THE TRIAD SYNDROME OF PRUNE-BELLY: AN AGONY TO PARENTS AND MEDICAL FRATERNITY

Avir Sarkar¹, JC Sharma², Inderkant Jha³, Basanti Mazumdar⁴

¹Senior Resident, Department of Obstetrics and Gynecology, ESIC Medical College and Hospital, Faridabad

²Professor and Head, Department of Obstetrics and Gynecology, ESIC Medical College and Hospital, Faridabad

³Department of Pediatrics, ESIC Medical College and Hospital, Faridabad

⁴Department of General Surgery, Institute of Medical Sciences, BHU

Conflicts of Interest: Nil

Corresponding author: Dr. Avir Sarkar

DOI: <https://doi.org/10.32553/ijmsdr.v5i4.779>

Abstract:

Being a rare clinical syndrome, Prune-Belly is a congenitally acquired disorder with a broad spectrum of abnormalities, ranging from incompatibility with life to ageing normally and having children of their own. This case series reports 2 different cases of Prune-Belly Syndrome being diagnosed in a short time frame from the same hospital with varied presentations. It also critically highlights the role of interprofessional team in managing with this complex condition.

Keywords: Prune-Belly, syndromic triad, multidisciplinary team approach

Introduction:

Prune Belly Syndrome, also referred to as Eagle-Barrett Syndrome, is a rare congenital disorder characterized by a triad of deficient abdominal musculature, cryptorchidism and urinary tract abnormalities [1]. With a contemporary incidence of 3.6 to 3.8 per 1 lakh live births [2], almost 95% of this syndrome is detected in males [3]. We hereby report two different cases of Prune-Belly Syndrome being diagnosed in a short time frame.

Case Series

Case 1

A 22-year old primigravida presented at obstetric emergency at 39-weeks gestation in active labor. Index pregnancy was unbooked and unsupervised. On admission, fetal heart rate was normal and cardiotocograph trace was reassuring. However, clinical examination was suggestive of oligohydramnios which was confirmed by bedside ultrasound. Fetal umbilical artery dopplers were, however, normal. There was no evidence of fetal growth restriction. Maternal blood pressure readings were within normal range. Labor progressed uneventfully and she

delivered vaginally a live baby girl of 2.6 kg. On gross examination, the neonate had a deficient abdominal musculature with only skin and subcutaneous tissue covering the peritoneum¹.

Figure 1: Gross pictograph of neonate 1 showing deficient abdominal musculature.

Baby had a poor muscle tone. A probable diagnosis of Prune-Belly Syndrome was considered. The was characteristic overriding of great toes over other toes in bilateral lower limbs

Figure

2.

Figure 2: Characteristic overriding of great toes over the other toes in bilateral lower limbs.

Although a plain skiagram at birth did not show any major bony dysplasias, however, ultrasound was advised to rule out major renal and urinary tract abnormalities ^{Figure 3}.

Figure 3: Plain skiagram at birth showing no major bony dysplasias.

Case 2

A 25-year old third gravida with previous 2 full-term vaginal deliveries, got referred from a district hospital at 37-weeks gestation due to fetal growth restriction and oligohydramnios (estimated fetal weight of around 2 kg and Amniotic Fluid Index of around 3 to 4 cm). Although maternal blood pressures were normal throughout pregnancy, serial growth lags were

noticed since late second trimester. There was no history suggestive of any antenatal fever, dehydration or watery discharge per-vaginum. Fetal umbilical artery S/D ratio was elevated (3.8 to 4.0) and so parents were explained about the need for early induction of labor. With proper documentation of written informed consent and timely arrangement of NICU backup, induction was done with cerviprime gel (3 doses at 8 hourly interval). Labor progressed well and she delivered vaginally a live baby boy of 2.2 kg. Gross examination revealed absent anterior abdominal wall musculature and cryptorchidism ^{Figure 4}.

Figure 4: Clinical pictograph of neonate 2 showing deficient anterior abdominal wall musculature.

An ultrasound of abdomen showed posterior urethral valve with bilateral hydroureteronephrosis. Baseline creatinine was 0.8 mg/dl. Parental counselling was necessary regarding the prognosis of the baby and need for timely renal replacement therapy and so a pediatric surgery referral was done.

