

Prune Belly Syndrome

What is Prune Belly Syndrome (Eagle-Barrett Syndrome)?

Prune belly syndrome (PBS) is a condition that affects approximately 1 in every 30,000 to 40,000 live births. Boys account for 95% of the cases. Babies with this rare condition are born with poorly developed or missing abdominal muscles, undescended testicles (in boys), and an abnormal, expanded bladder.

Symptom severity ranges from mild to severe. Most infants with PBS also have urinary tract abnormalities, including:

- » An undeveloped urethra
- » Enlarged kidneys
- » Poorly developed kidney tissue
- » Enlarged and twisted ureters
- » A large bladder with irregularities in the bladder wall.

These issues can lead to a bladder which does not empty very well, so children must be monitored for the development of chronic kidney disease. Defects of the skeletal system, lungs, intestines, and heart can also be associated with PBS.

While the cause of PBS is often unknown, rarely is it a genetic cause.

How Is Prune Belly Syndrome Diagnosed?

PBS is often diagnosed before birth during a routine fetal ultrasound.

When you are referred to the SSM Health Cardinal Glennon St. Louis Fetal Care Institute with a suspected diagnosis of prune belly, our dedicated and multi-disciplinary team focuses on monitoring you and your baby's health and preparing for delivery. This includes performing ultrasounds and carefully watching for any signs of fetal distress.

A Cardinal Glennon St. Louis Fetal Care Institute nurse is available 24 hours a day, seven days a week to discuss referrals with physicians and potential families by calling 314-268-4037, option 2.

Phone	314-268-4037, option 2
Toll-free	1-877-SSM-FETL (776-3385)
Web	stlouisfetalcare.com
Email	fetalcare@ssmhealth.com
Facebook	facebook.com/fetaldocs

Prune Belly Syndrome

During your pregnancy, you will meet with members of our multidisciplinary team from:

- » Maternal Fetal Medicine
- » Neonatology
- » Pediatric urology
- » Pediatric nephrology
- » Pediatric surgery
- » Genetics

This team will work with you to decide the best treatment for your child after delivery. You will also have the opportunity to visit our neonatal intensive care unit (NICU) where your baby will receive the necessary care after delivery.

How Will Prune Belly Syndrome Affect My Baby After Delivery?

Every baby's situation is different, and the Cardinal Glennon St. Louis Fetal Care Institute works with each family to prepare for their baby's delivery and care after birth.

Babies born with a mild case of PBS may need simple follow-up care and monitoring from a pediatric urologist, while those with a severe case may require surgical treatment to rebuild the abdominal wall and urinary tract.

After delivery, the following tests may be needed to determine the severity of your baby's condition.

- » Renal bladder ultrasound to provide images of the bladder and kidney.
- » Voiding cystourethrogram to help determine whether a blockage in the urethra is preventing urine from passing through.
- » MAG III renal scan to provide insight into kidney function and the degree of any blockage.
- » Magnetic resonance urography for detailed pictures of the kidneys, ureters, and bladder.
- » Video urodynamic study to assess how well the bladder stores urine and contracts to empty it.
- » Blood tests to measure kidney function.

What Are Treatment Options for Babies with Prune Belly Syndrome?

Treatment options for babies with PBS depend on the severity of symptoms. A team of specialists, including nephrologists and urologists, can treat other birth defects associated with the condition.

Other babies may require dialysis if the kidneys are not functioning properly. Abdominal wall reconstruction, surgery for undescended testicles, and other procedures to correct anatomical abnormalities may also be necessary.

How Will Prune Belly Syndrome Affect My Baby Into Childhood?

There is no cure for PBS, though treatment options and our understanding of the condition are consistently growing. The prognosis for newborns with this condition varies and depends on the severity of symptoms and kidney function. In severe cases, dialysis or a kidney transplant may eventually be required. Overall, many people with PBS have good physical and mental health.

From your first appointment to your last, our fetal and baby specialists are available to care for you and your baby's complex needs. Our multi-disciplinary team collaborates across our hospital to provide your baby the specialized care they require.

We understand that prune belly can be a scary diagnosis. That's why we're available to help 24 hours a day, 7 days a week. For more information or to schedule an appointment, call us at 314-268-4037 or toll free at 877-SSM-FETL (877-776-3385).

While we can't change the diagnosis, we can provide you with expert care and support, helping your baby get the most out of treatment and life.
