IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 16, Issue 6 Ver. VI (June. 2017), PP 26-29
www.iosrjournals.org

Case Study on Management of Prune Belly Syndrome (Triad Syndrome)

Dr. Rajnish Kumar[1],Dr. Nameer Faiz[2], Dr.Amarendra Kumar[3],
[1]*Senior Resident, Department Of Surgery, Indira Gandhi Institute Of Medical Sciences, Patna, Bihar, India*
[2]*Senior Resident, Department Of Surgery, Indira Gandhi Institute Of Medical Sciences,Patna, Bihar, India*
[3]*Professor, Department Of General and Pediatric Surgery, Patna Medical College And Hospital, Patna, Bihar India*
Department Of General Surgery, Nalanda Medical College And Hospital, Patna, Bihar, India

Abstract: *Prune Belly syndrome, a group of birth defects has three abnormalities;- (1) poor abdominal muscular development causing the abdominal skin to wrinkle like a prune, (2) undescended testes (cryptorchidism) ,(3) urinary tract defects involving kidney, ureter and bladder.*
The defect is being diagnosed now in antenatal check-up but its clinical management has no benefit. Some new born remain undiagnosed and survived after birth presented with recurrent urinary tract infection. It is a serious problem and the survival depends upon the time of diagnosis and further management. All cases with prune belly syndrome should be evaluated thoroughly for extra abdominal abnormalities resulting on ectodermal and endodermal development. In this case the patient was diagnosed at the age of 10 years of age and managed surgically (urinary diversion and subsequent reconstruction of defects).
Keywords*:Prune-BellySyndrome; Cryptorchidism; Mega-Ureter; Splenomegaly;Surgical Repair;Abdominoplasty; Abdominal muscle deficiency*

I. Introduction

Prune-belly syndrome (PBS) represents a constellation of anomalies with variable degrees of severity. The three major findings are a deficiency of the abdominal musculature, bilateral intra-abdominal testes, and an anomalous urinary tract. The characteristic abdominal wall was first described by *Frolich*in 1839 and the full triad of anomalies by *Parker* in 1895. Osler's vivid description of the abdominal wall of an infant with the characteristic findings led to the term prune-belly syndrome [1]. Other names that have been applied to this syndrome include triad syndrome, Eagle-Barrett syndrome, and abdominal musculationsyndrome.About 300 cases have been reported in the literature and the incidence is one in 50000 live birth [2]. The diagnostic criteria are : poor or rather absence of abdominal musculature wrinkling to be a prune belly, bilateral cryptorchidism and urinary tract involvement-dysplastic/hydronephrotic , megauretar or dilated tortuous ureter, vesico-urethral construction leading to hypertrophy of bladder and urachal diverticula formation. Many infants with prune belly syndrome are either stillborn or die within the first few weeks of life from severe lung or kidney problem or combination of birth problems. Some new born survive but continue to have problemsFurther survival depends upon magnitude of renal involvement. Surgical intervention for correction of anatomical aberration, improve the quality of life. Although the cases are complicated by by associated anomalies like club foot, pulmonary hypoplasia, potter-facies and such cases are usually presented with still birth[4][8].Associated defects like persistent cloaca and preductalcoarctation of aorta have also been reported [5]. The causes of prune belly syndrome remain unknown. Some have suggested that both the abdominal wall defect and the intra-abdominal cryptorchidism are secondary to distension of the urinary tract during early fetal development. Obstruction in posterior urethra is possible cause of this distension and would explain the predominance of male patients[6].This theory suggest that testicular descent is blocked by distended bladder and that the abdominal wall defect is secondary to either urinary tract distension or fetal ascites. However other types of severe obstruction, such as posterior urethral valve, do not result in either similar abdominal wall defect or same type of urinary tract obstruction**.** [3]

Corresponding Author-
Dr. NameerFaiz, MS,MRCS
Senior Resident, Department of General Surgery,
Indira Gandhi Institute of Medical Sciences,
Patna, Bihar, India
drnameerfaiz@gmail.com

