Case Report

Non-Operating Room Anesthesia (NORA) Management And VACTERL Syndrome: A Case Report

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ABSTRACT

Non-Operating Room Anesthesia (NORA) Management And VACTERL Syndrome: A Case Report.

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VACTERL syndrome presents many challenges related to anesthesia management due to multiple congenital malformations that may include vertebral, cardiac, tracheo-esophageal, renal and extremity anomalies. This case report presents the preparation and management of non-operating room anesthesia (NORA) in an 8-year-old patient diagnosed with VACTERL, followed up with cardiac anomaly (large secundum ASD), right renal agenesis, operated tracheoesophageal fistula and esophageal atresia. With successful preoperative preparation and perioperative management, a safe environment was provided in NORA, and the patient was discharged to the ward without complications. Herein, we wanted to emphasize that in NORA procedures, the frequency of which is increasing nowadays, all precautions against airway management and risks should be taken entirely in the preoperative period and that a multimodal, multidisciplinary approach and good communication between teams should be approached, especially in this high-risk patient group.

Abbreviations: Non-operating room anesthesia (NORA), Atrial septel defect (ASD), Esophageal atresia (EA), Tracheoesophageal fistula (TEF), Colonic transposition (CT), Echocardiography (ECHO), Percutaneous angiography (PA), Intravenous (IV), The post-anesthesia care unit (PACU), Total intravenous anesthesia (TIVA)

Keywords: VACTERL syndrome, non-operating room anesthesia, preoperative management, difficult airway, multiple anomalies.

INTRODUCTION

VACTERL syndrome was first defined as VATER syndrome (abbreviation for **V**ertebral anomalies, **A**nal atresia, **T**racheoesophageal fistula with **E**sophageal atresia, **R**adial and **R**enal dysplasia) by Quan and Smith in 1973 and was finalized by Kaufman and Nora in 1975 with the addition of cardiac anomaly (by adding the letter "**C**") and anomaly of the extremity (by adding the letter "**L**"). The affected organs are not random, suggesting a developmental defect originating from the embryonic mesoderm at the stage of blastogenesis (2-4 weeks of gestation)[1].

The diagnosis of VACTERL is based on exclusion criteria and should be considered in the presence of at least three congenital defects and the absence of clinical or laboratory evidence of many other similar conditions. In addition, these children have normal development and intelligence[1].

Vertebral and vascular anomalies are seen in 60-80%, anal atresia 55%, cardiovascular anomalies 40-80%, esophageal atresia (EA) with tracheoesophageal fistula (TEF) 32%, renal anomalies 50-80%, and extremity anomalies 70%[2,3].

Anesthesia and airway management are critical in these children. Because of craniofacial-vertebral deformities accompanying cleft palate or facial microsomia, maintaining the airway becomes a complete struggle for survival[4]. Herein, we aimed to present our NORA preparation and management in a case with a difficult airway and many accompanying anomalies such as VACTERL syndrome.

CASE PRESENTATION

Ethics committee approval and the patient's relatives written and informed consent for the publication of this case report were obtained.

An 8-year-old male patient (18 kg, 128 cm, Body Mass Index (BMI) 11 kg/m2) applied for the repair of a known atrial septal defect (ASD) (Figures 1 and 2). In the pre-procedural evaluation, it was learned that the patient was followed up with VACTERL syndrome. In the antenatal period, right renal agenesis, cystic structures in the left kidney, and ASD were detected, and no anomaly was seen in the amniocentesis. He was born at term with a cesarean section weighing 2600 grams. The patient was operated for EA when he was 1 day old, and gastrostomy was opened on the second day with the percutaneous endoscopic method. He was operated for colonic transposition (CT) and EA at 1 year old and for hypospadias at 4 years old. There was no history of fatigue, palpitations, and syncope at admission. He stated that while eating, swelling developed in the area above the left clavicle, where an attempt was made for CT. His development and intelligence were evaluated to be appropriate for his age. In addition, he was followed with a 10x5 mm cyst and grade 2 hydronephrosis in

the left kidney. Pre-procedural laboratory findings were normal, and echocardiography (ECHO) revealed a large secundum ASD with a left-right shunt.

Figure 1. Patient preparation and initiation of the procedure.

