

ANESTHESIOLOGY™ 2014

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When It Rains It Pours; Large AVM Heart Failure Pulmonary HTN and Now There is V-tach!

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Disclosures: This presenter has no financial relationships with commercial interests

Stem Case and Key Questions Content

You have been assigned to interventional radiology (IR) today. The new IR attending that you have not worked with before walks into the control room and tells you about a baby that she would like to post for the schedule today. She would prefer to do the case within the hour. When you ask her about the baby she informs you that the infant is now 8 days old with a large cerebral arteriovenous malformation (AVM) that she would like to embolize and that in her assessment he is doing well.

What does an interventionalist do when he/she is performing an AVM embolization? How would you respond to this request? Is there any additional information you would like to obtain about the baby? As you start looking up information about the infant you call the anesthesia techs to start setting up the suite. Is there any specific equipment or monitors you want?

Upon review of the child's electronic health care record, you learn that the child was born at an outside hospital and stayed there for five days. He is a well-developed infant that had a prolonged hospital stay after birth due to poor feeding. When the pediatrician examined the child, he noted a large head with a bounding occiput to palpation.

What is in your differential diagnosis for this child's poor feeding and large pulsatile head?

The pediatrician was concerned and ordered a further work-up. The chest radiograph showed an enlarged heart with congested lungs. The head ultrasound showed a large occipital AVM in the posterior third of the sagittal sinus.

Does this change your differential diagnosis? Does this solidify a diagnosis for you?

The family was informed of the findings and the infant was transferred to your hospital. On arrival in the neonatal intensive care unit (NICU), the infant was very tachypneic with cool mottled extremities. The NICU team intubated the infant for respiratory support.

What additional information would you like to know about this patient? Is there any imaging or tests you would like to review?

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The patient's vital signs are as follows:

HR – 152, BP 73/40, RR – 47, SpO₂ 100% on n FiO₂ 0.52% Intubated, and Temp 37 Ax, Wt. 4.315 kg

Echo - The echocardiogram demonstrated supra-systemic pulmonary hypertension (with an right ventricle (RV) systolic pressure of 97 mm Hg), severe tricuspid regurgitation, largely dilated right ventricle, small atrial septal defect (ASD), small patent ductus arteriosus (PDA), reduced RV function and high out-put congestive heart failure. The NICU started him on a milrinone infusion, digoxin, and an epinephrine infusion to support his cardiac output. After spending three days in the NICU, this morning the patient's epinephrine infusion (gtt) is weaned off. The patient only started urinating after the initiation of the epinephrine according to the NICU team. (Creatinine at the outside hospital was 1.59.)

Imaging - The head US showed a 2.8 x 5.5 x 3 cm large dural AVM of the most posterior sagittal sinus, and enlargement of the transverse and sigmoid dural sinuses resulting in significant shunting. This morning's chest x-ray continues to show an enlarged heart, increased perihilar vascular markings in the lung fields, and a left clavicle fracture.

Labs: WBC 7.5, Hgb 15.5, Hct 45, Plt 263, Na 143, K 3.5, Cl 102, HCO₃ 34, BUN 51, Cr 0.51, Glucose 89, Art PCO₂ 66.9

Exam: He is a well-formed large infant. He has a warm plethoric head with a pale body and cool extremities. A loud bruit is audible and palpable when auscultating the back of his head.

Auscultation of the infant's lungs is clear with some rales at the bases. He spontaneously opens his eyes and moves all his extremities.

Would you proceed with the anesthetic? If not, how long would you postpone the procedure?

What measurable goals do wish to achieve before proceeding?

