CPB FMEA #9: Hemodynamic instability due to the presence of hypertrophic cardiomyopathy (HCM), idiopathic hypertrophic subaortic stenosis (IHSS) and its variants.

The AmSECT Safety Committee

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Friends-

This week’s FMEA deals with hypertrophic cardiomyopathy (HCM) patients. This is not the acquired form of hypertrophy or dilated cardiomyopathy. Rather it is a congenital form of HCM seen mostly in children and young adults. In the city where I live it probably accounts for the death of at least one young athlete annually. My experience with HCM patients goes back over four decades. In the early years the outcomes of these operations were frequently disastrous; the patients often died in the OR. As the years passed, better ventricular support gradually became available. Today nobody with HCM dies in the OR. They spend time on mechanical support in the ICU instead. Over the years, as ventricular support was being developed, I worked on preventing the need for mechanical support by devising a way to protect hypertrophic hearts. Unfortunately this was a trial an error process. There was a lot written about HCM and its variants, but there were no real specifics for perfusionists concerning methods to protect these hearts during surgery; at least that I was aware of. As time passed the method described in this FMEA developed from my experience and was usually successful for infants, children and adult patients. Of course these methods require cooperation on the part of the surgeon. But over the last 20 years I have been older and more experienced than most surgeons with whom I worked. So they would usually agree to the methods I requested. I don’t claim that my method is the best. Only that it is the best that I know of. I should also mention that several excellent suggestions have been added to this FMEA by the retired perfusionists who help to review these FMEAs for me. Let’s make this FMEA even better by combining all our experience to protect these fragile hearts and patients. Now it is time for you to offer your own experience with these patients. Send any suggestions you may have to me; [garygrist@comcast.net](mailto:garygrist@comcast.net) .

This week’s Failure Mode is below:

1. FAILURE: Hemodynamic instability due to the presence of hypertrophic cardiomyopathy (HCM), idiopathic hypertrophic subaortic stenosis (IHSS) and its variants.

II. Potential Effects of Failure:

1. Hemodynamic instability

2. Iatrogenic myocardial damage

3. Failure to wean from CPB

4. The need for extended ventricular support

5. Death

1. Potential Cause of Failure:
2. HCM is a form of congenital [cardiomyopathy](file:///C:\Users\Gary%20Grist\Documents\1.07%20REVISION\001105.htm); a condition in which the heart ventricular muscle becomes abnormally thickened without a compensating increase in perfused capillary density (PCD).
3. The thickening makes it harder for the heart to work and the reduced PCD impairs oxygen distribution to the myocytes.
4. HCM causes the size of the ventricular chamber to shrink. So the heart must work harder to pump a normal amount of blood per minute because there is a smaller stroke volume.
5. The thickening of the heart muscle may, at times, completely block the normal flow of blood out of the heart.
6. This is called IHSS and is a variant of HCM that involves the thickening of the interventricular septum.
7. HCM may also make it harder for the heart valves to work by obstructing their function.
8. The condition is seen in people of all ages.
9. Younger people are likely to have a more severe form of HCM.
10. In people over age 60, HCM is often associated with mild hypertension.
11. Patients with this heart condition have extremely fragile myocardium that must be carefully protected.
12. Despite the hearty appearance of the thickened myocardium, these hearts are quite fragile and do not tolerate periods of hypotension from general anesthesia or ischemia such as during aortic cross clamping with cardioplegia.
13. After cross clamping, the excessive bulk of the myocardium makes it difficult for the cold cardioplegia to cool the heart effectively.
14. The ambient temperature within the chest and relatively warmer venous blood returning to the heart and any pulmonary collaterals add to the difficulty in keeping the myocardium cool.

IV. Interventions to Prevent or Negate the Failure:

PRE-EMPTIVE MANAGEMENT: Do not attempt surgery on these patients without the availability of adequate mechanical ventricular support and necessary back-up personnel.

1. Myocardial protection is difficult with HCM.
2. Management on CPB involves maximizing oxygen delivery to the heart, optimizing the electrolytes and maximizing myocardial protection during cross clamping.
3. Weaning from CPB may prove difficult.
4. External defibrillation pads should be attached prior to prep and drape.
5. Hypotension during anesthesia induction can reduce coronary artery perfusion and cause fibrillation.
6. Consideration should be given to using a blood prime to prevent excessive hemodilution.
7. Use bi-caval cannulation to reduce warm blood return to the heart.
8. Increase coronary perfusion pressure to at least over 40 mmHg before and after cross clamping.
9. Expect to give cardioplegia at a greater frequency and possibly in larger amounts.
10. This may require hemoconcentration to reduce hemodilution from cardioplegia crystalloid.
11. Patients with a secondary diagnosis of coronary artery disease, even if it is not clinically significant, may benefit from frequent retrograde CP administration.
12. The protective effect of single dose CP solutions like HTK in HCM patients is unknown.
13. If cooling the patient, delay cross clamping until the target temperature is reached to aid in cooling the myocardium.
14. Prior to cross-clamp removal, increase the K+ to 4.5 mEq/L and the iCa to 1.4 mmoles/L.
15. Use 100% oxygen in the oxygenator sweep gas to maximize capillary oxygen distribution vectors in the myocardium particularly if the hemoglobin is low.
16. Increase the hematocrit to 40% before termination of CPB. This will require the availability of extra PRBC units, particularly in the larger patient.
17. Anticipate the need for higher than normal ventricular filling pressures; =/> CVP 15 mmHg.
18. Be prepared for ventricular support in the form of an intra-aortic balloon pump, VAD/biVAD or ECMO.

MANAGEMENT:

1. Failure to wean from CPB will necessitate ventricular support for an indeterminate period.
2. If patient weans from CPB and transits to the ICU, be prepared for extracorporeal support (ECPR) should patient have sudden cardiac arrest in the post-op period.

V. Risk Priority Number (RPN): (select the number from each category that you feel best categorizes the risk).

A. Severity (Harmfulness) Rating Scale: how detrimental can the failure be:

1) Slight, 2) Low, 3) Moderate, 4) High, 5) Critical

(The severity problems that this failure causes are usually 5) critical.)

B. Occurrence Rating Scale: how frequently does the failure occur:

1) Remote, 2) Low, 3) Moderate, 4) Frequent, 5) Very High

(Occurrence is usually 2) low.)

C. Detection Rating Scale: how easily the potential failure can be detected before it occurs:

1) Very High, 2) High, 3) Moderate, 4) Low, 5) Uncertain

(Although the diagnosis is usually known, it is difficult to know if any particular HCM heart will tolerate anesthesia, CPB and myocardial protection. So the ability to detect the failure to wean from CPB is 4) low.)

D. Patient Frequency Scale:

1) Only a small number of patients would be susceptible to this failure, 2) Many patients but not all would be susceptible to this failure, 3) All patients would be susceptible to this failure.

(This is a rare diagnosis. So the Patient Frequency RPN should be a 1.)

Multiply A\*B\*C\*D = RPN. The higher the RPN the more dangerous the Failure Mode.

The lowest risk would be 1\*1\*1\*1\* = 1. The highest risk would be 5\*5\*5\*3 = 375. RPNs allow the perfusionist to prioritize the risk. Resources should be used to reduce the RPNs of higher risk failures first, if possible. (The RPN for this failure is 5\*2\*4\*1 = 40.)