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# Anesthesia without Muscle Relaxants in an Infant with Prune Belly Syndrome: A Case Report

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# Case Report

#### **Abstract**

Prune Belly Syndrome (PBS) is a rare congenital disorder characterized by the absence of abdominal muscles, urinary system anomalies, and undescended testes. In this case, an 8-month-old male patient with PBS underwent laparoscopic orchiopexy, and anesthesia management was carefully planned to maintain respiratory and cardiovascular stability. The avoidance of muscle relaxants during anesthesia minimized postoperative complication risks and facilitated rapid recovery.

### Keywords:

Prune-Belly syndrome; anesthesia; pediatric; bilateral undescended testes

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# Introduction

Prune Belly Syndrome (PBS) is a congenital triad characterized by partial or complete absence of abdominal muscles (giving the appearance of a prune-like abdomen), renal malformations, and undescended testes [1]. Additional abnormalities may involve the cardiovascular system, aplasia or hypoplasia of the lungs, rib anomalies, musculoskeletal system, gastrointestinal tract, and central nervous system [2]. It is more common in males, with a prevalence of 1 in 30,000–40,000 live births and a mortality rate of 20% [3]. In this case, we aimed to present the anesthesia management of a patient with PBS who underwent surgery for urogenital anomalies.

# **Case Report**

An 8-month-old male patient, weighing 8 kg, diagnosed with PBS, was scheduled for laparoscopic orchiopexy due to bilateral undescended testes. During the preoperative evaluation, the characteristic prune-like abdominal appearance was observed (Figure 1). On physical examination, the patient was conscious, with an arterial blood pressure of 85/56 mmHg and a heart rate of 142 bpm. Echocardiography showed normal-sized right and left heart chambers with patent ductus arteriosus and a secundum-type atrial septal defect. Ultrasonography revealed renal enlargement. Laboratory values were within normal limits.

The patient was classified as American Society of Anesthesiologists (ASA) III and taken to the operating room. Standard monitoring was applied. For anesthesia induction, sevoflurane

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was used, followed by peripheral IV cannulation. After administering 2% propofol (3 mg/kg), fentanyl (2  $\mu$ g/kg), and remifentanil infusion (0.05  $\mu$ g/kg/min), endotracheal intubation was smoothly performed with a 4.5 mm inner diameter cuffed endotracheal tube. No muscle relaxants were used during intubation or anesthesia maintenance.

The lungs were mechanically ventilated with  $O_2$ –air (40–60%) and sevoflurane. Tidal volume and ventilator frequency were adjusted to maintain normocapnia (EtCO<sub>2</sub> 40  $\pm$  4 mmHg) with 5 cmH<sub>2</sub>O positive endexpiratory pressure (PEEP). Pneumoperitoneum was established by insufflation of CO<sub>2</sub>, and intra-abdominal pressure was kept below 12 mmHg. Postoperative pain management was achieved with acetaminophen (15 mg/kg).

The surgery lasted 75 minutes and was completed without complications. The patient was extubated smoothly. After receiving oxygen therapy by mask in the recovery unit, he was discharged to the ward after two hours and sent home without issues 48 hours later.

### Discussion

Prune Belly Syndrome, also known as Eagle-Barrett Syndrome, is a rare congenital anomaly characterized by the classic triad of abdominal wall muscle deficiency, genitourinary tract anomalies, and bilateral cryptorchidism, with an estimated incidence of 1 in 30,000–40,000 live births [4]. The pathophysiology is thought to be related to early urinary tract obstruction, leading to oligohydramnios and secondary pulmonary hypoplasia, abdominal wall hypoplasia, and Potter-like facial features [5].

Anesthesia management in PBS requires special consideration due to multisystem involvement. The absence or severe hypoplasia of abdominal musculature affects not only intra-abdominal pressure

dynamics but also respiratory mechanics, with reduced cough efficacy, diaphragmatic dysfunction, and a predisposition to atelectasis and respiratory infections [6].

Intraoperatively, airway management and neuromuscular blockade decisions are critical. Although PBS is not classically associated with a difficult airway, individual case reports have described airway challenges due to associated craniofacial features (e.g., Potter facies) [7]. In a case by Baris et al., difficult intubation was encountered, and a supraglottic airway device (LMA<sup>TM</sup>) was successfully used after multiple failed attempts [8]. Minimizing airway instrumentation and stimulation may therefore be beneficial in select PBS patients, especially those with craniofacial anomalies.

The use of neuromuscular blocking agents (NMBAs) in PBS is controversial. Due to hypoplastic abdominal muscles, the pharmacodynamics of NMBAs may be unpredictable, and residual neuromuscular blockade may significantly impair postoperative respiratory function [9,10]. Moreover, PBS patients may exhibit altered distribution and metabolism of these agents, particularly in the presence of renal anomalies. Hence, many authors recommend avoiding muscle relaxants when feasible, particularly in short or minimally invasive procedures [11].

In our case, to mitigate these risks, we avoided NMBAs and instead used a combination of remifentanil infusion and sevoflurane, which provided adequate surgical relaxation and hemodynamic stability throughout the laparoscopic procedure. This approach has also been reported as effective in other pediatric patients where neuromuscular blockade was contraindicated or undesirable [12]. Although laparoscopic procedures inherently increase intra-abdominal pressure and may challenge respiratory mechanics, we prioritized spontaneous recovery of neuromuscular function and

reduced postoperative pulmonary risk by omitting muscle relaxants. This decision aligns with reports suggesting individualized anesthetic strategies based on the patient's neuromuscular and pulmonary reserve [13].

### Conclusion

In this case, we emphasized the importance of individualized anesthesia planning in patients with particularly Prune Bellv Syndrome, during laparoscopic procedures. Avoidance of neuromuscular blocking agents may reduce postoperative respiratory especially complications, in patients with compromised abdominal wall function. A tailored approach using short-acting opioids and volatile agents can provide effective intraoperative conditions while preserving spontaneous recovery, underscoring the need for vigilant perioperative assessment and planning in this unique patient population.

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# Ethics

#### Informed Consent

The patient provided consent for the clinical information pertaining to the case to be published in a medical journal.

#### **Author Contributions**

Surgical and Medical Practices – N.M., F.C.; Concept - N.M., F.C.; Design - N.M., F.C.; Data Collection and/or Processing - N.M., F.C.; Analysis and/or/Interpretation - N.M., F.C.; Literature Review - N.M., F.C.; Writing - N.M., F.C..

#### **Declaration of Interests**

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