

Neuraxial Anesthesia for a Cesarean Delivery of a Parturient with **VACTERL** Association

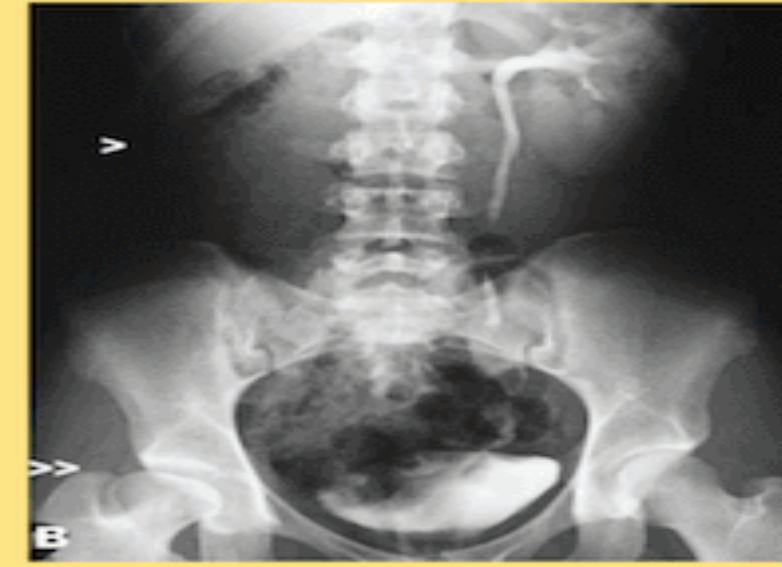
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- **VACTERL association** is a rare multi-system congenital disorder with an estimated incidence of 1/10,000 – 1/40,000 births
 - **Vertebral Defects**
 - **Anal atresia**
 - **Cardiac defects** (VSD, TOF, ASD, PDA)
 - **TracheoEsophageal Fistula**
 - **Renal abnormalities**
 - **Limb abnormalities**
- Diagnosis requires presence of 3 or more abnormalities in the absence of other congenital abnormality – intelligence is normal
- With advances in early care, particularly with surgical correction of cardiac and TE abnormalities, more women with VACTERL are living to adulthood and having children
- The features of VACTERL make pregnancy, delivery, and anesthesia more complicated
 - safe care requires a multi-disciplinary approach

VACTERL ASSOCIATION

The term 'VACTERL' describes a group of anomalies which often occur together in newborn babies.

To qualify as a 'VACTERL' child, 3 of the 7 anomalies mentioned below must be present.

	Vertebral defects - Affect between 60-80% - Hypoplastic vertebrae - Hemivertebrae - Congenital scoliosis - Spina bifida
	Anorectal anomalies - Affect between 60-90% - Low type - opening is present but in abnormal position or covered by a layer of skin/membrane - High type - no anal opening present - May have openings to the urethra, bladder, base of penis or vagina
	Cardiac defects - Affect between 40-80% - Ventricular Septal Defect - Atrial Septal Defect - Tetralogy of Fallot
	Tracheoesophageal fistula (TOF) with/without esophageal atresia (OA) - Affect between 50-80% - TOF - part of the oesophagus is joined to the trachea - OA - a short section at the top of the oesophagus has not formed properly and is not connected to the stomach
	Renal abnormalities - Affect between 50-80% - Lack of development of one or more kidneys (renal aplasia) - Malformation of one or both kidneys (renal dysplasia) - Displaced or malpositioned kidneys (renal ectopia)
	Limb defects - Affect around 50% - Underdevelopment of the radius (radial aplasia) - Extra digits (polydactyly) - Webbing of digits (syndactyly) - Lower limb malformations - clubfoot, hypoplasia of tibia/great toe

