Case Report:

Cleft palate repair, tongue tie release with anterior sagittal anorectoplasty in a patient with VACTERL association: An anaesthetic challenge

Dr. Abhishek Choudhary¹, Dr. Nitin Johar¹, Dr. Rahul Gupta², Dr. Rambabu Sharma¹, Dr. Shyam Bihari Sharma³ & Dr. Meenaxi Sharma⁴

¹Resident, Department of Anaesthesia, ²Assistant Professor, M.Ch. Paediatric Surgery, ³Professor, M.Ch. Paediatric Surgery, ⁴Professor & HOD, Department of Anaesthesia.

Name of the Institute/college: Department of Anaesthesia and Paediatric Surgery division of Department of General Surgery, NIMS University Medical College, Shobha Nagar, Jaipur, Rajasthan.

Corresponding author: Dr. Rahul Gupta

Date of submission: 05 Aug 2014 ; Date of Publication: 25 September 2014

Abstract:

VACTERL Association is a set of congenital anomalies which often occur together in diverse combination. Repair of cleft palate and anorectal malformation could be challenging for an anaesthetist in view of difficult airway. Here we report a rare case of a 2-year-old female child who presented with anteriorly placed anus, cleft palate, tongue tie and congenital absence of a right thumb. On evaluation, there was segmentation defect of the sacral spine and VACTERL association was established. Airway assessment showed Mallampati Classification grade III. Successful anaesthetic management of cleft palate repair, tongue tie release with Anterior Sagittal AnoRectoPlasty was performed in a single setting.

Key words: VACTERL Association; cleft palate; anorectoplasty; difficult airway; segmentation defect.

Introduction:

VACTERL Association is a group of congenital anomalies which often occur together in different combination. To qualify for VACTERL child, they must have at least two or more of the following anomalies: V- Vertebral, A- Anal, C- Cardiovascular, TEF- Tracheoesophageal Fistula, R- Renal and L- Limb anomalies¹-³.

In addition to these anomalies they may have other characteristics which occur more frequently in affected child than the rest of the population, and they are ear abnormalities, genital anomalies, cleft lip or palate, thumb abnormalities and presence of single artery in the umbilical cord¹-³.

Due to multiple congenital anomalies of different system, anaesthetic management of these patients can be complicated. Here we report the successful anaesthetic management of a rare case of a 2-year-old female child with VACTERL association for cleft palate repair with tongue tie release with Anterior Sagittal AnoRectoPlasty (ASARP).

Case Report:

A 2-year-old female child came to the pediatric outpatient department with chief complaints of anteriorly placed anal opening and defect in the palate since birth. The patient regurgitated liquids from nose since birth. The patient had small anteriorly placed anal opening, just posterior to the vaginal vestibule and used to pass stools once in 2 to 3 days, semisolid in consistency. The child also had ankyloglossia (tongue tie), segmentation defect of the sacral spine and had congenital absence of a right thumb.

The child was born full-term with normal vaginal delivery, and baby cried immediately after birth. The patient regurgitated liquids from nose since birth. The patient had small anteriorly placed anal opening, just posterior to the vaginal vestibule and used to pass stools once in 2 to 3 days, semisolid in consistency. The child also had ankyloglossia (tongue tie), segmentation defect of the sacral spine and had congenital absence of a right thumb.

