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Prune Belly Syndrome: A Ten-Year Single Tertiary Centre Experience in South-South, Nigeria

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Abstract

Background: Prune belly syndrome (PBS) is a congenital anomaly that consists of a triad of abdominal wall defect, bilateral cryptorchidism, and urinary tract dilation. The disease is of varying severity. This study aims to highlight the challenges and peculiarities in the management of PBS in a resource-poor setting. **Materials and Methods:** This is a ten-year retrospective study conducted at the University of Port Harcourt Teaching Hospital. Ethical approval for the study was sought and gotten from the hospital's ethical committee. The information gotten included history, duration of symptoms, examination findings, age of the patient, category of disease, and intraoperative findings. The data from the folders were collected and evaluated. Frequencies, percentages, the mean and standard deviation were used to summarize the data as appropriate. **Results:** Fifteen patients were included in the study. The hospital incidence of PBS was 112/100,000, twelve males and three females. The age range was from 1 day to 15 years, mean age was 14 months ± 2.3 months. Most patients presented between 3 months and 2 years and 11 months. Twelve patients had category three PBS and five patients had associated anomalies. Eleven male patients died after 5 years of follow-up from progressive renal deterioration. The female patient fared better than the males. **Conclusion:** PBS is rare, most patients with the condition present late. The most common cause of mortality was progressive renal deterioration.

Keywords

Prune Belly Syndrome, Renal Deterioration, Late Presentation

1. Introduction

Prune belly syndrome (PBS) is a congenital anomaly that consists of a triad of

abdominal wall defect, bilateral cryptorchidism, and urinary tract dilation [1]. The disease is of varying severity [2]. PBS is also called Eagle-Barrett syndrome, Orbinsky syndrome, abdominal musculation syndrome and triad syndrome. The disease was named PBS because of the wrinkled appearance of the anterior abdominal wall muscle due to the deficiency of muscle cells. The most severely affected die during the neonatal period while the least severely affected grow up with minor defects [3]. Males are more affected than females.

Prune belly syndrome is a rare congenital disease of unknown aetiology that is present in one in every 40 thousand live births worldwide [4]. Several theories have been postulated to explain the aetiology of PBS. These theories propounded include mesenchymal developmental defects and an obstruction to urine outflow in utero [1]. The aetiology of PBS is largely unknown. However, the high male-to-female ratio and occurrence of the disease amongst siblings, relatives and twins suggests a genetic basis to PBS. PBS has been associated with trisomy 13, trisomy 18, turner syndrome and monosomy 16 [5].

The urinary tract anomaly appears to be the most important determinant of survival in these patients [5]. Individuals with PBS presented with varied symptoms with the urogenital syndrome mostly affected. Patients presented with renal dysplasia, hydronephrosis, ureteropelvic junction obstruction, hydroureters, vesicoureteric reflux, patent urachus and distended bladder [6]. The prostate may be hypoplastic, and the vas deferens and seminal vesicles are either dilated or atretic [7]. The tests lie intraabdominal [7]. Extra genitourinary anomalies include abdominal wall anomalies, pulmonary [1], gastrointestinal [8], orthopaedic [1] and dental anomalies [9].

In Africa, there are not too many publications on the subject [10] [11] with most of the available publications being case reports. In Nigeria, few studies on PBS have been carried out [12]-[17]. We are currently unaware of any study on PBS conducted in Port Harcourt, Nigeria. Treatment of PBS is also a challenge [12] because of poor access to health care and cultural beliefs [10]. This study will evaluate the presentation, and management of patients with PBS in Port Harcourt, Nigeria.

2. Materials and Methods

This was a ten-year retrospective study. All patients who presented with features suggestive of PBS between January 2011 and December 2020 at the Paediatric surgery unit of the University of Port Harcourt Teaching Hospital UPTH were included in the study. Ethical approval for the study was sought and gotten from the hospital's ethical committee.