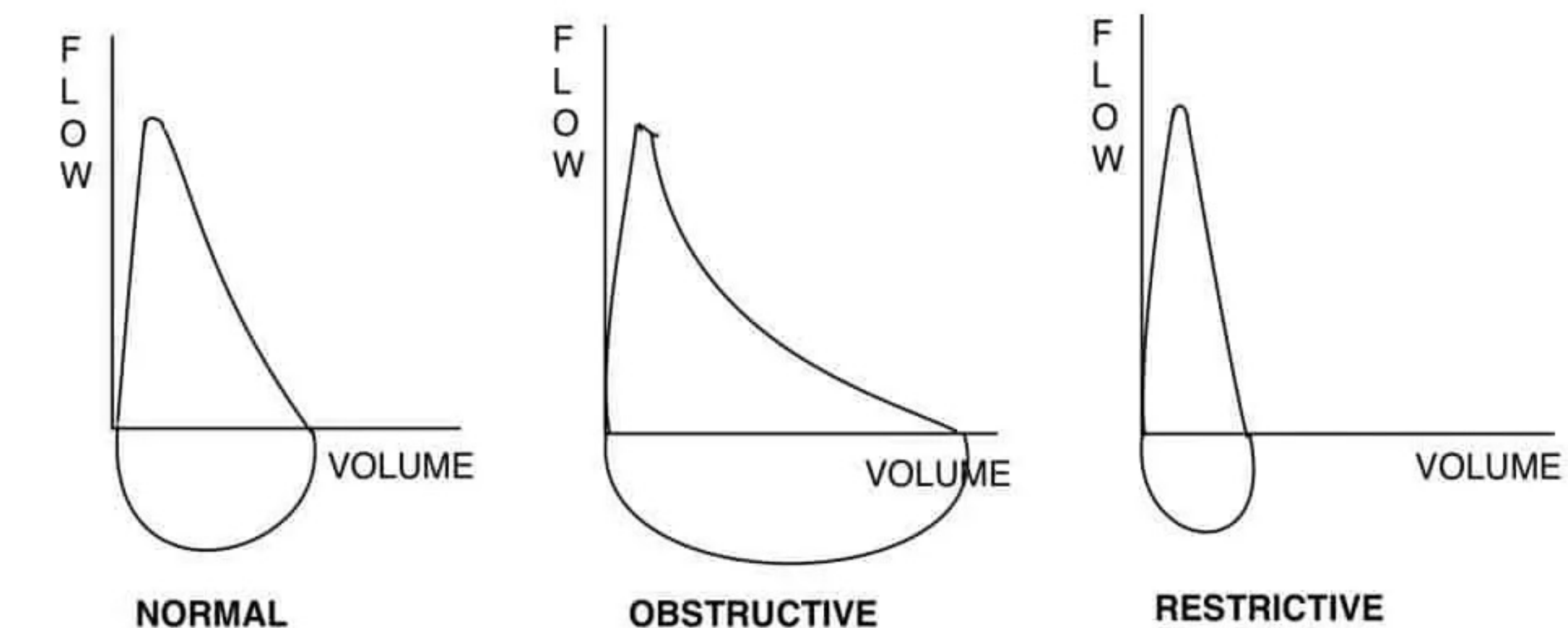
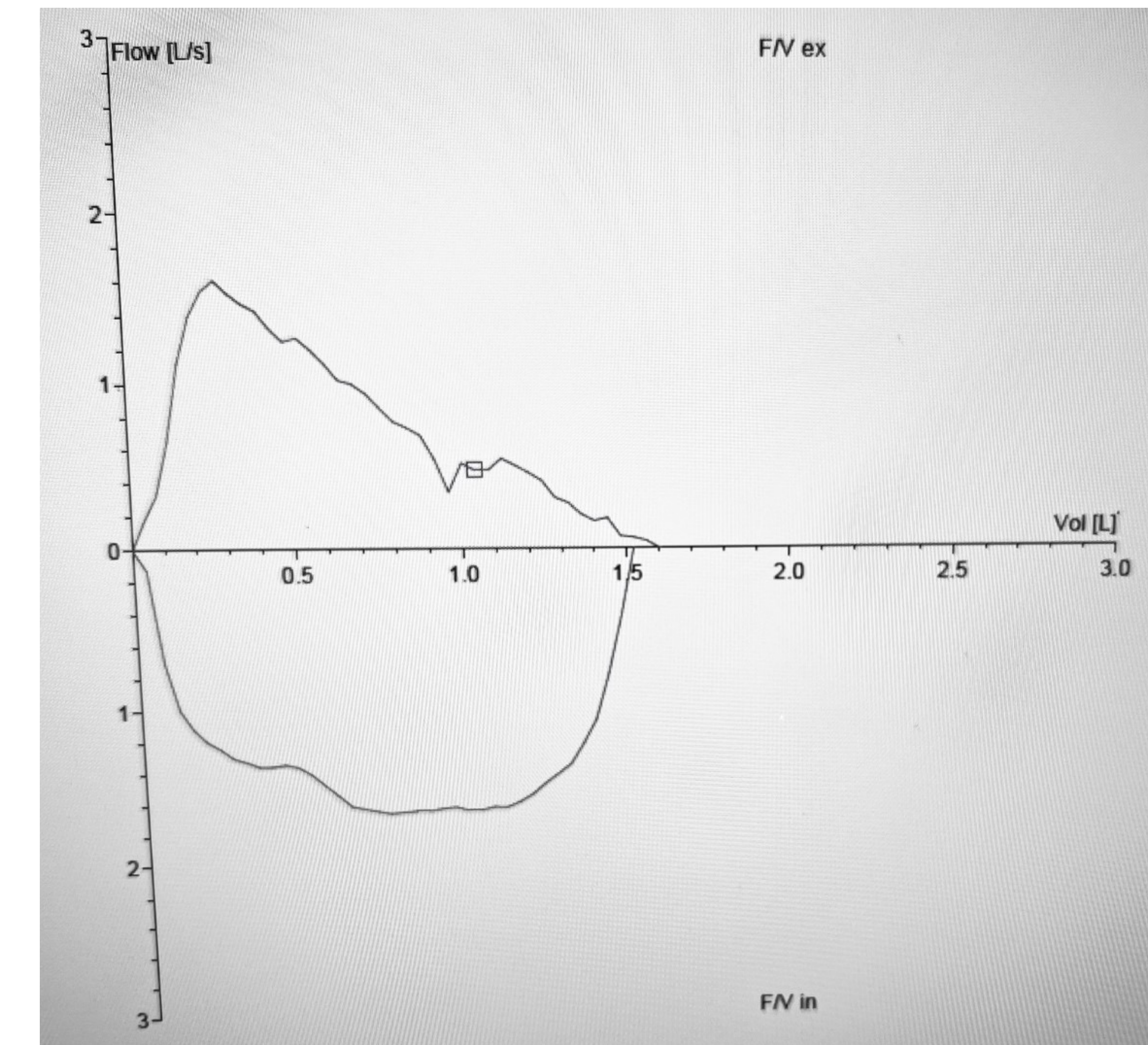
Case Presentation

