Department of Anesthesiology
Perioperative, Critical Care
and Pain Medicine

Cincinnati Children's

Midnight VACTERL Challenge: A Complicated Cesarean Delivery After Dark

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- VACTERL is a rare collection of congenital abnormalities that pose significant challenges throughout life with some complicating care during pregnancy and delivery.

- 23yo G2P0101 at 34wks gestation
- Presented overnight with painful contractions and spontaneous rupture of membranes on arrival.
- Chronic hypertension, VACTERL:
  - Congenital cervical fusion with subluxation of C4.
  - Levoconvex lumbar curvature with coccygeal agenesis
  - Imperforate anus s/p surgical repair
  - Neurogenic bowel s/p Malone Antegrade Colone Enema (MACE) procedure
  - TOF w/ chronic severe PI
    - Pre-Pregnancy ECHO: severe pulmonary regurgitation, severely dilated RV, normal LVEF
  - Chronic renal insufficiency
  - Left radial hypoplasia, left thumb aplasia

A group of anomalies that occur in the newborn
- Approximately **1 in 10,000 to 40,000 births**
- The cause of VACTERL association is **unknown**
- The mode of inheritance is **sporadic**
- Greater frequency in **infants of diabetic mothers**

DIAGNOSIS
- VACTERL is a **clinical diagnosis**
- **≥ 3 features** required for diagnosis
- **Diagnosis of exclusion** – other possible causes of birth defects need to be ruled out

Vertebral defects
- Present in 60-80% of cases
- Includes hemivertebrae, hypoplastic vertebrae, extra or missing vertebrae, congenital scoliosis, spina bifida
- Can cause back pain later in life

Anal atresia
- Present in 60-90% of cases
- Narrowing or lack of patency of the anus
- May be accompanied by genitourinary abnormalities
- No passage of meconium, abdominal distension, vomiting

Cardiac defects
- Present in 40-80% of cases
- Subtle to life threatening defects
- Common defects: ventricular septal defect, atrial septal defect, tetralogy of fallot, patent ductus arteriosus

Tracheo-esophageal fistula
Esophageal atresia
- Present in 50-80% of cases
- TEF is an abnormal connection between the trachea and the esophagus
- EA is a congenital condition characterized by the incomplete development of the esophagus
- TEFs are most often found in conjunction with EA
- May present with vomiting, copious oral secretions, feed intolerance, coughing, cyanosis, or respiratory distress

Renal anomalies
- Present in 50-80% of cases
- Renal aplasia, renal dysplasia, vesicoureteral reflux, displaced or mal-positioned kidneys
- May not cause problems immediately
- Nephrology and urology consultation may be necessary

Limb abnormalities
- Present in 50% of cases
- Radial aplasia – underdevelopment of the radius
- Polydactyly (additional digits), missing digits, lower limb hypoplasia, syndactyly (webbing)

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Urgent repeat cesarean section indicated

- Extensive pelvic floor reconstruction
- Prior c/s under neuraxial. Difficult placement but successful.
- Peds Cardiac Anesthesiology consulted