II. Case Presentation

A male child of 10 years old,presented to us with feature of run down health (poor mile stone) with history of recurrent fever and a protruded belly **(Fig-1).** On physical examination there was prune belly,bilateral cryptorchidism, enlarged liver and spleen and palpable to the lower abdomen and there was no musculature over anterior abdominal wall and underdeveloped scrotum without testis.Haematological parameters showed haemoglobin 7.5 gm%,leucocytosis, neutrophilia, serum creatinine 2.5gm/dl and associated hypercalcemia. Urine culture showed the growth of E. coli. Ultrasonographic examination showed normal contour of kidney but ureter and bladder were not outlined properly, whole abdomen was filled with cystic lesions containing debris. Inguinal canal had well visualized testes. Intravenous urography (25 ml of non-ionic contrast 300gm%) with no excretion of dye was noted on either side of ureter and bladder even after 12 hrs of injection, uroflometry (max peak flow rate was 5ml/sec).

III. Management

1. To restore renal function-**temporary diversion**, bilateral pyelostomy**(Fig 2)** and supportive treatments
2. Definitive treatment - after normal renal function i.eapprox six months later of pyelostomy, definitive reconstruction of the defect was taken by abdominal incision for excision of epidermis and dermis of central abdomen except around umbilicus. The abdomen wall central plate was incised at lateral border of the rectus muscle on the either side.
3. Transperitonial**genito-urinary procedures** were conducted by excision of the redundant pelvis and ureter , pyeloplasty , ureteroplasty and transtrigonalvesicouretralreimplantation of neoureter**(Fig 3)**. Excision of the bladder diverticula then closer of bladder over Malecots catheter then bilateral orchidopexy.
4. Completion of **abdominoplasty** by scoring the parietal peritoneum overlying the lateral abdominal wall musculature. Edges of central plate were sutured to the lateral abdominal musculature along the scored line. Lateral flaps were brought together in the mid-line with closed suction drain placed between the lateral flaps and central plate. Skin was then brought together in midline enveloping the previously isolated umbilicus.

The post-operative recovery was uneventful

Follow-up: Subsequent follow-up for 2 yrs, after operation showed improved general health and abdominal countour and strength **(Fig 4**), improved hematological, biochemical parameters uroflometry –normal and usg findings ofwell outlined kidneys Right-(6.5 cms× 3.7 cms), Left-(6.0 cms × 3.7 cms), Ureters patent along the whole length and Bladders well filled with urine.

IV. Discussion

The patient has classical defects of prune-belly syndrome i.e poorly developed abdominal musculature causing skin of the belly area like winzed prune, undescended testis and urinary tract problems resulting in recurrent urinary tract infection and mild azotemia with poor milestones. During examination besides above features it was found that no typical uro organs except dilated multiple cystic spaced areas containing debris in the abdomen with the presence of spleen, liver and testis were absent from the under developed scrotum and IVU (intravenous urography) with no excretion of dye noted either side even after 12 hours of injection and uroflometry (pathological result- max peak flow was 5 ml/sec)

Here the evidence was of obstruction and recurrent infection so in this case bilateral pyelostomy was done to relieve the obstruction. Although***Ducket***has advocated cutaneous vesicotomy[4],which provides expedient and effective bladder drainage. In this case high proximal drainage that is bilateral pyelostomy was chosen because of multiple cystic spaces on sonography. It has been also advocated by reviews of pediatric urology[7].The final goal of the surgical care of children is to assure as normal an adult life as possible. The pediatric urologist should continue to act as consultant even after the child enters adulthood for boys with prune-belly syndrome or posterior urethral valves and for children of either sex born with bladder or cloacalexstrophy. As the children grow into adults, the pediatric team must develop a liaison with a skilled, interested adult team. In this way, a lifetime plan of care may be designed and carried out to ensure well-coordinated urologic therapy that is capable of addressing the complicated problems unique to this special group of patients

V. Conclusion

Prune Belly Syndrome is a rare congenital anomaly which has no known prevention other than the routine use of screening for fetal anomalies. Routine antenatal care with ultrasonography will help in detecting renal anomalies early and optimal treatment thereafter can avoid fatal course of this condition

References

[1]. Osler 1901. Osler WO: Congenital absence of abdominal muscles, with distended and hypertrophied urinary bladder. Bull Johns Hopkins Hosp 1901; 12:331-333.

