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I Thought Three Pumps Were Bad but ThisPatient With a Failing Fontan for Emergency Exploratory Laparotomy

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Stem Case and Key Questions Content

9 year old male presents for Ross-Konno procedure with cardiopulmonary bypass. He has a past history of discrete left ventricular outflow obstruction and coarctation of the aorta s/p aortic valvuloplasty and coarctation repaired 4 years ago. He now has severe sub-aortic stenosis with a peak gradient of 85 and dyspnea and fatigue with exertion. He is otherwise healthy, though he complains of a recent cough that his PMD attributed to a mild viral upper respiratory illness. Labs are all within normal limits and echo is notable for the findings as above.

Questions:
What is the Ross-Konno procedure and what are your pre-operative considerations?
What are your concerns regarding his recent upper respiratory illness? Would you postpone the case?

Case discussion:
Induction, placement of lines, and pre-bypass period is uneventful. Cardiopulmonary bypass is initiated and is unremarkable other than a long total CPB time at 4 hours. Upon weaning from bypass after completion of Ross-Konno repair, right ventricular dilation and severe systemic hypotension is noted immediately.

Questions:
What would your concern be? What are some immediate post-operative complications of the Ross-Konno procedure?

Case discussion:
The surgeon states the problem is likely the length of the pulmonary graft and the patient is placed on cardiopulmonary bypass again for further repair. After repair, attempts to wean a second time are complicated by severe tricuspid regurgitation on TEE of unknown etiology. Pt is placed back on CPB for a third time for tricuspid valve repair.

Questions:
Is there a physiologic difference between one cardiopulmonary bypass runs and multiple runs? How does overall time on cardiopulmonary bypass impact post-operative complications and bleeding?

Case discussion:
He is successfully weaned from CPB on the third attempt and post-operative TEE demonstrates good atrio-ventricular valve and ventricular function. As you are weaning off of CPB you start transfusing platelets. He does continue to have significant bleeding after bypass wean and you begin to give additional blood products as well as cell saver. Protamine is given. Shortly after protamine is given, oxygen saturations are noted to fall to 80%. The patient is hypotensive with mean arterial pressures of 40. You check your endotracheal tube and it is noted to be filled with pink, frothy fluid.

Questions:
What is your next step? Is there utility to suctioning the endotracheal tube? How do you treat pulmonary edema in the OR?
What is your differential diagnosis for pulmonary edema?
How would your monitors help to differentiate between these causes?
Is there utility to routine administration of platelets while weaning off CPB?
What is TRALI? What is the incidence?
What are risk factors for the development of TRALI?
Could this be a protamine reaction?

Case discussion:
Bleeding is improving, but he still remains hypotensive. Hematocrit on most recent ABG is 29. The surgeon initiates a discussion about possible delayed sternal closure.

Questions:
What are considerations in considering delayed sternal closure? What are the associated risk factors?
How could delayed sternal closure benefit the patient?
If you are concerned about TRALI how would you handle further administration of blood products? What would your workup be with those blood products already administered?
If this is a protamine reaction, what would be your advice for further cardiac surgery? What is the usual timeline to resolution of symptoms with a protamine reaction?
How would your post-operative ventilation strategy differ depending on your differential diagnosis?

Model Discussion Content

The Ross-Konno procedure is a surgical procedure performed in patients with left ventricular outflow obstruction wherein the aortic valve is replaced with the patient's native pulmonary valve and a homograft is utilized for the pulmonary valve. This avoids the need for chronic anticoagulation and is an option for infants and children with severe multilevel aortic stenosis. Complications of the procedure include autograft insufficiency, right ventricular outflow tract obstruction, and bleeding.[i]

Post-operative pulmonary and cardiovascular insufficiency is common after cardiopulmonary bypass for congenital heart surgery. CPB initiates a systemic inflammatory response that has a profound effect on end-organ function. This inflammatory response involves activation of

complement, humoral and cellular cascades. There are several factors that contribute to this, including mechanical and shear stress, hemodilution, cellular activation due to contact with foreign material, and hypothermia. Studies have demonstrated that the pulmonary system seems to be at greatest risk of injury after cardiac surgery with CPB, though multi-organ dysfunction is prominent, especially with prolonged CPB runs and complicated surgical procedures.[2] "Pump-lung" refers to the post-operative pulmonary injury that is common after CPB, and manifested by decreased pulmonary compliance, atelectasis, pulmonary edema, microvascular leak and increased alveolar-arterial oxygen gradient.[2] Though pulmonary injury is one of the major causes of post-operative morbidity in children undergoing CPB, end-organ dysfunction is also present in the form of neurologic injury, myocardial damage, renal failure, and gastrointestinal complications.

There is also a strong correlation between the initiation of an inflammatory response and the development of a coagulopathy post-bypass. In addition, cardiac surgery still consumes 20% of the nation's blood supply, and most children undergoing CPB will necessitate a blood transfusion. Blood transfusion reactions are oftentimes difficult to discern under general anesthesia. Transfusion reactions include acute hemolytic reaction, febrile non-hemolytic reaction, bacterial contamination, allergic reaction, transfusion-related acute lung injury (TRALI), and transfusion-associated circulatory overload. Acute transfusion reactions occur in anywhere between 0.2-10% of blood transfusions.[4] One recent study cited an overall risk of adverse transfusion reaction of 1.6% in a pediatric ICU population. [4] The most common reaction cited was a febrile non-hemolytic reaction, which is attributed to the recipient's antibodies binding to donor white blood cells and/or accumulation of complement or cytokines in blood product. The second most common reaction was a minor allergic reaction, due to type 1 hypersensitivity, followed by isolated hypotension and bacterial contamination. More severe complications such as anaphylactic shock and TRALI are exceedingly rare.

