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The Blue Baby Blues: A Rare Case of Cyanosis in the Newborn

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CHIEF COMPLAINT

JS presented at birth with cardiorespiratory failure of unknown origin and has failed to respond to conventional medical therapy.

HISTORY OF PRESENT ILLNESS

JS was a full term large for gestational age (LGA) male born to a 21 year old gravida 2 para 1 mother at a referring hospital. His mother received appropriate prenatal care and the pregnancy was uncomplicated. Maternal labs revealed that she was RPR negative, HBsAg negative, Rubella immune, HIV negative, and Group B Strep (GBS) unknown. JS was delivered via spontaneous vaginal delivery seven hours after rupture of membranes (ROM) for clear amniotic fluid. The delivery was uncomplicated with a reassuring fetal heart rate tracing throughout. At birth, the infant was found to be cyanotic with poor respiratory effort. He received bag and mask ventilation, but did not improve, thus he was intubated in the delivery room. He did not require chest compressions or medications as he maintained a heart rate greater than 60 beats per minute (bpm) throughout the resuscitation (Kattwinkel, 2006). Apgar scores were 5 and 6 at 1 and 5 minutes of life.

JS was immediately transferred to the neonatal intensive care unit (NICU), placed on mechanical ventilation with 100% oxygen, and umbilical venous and arterial lines were inserted to appropriate placement. Oxygen saturations (SaO₂) on admission were found to be in the 50s and his initial arterial blood gas (ABG) was significant for a mixed respiratory and metabolic acidosis (pH 6.95, PaCO₂ 72 mmHg, PaO₂ 12 mmHg, HCO₃ 13 mEq/L, base deficit -16). Over several hours, respiratory support was increased to maximal settings on conventional ventilation and inhaled nitric oxide (iNO) was initiated at 20 parts per million (ppm) without improvement. Multiple normal saline (NS) boluses and doses of sodium bicarbonate were administered with minimal improvement in blood pressure (BP), SaO₂, or ABG. Dopamine was initiated and titrated up to 10 micrograms/kilogram/minute (mcg/kg/min) for management of hypotension, but also had little effect. A blood culture was drawn and empiric antibiotic therapy was initiated.

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Given the infant's critical state and inability to stabilize despite the described measures, at approximately 10 hours of life transport was arranged to the nearest children's hospital with extracorporeal membrane oxygenation (ECMO) for suspected persistent pulmonary hypertension of the newborn (PPHN). Alprostadil (Prostaglandin E1) was initiated during transport at 0.05 mcg/kg/min due to concern for a possible ductal-dependent cardiac lesion.

On arrival, JS was placed on high frequency oscillatory ventilation (HFOV) and remained on 100% oxygen and iNO at 20 ppm. His ABG revealed pH 6.98, PaCO₂ 55 mmHg, PaO₂ 27 mmHg, HCO₃ 12 mEq/L, base deficit -20. An arterial lactate was 16.1 mmol/L (reference range 0.5 – 1.6) (Gunn & Nechyba, 2002). A basic metabolic panel (chemistries, blood urea nitrogen and creatinine) and complete blood count (CBC) with differential were normal. Additional NS and sodium bicarbonate boluses were administered and dobutamine was started and titrated to 10 mcg/kg/min. A chest x-ray revealed the endotracheal tube and umbilical venous and arterial lines were in appropriate placement, decreased pulmonary vascular markings, and a mildly enlarged cardiac silhouette with no other abnormalities. Despite escalation in both respiratory and inotropic support, JS's pre and postductal SaO₂ remained in the 40s–50s and he remained hypotensive.

FAMILY HISTORY

JS's mother reports that both parents and one sibling are healthy without any significant medical history. There was no known family history of neonatal death, birth defects, or childhood illness.

PHYSICAL EXAMINATION

JS's birth weight was 4270 grams, length was 52 centimeters (cm), and head circumference was 36 cm. His temperature on arrival was 97.2 degrees Fahrenheit, heart rate 172 bpm, respiratory rate 76 breaths per minute, and arterial blood pressure 57/21 mmHg with a mean of 33 mmHg. The general impression is of a non-dysmorphic, hypotonic LGA male with central cyanosis and respiratory distress. Evaluation of the respiratory system revealed tachypnea and retractions with bilaterally clear and equal lung sounds. Cardiovascular assessment revealed a tachycardic and poorly perfused infant with weak peripheral pulses and delayed capillary refill. Precordial exam was significant for a grade 2/6 pansystolic murmur with an active precordium. No bruits were heard over the liver or anterior fontanelle. Abdominal exam was remarkable only for a liver edge palpable 2 cm below the right costal margin.

