Congenital Amniotic Constriction Band of the Pelvis: First Reported Case and Proposed Classification Criterion

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Summary: Congenital amniotic band syndrome (ABS) is a complex disorder characterized by a spectrum of rare congenital anomalies, with constriction bands as their hallmark manifestation. Constriction bands about the thorax, abdomen, or pelvis are rare. There is no consensus in the classification criterion for constriction bands of the thorax, abdomen, or pelvis in the literature. We present the first true pelvic constriction band in a newborn female with ABS. This case adds valuable information to the literature on ABS. The authors propose a classification criterion to further classify constriction bands of the thorax, abdomen, and pelvis using accepted and reliable anatomical landmarks. (Plast Reconstr Surg Glob Open 2019;7:32336; doi: 10.1097/GOX.0000000000002336; Published online 25 July 2019.)

Congenital amniotic band syndrome (ABS) is a complex disorder characterized by a spectrum of rare congenital anomalies, occurring in about 1 of every 10,000 live births. While there are many diverse manifestations of ABS, constriction bands about the extremities are among the most common, with consequential lymphedema, vascular and neural deficits, and aesthetic loss. Few, rare cases of constriction bands involving the thorax and abdomen have been reported. Although some of these cases were reported as pelvic constriction bands, none of them described a constriction band within the pelvis. Cases involving bands around the abdomen near the umbilicus are reported as both abdominal and pelvic constriction bands, with a lack of consensus in classification criteria. To date, there are no true constriction bands encompassing the pelvis reported within the literature. Therefore, we describe the first case of a true pelvic constriction band in a newborn female with ABS and propose a classification criterion to help future providers accurately classify congenital constriction bands of the thorax, abdomen, and pelvis.

CASE REPORT

We were called to consult on a few hour-old newborn female, born at 37 and 2/7 weeks. She was prenatally diagnosed with an amniotic band involving her right pelvis. Additionally, she was diagnosed with left multicystic dysplastic kidney, ventriculomegaly, and a single umbilical artery on her prenatal ultrasound at 15 weeks. Her mother was an otherwise healthy 31-year-old G5P3104. Mother’s medications were prenatal vitamins and denied alcohol, tobacco, and any recreational drug use. Mother’s previous children were all healthy except 1 with autism born 5 weeks premature.

On exam the newborn had a circumferential, obliquely oriented constriction band overlaying the right pelvic rim extending over the mons pubis along the left groin crease and dividing the left gluteal muscles as it extended posteriorly. Additionally, there were constriction bands around her first, second, and third toes of her right foot. Her right leg was held in an open frog leg position with concerns for hip dysplasia or frank dislocation. She had significant swelling and lymphedema of the soft tissues distal to the constriction band that extended to the ankle. Spontaneous movement, and ultrasound, confirmed arterial flow were present in the leg, foot, and toes. Genetic testing was normal.

On radiographic imaging, her pelvis showed bilateral developmental dysplasia of the hip, with an abnormally positioned left ischium more superior and medial. Lower extremity ultrasound showed small caliber vessels but a
patent deep venous system of her right leg with slowed outflow. Due to the significant swelling of her right leg and concern for limited venous outflow, she was taken to the operating room on her second day of life (Fig. 1).

Under general anesthesia, the anterior portion of the constriction band was excised in its entirety and closure was performed with multiple sequential Z-plasties. Postoperatively, there was significant improvement of right leg lymphedema. The posterior aspect was also excised and closed in similar fashion 8 weeks later because of mild persistent lymphedema.

The obliquely oriented constriction band was noted to extend down to the junction of external iliac and femoral vessels anteriorly extending over the pubic symphysis and through the left groin crease lateral to left ischial tuberosity. The band transected the left gluteus maximus and medius muscles extending down to sacral bone before wrapping around superiorly over the iliac crest and medially to the ASIS where it rejoined forming a complete band.

The postoperative course of each surgery was uncomplicated, and at 6- and 12-month follow-up appointments the patient had good resolution of lower leg lymphedema with improved contour (Fig. 2). There remains some excess tissue and symmetry issues which will be addressed once the child is older.

**DISCUSSION**

This case presented with a unique challenge as it was the first to involve the pelvis and its particular anatomy including the external genitalia, ilium, gluteal muscles, and iliac vessels. It was also unique from abdominally based
constriction bands as it neither followed a single segment nor was higher on the back. Instead, the high side was laterally based over the right iliac crest. The unique band location in the pelvis was also causing lymphovascular obstruction, requiring a more urgent surgical intervention compared with abdominal-based constriction bands, which can be managed in a more delayed fashion.

A source of confusion within the literature of ABS is the lack of consensus in accurately classifying truncal constriction bands. In the literature concerning constriction bands of the trunk, 4 cases claim to be pelvic constriction bands. Upon review, all 4 cases were within the periumbilical region and were identical in presentation to other cases of abdominal constriction bands. Various methods for classifying these constriction bands were also used. The confusing and subjective nature of these reports show why there are cases of similar constriction bands reported as both abdominal and pelvic in the literature. Thus, there is a need to distinguish between constriction bands of the thorax, abdomen, and pelvis.

Thoracic, abdominal, and pelvic constriction bands should be distinguished using predetermined anatomical planes and landmarks. We propose classifying bands located between the clavicles and transpyloric plane as thoracic constriction bands, bands located between the transpyloric plane and intertubercular plane as abdominal constriction bands, and bands located between the intertubercular plane and ischial tuberosities.
plane and ischial tuberosities as pelvic constriction bands (Fig. 3).

The transpyloric plane passes through the vertebral body of L1, whereas the intertubercular plane passes through the body of L5 and iliac tubercles. Compared to previous methods, these landmarks are more reliable, anatomically accurate, and are visualized with imaging, making it easier and more reproducible for others to precisely identify and classify the constriction band. Using this criterion, the 4 previously reported pelvic constriction band cases are more accurately classified as abdominal constriction bands.

CONCLUSIONS

This is the first reported case of a true constriction band about the pelvis. A criterion for classifying thoracic, abdominal, and pelvic constriction bands was also proposed. Thoracic constriction bands should be located between the clavicles and transpyloric plane, abdominal constriction bands between the transpyloric and intertubercular planes, and pelvic constriction bands between the intertubercular plane and ischial tuberosities.

Using this criterion adds clarity when identifying constriction bands about the thorax, abdomen, and pelvis.

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