[2]. GarlingerandOtt, 1974. Garlinger P, Ott J: prune belly syndrome. Possible genetic implications. Birth Defects Orig Artic Ser 1974; 10:173-180.

[3]. Campbell-Walsh Urology, 9th ed

[4]. Ducket1976DuckettJr JW: Theprune belly syndrome. In: Kelalis PP, King LR, Belman AB, ed. Clinical PediatricUrology, Philadelphia: WB Saunders; 1976:615-635.

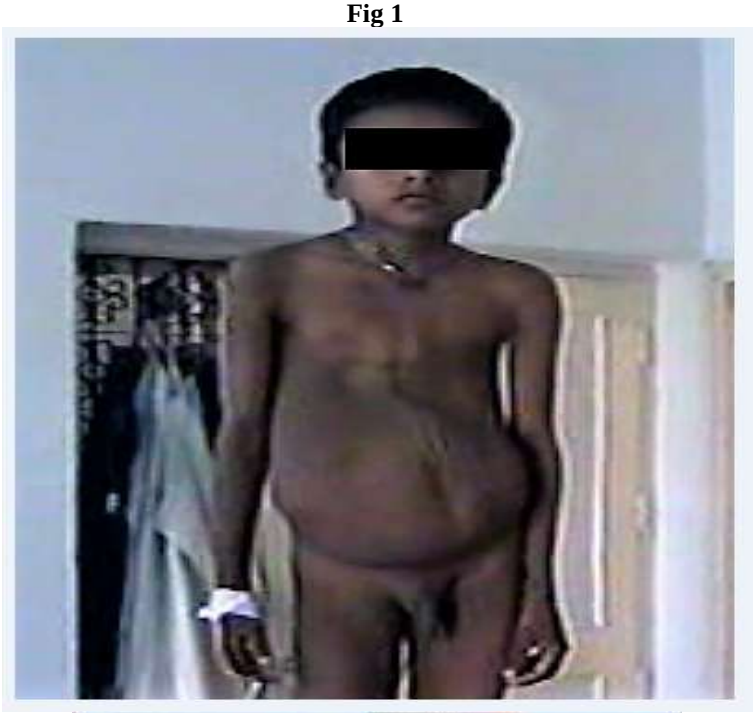
[5]. D Rupshikha et al, Prune belly syndrome, Journal of College of Medical Sciences-Nepal, 2011, Vol-7, No-3, 48-52

[6]. Nakayama et al., 1984. Nakayama DK, Harrison MR, Chinn DH, et al: The pathogenesis of prune-belly. Am J Dis Child 1984; 138:834-836.

[7]. Williams, D.I., and parker, R.M.: The role of surgery in the prune-belly syndrome: Reviews of pediatric urology. Amsterdam, ExcerptaMedica,1974,pg 315-331.

[8]. J. C. Manivel, G. Pettinato, Y. Reinberg, R. Gonzalez, B. Burke, and L. P. Dehner, "Prune belly syndrome: clinicopathologic study of 29 cases," Pediatric Pathology, vol. 9, no. 6, pp. 691–711, 1989

