

# CASE REPORT

## Prune belly syndrome: A rare congenital malformation in a 6-year-old Ugandan boy

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### Abstract

A 6 year male presented with history of recurrent urinary tract infections since birth. At the time of presentation he had an active urinary tract infection associated with deranged renal function tests, a soft wrinkled abdomen and bilateral undescended testis. Further evaluation with an abdominal ultrasound scan confirmed a diagnosis of prune belly syndrome. He was managed for the urine infection, had a urethral catheter for ureter decompression inserted and put on maintenance antibiotics. There are few reports about this condition in Uganda and hence this case can advance knowledge on this rare syndrome.

**Keywords:** prune belly syndrome, cryptorchidism, abdominal wall muscle deficiency

### Introduction

Prune belly syndrome (PBS) is a figurative term coined by Osler for the wrinkled appearance of the abdomen, like a prune, resulting from agenesis of abdominal musculature<sup>1, 2</sup>. Parker (1895) noted the triad of congenital anomalies associated with PBS (cryptorchidism, deficient abdominal wall musculature and urinary tract anomalies). Originally, it was described by Frohlich in 1839, who described that this triad doesn't mean the total spectrum of the syndrome, it may also involve cardiopulmonary, gastrointestinal, musculoskeletal and CNS<sup>4, 5</sup>. It is estimated to affect 1 in 30,000-40000 live births worldwide with more than 90% of the cases occurring in boys.<sup>1</sup> There are no obvious racial predilections, but common with twin pregnancies, trisomy 18 and 21.<sup>1</sup>

Children with prune belly syndrome can present with a myriad of renal, ureteral and urethral abnormalities with complications ranging from urinary tract infections to chronic kidney disease<sup>1</sup>. A lack of abdominal muscles leads to a poor cough mechanism which in turn leads to increased pulmonary secretions. The weak abdominal wall muscles also lead to constipation due to inability to perform Valsalva manoeuvre which helps push the stool out of the rectum during defaecation<sup>2</sup>. The prognosis may vary from stillbirth to a normal life expectancy depending on the degree of pulmonary and renal compromise; mortality is 20% within first month and 50% within 2 years<sup>14</sup>. Major cause of morbidity in PBS is recurrent urinary and pulmonary complications that require meticulous preoperative monitoring, antibiotic

coverage, active pulmonary physiotherapy and avoidance of any respiratory depressant drugs<sup>4, 6, 14</sup>

We hereby describe a case of prune belly syndrome in a 6 year old male who presented with a wrinkled abdomen and cryptorchidism who has been getting recurrent urinary tract infections.

### Case presentation

He is a 6 year old male, who lives in Omotorok village in Kaberamaido district in north eastern Uganda about 400kms from Kampala.

He presented with absence of testicles since birth and abdominal pain for 4 days. The abdominal pain was, mild, constant, more concentrated in the flanks and suprapubic regions. It was associated with low grade fever, burning micturition and dribbling of urine but there is no frequent micturition, hesitancy or hematuria. The father also reported that the child's abdomen was always distended with wrinkles and abnormally soft, he however reports normal bowel habits without constipation or diarrhoea. The father reports that the child has been treated for urinary tract infections several times in the last 2 years of life. He also reports occasional palpitations but with no associated cough, body swelling, and difficulty in breathing on lying flat. He had no other complaints.

No significant surgical history prior to this presentation

On the birth history, the mother attended antenatal care three times during pregnancy at a nearby Health centre she



**Figure 1.** Six-year-old boy with prune belly syndrome

had no issues during pregnancy and no **ultrasound scan was done**. He delivered at term from home without any complications and the child was in good condition and breastfed immediately. He took him to hospital for immunization after one week. He was fully immunized and has been growing well and comparable to his age mates and is expected to begin school the following year. He is the third born in a family of 5 children, other children are fine. No known chronic familial diseases or history of this condition in the family. The mother and father neither smoke nor take alcohol.

Clinically, he was a young child in fair general condition, febrile ( $T = 37.7^{\circ}\text{C}$ ), not pale, no jaundice, no oedema, no lymphadenopathy, no digital nail clubbing, no cyanosis and not dehydrated.

The abdomen was moderately distended with wrinkled skin, asymmetrical with bulges in flanks but moving with respiration. On palpation, the abdomen was soft with absent abdominal muscles and easily palpable intestines. The kidneys could be palpable bilaterally with renal angle tenderness. No hepato-splenomegaly. On pelvic examination, the scrotum was well distended but without testis bilaterally.

The chest wall was symmetrical and moving with respiration. He was not in distress,  $\text{RR} = 20$  breaths per minute. Resonant percussion and bilateral breath sounds with no added sounds.

Pulse rate was 100 bpm, regular, full volume. Blood pressure was 90/70 mmHg, no distended neck vessels. Heart sounds I and II were present with no added sounds. Other systems were normal.

Abdominal ultrasound scan revealed the urinally bladder which is fully distended with echogenic contents. The ureters were tortuous and enlarged with diameters of 1.33cm and 2.03cm for the right and left ureters respectively. The kidneys were also enlarged with dimensions of  $10.2 \times 4.7\text{cm}$ . The abdominal wall was thin and flabby and there were no testicles bilaterally in the scrotum. In conclusion the scan found bilateral hydronephrosis, urinary bladder outlet obstruction and possible vesico-ureteric reflux, plus a gallstone.

