

Prune belly syndrome: Early management outcome of nine consecutive cases

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Abstract

Background: Prune belly syndrome (PBS) is a rare congenital malformation of unclear etiology. The disease progress and outcome in developing countries are not clear as most reports are isolated case reports.

Materials and Methods: A review of 9 patients managed for PBS in 5 years.

Results: There were 7 males and 2 females, aged 30 min-11 days (median = 5 days) at the time of presentation (a child presented as neonate, defaulted from follow-up and represented at 10 years of life). Their weights on admission were 2.5-4.2 kg (median = 3 kg). Maternal age range was 26-37 years (median = 32 years), with five mothers being above 30 years. Seven mothers had febrile illness in the first trimester and took antimalarial drugs or antibiotics. Intestinal malrotation was the most common associated anomaly. The degree of the anterior abdominal wall and the urinary tract morphology varies from patient to patient. Urinary tract anomalies were initially managed conservatively. Two infants however later had cutaneous ureterostomy due to worsening renal function and recalcitrant urinary tract infection (UTI). Four infants had abdominoplasty at the 2nd week, 6th week, 3rd year and 10th year of life. Seven orchiopexies were done. Four were done by Fowler-Stephen's method while the rest were via the inguinal route. Of the former, 3 testicles have normal volume 6 months after, whereas one atrophied. Post abdominoplasty, there was a significant reduction in the frequency of respiratory tract infection (RTI), UTI and post void urine volume in three infants. In addition, there was improved peer interaction and academic performance in the 10-year-old child. One infant died of pulmonary hypoplasia and two others from worsening urosepsis and progressive renal failure.

Conclusion: PBS presents with a spectrum of features. Initial conservative management of the urinary tract was beneficial. Abdominoplasty and orchiopexy have both physiological and improved quality of life benefits. Early Parental education helped in reducing defaults from follow-up.

Key words: Abdominoplasty, congenital, Nigeria, prune belly syndrome, undescended testis

Date of Acceptance: 07-Nov-2013

Introduction

Prune Belly Syndrome (PBS) (Triad syndrome, Eagle-Barret syndrome) is a rare congenital anomaly^[1-11] characterized by a triad of deficient abdominal wall muscle, cryptorchidism and urinary tract anomalies.

The disease pattern and management outcome are not certain in developing countries as reports are few. Majority

of the reports are case reports and consist of findings made at presentation without sufficient follow-up to determine the course of the disease.^[4-9]

This is a report of our experience in the management of a series of patients with PBS and is intended to raise awareness and highlight the disease characteristics and outcome in this setting.

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Access this article online

Quick Response Code:



Website: www.njcponline.com

DOI: 10.4103/1119-3077.134012

PMID: 24909464

Materials and Methods

In the period of January 2008-December 2012, 9 consecutive patients with PBS were managed at the Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria. The hospital records of these patients have been reviewed and form the basis of this report. Records for the first two patients were obtained retrospectively and prospectively for the last seven patients.

The morphology of the urinary tract was assessed with ultrasonography, intravenous urography and micturating cysto urethrography. Repeat investigations were individualized based on each case presentation and progress. Regular outpatient follow-up was maintained for all the cases. Compliance to follow-up was enhanced by regular counseling and by encouraging the parents to use telephone communication for easier access to information and direction from the senior members of the managing team. Neonatologists, pediatric nephrologists, urologist, radiologist, histopathologists and social workers were all involved in the care as necessary.

Results

There were 7 males and 2 females, aged 30 minutes-11 days (median 5 days) at the time of presentation. Their weights on admission were 2.5-4.2 kg (median = 3 kg). A child presented as neonate, defaulted from follow-up and represented at 10 years of life. All were delivered at term to parents of middle and lower socio-economic class. No familial case involvement was recorded.

Prenatal ultrasound was done in six mothers, but suspicion of abdominal wall defect was made in only two cases. The sonologists actually misinterpreted the defect as omphalocele in one child. Maternal age range was 26-37 years (median = 32 years), with five mothers being above 30 years. Seven mothers took antimalarial drugs or antibiotics in the first trimester for a febrile illness. Four accepted taking herbal drugs within 3 months of conception. All but one child was delivered vaginally.

