

Prune Belly Syndrome: Errors in Management and Complications of Treatment

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Abstract: Prune Belly Syndrome is a condition in which the abdominal wall, urinary tract and testes are affected resulting in a severely curtailed life span. With proper management these patients could have a normal, productive life.

Key words: Abdominal muscular dystrophy, Prune Belly Syndrome, Urinary tract anomalies

Abbreviations: CIC-Clean intermittent bladder catheterization, IVP-Intravenous pyelogram, PBS-Prune Belly Syndrome, MAG3-Tc99m-mercaptoglycine, PUV-Posterior urethral valves, UTI-Urinary tract infection, VCUG-Voiding cystourethrogram, VUR-Vesicoureteric reflux

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Introduction

Prune Belly Syndrome (PBS) is a congenital anomaly that, by definition, occurs only in males. These boys have abdominal muscular dystrophy (prune belly), urinary tract anomalies and intraabdominal testes. Although the basic combination of anomalies is well defined, their severity varies considerably from patient to patient. The abdominal wall defect may range anywhere from a mild "pot belly" to an apparent total lack of abdominal wall musculature with intervening presentations that may involve either the entire abdominal wall or parts of it. The urinary tract may be obstructed, there may be vesicoureteric reflux (VUR) or only urinary stasis. Similarly, the severity of involvement of the kidneys, ureters, bladder and urethra demonstrate a very wide spectrum. In addition, since these patients have a "bell-shaped" chest with a flattened diaphragm they have an ineffective cough thus making them susceptible to respiratory infections. Their intestines are suspended in the abdomen from a universal mesentery, which may make them susceptible to volvulus. Anorectal anomalies, spinal defects, compression defects of the lower extremities are other known associations. Finally,

there are patients with an incomplete form of the syndrome, also known as "Pseudoprunes". This group consists of:

1. Females with typical abdominal wall and urinary tract abnormalities.
2. Males or females with abdominal muscular dystrophy without urinary tract involvement.
3. Males or females with typical urinary tract findings and a normal abdominal wall.

It is thus apparent from all these variations that there is considerable scope for misdiagnosis or incorrect intervention.

Our experience in managing 37 children with PBS has provided us with some insight into their management.

Historical Errors

In the 1950s many authors, incorrectly equated urinary tract dilation with obstruction and, advocated early, intubated urinary diversion.^[1,2] Use of tubes for urinary diversion invariably leads to the expected problems of dislodgment or obstruction of the tube and urinary infections due to this indwelling foreign body. The belief that PBS urinary tracts were obstructed, resulted in the next step in management, in the 1970s, when early complete reconstruction of the urinary tracts of all PBS patients was advocated.^[3,4] At the same time others were coming to the exact opposite conclusion that minimal or no interference was the way to go.^[5,6]

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Welch and Kearney^[7] made the first attempt at classifying these patients into therapeutic groups. A better classification was subsequently proposed by Berdon *et al.*^[8] In our hands, the latter classification, with a minor modification of our own, provides the best guideline for managing these patients.

Classification of Berdon *et al.*^[8]

They placed patients into three groups:

Group I: Potter's syndrome (oligohydramnios)

Group II: Severe neonatal and infantile urinary tract involvement

Group III: Mild involvement

Group I patients have severe pulmonary hypoplasia and renal dysplasia at birth. Their limbs often demonstrate the effects of intrauterine compression. They die at birth, usually of respiratory failure. Group II patients also have severe respiratory and urinary tract problems, however, both systems are not so severely affected that the child succumbs at birth. Group III patients typically present because of the abnormal appearance of the abdominal wall or for undescended testes. Imaging demonstrates a typically abnormal urinary tract.

Berdon's classification^[8] suggests that all patients in Group II require surgical correction. That is not our experience. We further subdivide Group II into three subgroups depending upon whether they have urinary obstruction, VUR or stasis. Patients with urinary obstruction invariably require urinary tract reconstruction, those with VUR require aggressive medical management and often surgical reconstruction is eventually required, whereas those with stasis either do not require surgical intervention or, at most, their urinary drainage is improved by bladder domectomy (to improve detrusor function) and abdominoplasty (to enable a more effective Valsalva maneuver). In our view the difference between Group II stasis and Group III patients lies in the fact that the latter never presents with urinary tract infections or deteriorating renal functions.

