

A Novel Technique of Abdominoplasty for Prune Belly Syndrome

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Abstract

Prune belly syndrome is an extremely rare congenital condition occurring predominantly in males. This triad syndrome comprises of partial or complete deficient abdominal wall muscles, undescended testes and dilated urinary collecting system. We present the case of a 2-year-old male patient, who presented with classic prune belly syndrome, operated with modification in classical Monfort technique of abdominoplasty. The basis of this approach was to save and use the supraumbilical abdominal wall which has better tensile strength. The infraumbilical skin was mostly discarded. The outcome was better in terms of cosmetic appearance as the scar was limited to the lower abdomen and better anterior abdominal wall strength.

Keywords: Abdominoplasty, prune belly syndrome, undescended testis

INTRODUCTION

Prune belly syndrome is an extremely rare congenital anomaly with incidence of about 1:30,000 live births. This almost always occurs in boys.^[1] The three components of this anomaly are partial or complete absence of abdominal muscles, bilateral cryptorchidism and urinary tract abnormalities mainly, vesicoureteric reflux and hydroureteronephrosis.^[2] Due to deficit of abdominal muscles, there is decreased support and compression of intra-abdominal organ and reduced respiratory effort. There are several techniques to repair abdominal wall weakness in these patients. In this article, we have used modified abdominoplasty, which had improved aesthetic and strength of abdominal wall.

PATIENT PROFILE

Our case was a 2-year old male child who presented with gross laxity of abdominal muscles, more in infraumbilical aspect [Figure 1a and 1b] with bulging out of intra-abdominal organs, bilateral undescended testis and laxity of skin over the phallus also. The patient was not having any pulmonary problem. The urinary stream and frequency were normal. Blood reports were normal. Ultrasound abdomen was suggestive of mild bilateral hydronephrosis and bilateral intra-abdominal testis. Retrograde urethrogram was suggestive of dilated anterior urethra [Figure 1c].



Figure 1: (a) Laxity of the anterior abdominal wall with bilateral undescended testis and megalourethra. (b) Note that the laxity of skin is more on the lower abdominal wall than that in the upper abdominal wall and dilated anterior urethra. (c) Retrograde urethrogram showing dilated anterior urethra

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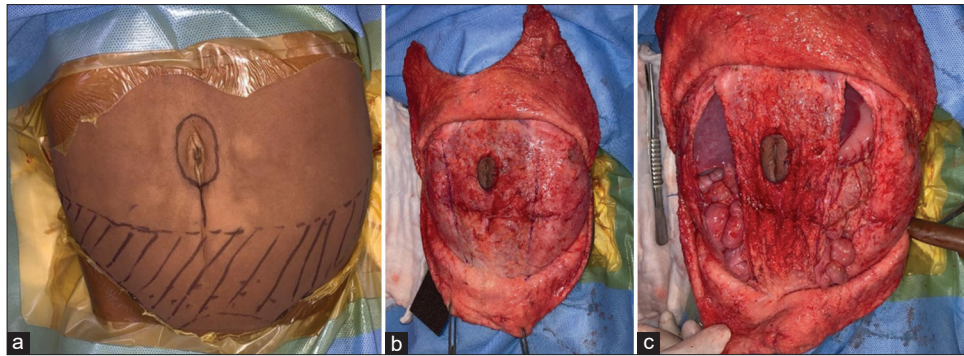


Figure 2: (a) Skin marking of the excision plan. (b) Skin flap of the anterior abdominal wall elevated superiorly and inferiorly with a rim of skin left in the peri-umbilical region. (c) Creation of the rectus muscle done

