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Case Report / Olgu Sunumu

Anaesthetic Management of Laparoscopic Surgery in Prune-Belly Syndrome: Report of Two Cases

Laparoskopik Cerrahi Uygulanan Prune-Belly Sendromunda Anestezi Yaklaşımı: İki Olgu Sunumu

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Abstract / Özet

In prune-belly syndrome, which comprises the congenital triad of abdominal muscle defects, urinary tract anomalies and bilateral cryptorchidism, several surgical interventions may be needed due to these anomalies. Complications of pulmonary hypoplasia and end stage renal disease are the major prognostic factors. Pre-anaesthetic, anaesthetic and post-anaesthetic monitoring should be carefully performed in these patients. When planning the anaesthetic technique, anomalies of the renal or cardio-pulmonary system must be considered. In this case report we present the use of the Proseal LMA for anaesthetic management of two patients with prune-belly syndrome who underwent laparoscopic surgery.

Key Words: Prune-Belly syndrome, Laryngeal Mask Airway, laparoscopic surgery

Prune-Belly sendromu, karın kaslarının yokluğu, üriner sistem anormallikleri ve çift taraflı inmemiş testis üçlemesinden oluşur. Mevcut anormallikler nedeni ile cerrahi işlemlere ihtiyaç duyulabilir. Ana prognostik faktörler pulmoner hipopolazi ve son dönem böbrek yetersizliğine bağlı komplikasyonlardır. Bu hastalarda anestezi öncesi, sırasında ve sonrasında takip dikkatle yapılmalıdır. Anestezi tekniği planlanırken böbrek ve kardiyo pulmoner sistem anormallikleri göz önünde tutulmalıdır. Biz bu yazıda, laparoskopik cerrahi uygulanan Prune Belly sendromlu hastaların anestezi uygulaması için prosil LMA kullandığımız deneyimlerimizi sunduk.

Anahtar Kelimeler: Prune Belly sendromu, Laringeal Mask Airway, laparoskopik cerrahi

Introduction

Prune-belly syndrome is a congenital disorder that is characterized by the wrinkled appearance of the abdomen. This syndrome consists of the triad of congenital urinary tract anomalies, defects in the abdominal wall musculature and bilateral undescended testicles. The syndrome may be accompanied by cardiovascular, skeletal, gastrointestinal, respiratory and central nervous system anomalies. Several surgical interventions may be required due to the abnormalities present in prune-belly syndrome. Medical and surgical issues may be encountered by these patients due to anatomical defects (1). In this case report we present the use of Proseal LMA for anaesthetic management of patients with prune-belly syndrome who underwent laparoscopic surgery.

Case Reports

Case 1

A 5-year-old, 20 kg boy with prune-belly syndrome was admitted to the Department of Paediatric Surgery for placement of a peritoneal dialysis catheter. The patient had a past medical history of mild aortic stenosis and chronic renal failure. On physical examination, the typical prune-belly appearance of the abdomen and face was observed (Figures 1, 2). The patient's laboratory values showed creatinine 1.9 mg dL^{-1} and urea 75 mg dL^{-1}. Other laboratory values were within the reference ranges. The patient was classified as ASA III by the American Society of Anesthesiologists' (ASA) score system and informed consent was obtained from the parents. The patient was admitted to the operating room without premedication, and three-lead ECG, peripheral O_2 saturation (SpO_2) and non-invasive blood pressure monitoring were applied. Preoperatively the patient's heart rate was 120 beats min^{-1}, blood pressure (BP) was 90/60 mmHg and SpO_2 was 97%. Anaesthesiology was induced with 20 µg fentanyl and 50 mg propofol. Neuromuscular blockers were not used. A size 2, Proseal LMA was inserted. A mixture of 50% O_2, 50% medical air and 2% sevoflurane was used for maintenance of anaesthesia. Perioperative fluid replacement consisted of a 1/3 mixture of isotonic-5% dextrose. Laparoscopic insertion of a peritoneal dialysis catheter was completed in 55 minutes, and the patient was stable during surgery. Spontaneous breathing and respiratory reflexes were restored after termination of anaesthesia, and the Proseal LMA was removed with no complications. The patient was sent to the Paediatric Surgery Clinic after 1 hour of postoperative monitoring.

