

Case Report

Risk factors in the development of prune belly syndrome in a Nigerian child

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Abstract

Prune Belly Syndrome is characterized by deficient anterior abdominal wall, obstruction in the urinary tract and failure of testicular descent. It occurs in 1 in 40,000 live births with a male preponderance. Its presenting features can sometimes be missed by new and inexperienced clinicians, and more so by traditional birth attendants and parents. Such cases usually will present late with complications. This report on a 3 week old male newborn that presented with typical features of the syndrome is aimed at highlighting the difficulties associated with the management of PBS in Benin City. We observed that young maternal age, male sex and use of herbal concoctions in pregnancy were possible risk factors in developing PBS. We therefore recommend that all women of child bearing age should be taught to avoid using herbal concoctions and non-prescription drugs in pregnancy.

Keywords: Prune Belly Syndrome, Risk, Factors, Nigeria.

INTRODUCTION

PBS is a distinct syndrome of birth defects comprising deficient anterior abdominal wall; obstruction in the urinary tract and failure of testicular descent (Elder JS, 2007). There are few reports on some aspects of PBS in Nigerian children (Adeyokunnu et al., 1975; Egri-Okwaji et al., 2004; Okeniyi et al., 2005; Ibadin et al., 2012) but the exact frequency and associated epidemiological factors are yet to be ascertained. Consequently, awareness of PBS is low leading to missing of many cases. Hence, there is need to constantly highlight the clinical presentations of the syndrome and the difficulties associated with its management in Nigeria. For that reason, we hereby describe our experience with a case of PBS to further illustrate the challenges that are frequently encountered in children presenting with PBS in the country.

CASE REPORT

This four week old boy presented at Modic Medical Centre on the 28 of April, 2011 with fever and poor weight gain, despite being adequately fed with breast milk and artificial formula. From birth his micturition had been characterized by small volume urine, poor stream and scattered flow. Despite these, his mother did nothing until he developed high fever with marked decrease in urine production; his urine was described as 'whitish'. Antenatal care and delivery were supervised by a traditional birth attendant that also treated his mother with topical and herbal concoction during pregnancy.

He was the second of two siblings in a monogamous setting, the other a healthy 3 year old girl. His mother is a 25 year old petty trader with some primary school education and the father a 32 year old with some secondary school education and worked as a youth leader in a political ward.

He was chronically ill-looking, pale but not jaundiced, drowsy and dehydrated with depressed anterior fontanel and sunken eyeballs. He had marked loss of subcutaneous fat, and ulcers at different stages of healing on trunk and limbs. He weighed 2.8kg with a length of 50cm, his occipitofrontal and chest circumfe-

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Figure 1. Reveals flabby abdomen, flank fullness, wrinkled skin

rences were 35 cm and 33 cm respectively. He was tachypnoeic with a respiratory rate of 68 cycles per minute and had deep sighing breaths. His pulse rate was 168 beats per minute, regular with moderate volume. Auscultation revealed transmitted breath sounds and normal heart beats.

His abdomen was flabby, full at the flanks and covered by wrinkled skin. His kidneys were ballotable. The phallus and meatus were normal, the scrotum was hyperpigmented and empty, there were no palpable testes in the inguinal canal. He was diagnosed with Prune Belly syndrome complicated by pyelonephritis, septicemia, dehydration and renal failure.

Abdominal ultrasound showed bilateral hydronephrosis and dilated prostatic urethra. Using a size 6-French feeding tube as catheter, 3.5mls of milk coloured urine was drained from his bladder. Urine microscopy and culture revealed numerous pus cells and growth of *Klebsiella*. The hemogram showed a PCV of 40%, CBC of 19.5×10^9 , neutrophil of 85% and lymphocyte of 15%. The results of the electrolytes were: Urea - 10mmol/L; Sodium - 138mmol/L; Potassium - 6.5mmol/L; Chloride - 100mmol/L; Bicarbonate - 14mmol/L and Creatinine - 85micromol/l.

He was started on intravenous Ceftriaxone and 8% dextrose saline infusion. Despite counseling and persuasion, parents insisted on discharge against medical advice on the second day of admission. Subsequent visits by the Social Welfare Unit of the hospital revealed he died on the same day he was taken from the hospital (See Figures 1, 2 and 3).

DISCUSSION

This child illustrates the striking characteristics of Prune Belly Syndrome including the epidemiological features, physical manifestations and laboratory findings. Our patient is a male and thus agrees with the observation that PBS is a condition that more commonly afflict males than females (Elder JS, 2004, Baird, 1981). The reason for the overwhelming predilection of males to the affliction is not certain but may be related to genetic factors (Elder JS, 2004, Stephens et al, 1994; Metwally, 2008; Ibadin et al 2012).

It was worthy of note that the parents of our patient were of young age; thus agreeing with the description of PBS in children born to young parents, particularly to



Figure 2. Ulcers on anterior abdominal wall at different stages of healing



Figure 3. Flabby and full abdomen, ulcers, marked wasting and dehydration

mothers aged 25 years or less (Baird, 1981; Stephens, 1994) The reason for the predilection of children of young mothers to PBS has not been ascertained.

Because the mother of this child had very limited

primary school education, we reason that she was not sufficiently empowered to make appropriate decisions regarding the use of antenatal care and herbal concoctions in pregnancy (Osaghae, 2012). It can be

argued that with a limited secondary school education, the father was also not well positioned to make better decisions on these matters. Hence this child was exposed to various herbal concoctions during fetal life. While the exact cause of PBS is not fully known, we reason that the use of herbal concoctions that may have possible teratogenic effect may have contributed to the embryonic disorders in the genitourinary system of this child with subsequent development of PBS.

In addition, the late presentation of this case meant that much could not be done for the child. Despite, overt urinary symptoms since birth, parents failed to recognize the danger signs in this child and did not take him to hospital early enough. It can be contended that if this child had been born in a hospital, the condition would have been identified much earlier with prompt early intervention. The young maternal age, low level of education coupled with no antenatal care and supervision by a traditional birth attendant meant that the condition could not be identified earlier leading to development of complications.

Other reports have also highlighted late presentation of PBS (Adeyokunnu et al, 1975; Egri-Okwaji et al, 2004; Okeniyi et al 2005; Ibadin et al 2012), in contrast to early presentation in countries with well-organized health care and antenatal facilities (Elder JS, 2004; Baird, 1981; Stephens, 1994; Metwally et al, 2008). In such countries, PBS is usually recognized before birth by ultrasonography or at birth during baby checks in the hospital.

With the observation that there may be poor awareness by the public regarding PBS in Benin City, we recommend that the Paediatric Association of Nigeria (PAN) and the Society for Obstetricians and Gynecologists of Nigeria (SOGON) should embark on an enlightenment campaign on PBS for all women of child bearing age and traditional birth attendants.

Additionally, providing free antenatal care services can go a long way in encouraging mothers to utilize hospital facilities. This could translate to early detection of PBS and other congenital abnormalities with ultrasonography.

The discharge against medical advice as seen in this case though not surprising can be minimized by providing free health care and health education for young couples.

CONCLUSION

This presentation documents a typical case of PBS seen in a four week old boy in Benin City. It found that low maternal level of education, use of herbal concoction in pregnancy were possible risk factors in developing PBS.

RECOMMENDATIONS

There is need to have central and properly documented cases of PBS and other congenital disorders in Nigeria.

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