You discuss the patient with the NICU attending, who reports that the baby has improved since his arrival 3 days ago. He is less tachypneic and they have been able to wean his oxygen requirement. He has had multiple episodes of supraventricular tachycardia (SVT) that required adenosine to treat, including an episode approximately 1 hour prior to your visit. However, the NICU faculty reports the bouts of SVT are becoming less frequent. They were also able to wean off the epinephrine infusion and the infant continued to have normal urine output and remained hemodynamically stable. He remains on his milrinone infusion. The interventionalist tells you that the baby will never get better from the high output cardiac failure until she treats the AVM. She expresses an urgent desire to start the procedure and tells you she doesn't understand why we are so concerned. She explains that she plans on feeding a catheter from the groin into the AVM and then inject some cyanoacrylate glue into 4-6 vessels to start embolizing the AVM. You agree to proceed. *How will you monitor the child? Would you request any special medications or infusions to be available? How would you induce and maintain this child's anesthetic? What are your concerns specific to the condition of the child? Do you have any additional concerns because you are in a remote location (3 floors away from the main OR)? How may this affect your anesthetic plan?*

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After transferring the child from the NICU to the interventional radiology suite and placing your ASA monitors and NIRS (Near Infrared Spectroscopy) monitor on the patient you begin your induction. You slowly give 3 mcg/kg of fentanyl, 0.5 mg of midazolam, and 10 mg of rocuronium. You successfully position the patient and confirm bilateral breath sounds. You continue the milrinone infusion at 0.5 mcg/kg/min and start the infant on 1/2 MAC of sevoflurane and a fentanyl infusion of 3 mcg/kg/hr. Your fellow successfully places a 22 g right radial art line in the infant on the first pass. Access to the baby is very limited and the breathing circuit is distended to allow room for the radiology equipment.

As the case proceeds and the interventionalist shoots several rounds of dye you note occasional episodes of hypotension and desaturations that always quickly self resolve. However, approximately two hours into the procedure you start noticing that every injection of "glue" by the interventionalist is followed by 20-30 seconds of desaturation to the low 80'.

What is your differential? What are you going to do?

You inform the interventionalist of your observation and concerns. She informs you that she has embolized three vessels and things are going well from her point of view. You inquire about the possibility of an air embolus with the injections. She assures you she is not injecting any air and she feels the cardiorespiratory reaction of the child has nothing to do with what she is currently doing. She plans on embolizing at least 1-2 more vessels today.

Do you change your anesthetic?

You instruct your fellow to increase the FiO₂ and watch for any changes in the patient. As you are telling your trainee this, you see an 8-beat run of ventricular tachycardia (V-tach) that correlates with another injection of cyanoacrylate. You also note a prolonged desaturation into the 80's on a FiO₂ of 100% and a transient decrease in BP's.

You watch as the interventionalist again injects the glue compound resulting in a short run of V-tach that self resolves after 8-beats. The arrhythmia is again accompanied by a desaturation lasting approximately 1 minute and a decrease in systolic BP to the 40's before self-resolving.

What is your differential diagnosis now? How can you intervene?

You notify the radiologist of your concerns and ask her to stop the procedure. You restart the epinephrine to support the heart. The interventionalist seems incredulous that this could be the result of her procedure and wishes to see for herself. She argues this patient really needs to be treated for the AVM. She proceeds to inject cyanoacrylate two more times into the AVM but this time also notes the V-tach. You note that these last two times the hypotension, desaturation, and tachycardia are more prolonged and now the child is saturating in the low 80's without recovery.

Has your differential diagnosis changed? What are your concerns? When do you call it quits?

You insist the interventionalist stops the procedure for the day. You inform her that the child has become unstable and you are worried that the child will soon code and require CPR if she continues. The interventionalist physician agrees but reports she was only able to accomplish 4 of the 6 vessels she had planned for the day. You again ask the radiologist if she may have injected air or if the glue could have migrated. She tells you that it is very unlikely but agrees to fluoro the chest to show you. The image of the chest shows multiple new lesions in the chest and she is forced to acknowledge that the glue has embolized into the pulmonary vasculature.

Does this change your differential diagnosis? Does this change your anesthetic management? Do you continue the procedure? If not, do you extubate this child?

You elect to turn on the nitric oxide (iNO) you had brought to the IR suite. The patient's saturations improve to the low 90's and the systolic pressures are now in the 70's. You elect to transport the infant to the NICU intubated and sedated. Your report to the NICU emphasizes the embolization of the glue into the pulmonary vasculature worsening the pulmonary hypertension and cardiac irritability.

