

A patient with VACTERL association for caesarean delivery

Sir,

VACTERL association (V: Vertebral anomalies, A: Anal malformation, C: Cardiovascular defect, TE: Tracheal and esophageal malformation, R: Renal agenesis, L: Limb anomalies), is a cluster of congenital malformations and is most likely caused by multiple factors.^[1] VACTERL is typically defined by the presence of at least three of the congenital malformations.^[2] The incidence is estimated approximately as 1 in 10,000-1 in 40,000 live-born infants.

Here, we share our experience of anaesthetic management of a pregnant patient with VACTERL syndrome for cesarean delivery.

A 30 year old primigravida was admitted for elective cesarean delivery at 38 weeks of gestation. She was a known case of VACTERL syndrome and was detected to have multiple anomalies like vertebral fusion of L3 and L4, anorectal anomaly, cardiac abnormality with ventricular septal defect (VSD), infundibular stenosis, and preaxial polydactyly in left hand. She had undergone multiple corrective surgeries in the past. At 6 months of age, cut back anoplasty; at the age of 12, VSD closure and infundibular resection and at the age of 13, ureteral reimplantation were done.

Her height was 148 cm and weight 64 kg. Airway examination showed Mallampatti Class I. Her preoperative blood workup was normal. An X-ray abdomen, done 2 years back, showed L3–L4 vertebral fusion [Figures 1 and 2]. Echocardiogram at 36 weeks of

her gestation showed ejection fraction of 50%, adequate left ventricular function, and no residual shunt in the ventricular septum. Electrocardiogram showed right bundle branch block. Her cardiopulmonary reserve seemed good with a good effort tolerance. Due to her history of multiple corrective surgeries, a cardiologist and urologist were involved in the preoperative workup.

After preanaesthetic work up and a detailed discussion of the plan of management with the obstetrician, patient, and her spouse, an elective cesarean section under general anaesthesia was scheduled and an informed consent was taken. After premedication with injection ranitidine 1 mg/kg, the patient was shifted in left lateral position to operating room. Under standard monitoring and after preoxygenation for 5 min with 100% oxygen, rapid sequence induction of general anaesthesia was started with thiopentone 4 mg/kg and succinylcholine 2 mg/kg. Anaesthesia was maintained with sevoflurane (1 MAC) and atracurium 0.5 mg/kg. After 6 min of induction, the baby was delivered and handed over to the pediatrician. Transverse abdominal plane (TAP) block under ultrasound guidance was given for postoperative analgesia. Patient was extubated on table and shifted to the postanesthesia care unit for observation. The entire procedure was uneventful. The mother and the baby were safe and discharged in good health on the 4th postoperative day.

The management of patients with VACTERL association is complex. Due to the rare incidence of VACTERL association, not many cases have been reported.^[3,4] The congenital malformations associated can result in long-term sequelae. The obstetrician opted for an elective cesarean delivery taking into consideration the corrective surgeries in the past (pelvic floor surgery).

Our options were to choose between general or regional anaesthesia or a combination of both. We were concerned about using a neuraxial block as the primary anaesthetic technique for cesarean delivery for a number of reasons. Literature suggests that 25% of patients who receive spinal anaesthesia for cesarean delivery have a probability of extension of sensory blocks to the cervical dermatome region.^[5] Due to the vertebral column abnormalities, these patients have a higher incidence of difficult neuraxial block placement and failure of the same.^[6] In our patient as the urologist (called for co-management)



Figure 1: X-ray spine (Lumbosacral)

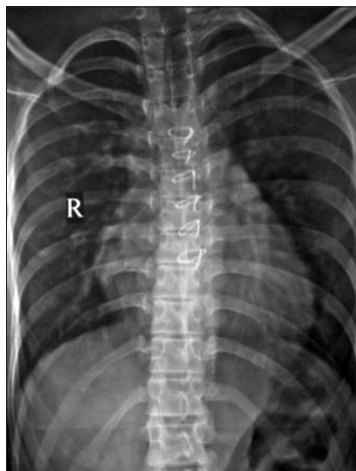


Figure 2: X-ray chest

and obstetrician decided for a midline abdominal incision, we decided that a general anaesthesia would be more ideal. After detailed discussion and multidisciplinary specialist input, we chose to use general anaesthesia for cesarean delivery along with ultrasound-guided TAP block for postoperative analgesia. The successful obstetric and anaesthetic outcomes for our patient were possible as a result of the specialist multidisciplinary review and clinical care that she received during the antenatal, peripartum, and postpartum periods.

The report advocates that women with potentially serious medical conditions should receive immediate and appropriate multidisciplinary specialist care.^[7] As exemplified in this case report, an individualized approach should be considered for the anaesthetic management of high-risk pregnant patients with complex and multiple medical and surgical morbidities undergoing labour and delivery.

**Kamalakkannan Thulasidoss,
Aparna Duraisamy¹, Muruges Babu**

Department of Anaesthesiology, ¹Department of Obstetrics and Gynaecology, Apollo Hospitals, Karapakkam, Chennai, Tamil Nadu, India

Address for correspondence:

Dr. Kamalakkannan Thulasidoss,
130, University Staff Quarters, Palkalai Nagar,
Palavakkam, Chennai, Tamil Nadu, India.
E-mail: kamprat15@gmail.com

REFERENCES

1. Hersh JH, Angle B, Fox TL, Barth RF, Bendon RW, Gowans G. Developmental field defects: Coming together of associations and sequences during blastogenesis. *Am J Med Genet* 2002;110:320-3.
2. Shaw-Smith C. Oesophageal atresia, tracheo-oesophageal fistula, and the VACTERL association: Review of genetics and epidemiology. *J Med Genet* 2006;43:545-54.
3. Luce V, Mercier FJ, Benhamou D. Anesthetic management for a parturient affected by the VACTERL association. *Anesth Analg* 2004;98:874.
4. Hilton G, Mihm F, Butwick A. Anesthetic management of a parturient with VACTERL association undergoing Cesarean delivery. *Can J Anaesth* 2013;60:570-6.
5. Russell IF, Holmqvist EL. Subarachnoid analgesia for caesarean section. A double-blind comparison of plain and hyperbaric 0.5% bupivacaine. *Br J Anaesth* 1987;59:347-53.
6. Vercauteren M, Waets P, Pitkänen M, Förster J. Neuraxial techniques in patients with pre-existing back impairment or prior spine interventions: A topical review with special reference to obstetrics. *Acta Anaesthesiol Scand* 2011;55:910-7.
7. Cantwell R, Clutton-Brock T, Cooper G, Dawson A, Drife J, Garrod D, *et al.* Saving Mothers' Lives: Reviewing maternal deaths to make motherhood safer: 2006-2008. The Eighth Report of the Confidential Enquiries into Maternal Deaths in the United Kingdom. *BJOG* 2011;118 Suppl 1:1-203.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/0019-5049.156896