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Anesthetic Management of a Patient with Associated VACTERL Anomalies and Uncorrected Tetralogy of Fallot Posted for Appendicectomy

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Abstract: ***VACTERL association includes multiple congenital anomalies like vertebral defects, anal atresia, cardiac defects, tracheo-oesophageal fistula, renal anomalies and limb abnormalities. But its association with Fallot's tetrology is rare case. Anesthetic management in such cases becomes challenging. Here we are discussing anesthetic management in a case with VACTERL anomalies and uncorrected Tetrology of Fallot undergoing open appendicectomy in terms of preanaesthetic evaluation, induction and maintenance of anesthesia with postoperative follow up.***

Keywords: VACTERL association, anesthesia, Tetrology of Fallot

1. Introduction

VACTERL association is a sporadic disorder and a positive family history requires careful differential diagnosis with other genetic conditions. Though the exact cause is unknown, it is thought to be multifactorial in etiology, with environmental triggers, including teratogens

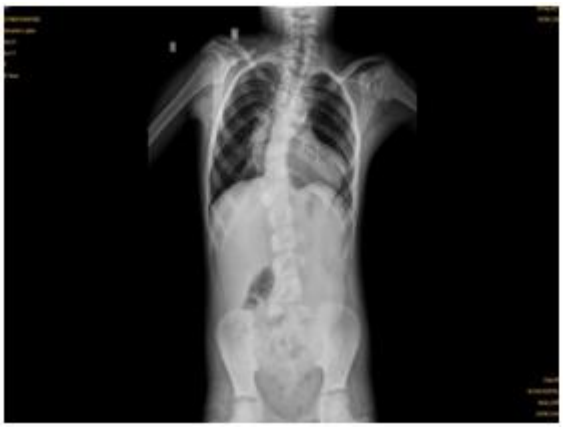
Interacting with a genetically susceptible genome. The cardiac anomalies in the VACTERL association account for 40-60% of cases. The ventricular and atrial septal defects are common cardiac anomalies. The occurrence of TOF is relatively rare and hence is being reported.

VACTERL stands for Vertebral defects, Anal atresia, Cardiac defects, Tracheo-oesophageal fistula, Renal anomalies, and Limb abnormalities. The diagnosis is made clinically when 3 or more congenital defects are present.

Tetralogy of Fallot is the most common cyanotic congenital heart defect, accounting for about 10 % of all congenital heart disease but its association with VACTERL anomalies is rare. It includes ventricular septal defect, right ventricular outflow tract obstruction (pulmonary stenosis), right ventricular hypertrophy, and overriding of aorta. Patients with Tetralogy of fallot undergoing non cardiac surgeries is challenging owing to effects of hypoxia, decreased pulmonary blood flow, decrease systemic vascular resistance and acidosis. Anesthetic considerations must focus on minimizing the hemodynamic changes that would increase right to left shunt.

2. Case Report

A 12 yr, 20 Kg, female child presented with complaints of fever, abdominal pain, and vomiting for two days. Patient was a known case of Tetralogy of Fallot with associated VACTERAL anomalies. She underwent multiple surgeries including pulmonary valvotomy with MBT shunt in 2016 which was not corrected completely, craniotomy (Burr hole) for Brain abscess in frontal lobe in 2020. Patient had right hemiparesis with right facial nerve palsy with acute infarct in fronto parietal region. She was on Tab Ecosprin 75 mg and Tab Propranolol 20 mg OD.



On general examination, patient was severely malnourished, pale, febrile with pulse rate 63/min, RR- 26/min, peripheral and central cyanosis, grade IV clubbing in nails of both upper and lower limbs.

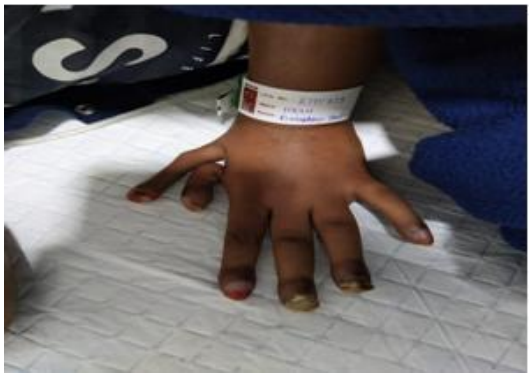
Polydactyly in the left hand, Kyphoscoliosis of the spine was also present.

