

Prune Belly Syndrome

What is prune belly syndrome?

Prune belly syndrome consists of three abnormalities:

1. Deficiency of the abdominal (belly) wall muscles that create a wrinkled appearance
2. Both testicles are undescended (have not dropped into the scrotum)
3. Dilated urinary tract (Kidneys, ureters, bladder, and urethra: the organs that produce, store, and excrete urine).

The majority of children with prune belly syndrome are male (>95%).

What are the symptoms of prune belly syndrome?

The symptoms of prune belly syndrome vary in severity and can be categorized on a scale from I-III.

Category I patients are the most severe and have poor lung and kidney development. These children usually do not survive more than a few days after birth.

Category II patients do not have lung problems but have significant problems with the urinary tract. They may develop kidney failure overtime.

Category III patients do not have lung problems and their kidney function is not severely impaired.

How is prune belly syndrome diagnosed?

Prune belly syndrome may be suspected due to the appearance of a dilated urinary tract on prenatal ultrasound. Once the child is born the combination of physical exam (wrinkled appearance of the belly wall and the presence of undescended testicles) and radiographic studies to identify the dilated urinary tract (ultrasound and voiding cystourethrogram) make the diagnosis.

How is prune belly syndrome managed?

The goal for management of prune belly syndrome is to preserve kidney function and prevent urinary tract infection. Sometimes surgical intervention for reconstruction of the urinary tract and the abdominal wall is required. Children may be placed on prophylactic antibiotics to help prevent urinary tract infection. Male children with prune belly syndrome will also require surgery to place the testicles down in the scrotum. All patients require lifelong surveillance because they are at risk for urinary tract infection and decline in kidney function overtime. Instrumentation of the urinary tract (including catheterization) should be performed with caution and avoided when possible since this places the child at risk for urinary tract infection.

See the next page for contact information.

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