CONDITION

Prune belly syndrome

Prune belly syndrome is a congenital condition affecting the abdominal muscles, urinary tract, and usually the testes.

WHAT IT IS
Congenital abdominal muscle and urinary tract condition

BODY SYSTEM
Urinary, muscle, and reproductive systems

MAIN SYMPTOMS
Wrinkled abdomen, urinary infections, weak cough, undescended testes

WHAT CAUSES IT
Unknown, with developmental and possible genetic factors

HOW IT IS DIAGNOSED
Prenatal ultrasound or examination with urinary tract imaging

MAIN TREATMENT
Urinary, kidney, surgical, and supportive care

CAN IT BE CURED?
No - congenital organ changes need ongoing care

IS IT AN EMERGENCY?
Sometimes - fever with urinary symptoms or greatly reduced urine

LONG-TERM OUTLOOK
Best when kidney and lung function remain strong

Overview

Prune belly syndrome is a rare condition that develops before birth. It affects how the abdominal muscles and urinary system form. Nearly all people with the condition are male, although a similar pattern can occasionally occur in females.

The name comes from the loose, wrinkled skin that may cover the abdomen. This appearance is caused by abdominal muscles that are very thin, weak, or partly absent. The condition is not caused by eating prunes or by anything done during pregnancy.

Prune belly syndrome usually involves three main features:

The urinary tract includes the kidneys, ureters, bladder, and urethra. The ureters are the tubes carrying urine from the kidneys to the bladder. In prune belly syndrome, these structures may

be stretched, widened, or unable to move urine normally.

The effects range from relatively manageable urinary problems to serious kidney and breathing difficulties. The outlook is influenced mainly by kidney development, urine drainage, and lung function.

- Weak or missing abdominal muscles
- An enlarged or unusually formed urinary tract
- Testes that have not moved into the scrotum in males

Symptoms

The most noticeable feature is often a soft, wrinkled abdomen. Because the abdominal muscles are weak, the belly may look enlarged. A child may have difficulty sitting up, coughing strongly, or clearing mucus from the lungs.

Urinary symptoms depend on how well urine drains. Some babies and children have few outward symptoms at first. Others develop:

Urine may flow backward from the bladder toward the kidneys. This is called vesicoureteral reflux. Urine may also remain in widened ureters or in the bladder, increasing the chance of infection.

In males, both testes are usually undescended. This means they remain in the abdomen rather than moving into the scrotum before birth.

Some babies have underdeveloped lungs. This is more likely when severe urinary tract problems reduce the amount of amniotic fluid surrounding the baby during pregnancy. Low amniotic fluid can limit lung growth.

Other possible features include constipation, differences in the spine or limbs, and heart or digestive tract abnormalities. Not everyone has these problems.

- Repeated urinary tract infections
- Fever, irritability, or poor feeding during an infection
- A weak urine stream or difficulty emptying the bladder
- A swollen abdomen from a large bladder
- Reduced urine production
- Poor growth when kidney problems are significant

Causes and risk factors

The exact cause is not known. Prune belly syndrome begins early in fetal development, when the abdominal wall and urinary tract are forming.

One explanation is that a blockage or functional problem in the developing urinary tract causes the bladder and ureters to expand. This stretching may interfere with abdominal muscle development. Another explanation is that the muscles and urinary tract are affected by a broader developmental change from the beginning.

Genetic factors may contribute in some families. Changes in several genes have been reported, but no single genetic change explains most cases. The condition usually occurs without a known family history.

There is no good evidence that it results from a parent's diet, activity, or ordinary actions during pregnancy. Parents should not assume that they caused it.

Diagnosis

Prune belly syndrome may be suspected during pregnancy. An ultrasound may show a very large bladder, widened ureters, enlarged kidneys, or low amniotic fluid. The abdominal wall may also appear unusually thin.

After birth, diagnosis is based on the child's physical features and tests of the urinary system. There is no single blood test that confirms every case.

Testing may include:

A voiding cystourethrogram uses X-rays while the bladder fills and empties. It can show whether urine travels backward toward the kidneys or whether there is an obstruction.

Doctors may also assess breathing, heart structure, bowel function, bones, and spine. Genetic testing may be offered in selected cases, especially when there are other congenital differences or a family history. A negative genetic test does not rule out the condition.

- Kidney and bladder ultrasound to examine their structure
- Blood tests to assess how well the kidneys filter waste
- Urine tests to look for infection or kidney problems
- A voiding cystourethrogram to show urine flow and reflux
- A kidney scan to measure drainage and function in each kidney

Treatment and management

Care is matched to the organs affected and the severity of the urinary problems. A child may be supported by specialists in children's urinary surgery, kidney care, respiratory care, and rehabilitation.

The first priorities after birth are breathing and safe urine drainage. A baby with underdeveloped lungs may need oxygen or other breathing support. If the bladder cannot empty, temporary catheter drainage or another procedure may be required.

Urinary tract infections are treated with antibiotics like amoxicillin or doxycycline . Some children at high risk of repeated infection may be offered preventive antibiotics, but this is an individual decision based on urinary anatomy, kidney function, and previous infections.

Regular monitoring commonly includes urine tests, blood pressure checks, kidney blood tests, and ultrasound scans. These checks can identify infection, poor drainage, or declining kidney function before symptoms become obvious.

Surgery may be considered to:

The operation that moves a testis into the scrotum is called orchiopexy. It may support examination and future reproductive care, although fertility can still be affected.

Abdominal wall reconstruction may improve support, appearance, coughing, or movement in selected children. It does not correct kidney damage.

Some children need help emptying their bladder, such as intermittent catheterisation. This involves passing a small tube into the bladder at planned times. Bowel treatment, physiotherapy, and breathing exercises may help with constipation, posture, mobility, and mucus clearance.