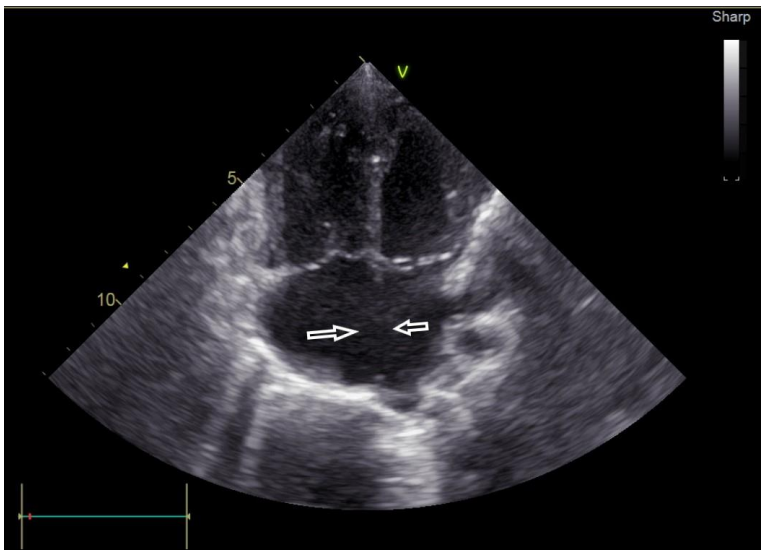

Figure 2. Large secundum ASD on the ECHO.

The pediatric cardiology team decided on percutaneous angiography (PA). No cranio-facial and vertebral anomalies were detected in the preoperative evaluation. Active communication was maintained with the team that would perform the angiography, and it was decided to perform this procedure under deep sedation. A transparent ventilation mask was prepared to monitor the mouth's contents, and different sizes of cuffed intubation tubes, laryngeal mask airways, stylets, bougies, and a video-laryngoscope were ready for this patient group whose airway management is difficult. A double system vacuum device was installed for the patient, who was remembered to have developed neck swelling after food consump-

tion and had an increased risk of aspiration. In this patient with a high risk of regurgitation, the preoperative fasting period was determined as 12 hours, and blood glucose level was monitored. Routine hemodynamic monitoring (electrocardiography, non-invasive blood pressure, pulse oximetry) was applied to the patient who was taken to the operation room. Heart rate was 124/min, SpO_2 was 96%, and non-invasive blood pressure was 110/70 mmHg. Oxygen support at 4 L/min was administered with a nasal oxygen cannula. A 24 G peripheral IV catheter was placed. At the beginning of the procedure, midazolam 2 mg, propofol 20 mg, and ketamine 10 mg IV were administered. The patient's alertness was monitored with the Ramsey Sedation Scale, and a propofol 0.25 mg/kg IV additional dose was administered if the level was 4 or below.

The angiography team performed right femoral vein catheterization, and the procedure was started. A large secundum ASD up to 2 cm and a left-right shunt were detected. It was decided that the ASD could not be closed with percutaneous angiography, and the procedure was terminated (Figures 3 and 4). The process took about 45 minutes. The patient, who was taken to the post-anesthesia care unit (PACU), was transferred to the service after the Richmond Agitation-Sedation scale score was evaluated as 9. The patient, who was stable on the first day after the procedure, was discharged, and it was decided to be evaluated in the council with pediatric cardiovascular surgery for further treatment.

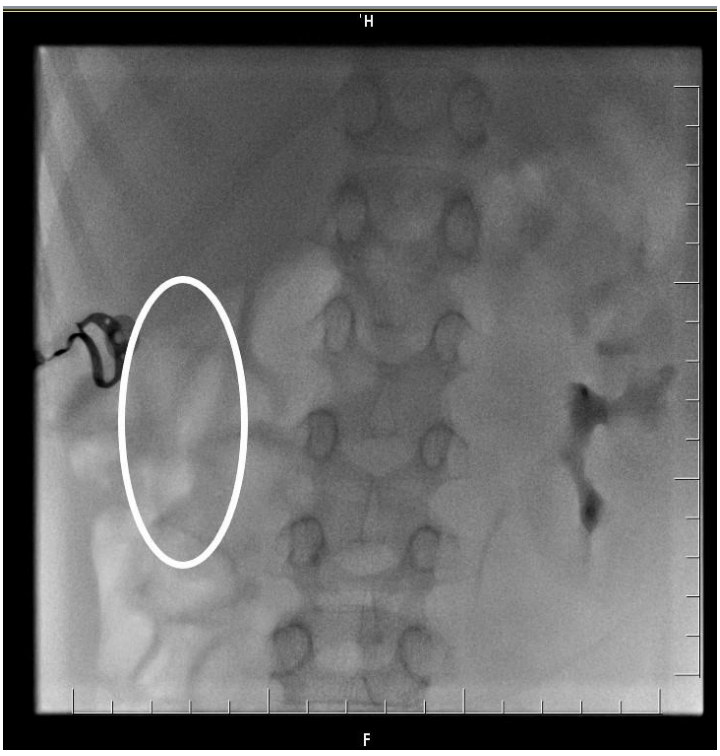

Figure 3. Right renal agenesis on the PA.

