

Amniotic Band Syndrome With Pseudoarthrosis of Tibia and Fibula A Case Report



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ARTICLE INFO

Level of Clinical Evidence: 4

Keywords:

amniotic band
pseudoarthrosis
preterm infant
rudimentary digit
Syme's amputation

ABSTRACT

Constriction of the lower limb by a congenital amniotic band has been proposed to explain the development of pseudoarthrosis of the tibia and fibula. We report a case of amniotic band syndrome in a preterm female infant with the presence of the tibia and fibula. She was born at 29 weeks of gestation with congenital amniotic bands and was noted to have a severely edematous left foot distal to the constricting band with rudimentary digits. The skin was pink and well perfused with palpable pulses. Radiography demonstrated pseudoarthrosis of the tibia and fibula. The limb deformities were managed with splints, positioning, and physical therapy. She underwent Z-plasty with soft tissue release on the left lower leg on day 7 of life. At 36 weeks of postmenstrual age, a repeat radiograph showed markedly improved growth of the tibia and fibula with mature new bone formation, which avoided the need for further surgical intervention. During the follow-up period, she underwent left Syme's amputation at 18 months. At 29 months of age, the child was able to walk and run without support. The findings from our case confirm the potential for bone growth in patients with amniotic band syndrome, once the constricting band has been released. Simple release of the constricting band with Z-plasty resulted in growth of mature bone, replacing the pseudoarthrosis and, hence, the patient did not require surgical amputation. Thus, one should be cautious when deciding on surgical amputation, even in the presence of pseudoarthrosis, especially in preterm infants. Early limb-preserving surgery with release of the constricting band with an intention to salvage the limb appears appropriate.

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Amniotic band syndrome (ABS) is a rare congenital malformation characterized by gross deformity of the limbs distal to the constricting band and pseudoarthrosis of the affected bones was considered one of the indications for surgical amputation of the affected limb.^{1,2} We report a case of ABS in a preterm infant (born at 29 weeks of gestation), in whom simple release of the constriction band with Z-plasty resulted in the growth of mature bone in the tibia and fibula, replacing the pseudoarthrosis and avoiding a knee-level surgical amputation. She only required amputation at the ankle level at the age of 18 months, received an ankle prosthesis at 22 months, and at the age of 29 months was able to walk and run

independently. The present study was performed in accordance with the ethical standards set forth in the 1964 Declaration of Helsinki and its later amendments. The parents of the present patient provided written informed consent for inclusion in the study. Details that might disclose the patient's identity were omitted.

Case Report

A preterm female infant was born at 29 weeks of gestation with a birth weight of 1384 g to a primigravida mother by nonelective cesarian section, because of fetal distress. An antenatal ultrasound scan at 20 weeks had detected the presence of amniotic bands on the upper and lower limbs. At birth, she was noted to have limb abnormalities, which included constrictive bands around the right ring and little fingers and evidence of auto-amputation of the right and left index and middle fingers with pseudosyndactyly. In the lower limbs, amniotic bands were present around the left lower leg with a severely

Financial Disclosure: None reported.

Conflict of Interest: None reported.

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Fig. 1. Photograph showing amniotic bands around the left lower leg with a severely edematous foot and constrictive bands around the second toe.

edematous, and the second toe of the right foot was also affected (Fig. 1). However, the skin was pink and well perfused, and the posterior tibial artery pulsations were easily palpable. A constrictive band was present around the right second toe with auto-amputation of the great toe. Apart from the limb abnormalities, the infant was healthy, without any associated anomalies. Regular monitoring of the limbs did not reveal evidence of acute compartment syndrome.

A skeletal survey performed on day 3 of life showed constriction of the left lower leg, with deficiency of the mid to distal tibia and fibula with pseudoarthrosis. The left foot was markedly swollen. Only the great toe phalanges were present, suggesting failure of development or auto-amputation of the other digits (Fig. 2). In both hands, constrictions were present at the level of the proximal interphalangeal joints of the index and middle fingers, with auto-amputation distally.

The limb deformities were managed with splints, positioning, and physiotherapy. On day 7 of life, with the patient under general anesthesia, the sloughy tissue over the left lower leg was removed, unhealthy skin edges were excised, and Z-plasty was used for closure with 5-0 plain gut suture. The area was covered with Xeroform, gauze, a crepe bandage, and a plaster-of-Paris cast. Postoperatively, the leg was kept elevated for 72 hours and the plaster-of-Paris cast was removed after 1 week. This resulted in a significant reduction in the edema of the affected area.

