

CASE REPORT

A rare case report of prune belly syndrome with malnutrition

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Abstract

Prune belly syndrome (PBS) is a rare congenital malformation of unknown etiology characterized by a triad of deficient abdominal wall musculature, undescended testicles and urinary tract malformations. Most of the patients have pulmonary, cardiac, skeletal and gastrointestinal tract anomalies. Lack of abdominal muscles leads to constipation due to inability to perform Valsalva maneuver, which helps push the stool out of the rectum during the defecation. Additionally, frequent respiratory tract infections, persisting constipation and urinary tract infections lead to the development of malnutrition in children. We report this case to raise the awareness of low socioeconomic and low-resource medical settings that malnutrition could be existed or caused by PBS. We also encourage the expansion of pediatric surgery and family medicine training to increase the number of specialist (family medicine) to report and refer PBS in earlier phase, while working in rural areas and remote provinces.

INTRODUCTION

The World Health Organization defines malnutrition as ‘the cellular imbalance between the supply of nutrients and energy and the body’s demand for them to ensure growth, maintenance and specific functions’. Malnutrition can be classified as acute (marasmus and kwashiorkor) and chronic. In 2019 globally, 47 million children <5 years of age were wasted, of which 14.3 million children were severely wasted. This translates into a prevalence of 6.9% and 2.1%, respectively. Afghanistan has one of the world’s highest rates of stunting in children <5 years of age (41%) and severe acute malnutrition (9.5%). Only half of Afghan babies are breastfed in their first 6 months, and only 12% of Afghan children aged 12–24 months receive the right variety of food in the quantity needed for their age [10].

Prune belly syndrome (PBS), or the triad syndrome, is a rare entity, usually described in male neonates. It comprises complex urinary tract anomalies, bilateral undescended testes and absence of anterior abdominal wall muscles [1, 3]. While the cause of PBS is unknown, several aspects of the syndrome, including a high concordance rate in twins (12.2 per 100 000 live births), monozygotic male twin case reports, familial case reports and a higher incidence in males, suggest that it is influenced by a sex-linked genetic factor, although some have suggested an autosomal recessive mode of inheritance [6, 7, 8]. The anomaly occurs in 1 in 30 000 to 1 in 40 000 live births, with > 95% occurring in males and 3–5% in females [1, 4]. Clinical diagnosis and radiological investigations show only the extent of renal dysplasia and/or dysfunction [1, 5]. Increased intra-abdominal pressure, lax abdominal wall, secondary infection together will cause failure of child to thrive and lead to malnutrition.

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Figure 1: patient image on the bed.

CASE REPORT

A 4-month-old male infant was admitted in the pediatric ward of our hospital due to signs/symptoms of malnutrition. The patient was suffering from muscles wasting, weight loss and respiratory tract infection. Perinatal and family history was unremarkable. Due to insufficient breast milk, the child was also fed with bottle milk. Upon admission, the patient weight was 4.1 kg, height was 54 cm and he was suffering from severe muscle wasting. On the next day of admission in pediatric ward, pediatric surgery team was consulted for the child distended thin-walled abdomen with visible bowel loops (suspicion of bowel obstruction) and urine dripping. The pediatric surgery team saw the patient and examined him physically with relevant examinations. On examination the abdominal wall was lax and distended (Eagle Barrett feature) with both retractile testicles and external urethral meatus in position but stenotic. Belly button was intact and muscle waste was noted in upper and lower limbs Fig. 1. Ultrasonography of the patient performed by the pediatric surgery team, which confirmed left renal second-grade parenchymal disease, mild hydronephrosis of upper one-third of ureters, large distended urinary bladder (megacystis) and mild ascites in left abdomen. Since the patient was suffering