Family history showed no consanguinity and no history of congenital malformation in any other family members. The weight of child was 8 kg and height 76 cm and was rated PEM grade II. Baby
was never breastfeed and was given top feeds with rubber nipple.
The baby was evaluated and diagnosed with VACTERL Association, having anorectal malformation, cleft palate, ankyloglossia, segmentation defect of sacral spine and congenital absence of right thumb. The paediatric surgeon planned for cleft palate repair, tongue tie release with ASARP. A complete pre anaesthetic evaluation was performed. General physical examination was normal, except for the nasal discharge and altered texture of the hair. Systemic examination, revealed normal heart sounds, S1 and S2 and bilateral air entry in the lung was equal and clear. The blood investigations revealed haemoglobin-12.1 gm%, TLC-8600, serum urea-19.2mg/dl, serum creatinine-0.43 mg/dl, random blood sugar-96 mg/dl, bleeding time-2min 5sec and clotting time-4min 6 sec, and blood group-B+ve. Chest X-ray and ECG were normal, however Magnetic Resonance Imaging (MRI) spine showed segmentation defects of the sacral spine. Airway assessment showed Mallampati Classification (MPC) grade III. Patient was classified as American Society of Anaesthesiologists (ASA) grade III in view of multiple anomalies. Patient had no cardiac and neurological abnormalities.
Patient was kept NPO for 6 hours before operation. Patient was taken to operating room and baseline monitors were attached including pulse oximetry, noninvasive blood pressure (NIBP) and electrocardiography (ECG). A Foley’s catheter was inserted and connected to urinary bag. An intravenous line with 24 G intravenous cannula was secured and iv fluids (Isolyte P) were started as per Holliday and Segar formula. Anticipating difficult intubation a difficult airway cart was kept ready. Inhalational induction was planned anticipating difficult intubation. Patient was pre oxygenated with 100% oxygen for 3 minutes and induced with Sevoflurane 5% along with O2, N2O. Once the patient was induced, laryngoscopy was performed using a gauge piece between the gap to prevent trauma to the underlying tissue. Trachea was intubated with endotracheal tube (ET) #4 with the help of stylet and BURP (Backwards Upwards Rightwards Pressure) manoeuvre in the second attempt. Bilateral air entry was checked and the tube was fixed in the midline of the lower lip. A throat pack was inserted using Magill’s forceps.
Inj. Fentanyl 2microgm/kg was given iv and inj. Vecuronium 0.1 mg/kg was given as a neuromuscular relaxant. Anaesthesia was maintained on O2, N2O and Sevoflurane. End tidal CO2 probe was connected to the ET and normocapnia was maintained. ET Tube position was checked after the patient was positioned and following the insertion and opening of the Dingman mouth gag. First cleft palate repair was performed, then tongue tie was released and then ASARP was performed. The surgery went uneventfully for four hours. Infraorbital nerve block and Caudal block was given with 2ml of 0.25% Bupivacaine for postoperative analgesia. After the surgical procedure was over neuromuscular blocked was reversed with inj. Neostigmine 0.05mg/kg and inj. Glycopyrrolate 0.01 mg/kg. The patient was hemodynamically stable intraoperatively and was extubated on table. A gentle laryngoscopic examination and suction was done to rule out oedema and to remove clots. Patient was shifted to paediatric ICU with O2 by ventimask@2litre/min in stable condition. She had a smooth and uneventful recovery and is under follow up.
Discussion:
VATER / VACTERL Association is a set of congenital malformation occurring together in newborn babies. The incidence is estimated at approximately 1 in 10,000 to 1 in 40,000 live-born
infants. VACTERL / VATER Association was first termed by Quan & Smith in 1973[^4].

In 1975 the VATER Association was changed to VACTERL Association which includes V-Vertebral anomalies 70%, A-Anal atresia 80%, C-Cardiovascular defect 53%,[^5] TE-Tracheoesophageal malformation 70%, R-Renal anomalies 53%,[^6] and L-Limb anomalies 65%. In addition to these core component features patient may have other characteristics such as ear abnormalities, genital anomalies, cleft lip/palate, thumb abnormalities and presence of in the umbilical cord. To qualify as a ‘VACTERL’ child three of the seven core component should be present.

One explanation for the group of anomalies in a new born is the “developmental field defect” in which malformation that occur in blastogenesis tend to result in polytrophic anomalies, or birth defects affecting multiple organ system.[^7-8] The malformation predominantly occurs in association and with sporadic presentation in families without previous history.

Our patient had anorectal malformation,[^9] segmentation defect of sacral spine, absence of right thumb, cleft palate and tongue tie and was included into VACTERL group. There were no cardiac and renal anomalies. Both cleft palate repair and anterior sagittal anorectoplasty was performed in a single setting and was an anaesthetic challenge because of “trouble at both ends”, ASA grade III, long intraoperative time in a paediatric patient and index case (VACTERL Association) for the institution. Anticipating difficult airway a difficult airway cart was kept ready. Positive pressure ventilation was avoided to prevent regurgitation of gastric content. Inhalational agents were used to induce the patient in view of difficult intubation. The patient should be successfully intubated in one or few attempts with the help of a stylet and BURP manoeuvre.[^10]

Intraoperative hypothermia was taken care and monitored vigilantly. For postoperative analgesia, infraorbital block and caudal block should be given to patients for painless postoperatively course[^11].

**CONCLUSION:**

In conclusion the patient with VACTERL anomaly with potential risk of regurgitation and difficult airway demands careful pre-operative assessment, use of skilful anaesthetic technique and close monitoring to avoid fatal complications.
Figure-4: Intraoperative photograph showing on left- mobilisation of anorectum, right- completion of Anterior Sagittal AnoRectoPlasty.

References: