CASE REPORTS

Pseudoprune Belly Syndrome; Its' Associations and Management Challenge In a Developing Country: A Case Report

*Olori S *MBBS* **Ameh E A *FWACS, FACS* **Mshelbwala P *FWACS* **Gomna A *FWACS*
*Paediatric Surgery Unit, Department of Surgery, Jos University Teaching Hospital (JUTH), Jos, Nigeria.**Division of Paediatric Surgery, Department of Surgery, Ahmadu Bello University Teaching Hospital (ABUTH), Zaria, Nigeria

Abstract

Background: *Pseudo prune belly syndrome is an incomplete expression of the triad syndrome. Its incidence is poorly documented worldwide. We are not aware of any documented cases in Nigeria in recent times. Diagnosis is clinical; however, ultrasound scan plays key role in the overall assessment of the patient.*

Method: *It is a report of an 8day old boy who had pseudo prune belly syndrome with associated micro colon and rectal atresia managed at the Ahmadu Bello University Teaching Hospital (ABUTH), Zaria, in October, 2005.*

Conclusion: *Management of this patient was challenging due to lack of frozen section facility, parenteral nutrition and finance.*

Awareness of the associated conditions and how to manage them is emphasized for good outcome.

Key words: pseudoprune belly syndrome, parenteral nutrition, frozen section, electrolytes

Date accepted for publication 11th March 2008

Nig J Med 2008; 215 - 217

INTRODUCTION

Prune belly syndrome which goes by several other names is a rare congenital disorder with an incidence of 1 in 35000 to 1 in 40000 live births. It is commoner in twins, blacks, children of young mothers [1] and its genetic basis is not established [2]. It is a triad of absence or deficient anterior abdominal wall muscles, cryptorchidism and urinary tract anomalies [dilated collecting ducts, distal ureters, megacystis].

Pseudoprune belly syndrome[1, 9]; an incomplete expression of the triad syndrome is less common, and its incidence is poorly documented. Males with pseudoprune belly syndrome may be fertile if testes are descended and prostate is healthy as against infertility with full blown prune belly syndrome.

Outcome varies from early death due to the severity of renal impairment and pulmonary hypoplasia, to near normal life expectancy.

CASE REPORT

An 8day old boy was referred to the Ahmadu Bello University Teaching Hospital, Zaria in October, 2005. He was a product of a supervised term pregnancy delivered per vaginam at the referring hospital. At birth, he had a left flank swelling that later became generalized. There was delayed passage of meconium and vomiting, but, no fecaluria, pneumaturia, oligo- or poly-hydramnios, maternal illness, nor ingestion of unprescribed and herbal medications. The mother had no exposure to radiations, and antenatal ultra sound scans (USS) were normal.

He was small for age and weighed 1.25kg. The abdomen was asymmetrically distended and wrinkled with prominent visible and palpable peristaltic waves globally. The abdominal wall muscles were grossly deficient. The umbilical stump had healed and intra abdominal organs were not palpably enlarged. He had bilateral reducible inguinal hernia and a well formed scrotum with both testes in situ. There was good anal sphincteric tone but, the anorectum ended blindly 5cm from the anal verge. He had symmetrical polydactyly in both hands involving the little fingers, which had been tied off with a string. Other systems examined were normal. A diagnosis of neonatal intestinal obstruction in a patient with multiple congenital anomalies consisting of pseudo-prune belly syndrome, rectal atresia, and polydactyly was made.

Abdominal ultrasound reported normal urinary bladder but silent on the ureters. He was resuscitated with intravenous fluids. Antibiotics and vitamin-K were given. He was then prepared for and had exploratory laparotomy. Finding at operation included a micro-colon extending from the rectum to the caecum with a normal looking appendix about 12.5cm long, and 20cm length of contracted terminal ileum with distended proximal part. No intra luminal masses could be felt at the transition between dilated proximal and the distal pathologic portions of bowel. Attempt at milking the content of the proximal bowel towards the distal portion so as to assess patency was not successful. An intra operative diagnosis of associated Hirschsprung's

Correspondence: Dr S. Olori, e-mail samsonolori@yahoo.com

disease [9] to rule out a type 1 ileal atresia was made. An ileotomy was made on the dilated ileum proximal to the transition between the dilated and contracted segments. A size 6 feeding tube was then passed distally through the ileotomy, the tip of tube being short of the transition point. Normal saline was then injected through the tube to test for patency of the transition point and distal bowel. The distal bowel was filled with the saline, but, drainage through the anus was not demonstrated. He had appendectomy and biopsy from the transition zone which was sent for histology. The normal ileum was brought out as the ileostomy while the distal limb as a mucus fistula. On opening the resected segment, no membrane, web or concretion of meconium was seen. The histology report revealed normal ganglion cells in both submucosal and myenteric plexuses of both the ileum and appendix.

