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PBLD: Anesthetic Misadventures with Long QT Syndrome

7 year old, 22Kg, female with congenital Long QT Syndrome type 2 is admitted due to syncope while running at school. She had a pacemaker placed on day of life 6 for LQTS with intermittent AV block 2:1, and is being paced at 100BPM. Interrogation fo the pacemaker reveal an episode of ventricular fibrillation. She continues to have life threatening arrhythmias difficult to control with medications. She is scheduled for a left thoracic sympathectomy. The patient is on nadolol 20mg po qd , a magnesium infusion and a lidocaine infusion for suppression of arrhythmias, and on vasopressin for treatment of hypotension. She is in the intensive care unit on room air with 2 peripheral IVs and a left dorsalis pedis arterial line. Her mental status is somnolent. Vital signs: BP 70/51 HR 100 SpO2 98% on room air.

1 What is Long QT Syndrome? How is is diagnosed?

2 How is it treated?

3 Why is she somnolent and hypotensive?

4 How would you induce and maintain anesthesia on a patient with LTQ syndrome?

5 Are there any drugs you should avoid?

6 How would you manage the pacemaker?

You induce anesthesia and intubate. The vital signs following : BP 45/39, HR 100, SpaO2 100%, ETCO2 28 on mechanical ventilator set at TV 200ml RR 20

6 How would you treat hypotension in the setting of long QT syndrome?

7 How do you maintain anesthesia in the setting of long QT syndrome?

Following you BP management interventions, the arterial line becomes non pulsatile and the BP cuff is not reading. EKG still shows paced rhythm. SPO2 is not reading. ETCO2 has a normal waveform at 32 mm CO2. You are unable to draw from the arterial line.

8 What is your next step?

Femoral and axillary arterial lines are attempted. A right axillary arterial line is placed under ultrasound guidance. BP on axillary arterial line is 134/84. BP on dorsalis pedis arterial is still not pulsatile. Blood gas from the axillary line reveals: pH 7.3 CO₂ 43 O₂ 52 BE -4.2 Sat 85% on 100% FiO₂

9 What is your next step?

A chest x ray is performed. Pediatric cardiology is called for a TEE. TEE reveals moderate biventricular dysfunction with moderate to severe mitral regurgitation. No atrial septal defect or patent foramen ovale is seen. Rest of exam is normal.

10 What could be the cause of the new finding of moderate to severe mitral regurgitation?

11 What do you expect to find in the chest x ray?

12 Would you proceed with the case?

The thoracic surgeon is going away for a week. The case will have to wait that long if postponed. Cardiology deems the arrhythmia as life threatening and would like to the procedure done before the surgeon leaves.

13 Is there any treatment you could attempt improve her A-a gradient so that one lung ventilation is feasible?

You decide to wait in the OR for and treat the pulmonary edema for at least half an hour. Intravenous furosemide is administered. A milrinone infusion is started at 0.3mcg/kg/min after a 25mcg/kg load. TEE reveals improved biventricular function and mild MR. Repeat arterial blood gas is pH 7.37 CO₂ 42 O₂ 142 sat 100% on 100% FiO₂. You decide to proceed with the case.

14 Would you proceed with the case?

15 How would you achieve lung isolation?

A trial at one lung ventilation is done. Arterial blood gas is: pH 7.47 CO₂ 35 O₂ 104 Sat 98% on 100% FiO₂. The case proceeds without further incident. The patient was transferred to the ICU on a propofol infusion. She was extubated the following day.

16 How would you discuss the events with the parents?

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