease makes these cases very challenging. The surgeon frequently is faced with a bulbous portion and deep constriction rings. Fingers are short and often distorted. The surgical goal in these cases is early separation combined with commissural deepening. Further revision and deepening may be performed at a later date to optimize function. Digit lengthening procedures may be considered in patients with severe acrosyndactyly.

Surgical Technique

Numerous techniques have been described for separation of syndactyly, but all share general principles as presented by Flatt:[6] (1) create a broad commissural space from a local (usually dorsal) skin flap, (2) defat fingers to aid in closure, (3) close the sides of the fingers with zigzag local flaps, (4) cover bare areas with full-thickness skin grafts, (5) correct skeletal abnormalities, (6) release only one side of a finger at a time, (7) use magnification and meticulous technique, and (8) use postoperative dressings designed specifically for the pediatric patient to protect the repair.

Acrosyndactyly caused by constriction bands can be challenging to repair because of the associated rings and distal deformity. Each case is unique and must be individualized. In general, the fingers are separated using carefully planned Z-plasty skin flaps, and a broad commissural space is created based on a dorsal skin flap. The sinus tract is excised and may be used as a skin graft. This tract is generally not well suited for a commissural space because of its distal location and narrow space.[40] Skin flaps are mobilized, and the tissue is defatted. Fingers are separated proximally to the transverse intermetacarpal ligament.[30] Blood vessels and nerves are preserved, and the skin flaps are sewn down, with a full-thickness skin graft covering the bare area. The hand is dressed and inspected at 2 weeks postoperatively. Further dressing for 7 to 10 days is followed by removal and complete mobilization of the hand. When graft failure occurs, the area should be regrafted to avoid scars because these tend to tether the joint as the patient grows. Patients should be followed because they frequently require commissural revision in adolescence[6] (Figure 7).

Digital Aplasia

Clinical presentations of digital aplasia include thumb aplasia and cleft hand, as well as the monodactylous (one remaining digit) and peromelic (no remaining digits but intact metacarpals) types. Many procedures have been described, including on-top plasty, toe phalanx transplantation, metacarpal distraction, web space deepening, and thumb lengthening.

On-top plasty is used for the deficient thumb, or index or long finger. The distal end of the index or ring finger metacarpal is transposed with the proximal phalanx on a neurovascular pedicle.[30,41] This digital transfer lengthens the digit and deepens the web space when combined with index metacarpal shortening.

Web space deepening (ie, phalangization) is effective with thumb involvement. This deepening, as well as syndactyly release, allows for opposition. For the patient presenting with cleft hand, the rudimentary digits are removed, and the first web space is created or improved. Stabilization of boneless digital stumps can be accomplished with toe phalangeal transplants.