His post operative recovery was uneventful. The parents were taught ileostomy care, and he was discharged on request for financial reason after 2weeks. He was being planned for posterior sagittal anorectoplasty, but represented 2weeks after discharge with vomiting, refusal of feeds, and a fever of 2 days duration. He was investigated and managed for sepsis and dehydration. Biochemical parameters were sodium 138, potassium 3.2, bicarbonate 24, chloride 98 and urea 19 (mmol/l). He had correction for hypokalemia, and hydration with intravenous fluids, and after 2 days, the blood chemistry was normal. A two dimensional echocardiography was normal.

A distal loopogram was to be done to ascertain the patency and calibre and rule out any fistula with the urinary tract, however, this could not be done because of financial constraints. However, the patient began to lose feeds [breast milk] through the ileostomy, and despite efforts at modifying the feeds, he developed progressive hypokalemia, hyponatremia and metabolic acidosis which did not respond to correction, and he subsequently died.

DISCUSION

Pseudo prune belly represents an incomplete expression of the triad syndrome[1, 9].Its incidence and associated anomalies are generally poorly documented especially in the West African sub region and the authors are not aware of recent documented cases especially in Nigeria[5]. Ultrasound scan [USS] plays a key role in both prenatal and postnatal diagnosis, and follow-up. In our patient, prenatal diagnosis was missed perhaps due to inexperience of the sonologist. A plethoria of associated anomalies are documented with prune belly syndrome viz; inguinal hernia, malrotation with or without volvulus, and atresia. In this patient we found bilateral inguinal hernia, rectal atresia, micro colon and distal micro terminal ileum (histology ruled out Hirschsprung's[10]). Strangely, the proximal colon to the atretic rectum was not dilated.

The management challenges were; inadequate nutrition for which parenteral nutrition would have been helpful since the patient had a very low birth weight and malabsorption of feeds, but, this was not available in our center, finance was a great limiting factor for investigations. The lack of frozen section to rule out total colonic aganglionosis compelled us to do ileostomy rather colostomy. Our patient did not have bilateral herniotomy done while having the ileostomy because we felt his clinical state was not optimal for a prolonged anaesthesia. However, this would have been done at the ileostomy take down when his anesthetic risk would have been less.

Generally, management of pseudo prune belly syndrome akin to that of prune belly syndrome is multi-disciplinary. There are two approaches of care which include the aggressive surgical and conservative intervention[6,7,9], the former commencing as early as 10days of life to repair the deficient anterior abdominal wall, effect orchidopexy and correct any associated condition. In the conservative approach the abdominal wall defect may be left alone to improve its musculature and function as the child grows, or is aided by corsets or binders. However, if defect is severe, abdominoplasty may be affected at which setting orchidopexy may be done for cases of cryptorchidism. Early orchidopexy is usually not done because of an almost hundred percent infertility[4, 8] in survivors.

Constipation, which may be a problem in early childhood may resolve as the abdominal muscle integrity improves and patient is able to bear down while defecating.

Urinary tract anomalies should be managed resiliently. Urinary diversion may be indicated in rare cases with grossly dilated poorly draining ureters or distal obstruction. Renal dysplasia presents with progressive depreciation of renal function as against early renal shutdown which may be consistent with obstruction from urethral valve or distal ureters. A prolonged suppressive antibiotic therapy is indicated for underlying urinary tract infection. Pulmonary toileting may be required in patients with pulmonary hypoplasia[4]. Pulmonary physiotherapy which helps in clearing the air

ways of secretions and improving respiration has a useful role in managing respiratory tract infection which could decimate the patient.

Prenatal diagnosis[3] has a role in planning postnatal care, and in some cases, elective termination of pregnancy where anomaly is considered severe, or in association with other congenital anomalies not compatible with life.

Prognosis is limited by the degree of renal dysplasia and pulmonary hypoplasia[8].

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