TRALI has now become the leading cause of transfusion-related death in the United States. All blood products have been implicated in the development of TRALI. The incidence of TRALI is currently noted to be 1:5000 blood and blood-containing components, 1:2000 plasma containing components and 1:7900 units fresh frozen plasma and 1:432 whole blood derived platelets. [5] Increased pulmonary micro-vascular permeability with increased protein in edema fluid is the hallmark of TRALI, but the diagnosis remains clinical. Increased awareness has lead to improved diagnosis, however overall TRALI continues to be under-diagnosed and under-reported.

Two possible causes of TRALI have been hypothesized. One is that leukocyte antibodies in donors activate recipient neutrophils in pulmonary capillaries and may cause pulmonary damage and capillary leak. The other hypothesis is that TRALI is caused by two events, one being linked to the patient's condition at the time of transfusion that leads to the sequestering of neutrophils in the lungs. The second event is the transfusion of biologically active substances such as lipids or cytokines that activate those neutrophils causing lung damage and capillary leak.[6] Risk factors for TRALI include sepsis, high peak inspiratory pressures, severe liver

disease, cardiopulmonary bypass, hemataologic malignancy, cardiovascular disease, and mechanical ventilation.[5-6] TRALI should be suspected with the development of acute lung injury within 6 hours of a transfusion, however, in the presence of other risk factors the etiology of the acute onset of pulmonary edema can oftentimes be difficult to discern. Treatment is largely supportive, and mechanical ventilation should be aimed at a low tidal volume strategy similar to the treatment of acute respiratory distress syndrome. Diuretics are oftentimes used in post-operative management. No evidence exists in support of the use of corticosteroids. Suspected transfusion reactions should be reported to the blood bank for further identification and removal of suspected donors with antibodies.[6]

In addition to TRALI, protamine has been implicated in the development of post-bypass noncardiogenic pulmonary edema, though this reaction is rare. [7] Protamine reactions have many clinical presentations, and risk factors include prior reactions or exposure to the drug, allergies to other medications, prior exposure to protamine-containing insulin, and fish allergy.[8] The most common reaction to protamine is transient systemic hypotension, and in one pediatric study this was the only identified adverse event, with an incidence of 2.88%.[8] More severe complications have been described in adult studies, including severe pulmonary hypertension with associated right ventricular failure, bronchospasm, and non-cardiogenic pulmonary edema. Treatment of suspected reaction is largely supportive, with consideration given to administration of diphenhydramine and corticosteroids. Management of a patient with a previous suspected allergic reaction to protamine is difficult, as currently there are no available alternatives to protamine. Consideration should also be given to pre-operative allergen testing in the case of a patient with a prior suspected protamine reaction. [9]

Delayed sternal closure (DSC) has long been a consideration in neonatal and infant cardiac surgery. This is primarily due to concerns that sternal closure in some patients decreases cardiac output and diastolic filling. Reasons for DSC include prolonged cardiopulmonary bypass time, pulmonary edema, complex surgical procedures, small chest cavity, and uncontrolled bleeding. DSC has been associated with overall increased post-operative infection rate, though not an increased rate of mediastinal infection.[10] This increased post-operative infection rate may also be due to numerous other co-morbidities and complications in the patient population necessitating DSC, and it is unclear whether this is directly related to the open sternum. Additionally, closing the sternum alters lung mechanics and respiratory compliance. If concern for ability to ventilate exists in the setting of high airway pressures, DSC may be considered. Overall, the decision for DSC varies greatly by center and remains an option for those patients in whom hemodynamic instability with chest closure remains a concern.

In summary, intra-operative pulmonary edema after cardiac surgery and CPB has many potential etiologies and can represent a diagnostic challenge. As most cardiac surgeries necessitate the transfusion of blood products, it is vital to understand the risks of blood transfusion and identification of intra-operative reactions under general anesthesia.

References

1. Hraska V, et al. Ross-Konno operation in children. Multimedia manual of cardiothoracic surgery 2008 (Jan): 1-6
2. Brix-Cristensen, V. The systemic inflammatory response after cardiac surgery with cardiopulmonary bypass in children. Acta Anaesthesiol Scand 2001 (45): 671-9.
3. Iyenger A et al. Association of complications with blood transfusions in pediatric cardiac surgery patients. Ann Thoracic Surg 2013 (96): 910-16.
4. Gauvin, F. et al. Acute transfusion reactions in the pediatric intensive care unit. Transfusion 2006 (46): 1899-1907.
5. Toy, P et al. Transfusion-related acute lung injury: Definition and review. Critical care Med 2005 (33): 721-6.
6. Vlaar, A Jeffermans, N. Transfusion-related acute lung injury: a clinical review. Lancet 2013 (382): 984-94.
7. Wong, M Halliwell J. Non-cardiogenic pulmonary edema in a cardiac surgery patient: never a welcome sight for the anesthesiologist. J Cardiothoracic and vascular anesthesia 2006 (26): 178-9.
8. Siefert, H et al. Adverse events after protamine administration following cardiopulmonary bypass in infants and children. Anes Analg 2003 (97) 383-9.
9. Urdaneta F et al. Non-cardiogenic pulmonary edema associated with protamine administration during coronary artery bypass grafting surgery. J Clinical Anesthesia 1999 (11): 675-681.
10. Erek E, et al. Indications and risks of delayed sternal closure after open heart surgery in neonates and infants. World Journal for Pediatric and Congenital Heart Surgery 2012 (3): 229-34.