

# Congenital amniotic band constriction of the proximal tibia: A Yucatan project case report

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The Yucatan peninsula, in southeastern Mexico, is home for a high incidence of various physical and neurological deformities. This provided Dr. Charles Southerland of Barry University School of Podiatric Medicine to establish the Yucatan Crippled Children's Project with the main goal of providing care for a community with limited resources. Over the years, the project has evolved into a crucial tool of academics and medical research for students and physicians and is partially sponsored by the International Foot & Ankle Foundation for Education and Research [1]. This clinical case report from the Yucatan Project addresses a rare condition of an in utero fibrous band composed of amniotic fluid constricting the proximal tibia of the fetus.

**Keywords:** amniotic band, proximal tibia, Z-plasty

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Proper development of the embryo involves a delicate sequence of spatiotemporal gene expression that governs tissue development and pattern formation [2]. At the beginning of the 2nd week of development, the blastocyst begins the process of implantation within the uterine wall. The embryoblast reorganizes into a bilaminar disc made up of two cell layers, the epiblast, and hypoblast [3]. The embryo becomes covered by two thin membranes derived from extraembryonic ectoderm and mesoderm, the amnion and the chorion. Fluid accumulates between the epiblast and the trophoblast cells lining the wall of the blastocyst, creating the amniotic cavity. Epiblast lines the new cavity forming the amniotic membrane. The extraembryonic mesoderm, derived from the hypoblast and primary yolk sac, proliferates and fills the cavity of the blastocyst. The extraembryonic mesoderm surrounding the yolk sac splits into two layers, creating the chorionic cavity [4]. The chorionic cavity separates the embryo and amniotic cavity from the outer wall of the blastocyst which is lined by the

chorionic membrane. (Larsen, 2015). The amnion produces fluid causing expansion of the amniotic cavity, eventually obliterating the chorionic cavity. This results in the fusion of the amnion with the chorion [5]. Folding of the embryonic disc pulls the amniotic cavity ventrally, enclosing the developing embryo within the amniotic sac (fused amnion and chorion). This allows the embryo to be suspended in a liquid environment which will allow for growth while protecting against mechanical injuries and adhesions [3, 6].

Development of the lower limb involves limb positioning and outgrowth, rotation, and the development of region-specific morphology. These events are governed by gene expression and morphogen gradients [3, 7]. The lower limb buds begin to appear late in the 4th week of gestation on the ventrolateral body wall, opposite the lumbar somites. The lower limb buds consist of an undifferentiated mesenchymal (mesoderm) core, derived from the lateral plate mesoderm, covered by

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an epithelial layer derived from ectoderm. The epithelium over the distal margin of the limb bud thickens forming the Apical Ectodermal Ridge (AER). The AER is a major signaling center that guides the proliferation and elongation of the underlying mesoderm through reciprocal signaling [7].

The limb bud core initially contains cells from the lateral plate mesoderm that will give rise to the skeleton, vasculature and the connective tissue of the future limb. Migrating neural crest cells will give rise to the sensory nerves, Schwann cells, and melanocytes [3]. During the 4th week, myogenic mesodermal cells begin migrating from the somites into the lower limb bud, creating the flexor and extensor muscle masses that will surround the future bone. As the limb bud enlarges, muscle mass will increase through mitosis until the mid-fetal period. During the 5th week, the paddle-shaped foot plates develop. Skeletogenesis begins during the 5th week establishing models for future limb bones. During the 6th week, lower limb segments become recognizable and most of the mesenchymal models begin the process of chondrification. By week 7, the digital rays of the foot plate become visible. Primary ossification of the cartilaginous templates begins as early as the 8th week in the lower limb [8, 9]. Vascular supply to the embryonic limb bud is primarily through the axial artery. During the 8th week, the axial artery involutes and the femoral artery system becomes the major vascular supply to the lower extremity, establishing the adult pattern of vasculature. Between the 6th to 8th week, the limb buds undergo long axis medial rotation and assume a more ventral position on the body wall. By week 7, the feet are located primarily in the sagittal plane. The tibial (pre-axial) borders are oriented cephalically with the extensor/dorsal surfaces facing laterally and the flexor/plantar surfaces facing medially. Rotation continues through the fetal period, eventually orienting the knees cephalically the extensor surface anteriorly/superiorly and the flexor surface posteriorly/inferiorly. During the fetal period, the thigh continues to rotate internally, while the foot dorsiflexes and pronates [4, 10].