At 36 weeks of postmenstrual age, a repeat radiograph showed markedly improved growth of the tibia and fibula with mature new



Fig. 2. Radiograph showing only the presence of the great toe phalanges, suggesting failure of development or auto-amputation of the other digits.

bone formation (Fig. 3). Hence, the parents were reassured, and she was discharged home with a plan for regular follow-up examinations with the orthopedic and plastic surgery teams.

Subsequent to discharge from the neonatal intensive care unit, she underwent release of syndactyly of the fingers, release of the constriction rings of the leg, and amputation of the left big toe at 8 months of age (corrected age 5.5 months). At a corrected age of 1 year, the infant had normal scores on the developmental assessment in all domains, including personal, social, hearing, speech, hand and eye coordination, and performance. She was able to crawl, pull herself to a standing position, and tall kneel through both legs, all of which were age-appropriate milestones, given the limitations imposed by the problems with the left lower limb.

At the age of 18 months (corrected age 15.5 months), left Syme's amputation was performed with the patient under general anesthesia. A fish-mouth incision was premarked, leaving a generous heel pad. Careful dissection down to the bone was performed, and the talus and calcaneus were shelled out. The tendo-Achillis and plantar fascia remnants were preserved. The tourniquet was released, and the incision over the drain was closed. The drain was removed 24 hours later. A radiograph performed 4 weeks after the amputation showed good results (Fig. 4). Four months later, an ankle prosthesis was inserted, with good effect.

At the age of 24 months (corrected age 21.5 months), she underwent release of the syndactyly of the fingers, release of the constriction bands of the right ring finger and little finger, revision of the stump scar on the left leg, and excision of the constriction band over the right second toe with Z-plasty.

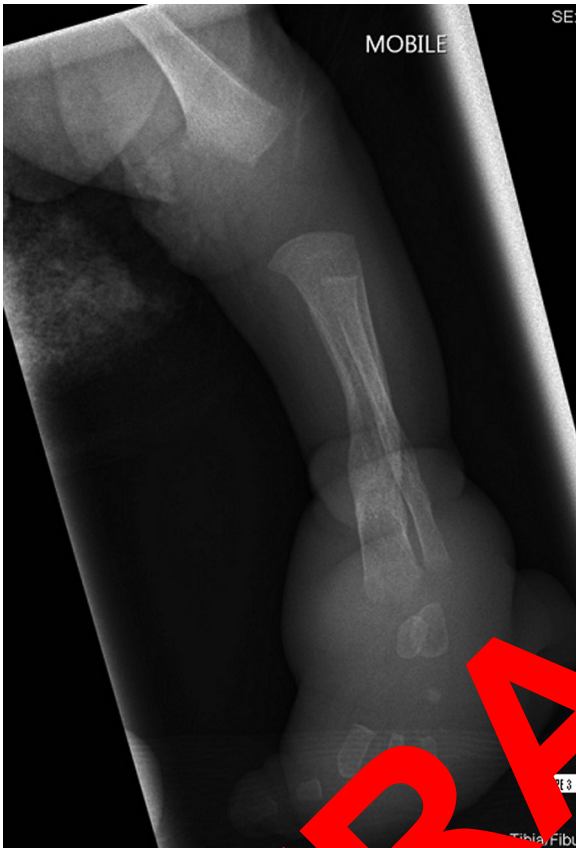


Fig. 3. Radiograph showing markedly improved growth of the tibia and fibula with mature new bone formation.

She underwent developmental assessment using the Bayley Scales of Infant Development – version III, at the age of 25.6 months (corrected age 23.1 months). At that time her physical growth was appropriate for her age, with a weight of 14.2 kg (98th percentile), length of 88.5 cm (98th percentile), and head circumference of 51.4 cm (98th percentile). The Bayley Scales of Infant Development cognitive scores (75th percentile), language scores (42nd percentile), motor scores (27th percentile), and social-emotional score (75th percentile) suggested the achievement of age-appropriate milestones. She was able to stand and throw a ball, walk independently, walk up and down stairs with support, and manipulate objects with her hands (e.g., grasping, picking up toys from the floor, building blocks, and drawing). She was able to recognize other's feelings at a level appropriate for her age. She was able to understand and respond appropriately to words and requests, and her expressive communication through words and gestures (expressive skills) were within the expected parameters for her age. She was able to complete puzzles and activities that required her sensory motor development, exploration and manipulation of objects, concept formation, and memory at a level appropriate for her age. At the age of 29 months (corrected age 26.5 months), the child was able to walk and run without support. Her parents were quite happy with her overall progress.

