Congenital Vascular Causes Of Airway Compression: A Report Of Three Cases

A Thallaj

Abstract

Double aortic arches (DAA) are a group of congenital anomalies of the aortic arch system in which the trachea and the esophagus are completely encircled by a vascular ring causing compression of both structures. Other aortic arch anomalies like aberrant right or left subclavian arteries can form complete or partial vascular rings. We would like to present three cases which were operated at King Khalid University Hospital in Riyadh, Saudia Arabia.

CASE REPORTS

CASE 1

A 44 days old female infant with 2.3 kg body weight was admitted with a history of weak cry and respiratory distress since birth characterized by inspiratory retraction of the ribs and prolonged expiration. Several attempts to introduce a nasogastric tube were unsuccessful as every time child became cyanosed. Initial diagnosis was in favor of tracheoesophageal fistula (TOF). Further workup was done which ruled out TOF.

Investigations included:

1. Barium oesophagography which showed two areas of compression in the oesophagus.

2. Computerized tomography scan (CT) supported the diagnosis of vascular ring due to DAA.

3. Cardiac catheterization revealed DAA with dominant right arch and atretic left arch.

The patient was admitted to the operation room with the trachea intubated and ventilated. Sedation was achieved with midazolam 5 mcg/kg/min.

Intraoperative monitoring included ECG, two pulse-oximeters on both left and right upper limbs, non invasive blood pressure (BP), temperature, urinary output, ETCO2, and intrathoracic pressure. Anesthesia was maintained with O2/air/Isoflurane, atracurium 0.3mg/kg and fentanyl 5mcg/kg body weight.

Through left posterolateral thoracotomy approach, the lung was retracted and the pleura overlying the descending aorta was opened. Carotid pulses were palpated and pulse oximeters were checked after clamps were applied and before arch division. The left aortic arch was divided between vascular clamps and the patent ductus arteriosus (PDA) (not seen during cardiac cath) was ligated.

The surgical procedure took 1.5 hr and surgery as well as anesthesia were uneventful. The patient was sent to the pediatric intensive care unit (PICU) ventilated. The trachea was extubated on the second postoperative day and the respiratory stridor had resolved. The patient was discharged from the PICU on the third postoperative day.

CASE 2

A 5 months old full term male with 5kg body weight infant had a history of cesarean section with subsequent noisy breathing, stridor and feeding difficulties since delivery. Three months later, he developed respiratory distress due to bronchial aspiration. He was referred to our hospital sedated, with intubated trachea and ventilated with a history of failed extubation.

A CT-scan (3D-reconstructional) showed double aortic arch with left dominance and right atresy with PDA (Figures 1, 2).
Congenital Vascular Causes Of Airway Compression: A Report Of Three Cases

Figure 1
Figure 1: DAA forms ring encircling trachea & oesophagus

Preoperatively, the patient was sedated (1mg midazolam-5mcg fentanyl/h) and ventilated on synchronized intermittent mandatory ventilation (SIMV) mode, maintaining satisfactory arterial blood gases (ABG) with normal chest x-ray and laboratory investigations.

Intraoperative monitoring was achieved with 2 pulse-oximeters, invasive BP, ECG, Temperature, ETCO2, respiratory parameter and intrathoracic pressure. Anesthesia was maintained with (O2/air/sevo) and incremental doses of atracurium and fentanyl when required. Surgery was performed through right posterolateral video-assisted thoracoscopy (VATS) with CO2 insufflation at intrathoracic pressure of 8mmHg. The right aortic arch was delineated and non crushing vascular clamps were applied. The carotid pulses were palpated and pulse oximeters and invasive blood pressure were checked after clamping, then four clips were inserted. The right aortic arch was divided distal to the right subclavian artery. The surgery and anesthesia were smooth and uneventful, except for high PaCO2 (70mmHg). Hyperventilation was instituted to reduce the CO2 and gas insufflation was stopped during surgery.

Patient was transferred to the PICU while being ventilated. The trachea was extubated 48 hours later. His respiratory stridor had resolved completely.

CASE 3
A 10 months old full term female infant with 8.8kg body weight was admitted with a history of vomiting and choking that started 6 months ago when solid food was introduced. The patient had also convulsions diagnosed as complex partial seizures treated with Phenobarbitone (4.5mg/kg/day). A barium oesophagography showed an area of compression in the oesophagus. A CT-scan (3D-reconstructional) confirmed the diagnosis of a vascular ring due to an aberrant right subclavian artery (Figures 3, 4).