Advanced kidney failure may require dialysis or a kidney transplant. These treatments are needed only when the kidneys can no longer manage the body's fluid and waste adequately.

- Improve drainage from the bladder or ureters
- Reduce severe urine reflux or obstruction
- Reconstruct the abdominal wall
- Move undescended testes into the scrotum

Complications

Urinary tract infections are common because urine may drain slowly or remain in the bladder

and ureters. An infection can sometimes spread to a kidney or the bloodstream.

Kidney damage may be present from birth or develop over time. Possible consequences include high blood pressure, abnormal salt or fluid balance, chronic kidney disease, and kidney failure.

Weak abdominal muscles can reduce the strength of a cough. This may make chest infections harder to clear, especially in babies who also have underdeveloped lungs.

Other possible complications include:

Not every child develops these complications. Routine follow-up focuses on finding treatable changes early.

- Bladder emptying problems and urinary leakage
- Kidney stones
- Constipation
- Reduced core strength or changes in walking and posture
- Fertility problems related to undescended testes
- Testicular cancer risk associated with testes that remain undescended

Prevention

Prune belly syndrome cannot usually be prevented because it develops before birth for reasons that are not fully understood.

Prenatal ultrasound can sometimes identify the condition, but screening does not prevent it. In selected pregnancies, fetal urinary procedures may be considered when there is severe obstruction. These procedures carry risks and do not reliably prevent all kidney, lung, or abdominal effects.

If there is a family history or a suspected genetic cause, genetic counselling can explain what is known about recurrence. In most families, an exact recurrence risk cannot be determined from the diagnosis alone.

Living with it and what to expect

The first months of life often involve close attention to breathing, urine drainage, infection, and kidney function. Children with severe lung underdevelopment or major kidney abnormalities may need prolonged hospital care. Those with better lung and kidney function may leave

hospital with a plan for regular monitoring.

Over childhood, urinary anatomy usually remains abnormal even when surgery improves drainage. Kidney function may stay stable for years or may gradually decline. Regular testing remains important because early kidney changes do not always cause symptoms.

Some children have repeated procedures or hospital visits. Others need mainly clinic appointments, imaging, and prompt treatment of infections. School attendance, play, and family activities are often possible, although care routines and physical limits may need to be accommodated.

Weak abdominal muscles may continue to affect posture, coughing, exercise, and bowel movements. Physiotherapy and an activity plan can support strength and mobility. Contact sports may require individual advice if the kidneys or urinary tract are enlarged or vulnerable.

During adolescence and adulthood, care should move to clinicians experienced in adult kidney and urinary conditions. Follow-up may include blood pressure, kidney health, bladder emptying, sexual health, and fertility.

The long-term outlook cannot be predicted from abdominal appearance alone. Kidney and lung function are much more important. People with preserved kidney function may live well into adulthood, while severe kidney or lung involvement can shorten life or require intensive long-term treatment.

When to get medical help

Contact the usual healthcare team promptly for possible urinary infection symptoms, including fever, pain or crying during urination, cloudy or strong-smelling urine, new wetting, back pain, poor feeding, or unexplained irritability. Children with urinary tract abnormalities may need testing even when symptoms seem mild.

Arrange routine medical review for:

Seek urgent medical care if there is fever with urinary symptoms, repeated vomiting, marked sleepiness, significantly reduced urine, or inability to pass urine.

Call emergency services for severe difficulty breathing, blue or grey lips, collapse, or a child who is difficult to wake. These are emergency signs rather than routine features of prune belly syndrome.

- Changes in urine stream or bladder emptying
- Increasing constipation or abdominal swelling

- New urinary leakage
- Poor growth, tiredness, or reduced appetite
- Missed kidney, blood pressure, or imaging follow-up

Common questions

Can prune belly syndrome be cured?

There is no treatment that makes all affected muscles and urinary organs develop normally. Surgery and supportive care can improve urine drainage, abdominal support, and other specific problems. Ongoing kidney and urinary monitoring is usually still needed.

Why does the stomach look wrinkled?

The skin wrinkles because the abdominal muscles underneath are thin, weak, or missing. The bladder and urinary tract may also have stretched the abdomen before birth. The wrinkles themselves are not painful.

Can a child with prune belly syndrome walk and go to school?

Many children can walk, attend school, and take part in everyday activities. Weak core muscles, medical appointments, infections, or kidney treatment may affect individual routines. Physiotherapy and school support can help when needed.

Will my child need surgery?

Many children have surgery, but the type and timing are not the same for everyone. Procedures may address urinary drainage, the abdominal wall, or undescended testes. Decisions are based on kidney function, infections, bladder emptying, and overall health.

Can people with prune belly syndrome have children?

Fertility is often reduced in affected males because the testes developed in the abdomen and may not produce sperm normally. Some adults may still have biological children, sometimes with fertility treatment. Sexual function and fertility are separate issues and should be assessed individually.

Does prune belly syndrome get worse with age?

The congenital anatomy does not spread through the body. However, repeated infections, poor urine drainage, or abnormal kidney development can lead to worsening kidney function over time. Regular follow-up is intended to detect these changes early.

Sources

- MedlinePlus Genetics: Prune belly syndrome , U.S. National Library of Medicine
- National Organization for Rare Disorders: Prune Belly Syndrome , Rare Disease Database
- Arlen AM, Nawaf C, Kirsch AJ. "Prune belly syndrome: current perspectives." , Pediatric Health, Medicine and Therapeutics , 2019;10:75-81
- Zugor V, Schott GE, Labanaris AP. "The Prune Belly syndrome: urological aspects and long-term outcomes of a rare disease." , Pediatric Reports , 2012;4(2):e20