Errors in Antenatal Diagnosis and Therapy

Since antenatal ultrasonography became universally available in the 1980s PBS has often been diagnosed and also misdiagnosed as posterior urethral valves (PUV). Initially unnecessary antenatal interventions occurred in many PBS babies since ultrasonographers were unable to distinguish them from PUV. Hydronephrosis, megaureters and a thick-walled bladder are seen in both conditions, however, there are differences. In PBS the abdominal wall tends to be lax and the urinary bladder is massive, whereas in posterior urethral valves the abdominal wall may be distended but it is not lax and the thick walled bladder is small. Finally, the posterior urethra in PBS is dilated, elongated and "heart-shaped" while

in PUV the bladder and urethra have a "key-hole" or "pear-shaped" appearance. Subsequently, even though PBS was diagnosed in utero vesicoamniotic shunts were placed because of the erroneous impression that the urinary tract was obstructed. Shunting did not improve their status.^[9]

Errors in Initial Evaluation and Management

When a baby is born with PBS, the initial focus should be in determining his respiratory capacity and administering respiratory support as required. A Group I patient will probably succumb despite treatment whereas a Group III patient will require minimal, if any, supportive care. Group II patients will respond to intensive respiratory care. The appearance of the abdominal wall does not correlate at all with the severity of urinary tract abnormalities (Fig. 1 and Fig. 2). Potter's facies,



Fig. 1. Evaluation of this child with severe lack of abdominal musculature revealed only urinary stasis. All he required was a bladder domectomy along with orchidopexy and abdominoplasty. Note the bell-shaped chest



Fig. 2. This patient with very mild abdominal wall involvement had severe bilateral ureteric obstruction and borderline renal functions. Early cutaneous pyelostomies were carried out followed by urinary tract reconstruction. By 9 years of age he required a renal transplant. It is to be noted that he has a bell-shaped chest and a megalourethra. There is a known association between PBS and megalourethra

a bell-shaped chest and compression deformities of the lower extremities all indicate maternal oligohydramnios due to poor renal function or urinary outflow obstruction. Urinary ascites and/or a patent urachus occur due to lower urinary tract obstruction. These obstructed patients require urinary drainage in addition to respiratory support. Urinary ascites is aspirated to permit improvement in respiratory functions. Fortunately, leak from the collecting systems which causes urinary ascites helps maintain low intra-renal pressures, thus protecting the kidneys from further damage. These children improve with adequate drainage of the urinary tract. If patent, the urachus is a convenient site to insert a large catheter for drainage. In this initial stage evaluation of the urinary tract should be confined to blood tests and ultrasonography. In all other patients, unless it is necessary to measure urine output, it is best to avoid catheterizing the neonate for fear of precipitating a urinary tract infection (UTI). Radiographic and nuclear studies should be delayed until the child is stable and they must be evaluated with care since they always demonstrate a dilated urinary tract that retains contrast for long periods of time. Similarly, renal function studies also do not give the true picture. Furthermore, during follow-up, changes in contrast studies and blood tests may lag behind true functional deterioration. In our experience the child's clinical status with regard to appetite, general well-being, somatic growth and development and control of UTIs are much more sensitive indicators of deteriorating renal function than radiographic or chemical tests.

At this early stage we feel that all PBS patients should be placed on urinary suppressive antibiotics as we do not know what their eventual status would be. Initially we like to use an antibiotic from the penicillin group. After the baby is 6 weeks old a sulfa drug or nitrofurantoin may be used. Nitrofurantoin is an excellent drug, however, children often do not like its taste.