Data from all patients listed in the medical records department as having been treated for PBS during the study period were retrieved. Also, data were obtained from ward admission registers, theatre, and discharge records. The data included history, examination findings, patient age at presentation, intraoperative findings, and post-operative complications. Patients were also categorized into three

categories based on the severity of the symptoms. Category 1 had severe renal dysplasia, oligohydramnios, pulmonary hypoplasia and Potters facies. Category 2 patients presented with Full triad features, moderate or unilateral renal dysplasia, no pulmonary hypoplasia, and slightly abnormal renal function. Category three patients presented with incomplete or mild triad features, mild uropathy, no renal dysplasia, stable renal function and no pulmonary hypoplasia. patients with incomplete records were also excluded from the study.

Each patient had an intravenous urography, urinalysis/ microscopy culture and sensitivity, full blood count, electrolyte urea, and creatinine before surgery.

The data from the folders were collected and entered using Microsoft Excel 2016 version and transferred into the statistical package for social sciences (SPSS) for windows (version 25) (IBM SPSS Inc. Chicago, IL) for analysis. Ninety-five per cent confidence interval and a p-value less than 0.05 was considered significant. Frequencies, percentages, the mean and standard deviation was used to summarize the data as appropriate. Categorical data were presented in the form of frequencies and percentages using tables. Continuous variables were presented in means and standard deviation. Results were presented in tables and charts.

3. Results

Of the 13,343 patients seen during the study period, 15 with PBS were seen at the paediatric surgery clinic and Paediatric surgery ward, giving a hospital incidence 112/100,000 (Table 1).

4. Discussion

Since the first description of the disease between 1839 and 1895 PBS data have been composed primarily of case reports and small case series [13]. Prune Belly Syndrome is quite rare as shown in this study, and only 15 patients were diagnosed with this condition in 10 years of study with a hospital incidence of 112/100,000. Many other studies attest to the rarity of this syndrome [1] [14] [17]. The sex most affected in this study was male sex with 12 males and only 3 females as shown in Table 2. The literature on this subject is also in agreement with this finding [2] [14] [17] [18]. The mean age of presentation was 14 months ± 2.3 months.

Table 1. The age at presentation of patients to the hospital.

Age at presentation	Frequency	Percentage
Below 3 months	2	13.33
Three months to 2 years 11 months	11	73.34
Three years and above	2	13.33
Total	**15**	**100**

Table 2. The sex distribution of patients with PBS.

Sex distribution	Frequency (n)	Percentage (%)
Male	12	
Female	3	
Total	**15**	**100**

Sex distribution of patients with PBS, three females and 12 males. Age range = 1day to 15 years. Mean age = 14 months ± 2.3 months.

In utero, the amniotic fluid is composed mainly of urine, in situations of poor production of urine, there are oligohydramnios and poor development of the thoracic and musculoskeletal system. Also, the development of the kidneys in the presence of urinary tract obstruction can lead to dysplasia and renal compromise [5]. Severe oligohydramnios and renal dysplasia is associated with poor outcome. No patient in this study presented with respiratory difficulties, even the day-old neonate in **Figure 1** had no respiratory difficulties.

In Nigeria, there is poor antenatal care [19] [20], while some of these cases of PBS are diagnosed before birth in developed countries as early as 14 weeks gestational age with the use of an obstetric scan [1] [18]. The findings on ultrasound include abnormal dilatation of the bladder, reduced amniotic fluid, and bilateral hydronephrosis [18]. In Nigeria, the finding is different; late presentation and diagnosis are common. In this index study, only 2 presented before 3 months of age as shown in **Table 2**. These two patients were delivered at the University of Port Harcourt Teaching Hospital and the syndrome was promptly diagnosed by the paediatricians. In Africa and other developing economies, health care is largely unavailable [21] [22]. If these children were delivered by a traditional birth attendant or at home the syndrome may not have been diagnosed that early. Case reports of patients with PBS treated prenatally abound [18].