CASE STUDY QUESTIONS

1. What differential diagnoses are you considering for this neonate?
2. What is your plan, including diagnostic tests and consultations with specialty services?
3. What specialized teaching should be included in the discharge preparation for this family?
4. What outpatient follow-up services are necessary for this infant after discharge?

QUESTIONS & ANSWERS

1. What differential diagnoses are you considering for this neonate?

The clinician should consider differential diagnoses originating from an infectious, respiratory, or cardiac cause. Although sepsis can present with multi-organ failure and cannot be ruled out at this point, several pieces of information suggest the clinician should continue investigating.

First of all, there are few risk factors for sepsis. The maternal GBS status was unknown, but ROM occurred only seven hours prior to delivery, amniotic fluid was clear, all other labs were negative, and JS's mother reports no history of recent illness or symptoms of infection during delivery. Additionally, the clinician can be reassured by the normal CBC with differential. Empiric antibiotics should be continued until blood culture is confirmed negative, but the clinician should consider another etiology.

JS presented with a number of symptoms of respiratory failure, including cyanosis with respiratory distress, an extremely low SaO₂, and a mixed respiratory and metabolic acidosis, which might suggest aspiration pneumonia or PPHN. Aspiration pneumonia is unlikely in this case because there was no history of perinatal compromise, there was clear amniotic fluid, and the chest x-ray (CXR) was clear. PPHN, on the other hand, is a definite possibility and can often present with respiratory failure, decreased pulmonary vascular markings (PVMs) on CXR, a cardiac murmur, cardiomegaly, and cardiac failure. The two signs that suggest PPHN may not be the diagnosis are that the pre and postductal SaO₂ are equally low. Usually with PPHN, the preductal SaO₂ (from the right radial artery) is higher than the postductal SaO₂ (from the left radial, umbilical, or tibial arteries) by >5%, secondary to right-to-left shunting at the level of the patent ductus arteriosus (Gomella, Cunningham, Eyal, & Zenk, 2004). Additionally, PPHN usually responds to iNO, which in this case was not appreciated. At this point, there is not enough information to distinguish PPHN from congenital heart disease (CHD).

The differential diagnosis of CHD is supported in this case by several indicators of cardiac failure, the lack of response to either 100% oxygen or iNO, and equally low pre and postductal SaO₂. Diagnosis of CHD must first distinguish between acyanotic, meaning normal SaO₂, or cyanotic, as in this case with a low SaO₂. Cyanotic CHD can then be differentiated by increased or decreased pulmonary blood flow, which is determined by evaluation of the PVMs on CXR (Polin, Yoder, & Burg, 1993). In this case it was noted that there were decreased PVMs, which suggests one of the cyanotic CHD lesions with decreased pulmonary blood flow, specifically total anomalous pulmonary venous return (TAPVR) with obstruction, transposition of the great arteries (TGA), pulmonary atresia with intact ventricular septum, or severe tricuspid insufficiency. Definitive diagnosis requires evaluation by echocardiogram (ECHO).

2. What is your plan, including diagnostic tests and consultations with specialty services?

Maximal respiratory support via HFOV with 100% oxygen and iNO at 20ppm, vasopressor infusions, Alprostadil, and empiric antibiotics were continued. The ECMO and Pediatric Cardiology teams were consulted. A 12 lead ECG revealed normal sinus rhythm, right axis deviation, possible right ventricular enlargement, and a non-specific T wave abnormality. ECHO demonstrated absence of flow through the pulmonary valve consistent with pulmonary atresia, an intact ventricular septum, right ventricular hypertrophy, an abnormal tricuspid valve with a large amount of tricuspid regurgitation, and right to left flow through a tiny patent ductus arteriosus and patent foramen ovale.

JS was emergently taken to the operating room for a central aortopulmonary shunt and pulmonary valvotomy. The surgical procedure on cardiopulmonary bypass was uncomplicated. Sternal closure was delayed due to diffuse swelling. He returned to the NICU with a SaO₂ of 85–90%. Shortly after return to the NICU, JS became profoundly hypotensive and desaturated requiring multiple NS boluses and sodium bicarbonate. He then became bradycardic, requiring epinephrine twice. At this time, the cardiothoracic surgical team performed an emergency thoracotomy and suctioned a large amount of blood from his pericardium to relieve a cardiac tamponade. Despite relief of the hemopericardium and maximal inotropic support, he remained acidotic and hypotensive.