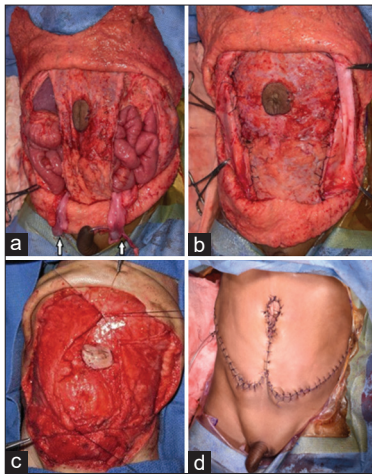


Figure 3: (a) Fowler–Stephen single–stage orchidopexy. (b) Apposition of the lateral borders of the recti to the undersurface of the ipsilateral musculofascial layer of the anterior abdominal wall. (c) Double breasting of the musculofascial layer anterior to the created rectus. (d) Final outcome after removal of the excessive skin



Figure 4: Figure 4 Follow up images showing (a) Taut abdominal wall and (b) Testis well descended in the scrotum

TECHNIQUE

The patient was positioned in supine position with marking of the incision over the infraumbilical part of the abdominal wall. An inverted T-shaped incision from the tip of the 12th rib of

one side to the opposite side in a curvilinear pattern was made about 5 cms inferior to the umbilicus. The vertical axis of the inverted T was from the pubic symphysis up to the umbilicus, with preserving it, leaving a rim of skin of about 2 cm around the umbilicus [Figure 2a].

Full-thickness skin was lifted up with exposure of muscles and fascia from the xiphoid to the pubic symphysis longitudinally and up to the Anterior Superior Iliac Spine (ASIS) transversely with preserving the umbilicus [Figure 2b]. There were no separate and distinct muscle components of the anterior abdominal wall. Virtual lines were marked over the anterior abdominal wall laterally on both sides of the midline about 5 cm from the midline for creation of rectus muscle. The peritoneum was opened via markings of the lateral border of the rectus muscle on both sides [Figure 2c]. Bilateral single-stage Fowler Stephen’s orchidopexy was performed, and the testis was brought down into the scrotum [Figure 3a]. Double breasting of the musculofascial layer was done on both sides by using continuous running stitch of absorbable sutures to join the rectus to the inner surface of the lateral musculofascial layer and then opposing the margins of the lateral musculofascial layer of both sides in the midline and suturing it to each other and the umbilicus [Figure 3b and c]. Trimming of excessive skin was done on the lower and lateral aspects, accommodating the umbilical skin. For accommodation of the umbilical skin, a small cut was made in the midline in the reflected upper abdominal wall skin.

The upper reflected abdominal wall skin was opposed to the skin of the lateral abdominal wall, groin and suprapubic region [Figure 3d]. In the postoperative period, there was no complication except some hypertrophic scar, which will be revised subsequently. On a follow-up of 14 months, the patient is doing well without any urinary complaints [Figure 4]. Renal function and viability of the testis were assessed in the follow-up visits and were found to be normal.

DISCUSSION

Prune belly syndrome is an extremely rare congenital disease affecting multiple body organs, and adversely affects patient’s quality of life. Abdominal muscle weakness is the most

consistent feature. Muscle weakness is not uniform in these patients; the weakness is more in the infraumbilical part than that in the supraumbilical and more in central than in the periphery of the abdomen and is also found on using electromyography.^[3] The aim of abdominoplasty is to increase abdominal wall strength, to normalise abdominal shape and to normalise the pulmonary functions which are severely compromised by weakness of abdominal muscles.^[4] Randolph *et al.* used elliptical excision of infraumbilical redundant skin and fascia but failed to correct lateral bulging adequately.^[3] Lesavoy *et al.* described a new technique with double breasting of muscles and fascia with preservation of umbilicus.^[5] In contrast to the above studies, we have used an inverted T-shaped incision preserving the umbilicus and performed excision of excessive infraumbilical skin, with medial approximation of musculo-aponeurotic fascia by double breasting. Our technique completely preserves the musculo-aponeurotic fascia with the umbilicus along with their vascularity and hence provides excellent reinforcement to the mid-abdominal wall. This technique also offers better functional and cosmetic results as the scar is infraumbilical, with the horizontal axis easily getting hidden under the clothing.

CONCLUSION

Although several techniques have been described for managing abdominal wall weakness, our technique provides better vascularisation and umbilical preservation and good cosmetic results. Orchidopexy and urinary tract surgeries can be performed concomitantly.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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