What is the mechanism of action of iNO? Could there be a disadvantage in using it in this situation?

The infant takes approximately one week to recover from his initial embolization and eventually undergoes several more treatments for his AVM.

Model Discussion Content

Arteriovenous malformations (AVM's) may be the result of trauma, may be hereditary such as hereditary hemorrhagic telangiectasia, or may be congenital in origin. The embryonic vascular system forms between weeks 3 and 10 of gestation and congenital cerebral AVM's are lesions resulting from an abnormal development of the arteriolar-capillary network prior to 11 weeks of gestation. The cause still remains unclear.

The incidence of cerebral AVMs is estimated to be less than 1% and affects males more commonly than females. AVM's are considered a fast flowing vascular lesion that generally grows with the child. As a result of the AVM malformation, there is a large left to right shunt from the arterial side to the venous side. This shunt leads to possible overcirculation, ischemia to surrounding tissue secondary to a steal affect, and possible vascular congestion, obstruction, and abnormal vascular autoregulation. The presenting symptoms may include seizures, hydrocephalus, macrocrania, neurodevelopmental delay, hemorrhage, stroke, and rarely congestive heart failure and pulmonary hypertension. Most AVMs become symptomatic in adulthood, with only 18% presenting prior to the age of 15. The mortality for all patients is 1 -2 %. There is a rate of 10% instant death with the first hemorrhage of an AVM, with the mortality of subsequent bleeds being 30% .⁷ Less than 1% of all AVM's present with congestive heart failure, primarily in infants. The mortality of an infant presenting with an AVM in congestive heart failure that is left untreated is greater than 90%⁴.

Cerebral AVMs that result in congestive heart failure are most commonly associated with the deep vascular structures such as the Vein of Galen, the dural vessels, or the superficial pial vessels.⁹ Large cerebral AVMs, with their high left to right shunt volume create an increased volume load on the right ventricle of the heart. This can lead to right atrial dilatation, right ventricular hypertrophy and dilation and overcirculation of the pulmonary vasculature resulting in pulmonary hypertension. AVM's also result in a decrease in systemic vascular resistance, increased heart rate, stroke volume, and increased overall plasma volume.⁹ Left untreated the

increased cardiac output and increased volume returning to the heart will lead to high output congestive heart failure, cardiogenic shock and eventually to multi-organ failure and death. Confounding factors in the neonatal period may include a patent foramen ovale (PFO), patent ductus arteriosus (PDA), a stiff right ventricle, and increased pulmonary vasculature resistance which may result also in a right to left shunt and cyanosis of the newborn. In neonates the persistent high pulmonary vascular resistance may prevent the transition from fetal to neonatal physiology, so that even after treatment the child may suffer from persistent pulmonary hypertension. The initial presentation of impending congestive heart failure secondary to a cerebral AVM may, as in our case, maybe as subtle as feeding intolerance, tachypnea and tachycardia. The progression to cardiogenic shock is marked by the development of oliguria, hepatomegaly, and cardiovascular instability requiring vasoactive support.

On physical exam, the patient may have a bruit over the vascular malformation due to the lack of laminar flow. The patient should be inspected for any signs of vascular congestion, hydrocephalus or macrocephaly. Arteries proximal to the malformation tend to be enlarged and may have a thrill or prominent pulse on palpation while vessels distal to the malformation may have diminished pulses. Therefore, in cerebral AVM's, the carotid pulse may be bounding and the femoral pulses absent or significantly diminished. The exam may note a hyperdynamic or enlarged heart and possible cardiac murmur.⁹

