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Review Article

# A review on prune belly syndrome

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ABSTRACT

The trinity of cardinal symptoms that comprise Prune Belly Syndrome (PBS), an uncommon but severe congenital condition, include cryptorchidism, urinary tract dilatation, and flexibility of the abdominal wall musculature. In 1839, Frölich made the first mention of the prune belly condition. Osler gave the disorder its name in 1901; it relates to the wrinkly look of the abdomen brought on by the lack of muscle. Eagle-Barrett syndrome is another name for prune belly syndrome. Although aspiration pneumonia was identified as the immediate cause of death, the autopsy confirmed the PBS diagnosis.



## 1. Introduction

An uncommon multisystemic condition known as prune belly syndrome (PBS) is characterised by a lack of abdominal wall musculature, abnormalities of the urinary tract, and bilateral intra-abdominal testes. PBS in females is known as pseudo-prune belly syndrome (PPBS) and is characterised by abnormalities in the urinary system and flexibility of the abdominal wall. We report three male and one female stillborn piglets with hypoplastic abdominal wall musculature, urinary tract abnormalities, and abdominal distension. Males with bilateral cryptorchidism were noted.[1]

A trifecta of defects known as prune belly syndrome (PBS) involves flaccidity of the abdominal wall, urologic malformations, and bilateral cryptorchidism in males. Hypoplasia of the abdominal musculature puts a person at risk for respiratory issues, respiratory infections because of a compromised cough mechanism, and persistent constipation because of a weak valsalva reflex. In this article, the authors describe an infant with prune belly syndrome who experienced respiratory and gastrointestinal issues that went away after wearing a corset. According to the authors' knowledge, this is the first instance of a newborn infant being treated for PBS with a corset.[1]

The three primary symptoms of prune belly syndrome (PBS) are flaccidity of the abdominal wall, urinary abnormalities, and cryptorchidism. Urologists must therefore determine whether to carry out the procedures in a single comprehensive approach or in many steps, as well as take into account the future repair of the abdominal wall flaccidity and urinary tract anomalies, as well as the required correction of cryptorchidism.[2]

## 2. Synonyms

Abdominal Muscle Deficiency Syndrome. Congenital Absence of the Abdominal Muscles. Deficiency of abdominal musculature. Eagle-Barrett Syndrome. Obrinsky Syndrome. Triad syndrome.[3]

### *2.1. Sign and symptoms*

It is unclear what specifically causes prune belly syndrome. Most boys are affected by the illness. The developing baby's

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abdomen fills with fluid while it is within the womb. A urinary tract issue is frequently the root cause. After giving birth, the fluid drains, leaving a prune-like abdomen with wrinkles. The absence of abdominal muscles makes this impression more obvious.

Having weak abdominal muscles can lead to:

Constipation

Walking and sitting slowly

Having trouble coughing

Urination difficulties can result from urinary tract issues.

## 3. Case Presentation

A 39-year-old unregistered G5P4L4A0 who was 39 weeks and 5 days pregnant and complaining of labour pains was admitted to our labour room. She had four healthy vaginal births in the past. She had never had antenatal care before, and her tribal tribe had a lower socioeconomic position. Her husband, a 42-year-old daily labourer, had a history of consanguineous marriages. In this pregnancy, there was no history of drug use, radiation exposure, or fever with rashes. She did not have diabetes, and neither did her family have a history of genetic or congenital abnormalities.

Her cervix was fully dilated and taken up, and she was in the second stage of labour with a cephalic presentation and a regular foetal heartbeat. Her term 2700-g girl baby's Apgar scores at 1 and 5 minutes were 7 and 9, respectively. Upon physical examination, the infant's hypotonia was evident. A swollen abdomen with thin, wrinkled skin, evident peristalsis, and palpable kidneys and bladder were found during an abdominal exam. Little rugae and no testes in the sac could be seen on the scrotal skin. Additional systemic testing turned up nothing unusual. Abdominal ultrasonography revealed bilateral gross hydronephrosis and a megaureter. The infant was sent to a higher centre's paediatric surgery department for additional care. [4]

### *3.1. Causes*

It might be brought on by a flaw in the bladder's foetal development. Urine buildup can cause the kidneys, ureters, and bladder to enlarge. The abdominal muscles waste away (atrophy) when the bladder gets bigger. A big bladder or the obliteration of the groyne (inguinal) canals may be to blame for the retention of the testes in the abdomen (cryptorchidism). The obstruction at the bladder outlet or the urethral obstruction may have been cleared up by the time the baby was born, making it impossible to spot any mechanical obstacles later on. Other experts believe that the inadequate development of the abdominal muscles is a secondary cause of the urinary problems. Due to incomplete bladder emptying, infections and urine retention may develop. Symptoms and constipation. [5]