Errors in Management After the Neonatal Period

Severity of involvement in these children becomes apparent after the first few weeks. By then Group I patients have succumbed to respiratory failure. All other patients tend to stabilize. The urinary tract should now be evaluated with the child on antibiotics to prevent urinary infection after instrumentation. We prefer a Tc99m-mercaptotriglycine (MAG3) renal scan since in a single test we obtain information regarding the amount of functional renal tissue and outflow obstruction. The test must always be done with a draining bladder catheter in place to minimize confusion caused by urinary stasis and/or VUR. An intravenous pyelogram (IVP), also with a draining bladder catheter, would also give the same information regarding renal concentrating ability and drainage but one is better able to quantify the results of a renal scan (Fig. 3). It is essential to remember that the renal

scan and IVP are guidelines for management and no decision should be made based solely upon the tests. Invariably, a borderline obstructive pattern will be apparent in all PBS patients even after administration of diuretics since the ureters and bladder have such a great capacity. A radiographic voiding cystourethrogram (VCUG) is valuable in determining the presence of vesicoureteral reflux (VUR) (Fig. 4), the shape and emptying capacity of the bladder, the appearance of the urethra and the presence of urethral obstruction. Approximately 70% of PBS patients have VUR and they should stay on suppressive antibiotics. The bladder often has an hourglass shape with a urachal cap, which traps urine when the detrusor contracts (Fig. 5). The posterior urethra is "heart-shaped" and there is an area at its junction with the bulbous urethra where it narrows suddenly. This narrowing is usually insignificant, however, it could form a urethral ring that requires division or one may find Type IV valves.^[10]

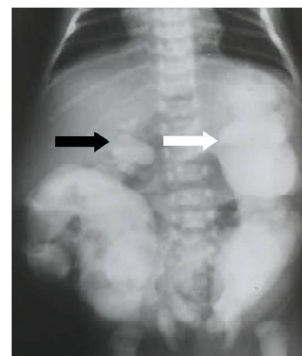


Fig. 3. The massively dilated ureters visible on this intravenous pyelogram were indeed obstructed at the lower end. However, such an appearance is also possible when only stasis is present. In this case the left kidney is extremely hydronephrotic and dysmorphic (white arrow) while the right kidney (black arrow) is only minimally involved. The ureters often absorb back-pressure in an obstructed system, thus preserving renal function. This patient required complete urinary reconstruction

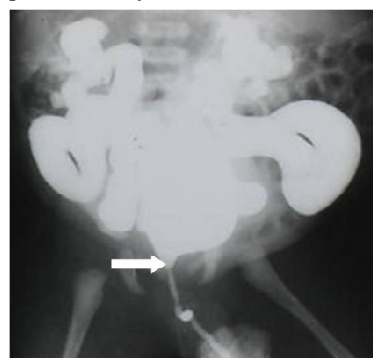


Fig. 4. Massive bilateral VUR is demonstrated in this VCUG. This child had complete urinary reconstruction at 6 weeks of age. White arrow points to the narrow junction between the posterior and the bulbous urethra

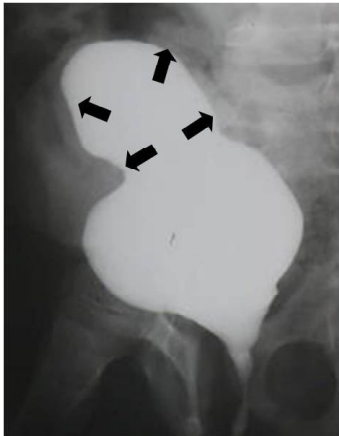


Fig. 5. VCUG demonstrating an hourglass shaped bladder due to a large urachal diverticulum (black arrows)

Once the urinary tract has been evaluated, Group II VUR should be left on suppressive antibiotics. Their follow-up consists of monthly urinalyses and urine cultures to maintain sterile urine. In addition, renal ultrasonography every 3 months or so is valuable in evaluating dilation of the urinary tract, as it demonstrates efficacy of drainage. Patients with stasis alone are typically placed on suppressive antibiotics initially. Antibiotics are discontinued if random monthly urine cultures on three occasions are sterile. They are then followed clinically and if they continue to thrive, they will be placed in Group III. If urinary infection recurs, the child is considered to be Group II stasis and consideration is given to determining whether bladder drainage is adequate. On occasion, clean intermittent bladder catheterization (CIC) is helpful in this group. Group II patients with urinary obstruction have the worst prognosis. They go into renal failure early and they are also particularly susceptible to UTIs. In our experience, keeping a close watch on their general well-being is the best way to catch changes early. If the obstruction is worsening, the child will not thrive and the ultrasound will demonstrate further dilation of the urinary tract. Renal scans and blood chemistries are carried out but they are slow in identifying deterioration. Group II obstructed patients invariably require surgical reconstruction.