An ECHO was repeated at this time to evaluate cardiac function, which demonstrated no antegrade flow through the pulmonary valve, filling of the pulmonary arteries via the central shunt, and a flail tricuspid valve with severe tricuspid insufficiency. JS returned to the operating room for a tricuspid valve repair. Direct inspection of the tricuspid valve revealed a flail anterior leaflet with a sclerotic and necrotic papillary muscle tip that had ruptured from a fibrotic chord. The valve was repaired by reimplantation of the anterior leaflet chordae. The postoperative ECHO demonstrated antegrade pulmonary blood flow with mild tricuspid regurgitation. He was successfully weaned from CPB without complication and again returned to the NICU. The postoperative course thereafter was uncomplicated. Following extubation on day 9, the neurology team was consulted secondary to asymmetrical facial movement during crying and concern for hypoxic injury given a history of profound hypoxia and acidosis. It was determined that the asymmetrical facial movement was due to congenital absence of the depressor anguli oris, a facial muscle involved in crying and smiling. Although this is a minor congenital anomaly seen in only 0.5–1% of newborns, it is associated with congenital defects of the cardiovascular system (Rioja-Mazza, Lieber, Kamath, & Kalpathi, 2005). Magnetic resonance imaging of the brain revealed no evidence of acute infarct, hemorrhage, or masses. He passed a newborn hearing screen and was discharged home on day of life 20 requiring feeding supplements via nasogastric tube and diuretic therapy, but otherwise well.

Flail tricuspid valve is a rare, but potentially lethal, cause of cyanosis and hypotension in the newborn infant. More commonly presenting in the adult patient with chest trauma after motor vehicle accident (Varahan, Farah, Caldiera, Hoit, & Askari, 2006), flail tricuspid valve results from rupture of one of the chordae or papillary muscles that holds the valve in place, causing severe and potentially life threatening valvar insufficiency and congestive heart failure. In the neonate, the etiology of flail tricuspid valve cannot always be determined, but is likely due to either an intrapartum hypoxic event, traumatic rupture during the birthing process, or a thromboembolic event resulting in necrosis and rupture of the papillary muscle (Arrington, Kouretas & Mart, 2005). This can occur at either the level of the mitral or tricuspid valve, but the anterior papillary muscle is especially vulnerable because of its high oxygen requirement, location at the distal end of the coronary circulation, and perfusion only during diastole in the setting of systemic right ventricular pressures (Arrington et al., 2005).

Typical presentation of the infant with flail tricuspid valve is similar to the case presented here. The infant is often full term after an uncomplicated pregnancy. Although some cases are associated with evidence of perinatal compromise (Benvenuti, Aiello, Cury & Ebaid, 1992; Pinto et al., 1992), the majority cannot be attributed to a perinatal event. The infant presents immediately or shortly after birth with profound cyanosis and refractory hypotension, respiratory distress, and a grade 2–3/6 murmur with an active precordium (Lim, Huh & Jun, 2004). The infant typically will not respond to a hyperoxia test, will have diminished PVMs on CXR, and an ECHO will reveal tricuspid insufficiency with functional pulmonary atresia, meaning absence of flow through the pulmonary valve as a result of complete tricuspid insufficiency (Boon, Hruda & Schoof, 2007). In most cases, the infant will not respond, or will only respond minimally, to maximal respiratory and inotropic support and the only life saving intervention is surgical repair of the valve. In order to improve chances of survival, it is imperative to quickly come to a diagnosis and transfer the infant to a tertiary care facility with pediatric cardiothoracic surgical intervention and/or ECMO for stabilization until surgical intervention can be arranged (Alkalay et al., 1988; Arrington et al., 2005).

3. What specialized teaching should be included in the discharge preparation for this family?

Discharge preparation for the infant with CHD should first focus on the standard education provided to new parents upon discharge of a newborn, including diaper changing, umbilical cord care, temperature measurement, prevention of infection, appropriate car seat use (AAP,

2007a), and positioning the infant on his back to sleep (AAP, 2007b). Families should then be educated on the special needs of the infant with CHD with regards to feeding, bathing, wound care, handling, dressing, cardiopulmonary resuscitation, medication administration, and when to call the primary care provider (PCP) or emergency medical services. Each of these unique needs is discussed briefly. Please consult Pye & Green for more information (2003a) and for a family teaching resource (2003b).