No patient presented with category 1 PBS in this study as shown in **Table 3**. The reason may be that even if patients with the severe form of PBS were delivered, they will most likely die within the first few hours because of poor health care in developing economies. Even in developed healthcare systems, early mortality for patients with severe PBS is not uncommon [18]. Most of the patients (12.80%) presented with category 3. Female patients tend to have a milder form of the disease as seen in **Figure 2** and **Figure 3** with these female patients having lax abdominal wall musculature on the left and right side respectively. We don't know if this one-sided laxity of the anterior abdominal wall leads to a better prognosis in these females. Men tend to have a more generalized laxity of the anterior abdominal wall as seen in **Figures 4-7**. The older patients tend to develop a pot belly as seen in **Figure 7**, which revealed a 15-year-old male with PBS.

PBS is associated with urogenital and extra-urogenital anomalies. The extra urogenital anomalies include cardiopulmonary, gastrointestinal and musculoskeletal and orthopaedic anomalies [1] [18] [23] [24] [25]. In this study, 2 patients had patent urachus as shown in **Figure 8** and **Figure 9**. A patent urachus tends to reduce the pressure on the developing kidneys, reducing urinary tract

obstruction's effect on the developing renal system. Each patient had an intravenous urography which revealed hydroureteronephrosis of various degrees as shown in Figure 10.

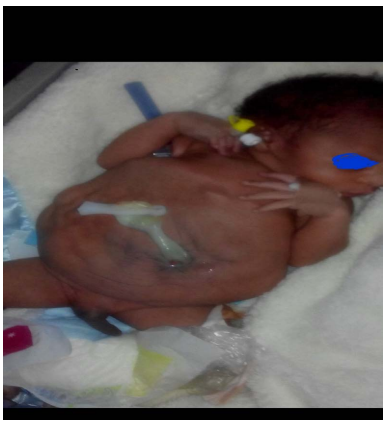

Figure 1. A day-old male with PBS, note the wrinkles on the whole anterior abdominal wall.

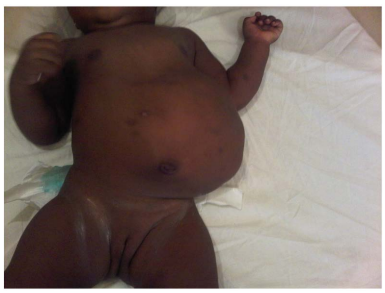

Figure 2. A 45 days-old female infant with PBS, note the defect more on the left Side of the abdomen.

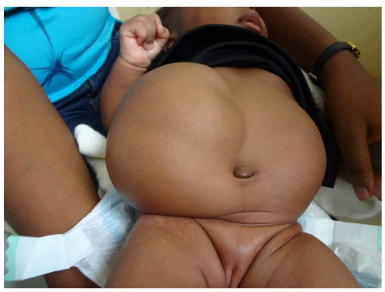

Figure 3. A 4 month old female infant with PBS and the abdominal wall defect is more on the right.

Table 3. The different categories of presentation. None presented with the most severe form of PBS.

Category	Frequency	Percentage %
1	0	0
2	3	20
3	12	80
Total	**15**	**100**

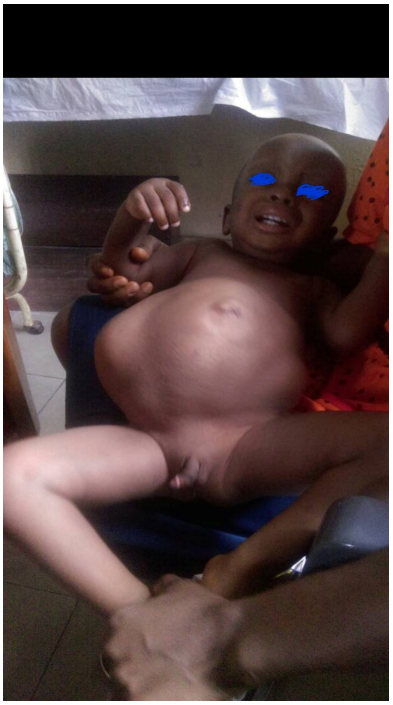

Figure 4. A two-year-old male with PBS, note the undescended testes and wrinkled abdominal wall.