طول (cm)	وزن (kg)			
		(-2 SD)	(-2 SD)	(-1 SD)
45	1.9	2	2	2.2
46	2	2.2	2.2	2.4
47	2.1	2.3	2.3	2.5
48	2.3	2.5	2.5	2.7
49	2.4	2.6	2.6	2.9
50	2.6	2.8	2.8	3
51	2.7	3	3	3.2
52	2.9	3.2	3.2	3.5
53	3.1	3.4	3.4	3.7
54	3.3	3.6	3.6	3.9
55	3.6	3.8	3.8	4.2
56	3.8	4.1	4.1	4.4
57	4	4.3	4.3	4.7
58	4.3	4.6	4.6	5
59	4.5	4.8	4.8	5.3
60	4.7	5.1	5.1	5.5
61	4.9	5.3	5.3	5.8
62	5.1	5.6	5.6	6
63	5.3	5.8	5.8	6.2
64	5.5	6	6	6.5
65	5.7	6.2	6.2	6.7
66	5.9	6.4	6.4	6.9
67	6.1	6.6	6.6	7.1
68	6.3	6.8	6.8	7.3
69	6.5	7	7	7.6
70	6.6	7.2	7.2	7.8
71	6.8	7.4	7.4	8
72	7	7.6	7.6	8.2
73	7.2	7.7	7.7	8.4
74	7.3	7.9	7.9	8.6
75	7.5	8.1	8.1	8.8
76	7.6	8.3	8.3	8.9
77	7.8	8.4	8.4	9.1
78	7.9	8.6	8.6	9.3

Figure 2: male growth chart with standard deviation (SD).

سن (cm)	وزن (kg)			بخترا
	(-3 SD)	(-2 SD) ✓	(-2 SD)	
45	1.9	2.1	2.1	2.3
46	2	2.2	2.2	2.4
47	2.2	2.4	2.4	2.6
48	2.3	2.5	2.5	2.7
49	2.4	2.6	2.6	2.9
50	2.6	2.8	2.8	3.1
51	2.8	3	3	3.3
52	2.9	3.2	3.2	3.5
53	3.1	3.4	3.4	3.7
54	3.3	3.6	3.6	3.9
55	3.5	3.8	3.8	4.2
56	3.7	4	4	4.4
57	3.9	4.3	4.3	4.6
58	4.1	4.5	4.5	4.9
59	4.3	4.7	4.7	5.1
60	4.5	4.9	4.9	5.4
61	4.7	5.1	5.1	5.6
62	4.9	5.3	5.3	5.8
63	5.1	5.5	5.5	6
64	5.3	5.7	5.7	6.3
65	5.5	5.9	5.9	6.5
66	5.6	6.1	6.1	6.7
67	5.8	6.3	6.3	6.9
68	6	6.5	6.5	7.1
69	6.1	6.7	6.7	7.3
70	6.3	6.9	6.9	7.5
71	6.5	7	7	7.7
72	6.6	7.2	7.2	7.8
73	6.8	7.4	7.4	8
74	6.9	7.5	7.5	8.2

Figure 3: female growth chart with SD.

from chest infection, appropriate antimicrobial treatment along with oxygen support had already been started along with nutritional follow up charts (patient growth charts and patient weight curve) in pediatric ward (Figures 2-4). Both teams (pediatric and pediatric surgery) agreed to decide on definite surgical treatment after the infection suppress and the child condition returns to normal but after 48 hours of inpatient stay at hospital, the child was expired due to secondary chest infection and respiratory failure.

DISCUSSION

PBS is a congenital disorder characterized by abdominal wall musculature deficiency, urinary tract anomalies and bilateral cryptorchidism. Developmental delays and growth retardation have also been reported [9]. The massive bladder distension and urinary ascites due to severe obstructive uropathy lead to degeneration of the abdominal wall musculature and failure of testicular descent. The impaired elimination of urine from the bladder leads to oligohydramnios and pulmonary hypoplasia [8].