Figure 2
Figure 2: Ascending, descending & DAA
A VATS division of vascular ring was planned. Induction of anesthesia was achieved with (O2 / sevoflurane); the external jugular vein was cannulated with a 20G iv cannula. Induction of anesthesia was achieved with fentanyl 10mcg, atracurium 3mg i.v, then direct laryngoscopy performed and the right main bronchus was intubated with 4mm endotracheal tube (ETT) and controlled ventilation with pressure control mode (PCV) was started. Anesthesia was maintained with O2/Air/sevoflurane. Intraoperative monitoring was achieved with two pulse oximeters (left and right upper limbs), left radial invasive blood pressure, temperature, ETCO2, and intrathoracic pressure. The patient was positioned in left posterolateral (semi-prone) position. Three ports were inserted and thoracoscopy set was adjusted with a pressure of 4 mmHg and flow was 0.2L/min to create pneumothorax, then the pressure was increased to 8 mmHg and the flow to 1L/min. The aberrant right subclavian artery was arising from the aortic arch distal to the left subclavian artery. Using a non crushing forceps the aberrant artery was clamped for 2 minutes and changes in SPO2 (desaturation) were detected in the right upper limb pulse oximeter. Then, four clips (10 mm) were applied and the artery was divided.
in between. The saturation returned to normal values after 20 minutes. During the procedure, hypercapnia occurred (PaCO2 60mmHg) and was managed by increasing the respiratory rate to 40/min. A chest drain tube was inserted. The procedure took 70 minutes and the patient was extubated on table fully awake and transferred to the PICU. The patient was discharged to the ward 24 hours later.

**DISCUSSION**

The left 4th arch forms the aortic arch, while the right 4th arch usually regresses and forms the proximal portion of the right subclavian artery. DAA is formed when both 4th arches and both dorsal aortas remain present. In about 80% of patients with DAA, the right arch is dominant, when the minor arch is atretic. The atretic segment is almost always distal to left subclavian artery. In approximately 20% of the patients, the left arch is dominant; in these patients the minor right arch typically is patent. DAA usually occurs without associated cardiovascular anomalies, but if present, tetralogy of fallot is the most common associated defect. The anomaly of left aortic arch with aberrant right subclavian artery results from early regression of the right fourth aortic arch between the right carotid and right subclavian arteries. Symptoms of external tracheal obstruction such as wheezing, stridor (biphasic), respiratory distress, aspiration and dysphagia are the most common symptoms (\( \text{\textsuperscript{1}} \)).

Several methods can be employed to diagnose and evaluate the underlying vascular and tracheobronchial anatomy. Barium oesophagography, echocardiography, cardiac catheterization, and bronchoscopy are used to evaluate the functional component of the compression and detect airway abnormalities (\( \text{\textsuperscript{2}} \)). CT scan (3D-reconstruction) has been reported as the best tool to evaluate airway associated with congenital cardiovascular diseases with the advantage of extremely short scanning time (\( \text{\textsuperscript{3}} \)). Magnetic resonance imaging (MRI) also has been used to evaluate the abnormalities related to tracheo-bronchial tree and cardiovascular status. However, MRI studies are too long and need the infant to be sedated for long time (\( \text{\textsuperscript{4,5,6}} \)).

Surgery is indicated in all symptomatic patients, the aim is to divide the vascular ring and relief tracheal and oesophageal compression. Surgical repair is generally performed through left lateral thoracotomy; although, some cases (2nd case report) required right lateral approach. Infants with tracheomalacia may have airway obstruction because of tracheal collapse during induction of anesthesia. Vascular rings may cause tracheal compression similar to anterior mediastinal mass. Careful inhalation induction followed by administration of assisted or controlled ventilation before giving muscle relaxant has been recommended. A rigid bronchoscopy should be available in the OR, because it may be lifesaving in case of tracheal collapse (\( \text{\textsuperscript{7}} \)). The ETT should pass the stenotic area to ensure adequate ventilation. Appropriate i.v and arterial cannulation should be performed and blood should be available before surgery.

In most cases, thoracic surgery is performed by thoracotomy, and anesthesiologist ventilates both lungs with an ordinary ETT. Recent advances in surgical techniques and technology, including high-resolution microchip cameras and endoscopic instruments, have made VATS feasible in infants (\( \text{\textsuperscript{8,9}} \)). Advantages of VATS are; smaller surgical incision, less pain, early postoperative recovery and reduce the possibility of chest wall deformity (long term complication) (\( \text{\textsuperscript{10,11}} \)). VATS can be performed while ventilating both lungs and CO2 insufflation, or by achieving one lung collapsed ventilation (OLCV) which is more desirable (\( \text{\textsuperscript{12}} \)). Techniques that can be used for OLCV in infants are; bronchus intubation using ordinary ETT, bronchial blocker (Fogarty cath), Univent tube or double lumen tube (DLT) (\( \text{\textsuperscript{13}} \)).

**CONCLUSION**

In conclusion, operative division of a vascular ring in pediatric anesthesia is challenging. It requires skills of cardiac as well as thoracic anesthesia, namely one lung isolation techniques.

**References**

Author Information

Ahmed Thallaj
Consultant Anesthesiologist, Department of Anesthesia, King Khalid University Hospital