Acute Autonomic Nervous System Dysfunction with Cardiac Decompensation in a Child with Dystonia After Administering a Short Acting Anticholinergic Agent

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Goals:

1. Discuss the causes as well as the medical and surgical treatment of dystonia in children.

2. Discuss the normal function of the autonomic nervous system

3. Increase awareness and appreciation for the potential complications that might be associated with the administration of anticholinergic agents.

4. Review the determinants of myocardial oxygen supply and demand and the physiologic implications of these factors in the care of pediatric patients.

5. Understand the pathophysiologic effects of unopposed adrenergic system activity and implement effective treatment responses to this pathophysiologic state.

Case Presentation

An 8 year old female with a known history of seizures and paroxysmal dystonia presents for an MRI of her brain to evaluate a recent acute exacerbation of her dystonia and severe fluctuating dysarthria.

Questions

What is dystonia? What is the incidence of dystonia in pediatrics? How are dystonias classified? What are some significant clinical manifestations associated with dystonias? How do you grade the severity of dystonia? Is there a common pathophysiologic pathway that connects all dystonias? What is the natural history of dystonia?

Case Continuation

Her medications include Baclofen, Bromofed, Sinemet, Keppra, and Artane.

Questions

Can you describe what each of these medications are and how they work? How does Bromofed work? How does Baclofen work in treating dystonia? How does Artane work in treating dystonia? How does Sinemet work in treating dystonia? Are there any non-pharmacologic therapies employed to treat dystonia? How do the other modalities work to treat or control dystonia?

Case Continuation

When you see the child in the preoperative assessment area her vital signs are:

BP 107/72 **HR** 56 **RR** 16 **SpO2** 100% on room air

Questions

What do you think of her heart rate? Is this a normal or acceptable heart rate in a patient this age? Do you have any anesthetic management concerns before inducing this patient for her diagnostic study? What are your anesthetic management or sedation options? How will you induce this patient? What is your anesthetic maintenance plan? Could you do this MRI with intravenous sedation only? What agents would you choose? Is general anesthesia required for this case? What are the advantages and disadvantages of each approach in pediatric patients?

Case Continuation

The patient undergoes an unremarkable general anesthetic including induction and maintenance with sevoflurane, placement of a laryngeal mask airway, and standard monitoring. Her vitals remain stable throughout the 45 minute study and her LMA is removed deep immediately after the study. She is transported to the PACU and report is given. She experiences episodes of bradycardia in PACU and her heart rate reaches a nadir of 34 beats per minute approximately 15 minutes after arrival.

Questions

What might be the possible causes of her decrease in heart rate? What role does heart rate play in determining blood pressure and cardiac output? How low are you comfortable letting a pediatric patient's heart rate go before you feel compelled to intervene? What agent would you give when you treat a mild to moderately decreased heart rate?

Case Continuation

After receiving a modest dose of glycopyrrolate the patient develops a hyperdynamic hemodynamic picture including a maximum heart rate of 220 bpm, a maximum blood pressure of 169/136 and an EKG pattern consistent with ventricular tachycardia.

Questions

Can this response be explained by the pharmacologic effect of 10 mcg/kg of glycopyrrolate? What other contributing factors might play a role? How does glycopyrrolate cause an increase in heart rate? Can glycopyrrolate have a synergistic effect with any of the medications this child is already receiving? Why did this child develop ventricular tachycardia? What are the determinants of myocardial oxygen supply and myocardial oxygen demand?

Case Continuation

The child now begins to desaturate. She is determined to have extremely weak pulses and CPR is initiated while lidocaine is administered and bag mask ventilation is started. When equipment is available, the patient is intubated. Pink, frothy fluid issues from her endotracheal tube.

Questions

Should we give any sedatives to this patient before intubating her? Why does this patient develop cardiogenic pulmonary edema? What are your next immediate interventions?

Case Continuation

After intubation the patient is resuscitated and stabilized and transported to the PICU. She develops a hypodynamic picture requiring alpha agonists and inotropic support overnight. A cardiology consult is obtained and an echocardiogram performed. Her ejection fraction is noted to be 35.4%. She has prominent left ventricular dilation, significant regional wall motion abnormalities, and moderate mitral regurgitation.

Questions

Why does she develop ventricular hypokinesis and mitral regurgitation? What inotropic support would you administer? Does she need afterload reduction in addition to the inotropic support? The parents ask questions regarding why this anesthetic course was different than the previous perioperative courses. How do you respond? What would you tell the parents regarding the patient's prognosis at this point?

Case Conclusion

Over the next week, daily echocardiograms are performed and the patient improves on each subsequent evaluation, ultimately recovering completely back to baseline cardiovascular status and being discharged from the PICU to a transitional care unit within the hospital where her workup of her dystonia exacerbation continued.

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