### **Background on Congenital Amniotic Bands**

Congenital amniotic band constriction, also known as amniotic band syndrome and Streeter dysplasia, was first defined by Montgomery in 1832 [11]. The two proposed explanations are the intrinsic and extrinsic theories. The intrinsic theory, proposed by Streeter, describes the composition of the bands as tissue that

has been left behind due to a defect in the development of germ plasm [11]. However, the most widely accepted pathophysiology is the extrinsic theory proposed by Torpin in 1965 [12]. This theory explains that the fibrous bands occur due to the amnion separating from the chorion. The amnion further wraps around a part of the embryo constricting its growth and causing malformations. As previously mentioned, the amnion is responsible for creating amniotic fluid. Therefore, the developmental injury leads to a deficiency in amniotic fluid that contributes to the displacement of fetal limbs into the chorionic cavity, where compression occurs [12].

Patterson's classification divided the stages of these congenital amniotic bands based on the severity and outcomes of the constriction [13]. Stage A is classified as simple constriction rings. Stage B describes constriction rings with deformities of distal aspects of limbs with or without lymphedema. Stage C involves constriction rings with fusion of distal limbs, such as acrosyndactyly. Lastly, Stage D is the most severe and leads to intrauterine amputation. Additionally, two or more of these symptoms are needed to diagnose congenital amniotic band constriction. This classification system can provide insight into proper treatment options according to the extent of the constriction [13].

Although no concrete etiology has been identified, literature described a few possible causes of congenital amniotic bands such as local disruption of vasculature, constricted uterine environment caused by oligohydramnios or large benign tumors of the uterine myometrium.

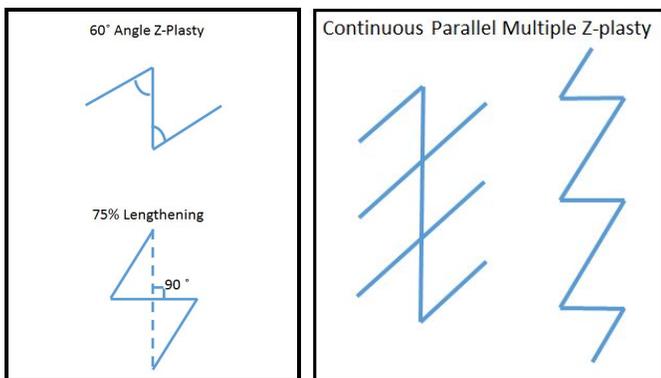
Constriction bands can lead to a myriad of problems such as vascular compromise leading to ischemia and venous congestion. More severe cases have been reported to cause natural limb amputation, pseudosyndactylism, anencephaly, craniofacial abnormalities and abdominal wall defects [14, 15].

### **Clinical Presentation**

A 4-year-old female presented to the Yucatan Project clinic in February of 2014. Neonatal records were not provided. Upon physical examination, a congenital band constriction was noted to the proximal tibia of the right leg. Skin texture is smooth with no signs of infection, hyperkeratosis or xerosis. Non-pitting edema was present in the right knee and leg both proximal and distal to the constriction band as seen in Figure 1a and 1b.



**Figure 1** a: Weight bearing anterior clinical view, showing right leg constriction band, and b: Non weight bearing clinical view.



**Figure 2** a: 60 degree Z-Plasty and, b: Multiple Parallel Z-Plasty.

According to the Patterson classification system, this patient would be categorized into Stage B with lymphedema.

### Surgical Procedure

Continuous parallel multiple stereometric Z-plasty surgical procedures were performed March of 2014 with the purpose of releasing the constriction band. The first step of the Z-plasty is to draw a line for the central arm incision directly along or parallel to the skin contracture. The additional two arms of the Z-plasty are then added to the edges of the central arm, forming a 60-degree angle between the central and additional arms. With a 60-degree Z-plasty, 75% lengthening should be achieved, as seen in Figure 2a. The length of the central arm should also have the same length as the two additional arms.