Discussion

ABS is a rare condition, with a reported prevalence ranging from 1:11,200 (3) to 1:50,579 (4). The spectrum is variable and includes limb deformities and auto-amputations at various levels, pseudo-syndactyly, polydactyly, and talipes. Possible facial anomalies include cleft lip, mid-facial clefts, nasal deformity, bony orbital clefts,



Fig. 4. Radiograph 4 weeks after amputation showing good results.

hypertelorism, eyelid colobomas, ptosis, ectropion, lacrimal outflow obstruction, and corneal opacities. Cranial deformations include hydrocephalus, microcephaly, encephalocele, exencephaly, acrania, acalvaria, and anencephaly. Neural tube defects and other internal organ anomalies such as heart defects and genitourinary abnormalities have also been well described (5–10). Familial ABS is associated with hereditary connective tissue abnormalities such as Ehlers-Danlos syndrome and osteogenesis imperfecta (11).

Vascular compromise, if present, distal to the amniotic band can lead to gangrene of the limb. Hence, careful and frequent monitoring and timely release of the constricting bands are essential (12–14). Although significant swelling of the soft tissue distal to the constricting band was present, our patient did not develop any acute vascular compromise.

Pseudoarthrosis of the tibia and fibula can cause problems with healing and reunion and might require multiple surgical procedures. Owing to the poor outcomes in the past, early amputation used to be given strong consideration (2). However, improved results in recent times suggest that early amputation might not be necessary. Instead, every attempt should be made to achieve reunion with excision of the pseudoarthrosis, bone grafting, and adequate fixation (15).

A newborn infant with ABS of the right leg with pseudoarthrosis of tibia was reported by Rajoo and Mennen (12). The foot was massively swollen and floppy with a bluish discoloration. No pulses could be palpated distally. The amniotic band that encircled the right leg distally and extending deep to the bone was excised urgently and the skin closed with multiple Z-plasties. Postoperatively, the bluish discoloration disappeared, and the distal pulses were palpable within 3 days. The radiograph showed complete remodeling of the tibia with no pseudoarthrosis after 6 weeks (12).

Ho et al (16) described the case of a 1-day-old neonate born at 28 ± 3 weeks of gestation with ABS of the forearm and pseudoarthrosis of the radius and ulna. Although the forearm and hand were soft

and viable initially, severe edema occurred after fluid resuscitation. This necessitated urgent operation at the bedside, with release of constriction bands using 2 longitudinal incisions over the forearm. Circulation was restored, and the pseudarthrosis healed with no further surgical intervention. Successful delayed reconstruction was performed when the baby was 7 months of age (16).

Zionts et al (17) described the case of twins with ABS. One twin had pseudoarthrosis of the radius and ulna with severe edema distally. The other twin had mild constriction around 1 leg. The affected infant's arm was immobilized with a plaster splint, elevated, and observed for 10 days. At 4 months of age, radiographic union was observed, and staged Z-plasty was successful at 7 months (17).

Martinot-Duquennoy et al (18) reported a case of congenital annular constricting band syndrome with open pseudarthrosis of the tibia and fibula in an older child aged 5 years. This was complicated by ischemia, a neurologic defect, and major lymphedema. The child underwent emergency treatment, combining external fixator placement and single-stage resection of the constricting band. Bone union was obtained within 4 weeks, with complete neurologic recovery at 6 months. The long-term results, with realignment and normal function, were satisfactory (18).

In conclusion, the results of our case have confirmed the potential for bone growth in patients with ABS, once the constricting band has been released. Hence, one should be cautious when choosing surgical amputation and determining the level of amputation, especially in the presence of pseudoarthrosis, especially in preterm infants. Early limb-preserving surgery with the release of the constricting band with an intention to salvage the limb appears appropriate.

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