

## CASE REPORT

### Prune Belly Syndrome - A Rare Presentation in Ghanaian Neonate

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**Prune Belly Syndrome (PBS) is a rare congenital anomaly characterized by deficiency or absence of abdominal muscles, cryptorchidism and severe urinary tract abnormalities. Although it is thought to be more common in people of African descent in the USA, there are few reports of the syndrome from countries in Africa including Nigeria and Rwanda. Prenatal diagnosis through ultrasonography where the cardinal signs of hydronephrosis, bilateral hydroureters, megacystis and oligohydramnios are detected is increasingly becoming the norm. However, in resource limited settings where prenatal ultrasound services are not readily accessible or available, late postnatal presentations with pulmonary hypoplasia are encountered. This study reports of a neonate who presented with difficulty in breathing and wrinkled abdomen to a tertiary center in the Northern region of Ghana.**

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#### INTRODUCTION

Prune belly syndrome (PBS), also known as Eagle-Barrett syndrome or the Triad syndrome, is a rare congenital anomaly characterized by congenital abdominal wall muscle deficiency, cryptorchidism and severe urinary tract abnormalities (Hassett *et al.*, 2012). The syndrome is sometimes termed pseudo-prune belly syndrome when it occurs in females, in view of the fact that by definition, affected females cannot have the complete triad and the urologic manifestation may be less severe (Fotter *et al.*, 2001; Kristoff *et al.*, 2012). The syndrome was first reported in the 19<sup>th</sup> century but only got its current name in the early part of the 20<sup>th</sup> century (Osler, 1901). The incidence has been reported to range between 1/29000 and 1/40000 in different studies (Woods and Brandon, 2007). About 95% of patients with PBS are male, making the syndrome an almost exclusively male disorder (Woods and Brandon, 2007; Ademola *et al.*, 2012) with the incidence being quadrupled in twins compared to single deliveries (Balaji

*et al.*, 2000). PBS is thought to be more common in African-American/Afro-Caribbean population than Caucasians in the USA although there are very rare reports of the syndrome from Nigeria (Okeniyi, 2005; Ademola *et al.*, 2012). The exact etiology of the syndrome is not well defined but there are a number of theories to explain its etiopathogenesis. The mesodermal developmental defect during early pregnancy and proximal urethral obstruction are the two main theories advanced (Straub and Spranger, 1981; Moerman *et al.*, 1984) but none of these theories fully explains the syndrome.

The most common mode of diagnosis is an obstetric ultrasound scan (USG) in the second trimester of pregnancy. Features that may point to the syndrome include hydronephrosis, bilateral hydroureters, megacystis and oligohydramnios (Hoshino *et al.*, 1998; Hassett *et al.*, 2012). The initial postnatal management depends on the presentation and whether or not there is pulmonary involvement, but all surviving patients will eventually require a multi-disciplinary approach in long term management. Prognosis depends on severity of renal impairment and degree of pulmonary involvement but these have significantly improved over the years

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(Woods and Brandon, 2007). About 1/3 of all cases surviving beyond the neonatal period will need dialysis and subsequent renal transplant due to chronic renal failure (Crompton *et al.*, 1994) making renal and urinary tract care a central part of the management of these patients. Furthermore, patients may benefit from cosmetic surgery to reduce the redundant, floppy skin over the abdomen (Kristoff *et al.*, 2012).

In order to create awareness of this rare syndrome and to sensitize medical practitioners and sonographers of the importance of early referral for expert review when abnormal findings are suspected on obstetric USG, a case report of a day old neonate who was presented to the Neonatal Intensive Care Unit (NICU) of the Tamale Teaching Hospital after delivery at home due to difficulty in breathing and abnormally-appearing abdomen was reviewed.

#### CASE REPORT

A day old male baby who was delivered at home by spontaneous vaginal delivery (SVD) at term was brought to the NICU of the Tamale Teaching Hospital by his parents on the same day because of difficulty in breathing and abnormally-appearing abdomen. The weight at presentation was 3.2 kg and according to the mother the baby cried immediately after birth although the baby had not been fed prior to presentation at the hospital.

The mother was 30 years old, (gravida 4 para 3), with regular antenatal clinic attendance but none of the routine laboratory investigations (e.g blood group, HBsAg, G6PD, retroviral screen and VDRL) was carried out during the pregnancy. A single obstetric ultrasound scan performed in the second trimester of pregnancy at a radio-diagnostic center reported an enlarged yolk sac with sufficient liquor.