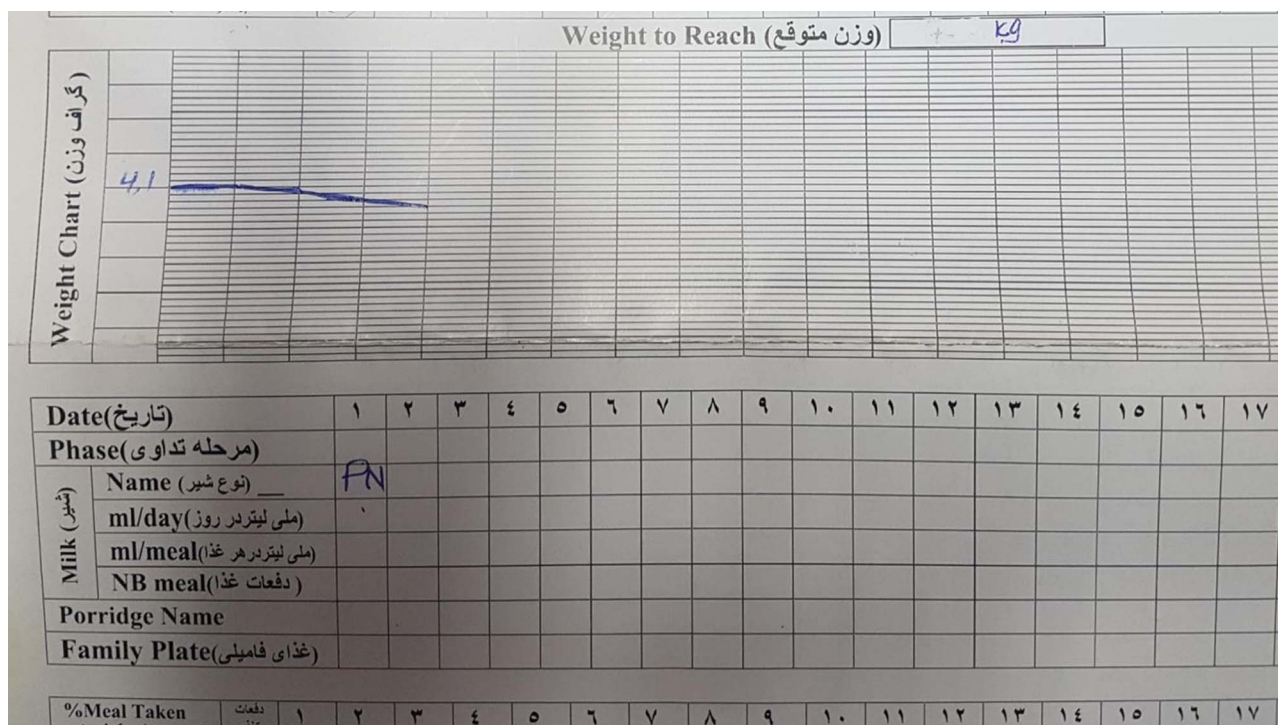


Figure 4: patient weight curve.

The association of PBS with malnutrition is not mentioned in literature, meaning that PBS itself may be an isolated condition. It could be diagnosed and treated in utero and after birth, as in developed nations, but in low-resource settings due to unawareness of the disease (because of the rarity of PBS) and late presentation of the patient due to home delivery in rural areas, the condition may have worsened and lead to malnutrition.

Treatment modality is surgical and medical. The prognosis of PBS is usually poor as many infants are either stillborn or die within the first few weeks of life due to pulmonary hypoplasia or renal failure or a combination of congenital anomalies. Chronic renal failure is the most common complication and found in 25–30% of cases [8]. Although the condition is usually incompatible with life due to visceral abnormalities especially that of renal function, there are cases who survived into adult life after urinary tract repair and abdominal reconstruction surgery [2].

Our case report reveals challenges in diagnosis of PBS before and after birth. As in Afghanistan due to unawareness of the disease and inaccessibility of the patient's family to a specialized medical center, the patient developed malnutrition as a result of causative factors. Meanwhile, the reason of reporting this case is to raise the public and medical staff awareness of rare anomalies and its resultant negative impact. We recommend further research in this regard.

CONCLUSION

Our research indicates to remind us about the challenges in prenatal/postnatal diagnosis and treatment of PBS as well as its resultant negative impact and life-threatening consequence in low-resource settings, especially Afghanistan. We recommend more research and investment on pediatric professionals and family medicine training, mostly at the level of referral hospitals

to increase the referral capacity of rare anomalies to specialized centers.

CONFLICT OF INTEREST STATEMENT

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

CONSENT

Informed consent was taken from the patient's parents.

GUARANTOR

T.H.

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