- 20 y/o G₂P₀ woman with **VACTERL** diagnosis
 - Severe, uncorrected thoracic scoliosis
 - Atrial and Ventricular Septal Defects → corrected early in life
 - Tracheoesophageal Fistula → corrected early in life
 - Hypoplastic right upper extremity
- Scheduled for primary Cesarean delivery secondary to small body and contracted pelvis
- Physical Exam
 - Pleasant and cooperative
 - Height 4'5", Weight 35 kg
 - Airway exam reassuring
 - Thoracic scoliosis with easily palpable spinous processes in the lumbar spine
 - Breath sounds clear but diminished bilaterally
- Echocardiogram
 - Normal biventricular function
 - No evidence of residual ASD nor VSD, no shunts

Pulmonary Function Test

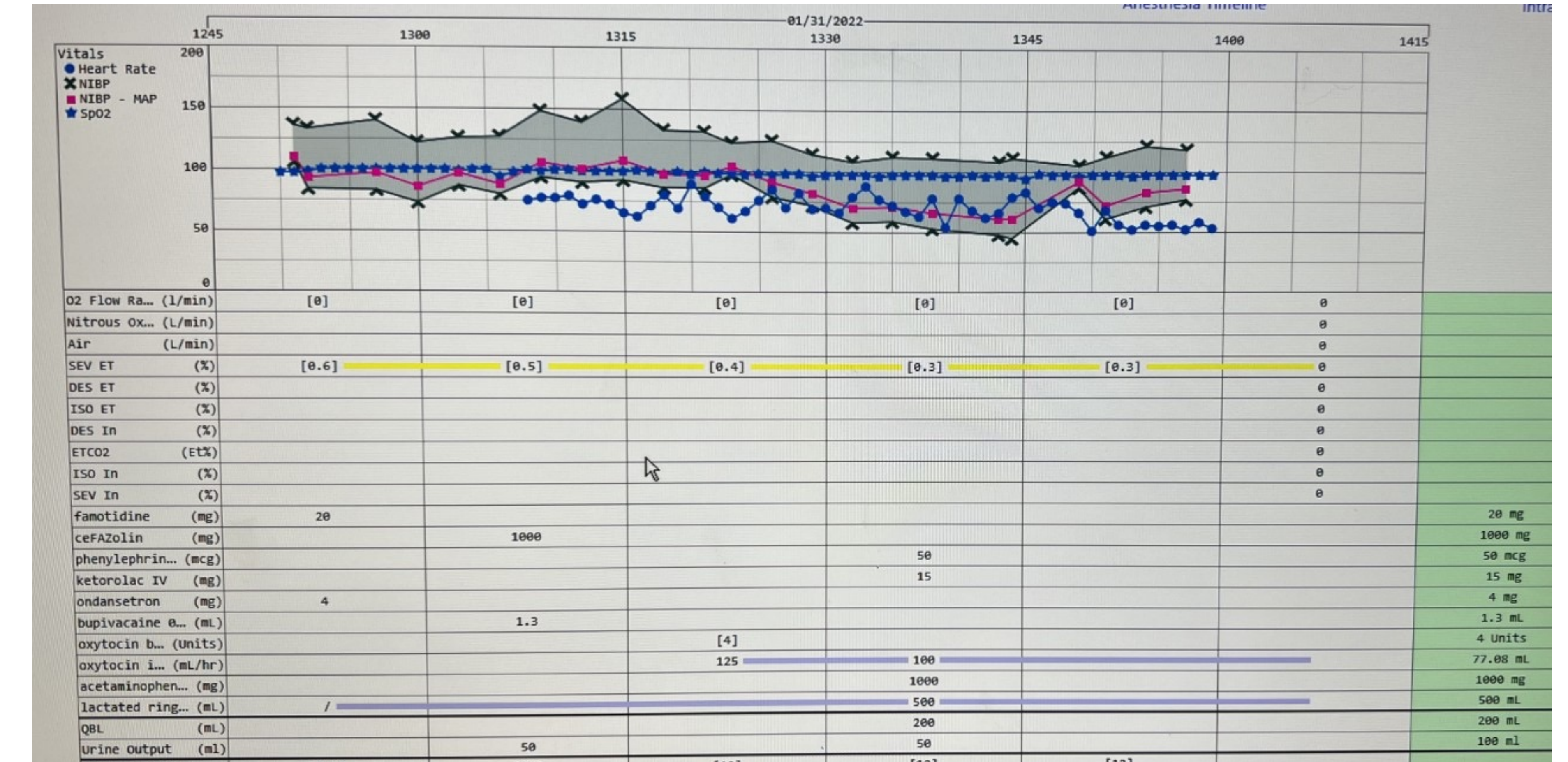
- FVC: 2.58 L (62% predicted)
- FEV₁/FVC 63% (67% predicted)
- VC 1.6 L (62% predicted)
- TLC 2.41 L (71% predicted)
- PEF 1.61 L/s (29% predicted)
- FRC 1.24 L (72% predicted)

*“ Results consistent with **co-existing obstruction and restriction with severe ventilatory defect**. The spirometry pattern discloses obstructive defect in the small airways. **Mid-expiratory flow rates are disproportionately decreased** and correspond to a hyperbolic shape of the expiratory portion of the flow volume loop. The lung volume pattern discloses the presence of a **restrictive lung defect**. The **Total Lung Capacity, Functional Residual Capacity and Residual Volume are decreased**. ”*



Anesthetic Considerations and Perioperative Course

- Neuraxial and general anesthesia each with potential difficulty:
 - **Neuraxial** – difficult placement with scoliosis, **high spinal risk** given short stature
 - **General** – **rapid desaturation** with ventilatory deficits, post-op respiratory depression, **suboptimal pain control** without neuraxial morphine