Infants with CHD often experience difficulties with feeding and many require increased calories and/or non-oral supplementation via nasogastric, orogastric, or gastrostomy tube. All caregivers should be taught the recipe for increased calorie formula and/or expressed breast milk, provided a hard copy for reference purposes, and should be observed to ensure understanding. Similarly, all caregivers should be taught and observed while placing and confirming correct position of nasogastric or orogastric feeding tubes and while administering non-oral feedings.

Bathing the infant with CHD after cardiac surgery should be delayed until the incision is fully healed. Until then, the infant may be given a sponge bath with a gentle, pH-balanced infant soap. The incision may be gently wiped with a soft cloth. The incision should be kept clean and dry and will heal with time. Nothing should be applied to the incision unless instructed by the cardiothoracic surgery team. Clothing that may irritate the incision should be avoided. When handling the infant, caregivers should be taught to avoid lifting the infant under the arms until the sternum is healed (Pye & Green, 2003a).

All parents of infants with CHD should be taught cardiopulmonary resuscitation according to the American Heart Association (AHA) guidelines (2006). If this service is not offered in the hospital, parents may refer to the AHA website to find courses in their community. Additionally, parents need to be provided with instruction on dosage and administration of medications. It is best if prescriptions are filled and brought in to the hospital prior to discharge to confirm correct formulation. Medication teaching must also include education regarding Palivizumab (Synagis) and bacterial endocarditis prophylaxis in those infants who qualify (AAP, 2003; AHA, 2007). When appropriate, parents should be provided a Bacterial Endocarditis Wallet Card, which is available in English and Spanish, and be instructed to present the card to any healthcare provider attending to the infant (AHA, 2007).

Finally, in addition to the standard teaching of when to contact the PCP, the family should be instructed on signs and symptoms specific to the infant with CHD that necessitate immediate medical attention. The PCP should be contacted immediately if there is redness, swelling, tenderness, or drainage from the incision site, fever over 101 degrees Fahrenheit, any change in breathing, color, feeding habits, or behavior such as increased fussiness, sweating, clamminess, vomiting, or diarrhea (Pye & Green, 2003b). Parents may also be provided with the handout "When Your Child Needs Emergency Medical Services" as a teaching tool for identifying and handling emergency situations (AAP, 2007c).

4. What outpatient follow-up services are necessary for this infant after discharge?

Prior to discharge, the Hepatitis B vaccine and, if appropriate, Palivizumab (Synagis) should be administered (CISP, 2007; AAP, 2003). Arrangements for visiting nurse services and home care supplies should be made as needed. Follow-up appointments with the PCP, the Pediatric Cardiologist, and any other necessary services, such as in JS's case, Pediatric Nutrition to follow growth and need for continued nasogastric supplementation, Pediatric Neurology, and the Neonatal Follow-Up Program, should all be arranged prior to discharge to ensure timely appointments. Finally, if the infant qualifies, Early Intervention (EI) should be contacted to arrange for services to promote optimal developmental outcome.

Although infants with CHD are often seen by a variety of health care professionals, the PCP is often the person who sees the infant most regularly, is the first contact when a parent has concerns, and is the ideal person to coordinate services to ensure the infant receives appropriate care and referrals. Therefore, it is imperative that he or she be aware of their unique needs. In addition to standard care, each visit should include a physical assessment, paying particular attention to assessment of the cardiorespiratory system and incision site, evaluation of growth parameters, development, and the infant-caregiver attachment relationship. Every visit should also include a review of the medications being administered, confirmation of appropriate dosing, and a discussion with the family regarding compliance. Decisions regarding activity restriction, immunizations, need for Palivizumab (Synagis) or bacterial endocarditis prophylaxis must be made according to the infant's cardiac defect. Any questions or concerns should be openly discussed with the cardiologist to ensure safe and appropriate care. A more thorough review of the primary care role in infants with CHD is beyond the scope of this article, but those interested in more information on this topic should refer to Smith (2001).

CONCLUSION

The infant with flail tricuspid valve is a rare event, however quick identification and appropriate surgical management is the key to improving mortality in these unique cases. Nurse practitioners (NPs) who care for infants in the first hours of life are the ideal providers to recognize these critical babies, provide life-saving stabilization, arrange for transport to appropriate centers for care, and then ensure appropriate discharge education and follow-up. Similarly, the primary care NP in the community has the potential to have a critical impact on health maintenance and long-term outcomes of these special children and their families.

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