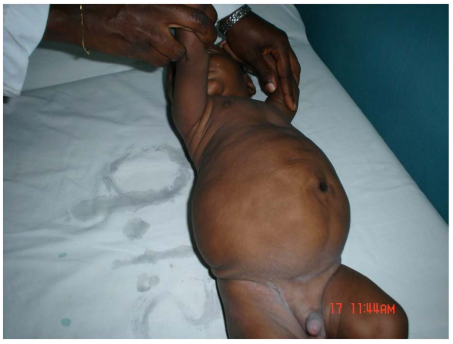

Figure 5. A 2-year-old male with PBS with generalized wrinkling on the abdominal wall.

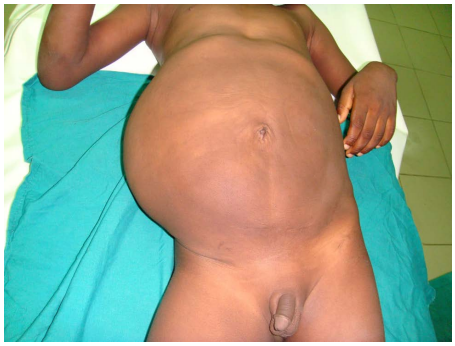

Figure 6. A 7-year-old male child with PBS, testes in the scrotum, note the scar above the inguinal region where he had orchidopexy.

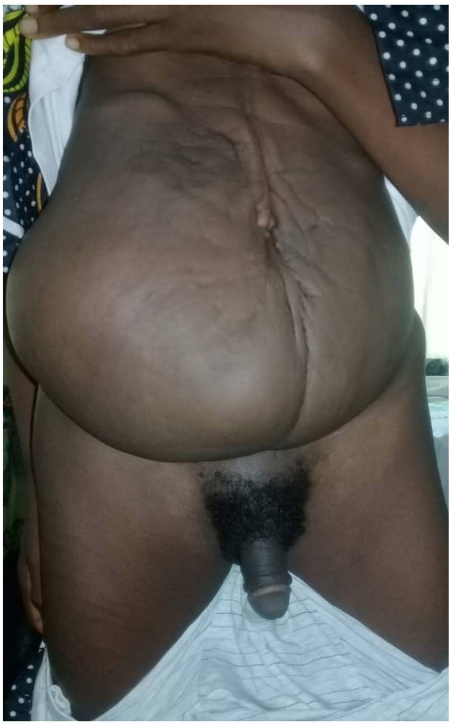

Figure 7. A 15-year-old male child with PBS who appears to have a pot belly. With ageing, the wrinkled abdominal wall may appear as a pot belly.

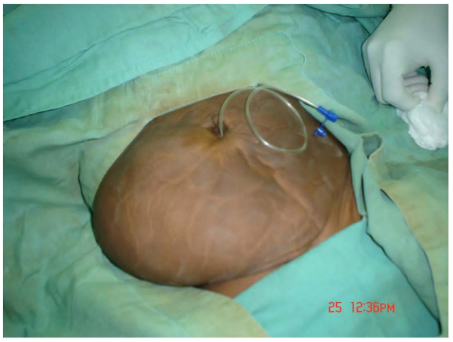

Figure 8. A patient with PBS with a patent urachus.

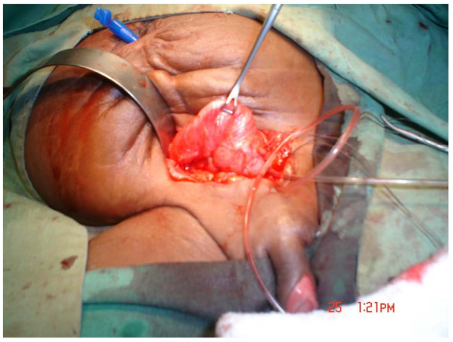

Figure 9. A feeding tube passed into the bladder from an opening in the umbilicus.