Case 2

A 3-year-old, 14 kg boy was admitted for surgery for undescended testicles. The patient had been diagnosed as having prune-belly syndrome when he was 2 months old and was operated for undescended testicles at the age of 1. On physical examination, the typical

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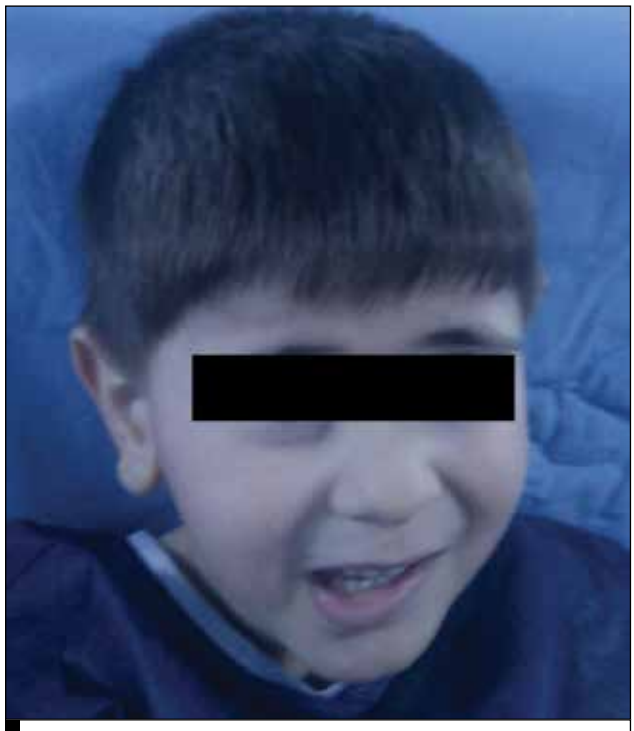

