

PBLD – Table #6

Kidney Transplantation in a Pediatric Patient with Fontan Physiology

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Objectives: After discussing this case the learner will be able to:

1. Describe how to evaluate and treat metabolic acidosis in a Fontan patient undergoing renal transplantation.
2. Identify how to evaluate adequate intravascular volume status in a patient with Fontan physiology and oliguria.
3. Construct a decision-making algorithm when faced with a complex patient requiring opposing therapies.
4. Explain why the treatment pathway for a patient status-post renal transplant conflicts with the long-term management of a patient with a Fontan.
5. Formulate a management plan for hypotension in a patient with Fontan physiology.

Case history:

A 9 year old, 18 Kg girl with horseshoe kidney and end-stage renal disease presents for cadaveric renal transplant with complex ureteral reconstruction. The patient also carries a history of tricuspid atresia s/p Fontan completion. Additional past medical history includes hypothyroidism, iron-deficiency anemia, short stature, and mild developmental delay.

Questions:

What are the etiologies of end-stage renal disease in children? What are the indications for renal transplant in children? Describe the anatomy of tricuspid atresia. How is this congenital heart defect surgically repaired? What is the Fontan palliation?

Case history and exam continued:

The patient presented at 2 weeks of age with cyanosis. The diagnosis of tricuspid atresia was made. The patient subsequently underwent a three-staged repair with a Blalock-Taussig shunt as a neonate, a bilateral, bidirectional Glenn procedure at 4 months of age, and then a modified Fontan with an extracardiac, fenestrated conduit when she was 2 years old. She is currently without cyanosis and reported to have good energy. At the time of diagnosis, it was discovered that the patient also had a horseshoe kidney and other urogenital sinus abnormalities. She has had multiple urologic procedures including a Mitrofanoff procedure, ureteral stent and

suprapubic tube placement, right colon urinary reservoir/pouch and ureterostomy. She is currently catheterized every 2 hours, retrieving 16 ml of urine per cath.

On exam, the patient appears pleasant, alert, smiling, and small for her age. She is afebrile with heart rate of 94, blood pressure of 104/65, SpO2 94% in RA. Her cardiac exam reveals a quiet precordium, RRR, S1 & single S2, and a 3/6 harsh holosystolic murmur. Her perfusion was good with full peripheral pulses, pink warm skin, and brisk capillary refill. The remainder of her exam was unremarkable.

Questions:

What other information do you require regarding this patient's cardiac disease? Additional history? Previous or additional studies? What additional information do you want regarding the patient's renal disease? Additional history? Labs? Would you wait for additional information to become available prior to taking this patient to the OR?

Case continued:

The patient was last seen in cardiology clinic 2 months prior to her current presentation. At that time the patient was reported to be doing well from a cardiac standpoint. Her ECG showed sinus rhythm at 74 bpm. There was a left axis deviation at -36 degrees. There was no chamber enlargement or ventricular hypertrophy. ST segments and T waves were normal. A complete echocardiogram demonstrated mild left ventricular dilation with normal systolic function. There was mild MR. The Fontan conduit was widely patent with laminar flow and no thrombus or obstruction. The bilateral Glenn anastomoses were patent with laminar flow into the right and left pulmonary arteries. No fenestration was seen on exam. Prominent collaterals were seen near the descending aorta and branch PA's.

This patient was not receiving dialysis. Her electrolytes from 2 months ago were Na 146, K 4.5, Cl 122, Glu 92, BUN 28, Cr 3.5, P 4.4. A new electrolyte panel was drawn on admission, but the results were not available upon transport to the OR. A CBC on admission revealed a WBC 6.2, Hgb/Hct 13.2/39.6, Plt 186.

The patient was brought to the operating room for induction of anesthesia and surgery.

Questions:

How does Fontan physiology effect the anesthetic management of patients with single ventricle CHD presenting for noncardiac surgery? What complications arise in these patients and how might they affect the operative course? Do you have concerns at induction, and which drugs would you choose? What are important considerations for providing a safe anesthetic? Ventilation strategies? Fluid management? Hemodynamic goals?

Our patient also has end-stage renal disease and is presenting for kidney transplant. What are the concerns regarding anesthesia for patients in kidney failure? Drug selection? Metabolic abnormalities? Fluid management? What are the anesthetic goals for patients undergoing kidney transplant? Fluid management? Hemodynamic control? Pain management?

The two major pathologic processes presenting in this patient may require conflicting therapeutic strategies. And the optimal treatment for one disease may oppose the optimal therapy for the other disease. What are some of these conflicting strategies? How do you balance this conundrum? What would be your plan for anesthesia for this particular patient? Induction, monitoring, maintenance, pain control, ventilator strategies?