At the initial physical examination on presentation to the NICU, the weight was 3.2 kg, axillar temperature was 37.3°C and SpO<sub>2</sub> was 37% in room air with central cyanosis. There was flaring of the alae nasi and lower chest in-drawing. Air entry was bilaterally reduced on auscultation of the chest. No cardiac murmurs were heard. The abdomen was soft, bulging to

the flanks and wrinkled (Figure 1) with visible peristalsis. The baby had normal external male genitalia with bilateral undescended testes.

The patient was diagnosed as having PBS with severe respiratory distress, probably secondary to pulmonary hypoplasia. His airways were immediately suctioned to clear it of secretions and he was placed on a bubble C-PAP (Pressure of 6 cm H<sub>2</sub>O) with which the SpO<sub>2</sub> rose to a maximum of 88%. The patient was also started on parenteral ampicillin (50 mg/kg, twice daily), gentamycin (3.5 mg/kg daily), intravenous fluids (1/5 normal saline) and a statum dose of 1 mg vitamin K administered intramuscularly. Eight hours into admission in the hospital, the baby's condition began to deteriorate and the SpO<sub>2</sub> dropped to less than 50% with bradycardia on the bubble C-PAP. Cardiopulmonary resuscitation was initiated immediately but was not successful and the baby passed away after 9 hours on admission.

#### DISCUSSION

PBS is a rare congenital disorder characterized by abdominal wall muscle defects, cryptorchidism and urinary tract anomalies. The condition is a predominantly male disorder as seen in the present case and others (Ademola *et al.*, 2012). It is thought to be more common in the African-American population in the USA but there has been very few reports in literature from Africa (Ademola *et al.*, 2012). The condition is commonly diagnosed through obstetric ultrasound during the second trimester of pregnancy and in most resource-limited settings this procedure is not generally available or readily accessible. Indeed, very little is known about the condition and this coupled with the fact of non-availability or ready accessibility of diagnostic tools may lead to misdiagnosis hence the resultantly low number of cases recorded in Africa. It could also be due to the negative socio-cultural beliefs that perceive babies with congenital anomalies to be bad omen to the family, precluding families from bringing them to health facilities (kotei, 1990; Okeniyi, 2005). An extensive search through available literature has yielded no published reports of PBS in Ghana and as such makes this case a novelty. Prenatal USG is by far the most common diagnostic method for this



Figure 1: Wrinkled abdominal wall resembling a dry prune in a patient with PBS

complex syndrome and can detect the presence of the syndrome as early as 12-14 weeks of gestation (Hoshino *et al.*, 1998; Papantoniou *et al.*, 2010). In formulating a diagnosis though, other causes of lower urinary tract obstruction which may lead to distended bladder, megaureters and hydronephrosis should be ruled out. With respect to the case under review, the scan was carried out in a peripheral diagnostic center by a technician sonographer who made a record of and reported an enlarged yolk sac but the pregnant woman was not referred for a more detailed scan by an obstetrician until delivery. Termination of the pregnancy may be offered if diagnosis is made before viability (Hoshino *et al.*, 1998; Agarwal, 2005; Papantoniou *et al.*, 2010) and rightly so, in this case, the prenatal diagnosis could have helped in taking a decision regarding termination of the pregnancy.

Pulmonary hypoplasia, associated with approximately 60% of all cases, is the most common respiratory condition encountered in PBS (Hassett *et al.*, 2012; Tonni *et al.*, 2013). It is thought to be secondary to oligohydramnios which was one of the features of this syndrome (Ome *et al.*, 2013). Although the only obstetric USG performed during the second trimester in this case did not report oligohydramnios, the patient presented hours after delivery with severe respiratory distress, central cyanosis and very low oxygen saturation in room air. Majority of newborns with PBS who have pulmonary hypoplasia would die within the first week of life and in conformity to this fact, the patient in the present case died 9 hours into admission due to severe respiratory distress.

In Prune Belly syndrome, the presence of pulmonary hypoplasia in addition to severe renal dysfunction is a predictor of very high mortality in early days after birth (Woods and Brandon, 2007; Hassett *et al.*, 2012; Tonni *et al.*, 2013). Home delivery, inadequate management and delayed presentation as was done in this case might have contributed significantly to the high mortality.

### CONCLUSION

Prenatal diagnosis through USG is the contemporary practice and needs to be enhanced particularly in resource-limited settings in order to increase availability and ready accessibility. Sonographers should be encouraged to refer suspicious cases for more detailed scans and expert opinions. In cases where early diagnosis is made in gestation, termination of pregnancy could be offered. Furthermore, early detection and management of pulmonary hypoplasia which is one of the predictors of early and high mortality in Prune Belly Syndrome is very vital to survival.

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### COMPETING INTERESTS

The authors declare that they have no competing interests.

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