“Thinking outside of the box”
Perfusion management and myocardial protection strategy for a patient with sickle cell disease

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Disclosures

• I have no disclosures
Overview

- Background on Sickle Cell Disease (SCD) and End Stage Renal Disease ESRD.
- Unique challenges for managing this patient at Vanderbilt University Medical Center.
- Strategies utilized to manage this patient on Cardiopulmonary bypass.
- Priming and Circuit modifications for this patient.
- The clinical course for this patient.
Sickle Cell Disease

• SCD is an autosomal recessive disease that affects approximately 1 of 500 African Americans in the United States.
• The $\beta$-globin gene is found on chromosome 11 and the Hemoglobin S (HgS) allele involves the substitution of valine for glutamic acid at the sixth amino acid position.
Sickle Cell Disease

- SCD is characterized by recurrent episodes of ischemia-reperfusion injury to multiple vital organs through a process termed sickle vaso-occlusion.
Sickle Cell Disease

• Triggers of sickling:
  – Hypothermia
  – Acidosis
  – Hypoxemia
  – Increased levels of 2,3 DPG
  – Hypertonic solutions
  – Capillary stagnation
End Stage Renal Disease

- The vaso-occlusive process in the kidneys leads to renal ischemia, micro-infarcts, and tubular abnormalities.
- If the tubular dysfunction occurs in the distal tubule, the ability to remove potassium becomes impaired and leads to hyperkalemia.
- In the kidneys, vaso-occlusion leads to ESRD and dialysis dependence.
Patient

- A 60-year-old African-American male with a height of 188cm and weight of 75kg and BSA of 1.98m$^2$.
- Diagnosed SCD with severe mitral valve insufficiency, congestive heart failure, left ventricular hypertrophy, moderate pulmonary hypertension, severe anemia, and ESRD requiring peritoneal dialysis.
- Cardiac echo displayed possible calcified mass leaflets suggestive of old endocarditis.
Standard Mitral Valve Approach at VUMC

• Minimally invasive approach through a right thoracotomy.

• Myocardial protection is achieved by cooling the patient to 28°C, and fibrillating the heart and keeping it empty during the procedure.
Pre-Operative Discussion

• Hurdle #1 surgical approach…..we can’t cool to 28°C and perform the standard minimally invasive approach via right thoracotomy.

• Hurdle #2 myocardial protection….we can’t give cold blood with high K⁺ due to sickling and vaso-occlusion.

• Hurdle #3 myocardial protection….how can we arrest the heart and minimize K⁺ use.
Pre-Operative Discussion

• Hurdle #1- It was decided to perform a median sternotomy with bicaval cannulation and stay normothermic.

• Hurdle #2- Cardioplegia discussion included possible continuous high K⁺ warm blood but was ruled out secondary to the inability to clear K⁺.

• Hurdle #3- It was decided that del Nido cardioplegia was the best option for this patient in terms of minimizing the total K⁺ dose.
del Nido Cardioplegia

- Normally given in a 1:4 ratio of blood to crystalloid.
- The crystalloid component of del Nido contains
  - one liter of Plasmalyte A which has a similar to composition to plasma and is pH balanced at 7.4
  - 16.3 milliliters of 20% Mannitol - osmotic properties can reduce cardiac myocyte edema
  - 4 milliliters of 50% Magnesium sulfate - calcium channel blocker
  - 13 milliliters of 8.4% Sodium bicarbonate - a buffer to help maintain intracellular pH
  - 13 milliliters of 2 mEq/ml Potassium chloride - rapid depolarized arrest
  - 13 milliliters of 1% Lidocaine - sodium channel blocker
Perfusion Strategy and Goals

- Goal: Avoid sickling and vaso-occlusion.
  - Strategies
    - Check in the room baseline Hct and HgS
    - Perform exchange transfusion immediately after initiating bypass- Primed with blood/blood products
    - Check new Hct and HgS after transfusion
    - Get HgS < 10%
    - Maintain warm cardiopulmonary bypass (36°C)
    - Keep SVO$_2$ > 80% (C.I. 2.5 to 2.9)
    - Target MAP > 60 (minimal vasopressor usage)
    - Maintain pH 7.40 – 7.45
    - Cardioplegia strategy: Flush with crystalloid del Nido (warm) then cold del Nido cardioplegia
The circuit was initially primed using 2,000 mL of Plasmalyte-A.

