


## Anesthesia Management in a Pediatric Patient with Prune Belly Syndrome; A Case Report

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### Abstract

Prune belly syndrome (PBS) is a rare congenital disorder, which consists of three symptoms: anterior abdominal muscle deficiency, cryptorchidism and genitourinary malformation. These patients have cardiovascular and musculoskeletal abnormalities, mental retardation, chest deformities and scoliosis that lead to pulmonary dysfunction. Anesthesia plan in these patient needs to consider mentioned anomalies. For airway management, laryngeal mask airway (LMA) is preferred in order to avoid the use of muscle relaxants. The use of short-acting anesthetics can accelerate recovery from anesthesia. To reduce postoperative pain, regional techniques are preferred. We report a 6 month-old boy with PBS, including airway management and anesthesia during surgery.

### Keywords

- Prune belly syndrome
- General anesthesia
- Regional anesthesia

## Introduction

Prune belly syndrome (PBS) is a rare genetic syndrome of unknown etiology. It consists of three symptoms: anterior abdominal muscle deficiency, cryptorchidism and genitourinary malformation.<sup>1</sup> These patients have cardiovascular and musculoskeletal abnormalities, mental retardation, chest deformities, and scoliosis that lead to lung hypoplasia and pulmonary dysfunction.<sup>2,3</sup> Incidence is 1:40,000 in live births and males are more commonly affected. Several surgical interventions may be required due to the abnormalities present in Prune belly syndrome.<sup>4</sup> Usually, patients with genetic syndromes have difficult airways, so it is an essential issue during anesthesia and needs special attention between pediatric anesthesiologists. Several Prune belly syndrome patients have cardiac abnormalities. In the presence of an abnormal cardiac examination additional diagnostic studies such as echocardiography is necessary.<sup>5</sup> Postoperative pain can be a major issue. Opioids can cause respiratory depression which can be problematic due to deficient abdominal muscles.

Regional blocks offer analgesia without having a depressant effect on respiration.<sup>4</sup>

## Case Report

A 6-month-old boy, weighing 6 kg was referred to our institute for orchiopexy of bilateral undescended testes and he was a known case of the prune-belly syndrome **Figure 1**. In primary evaluation, he was completely alert and oriented with no history of neurological diseases or seizures. In the evaluation of the airway, he had limited mouth opening and micrognathia. In physical examination, pectus excavatum was evident on the chest wall and abdominal muscles had diastasis **Figure 2**. In a cardiovascular evaluation by echocardiography, he had atrial and ventricular septal defects (ASD and VSD). Laboratory tests were normal. Premedication was performed using 6 mcg of IV fentanyl. Monitoring was carried out using pulse oximetry, ECG, and blood pressure monitoring. Preoxygenation was performed with 100% oxygen for about 3 minutes. Induction was carried out using 6mg of lidocaine and 18mg of propofol. Laryngeal mask airway (LMA) of size 1.5 was used for intubation and proper

placement was confirmed by auscultating the neck. Maintenance of anesthesia was achieved with N<sub>2</sub>O 50%, O<sub>2</sub> 50%, and sevoflurane. The caudal block was also given with 6 ml of 0.2% bupivacaine for

pain relief. He did not receive any muscle relaxants and had spontaneous ventilation. He did not experience any adverse effects during the operation and recovery.



**Figure 1:** Prune-belly syndrome in the reported case



**Figure 2:** Pectus excavatum on the chest wall and abdominal muscles in the reported case

## Discussion

Preparation for the management of anesthesia for these patients should start in the preoperative period. Anesthesiologists need to rule out any renal and pulmonary dysfunctions and cardiac abnormalities.<sup>6</sup> For airway management, LMA is preferred over the endotracheal tube to avoid the use of muscle relaxants

and decrease the chance of airway stimulation. Regional techniques for pain relief such as caudal, epidural and peripheral nerve block and the use of short-acting anesthetic agents such as remifentanyl and sevoflurane can accelerate recovery from anesthesia.<sup>7</sup> Anesthesia plan in patients with the prune-belly syndrome should make provisions for the associated

anomalies of the chest wall, lungs, renal, cardiovascular, central nervous systems and other organs, while avoiding use of muscle relaxants.

### **Conclusion**

Due to musculoskeletal abnormalities, chest deformities, and scoliosis in patients with Prune belly syndrome, it is better to use laryngeal mask airway instead of endotracheal tube for preventing endotracheal stimulation and muscle relaxant administration. In case of intubation, short-acting anesthetics such as remifentanyl and sevoflurane are of great acceptance.

### **Ethical Considerations**

This study was approved by Institutional Ethics Committee of Shahid Beheshti University of Medical Sciences and the approval ID is IR.SBMU.MSP.REC.1399.170.

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### **Conflict of interests**

There are no conflicts of interest.

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