Midgut malrotation was the most common associated

congenital anomaly and is present in all the 5 children that had a need for laparotomy for varying reasons, which include abdominoplasty, excision of persistent urachus and colostomy. The occurrence and frequency of associated congenital anomalies is higher in females with pseudo-PBS [Table 1]. The degree of abdominal wall involvement is variable ranging from mild wrinkling with mild redundant abdominal protuberance to severe wrinkling with gross redundancy [Figure 1]. The imaging of the urinary tract also revealed wide variations ranging from mild caliectasis to gross tortuosity of the ureters. Three babies with gross abdominal wall wrinkling had associated bilateral hydronephrosis with gross dilatation and tortuosity of the ureters and dilated flabby bladder. The two with localized right sided wrinkling has minimal urinary tract changes. Abdominal wrinkling tends to straighten out as the child grows as follow-up of 5 of the 6 survivors showed, but the flaccidity and redundancy persisted.

Four children had abdominoplasty at the 2nd week, 6th week, 3rd year and 10th year of life. Abdominoplasty was done after the repair of patent urachus in the 2-week-old child. Increasing abdominal protrusion and deep parental concern necessitated abdominoplasty in the 6th week child with partial abdominal wall wrinkling. Protuberant abdomen, recurrent respiratory tract infection (RTI) and urinary tract infection (UTI) were the indications for the other remaining two. The technique of abdominal reconstruction was by Montfort's procedure in 3 cases. Simple fascial closure and skin reconstruction was successfully done without residual protuberance in the child with partial wrinkling (case no. 1). Post-operative clinical and ultrasound assessment of the older children (with abdominoplasty done at 3rd and 10th year) showed a significant reduction in the frequency of episodes of RTI, UTI and post void urine volume. In addition, abdominoplasty also significantly improved sense of well-being evidenced by increased play activity, peer interaction and academic performance in the 10-year-old child.

Orchiopexy via the inguinal route was successful within the first year of life in 2 infants. Single stage open Fowler-Stephen Orchiopexy was done in another 2 infants (case no. 4 and 7). Of the four testicles fixed by Fowler-Stephen's method, three has normal volume and is well-situated in the scrotum



Figure 1: Showing gradations of abdominal wall wrinkling

Table 1: Summary of patients features

Patient	Age	Sex	Gestational age at birth	Maternal age	Abdominal wall	Genito-urinary anomalies	Other associated anomalies	Treatments	Complications	Outcome
1	14 days	M	Term	28	Moderately circumscribed right sided wall deficiency (wrinkled)	Left UT Dilated upper moiety calyx	Midgut malrotation	Orchiopexy and abdominoplasty Simple fascial closure at 10 weeks	Nil	Alive
2	7 days	M	Term	26	Flabby abdominal wall with mild wrinkling	Bilateral UT Bilateral gross hydronephrosis Dilated tortuous ureters (grade IV) Posterior urethral valve Dilated bladder with trabeculated posterior wall	Midgut malrotation	Valvotomy at 10 weeks Bilateral orchiopexy at 1 year of age Abdominoplasty at 3 rd year	Recurrent pneumonia and urinary tract infection	Alive Normal renal function Improving urinary tract morphology
3	30 min	F	Term	28	Gross wrinkling of the abdominal wall	Ambiguous external genitalia Vaginal atresia Bi cornuate uterus Normal ovaries and tubes Left multicystic dysplastic kidney Right hydronephrosis Dilated and tortuous ureters (grade V) Grossly distended bladder, smooth walled Persistent urachus Urethral atresia	Rectal atresia Malrotation syndrome Bilateral CTEV deformity Scoliosis Plagiocephaly	Cutaneous ureterostomy, Sigmoid colostomy and gender assignment at birth	Urosepsis	Died at 6 months of age
4	10 years	M	Term	27	“Pot bellied” flabby abdomen. No wrinkling Pushes up the abdomen to effect micturition	Bilateral UT Bilateral hydronephrosis Hydroureters (grade IV) Mega bladder	Midgut Malrotation	Bilateral Orchiopexy (Fowler-Stephens) Abdominoplasty	Normal	Alive Improved micturition (un aided) Better quality of life
5	24 hours	M	Term	33	Gross abdominal distension and severe wrinkling	Bilateral UT Gross hydronephrotic kidneys (thin parenchyma, poor function) Hydroureters (grade V) Mega bladder	Bilateral CTEV Midgut malrotation	Bilateral cutaneous ureterostomy	Progressive deterioration of renal function UTI	Died at 6 weeks of age
6	30 mins	M	Term	35	Flabby abdomen with bilateral flank distension and severe wrinkling	Bilateral UT			Poor respiratory effort	Died within 30 min of presentation

