CPB FMEA #35 Failure to prevent post pump chorea from developing after CPB using deep hypothermic circulatory arrest (DHCA).

This is another one of those failures for which there is no known etiology: post pump chorea after deep hypothermic circulatory arrest (DHCA). Although this is mostly seen in small children, it can occur in all ages including adults. While most frequently associated with DHCA, post pump chorea can occur after a variety of CPB procedures. In my opinion, the reason it is seen more commonly in pediatrics is because children are routinely cooled to lower temperatures for longer periods than most adults.

Since this is such a rare and mysterious problem, some would say that the average adult perfusionist need not worry about it. I disagree. This is just one more failure that perfusionists should educate themselves about to try to prevent whenever possible. The outcome can be devastating to a patient and heart breaking to a guilt-ridden perfusionist who has no idea why things went so badly days after an uneventful bypass procedure.

One can search the internet for “post pump chorea” to find letters from affected family members seeking advice and recourse. I have quoted one of those letters below because it best expresses a family’s anguish. I think the author may even be a doctor (MD). It was posted in 1997, but letters posted as late as 2015 can be found.

<http://www.medhelp.org/posts/Neurology/postpump-ch>

“Posted by CCF MD on November 26, 1997 at 12:01:09: In Reply to: postpump chorea posted by CBaeta on October 26, 1997 at 17:06:52: My 7 year old son underwent his second open heart, with cpb and hypothermia, on July 28th of this year. He went into the surgery a very neurologically healthy and bright little boy. He has been diagnosed with Postpump Chorea. We have a host of excellent doctors but no one knows how this happens or what to do now except wait. Our family is suffering terribly. My son now has dystonia, hypotonia, occular [sic] motor atresia, speech problems, academic problems, personality changes, is very emotional, and ADHD. He had a normal EEG and MRI two weeks post op. Would we expect these to change? What can we do? What is his long term prognosis? I have read everything I could find on this subject but can not [sic] figure out why this happens only in the very young, usually infants yet occurred in my seven year old. Does this happen in adults? Why or why not? I would love to hear from anyone with any interest or knowledge on this subject.”

How much of a role does perfusion practice play in post pump chorea? Nobody knows. Back in the 1970s and 80s we used bubbler oxygenators; essentially a hyperoxia/pH-stat process. We did not see much post pump chorea. Our procedures were not as complicated then, either. Then in the late 1980s, I noticed an uptick in certain problems, including chorea. Co-incidentally, this uptick occurred at about the same time as the changeover to membrane oxygenators and the development of higher risk procedures. However, the post pump chorea was not confined to the more complicated cases. Others noticed an increase in chorea during this time period as well (Wong, Barlow et al, 1992).

There were many good aspects to membrane oxygenators. Membranes were capable of using alpha-stat gas control with much lower paO2 levels. Back then I did not correlate the increase in chorea and other problems to using alpha-stat or lower sweep gas FiO2. (I tried to blame Anesthesia for any mysterious problems and Anesthesia tried to blame me!) As time passed I realized that the majority of pediatric patients, usually being cooler, tended to do better overall with pH-stat. And the majority of adults, usually being less cool, did better with alpha-stat. But that is by no means written in stone. It varies from patient to patient.

I don’t believe that there have been any new answers for ‘CCF MD’ since the 1997 letter was written. The closest I can come is this FMEA. It is written based on my own experience and the few articles available. If you disagree with my conclusions let me know and we will discuss it.

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FAILURE:

Failure to prevent post pump chorea from developing after CPB using deep hypothermic circulatory arrest (DHCA).

EFFECT:

1. The encephalopathy ranges from transient and mild to persistent and severe.

2. Develops within two weeks after CPB.

3. Causes delayed development in:

a. memory

b. attention

c. language skills

d. motor skills

e. IQ

4. Neurological symptoms:

a. choreoathetosis

b. dystonia

c. hypotonia

d. obtundation

5. Death

CAUSE:

1. Most commonly occurs in children but does occur in adults. (\* If the patient is a child the occurrence is about 1.2% of cases, so the Occurrence RPN should be increased to 2.)

2. The delayed onset may reflect the time required for the diffuse cellular atrophy to mature.

3. High risk patient characteristics:

a. often have pre-existing developmental delay.

b. undergo longer CPB times at lower temperatures.

c. are more likely to have had DHCA with a cooling time <20 minutes and alpha-stat pH management during CPB (Levin et al. 2005).

4. Exact etiology and pathophysiology is unknown, but it is thought that chorea and similar neurological complications most likely result from non-embolic hypoxia-ischaemia within diffuse cellular areas rather than global hypoperfusion of the brain.

5. Area of damage is thought to be near the basal ganglia, but multiple EEGs and MRIs have not confirmed this.

6. Hemodilution during CPB reduces the amount of hemoglobin-bound O2 for use.

7. At profound temperatures, the affinity of O2 for hemoglobin greatly increases and impairs the disassociation of O2.

8. Low CO2 concentration (alpha-stat gas control) further impairs the disassociation of O2 from hemoglobin.

9. Dissolved O2 is the primary source of O2 for brain tissue at profound hypothermia (Dexter 1997).

10. Low FiO2 sweep gas will fully saturate hemoglobin but reduces dissolved O2. This may result in diffuse cellular hypoxia even in the presence of hyperoxemia. (du Plessis et al, 1995).

PRE-EMPTIVE MANAGEMENT:

1. pH-stat gas strategy with hyperoxia during CPB cooling maximizes capillary perfusion and shifts the oxy-hemoglobin dissociation curve to the right for better oxygen release to the tissues. This allows maximum tissue oxygen loading prior to arrest.

2. Pre-arrest preparation by oxygen loading of tissues in combination with pH stat gas strategy can delay the conversion from aerobic to anaerobic metabolism, thereby extending the safe DHCA time.

3. pH-stat gas strategy with hyperoxia during CPB cooling results in 85% less acid production during the hypothermic arrest period than normoxia with alpha-stat gas strategy. (Pearl at al, 2000).

4. Traditional methods of DHCA rely on reaching a specified temperature without consideration of impaired O2-hemoglobin disassociation, dissolved O2 utilization or tissue oxygen loading.

5. Cooling for at least 20 minutes, reaching a temperature of 18C and a pvO2 of 300+mmHg may provide the most favorable conditions for DHCA of the brain.

6. Rewarming with normoxia may reduce the risk of reperfusion injury in an acidotic brain after a prolonged circulatory arrest.

MANAGEMENT:

Since post pump chorea develops several days after surgery and because there is no clear cut way to determine if it will develop during CPB, there is no perfusion management strategy.

RISK PRIORITY NUMBER (RPN):

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

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

(I would give this failure a Moderate RPN, 3.)

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

1) Remote, 2) Low, 3) Moderate, 4) Frequent, 5) Very High. (The Occurrence is Remote with the exception of pediatric patients. So the RPN would be a 1\* unless it is a child. Then the Occurrence RPN would be 2.)

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. (The Detectability RPN equals 5. The potential for post pump chorea is not be detectable prior to surgery or during CPB.)

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. (Only patients undergoing moderate to profound hypothermic CPB would be potentially at risk. So the Frequency RPN would be 2.)

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 total RPN for this failure is = 3\*1\*5\*2 = 30. If this were a pediatric patient undergoing moderate to profound hypothermic CPB, the total RPN would be 3\*2\*5\*2 = 60.)