

Prune Belly Syndrome In A Nigerian Child

J Okeniyi, T Ogunlesi, I Dedেকে, O Oyelami, G Oyedeji

Citation

J Okeniyi, T Ogunlesi, I Dedেকে, O Oyelami, G Oyedeji. *Prune Belly Syndrome In A Nigerian Child*. The Internet Journal of Pediatrics and Neonatology. 2004 Volume 5 Number 2.

Abstract

Prune belly syndrome (PBS), a rare congenital anomaly of uncertain aetiologies has been seldom reported among Nigerians. We report a 3-day old term male Nigerian neonate with PBS, conjugated hyperbilirubinaemia and features suggestive of Down syndrome who died at age 19 days. The socio-cultural factors which led to a grave outcome are discussed.

INTRODUCTION

Prune belly syndrome (PBS) also called Eagle-Barrett syndrome,¹ Obrinsky syndrome² and triad syndrome, consists of congenital urinary tract abnormalities, bilateral cryptorchidism and wrinkled anterior abdominal wall due to deficits in the musculature.³ It occurs in about 1 in 50,000 live births and the aetiology is uncertain.⁴ PBS is reportedly more common among Black Americans.⁵ Contrariwise, among Nigerians the most populous black nation, it is seldom encountered and has been reported only twice.^{6, 7} Thus, we report this case which was associated with conjugated hyperbilirubinaemia and features consistent with Down syndrome with emphasis on the confounding socio-cultural factors that lead to an early demise.

CASE REPORT

On the 14th of November 2004, IB, a 3 day-old male was brought because of neonatal jaundice to the special care baby unit of the Wesley Guild Hospital, Ilesa, Nigeria from the private hospital where he had been delivered by emergency caesarean section performed because of hand prolapse. His APGAR scores were 6 and 10 at 1 and 5 minutes respectively. He had only been given few sips of glucose drinks prior to presentation. His 38 year old mother, Para 4⁺³, had had three preceding first trimester spontaneous abortions. She had no prenatal care. It was her 45 year-old husband, a peasant farmer who complained that the boy was 'not normal'.

On physical examination, the patient was a mature baby who weighed 3.05 kg and had a full length of 46.0 cm. He was mildly pale, acyanosed, moderately icteric and afebrile (rectal temperature was 36.5°C). The head circumference

was 34.0 cm with a third fontanelle and global hypotonia. He had a flat facial profile, Mongolian eye slants (with bilateral purulent discharges), low set ears, excessive nuchal skin folds and a right-sided simian crease. His respiratory rate was 42 cycles/ minute with normal breath sounds. The heart rate was 130 beats/ minute, only the 1st and the 2nd heart sounds were heard but the pulmonary component was loud. His abdomen was flabby with a wrinkled appearance and visible peristalses and the organs were easily palpable. The liver and the spleen were palpable to 5 and 3 cm below the costal margins respectively and the left kidney was also palpable. The scrotum was hypoplastic and poorly rugous with bilateral cryptorchidism. The hips demonstrated positive Ortolani test and the left foot was clubbed with a wide sandal gap. Figure 1 shows the baby. The presumptive diagnoses were Prune belly syndrome with Down syndrome, mild birth asphyxia, moderate neonatal jaundice, ophthalmia neonatorum, hypoglycaemia and septicemia.

His random blood sugar was low (1.7 $\mu\text{mol/L}$), venous haematocrit 57% and leucocytes counts 7,000 cells/ mm^3 (lymphocytes 18% and neutrophils 82%). Serum total and conjugated bilirubin were 360 and 148 $\mu\text{mol/L}$ respectively. His blood group was B and his mother's group O, both were Rhesus positive and the Direct Coomb's Test and Hepatitis B and C viral studies were negative. The Glucose-6-Phosphate Dehydrogenase status and the serum Alkaline phosphatase (176 IU/L) were normal but both Aspartate and Alanine aminotransferases were elevated (110 and 53 IU/L respectively). Urinalysis showed bilirubin and urobilinogen. Staphylococcus aureus sensitive to Gentamicin was obtained from eye swab but the blood culture yielded no growth. Abdominal ultrasound showed left sided hydronephrosis, normal spleen, liver and hepatobiliary architecture with no

ascites. Chest x-rays was normal.

The initial management included 10% Dextrose-in-water infusion, intravenous Ampicillin and Gentamicin, vitamin K1, saline eye irrigations, topical Gentamicin eye instillation and phototherapy. His father was counselled on the diagnoses and he displayed enthusiasm. However, five days after admission, following consultations with his own father who is a native doctor and a traditional religion (Ifa) ⁸ priest, he abruptly requested for the baby's discharge from the hospital against medical advice citing financial reasons. In the interim, the patient's mother was still recuperating at the referral centre following caesarean section.