Recurrent UTIs are common in PBS patients. Infections can be difficult to clear since the tortuous ureters form pockets of pus. We failed to diagnose a urinary infection in one of our Group II patients. He died of overwhelming sepsis, from resistant E.Coli, 48 hours after total urinary reconstruction. Another Group II patient presented semicomatose with high serum ammonia levels suggestive of Reyes syndrome. On further evaluation it became apparent that the culprit was an ammonia-producing organism in the urinary tract. Finally, one has to maintain a careful balance because clearing pathogens from a urinary tract in which stasis is always present often results in overgrowth of yeast. On occasion the patient may

go back and forth between growth of urinary pathogens and yeast.

Errors in Decisions Pertaining to Urinary Diversion

Intubated nephrostomy or pyelostomy: We have already pointed out that the earlier view that every PBS patient is obstructed and requires early urinary diversion is clearly faulty. In addition a nephrostomy or pyelostomy tube tends to get dislodged, kinked, plugged with debris or it becomes the source of infection. Intubated diversions are only indicated as an emergency and for the short-term to resolve an acute problem. They are best performed percutaneously under radiological guidance. The smallest pigtail catheter that would adequately drain the kidney should be placed in a lower pole calyx. If possible, the catheter should be inserted through a part of renal cortex that has the least parenchyma.

Ureterostomies: Ureterostomies are mentioned to be condemned. First, stasis is not avoided since urine still has to pass through these dilated aperistaltic proximal ureters. Secondly, urine has to drain against gravity when ureterostomies are brought out on the abdominal wall (Fig. 6) particularly in infants who spend most of the time on their backs. Thirdly, ureterostomy stomal stenosis is very common. Finally, the longitudinal blood supply of the ureter is placed in jeopardy and the distal ureter may be devascularized when a ureterostomy is taken down.



Fig. 6. Bilateral loop cutaneous ureterostomies did not improve drainage in the dilated aperistaltic ureters in this boy because the location of the ureterostomies required anti-gravity drainage of urine. He responded well to total urinary reconstruction

Internal Stents: Internal stents are only of value for obstruction at the ureteropelvic junction or upper ureter and that is rarely, if ever, the case in PBS. Furthermore, stents create free reflux from the bladder, thus compounding stasis and since they are foreign bodies they increase the chance of UTI.

Vesicostomy: A vesicostomy also serves no purpose since urine still has to traverse the full length of the atonic ureters. It does permit continuous drainage from the bladder, but high intravesical pressure is not an issue in these children. In addition, these large, flabby bladders tend to prolapse through the stoma even if a Blocksom vesicostomy^[11] is carried out. A suprapubic cystostomy is even worse since the tube is prone to dislodgment and obstruction in addition to being a source of infection.

Perineal urethrostomy: A perineal urethrostomy is never indicated, as it does not bypass any part of the urinary tract.

Indications & Techniques of Urinary Diversion

The only indication for emergent urinary diversion in PBS is the child who develops a severe urinary infection that is not responding to treatment. In such a situation a percutaneous nephrostomy placed under radiographic guidance is the least invasive technique and is easily reversed upon removal of the catheter. While the catheter is in place it carries the already mentioned risks of infection, dislodgment, and obstruction by debris.

Diversion for an extended period is indicated if the kidney has very limited function and one is trying to delay its eventual failure. One or both kidneys can be diverted. A high diversion under such conditions also permits accurate evaluation of individual renal functions.