Four units of type O+ packed red blood cells were washed in a cell saver. The washed packed red blood cells were used to “chase” the Plasmalyte through the circuit.

Four units of fresh frozen plasma, 50 ml’s 25% mannitol, 5,000 units of heparin, 50 meq of NaHCO₃ 8.4%, and 500 mg of methylprednisolone were added to the “blood” prime.
Circuit Modification

- A $\frac{1}{2} \times \frac{1}{2} \times \frac{1}{4}$ ‘Y’ connector was inserted prior to the venous reservoir in with an empty plasmalyte bag attached via $\frac{1}{4}$ inch line to the $\frac{1}{4}$ inch part of the “Y” in order to perform an exchange transfusion as cardio-pulmonary bypass was initiated.
Clinical Course

• In the operating room, general anesthesia was administered with endotracheal intubation and the patient was prepped and draped. A trans-esophageal echo probe was inserted for assessment of ventricular function and valvular anatomy.

• The patient was anemic upon entering the operating room with a baseline hematocrit (HCT) of 17 with a HgS concentration of 16.7%
Clinical Course

- Twenty-three thousand units of heparin (300u/kg) were administered.
- The ascending aorta was cannulated with a 21fr cannula.
- A 14fr antegrade root cannula with vent was placed.
- The patient was bicavally cannulated with 24fr and 28fr angled plastic tip venous cannula for the SVC and IVC, respectively.
Clinical Course

- Once an activated clotting time of > 480 seconds was verified, CPB was initiated and as blood was transfused to the patient via the heart lung machine, an exchange transfusion was performed with approximately 750 ml’s of patient blood removed via the ¼ inch line “Y” connector to the empty plasmalyte bag.

- Initial HCT on CPB after the exchange transfusion was CPB 22% and the HgS concentration decrease from 16.7% to 9.1%.
Clinical Course

- The aorta was cross-clamped proximal to the arterial cannula and 1L of del Nido cardioplegia solution without the blood component was administered as a single dose at a flow of approximately 300 ml’s/min in an antegrade fashion.
- The first 250ml’s were delivered warm and then was delivered cold at approximately 6°C for the remaining 750ml’s.
- Full electrical arrest was achieved at approximately 700 ml’s total volume and was maintained throughout the surgical procedure with a single dose
Clinical Course

- The baseline K$^+$ was 5.1 meq/L and after cardioplegia the K$^+$ was approximately 8.5 meq/L. A combination of CUF and Z-BUF was initiated. The final K$^+$ coming off bypass was 5.3 meq/L.
- Flows were utilized to maintain a Cardiac Index between 2.5 to 2.9 in order to achieve the targeted MAP between 60 mmHg and 70 mmHg with minimal vasopressor usage.
- Sodium bicarbonate was given as needed throughout the case to maintain a pH of 7.40 to 7.45.
- After an arrest time of 120 minutes the mitral valve replacement was completed.
Clinical Course

• Prior to cross clamp removal, The Quest system was then flushed with 500ml’s of warm NS.
• 100mg of Lidocaine and 2mg Magnesium Sulfate was delivered per protocol prior to cross clamp removal.
• Warm blood was then given at 300ml/min via the aortic root cannula. After approximately 1500ml’s of warm blood was delivered the patient spontaneously converted to normal sinus rhythm, and the cross clamp was released.
• Five minutes after cross clamp removal 1g CaCl$_2$ was given.
• The patient did not require defibrillation or cardioversion.
Clinical Course

• TEE studies of wall motion and EF were unchanged compared to pre operative study.
• Total myocardial ischemic time was 120 min. After successful de-airing of the heart the patient was slowly weaned from CPB. Total CPB time was 173 min.
• The patient was transferred to stepdown ICU on post-op day 6 and discharged on post-op day 22.
Take Home Points

• Know what your patients baseline HCT and HgS values are and have a plan.
• Adjust your plan for CPB to manage your patient without hypothermia.
• Have a plan in place to provide myocardial protection that may not be your standard protocol.
Questions?