On auscultation, there was clear audible S1 S2 with pansystolic murmur all over the chest. Air entry was equal on both sides with adventitious sound, SpO_2 in right foot and right arm was 80% with oxygen face mask at 6-7 liter /min.

SpO_2 on room air was 56%.

On investigation, Hb was 19.3, which shows haemoconcentration, TLC- 7790, Neutrophils- 65%. Lymphocytes- 25%, Platelets- 1,20,000 lakhs, Na- 146, K- 4.9 with normal KFT and LFTs. 2D Echo showed Levocardia, S/P mBT shunt with open pulmonary valvotomy, functioning left mBT shunt with respiratory variation. Tetralogy of Fallot 19 mm perimembranous VSD with aortic override, fair size PA branches, mild PS, PG-28 mm Hg. Normal Biventricular function. Chest X- ray revealed boot shaped heart.

She was posted for open appendectomy. High risk consent was taken and the patient was accepted under ASA grade III GA high risk. Infective endocarditis prophylaxis was given with Inj. Gentamicin 2gm IV. All emergency drugs and defibrillator were kept ready.



Monitors for ECG, NIBP, EtCO2 and SpO2 were connected. 22G IV cannula was put in and the patient was given general anesthesia. Premedication given was Inj. Glycopyrolate 0.1 mg, Inj. Fentanyl 25 mcg , Inj Midazolam 0.5 mg. Induction was done with Inj. Ketamine 50 mg and Sevoflurane at MAC - 4 Inj. Scoline 40 mg IV was given. Scopy was done using McCoy blade no.2 and uncuffed endotracheal tube no. 5 so was inserted under vision. Intermediate acting muscle relaxant Inj. Atracurium 10 mg was given. Inhalational agent Sevoflurane with Oxygen and air at 50:50 ratio were given to maintain adequate depth of anesthesia. Inj. Atracurium 2.5 mg and Inj. Ketamine 10 mg IV was given intraoperatively. Patient was catheterized.

Surgery lasted for 20 min, the patient was reversed with Inj. Glycopyrrolate 0.1 mg with Inj. Neostigmine 1.25 mg and smooth extubation was done after eye opening, adequate neck holding, obeying of oral commands and maintenance of Spo_2 80% by the patient, IV Paracetamol 300 mg given for postoperative analgesia. Pulse rate was maintained between 70-90 beats/ min and SpO2 between 75-80%. Urine output was adequate and 100ml Ringer lactate was given intraoperatively. Patient shifted to PICU for observation post surgery. Patient was discharged on 3rd post operative day.

3. Discussion

VACTERL- Associations

1) Babies associated with VACTERL anomalies are born small or have low birth weight and they have difficulty in gaining weight. Our case had severe malnourishment.
2) About 70% of patients with VACTERL association will have vertebral anomalies or defects of the spinal column. In early life, these do not cause any difficulties but in later life these spinal column abnormalities may put children at risk for developing kyphoscoliosis, scoliosis or curvature of spine. Here the patient had kyphoscoliosis.
3) Anal Artesia or imperforate anus is seen in about 55%of patient with VACTERL association. These anomalies are usually noted at birth and often require surgery in the first days of life. In our case, the patient had undergone Anorectal malformation correction in childhood.
4) Up to 75% of patients with VACTERL association have been reported to have congenital heart disease. Most common heart defects seen with this are VSD, ASD and TOF. Here patient had TOF – Tetralogy of Fallot.
5) Esophageal Atresia with TEF is seen in about 30% of patients with VACTERL association. Our patient didn't have TEF.
6) Renal or kidney defects are seen in 50% of patients with VACTERL association. 35% of patients with this have a signal umbilical artery which can often be associated with kidney or urological problems. These defects can be severe with incomplete formation of one or both kidneys. Here the patient had right renal agenesis.
7) Limb defects occur in up to 70% of babies with VACTERL anomalies. These defects include polydactyly, syndactyly, absent or displaced limb. Babies with limb defects on both sides tend to have kidney or urologic defects on both sides. Babies with limb defects on one side of the body tend to have kidney problems on that same side. Here the patient had Polydactyly on the left side with right renal agenesis.

