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EDİTÖRE MEKTUP / LETTER TO THE EDITOR

Anesthesia management in a patient with Dandy-Walker Syndrome and Fontan physiology

Dandy-Walker Sendromu ve Fontan fizyolojili bir hastada anestezi yönetimi

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To the Editor,

Dandy-Walker Syndrome (DWS) was first defined in 1914. The classical triad of DWS is hypoplasia or agenesis of the cerebellar vermis, dilatation of the fourth ventricle, and hydrocephalus. The incidence of the syndrome is 1:25,000–30,000 of live births. Patients with DWS have different malformations of the heart, face and gastrointestinal or genitourinary system[1], and unfortunately only around 20 to 23% of these patients could reach adult life[2]. Fontan procedure first described in 1971[3]. The method that diverted all essential venous blood into the aspiratory courses, short of the intercession of a ventricle, by way of a careful relief of tricuspid atresia. Here, we presented anesthetic management on a patient with Dandy-Walker syndrome and Fontan physiology

A six years old, 18 kg patient was accepted to provide oral hygiene. She was born with DWS. When she was 1 year old, she underwent ventricular-peritoneal shunt surgery for hydrocephalus and diagnosed with double-entry left ventricle + restrictive ventricular septal defect + secundum atrial septal defect all at once. Then, when she was 18 months of age, Glenn procedure + pulmonary banding were applied. Her cleft palate lip surgery was performed 5 months after that. She underwent a Fontan operation as she was 5 years old but unfortunately, she had intracranial hemorrhage two times before the operation. In the pre-anesthetic investigation, she was aware and afebrile. Her rhythm was 102 beats per minute and regular, blood pressure was 120/60 mm Hg, SpO_2 82. Cleft lip operation and median sternotomy scars were present, she had slight hydrocephaly and left hemiplegic. Cardiovascular system investigation indicated a regular rhythm, a grade 3^o systolic murmur on the second intercostal space. She had hepatomegaly, the remainder of the systemic investigation was unremarkable. Preoperative laboratory explorations were unexceptional. ECG displayed normal sinus rhythm. Acetylsalicylic acid was changed to low-molecular-weight heparin before operation. When INR<1.5 she was operated on. Infective endocarditis prophylaxis was given. It was applied to her regular monutarization in operation room. Then, the patient was pre oxygenated for 3 minutes. SpO_2 is reach 92 and decreased to less than 90 during the entire anesthetic period. Anesthesia was induced by i.v propofol 1.5 mg/kg with lidocaine 1mg/kg and 0.6 mg /kg rocuronium and then continued with sevoflurane 0.8-1.2 % in 50% nitrous oxide/oxygen mixture. Intubation was done without complications and easily by a 4.5 size uncuffed endotracheal tube. An invasive pressure line was introduced after anesthesia. She was ventilated with tidal volumes of 7ml/kg and breathing frequency of 12 to 14. The $ETCO_2$ was maintained within the range of 30-35mmHg. Her blood pressure and rhythm were constant over the surgical procedure. The procedure lasted 45 minutes. 10 mg/kg i.v paracetamol, i.v ondansetron 0.1 mg/kg postoperative were given. Neostigmine 0.03mg/kg and 0.01 mg/kg atropine were given at the finish of the surgery, and extubation was performed successfully. She was observed in the pediatric critical

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care for 12 uneventful hours after the operation, then she was shifted to the ward. She was discharged from the hospital in the following day.

The management of patients with DWS is already challenging for anesthesiologists. The intertwining of this by Fontan's procedure more challenging for anesthesiologists and requires a lot more effort. First of all, airway management may be difficult due to hydrocephaly in DWS patients but mask ventilation and intubation were done without any complications. Cardiac anomalies can range from ventricular septal defect to pulmonary stenosis or complex cardiac anomaly in DWS, as in our patient[4]. And detailed physical examination, especially cardiovascular, baseline status should always be investigated even before a minor surgery and necessary measures should be taken for either the DWS patient or solely the Fontan's procedure patient[5]. Myocardial dysfunction, heart failure, and atrial arrhythmias develop in about 45%- 70% of Fontan's physiology patient[6]. Invasive cardiac monetarization was not applied as it is not mandatory for non-cardiac surgery[7]. Since the blood circulation is slow, arrhythmias and hypercoagulability increase the risk of thromboembolism in Fontan's physiology patient[6]. These patients are predisposed to both thromboembolism and hemorrhage, and so are given antithrombotic therapy. When compared to these patients, she was more complicated because of the past cerebra vascular event and epilepsy caused by DWS. For patients receiving warfarin in combination with antiepileptic drugs (As in this patient), INR should be closely monitored. And the anesthesiologist should know the potential interactions of antiepileptic drugs with anesthesic drugs for the management of patients with epilepsy[8].

There are several reasons for the reduction of venous return such as hypoxia, hypothermia, hypercarbia, acidosis, positive pressure ventilation, PEEP, and pulmonary vascular resistance; which increase sympathetic activity and may threaten life in patients with Fontan physiology[9]. Blood gases CO_2 within the range of 35-45 mmHg and SpO_2 up to 80mmhg were maintained. Since this situation affects the increase in intracranial pressure, it is very important for patients with DWS too. A disruption of the patient's blood gases during the surgery time was not seen. Sedation may cause a rise in pulmonary vascular resistance via hypoventilation and hypercarbia[9]. General anesthesia in patients with Fontan physiology and pressure-controlled ventilation applications increases intrathoracic pressure but decreases pulmonary blood current. In this respect, she did not have any difficulties, and no complications developed. She was extubated in the operation room, then transferred to pediatric intensive care. For DWS in addition to all the above-discussed factors, the degree of hydrocephalus, whether the shunt is working, presence of additional anomalies should be known and situations that increase intracranial pressure should be avoided.

In conclusion, normovolemia should be sustained, and hypercarbia, hypoxia, and acidosis in either Dandy Walker syndrome or Fontan circulation should be avoided. Minor surgical procedures can be safely accomplished on a daycare basis.

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