Title: Anesthetic management of a toddler with VACTERL complex and chronic renal failure for the PSARP procedure

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Educational Objectives: At the conclusion of this session the participant will be able to:

1. Identify the common elements of the VACTERL complex
2. Describe basic elements of cloacal anomalies, imperforate anus and their surgical management
3. Describe common causes of acute renal failure in neonates and infants
4. Delineate the typical features of chronic renal failure (CRF) in infants and toddlers, including management strategies
5. Develop an anesthetic plan for this patient which includes the intra-operative correction of electrolyte disturbances.
6. Define a strategy for intra/postoperative pain control for this patient

Case Description:
A sixteen-month old girl is scheduled for posterior sagittal anorectoplasty (PSARP) as an initial step in the correction of her complex cloacal abnormality. She has VACTERL complex, and chronic renal failure. She was Twin “B” of a gestation complicated by preterm labor and delivery. However, prenatal ultrasound revealed the absence of the left kidney and the bladder, a dysplastic right kidney, and an imperforate anus. Preterm at 33 weeks, she weighed 1800 grams, and had APGAR’s of 3 and 8. Her associated malformations included a duplicate uterus and vagina, a fistula connecting the mid-colon to the cloacal cavity, and a right ureter which drained into the ipsilateral uterus.
Her previous surgeries had gone well, including an initial diverting colostomy, and a subsequent cutaneous ureterostomy. In addition to her complex urogenital malformations, her current problems include latex sensitivity, hypertension, chronic anemia and Type-2 renal tubular acidosis. Her growth has been poor (6.38 Kg), although she has met expected cognitive developmental milestones. She has been followed closely by the pediatric nephrology service, who are aware of the planned surgery.

The surgery is scheduled to last six hours. **How would you proceed?**

**Discussion Outline**

**Case History:**
A sixteen-month old girl is scheduled for posterior sagittal anorectoplasty (PSARP) as an initial step in the correction of her complex cloacal abnormality. She has VACTERL complex, and chronic renal failure.

**Questions:**
What is the VACTERL complex? Of the various anomalies of the VACTERL complex, which have the most impact on your anesthetic technique or plan? How? Is a neuraxial technique contraindicated in a child with VACTERL complex? If so, what kind of defect would prevent you from using a regional technique? What information would you require to make that decision: history? X-ray? MRI? Would you proceed without imaging?

What is a cloacal anomaly? What are the issues to address surgically? How does it impact anesthetic management?

**Case History...continued:**
She was Twin “B” of a gestation complicated by preterm labor and delivery. However, prenatal ultrasound revealed the absence of the left kidney and the bladder, a dysplastic right kidney, and an imperforate anus. Preterm at 33 weeks, she weighed 1800 grams. Her associated malformations included a duplicate uterus and vagina, a fistula connecting the mid-colon to the cloacal cavity, and a right ureter which drained into the uterus.

**Questions:**
In reviewing the patient’s old anesthetic records, you notice your name on the previous record! She had a g-tube placed for poor growth under GA at 5 months of age. Did you discharge her home from the PACU? What is the incidence of post-op apnea in a former pre-term infant at about 50 weeks AGA?

**Case History and physical examination (continued):**
Her previous surgeries had gone well, including an initial diverting colostomy, a subsequent cutaneous ureterostomy, and the g-tube placement. In addition to her complex urogenital malformations, her current problems include latex sensitivity, hypertension, chronic anemia and Type-2 renal tubular acidosis (RTA). Her growth has been poor (6.38 Kg), and she has been followed closely by the pediatric nephrology service. Her medications include amlodipine, vitamin D, Epogen, and prevacid. Physical examination reveals an anxious toddler with the following vital signs: BP 120/72, HR 170, R 28, O2 sat 96%.
Questions:

**Latex Sensitivity:** Why would she be labeled as latex sensitive?

**Hypertension:** How do you define hypertension in a toddler? How common is hypertension in children with chronic renal failure? Explain why these patients are often on an ACE-inhibitor – is this evidence based?

**Anemia:** Why is she anemic? How low would her Hematocrit be to label her as anemic? What Hematocrit value would raise concern for the anesthetic and upcoming surgery?

**Type-2 RTA:** What is type-2 RTA? In browsing the nephrologist’s last clinic note, you see she is labeled as having “Stage 4 CRF” – does that worry you? Isn’t that better than Stage 10? What are the peri-operative implications of this diagnosis? What pre-operative labs would you like and what would they show?

**Preoperative Studies:**
There is an echo from the neonatal period showing a PFO, small to moderate PDA, and an otherwise structurally normal heart. Chemistry from the previous month shows: Na 140, K 4.2, Cl 106, HCO3 19, BUN 18, Creat 1.08, and normal glucose. Hematocrit was 31. MRI of the spine was normal. The surgery is scheduled to last six hours.

Questions:
Would you like any other information before proceeding to the OR? Do you need repeat labs? Mom is tearful and explains her baby is a “difficult stick” – why not check labs after induction? What is the anesthetic plan you explain to her parents? Does it involve an arterial line? Are there any risks that are inherent to this case that should be discussed with the parents? What would be the advantages or disadvantages of placing a caudal versus lumbar epidural catheter?

**Intra-operative care:**
After a smooth inhalation induction, a 22-gauge peripheral IV is placed. Unfortunately, someone forgot to fill up the sevoflurane vaporizer and the patient begins to cough, then have upper airway obstruction, and quickly there is no ETCO2 and you’re unable to mask ventilate the patient. Her oxygen saturations plummet. What do you give to treat her laryngospasm? Is succinylcholine your first choice? Your IV is patent – what dose? She is improved and you go to intubate the patient – do you use muscle relaxant? Why or why not?

The airway is secured and the patient is turned lateral – you perform a one-shot caudal using a 20-gauge angiocatheter. What dose of Bupivacaine 0.25% with epinephrine do you give for the test dose? What dose do you use to bolus for an appropriate surgical level? Do you need to decrease your dose of local anesthetic given this patient’s renal insufficiency? A lumbar epidural is then performed and catheter is secured out of the surgical field.

You struggle at placing an arterial line and the surgeon is pacing the OR room. You abandon the arterial line and the surgery is underway in the prone position. You check a chemistry (venous sample) one hour into the case and it shows: Na 139, K 4.9, Cl 109, HCO3 14, BUN 13, Creat 1.15, lactate 1. How do you proceed? For the next gas, you struggle under the drapes to obtain another venous
sample and the K is 5.5. The EKG appears normal and the patient is hemodynamically stable. Do you treat hyperkalemia? If yes, how?

Her final VBG 1 hour prior to conclusion of the surgery shows: pH 7.16/CO2 30/ HCO3 10.7/ Lactate 1.4/ K 4.2. She has had multiple boluses of NaHCO3. Do you plan to extubate her at the end of the case?

**Post-operative Care:**
She recovered uneventfully in the PACU and the next day the pediatric nephrology service started her on oral bicarbonate replacement therapy. Her parents thank you for the wonderful anesthetic their little girl received.

**References:**