Anaesthetic Management of Laparoscopic Morgagni Hernia Repair in a Patient with Coexisting Down Syndrome, Patent Foramen Ovale and Pectus Carinatum

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Introduction

Hereditary diaphragmatic hernias are congenital anomalies that result from inadequacies in the closure of pleura-peritoneal membranes. Morgagni hernia (MH) is the rarest defect of the diaphragm among the inherited diaphragmatic hernias (1, 2). Unless complicated, it can remain asymptomatic until adolescence; however, the general approach is to apply surgical treatment when it is diagnosed (3). Despite its advantages compared to open surgical techniques, the safety of laparoscopic surgery in children with inherited heart disease has been controversial. In addition to complications such as pneumothorax and pneumomediastinum that might occur during pneumoperitoneum in laparoscopic surgery, paradoxical air embolism (PAE) makes the anaesthesia special in the presence of patent foramen ovale (PFO). In this case, anaesthetic management in a case of laparoscopic hernia repair in a 13-month patient who was incidentally diagnosed with MH accompanied by Down’s syndrome, PFO and pectus carinatus is presented.

Case Presentation

A 8.5 kg, 13-month-old female patient with a diagnosis of Down's syndrome was evaluated preoperatively to apply general anaesthesia for laparoscopic repair of MH. It was observed from the patient’s history that she had been hospitalized and had received antibiotic treatment twice because of recurrent pulmonary infections and also had been diagnosed with MH from anteroposterior and lateral chest radiographs taken because of the suspicion of bowel loops seen in front of the heart in an echocardiography taken in this period. There were pectus carinatus in the preoperative physical examination, PFO (3 mm) and a shunt flow from left to right in the echocardiography. There were no pathological findings in the preoperative examination of the respiratory system. The results of the biochemistry, blood count and coagulation tests were within normal limits. A polideks 1/3 (3.33% dextrose - 0.3% sodium chloride, Polifarma Pharmaceutical Industry and Trade Inc., Istanbul, Turkey) infusion was started in the patient who was taken to the operating room without premedication. ECG, oxygen saturation (SpO₂), end-tidal CO₂ pressure (EtCO₂), non-invasive blood pressure, body temperature and precordial stethoscope monitoring were performed. After the preoxygenation, propofol 2.5 mg kg⁻¹ and fentanyl 2 µg kg⁻¹ were used in the induction of anaesthesia. After adequate muscle relaxation with rocuronium 0.6 mg kg⁻¹, endotracheal intubation was performed at first attempt without any difficulty encountered with an uncuffed tube with a 3.5-mm inner diameter. In the maintenance of anaesthesia, sevoflurane was used in a 50% O₂+50% air mixture by being titrated according to the haemo-
dynamic parameters. The patient was placed in a 30-degree reverse Trendelenburg position for the laparoscopic repair. 1 L min⁻¹ and a maximum 8 mmHg of CO₂ was sent into the abdomen through a Veres needle and a pneumoperitoneum was created. The ETCO₂ value remained between 30 and 35 mmHg by means of pressure-controlled ventilation during the operation. Clinical signs of oxygen desaturation or low cardiac output were not observed. No additional dose of muscle relaxant was needed during the surgery, which lasted 50 min. At the end of the surgery, after decurarisation with atropine 0.03 mg kg⁻¹ and neostigmine 0.06 mg kg⁻¹, the patient having sufficient spontaneous respiration was extubated. The patient was then taken to the postoperative recovery unit. Contramal 2 mg kg⁻¹ was administered intravenously for the purpose of postoperative analgesia. No complications were observed in the patient who was monitored with ECG, non-invasive blood pressure and a pulse oximeter in case of potential complications in the postoperative period during 24 h. The patient was discharged without any problems 2 days later. The required written consent was received from the parents of the patient so that the patient’s information could be used for academic purposes.

Discussion

Morgagni hernia is a herniation of the abdominal organs into the chest cavity through a retrosternal defect that develops as a result of a junction defect of the transverse septum located on the ventral diaphragm during embryonic development with costal arches on the chest wall. MH is quite rare and only constitutes 1%–5% of all diaphragmatic hernias (1, 2). Although there is no evidence that MH develops as a result of a genetic defect, it may be accompanied by inherited heart diseases and other genetic anomalies at a rate of 34%–50% (3). Down’s syndrome with MH association was reported to be 20%–30% in a large series, and it was suggested that muscle hypotonia plays a role in this relationship (3-5). The clinical presentation of MH is variable in the paediatric age group; just as it may remain asymptomatic for many years, usually until it is found incidentally during examinations; for instance, it can be diagnosed in the emergency department due to incarceration or strangulation in large defects. Al-Salem et al. (5) stated that the most common complaint in a group consisting of 53 infants and children was recurrent pulmonary infections. Diagnosis can be made in most patients because of the bowel loops or air-fluid levels seen in the thoracic cavity in thoracic radiological examination. Down’s syndrome, pectus carinatus and PFO accompanied MH in our patient, and the diagnosis of MH was made by chance in anteroposterior and lateral chest radiographs that were taken when the bowel loops were seen in front of the heart in an echocardiography.