The father brought back the baby at age 19 days, still icteric and floppy. He weighed 2.75 kg. Closer and more intimate interaction with him brought out his opinion that the baby's condition was probably related to his wife's marital infidelity. He sought further medical or surgical interventions but declined our offer of re-admission and the baby was lost to follow up. A visit to their home revealed that the boy died 'suddenly' on the journey back home from the hospital that same day and has since been buried. His mother has also returned home.

Figure 1

Figure 1: The boy with PBS and Down syndrome.



DISCUSSION

Prune belly syndrome (PBS) is a congenital anomaly almost exclusive to males. ³ The features of PBS observed in our case are similar to those reported from developed countries. ^{1,2,3,4,5} Although rare, reportedly PBS is more common among Black Americans. ⁵ If there is actually a racial factor, many more cases ought to have been reported in Nigeria being the most populous black nation. Its rarity

may in part be due to reduced awareness and missed diagnosis at the maternity centres and referral hospitals. In situations whereby pregnancies and deliveries are often unsupervised many more cases may occur unnoticed. These perhaps are some reasons why PBS has been sparingly reported from Nigeria. ^{6, 7} Indeed a Cameroonian case series cited 13 cases ⁹ and Cameroon is geographically contiguous to Nigeria.

Adeyokunnu and Familusi's earlier report ⁷ suggests possible genetic basis for the anomaly as the cases cited were all members of the same extended family. The male preponderance of PBS ^{1, 2, 3} and an over four-fold increased incidence among twins ¹ are further pointers to a genetic basis. PBS has been associated with young mothers ^{5, 9} contrasting the maternal age in this present case. The mother in the present report had had recurrent spontaneous abortions. If these were due to chromosomal abnormalities, the index baby born at maternal age 38 years with features of Down syndrome lends credence to a chromosomal basis. Also, Down syndrome has been associated with PBS ¹ just as the relationship of Down syndrome, neonatal jaundice and elderly maternal age has been well established. ¹⁰ Although current theories of the pathogenesis of PBS suggest some yet unknown mesodermal injury and or in utero urinary tract obstruction, ¹¹ a genetic cause may also be operative. The lack of prenatal care prevented the analysis of the family pedigree and possible prenatal diagnoses of both syndromes. ⁹ Recently, Al Harbi reported very similar case of PBS and Down syndrome in a girl who died presumably of sepsis at an age of 29 days. ¹²

The prognosis of PBS is poor with still births and early infant deaths being common. ² Yet, recent surgical advances have dramatically improved the prospects. ¹ These are however not available in underdeveloped societies. Dysmorphism is yet a poorly understood issue in most traditional African settings where people readily explain away illnesses and diseases in terms of extra-terrestrial forces and situations surrounding childbirth. Thus, babies with anomalies are not socially accepted and stand risks of abuse, neglect and even infanticide all of which may account in some way for the apparent rarity of PBS (when such babies are not presented. The baby's paternal grandfather, a native doctor and a traditional religion priest, ⁸ strongly influenced his premature discharge from the hospital on the grounds that the anomaly was a bad omen. Was this baby killed? Although the father brought the patient back to the hospital for care after discharge against medical advice, his

refusal of further therapy raises questions. On the other hand, his explanation of the disorder as a manifestation of marital infidelity is a potential cause of disruption in the family capable of resulting in tremendous suffering to its members.

There is need for an improved social welfare system as well as better funding of the health sector in the country. Such improvements will, among other benefits give facilities for educating the community on dysmorphism, improve parents knowledge and perception of their own children's disease and also provide funds to investigate and manage affected babies. We therefore, wish to create awareness about PBS in Nigeria and advocate for an efficient social security system and much better funding of the health sector.

ACKNOWLEDGEMENT

We wish to thank the entire medical staff of the department of Paediatrics, Mr. Toluwase of the department of Chemical Pathology and Drs. C.O Jegede and O.A Oyedeji all of the Wesley Guild Hospital, Ilesa for their assistance.

CORRESPONDENCE TO

Dr. JAO Okeniyi. Department of Paediatrics and Child Health, Obafemi Awolowo University, Ile-Ife. E-mail: akinyemiokes2@yahoo.com Telephone: +234 (0) 8034014280.

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Author Information

John A.O. Okeniyi

Lecturer, Department of Paediatrics and Child Health, Obafemi Awolowo University (OAU)

Tinuade A. Ogunlesi

Senior Registrar, Department of Paediatrics, Wesley Guild Hospital

Iyabode O.F. Dedeke

Senior Registrar, Department of Paediatrics, Wesley Guild Hospital

Oyeku A. Oyelami

Professor of Paediatrics, Department of Paediatrics and Child Health, Obafemi Awolowo University (OAU)

Gabriel A. Oyedeji

Professor of Paediatrics, Department of Paediatrics and Child Health, Obafemi Awolowo University (OAU)