Fig 1



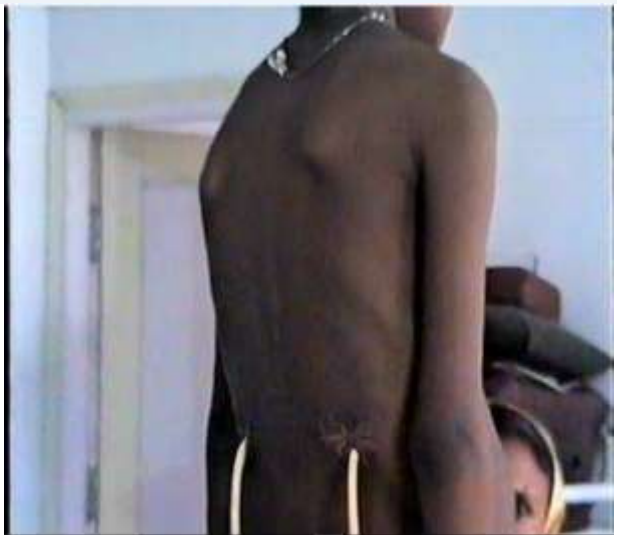


Fig 2

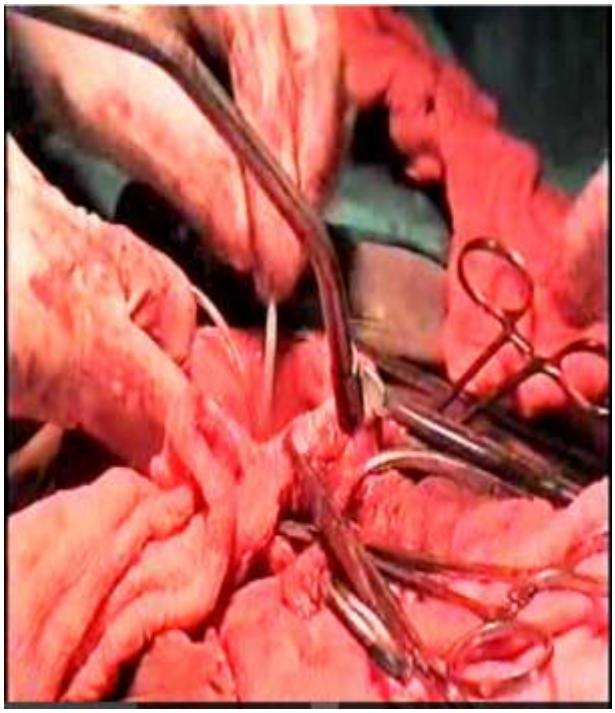


Fig 3

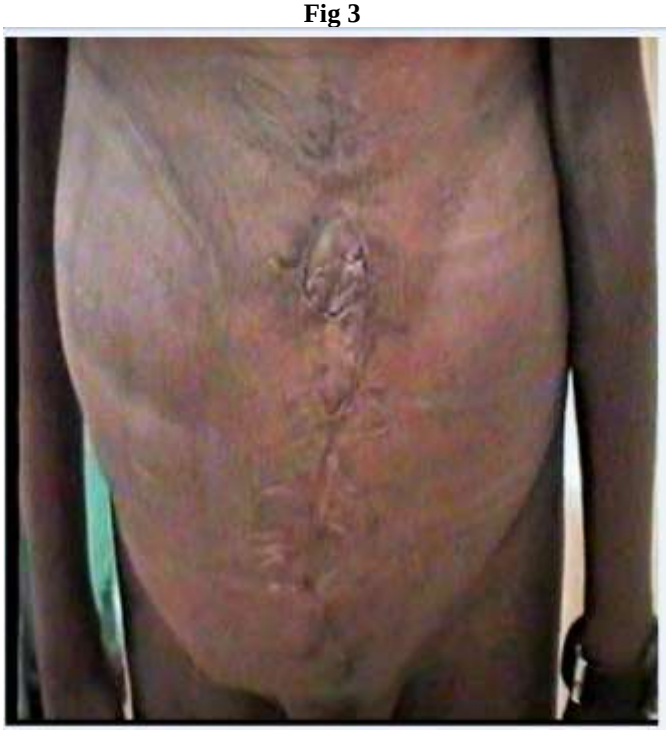


Fig 4