Table continued...

Table 1: Contd...

Patient	Age	Sex	Gestational age at birth	Maternal age	Abdominal wall	Genito-urinary anomalies	Other associated anomalies	Treatments	Complications	Outcome
7	7 days	M	Term	32	Flabby abdomen, moderate wrinkling	Bilateral UT Hydroureters (grade IV) Patent urachus Flabby urinary bladder, with wide connection to the umbilicus	Midgut malrotation Patent urachus Scoliosis	Excision of the urachus and repair of the bladder Orchiopexy Abdominoplasty	Mild residual abdominal wall redundancy	Alive
8	3 weeks	F	Term	37	Right sided flabby, lax anterior abdominal wall with wrinkling	Normal female external genitalia Mid calyceal dilatation	Microcephallus Micrognathia Bilateral claw hand deformity Pectus carinatum Umbilical hernia	Non-operative	Normal	Alive
9	10 days	M	Term	32	Moderate wrinkling	Parents refused investigations	Bilateral CTEV	Defaulted from treatment and follow-up		

UT=Undescended testis, CTEV=Congenital talipes equinovarus, UTI=Urinary tract infection

after 6 months. One testicle atrophied. The indications for cutaneous ureterostomy were urethral atresia (case no. 3), and worsening urosepsis and renal failure (case no. 5). Case no. 3 also had a defunctioning colostomy for associated ano-rectal atresia.

The most common complication during follow-up was UTI and Pneumonia. Four children with clinical and bacteriological UTI received suppression antibiotics with amoxicillin 250 mg daily. Parental compliance was however generally poor. Significant reduction in the frequency of episodes of UTI was observed in only one child with a better compliance.

Three infants died. One with gross abdominal wrinkling died within 30 min of presentation from worsening breathing difficulties and non-improvement of oxygen saturation during resuscitation. Though autopsy was not done pulmonary hypoplasia is suspected as the cause of death. The other two infants (both with cutaneous ureterostomy) died at 6 weeks and 6 months of age from urosepsis and progressive renal failure. Of the six survivors, five have maintained a satisfactory biochemical renal function over 6 months-4 years of follow-up. Serial abdominal ultrasound and intravenous urography have demonstrated improving ureteral and bladder morphology. A child was lost to follow-up.

Discussion

The etiology of PBS is yet to be fully understood. Theories to explain the Pathophysiology of PBS include:

The abdominal mesodermal maldevelopment theory,^[3,12] which is due to arrested or defective development of the mesoderm of the anterior abdominal wall and urinary

tract between 6th and 10th week of gestation; Urethral obstruction malformation complex,^[13] where a transient obstruction of the proximal urethra or obstruction at the junction of the glanular and penile urethra leads to dilated urinary tract and distended abdomen.

Recent findings of deletion of hepatocyte nuclear factor-1beta (HNF-1β), a transcription factor required for visceral endoderm specification during embryogenesis, in PBS patients increasingly also supports a genetic basis.^[14-16] Though rare, cases of familial PBS have been reported.^[17,18] None of our patients however has any positive family history.

