

## Case report of prune belly syndrome: An airway challenge too?

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### ABSTRACT

Prune belly syndrome (PBS) is a rare congenital disease presenting a characteristic triad of abdominal muscle deficiency, urinary tract abnormality, and cryptorchidism with associated anomalies of various systems. We report the anesthetic management of an infant with PBS with no obvious facial deformity in whom we encountered an unanticipated difficult airway. Multiple attempts to secure the airway with different supraglottic airway devices were unsuccessful and the trachea could be intubated only on the second attempt. The patient had deficient abdominal muscle and thus a prolonged duration of action of non-depolarizing muscle relaxant.

**Key words:** Block, Difficult airway, Muscle relaxant, Prune belly syndrome

**P**runo belly syndrome (PBS) is a rare congenital disease with an incidence of 1:40000 live births [1,2]. It presents with a characteristic triad of abdominal muscle deficiency, urinary tract abnormality, and cryptorchidism with associated anomalies of the cardiorespiratory, skeletal, gastrointestinal, and central nervous system in 75% of the cases [3-5]. The PBS is associated with ineffective coughing and prolonged duration of action of muscle relaxants but a difficult airway.

Since the syndrome has rarely been reported in the literature, we wish to highlight the occurrence of a difficult airway in a patient with PBS that too without any associated facial anomaly.

### CASE REPORT

A 3-month-old and 6 kg male infant, a diagnosed case of PBS, presented with a classical triad of wrinkled abdominal skin due to absent muscles, cryptorchidism, and ureterovesical abnormalities.

The patient presented to the hospital with a lump in the lower abdomen of 5×4 cm in size. On pressing the lump, there was a passage of urine. There was a history of recurrent urinary tract infections due to the retention of urine. The child had missing testicles in the scrotum which was suggestive of cryptorchidism.

The child was posted for the Mitrofanoff procedure with bilateral orchidopexy and ureteroneocystostomy. There were no features suggestive of the involvement of the cardiopulmonary system, no obvious facial deformity, or predictors of the difficult airway. All routine investigations and the 2D echocardiogram were normal (Fig. 1a).

General anesthesia using a supraglottic airway device (SGD) supplemented with a caudal epidural block (CEB) was planned. After obtaining the guardian's consent, the infant was pre-oxygenated and general anesthesia was induced with an injection of fentanyl 12 µg and an injection of propofol 30 mg was administered intravenously. Mask ventilation was optimal but three attempts with weight-appropriate sizes of blockbuster LMA and i-gel<sup>®</sup> were unsuccessful in securing the airway. Endotracheal intubation which we had wanted to avoid was imperative now. Therefore, an injection of succinylcholine 12 mg was administered intravenously and laryngoscopy was performed using a C-Mac<sup>®</sup> video laryngoscope. On laryngoscopy, the epiglottis was very floppy and the larynx was found to be extremely anterior and significantly deviated from the midline. An optimal external laryngeal manipulation maneuver was applied and the trachea was intubated using a 3.5 ID micro cuff endotracheal tube in the second attempt by an experienced pediatric anesthesiologist. Maintenance of anesthesia was done with oxygen in nitrous oxide and sevoflurane (2–3%). Landmarks of CEB could not be appreciated well, hence ultrasound guidance was used and a block was successfully placed, with 6 ml of injection bupivacaine 0.25% plus clonidine 12 µg. A single dose of injection atracurium 2 mg intravenously was given initially under neuromuscular monitoring that lasted for the entire 4 h duration of surgery.

In the Mitrofanoff procedure, the appendix was used to create a conduit between the skin and the urinary bladder for efficient emptying of the bladder through a skin opening near the umbilicus, with the help of a catheter. Testicles were found in the abdomen and orchidopexy was done. Intraoperative vitals remained stable. At the end of the surgery, anesthesia was terminated and when

#### Access this article online

Received - 03 July 2022  
Initial Review - 21 July 2022  
Accepted - 05 September 2022

DOI: 10.32677/ijcr.v8i9.3550

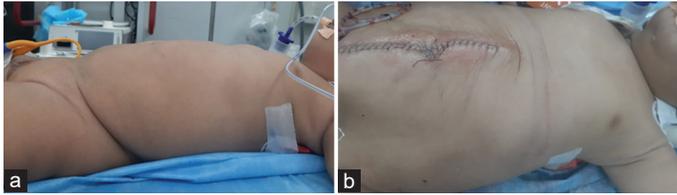
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**Figure 1: (a) Prune belly in the patient and (b) post-operative picture**

the infant was fully awake, the trachea was extubated. Chest physiotherapy and postural drainage were advised for the post-operative period (Fig. 1b).

## DISCUSSION

The term prune-belly donates the wrinkled appearance of the abdominal wall, due to hypoplasia of the abdominal muscles [5]. Furthermore, known as Eagle Barrett syndrome, its mortality may reach 50% before age of 2 years, depending on the type and severity of abnormalities [1,4,6]. As a consequence of urinary tract obstruction, there is oligohydramnios leading to pulmonary hypoplasia and perioperative pulmonary complications [1]. Oligohydramnios can also lead to Potter's faces with micrognathia, flattened nose, and malformed ear [1,6]. Deficiency of abdominal wall muscle and flat diaphragm can cause ineffective cough in these children leading to accumulation of pulmonary secretions and repeated chest infections [7-9].

There is scant literature regarding the association of PBS with a difficult airway and no report yet of the occurrence of unanticipated difficult intubation in a child of PBS with normal facies as was observed in our case. Baris *et al.*, reported a complicated airway in PBS in a child with Potter's facies but they were able to secure the airway with an LMA™ after multiple failed attempts of endotracheal intubation [6]. Lyon AJ reported a case of a neonate with PBS which was associated with laryngeal atresia. In this case, the airway could not be established and the neonate died within a few minutes of birth [10]. The inability to secure the airway with an SGD and an anterior larynx seen on video laryngoscopy in a seemingly normal airway in our patient could be due to anatomical variation as a result of the musculoskeletal involvement in this syndrome.

The presence of lax and deficient abdominal muscles can lead to a prolonged duration of action of non-depolarizing muscle relaxant (NDMR) as was reported by Yoon *et al.* and Hannington-Kiff [3,7]. However, Bösenberg suggested a normal duration of action of muscle relaxant unless the renal compromise is present [1]. We had planned the use of an SGD without the use of NDMR but since the trachea had to be intubated for securing the

airway, NDMR had to be given whose duration was found to be significantly prolonged.

Since it is advisable to restrict the perioperative use of opioids, which can further aggravate ineffective cough postoperatively in these patients [1,2,9], we planned a CEB to augment the general anesthesia. Difficulty in the identification of anatomical landmarks which we experienced may be attributed to vertebral skeletal abnormalities present in 45% cases of PBS [1]. Garg *et al.* have used quadratus lumborum block for regional anesthesia in PBS patients [2]. Ultrasound guidance facilitated the successful placement of the drug in the caudal epidural space in our patient.

## CONCLUSION

Our case highlights the association of a difficult airway in spite of seemingly normal facies in children of PBS. Perils of using NDMR in these patients should be considered and consequently, neuromuscular monitoring is advisable. Vertebral anomalies may preclude landmark identification for regional block; hence, ultrasound guidance is recommended.

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*Funding: Nil; Conflicts of interest: Nil.*

**How to cite this article:** Neha, Singh R, Agrawal G. Case report of prune belly syndrome: An airway challenge too? *Indian J Case Reports*. 2022;8(9):291-292.