VACTERL association defects are treated post birth with issues being approached one at a time. Because of multiple systems affected by this association, baseline investigations are necessary to rule out or determine the severity of the condition in addition to the usual physical observations including height and weight.

Vertebral anomalies – X-ray, Ultrasound and/or CT/MRI of the spine.

Anal Atresia – Physical examination/observation, abdominal ultrasound +/- additional testing for genitourinary anomalies.

Cardiac malformations – ECG (arrhythmias), Echocardiogram +/- Cardiac CT/MRI/Angiogram (exclude cardiac or vascular abnormalities; evaluate structure and function of heart). Paediatric cardiology consultation. CXR (cardiomegaly)

Tracheo-esophageal fistula – Physical examination/observation ; Chest X-ray – PA/lateral; CT/MRI (tracheoesphageal cleft) + Chest X- rayR (aspiration evidence) and Endoscopy.

Renal anomalies – Renal Ultrasound +/- voiding cystourethrogram; CT urogram.

Anesthetic management of patients with Fallot's tetralogy for non cardiac surgery is a challenge and requires a thorough understanding of pathophysiology, events and effects of medications which alters the magnitude of R- to- L shunt, and if it is associated with VACTERL anomalies the risk in terms of anesthesia increases more. Goal is to maintain normovolemia. The hypertrophied and poorly compliant RV is sensitive to hypovolemia which can lead to hypotension and increased Right to Left shunting. Avoiding techniques and agents that lower SVR and increase R- to- L shunting is important. Hypercarbia, hypoxemia, hypothermia and acidosis must be avoided as it can increase PVR and lead to increased R - to - L shunting. Excessive tachycardia and bradycardia should be avoided. Chronic hypoxia in such patients leads to cyanosis, secondary polycythemia, hyperviscosity and coagulation defects. Another problem in these patients is cyanotic spells which are usually triggered by 2 mechanisms: either decrease in SVR or spasm of cardiac muscle in the region of RVOT due to sympathetic stimulation (infundibular spasm). It responds to increase in volume and increase in SVR with alpha agonists such as Phenylephrine or Ephedrine and ceasing infundibular spasm with beta blockers like Propranolol or Esmolol.

Good pre-medication is important to reduce anxiety and smooth induction. This also reduces catecholamine release and avoids hypercyanotic spells in children with Fallot's Tetralogy. Here we have given Midazolam as pre-medications along with Glycopyrrolate to reduce hypersalivation. Care was taken to make sure that the IV line is free of air bubbles because even small amounts of air can lead to paradoxical embolism due to R- to- L shunt. Oxygen and air with sevoflurane was the preferred method of maintenance of anesthesia. Sevoflurane is an agent of choice for inhalation induction agent and Ketamine was used as IV induction agent as both increase SVR and thus decreases R-to- L shunt. Management of pain is an important concern because increase in sympathetic activity due to pain may trigger cyanotic spell in perioperative period. We have given Fentanyl intraoperatively and Paracetamol postoperatively for pain control. Patient with CHD coming for non cardiac surgeries are still under high risk postoperatively. Observing them in ICU will warrant assurance of catching arrhythmia, cardiac ischemia, dehydration, pain and other complications before they cause detrimental effects.

4. Conclusion

Careful administration of anesthesia after thorough understanding of physiology and pharmacology with meticulous planning combined with strict monitoring and vigilance can make a safe outcome in difficult cases. Our patient had cardiac anomaly which made anesthesia management challenging but we took all cautions in order to prevent possible hemodynamic disturbances, risk of regurgi-tation and aspiration. Close monitoring is crucial in perioperative and postoperative period. As findings are so variable in patients with VACTERL syndrome, each patient be carefully evaluated individually and anesthetic approach should be preferred according to the patient health conditions, risk factors and type of surgical treatment.

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