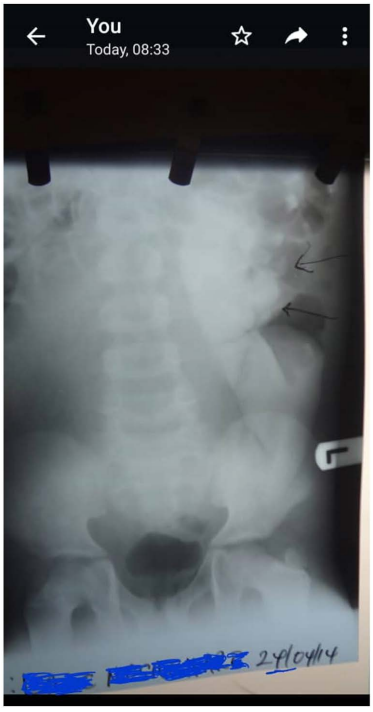

Figure 10. An intravenous urogram of a patient with PBS left hydroureteronephrosis and an elongated, dilated and tortuous left ureter.

Extra urogenital presentations of PBS are possible as shown in **Figure 11**. Two patients had malrotation of the gut. This was noted during the plication of the anterior abdominal wall. One patient had polydactyl. Ten patients with PBS had no other identified anomaly aside from the PBS.

Management of patients with PBS is quite tasking and should be multidisciplinary. In our centre, paediatricians, paediatric surgeons, urologists and radiologists are all involved. Preservation of renal function is a must. Nine patients had ureteric reimplantation, two had abdominal wall plication and one had a vesicostomy. One patient had orchidopexy in the study. The aim of these procedures was to attempt to make these patients lead a normal life. Despite the efforts of the managing team, eleven patients with PBS died during the study period as shown in **Table 4**. The female patients also seemed to fare better than their male counterparts. Other studies have reported good prognosis for patients in category 3 [3] [26]. Zugor *et al.* [27] feel that the level of renal dysplasia is the most significant factor affecting prognosis. The higher the degree of dysplasia, the worse the prognosis. We feel that despite the degree of dysplasia the poor health facilities in Africa and the late presentation contributed to the poor prognosis of the patients in this series. The most common cause of mortality in this study was progressive renal deterioration. Renal transplantation is an option for these patients if the resources are available and facilities are present [28] [29]. Renal transplantation is currently not carried out in Port Harcourt, Nigeria, so none of the patients in this study had renal transplantation.

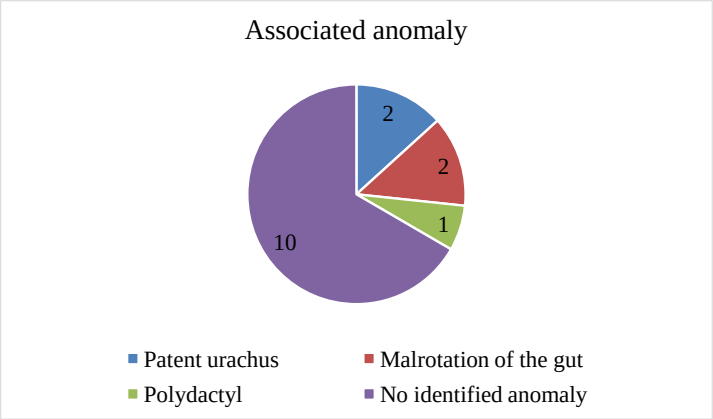

Figure 11. The anomalies associated with PBS in this study.

Table 4. The mortality of patients with PBS after a follow-up period of 5 years. More males (91.67%) tend to succumb to the disease than females.

Mortality	Frequency	Percentage
Male	11	91.67
Female	0	0
Total	11	

Follow-up is also important for these patients; the patients should have serial renal function tests. There are reports of developing renal failure and bladder malignancy later in life [25].

5. Conclusion

PBS is rare, most patients with the condition present late in Port Harcourt. Early antenatal visits can ensure prenatal diagnosis of the syndrome and hence proper and timely treatment. The most common cause of mortality was progressive renal deterioration. Adequate renal replacement therapy is also important in management.

Recommendations

Earlier registration for the antenatal clinic can lead to earlier diagnosis and better management.

Limitation of Study

This was a retrospective study. Records were poorly kept and this affected the sample size.

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Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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