In contrast to earlier reports that PBS is more common in young mothers,^[19] more than half of our patients were more than 30 years of age. Chromosomal defects are more common in advanced age groups. Associated trisomy 21 was found in a Nigerian girl with PBS born of a 32-year-old mother.^[20] The increased associated anomalies we noted especially in females with PBS may also point to chromosomal defects as a possible etiology of PBS in our environment.

There is no definite common consistent maternal risk factor from our study. Although most mothers accepted taken drugs for febrile illness in the first trimester of pregnancy, the cause of fever and the actual type of drug taken cannot be ascertained as the drug were often self-prescribed and procured.

There seem to be a direct correlation between the severity of flaccidity and wrinkling of the abdomen to the severity of the urinary tract dilatation. Those with severely flaccid and wrinkled abdomen tend to have gross dilation and tortuosity of the ureters while those with partial wrinkling have very

minimal urinary tract morphological derangement [Table 1]. The three deaths recorded all have severely wrinkled and redundant abdomen. On the other hand severe urinary tract involvement may occur with a minimal abdominal wall involvement.^[21] More cases however need to be studied to understand this relationship better.

Prior to abdominoplasty, as the child grows the abdominal wrinkling tends to smoothen out, mainly due to deposition of subcutaneous fat, but the flaccidity and protuberance remains. Due to ineffective vasalva and poor expectoration recurrent RTI is common. This and recurrent UTI are the two most common reasons for readmission of PBS children in our series. Abdominoplasty has both physiological and psychological benefits. By providing a firm support for effective vasalva, it improves the voiding pressure and chest expectoration. The improvement in the quality of life is very remarkable in the 10-year-old child who changed from being almost withdrawn to an active affable child [Figure 2]. Corresponding improvement in academic grade was also remarkable.

The two most common associations are gut malrotation and talipes equinovarus. From our study, we observed that VACTERL and other associations occur more commonly in females with Pseudo PBS [Table 1]. This corresponds with several previous case reports.^[8,22] The multiplicity of organs affected still suggests an early noxious insult inducement of mesodermal mal-development.

From our series, the early mortality rate of 33.3% compares with the worldwide mortality rate of 20-30%.^[2] Mortality in PBS is usually due to pulmonary hypoplasia and renal failure. Early aggressive management of the urinary tract and in extreme cases renal transplantatiwon have also been done with good outcome in advanced centers.^[23] Facilities for these are essentially lacking in developing countries like ours. We managed the urinary tract anomalies conservatively, intervening operatively only in selected cases with worsening renal function or recalcitrant UTI. Measurable positive outcome following conservative urinary tract management has been reported in a review of 34 cases



Figure 2: Before and after abdominoplasty

by Hubinois *et al.*^[3] Our early preliminary results suggests that many will do well on this method in our environment. However, a longer monitoring is needed for a better understanding of the long-term outcome.

Two major limitations in managing these children are financial constraints and superstitious beliefs. Multiple hospital visitations, admissions, investigations and surgeries are often so much burden on already impoverished parents. Poverty potentiates the erroneous belief that the children are evil and of bad omen. These may be responsible for the high number of discharge against medical advice and loss to follow-up as seen in earlier reports from developing countries.^[9] These babies possibly may have been left to die at home. We have tried to counter this by maintaining a sustained parental education and counseling. They were also given telephone access to the senior members of the managing team to enhance appropriate information delivery and to dissuade them from seeking counsel from the unqualified. These were highly effective as only one case default from follow-up was recorded.

Conclusion

PBS presents with a spectrum of features. Most of the patients did well with initial conservative management of the urinary tract; intervention reserved for those with worsening renal function. Abdominoplasty and orchiopexy have both physiological and improved quality of life benefits. Early Parental education and support helped them in coping with the challenges of treatment and significantly reduced default from follow-up.

Acknowledgement

We deeply appreciate Prof. Ameh EA of Ahmadu Bello University, Zaria for his inestimable support and guidance in producing this work.

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How to cite this article: Ekunife OH, Ugwu JO, Modekwe V. Prune belly syndrome: Early management outcome of nine consecutive cases. *Niger J Clin Pract* 2014;17:425-30.

Source of Support: Nil, **Conflict of Interest:** None declared.

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