Figure 1. Typical face appearance of the patient

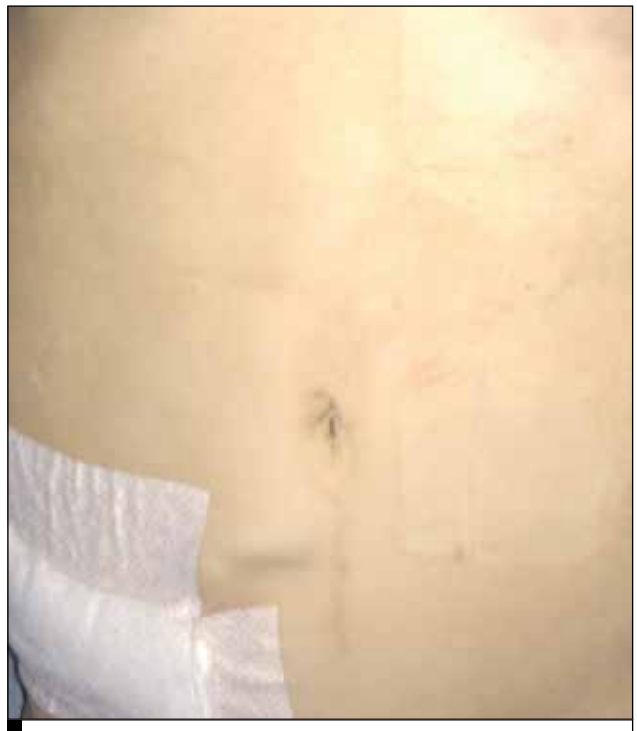

Figure 2. Typical appearance of abdomen in the patient

prune-belly appearance of the abdomen and face was observed. The patient's laboratory values were within the normal ranges. The patient was classified as ASA I by the American Society of Anesthesiologists' score system and informed consent was obtained from his parents. For premedication 0.5 mg kg^{-1} oral midazolam was given. The patient was taken to the operating room, and three-lead ECG, SpO_2 and non-invasive blood pressure monitoring were performed. The preoperative heart rate of the patient was 110 beats min^{-1}, BP was 80/50 mmHg and SpO_2 was 98%. Anaesthesia was induced with iv 15 µg fentanyl and 40 mg propofol. A size 1.5, Proseal LMA was inserted. Caudal block was performed by injecting 0.25% levobupivacaine at the dose of 1 mL kg^{-1} using a 20 G needle. Anaesthesia was maintained with 40% O_2, 60% medical air and 1% sevoflurane. A balanced crystalloid solution with dextrose (Isolite-P) was given for fluid replacement. Laparoscopy for undescended testicles was completed in 45 minutes; the patient was stable during the procedure. Spontaneous breathing and respiratory reflexes were restored, and the Proseal LMA was removed with no complications. The patient was sent to the paediatric surgery clinic after 1 hour of postoperative monitoring.

Discussion

Developmental delay and growth retardation are commonly seen in children with prune-belly syndrome. The incidence of this syndrome is approximately 1:40000 live births. Ninety-five percent of patients are male. Complications of pulmonary hypoplasia and end stage renal disease are the major prognostic factors, resulting in a mortality rate of 60% (2). Patients with prune-belly syndrome may develop renal failure and require insertion of a peritoneal dialysis catheter as a result of congenital renal dysplasia, hypoplasia, pyelonephritis or post-obstructive progressive injury. Recently, laparoscopic insertion of peritoneal dialysis catheters has been shown to be beneficial. Along with renal problems, pulmonary function is often compromised in prune-belly patients. Pulmonary dysfunction is mostly seen as restrictive lung disease as a result of pulmonary hypoplasia secondary to scoliosis and chest wall deformities (3). Prune-belly syndrome may be accompanied by congenital heart defects in some patients. In this case preoperative prophylaxis of infective endocarditis should be considered (4). One of the patients had aortic stenosis and received prophylactic therapy.

It is important to understand the pathophysiology of this rare congenital disease to allow successful anaesthetic management. Pre-anaesthetic, anaesthetic and post-anaesthetic monitoring should be carefully performed. When planning the anaesthetic technique, abnormalities of the renal or cardio-pulmonary system should be considered (1). In the case of impaired renal function, preoperative and postoperative fluid management should be performed carefully, and potential problems regarding the elimination of anaesthetics should be taken into consideration (4).

Although tracheal intubation has been recommended to facilitate ventilation and prevent aspiration in laparoscopic procedures, some prospective and retrospective studies have reported that classic LMA could be a good alternative. Proseal LMA is a new airway device, which unlike classic LMA has a drainage tube that facilitates passage of a gastric tube. When placed correctly, it provides protection against regurgitation and prevents gastric insufflation (5). Sinha et al. (6) suggested that Proseal LMA could be used for short, elective paediatric laparoscopic procedures. Our two prune-belly patients underwent laparoscopic procedures. We preferred the use of Proseal LMA given the possibility of difficult intubation and because this provided us with the opportunity to avoid neuromuscular blockers in case of respiratory difficulties. It has previously been reported that LMA was successfully applied in a case of difficult intubation with prune-belly syndrome. However, although airway control through LMA was adequate in these patients, alternative methods for intubation should be kept at the ready as well (7).

The defects in the abdominal wall musculature and flat diaphragm cause inadequate elimination of secretions by limiting effective

coughing. Furthermore, the depressant effects of general anaesthetics, the abnormal structure of the abdominal musculature and possible pulmonary anomalies increase the risk of infections, postoperative respiratory distress and atelectasis (4). No postoperative complications were encountered in either of our patients.

Conclusion

We suggest that the Proseal LMA can be used safely for laparoscopic surgery in patients with prune-belly syndrome.

Conflict of Interest
No conflict of interest was declared by the authors.

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Yazarlar herhangi bir çıkar çatışması bildirmemişlerdir.

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