

# Clinical Quiz

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## A neonate with lax abdominal wall and defective urogenital system plus dyspnoea

### Clinical Presentation

A 2-day old neonate was born with difficulty in breathing and Apgar score of 7. His birth weight was 800 gm. He was the third live birth to a closely related couple, with no history of any abnormality in the other 2 siblings. Examination revealed a dyspneic neonate with all accessory muscles of respiration in use (**Figure 1**). He also showed features of Pierre Robin syndrome. His abdominal skin was redundant and wrinkled (**Figure 2**), and the scrotum was empty. There were no other external significant findings. Blood tests revealed no significant findings, but ultrasound test of the abdomen showed an absent right kidney.



**Figure 1** - Neonate showing features of Pierre Robin syndrome and excessive use of respiratory muscles.



**Figure 2** - Typical Prune-Belly syndrome with lax wrinkled abdominal wall.

## Questions

1. What is the likely diagnoses?
2. What other organs of the body can be affected in this condition?
3. What is the prognoses of patients with the disease?

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## Answers

1. Prune Belly Syndrome.
2. The genitourinary system, kidneys, ureters, and the testes, in addition to the abdominal wall and the maxillofacial area..
3. The prognosis for patients with prune belly syndrome varies, depending on the severity of the disease, from stillbirth to a male with just undescended testicles and a minimal degree of abdominal wall laxity.

## Discussion

Prune-Belly syndrome was first described by Frolich in 1839, but Osler was the first to name it “Prune-Belly syndrome”,<sup>1</sup> because the abdominal wall in these infants typically looks like a “prune” which means old leather bag. It is also known as the Triad syndrome and Eagle-Barrett syndrome. The conditions consist of a triad of developmental defects of the abdominal wall musculature associated with urinary tract abnormalities and cryptorchidism.<sup>1,3</sup> Our patient demonstrated these 3 main features, in addition to Pierre Robin syndrome (**Figure 1**), which can, in addition to other orthopedic and cardiac anomalies, be present in some of these cases.<sup>4,5</sup> Prune-Belly syndrome affects males, as in our case, more frequently than females,<sup>1</sup> with a mortality rate of up to 20%. Our patient died due to respiratory failure because of severe chest infection resulting from poor cough mechanism secondary to inadequate movement of the weak abdominal muscles; this is well known in these patients.<sup>1</sup>

The etiology of Prune-Belly syndrome is not fully understood. Initially, it was postulated that the condition resulted from dilatation of the urinary bladder and ureters in utero secondary to obstructive uropathy, the dilated bladder then distends the abdomen resulting in atrophy of the abdominal wall muscles.<sup>1,5</sup> However, the most recent works on the subject concluded that the condition is actually due to mesodermal arrest, which would explain the involvement of the genitourinary tract, the testis, and the abdominal wall.<sup>1,5</sup>

## References

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