Thoracic Epidural for Nissen Fundoplication in a Child with Familial Dysautonomia

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Familial dysautonomia or Riley-Day syndrome is an autosomal recessive disorder limited mainly to children of Ashkenazi (East European) Jewish origin. The genetic defect, linked to chromosome 9, affects nervous system development and manifests as autonomic and sensory nervous dysfunction. Features include blood pressure lability, swallowing difficulties with excess salivation, gastric reflux, and impaired temperature control. Peripheral pain sensation is diminished while visceral pain sensation is usually intact.

Fifty percent of FD patients experience episodes of acute deterioration called "dysautonomic crises," which present as tachycardia, high blood pressure, sweating and vomiting. Precipitating factors include emotional and physical stress, pain and dehydration. Usually crises do not appear until age 3 years. The management of anesthesia in patients with FD poses a major challenge because many of these children suffer from lung disease, due to recurrent aspiration pneumonia, hypotonia, and diminished response to CO_2. The importance of pre- and postoperative respiratory physiotherapy in FD patients has been emphasized in the literature [1]. This is especially true for abdominal surgery, which is complicated by the impaired ability of patients to cough because of pain, and the occasional need for prolonged mechanical ventilation.

We describe the use of combined general and thoracic epidural analgesia in a patient with FD undergoing Nissen fundoplication.

FD = familial dysautonomia

Patient Description

A 14 month old girl weighing 9 kg was admitted for laparoscopic Nissen fundoplication and gastrostomy. This patient, born full-term, suffered from hypotonia and hyporeflexia. Before a diagnosis of FD was established at age 4 months, she had been hospitalized several times for recurrent aspiration pneumonia. Her last hospitalization was 4 weeks prior to the current admission. She had never experienced a dysautonomic crisis. Concurrent medications included cephalexin, ranitidine, inhaled budesonide, and eye drops.

Upon admission, the patient was tachypneic, with oxygen saturation (SpO_2) of 85–90% in room air. Chest X-ray revealed bilateral pulmonary infiltrates. Arterial blood gas analysis and pulmonary function tests were not performed. During the preoperative period the patient continued to receive inhaled budesonide, and intensive respiratory physiotherapy was begun. SpO_2 rose to 100% in room air, and there was partial improvement in the infiltrates on chest X-ray. The night before the procedure she was hydrated intravenously with crystalloids 5 ml/kg/hour. In the morning, she was premedicated with midazolam 4 mg per os. Following placement of routine monitors and pre-oxygenation with 100% oxygen, anesthesia was induced i.v. with ketamine 20 mg. Endotracheal intubation with the Sellick maneuver was facilitated with i.v. atracurium 4 mg, and general anesthesia was maintained with isoflurane 0.5–1.0% in 50% oxygen in air. Low dose dopamine infusion (1 µg/kg/min) was begun and continued until the end of the surgery. A radial arterial line and a urinary catheter were inserted for intraoperative monitoring. After endotracheal intubation and before beginning the procedure the patient was placed in the left lateral position. A median approach at the T11-12 epidural interspace was used. The correct positioning of the 18G Tuohy needle was ascertained by the loss of resistance method using a saline-filled syringe (Epidural Minipack Paediatric, Portex®, Hythe, UK). A 21G epidural catheter was then inserted at this level, and after a negative aspiration test, 0.2% ropivacaine 4 ml (8 mg) was slowly injected. Cardiovascular parameters remained stable, with systolic blood pressure 100–125 mmHg, diastolic 40–50 mmHg, and heart rate 100–115 beats per minute. Thereafter, infusion of 0.2% ropivacaine 0.1 ml/kg/hr was given, and laparoscopic surgery was started. The pneumoperitoneum and head-up position were hemodynamically well tolerated. Eighty minutes after beginning the operation, the surgeon decided to convert to open fundoplication because of technical difficulties. This part of the procedure was characterized by greater hemodynamic changes with lowering of systolic blood pressure until 81 mmHg, and diastolic 36 mmHg. Heart rate remained stable at approximately 130 beats/min. The periodic lowering of blood pressure was not caused by hemorrhage and was not correctable by fluid bolus. Titration of the dopamine infusion rate (from 1 to 4 µg/kg/min) resulted in normalization of the blood pressure. The patient did not experience ventilation difficulties, and SpO_2 remained 100% in 50% oxygen. Arterial blood gas and electrolytes remained within normal limits.

