Title: Congenital Complete Heart Block: A Neonatal Anesthetic Emergency

Author(s): D Almazan, MH Martin, C Ramamoorthy
Affiliation(s): Stanford University School of Medicine

ABSTRACT BODY:

To maintain cardiac output, the neonatal myocardium is heart rate dependant. In congenital complete heart block (CCHB), the heart rate is slow and the response to chronotropic medications is often variable (1). Therefore, early and sometimes emergent institution of cardiac pacing may be necessary. Reports of anesthetic management of CCHB are also scarce (2). We report the perioperative anesthetic management for emergent pacemaker insertion in a neonate with symptomatic CCHB.

A 34-week gestational age newborn presented with isolated CCHB. Due to maternal history of lupus, the neonate was diagnosed in utero by fetal echocardiography at 17 weeks gestation. A 2.2 Kg boy was delivered via scheduled caesarian section in the labor and delivery suite for worsening pericardial effusion and ventricular bigeminy. Following delivery, he was intubated for low Apgar scores and bradycardia in the 30’s. Epinephrine was given via endotracheal tube, CPR initiated, and transcutaneous pacing was attempted without capture. With ongoing CPR and inotropic support, the neonate was transferred and prepared for surgery in the main operating room (OR). Umbilical venous catheter was secured, but arterial access was not obtained prior to surgery. A subxiphoid incision was made and temporary epicardial right ventricular pacing wires were placed. The ECG rhythm was unclear with artifacts and an epicardial echocardiogram confirmed ventricular fibrillation with minimal myocardial contractility. The myocardium was defibrillated and then paced successfully in a VOO mode at 160 beats/minute. Aggressive resuscitation was continued with volume, inotropes and sodium bicarbonate. He also received 5mg of Ketamine, 2 mg of Rocuronium, 2 mcgs of Fentanyl and packed red blood cells for a low hematocrit. Right atrial and ascending aortic lines were placed for monitoring and blood sampling. Due to severe and persistent lactic acidosis, a plan was made to continue with temporary pacing wires and to place permanent leads when he was better resuscitated and stable. Therefore the patient’s chest was left open and transported to the intensive care unit.

CCHB was first described as “impaired atrioventricular syndrome” by Morquio in 1901 with an incidence of 1:22,000 live births (3,4). Mothers of children with CCHB usually have a connective tissue disorder where maternal immunoglobulins cross the placenta and damage the fetal cardiac conduction system (5). It can manifest as an isolated finding or be associated with other structural heart disease. Median age at time of diagnosis is 7 months, but can present during childhood or be asymptomatic until adulthood (6). However, neonatal diagnosis of CCHB is associated with high mortality (7). Emergency anesthesia for symptomatic CCHB can have significant hemodynamic instability.

Though rare, CCHB is a cause of sudden neonatal demise. For this case, preparation started months before, and a multidisciplinary team of health care professionals was available at the time of birth with an OR on standby. Airway and breathing were secured and aggressive CPR initiated early. Emergency drugs and drips were also prepared ahead of time. Although transthoracic pacing was unsuccessful, it is an essential first line treatment for CCHB, especially that transvenous pacing was not possible due to his small size. Having an echocardiogram and defibrillator readily available also proved useful in early diagnosis and treatment. Lastly, care was taken to avoid myocardial depressants, and acidosis and hypovolemia promptly corrected for a successful resuscitation of a neonate with symptomatic CCHB.

References: