ORIGINAL RESEARCH PAPER
Volume - 10 | Issue - 05 | May - 2021 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

CASE OF VACTERL ASSOCIATION WITHOUT THE "V AND L": ANAESTHETIC CHALLENGES

Anaesthesiology

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ABSTRACT

Aim & Objective: To highlight the importance of anesthetic management in a small neonate with cleft lip, cleft palate, single kidney, atrial septal defect, posted for trachea-oesophageal fistula repair.

Case description: A female baby born at 35 weeks of gestation to a 23-year-old primigravida mother by caesarean section in view of foetal distress with breech presentation .Baby had difficulty in breathing and froathing from mouth. Cleft lip and cleft palate was present. A nasogastric tube was not going beyond 11 cm per oral route. Chest x ray showed coiling of tube in upper esophagus.F/S/O Type C Tracheo esophageal fistula. . Plain X-ray abdomen showed presence of bowel gas. Ultrasonography of the abdomen showed right renal agenesis. The left kidney was normal. Echocardiography showed presence of atrial septal defect with left to right shunt, tiny patent ductusarteriosus, mild pulmonary arterial hypertension, mild tricuspid regurgitation.

Based on the presence of tracheoesophagealfistula, atrialseptal defect, unilateral renal agenesis and absence of features, suggestive of alternative diagnosis infant, meet criteria of vacteral association.

Discussion: VACTERL is a cluster of congenital malformations based on the non-random association of various congenital malformations in a single patient. Here “V” denotes vertebral defects or vascular anomalies (single umbilical artery), “A” anal atresia, “C” cardiac abnormalities, “TE” tracheoesophageal fistula, “R"renal (kidney) abnormalities and “L” for limb anomalies) Diagnosis of VACTERL association is done only when at least three of the above mentioned congenital malformations are identified in a patient.

Although 80% of these cases have vertebral defects, our case is unique as patient does not have one of the commonest occuring association i.e., vertebral anomalies.

The other highlight of this case is although reports say that VACTERL babies with ipsilateral renal disorder have the same side limb defects, our case has a renal anomaly with no limb anomaly.

Conclusion: Anaesthetic challenges were difficult airway, endotracheal tube placement, low respiratory reserve, small maximum allowable blood loss, long duration of surgery, risk of hypothermia, aspiratedlungs, risk of right to left shunt, difficulty in securing intravenous line and intra arterialline. this case needs continuous monitoring of ECG, invasive blood.

KEYWORDS

VACTERL, Anesthetic challenges, trachea-oesophageal fistula repair , V and L

INTRODUCTION

VACTERL is a cluster of congenital malformations based on the non-random association of various congenital malformations in a single patient. Here “V” denotes vertebral defectsor vascular anomalies (single umbilical artery), “A” anal atresia, “C” cardiac abnormalities, “TE” tracheoesophageal fistula, “R"renal (kidney) abnormalities and “L” for limb anomalies.

It is called an association, rather than a syndrome because the complications are not pathogenetically related and thought to be linked to embryonic mesodermal defects. [1]

Diagnosis of VACTERL association is done only when at least three of the above mentioned congenital malformations are identified in a patient.

Although 80% of these cases have vertebral defects, our case is unique as patient does not have one of the commonest association i.e., vertebral anomalies, but has all other associations and an additional cleft lip and cleft palate.

The other highlight of this case is although reports say that VACTERL babies with ipsilateral renal disorder have the same side limb defects, our case has a renal anomaly with no limb anomaly.

Finally VACTERL was considered in our patient in view of atrial septal defect, tracheo esophageal fistula, unilateral renal agenesis.

Patient was classified as ASA grade IV in view of multiple anomalies. Blood investigation were as follows: haemoglobin-20gm/dl, total leucocyte count-25,000/..total bilirubin-15.11,indirect bilirubin -13.70,urea/creatinine-53.8/0.52,uric acid-12.46,serum potassium-6.81,CRP<2.8,INR-1.7.High risk consent was obtained and patient shifted to operation theater in a head up position in a warmed isolate for ligation of fistula and end to end esophageal anastomosis. Electrocardiogram, oxygensaturatin probe, on invasive BP were attached. Peripheral IV cannulation with 24g cannula was done, DNS infusion was started.glycopyrrolate20mcg was given IV. Patient was preoxygenated with 100%oxygen for 5 min with head elevated at 30^0.patient was induced with sevoflurane and breathing spontaneously, no IPPV was given. Under direct laryngoscopy/fiberoptic, orotracheal intubation was done with 3.5 mm I.D ETT and after confirmation of bilateral air entery, atracurium 1 mg was given and tube was fixed and positive pressure ventilation was maintained. Endotracheal suction was done through endotracheal tube. Hydrocortisone 5 mg was given. Anaesthesia was maintained with oxygen(50%),air (50%), sevoflurane and atracurium intermittently. Central venous catheterization was done in right internal jugular vein under ultrasound guidance and right radial artery is cannulated for beat to beat monitoring. Aprecordial stethoscope was placed over left axilla. surgery was allowed to start as patient was stable and maintaining saturation 98%-99%. [2]

Blood pressure was maintained on higher range to avoid right to left shunt. Hypoxia, hypercarbia, metabolic acidosis was avoided to reduce pulmonary vascular resistance. All care to avoid hypothermia & fluid overload was taken in view of CRF. Intra –operatively vitals were stable. Total surgery time was 120 min. Patient was shifted to

PICU for elective ventilation.post operatively patient was kept on antibiotics.

AIM & OBJECTIVE

To highlight the importance of anesthetic management in a small neonate with cleft lip, cleft palate, single kidney, atrial septal defect, posted for trachea-oesophageal fistula repair.

MATERIALS AND METHODS

A female baby born at 35 weeks of gestation to a 23-year-old primigravida mother by caesarean section in view of foetal distress with breech presentation**.** Baby had difficulty in breathing and froathing from mouth. Cleft lip and cleft palate was present. A nasogastric tube was not going beyond 11 cm per oral route. chest x ray showed coiling of tube in upper esophagus. F/S/O Type C Tracheo esophageal fistula. Plain X-ray abdomen showed presence of bowel gas. Ultrasonography of the abdomen showed right renal agenesis. The left kidney was normal. Echocardiography showed presence of atrial septal defect with left to right shunt, tiny patent ductusarteriosus, mild pulmonary arterial hypertension, mild tricuspid regurgitation.

Based on the presence of tracheoesophagealfistula, atrialseptal defect, unilateral renal agenesis and absence of features, suggestive of alternative diagnosis infant, meet criteria of vacteral association.

DISCUSSION AND RESULT

VACTERAL association can cause problems during induction and maintenance of anaesthesia due to multiple anomalies.

VACTERAL association term was first developed by Quan and Smith in 1973. In 1975 VATER was expanded to VACTERAL.[3]

Our patient presented with respiratory distress and frothing from mouth. She also had cleft lip and cleft palate. Threat to life can occur during induction because inhaled air can bypass the lung through the fistula into the stomach causing hypoventilation and gastric distention. There is a continual risk of acidic stomach contents refluxing through the fistula back in to trachea causing aspiration pneumonitis. With the above anticipation, patient was kept nil per orally and head elevation was done. Glycopyrolate was given to avoid excessive secretion. Adequate pre oxygenation was done. Patient was kept on spontaneous ventilation till trachea was intubated and bilataeral air entry was confirmed. With this we could avoid soiling of lung and hypoventilation. To avoid the risk of aspiration, rapid sequence induction with suxamethonium is an ideal technique. Our patient was the case of a single kidney with hyperkalemia, so we could not use suxamethonium, due to risk of hyperkalemia leading to cardiac arrest. In this patient, care was taken to avoid hyperkalemia, fluidoverload, metabolicacidosis, hypoxia and hypercarbia to avoid decrease in pulmonary vascular resistance leading to right to left shunt. Blood pressure was maintained on higher side to keep systemic vascular resistance high with the use of phenyl epinephrine intermittently.[4]

After explaining the anaesthesia risk to the relatives with informed consent was accepted for surgery.

Patients with tracheo- esophageal fistula are susceptible to respiratory infections due to weak respiratory muscle and hyper reactive airways. These patients need active respiratory care with antibiotics, pre & post operatively. A silent aspiration is a great threat to life at induction of anaesthesia even if patient is adequately starved. This case demands vigilance & extra care due to cleft lip and palate which makes laryngoscopy more difficult.[5]

CONCLUSION

The pre-existing anomalies like ASD & single kidney with potential risk of difficult intubation and risk of aspiration in patient with VACTERAL association demands careful pre-operative assessment and use of skillful anaesthetic technique to avoid fatal complication.

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