We prefer cutaneous pyelostomy (pyelocutaneous anastomosis) for diversion. Exteriorization of the renal pelvis provides the shortest and most direct route for egress of urine, and it does not affect the blood supply of the distal ureter. A cutaneous pyelostomy is not without complications. The dysplastic kidney with thin parenchyma can turn inside out and prolapse through the stoma. The stoma could also be the route for an ascending infection and it is difficult to adequately place a device to catch the urine so close to the XII rib and spine. Parents have to place a second diaper around the waist to catch the urine. If bilateral diversions are carried out in the face of deteriorating renal functions the patient is undiverted prior to renal transplantation.

The cutaneous pyelostomy is carried out with the patient prone or in the lateral position. A 1cm transverse incision is made at the angle of the XII rib with the sacrospinalis muscle. The incision is deepened through all muscle layers. The highly mobile kidney can be manually pushed toward the incision and the posterior aspect of the renal pelvis is grasped and delivered out of the wound. A 1cm transverse incision is made on the posterior surface of the renal pelvis and the cut edges of the pelvis are sutured with interrupted absorbable sutures to the full thickness of the skin. It is worth repeating that the incisions in the abdominal wall and the renal pelvis

must both be no more than 1cm in length to avoid renal prolapse.

Total Urinary Tract Reconstruction

Indications: Past errors in management have already been discussed. In our experience, all Group II patients with obstruction and a majority of those with VUR require complete urinary tract reconstruction. Patients with an obstructed urinary tract typically become candidates for reconstruction within the first year of life. Occasionally, if their renal function is borderline, a preliminary cutaneous pyelostomy in preparation for reconstruction may be of value. Patients with VUR should initially be managed with suppressive antibiotics, awaiting spontaneous resolution of VUR by two to three years of age. However, if recurrent or persistent UTIs occur or VUR does not resolve, reconstruction is indicated.

Surgical Technique: If the urinary tract is reconstructed, simultaneous abdominoplasty and orchidopexy are carried out but they will be discussed separately. Fig. 7 illustrates the typical appearance of the urinary tract in PBS. The ureters are most tortuous and atonic in their lower halves hence, they are disconnected from the bladder. Any extra length is excised at the lower end and, if necessary, the upper ureters are straightened out. The ureters are reimplanted in the bladder after tapering. During ureteral mobilization it is essential that the peritoneum and periureteric adventitia on its medial aspect be pushed towards the ureter to maintain the blood supply. If a ureterostomy is being taken down, one must take extra pains to avoid damaging the longitudinal blood supply on the medial aspect of the ureter. The ureters are tapered by excising tissue on the lateral aspect and

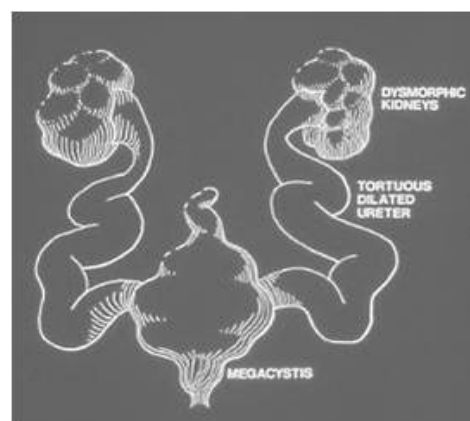


Fig. 7. Typical appearance of the urinary tract in PBS. The kidneys are dysmorphic and hydronephrotic. Renal dysplasia of varying degrees is evident on histologic examination. The ureters are large, elongated and tortuous, particularly at their lower ends. The bladder is large and thick walled with a wide bladder neck, and a urachal diverticulum. The prostatic urethra is "heart-shaped"

retubularization over a 10Fr. catheter. They are reimplanted in a 3-4cm cross-trigone submucosal tunnel. In these large low-pressure bladders reimplantation is relatively simple and results are excellent.

The bladder often has a urachal cap, which collects urine when the detrusor contracts. This cap is best removed by making a "fish-mouth" incision that runs in the anteroposterior direction. Once the cap is excised a spherical bladder with good detrusor function is obtained. Bladder domectomy is carried out at the beginning of the reconstruction and ureteral mobilization and reimplantation are carried out through this opening. It is important to remember that these children often produce large volumes of urine and it is crucial that an adequate bladder capacity be maintained (Fig. 8 and Fig. 9).