The assessment of an AVM can be performed by head ultrasound (in infants), CT, and MRI. However, the gold standard for evaluation is a cerebral angiogram. Additionally, a CXR, EKG, and Echocardiography exam (Echo) should be part of the preoperative workup. The EKG may show some ischemic changes and the Echo may show flow reversal in the aorta, which is pathognomonic for cerebral AVMs. The Echo may also demonstrate right ventricle (RV) dilatation, hypertrophy, and tricuspid regurgitation suggestive of pulmonary hypertension.³ Treatment options for cerebral AVMs include surgical excision or endovascular embolization. The latter is now considered the initial treatment of choice and is performed in the interventional radiology suite. The embolic materials used are isobutyl-2-cyanoacrylate (IBCA), n-butyl-2-cyanoacrylate (NBCA), barium impregnated polyvinyl alcohol particles (PBA), platinum microcoils and detachable balloons.⁴ The IBCA and NBCA have the advantage that they are in a liquid form. They can be injected through smaller flexible catheters allowing access to the most distal part of the vasculature. Embolization of proximal vessels bares the risk of recurrence of the AVM via recanalization and the development of collaterals. The disadvantage of cyanoacrylate is the need to polymerize after injection, which depending on the flow rate in the AVM bares the risk of migration.⁴ The embolization of AVMs is a staged procedure. The first treatment is aimed at reducing the flow rate through the AVM by approximately one third,² giving the cardiovascular system time to recuperate and adjust prior to subsequent treatments. When these infants present to the OR it is not uncommon for them to be on one or more pressors and on ventilatory support. The anesthetic management needs to be focused on preventing an increase in pulmonary vascular resistance, a decrease in cardiac inotropy, and maintenance of adequate cardiac output to ensure adequate perfusion and hemodynamic stability. This also involves maintaining appropriate ventilation and oxygenation. The ASA

standard monitors should be supplemented by NIRS (to monitor tissue perfusion) and an arterial line. The infant should be paralyzed to prevent any unanticipated movement. Positioning of the child and vascular access are crucial, since there will be very limited access during the procedure. You may want to consider having a type and cross in case of bleeding. Other aspects that require special vigilance are the potential for fluid overload from the dye load and flushes, hemorrhage from rupture of cerebral vessels, and pulmonary embolism from embolic material migration. The latter is especially true for high flow AVM's. In a retrospective review of 47 consecutive children undergoing therapeutic embolization of a cerebral AVM, 35% showed deposits of embolic material in the lungs post procedure. Two required endotracheal intubation and assisted ventilation and one developed ARDS. Both cases involved the use of cyanoacrylate glue. The symptoms resolved within 7 - 10 days with supportive care. In the animal model the embolization of cyanoacrylate resulted in medial and intimal necrosis of pulmonary vasculature and subsequent perivascular fibrosis after 2 months. This is thought to potentially be a cause of chronic pulmonary hypertension.

Since these procedures are performed in off site locations you should be well prepared and have all potential equipment and pharmaceutical needs available. This holds especially true for vasoactive drips, monitoring equipment, and nitric oxide, as well as skilled personnel to assist you. Nitric Oxide is a potent vasodilator that is especially useful in the treatment of acute pulmonary hypertension. It acts directly on the lungs with little systemic effect because of its short half-life in blood. Nitric Oxide activates guanylyl-cyclase in smooth muscle cells and platelets, increasing the levels of the intracellular messenger cyclic guanylyl phosphate (GMPc). This phenomena produces smooth muscle relaxation and platelet aggregation inhibition, presumably by reduction of the intracellular free Ca^{2+} concentration.⁶ In cases of right heart failure and associated pulmonary hypertension, Nitric Oxide can be beneficial by decreasing the PVR and therefore decreasing right heart workload. But in cases of left heart failure, Nitric Oxide will hasten decompensation of the left ventricle by increasing the volume load secondary to increased pulmonary perfusion. Since the type of heart failure may not always be clear, judicious use of iNO is recommended in the treatment of patients. The administration of iNO requires special equipment for it to be fed into the breathing circuit of the patient. Additionally the medication is expensive and the tank is cumbersome to transport.

This patient with his high-output heart failure, pulmonary HTN, arrhythmias, and multi-organ dysfunction provides a challenge to the skilled anesthesiologist. Placement of the case outside the operating room environment and in a remote location adds additional challenges and concerns. By understanding the patient's physiology, the underlying etiology of the disease, the needs of the interventionalist during the procedure, and being prepared for potential complications gives this infant the best chances for tolerating the procedure and anesthetic.

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