Intraoperative course:

The patient received a preoperative sedative of midazolam 0.1 mg/kg IV. She was brought to the operating room and underwent a smooth IV induction consisting of lidocaine 1 mg/kg, fentanyl 5 mcg/kg, and cis-atricurium 0.2 mg/kg. After direct laryngoscopy and tracheal intubation, a radial arterial line and right internal jugular venous central line were placed. Anesthesia was maintained with 0.8 – 1.2% isoflurane and periodic boluses of Fentanyl. Approximately 2 hours into surgery, the results of the preoperative BMP became available. Na 141, K 6.1, Cl 111, HCO₃ 13, BUN 59, Cr 3.6. Around this time, the patient also developed hypotension with BP dropping from 100/60 to 65/35. An I-STAT blood test revealed an ABG 7.03/50/158/13/-18. Repeat Na 140, K 4.4, Glu 116, I-Ca 1.22, Hct 33.

Questions:

What is your differential diagnosis in a patient with Fontan physiology and renal failure who presents with intraoperative hypotension and a primarily metabolic acidosis? What information would you use to sort through the differential diagnosis? How would you treat this patient?

Intraoperative course continued:

Anticipating that the volume resuscitation and afterload support required to maintain adequate perfusion of the transplanted kidney could have an adverse effect on her Fontan hemodynamics, the patient was empirically started on a milrinone infusion. With the discovery of severe acidosis, a transesophageal echocardiogram was performed.

Question:

How might the results of the TEE be helpful?

Case continued:

Treatment of the acidosis and hypotension included volume replacement and sodium bicarbonate administration. Fluid administration was guided by hemodynamic response, CVP readings, TEE findings, and close communication with the surgeons. Following reperfusion of the cadaveric kidney, additional hemodynamic support was provided with dopamine. The initial dose of 5 mcg/kg/min was quickly increased to 15 mcg/kg/min. At this time, an epinephrine infusion of 0.025 mcg/kg/min was added and the dopamine was decreased to its initial dose. The epinephrine gtt was discontinued toward the end of the case. The surgery was complete in 4 hours, and the patient had received NaHCO₃ 2.5 mEq/kg and 35 ml/kg of crystalloid. The final intraoperative blood gas was 7.30/35/109/18/-9. The patient received a total of 10 mcg/kg of fentanyl during the anesthetic and a total of 0.02 mg/kg of hydromorphone for post-operative pain control.

Questions:

What is your ventilation strategy at the end of the anesthetic? Where should this patient be admitted? What would be the goals of post-operative care, specifically volume management?

Case conclusion:

The patient's muscle relaxation was reversed and she was allowed to emerge from anesthesia. She was extubated without event in the operating room and transported comfortably to the Cardiac Intensive Care Unit. Resuscitation continued overnight in the CICU including careful volume and inotrope management, including a transfusion of pRBC. Her metabolic acidosis continued to improve, and her hemodynamics remained stable. Her new kidney gradually improved in function.

Discussion:

Patients with single ventricle Fontan physiology undergoing non-cardiac surgery present many challenges to the anesthesiologist. The Fontan procedure is the final procedure for single ventricle patients in their conversion from parallel to series circulation. Fontan physiology dictates that all pulmonary blood flow is supplied by cavo-pulmonary arterial anastomoses. As such, pulmonary blood flow, and thus, cardiac venous return is dependent of central venous pressure and the transpulmonary gradient. With the resultant increase in venous capacitance and increase in pulmonary vascular resistance, the Fontan heart is very sensitive to preload. Both dehydration and volume overloading can be detrimental to the patient with this physiology.

The clinical course of Fontan patients may be complicated by atrial arrhythmia, protein losing enteropathy, ascites, pleural effusions, plastic bronchitis, thrombosis, and ventricular dysfunction. Even patients who seem to be doing well clinically may not have enough cardiac reserve to tolerate the stress of major surgery and anesthesia. Fontan flow is dependent on unobstructed flow from the central venous circulation through the lungs and heart and out the systemic outflow tract. The anesthetic management must pay close attention to ventilation strategies, volume status, and hemodynamic management.

Generally, positive pressure ventilation will decrease the effectiveness of the Fontan circuit due to the increase in intrathoracic pressure and decrease in the transpulmonary pressure gradient. Spontaneous ventilation should be maintained whenever possible, but not at the expense of hypoventilation with its accompanying atelectasis and hypercarbia, both of which will increase PVR and decrease Fontan flow. When using PPV, one should pay particular attention to using only enough PEEP to maintain FRC and thus minimize PVR. Decreased inspiratory times and lower respiratory rates and slightly higher tidal volumes may allow more time for increase blood flow during expiration.