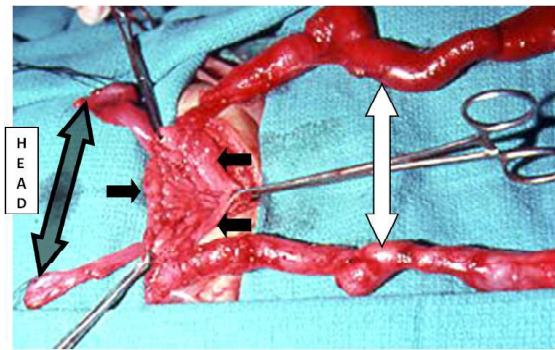


Fig. 8. Total urinary reconstruction. The patient's head is to the left. The megaureters have been dissected out and straightened (white arrow). The testes have been mobilized on the vessels of the vas deferens (shaded gray arrow). The dome of the bladder has been removed (small black arrows). The ureters will be shortened, tapered and reimplanted into the bladder which will be reconstructed so as to have a spherical shape. The testes will be brought down into the scrotum

In those rare instances when urethral outflow obstruction coexists, it is dealt with at the time of reconstruction. Valves should not be resected in a diverted or "dry" urinary tract lest a urethral stricture develops.

The ureters and bladder are stented for a two-week period after which the stents are removed if contrast studies demonstrate no leak from suture lines.

Orchidopexy

Invariably the testes are located intraabdominally, at the pelvic brim (Fig. 10). In Group II patients, orchidopexy is best carried out in conjunction with urinary reconstruction. In most instances the spermatic vessels are too short for the testes to reach the bottom of the scrotum. The spermatic vessels are divided close to the kidneys and mobilized with the blood



Fig. 9a

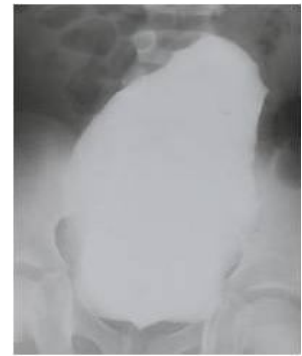


Fig. 9b

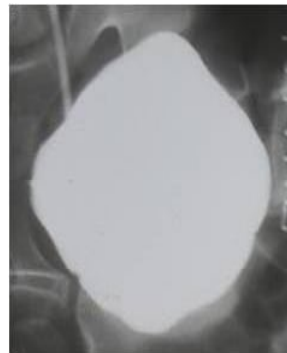


Fig. 9c



Fig. 9d

Fig. 9a. Intravenous pyelogram demonstrating a large bladder

Fig. 9b. Huge postvoid residue in the large bladder

Fig. 9c. Cystogram after bladder domectomy

Fig. 9d. Minimal postvoid residue after domectomy

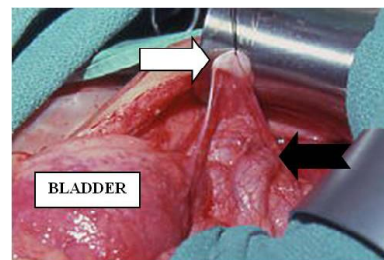


Fig. 10a

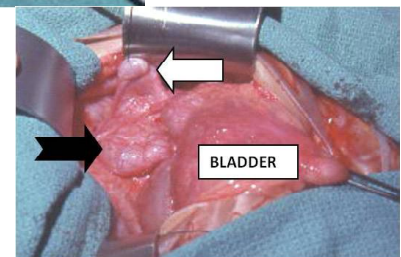


Fig. 10b

Fig. 10. Location of the intraabdominal testes at the pelvic brim. In both figures the bladder has been displaced downwards. The solid white arrows point to the testes. The notched black arrows demonstrate the megaureters