Urinalysis revealed 50-100 pus cells in urine and proteinuria of +1.

Complete blood count revealed significantly leucocytosis which was neutrophil dominant other parameters were in normal range.

$\text{HB} = 13.5\text{g/dl}$

$\text{WBC} = 16 \times 10^3$

$\#\text{NEU} = 9.2 \times 10^3$

#### Renal function tests

Urea = 76 (21-54mg/dl)

Creatinine = 2.2 (0.8-1.1mg/dl)

No other investigations could be done at this hospital level.

Urethral Catheterization was done for decompression of the ureters and reduction of backflow of urine to the kidneys

Treatment of urinary tract infection with antibiotics and putting the child on chronic antibiotics for prophylaxis

Counselling the parent about the child's condition and how to avoid abdominal trauma and likely complications a child might get.

## Discussion

### Prune belly syndrome

In this case, the boy had the classic triad of: urologic anomalies like mega ureters, hydro-ureters, hydronephrosis, vesicoureteral reflux and mega cystitis, bilateral cryptorchidism and absence of anterior abdominal wall muscles as described in several other studies<sup>1</sup>. Since the etiology and pathogenesis of this condition is not fully understood there were no identified risk factors in this case. This condition has no obvious racial predilections, but it is commonly associated with twin pregnancies, trisomy 18 and 21<sup>1,6,9</sup>. No features of these trisomies were identified in this child.

Clinically most patients are identified at birth with typical features of wrinkled abdomen and undescended testis<sup>1</sup>.

A lack of abdominal muscles leads to a poor cough mechanism which in turn leads to increased pulmonary secretions and chest infections. The weak abdominal wall muscles also lead to constipation due to inability to perform Valsalva manoeuvre which helps push the stool out of the rectum during defaecation<sup>1,2</sup>. Urologic anomalies are the commonest and patient presentation ranges from recurrent urinary tract infections to kidney failure mainly due to obstructive uropathy<sup>1</sup>. The urinary symptoms were the most obvious in this case and complications like renal failure due to excessive backflow of urine to the kidney had begun to set in.

Features that are usually present but not common include; Cardiac anomalies like patent ductus arteriosus, pulmonary abnormalities especially pulmonary hypoplasia which is the commonest cause of death immediately after birth, Orthopaedic anomalies like scoliosis and congenital hip dislocations and other gastrointestinal abnormalities which include malrotation, atresia, stenosis and volvulus<sup>3</sup>. None of these features were identified in this patient

The mother of this child never had a prenatal ultrasound scan but prenatal diagnosis can be done through ultrasonography where the cardinal signs of hydronephrosis, bilateral hydroureters, megacystis and oligohydramnios can be detected in utero<sup>9,10</sup>. Postnatally, ultrasonography of the abdomen was done and it usually identifies most abnormalities of the urinary system from the kidneys to urethra.

Other important investigations which were not done include cardiac echo to detect any associated congenital lung anomalies<sup>9,10</sup>, contrast voiding cystourethrogram which can be used to delineate the prostate-membranous urethra and bladder to detect presence of urachal remnants or vesico-ureteral reflux<sup>9</sup>, chest x-ray which can detect any underlying anomalies like pulmonary hypoplasia or heart anomalies and abdominal computerized tomography scan which can be used to assist in diagnosis of the abdominal anomalies<sup>10</sup>.

Laboratory studies done include urine analysis and renal function tests to assess the level of kidney function<sup>11</sup>.

Being in a resource limited setting, the child was managed with maintenance antibiotics and urethral catheter for the vesicoureteral reflux plus treating the underlying urine infection. Other non-surgical management may include chest physiotherapy and avoidance of respiratory depressants<sup>11</sup>.

Surgical management mainly involves orchidopexy<sup>17</sup> for undescended testis and then abdominal wall reconstruction for cosmesis and improvement of respiratory function and Valsalva manoeuvre. Placement of a percutaneous nephrostomy can be done in children with ureteropelvic obstruction.

## Prognosis

The prognosis may vary from stillbirth to a normal life expectancy depending on the degree of pulmonary and renal compromise; mortality is 20% within first month and 50% within 2 years. Major cause of morbidity in PBS is recurrent urinary and pulmonary complications.<sup>14</sup>

## Conclusions

We described a case of prune belly syndrome in a 6 year old boy who presented with recurrent urinary tract infections, wrinkled abdomen and undescended testis since birth with pending kidney failure who was managed in a rural and resource limited setting.

## Recommendations

Prenatal ultrasound scans are important for early diagnosis and every pregnant woman should have it done before delivery.<sup>9,10</sup>

Screening of other conditions like trisomy 18 and 21 should be done in such a case due to association with prune belly syndrome.<sup>1,2</sup>

Chest physiotherapy and avoidance of respiratory depressants are important in prevention of chest related complications of prune belly syndrome.<sup>11</sup>

Surgical procedures like orchidopexy and then abdominal wall reconstruction can be done to improve the quality of life of the patient.<sup>17</sup>

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