The single ventricle is, by definition, under more workload than a two-ventricle heart, because the one ventricle is providing the energy necessary for both the systemic and pulmonary circulations. Even in the face of seemingly normal systolic function, Fontan hearts often exhibit some diastolic dysfunction. With the resultant increase in venous capacitance and increase in

pulmonary vascular resistance that occurs, the Fontan heart is very sensitive to preload. Both dehydration and volume overloading can be detrimental to the patient with this physiology. Euvolemia is the goal of intraoperative fluid management. In addition, afterload reduction is a mainstay of care for the single ventricle heart. This both decreases the work on the ventricle and promotes antegrade flow through the entire system.

Patients with end-stage renal disease also pose their own set of unique challenges to the anesthesiology team. Volume status and metabolic balance may depend on timing of the patient's last dialysis. Hyperkalemia is common and can lead to ventricular dysrhythmia. Severe hyperkalemia is an indication for dialysis prior to surgery in patients on a dialysis regimen for renal failure. Metabolic acidosis may be due to the kidney's inability to retain bicarbonate. Renal patients often present with systemic hypertension and subsequent cardiac pathology including heart failure and valvular disease.

The anesthetic management of these patients for renal transplant must include attention to drugs that are metabolized and excreted by the kidney. The dosages of these medications must be adjusted where indicated in order to prevent prolonged and/or exaggerated effects. Some drugs can be avoided altogether in favor of those that are not metabolized by the kidney. For example, the use of cis-atricurium in place of pancuronium or rocuronium may avoid prolonged muscle relaxation and delayed emergence.

In an attempt to avoid delayed graft function and improve patient outcomes, the intra-operative and post-operative management of renal transplantation includes aggressive volume resuscitation often resulting in fluid overload. This can lead to pulmonary edema and respiratory failure necessitating mechanical ventilator support. The excessive fluid can also lead to cardiac compromise in susceptible individuals. The use of pressors to maintain adequate renal perfusion pressures is common practice.

In the patient presented here, the common therapies employed for renal transplantation may oppose the optimal management usually employed for Fontan patients.

1. While spontaneous ventilation is optimal for the Fontan circulation, it is not a reasonable expectation for this surgery. This patient's surgery was performed via a large right abdominal incision, and invasive positive pressure ventilation was certainly indicated. When using PPV in a Fontan patient, care must be taken to avoid excessive PEEP and MAP and to allow for maximum time. It is important to maintain normocarbida and avoid elevated PVR. However, hyperventilation may decrease blood flow through the Glenn by vasoconstricting the cerebral vascular bed. Oxygen should be administered to both maintain good oxygenation and keep PVR low. As our patient had stabilized by the end of surgery, it was decided to return the patient to spontaneous ventilation.
2. While aggressive fluid administration is standard therapy during renal transplantation, it may be detrimental to patients with single ventricle, Fontan physiology. The heart in

the Fontan patient carries a higher workload than a two-ventricle heart and may have limited functional reserve. The acute volume overload can tip the single-V heart into heart failure. Anticipating that our patient was going to require significant volume loading, we chose to empirically treat with milrinone in order to maintain good systolic and diastolic function. We also made a somewhat less aggressive fluid management plan, trying to balance the needs for renal perfusion and Fontan function.

3. Our patient developed hemodynamic changes and severe acidosis. The differential causes for these findings were broad, and include both renal/metabolic and cardiovascular etiologies. Renal failure can certainly lead to acidosis via metabolic derangements, such as bicarb wasting. The Fontan circulation will not function optimally in the face of severe acidosis. Myocardial function can be directly affected. The Fontan circulation can also be limited by pulmonary hypertension caused by acidosis. However, in this patient, an acutely failing Fontan could also lead to the findings of hypotension and acidosis. Consultation with pediatric cardiology and the use of intraoperative TEE helped to rule out a failing Fontan as the cause of these metabolic and hemodynamic findings. The TEE also was very valuable in guiding fluid management during the remainder of the surgery.
4. The use of inotropic drug infusions was indicated in order to maintain appropriate perfusion pressures in the transplanted kidney. Again, it was important to balance this against the desire to not provide excessive afterload to the single ventricle.

The patient presented here brought together the challenges of two very complex pathophysiological processes. Providing anesthesia for a patient with tricuspid atresia and Fontan physiology undergoing a renal transplantation for end-stage renal failure requires an understanding of a number of physiologic and pharmacologic principles. The management was further complicated by the fact that these two diseases necessitated therapies that were at times in opposition of each other. A carefully thought out plan made in close communication with the surgeons and consulting specialties, and the readiness to balance the anesthetic to the needs of the changing circumstance were key in the delivery of a safe and successful anesthetic.

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