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**Case Report**

# An interesting case report on obstructed labour with prune belly syndrome

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## ABSTRACT

Prune belly syndrome is a rare disorder of uncertain etiology characterized by a triad of deficient abdominal musculature, cryptorchidism and urinary tract abnormalities. This condition varies in its severity which makes diagnosis challenging during early antenatal ultrasound scan. 34-year-old G4P3L3 who was counselled for pregnancy termination at 20 weeks in view of multiple anomalies detected on antenatal ultrasound. At 34 weeks of gestation presented to our institution in active phase of labour and progressed to obstructed labour and was managed by ultrasound guided fetal abdominal tapping. We conclude that suspicion of such anomalies through an early antenatal scan require further follow-up with an experienced ultrasonographer and maternal–fetal medicine specialist for a decision to be made antenatally regarding the course of pregnancy and delivery management based on the severity of the condition in a tertiary center.

**Keywords:** Prune belly syndrome, Abdominal distention, Bladder outlet obstruction, Anhydramnios

## INTRODUCTION

Prune belly syndrome is a rare congenital disorder characterized by clinical trial of deficient abdominal musculature, cryptorchidism and urinary tract abnormalities.it has an incidence of 3.6-3.8 per 100000 live male birth.[1] The exact etiology remains unknown. It appears more predominately in males, and less than 5% of those diagnosed are females.[2] With high index of suspicion prune belly syndrome can be diagnosed antenatally by ultrasound.

Children born with this condition present on a broad spectrum ranging from incompatibility with life, to aging normally and having children of their own. The severity of renal dysplasia mostly determines the survival and prognosis among the survivors. Perinatal mortality ranges between 10 to 25% in contemporary studies and directly correlates to the severity of pulmonary hypoplasia as a result of oligohydramnios from reduced fetal urine production from renal dysplasia and urinary tract abnormalities leading to Potter sequence.[3]

## CASE REPORT

A G4P3L3 34 years with 34 weeks and 5 days (LMP-17/5/2023; EDD-24/2/2024) previous 3 full term normal vaginal delivery with last child birth 5 and half years came with abdominal pain admitted to labour room. Patient was hypothyroid and was started on thyronorm 12.5 mcg. There was no other significant illness or radiological exposure in present pregnancy. Patient had irregular antenatal checkups elsewhere and ultrasound at 21 weeks had shown Anhydramnios, Bilateral hydroureteronehrosis with renal dysplasia and massively obstructed bladder and was advised medical termination of pregnancy. But the patient and bystanders were not willing and continued the pregnancy. Patient came to our institution at 32 weeks. General physical examination was normal. Obstetric examination showed fundal height of 34 weeks, cephalic

presentation and fetal heart rate was 146 bpm. Obstetric ultrasound showed single live intrauterine gestation at 30w+2d with grossly reduced liquor, abdominal cavity abdominal cavity replaced by a large cystic structure probably overdistended bladder which limits the evaluation of the abdominal organs. Pericardial effusion. Risk of preterm delivery, obstructed labour, stillbirth, IUD and congenital anomalies were explained. Termination of pregnancy was advised. Patient and bystanders were not willing for termination. At 34 weeks patient came with complaints of abdominal pain and was admitted to labour room. On examination, general condition, fair, vitals, stable, other systemic examination was under normal limits. The fundal height was 36 weeks, with good uterine contraction. The fetal heart sounds good (146 bpm). Per vaginal examination: cervix fully effaced, 5-6 cm dilated, membranes absent, ppvx (-2), pelvis-adequate. Risk of obstructed labour, still birth and need for operative interventions explained. Within half an hour patient shifted to 2nd stage of labour room.

Head and shoulders were delivered. There was no further progress of labour. Delivery of the trunk was attempted but failed. After taking informed written consent patient was shifted to operation theatre. Senior obstetricians called for help. Under general anaesthesia urgent bedside ultrasound was performed which showed massive distension of fetal urinary bladder and fetal heart was absent. Patient was planned for USG guided transabdominal drainage after localization of placenta and thereby decompressing the fetal abdomen and delivering the rest of the baby. Taking all aseptic precautions; transabdominal drainage of fetal abdominal fluid was done by wide bore spinal needle? And intravenous drip set. About 1.5 l of straw-coloured fluid was drained out. After partial decompression of fetal abdomen delivery of the trunk was completed. Congenitally anomalous still born male fetus with grossly distended and decompressed abdomen was delivered. On detailed examination: chest was significantly small, consistent with lung hypoplasia secondary to anhydramnios. Fetus abdomen was distended with absent anterior abdominal wall musculature. External genitalia were abnormal. Lower limbs were short with talipes equinovarus. All features suggestive of prune belly syndrome. Parents refused for autopsy and karyotyping. Patient was started on broad spectrum IV antibiotics. Postpartum period was uneventful and she was discharged on the 5th postnatal day.