The skin dissection should only go with the extent of the deep fascia, preserving the vascular supply of the two newly formed triangular flaps, as shown in Figures 3 and 4 [16, 17]. The Z incision then allows for transposition of the triangular flaps to gain length along the longitudinal axis at the expense of the transverse skin narrowing.



**Figure 3** Intraoperative image following skin incision.



**Figure 4** Intraoperative image following Z-Plasty.

The multiple parallel Z-plasty are connected throughout the contracture line as seen in Figure 2b [18].

In November of 2014, the patient presented to the clinic for a 9-month postoperative follow-up visit. The incision site was healing well with no signs of infection. Edema was significantly reduced on the right proximal tibia where the surgery was performed. Mild discoloration was present at the scar site, as seen in Figure 5.



**Figure 5** Nine month postoperative view of the right leg.

The patient was noted to have mild residual foot drop on the right side, most likely due to damage of the common peroneal nerve from the previous constriction band or the surgical procedure itself.

### Discussion and Results

Congenital amniotic band constriction can occur in any area of the body due to disruptions in fetal development. This deficiency in amniotic fluid causing the displacement of body parts can be limb or life-threatening. If detected early, proper care can be given to avoid drastic results.

In the literature, several surgical procedures have been reported to help patients that suffer from congenital amniotic band constriction: Z-plasty, W-plasty, syndactyly release, skin and nerve grafting, and tendon transfers. The Z-plasty, as described above, still remains as the main skin lengthening surgical correction. The W-plasty is a description of multiple W-plasties, which may be necessary when the constriction is severe in order to effectively reduce the lymphedema [19]. Syndactyly or acrosyndactyly has been reported as a common complication of these bands, in which a syndactyly release has been performed. Skin grafting has been utilized to cover the area of tissue loss caused by syndactyly release

[11]. Following the aforementioned surgical procedures, one of the postoperative complications is nerve paralysis. A segment of the sural nerve can be used to replace the damaged nerve. Lastly, tendon transfers have helped to recover muscle function of previously limited range of motion [9].

Numerous lower extremity issues can also arise due to these congenital amniotic bands. In a study that involved 83 patients with congenital amniotic band constriction, 19 patients developed clubfoot deformities. Of these 19 patients, 10 patients had compression of the common peroneal nerve which led to paralysis of the clubfoot deformity [20]. Another case report identified a patient that had amniotic band constriction of both left and right lower extremities. The left lower extremity was constricted at the distal tibia and fibula to the point of autoamputation and there was no development of bones in the foot. A below the knee amputation had to be performed to the left lower extremity, as it was deemed unsalvageable. The right lower extremity constriction led to lateral bowing of the tibia and fibula, as well as a clubfoot deformity. A W-plasty and Z-plasty was performed on the right lower extremity to release the underlying tissue. The patient was eventually able to ambulate with the help of a prosthesis on the left side [21].

In the case report presented in this paper, the constriction band at the proximal tibia was categorized as Stage 2 according to Patterson's classifications. It did not lead to any malformations of the distal aspects of the lower extremity. The patient received a Z-plasty procedure and was able to ambulate with a mild residual foot drop. A future surgical option for the patient can be a sural nerve graft to replace the damaged common peroneal nerve as an attempt to recover dorsiflexion at the right ankle joint. Due to the lack of documentation provided with this patient, it is difficult to formulate the reasons that may have caused these amniotic constriction bands. Additionally, the patient did not return for further follow-up visits, which prevented documentation of the patient's progress or other complications of the surgery.

### Conclusions

Congenital amniotic constriction band is an in utero fibrous band that constricts the fetus potentially resulting in limb and life-threatening conditions. Identifying the presence of constriction bands early in pregnancy can reduce dangerous effects. However, if

the condition is not detected during pregnancy, surgical intervention is necessary to prevent further complications for the patient. Due to the rarity of congenital amniotic bands, documentation of affected patients is crucial to further expand health care professional's knowledge on the condition and treatment options.

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