Placement of CSE

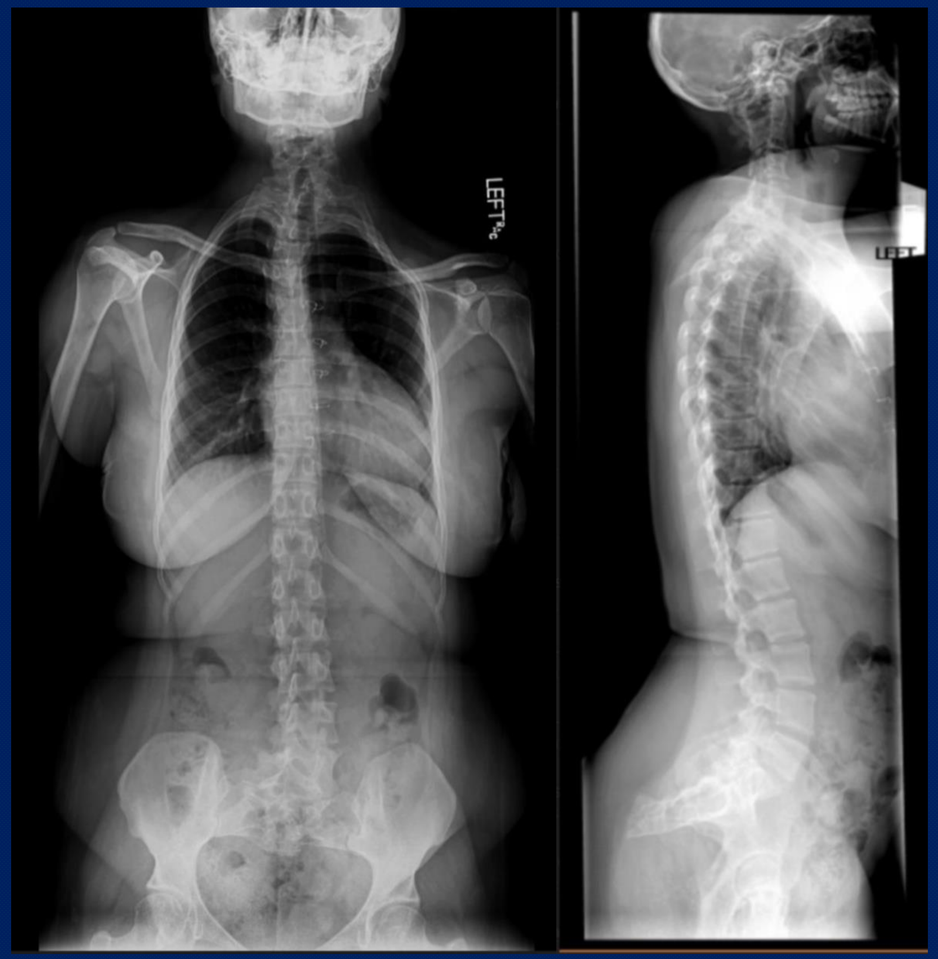
- Ultrasound guidance- identify midline landmarks and map spinal curvature
- T4 sensory level using 1.5ml hyperbaric bupivacaine, 150mcg PF morphine, 15mcg fentanyl.

Support RV, minimize pulmonary regurgitation

- Fluid restriction
- Ephedrine, diluted norepinephrine for hemodynamic support
- Avoid increases in PVR: avoid hypoxia, hypercarbia

Significant abdominal adhesions. Severe patient anxiety.

- Propofol boluses
- Successful delivery of viable male without any notable congenital abnormalities.



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VACTERL occurs in approximately 1 in 10,000-40,000 live births, defined by the presence of at least three abnormalities including vertebral, anal, cardiac, tracheoesophageal, renal, or limb defects (1).

Associated scoliosis may make neuraxial procedures challenging and resulting blockade can be unpredictable.

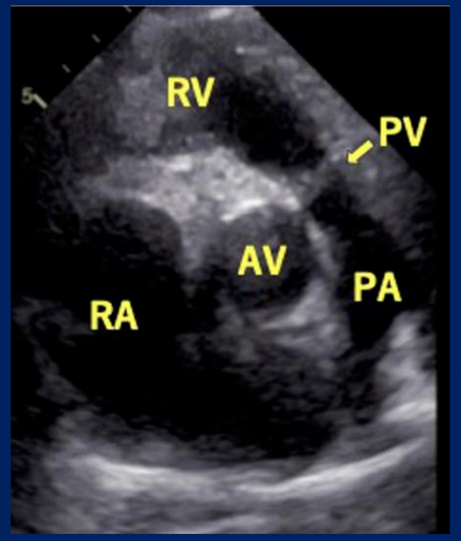
Cervical spine abnormalities, such as congenital fusion, may complicate endotracheal intubation.

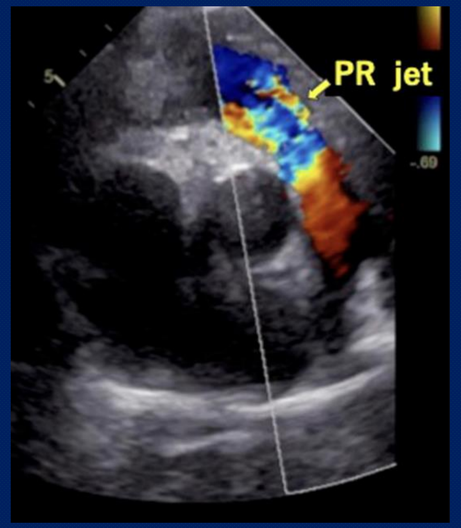
Although successful cesarean deliveries in patients with VACTERL have been reported, anesthetic management must be individualized (2,3).

Given the added cardiovascular burden in pregnancy, early workup is vital. Consider bedside ECHO in emergencies.

In this case, severe pulmonary regurgitation required fluid restriction, avoidance of increases in PVR, and maintenance of baseline sinus tachycardia (4).

This case highlights the complexity of anesthetic management in patients with VACTERL, particularly when urgent cesarean delivery is required overnight.





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References: 1- PMID: 21846383 2- PMID: 36798639 3- PMID: 23519725 4- PMID: 32491496