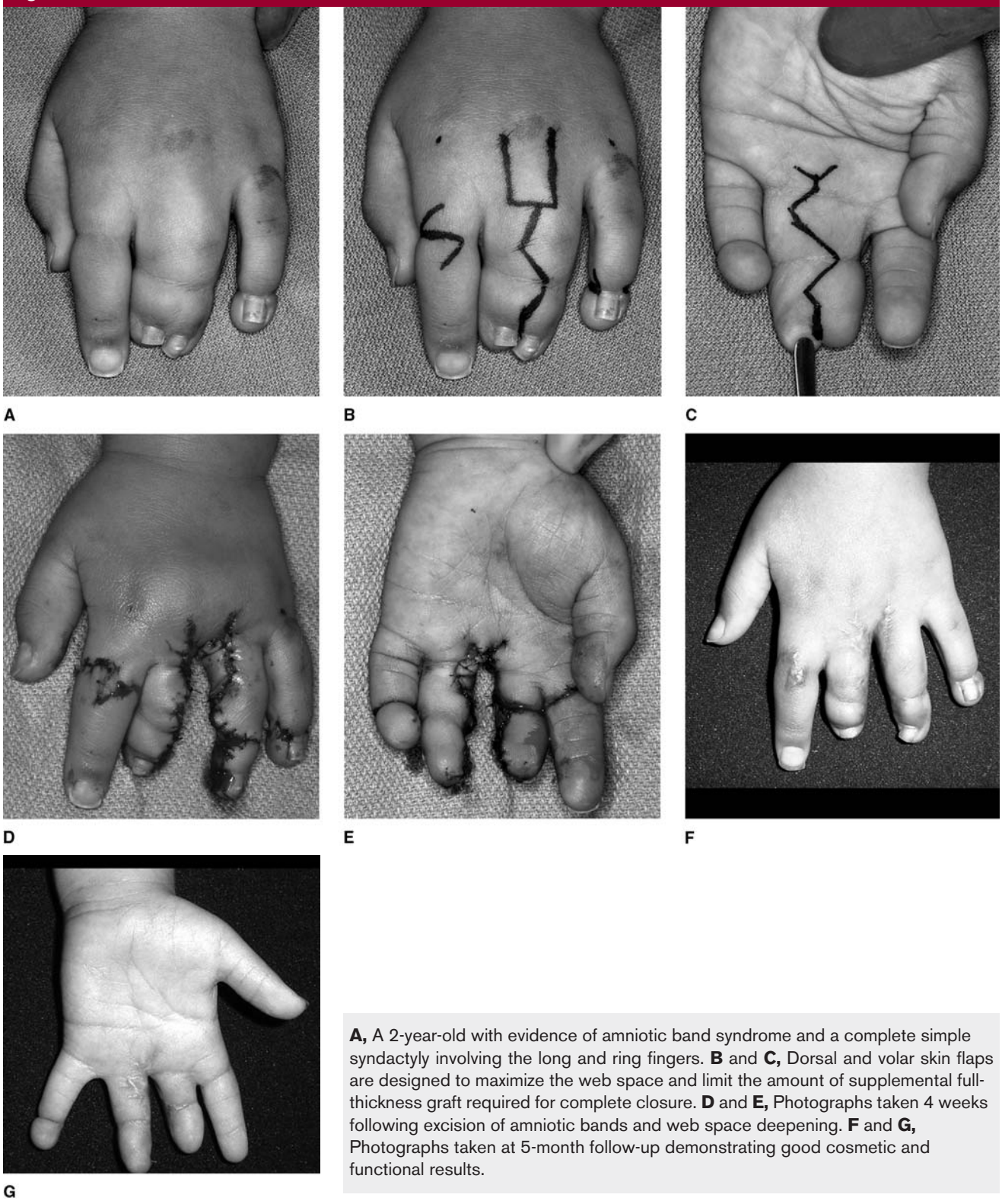
Monodactylous ABS is most commonly treated with vascularized toe transfer. This is done primarily at the ulnar side to provide pincer action. Limited mobility of the transferred digit allows the patient to use it as an ulnar post. When the thumb is the only digit involved, index finger pollicization or intercalary bone grafting with or without distraction osteogenesis can be done.[15]

Peromelic aplasia is an uncommon condition characterized by absent digits with intact metacarpals. Grip is possible only when the basal thumb joint is present. Some authors recommend thumb lengthening procedures using toe phalanx transfer.[42] Dual toe transfer may be preferable in some instances. The surgical goal is to provide a functional pincer group.[33] Prosthetics have been used as an alternative to toe transfer.

Treatment Recommendations for Digital Aplasia

The minimal requirement for reconstruction of a functional hand is the presence of at least two digits that can oppose with some power. One digit should be capable of motion so that it can grasp objects. The other digit need only act as a stable post against which the movable digit can pinch. To allow for prehensile movements, the digits require some form of cleft to divide them, which allows for the accommodation of objects. The digits must be sensate and pain free; otherwise, they will provide little benefit over a prosthesis.