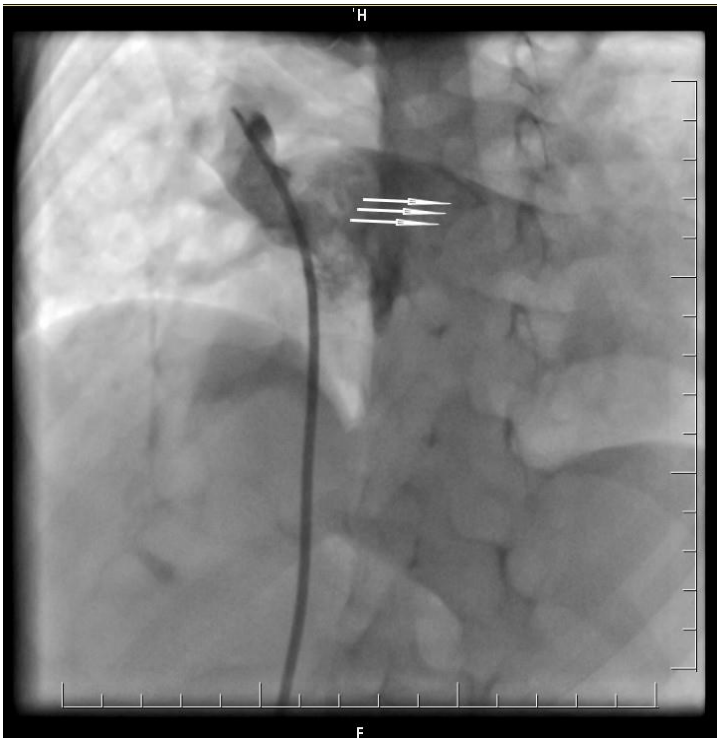

Figure 4 Left-right shunt with secundum ASD on the PA.

DISCUSSION

Although VACTERL syndrome is rare, it is an important phenomenon that requires multimodal follow-up and treatment when encountered. Due to different organ defects, the need for more than one operation and, as a result, the need for repetitive anesthesia arises. This makes these patients at high risk due to difficult airway with existing craniofacial-vertebral anomalies, high aspiration risk, and hemodynamic instability due to cardiac anomalies.

Khatavkar S. et al. emphasized airway management and risks in an 8-year-old patient with VACTERL syndrome who underwent ca-taract surgery under general anesthesia[5]. In our case, because of the high risk of aspiration and regurgitation, we applied preoperative fasting for 12 hours and prepared a transparent ventilation mask and a double-lumen vacuum system. In addition, due to the risk of difficult intubation, we prepared different sizes of cuffed intubation tubes, stylets, bougies, and video laryngoscopes.

In an article by Richards E., it was stated that there was no significant difference between total intravenous anesthesia (TIVA) and volatile anesthesia (including nitrous oxide) in these patients and that regional anesthesia should be decided according to the existing vertebral and extremity anomalies. Also, extensive preoperative preparation is recommended because of the high risk of aspiration[1]. Considering the procedure's type, duration and risks, we preferred deep sedation

as a method of anesthesia and provided induction with multimodal IV anesthetics (midazolam, propofol, ketamine) and used propofol for maintenance.

Herein, we wanted to emphasize that preoperative preparation is as essential as the intraoperative process in these and similar cases and that the preferred anesthesia method should be supported by preoperative laboratory and imaging methods. We think the preoperative fasting period should be decided by considering the existing anomalies, previous surgeries, and the patient's symptoms. Today, the need for NORA is increasing rapidly, and we think that in this and similar procedures, in high-risk patients like ours, preparation for a possible aspiration and intubation need should be complete, and the balance between sedation depth and hemodynamic stability should be closely monitored due to existing cardiac anomalies.

CONCLUSION

NORA procedures, the frequency of which is increasing nowadays, all precautions against airway management and risks should be taken entirely in the preoperative period and that a multimodal, multidisciplinary approach and good communication between teams should be approached, especially in this high-risk patient group.

Additional materials:

No

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UA: conceptualisation, data collection, paper draft, literature review, manuscript preparation and is the lead author. ÇN: paper draft, literature review. All authors approved the manuscript.

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The authors declare that they have no competing interests.

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