PBLD TABLE 15

Williams Syndrome Patient for Posterior Spinal Fusion with Profound Intraoperative, Refractory Hypotension

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Goals:

1. Recognize the pathophysiology and management of patients with Williams Syndrome
2. Recognize problems which may arise for patients with congenital heart disease undergoing non-CV surgery
3. Understand the limits and options for critical intraoperative monitoring during complex spinal surgical cases, as well as the anesthetic implications thereof
4. Recognize that complex spine cases require a multidisciplinary approach to optimize patient outcome & minimize complications
5. Understand guidelines for Mass Transfusions

Case History:

An 8 year old, 25 kg female is scheduled for PSF for 54° scoliosis. PMH is significant for Williams syndrome, s/p repair of AS/PS, coarctation, and recurrent arch PS. Patient coded during initial repair.

Questions:

What is Williams Syndrome, and what is the natural progression of disease? What are the anesthetic implications in Williams Syndrome?

Case History & Physical Exam:

Vitals were normal and stable. Patient had elfin features, 3/6SEM, developmental delay; remainder was normal. Medications included amlodipine bid.

Questions:

Discuss the medical and surgical management of Williams Syndrome? How should we prepare for intraoperative management considering this patient's previous operative history? Which other services should participate in the preoperative evaluation, and what other studies should be done?

Preoperative studies:

Cardiology follows this patient closely and manages medically. Preoperative echocardiogram showed no AS, mild PS, four tightly stenotic neck vessels. There is no note of arterial stenosis.

Questions:
Should this patient receive her normal medications perioperatively? Mother is tearful in discussing the previous critical intraoperative history (coding). In obtaining informed consent, what risks should be discussed? List risks specific to the procedure (PSF) as well as to Williams Syndrome. Would you require an ABG, PFTs or pulmonary consultation prior to surgery?

Case progression:
The patient was instructed to follow NPO guidelines as well as to hold her morning dose of amlodipine. The patient was type and crossed for two units of blood.

Questions:
What is the anesthetic plan? What IV access is optimal or necessary? What special monitors will be used and how will they affect the anesthetic management? What are the limitations of standard monitoring in Williams Syndrome? What are the hemodynamic goals during this case, and what are the repercussions of hemodynamic instability? Are fluid and blood management guidelines available?

Intraoperative care:

Preoperatively, amlodipine was ordered withheld. Inhalation induction, large bore PIV access, L radial arterial line, and endotracheal intubation were straightforward. Positioned prone, anesthesia was maintained with 0.5%Isoflurane, 40%O2/air, propofol (50mcg/kg/min), remifentanil (0.05-0.1mcg/kg/min) infusions. SSEPs/MEPs monitored spinal cord integrity throughout. Although induction of anesthesia was uneventful, the patient became progressively hypotensive. Keeping in mind hemodyanamic goals for this case, what initial therapies and then advanced therapies would you use to support this patient? This patient required phenylephrine and dopamine support, despite aggressive fluid resuscitation. About 2 hours into the case, profound hypotension became refractory; epinephrine was started. Infusions and inhalation agents were discontinued, and midazolam was administered to prevent awareness. What causes of profound hypotension could be contributing to this child's clinical picture? The patient's blood pressure remained low despite epinephrine infusion. Although SSEPs/MEPs were maintained throughout, the case was aborted.

Postoperative care:

The patient remained intubated. In PICU, RIJ CVL access was obtained and norepinephrine infusion was started. Parents revealed patient received amlodipine preoperatively. Cardiology was consulted and reported that BP was never obtained from the upper extremities, only the lower extremities. The patient was weaned off pressors, extubated, and amlodipine was held until completion PSF on POD5, which was uneventful. BP was monitored in LEs only, patient

was extubated, and discharged home after normal post-operative course, without neurologic deficit. Do you discuss the intra-operative problems with the patient and her family? Do you call risk management? What discussions should be held with the operative team for future planning?

Discussion:

Williams syndrome or Williams-Beuren syndrome (WBS) is a rare congenital disorder with an estimated prevalence of 1 in 20000 births. The features include elfin facies, outgoing personality, endocrine abnormalities (hypercalcemia and hypothyroidism), mental retardation, growth deficiency, and altered neurodevelopment. The cardiovascular problems include aortic coarctation, valvar or supravalvular aortic stenosis, and vessel narrowing of systemic, pulmonic, or coronary arteries. Other problems include hypertension, hypercholesterolemia, renal anomalies, liver disease, and inflammatory bowel disease. (4) A chromosomal deletion on the long arm of chromosome 7 causes the characteristic features. The protein elastin is altered by the chromosomal deletion and is associated with connective-tissue abnormalities and cardiovascular disease.

The cardiovascular anomalies of Williams syndrome like coronary artery anomalies or biventricular outflow tract obstruction can lead to sudden death or cardiac arrest in these patients under anesthesia. In many cases the cardiac arrest is refractory to the ACLS guidelines. Other cardiac issues include coarctation of the aorta, patent ductus arteriosus, peripheral arterial abnormalities, and intracardiac lesions like ventricular septal defects (VSD) and Tetralogy of Fallot (TOF). These patient's pre-anesthetic evaluations should include ECG, Echo, possible Holter monitor for any conduction issues or arrhythmia. A coronary angiography may also be needed for coronary ischemia. Epinephrine can have the potential to increase systemic vascular resistance which can decrease coronary blood flow with supravalvular aortic stenosis. As in this case, the history of coarctation of the aorta can lead to blood pressure difference between the lower and upper extremity. In many institutions, 2 anesthesiologists with cardiac anesthesia may need to be present for induction in these patients. These patients are on cardiac medications that needs to be evaluated to see which one to stop. Ace inhibitors, Angiotensin Receptor blockers, and diuretics are usually held on the morning of surgery to prevent severe hypotension intraoperatively.

Posterior Spine surgeries can be complex especially on neuromuscular scoliosis patients. The complications include extensive blood loss, coagulopathies, hypovolemia, alteration in cardiac and pulmonary function in the prone position. It is important to have adequate access in these patients to resuscitate them with fluids and blood. Appropriate monitors include arterial lines to measure constant blood pressure, central lines for drug infusion and central venous pressure

monitoring, and transesopheageal echo to evaluate the contraction of the heart. Techniques to help with massive blood loss include cell saver, use of antifibrinolytics in appropriate patient, and inotropes to maintain pressure.

In summary, patients with Williams syndrome have multi-organ disorders. In addition to the cardiovascular issues in these patients, there is also involvement of the renal, liver, endocrine systems. A thourgh pre-operative evalution should be conducted on these patients. Anesthetic help will also be useful in these complicated cases since Williams syndrome patients can have the risk of sudden death on induction. This is an interesting case of a complicated syndrome in a very challenging surgery.

References:

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