Discussion

Children born with this rare syndromic association have a broad spectrum of clinical manifestations. Although the exact etiology is clearly unknown, a number of postulates have been proposed. Possible defects in the lateral plate mesoderm or defects in the developing yolk sac could point to urinary tract abnormalities, thereby leading to bladder distension and hindrance to abdominal wall development and

testicular descent [4]. No single explanation has universal acceptance till date.

Perinatal mortality varies from 10 to 25% and is directly proportional to the degree of pulmonary hypoplasia (Potter sequence) resulting from oligohydramnios due to reduced urine output owing to urinary tract malformations [5].

The severity of defect in abdominal wall musculature is highly variable. It may range from very minute defects in few muscle fibers to complete absence of external and internal obliques and rectus abdominis muscles. In certain reported cases, skin, subcutaneous tissue and a single thin fibrous layer may be all that is found overlying the peritoneum.

Ureteral dilatation occurs generally at the distal third. Hydroureteronephrosis is secondary to posterior urethral valves and resulting vesico-ureteral reflux due to ineffective peristalsis [6]. Almost 50% of these babies have renal dysplasias of which, one-half will require renal replacement therapy at some point of time [7]. It is noteworthy to mention that unilateral normal appearing kidney in ultrasound and/or a baseline serum creatinine of less than 0.7 mg/dl during infancy may be indicators of a favourable outcome [5,8].

In the midst of diagnosing a case of Prune-belly infant, one must not forget to look for other abnormalities as these babies frequently harbor malformations involving other systems as well. About 58-60% have associated pulmonary hypoplasia, 25% have a patent ductus arteriosus, ventricular and atrial septal defects, 25% have midgut volvulus, anorectal malformations and small bowel atresia, and finally 20% babies show musculoskeletal defects like talipes equinovarus, scoliosis, hip dysplasias and overriding of great toe over other toes as in one of our index cases [5,6,9].

Conclusion

Syndromic associations like the index cases will require a multi-disciplinary team approach for their sustainable care. Physicians shall manage

the cases overall, while pediatric surgeon, urologist, nursing staff, social worker and primary health care providers at doorstep will play their vital roles as well. Coordination of ongoing care and timely counselling regarding surgery and post-op care and emotional support is also the need of the hour. Only through this type of collaboration can the interprofessional team drive outcomes in prune-belly syndrome to be successful, regardless of the severity of the particular cases.

References

1. Eagle JF, Barrett GS. Congenital deficiency of abdominal musculature with associated genito-urinary abnormalities: A syndrome. Report of 9 cases. *Pediatrics*. 1950;6(5): 721-736
2. Routh JC, Huang L, Retik AB, Nelson CP. Contemporary epidemiology and characterization of newborn males with prune belly syndrome. *Urology*. 2010;76 (1):44-48
3. Zugor V, Scott GE, Labanaris AP. The Prune Belly Syndrome: urological aspects and long-term outcomes of a rare disease. *Pediatr Rep*. 2012;4(2):e20
4. Wheatley JM, Stephens FD, Hutson JM. Prune-belly syndrome: ongoing controversies regarding pathogenesis and management. *Semin Pediatr Surg*. 1996;5 (2):95-106
5. Seidel NE, Arlen AM, Smith EA, Krisch AJ. Clinical manifestations and management of prune-belly syndrome in a large contemporary pediatric population. *Urology*. 2015;85(1):211-215
6. Nunn IN, Stephens FD. The triad syndrome: a composite anomaly of the abdominal wall, urinary system and testes. *J Urol*. 1961;86:782-794
7. Rogers LW, Ostrow PT. The prune-belly syndrome. Report of 20 cases and description of a lethal variant. *J Pediatr*. 1973;83(5):786-93
8. Noh PH, Cooper CS, Winkler AC, et al. Prognostic factors for long-term renal function in boys with the prune-belly syndrome. *J Urol*. 1999;162(4):1399-401
9. Hassett S, Smith GH, Holland AJ. Prune belly syndrome. *Pediatr Surg Int*. 2012;28(3):219-28