Figure 7



A, A 2-year-old with evidence of amniotic band syndrome and a complete simple syndactyly involving the long and ring fingers. **B** and **C,** Dorsal and volar skin flaps are designed to maximize the web space and limit the amount of supplemental full-thickness graft required for complete closure. **D** and **E,** Photographs taken 4 weeks following excision of amniotic bands and web space deepening. **F** and **G,** Photographs taken at 5-month follow-up demonstrating good cosmetic and functional results.

In most patients with ABS, the thumb can be preserved; thus, treatment is often directed at improving the function of the remaining fingers. In the absence of a thumb, a vascularized toe transfer provides the best means of recreating a sensate, mobile thumb that can grow with the patient. In ABS, vascular

and neural structures are usually of normal caliber proximal to the constricting band, making microvascular reconstruction possible.

Reconstruction of the thumb and a single digit will enable some prehensile grasp. For optimal function, however, an additional digit should be reconstructed. The preservation or reconstruction of the thumb and two digits allows for the possibility of chuck pinch and improved subterminal pinch. The presence of a third digit will confer lateral stability in power pinch. A third digit also allows the patients to perform hook grip and power grasp. Span grasp is also made possible because the functional palmar space is increased, allowing for grasp of larger objects. For digital loss at the level of the proximal phalanx or the metacarpophalangeal joint, a free second toe transfer often provides the best long-term longitudinal growth and recovery of interphalangeal motion.

Complications

Complications from repair of ABS include infection, hematoma, flap necrosis, graft necrosis, and distal circulatory failure. Use of surgical magnification to preserve vessels and meticulous surgical technique are needed to minimize vascular compromise. Hematoma is usually prevented with strict hemostasis, carefully applied postoperative dressing, splinting of the hand, and judicious use of drains. Once the flaps have been dissected, the tourniquet may be released to obtain hemostasis before closure. Flaps should be closed in a tension-free manner, and dressings lightly placed. Minimizing excessive movement of the reconstructed part results in a lower incidence of flap or graft necrosis. Preventive measures should be taken to minimize the aforementioned common complications.[38,43]

Prenatal Management

Advances in prenatal diagnosis and fetoscopic techniques have made prenatal treatment of ABS within the realm of possibility. Several investigators have reported on fetoscopic release of tight amniotic bands, with variable results.[10,11] In one case, difficulty with amniotic bleeding necessitated fetal abortion. In another report, two patients underwent fetoscopic Nd-YAG laser release of limb-threatening bands from two hands and one leg in each patient.[10,11] Preoperative Doppler ultrasound studies revealed loss of blood flow, which was restored after band release. Following delivery of the first patient, one hand was severely malformed and was amputated to fit a prosthetic. One leg of the same patient was salvaged, with only a surface impression of the band evident at birth. An arm was salvaged in the second patient, but persistent lymphedema necessitated postpartum band release.

These cases represent an exciting advancement in prenatal diagnosis and treatment; however, preterm premature rupture of membranes, chorioamnionitis, bleeding, preterm labor, and fetal loss remain real concerns. Further, prenatal screening ultrasounds are frequently performed during the second trimester, which is too late for intervention in most patients. Much more research is required to advance the technology used in prenatal management of ABS.[9-11]

Summary

ABS is a nonfamilial condition caused by aberrant strands of amniotic membrane exerting strangulating forces on the developing fetus. Its clinical presentation is varied, with frequent expression in the upper extremity. Deformities range in severity from mild contour deformities to severely debilitating limb malformations. The condition has a heterogeneous expression, with frequent lymphedema, neurovascular compromise, and acrosyndactyly.

Management of ABS is focused on increasing function and development while providing a more aesthetic appearance. Ideally, release should be performed within the first year of life to optimize proper neurologic and skeletal development. Parents and children must have realistic expectations and must understand that repeat surgery may be necessary. Research on prenatal diagnosis and treatment is ongoing.

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Citation numbers printed in **bold type** indicate references published within the past 5 years.

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