Fig. 10a. Right testis

Fig. 10b. Left testis

supply to the ureter. The testes are then brought down into the scrotum on the collateral blood supply from the vas deferens.^[12] On occasion, in the infant, it may be possible to bring the testes down into the scrotum with the spermatic vessels intact.^[4] Since there is no inguinal canal, an opening is made in the anterior abdominal wall over the pubic tubercle to develop the shortest possible route to the scrotum. The large bladder tends to get in the way and the opening has to be just lateral to it. Group III patients may require orchidopexy alone or in association with abdominoplasty. If abdominoplasty is planned, the orchidopexy is carried out through the same incision. If only orchidopexy is planned, the PBS bladder, which occupies the area of the lower abdomen behind the recti upto the umbilicus, has to be avoided (Fig. 11). We prefer bilateral transverse 3cm incisions extending anteriorly from the anterior superior iliac spines. Such incisions avoid the bladder, and place us directly above the testes. With this approach also, the spermatic vessels are divided and the testes are brought into the scrotum on the vessels from the vas, by the shortest route possible.



Fig. 11. category III patient with a large pot belly. The yellow structure outlined corresponds to the large bladder. The incisions extend anteriorly for 3 cm from the anterior superior iliac spine on either side

Abdominoplasty

Abdominoplasty is of great value to the majority of these patients. If the abdominal wall is tightened, respiratory functions, particularly coughing, and the ability to Valsalva to effectively evacuate feces and urine are improved. In addition we believe patients are better able to sit up from a supine position after abdominoplasty. Only a few patients in

Group III, with a pot-belly, do not require abdominoplasty. Various operations have been described for abdominoplasty.^[13-16] It is true that the infraumbilical rectus abdominis muscles demonstrate the greatest laxity, however, the deficiency may be worse on one or the other side, and it may extend to the supraumbilical recti and even the whole abdominal wall. If the defect is symmetrically located in the infraumbilical recti one obtains an excellent result from an abdominoplasty carried out through an infraumbilical "smile" incision (Fig. 12). On the other hand, the patient in figure 13 required a "reverse D" incision as his defect involved the supra- and infraumbilical portions of the right side of the abdomen while the left side was almost normal (Fig. 13). Another patient with an eccentric infraabdominal defect in which the "smile" incision was widened on the more affected side (Fig. 14). We believe that no single incision is adequate for all cases and it must be tailored to the defect. In our



Fig. 12a



Fig. 12b



Fig. 12c

Fig. 12a. The patient's defect was essentially infraumbilical and symmetrical on both sides

Fig. 12b. The incision has been marked

Fig. 12c. The appearance of the abdominal wall one week after surgery

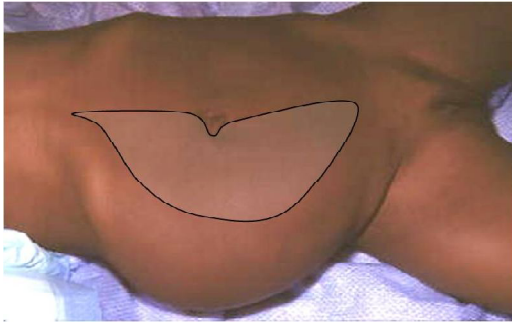


Fig. 13. This patient's major problem was in the upper and lower abdominal muscles on the right side. The reverse D shaped incision marked on the abdomen corrected the defect



Fig. 14. This patient had an eccentric infraumbilical defect involving more of the right side hence the "smile" incision was widened in the right infraumbilical area as marked in the photograph

experience careful physical examination in which the patient is made to contract his abdominal muscles is all that is required to delineate the defect. Electromyography is unnecessary, expensive and it scares the child.^[17]

Perioperative Management

It is essential that the entire team taking care of these children understand that that great care is required in their management. The patient's respiratory functions should be at their best prior to the operation and even the slightest respiratory infection should be grounds for postponement. The urine must also be sterile.

The immediate postoperative period is a critical time in which pulmonary infection and respiratory failure result in deaths. Vigorous pulmonary toilet and ventilator support are crucial at this time. Adequate postoperative pain control is essential for the patient to move, take deep breaths and cough. An epidural catheter maintained for 3-4 days is invaluable.

Conclusion

Prune belly syndrome is a complex previously lethal congenital anomaly. Numerous errors were made in the management of these patients in the past due to improper and inadequate understanding of its pathology. With careful and committed care these patients can go on to function very well in society.

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