**Fig. 1:** Prune belly syndrome: Distended abdomen, thin wrinkled skin with visible peristalsis

### *3.2. Diagnosis*

The diagnosis is typically clear from birth, but locating and counting the anomalies takes time and attention. Imaging procedures, including ultrasound, X-rays, and intravenous pyelograms (IVP), will be required to fully comprehend the difficulties and gauge the degree of the genitourinary tract's involvement. An IVP uses a dye to map the kidneys' and their ducts' level of involvement. [6]

### *3.3. Treatment*

The severity of prune belly syndrome will determine the course of treatment. If the foetal ultrasound has identified the birth defect, a neonatologist who will collaborate with the obstetrician will likely examine the newborn.

If kidney impairment is thought to be present, a nephrologist may need to be consulted. It could be necessary to determine how severely the ureters have been harmed by a paediatric urologist. The trajectory of the disorder's predicted development, the child's age, how well-tolerated certain treatments are, and other considerations will all affect the course of the treatment. A course of antibiotics may be suggested for a baby with a mild incidence of prune belly syndrome in order to manage and prevent urinary

The condition could occasionally need to be treated surgically. This could entail: Vesicostomy: To help the bladder empty of pee, a tiny hole is established through the abdomen. Orchiopexy is performed on boys with cryptorchidism to advance the testes into the scrotum. Cystoplasty or bladder reconstruction surgery if the bladder is big or deformed.

Detrusor augmentation: By employing the hip muscles to strengthen the abdominal muscles, the abdominal muscles can assist the bladder in performing regularly. If a donor

is easily available, kidney transplantation may also be recommended in situations of renal failure. However, finding child donors can be challenging.

In the majority of situations, early surgical intervention is advised if the severity of the symptoms necessitates it. 30% of infants with prune belly syndrome will need a kidney transplant, according to data gathered by the National Institutes of Health in the United States. The major problem for infants born with this condition is the high incidence of kidney failure.[7]

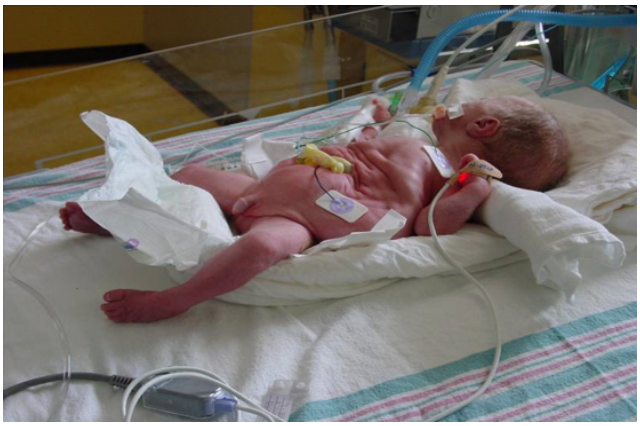

**Fig. 2:** Newborn with prune belly syndrome.

## 4. Conclusion

In a population-based study, we present epidemiological data from the present and demonstrate that the overall prevalence of PBS in Finland is 1 in 44,000. With admissions and hospital days exceeding 35 and 27 times those of the general paediatric population, respectively, PBS implies a considerable disease burden that is made worse by the 20% infant death rate.

## 5. Source of Funding

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## 6. Conflict of Interest

None.

## References

1. Martín-Alguacil LN. Prune Belly Syndrome in the Pig (Sus scrofa domesticus). *J Comp Pathol*. 2020;174:81–5. doi:10.1016/j.jcpa.2019.11.004.
2. Satar M, Özlü F, Yapıcıoğlu H, İskit S. Corset Usage for Gastrointestinal and Respiratory Problems in a Newborn with Prune Belly Syndrome. *The Indian J Pediatr*. 2016;83:717–26. doi:10.1007/s12098-015-1975-0.
3. Prune Belly Syndrome; 2023. Available from: https://rarediseases.org/?s=Prune+belly+syndrome&rdb-search=true&post_type%5B%5D=rare-diseases&post_type%5B%5D=gard-rare-disease.Retrievedon.
4. Mountsinai. Prune Belly Syndrome ; 2023. Available from: https://www.mountsinai.org/health.
5. Samal SK, Rathod S. Prune Belly syndrome: A rare case report. *J Nat Sci biol Med*. 2015;6(1):255–62.
6. Prune Belly Syndrome; 2023. Available from: https://rarediseases.org/rare-diseases/prune-belly-syndrome/.Retrievedon.
7. 2023. Available from: https://rarediseases.org/rare-diseases/prune-belly-syndrome/.Retrievedon.

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