As Down’s syndrome is the most common abnormality in medical practice and affects many organ systems, anaesthesia management requires particular consideration (6). Detailed assessment should be made in terms of the preoperative airway and respiratory and circulation systems. Because of the short neck, large tongue and tonsils, a difficult intubation should be considered in these patients. Because of subglottic stenosis, intubation should be applied with a tube smaller than the required size for the patient’s age, and over-extension and flexion of the neck should be avoided during laryngoscopy, in consideration of the possibility of atlantoaxial instability that can be seen in 20% of patients (6). In our case, the patient’s head was kept in the neutral position during laryngoscopy, and intubation tubes in different sizes and difficult airway tools were prepared.

The surgical approach in MH repair can be performed conventionally as a transabdominal or transthoracic approach or by laparoscopy. For MH repair in infant and child patients, the laparoscopic approach is preferable due to its advantages, such as less surgical trauma and postoperative pain, recovery of postoperative bowel functions in a shorter time, provision of early postoperative mobilization and a shorter duration of hospital stay (7). However, cardiorespiratory changes that occur depending on the pneumoperitoneum and the position created during laparoscopic surgery as well as issues around the absorbed CO₂ may cause complications in the anaesthetic applications in patients with hereditary heart disease. An increase in intra-abdominal pressure (IAP) causes an increase in systemic vascular resistance, a decrease in venous return and cardiac output. It was shown in a study performed with transoesophageal echocardiography during laparoscopic surgery in healthy children that when the IAP was 6 mmHg, the cardiovascular effects were minimal, but when it was 12 mmHg, the cardiac index decreased at a rate of 13% (8). In the presence of PFO, because the decrease in the heart index may cause desaturation and paradoxical air embolism (PAE) as a result of the reduction in shunt current, it is recommended that IAP should be kept under 12 mmHg in children with cardiac disease (9). In the present case, the patient was evaluated by the surgical team prior to the operation in terms of possible PAE that could occur during the laparoscopic repair, and IAP was limited to 8 mmHg to prevent the reversal of the shunt.

The increase of right atrial pressure or any increase in pulmonary artery through mechanical ventilation during laparoscopic procedures facilitates the direction of the air in the right heart towards the arterial circulation by changing the shunt flow from right to left through the foramen ovale. However, despite a high prevalence rate of PFO, around 20%–25% in healthy people, less frequently observation of PAE and cerebral complications can be explained by the high resolution of CO₂ or adhesion of platelets to the bubbles (10). Although transoesophageal echocardiography is the most sensitive and original monitor in the diagnosis of PAE, its implementation is not always possible depending on the conditions in routine operating rooms. A careful clinical follow-up and routine monitoring were applied in our patient and all the air bubbles
in the intravenous lines were removed and the use of nitrous oxide was avoided in order to prevent the possibility of PAE.

Because children have a more flexible abdominal wall and faster absorption of CO₂ from the peritoneum than adults, hypercapnia formation in children is more apparent than in adults (9). The reason for the greater carbon dioxide absorption is the short distance between the capillary and the peritoneum as well as the increased absorption area of the peritoneum relatively compared to the body weight. Hypercapnia may cause an increased sympathetic response, an increase in heart rate and blood pressure and ventricular arrhythmias (9). Hypercapnia can be overcome by increasing the minute ventilation and sometimes with postoperative ventilatory support. In our patient, normocarbia was provided, with pressure-controlled ventilation maintaining an EtCO₂ of 30–35 mmHg.

Subcutaneous emphysema, pneumothorax, pneumomediastinum and pneumopericardium may develop as a result of a diffusion of CO₂ into tissue spaces through the peritoneal, pericardial and pleural cavities with an effect of intraperitoneal pressure during the dissection and manipulation of the adhesions between the peritoneal sac and viscera, or when fallopian ligament damage occurs with the Veres needle. These complications should be considered intraoperative in the presence of crepitation in sternal notch, hypercarbia, hypoxia and increased airway pressure (11). To be able to instantly realize the potential complications that may occur in our patient, in addition to a careful clinical observation in the perioperative period, we applied EtCO₂ monitoring, airway pressure and pulse oximetry monitoring and chest auscultation with a precordial stethoscope. At the same time, at the end of the operation, extubation was realized after eliminating the possibility of pneumothorax or pneumomediastinum through taking a chest X-ray. The patient was discharged without any problem 2 days after the operation.

**Conclusion**

In the presence of hereditary abnormalities and heart disease accompanying MH in paediatric patients, laparoscopic surgery can safely be applied with low intra-abdominal pressures with a careful clinical observation and haemodynamic monitoring.

**Informed Consent:** Written informed consent was obtained from parents of the patient who participated in this case.

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