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Laparoscopic-assisted surgical reconstruction of a rare congenital abdominal wall defect in two children misdiagnosed with prune-belly syndrome

Andrew I. Fishman, Israel Franco*

Department of Pediatric Urology, New York Medical College, Valhalla, NY 10595, USA

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Abstract *Purpose:* Abdominal wall laxity is typically associated with prune-belly syndrome (PBS). Incomplete forms of PBS have been rarely reported with only the abdominal wall laxity. Herein, we describe a rare congenital abdominal wall defect that has been confused with PBS and illustrate the laparoscopic-assisted surgical technique used for reconstruction.
Materials and methods: Two boys with symmetrical, bilateral absence or hypoplasia of the internal and external oblique muscles and no genitourinary abnormalities underwent a laparoscopic-assisted abdominal wall reconstruction utilizing the technique previously described by Firlit. Each patient had a Ct scan which confirmed the absence of the oblique muscles. In one patient EMG data confirmed no electrical activity of the obliques. Radiologic evaluation of the urinary tracts revealed no abnormalities. The abdominal wall was plicated utilizing bilateral subcostal incisions.
Results: Both patients had excellent cosmetic and functional results with no weakness or bulging of the lateral abdominal wall and improvement of associated symptoms.
Conclusions: We believe these two cases and their congenital abdominal wall defects are a rare and often misdiagnosed muscular deficiency separate from PBS. The novel laparoscopic-assisted surgical technique illustrated is feasible and highly successful for these and possible other patients with similar rare congenital abdominal wall defects.

* Corresponding author. Pediatric Urology Associates, 150 White Plains Rd, Tarrytown, NY 10591, USA. Tel.: +1 914 493 8628; fax: +1 914 493 8564.
E-mail addresses: franco.pua@gmail.com, isifranco@gmail.com (I. Franco).

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Introduction

Abdominal wall laxity is typically associated with Prune-belly Syndrome (PBS). This syndrome has been classically described as an anterior abdominal wall laxity with muscular deficiencies of the rectus muscles with an ensuing coexistence of undescended testicles and genitourinary tract anomalies [1]. Pseudoprune-belly syndrome has been described as an incomplete form of prune-belly syndrome [2,3]. Pseudoprune-belly patients typically have the prune-belly urinary tract anomalies without the coexistence of abdominal wall or testicular abnormalities [2,3]. Incomplete forms of PBS have been rarely reported with only the abdominal wall defect as the only abnormality.

Herein we describe two children referred to the pediatric urologist with bilateral abdominal wall laxity. Both patients were diagnosed with congenital, bilateral abdominal wall defects from birth and labeled as having an incomplete form of prune-belly syndrome. An exhaustive search of the literature has led to the discovery of few cases with similar, but not identical features of these lateral abdominal wall defects. To the authors' knowledge it appears that this abnormality and its operative repair are previously undescribed and may be entirely separate from PBS, but may share a common etiology. Below we will describe the clinical characteristics and discuss a novel operative technique used to repair these congenital abdominal wall defects.

Methods

Patient 1

A 24 month old boy had symmetrical, bilateral, segmental atrophy or hypoplasia of the internal and external oblique muscles with normal intact functioning rectus muscles on physical exam. There was bulging primarily in both flanks, subcostal between the ribs and iliac spines. The sizes of the defects were approximately 5—6 cm wide (Fig. 1). This patient had pectus excavatum and bilateral descended testes. Most notably there was a history of in-utero megacystisis and urinary ascites that spontaneously resolved during the mother's pregnancy. This patient underwent extensive genetic and neurologic testing but had no other evidence of any other muscular defect.

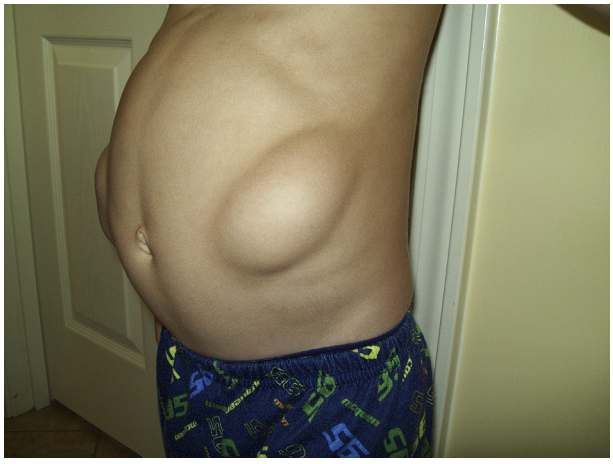

Figure 1 Abdominal wall bulges in the subcostal area in patient 1.

Patient 2

A 3-year old boy had the identical abdominal wall defect as Patient 1. He had symmetrical, bilateral, segmental atrophy or hypoplasia of the internal and external oblique muscles with normal intact functioning rectus muscles. There was bulging primarily in both flanks, subcostal between the ribs and iliac spines. The sizes of the defects were approximately 4—5 cm wide. This patient had pectus excavatum and bilateral descended testes. This patient underwent extensive genetic and neurologic testing but had no other evidence of any other muscular defect or any known in-utero abnormalities.

Both children were referred to the pediatric urologist because of congenital, bilateral abdominal wall defects seen at birth thought to be consistent with prune-belly syndrome. Both children were rediagnosed as not having prune-belly syndrome after radiologic imaging showed no signs of genitourinary tract abnormality. CT imaging revealed both children had attenuation of the internal and external oblique muscles with normal intact anterior rectus muscles. Both children had a history of chronic constipation, straining with bowel movements, but were able to perform sit-ups without assistance. Both children underwent a variation of Franco's [12] laparoscopic-assisted abdominal wall reconstruction to fix their congenital abdominal wall defects.

Operative technique

At operation, general anesthesia is administered and an epidural catheter is placed by the anesthesiologist for post-operative pain management prior to the positioning of the patient. Next, two inflatable blood pressure bags are placed on each side of the patient's flanks to be able to elevate each flank for easier access. The patient is placed in the supine position, prepped, and a foley catheter is inserted.

The first step of the operation is the placement of the laparoscope. A superior incision on the edge of the umbilicus is made and dissection of the subcutaneous tissue is carried down to the fascia carefully to identify any urachal remnant. This is a variation to the previously described procedure that placed the laparoscope subxyphoid [12,13]. A 5-mm trocar is placed into the abdomen and insufflated with carbon dioxide to a pressure of 12 mm Hg throughout the whole case. The peak airway pressure is monitored throughout the case to prevent over tightening of the abdominal wall. A 0-degree 5-mm lens is placed through the trocar and the abdomen is inspected to identify any evidence of bowel injury or adhesions to the anterior abdominal wall.

Once the abdomen is inflated with CO_2, the bilateral abdominal wall defects are clinically apparent and the lateral edges of the defects are marked out with a marking pen on the skin. A transverse skin incision is made at the most prominent point of the defect, and the skin is elevated, undermined and freed to the lateral edges of the marked defect. This will be done bilaterally for both