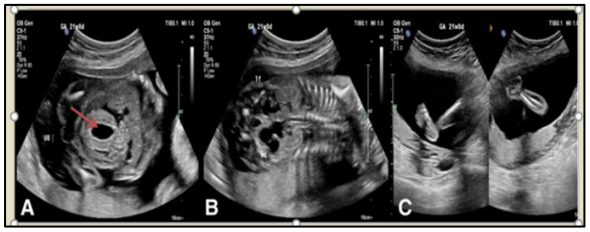


**Figure 1: USG showing, (A) fetal distended urinary bladder wall, (B) Distended ureters, (C) Bilateral clubfeet.**

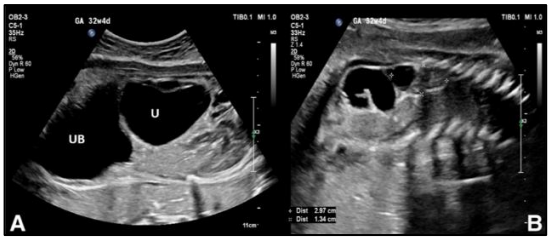


**Figure 2: (A) Hydronephrosis/Hydroureter/Bladder outlet obstruction (B) Oligohydramnios /Anhydramnios.**

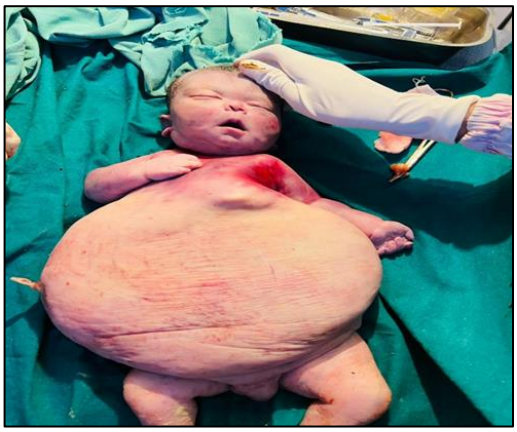

**Figure 3: Chest was significantly small, consistent with lung hypoplasia. Fetus abdomen was distended with absent anterior abdominal wall musculature.**

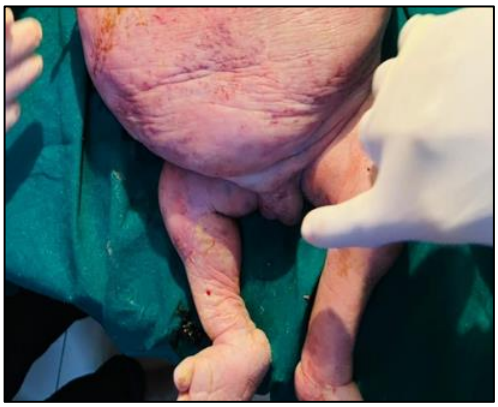

**Figure 4: External genitalia were abnormal. Lower limbs were short with talipes equinovarus.**

## DISCUSSION

It is also known as Eagle barret syndrome or Obrinsky syndrome or mesenchymal dysplasia or triad syndrome.[4] It is a rare congenital disorder characterized by the triad of deficient abdominal musculature, cryptorchidism, and urinary tract abnormalities. In addition to classical triad, associated musculoskeletal abnormality, cardiovascular abnormality and genital malformations are noted.[1,4] Exact etiology is not known. One theory is the urethral obstruction-malformation complex, and the proposal is

that a urethral obstruction during embryological development produces bladder distension that has secondary effects on the development of the urinary tract, abdominal wall, and testicular descent.[5]