- After discussion of benefits/risks patient prefers neuraxial
- Concern for high spinal given short stature
 - Reduced dose spinal? → may not last long enough, how much to reduce?
 - Epidural → slow onset, potential for asymmetric block
 - **CSE with reduced intrathecal dose** → allows for reduced chance of high spinal with epidural in-situ to prolong or augment spinal!!
- CSE placed at L2-3 with reduced intrathecal dose:
 - Hyperbaric bupivacaine 0.75% **0.8 cc**, fentanyl 15 mcg, morphine 0.1 mg
 - Remained sitting 2 minutes to prevent excessive cephalad spread and allow for adequate saddle anesthesia for vaginal prep and foley placement
 - Positioned supine with LUD → T3 sensory level to ice within several minutes with dense motor block in legs reflecting adequate block density
 - Case proceeded uneventfully with no need to dose epidural
- Ambulating well on POD 1 with pain 1/10



Conclusion: Patients with VACTERL present unique anesthetic challenges. In this case, restrictive lung disease made general anesthesia riskier, while scoliosis and small stature may have complicated neuraxial anesthesia. Given the patient's size, we opted for a very low-dose CSE to minimize the chance of high-spinal, while allowing for extension of the block via epidural catheter if needed. Thorough evaluation of the patient allowed for safe, awake delivery.

Reference

Hilton, G et al. (2013) Anesthetic Management of a parturient with VACTERL association undergoing cesarean delivery. *Can J Anesth*, 60(6), 570-576.
Aycan, IO et al. (2014) Anesthetic management in VACTERL syndrome. *Journal of Clinical and Experimental Investigations* 5(1): 10.-105