After completion of surgery, the tracheal tube was removed, and the patient was taken to the intensive care unit in stable

hemodynamic condition, fully conscious and free of pain. No respiratory distress or hemodynamic instability was reported during her stay in the ICU. Next day, the patient was transferred to the surgical ward. The epidural infusion of 0.2% ropivacaine 0.1 ml/kg/hr was maintained for 3 days. The postoperative period was characterized by excellent analgesia, permitting effective respiratory physiotherapy. She was discharged home on postoperative day 6 in good condition.

Comment

Familial dysautonomia is a multisystem disease, and the cardiovascular and pulmonary systems are of particular concern to the anesthesiologist. Patients with FD are unable to respond to hypovolemia and myocardial depressants owing to reduced endogenous catecholamine release, leading to hypotension intraoperatively. Stenquist and Sigurdsson [2] reported the successful control of intraoperative hypotension with inotropic support. In our patient, low dose dopamine infusion was used as a precautionary measure, based on their case report.

Several anesthesia techniques have been proposed to achieve better cardiovascular stability in FD patients. Moderate dose fentanyl anesthesia was described by Beilin et al. [3], but all patients required mechanical ventilation postoperatively for 4–14 hours. In FD patients with preexisting lung disease, such prolonged postoperative mechanical ventilation can aggravate pulmonary status and lead to further respiratory complications. It was claimed that cardiovascular instability made epidural anesthesia inadvisable. We assumed, however, that epidural anesthesia would involve less cardiovascular risk in our complex patient. First, it is well known that central neuroaxial block in patients under the age of 3 years is not associated with hypotension, presumably because of the less dominant role of the sympathetic nervous system. Additionally, preoperative fluid loading is usually unnecessary in healthy young children before regional anesthesia, due to the small blood volume in the lower part of the body. More than half the patients with FD who are scheduled for Nissen fundoplication have severe lung disease with significant X-ray findings.

Pulmonary problems include bronchial secretions, atelectasis, bronchiectasis, aspiration pneumonia, and need for prolonged postoperative mechanical ventilation. Nissen fundoplication has been shown to improve the nutrition and respiratory status of patients with FD, and is a common procedure in this population. Axelrod and co-workers [1] concluded that in the perioperative period, special attention must be paid to the intravascular volume of FD patients in order to achieve hemodynamic stability. They emphasized the frequent requirement for prolonged mechanical ventilation following abdominal surgery. Wilson et al. [4] in a comparison of opioid infusion and epidural analgesia in non-FD children with lung disease noted fewer major respiratory complications and postoperative pneumonia when epidural analgesia was used, and suggested that epidural analgesia might be associated with improved outcome in children with lung disease. The suggested inadvisability of epidural analgesia in FD patients was challenged by Challands and Facer [5], who successfully administered lumbar epidural analgesia for Nissen fundoplication in FD patients. The main indication for epidural analgesia was optimal intra- and postoperative pain control. They presumed that good pain management would lead to reduced catecholamine levels, which lessens the incidence of crisis and provides cardiovascular stability [5]. Based on the high incidence of preoperative and postoperative lung morbidity in FD patients, we postulated that diminishing pulmonary complications in this population is also an important goal for the use of epidural analgesia. Of note, only one of the three children described by Challands and Facer [5] suffered from lung disease at the time of operation. Our patient was known to have recurrent aspiration pneumonia and presented for surgery shortly after exacerbation of her lung disease. The indication for thoracic epidural analgesia was to provide good analgesia in order to permit early extubation, intensive respiratory physiotherapy, and consequently diminish pulmonary morbidity. The child was extubated and free of pain at the end of the procedure. Thoracic epidural analgesia permitted us to use a lower dose of local anesthetic as compared to the cases reported by Challands and Facer. We intentionally avoided the use of opioid adjuncts. The child was discharged home on postoperative day 6, sooner than the patients with lumbar epidural analgesia described by Challands and co-author.

To the best of our knowledge, this is the first use of thoracic epidural analgesia in a patient with FD in Israel. In conclusion, low thoracic epidural analgesia may be advisable for selected FD patients with preexisting lung disease undergoing upper abdominal surgery. It provides cardiovascular stability and excellent pain relief, permits early extubation and intensive respiratory physiotherapy, thereby preventing postoperative mechanical ventilation and other respiratory complications.

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ICU = intensive care unit