Diagnosis is done antenatally by ultrasound showing oligohydramnios or anhydramnios, hydronephrosis, hydroureter, bladder outlet obstruction.[6] In new born, the prune like abdomen usually leads to the diagnosis. The prognosis of PBS is usually poor as many infants are either stillborn or die within the first few weeks of life due to pulmonary hypoplasia or renal failure or a combination of congenital anomalies.[7] Treatment will depend upon the severity of the symptoms. Some children will require rather modest surgical procedures such as the creation of a small opening in the bladder through the abdomen (vesicostomy) that will facilitate voiding of urine, or a procedure to help the testicles descend into the scrotum (orchiopexy). More extensive surgical procedures such as bladder reconstruction (cystoplasty), surgical widening of the urethra, and augmentation of the muscles that contract the bladder (detrusor augmentation) using a paired graft of a hip muscle (rectus femoris) have been successfully undertaken on children with prune belly syndrome. In rare cases, kidney transplantation may be necessary.[8] The routine use of screening for fetal anomalies has resulted in more affected pregnancies being terminated.[9]

If an antenatal diagnosis of urinary obstruction is made, it may be possible to perform vesicoamniotic shunting and intrauterine surgery to prevent the development of prune belly syndrome.[10] But in developing countries the scenario is different. A substantial percentage of antenatal women especially from rural areas do not get antenatal check-ups and ultrasonography done and the anomaly in foetus goes undetected. Such antenatal women visit the health centre only when the labour sets in and labour course is obstructed and nothing can be done to prevent the perinatal mortality.

Similar case was reported by Swaroop N, et al and Cardoso et al which was also managed by foetal abdominal tapping.[11] The lessons obtained from this presentation are that termination of pregnancy is a very challenging and difficult decision to make in the presence of multiple fetal anomalies, a complete workup and detailed counseling are required assuring the survival rate based on the severity of the condition.

## CONCLUSION

Prune belly syndrome is a rare entity worldwide with wide variability in severity and clinical manifestations. Early detection and multidisciplinary management are crucial for optimizing outcomes. It presents as a spectrum of features that may be detected during early antenatal ultrasound and hence requires an experienced sonographer in a tertiary centre and referral to fetal medicine specialists for extensive counseling and management plan.

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## REFERENCES

1. Routh JC, Huang L, Retik AB, Nelson CP. Contemporary epidemiology and characterization of newborn males with prune belly syndrome. Urology. 2010;76(1):44-8.
2. Zugor V, Schott GE, Labanaris AP. The prune belly syndrome: urological aspects and long-term outcomes of a rare disease. Pediatr Rep. 2012;4(2):20.
3. Seidel NE, Arlen AM, Smith EA, Kirsch AJ. Clinical manifestations and management of prune-belly syndrome in a large contemporary pediatric population. Urology. 2015;85(1):211-5.
4. Nunn IN, Stephens FD. The triad syndrome: a composite anomaly of the abdominal wall, urinary system and testes. J Urol.1961;86:782.
5. Wheatley JM, Stephens FD, Hutson JM. Prune-belly syndrome: ongoing controversies regarding pathogenesis and management. Semin Pediatr Surg. 1996;5(2):95-106.
6. Hassett S, Smith GH, Holland AJ. Prune belly syndrome. Pediatr Surg Int. 2012;28(3):219-28.
7. Tagore KR, Ramineni AK, Vijaya Lakshmi AR, Bhavani N. Prune Belly syndrome. Case Rep Pediatr. 2011:1-3.
8. Prune belly syndrome-symptoms, causes, treatment. 2007. National Organization for Rare Disorders (NORD). Available at https://rarediseases.org/rare-diseases/prune-belly. Accessed on 17th March 2023.
9. Ellison L, Cendron M, Ornvold K. Early diagnosis of fetal bladder outlet obstruction. J Pediatr Surg. 2000;35:513.
10. Leeners B, Sauer I, Schefels J, Cotarelo CL, Funk A. Prune-belly syndrome: therapeutic options including in utero placement of a vesicoamniotic shunt. J Clin Ultrasound. 2000;28(9):500-7.
11. Swaroop N, Sharma P, Kumari K, Seth S. Obstructed labour caused by rare congenital anomaly of foetus: managed by foetal abdominal tapping leading to successful vaginal delivery. Int J Reprod Contracept Obstet Gynecol. 2015;1220-2.
12. Cardoso PM, Surve S, Pednecar G, Mello AD. A rare fetal anomaly causing obstructed labour. J Evol Med Dent Sci. 2014;3(56):12829-33.

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