

REVIEW

Navigating Neonatal Cardiovascular Emergencies: A Comprehensive Narrative Review on Assessment and Intervention Strategies

Stefana Maria Moisa^{1,2}, Criscentian Brinza^{3,4}, Mariana Floria^{3,5}, Alexandru Burlacu^{3,4}

¹ Department of Pediatrics, Faculty of Medicine, “Grigore T. Popa” University of Medicine and Pharmacy, Iași, Romania

² “Sfanta Maria” Clinical Emergency Hospital for Children, Iași, Romania

³ Faculty of Medicine, “Grigore T. Popa” University of Medicine and Pharmacy, Iași, Romania

⁴ “Prof. Dr. George I.M. Georgescu” Institute of Cardiovascular Diseases, Iași, Romania

⁵ Medical Clinic, “Sf. Spiridon” Emergency Hospital, Iași, Romania

ABSTRACT

The evaluation and management of congenital heart diseases (CHDs) in neonates are pivotal aspects of pediatric care given the substantial prevalence of this condition. This review emphasizes the urgency of timely recognition and intervention, as a notable proportion of affected neonates face critical illness shortly after birth. The multidisciplinary approach involving neonatologists, pediatricians, and obstetricians is essential to address the diagnostic and therapeutic challenges inherent in neonatal cardiac emergencies. The insights presented herein aim to guide medical practice, enhance early recognition, and ultimately improve outcomes for newborns grappling with critical heart conditions. Key proactive measures advocated include prenatal screening, facilitating early identification of potential cardiac anomalies. Advanced imaging techniques, such as fetal echocardiography, have a crucial role in enabling detailed examinations, contributing to the prenatal diagnosis of major CHDs. The review underscores the importance of preparing medical teams during the prenatal period for potential postnatal complications, emphasizing the necessity of a holistic approach to the care of neonates who survive critical episodes. Continued research and clinical advancements remain imperative to refine strategies and optimize care for this vulnerable patient population. The provided data serve as a valuable guide for healthcare professionals and could improve the outcomes of neonates confronting the challenges of CHD.

Keywords: neonatal cardiac emergency, cardiogenic shock, congenital heart malformation, newborn arrhythmia

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CORRESPONDENCE

Criscentian Brinza

Bd. Carol I nr. 50

700503 Iași, Romania

Fax: +40 232 219 270

Email: criscentian.brinza@d.umfiasi.ro

INTRODUCTION

The evaluation and management of congenital heart diseases (CHDs) in neonates is of particular interest, as 7–9 out of 1,000 live born children are affected by this condition and up to 25% become critical soon after birth.^{1,2}

Neonatologists and pediatricians need to be able to promptly recognize patients with congenital heart de-

fects, as their status can easily deteriorate, and late diagnosis can lead to death.^{2,3} Cardiac emergencies in newborns may pose significant diagnostic and therapeutic challenges due to the complexity of possible scenarios. Newborns may present as ‘gray babies’ in case of cardiogenic shock (due, for instance, to ductal dependent systemic circulation malformations), as ‘blue babies’ in ductal dependent pulmonary circulation malfor-

mations, or as 'pink babies' in congestive heart failure. Furthermore, any number of arrhythmias may present during neonatal life or even during fetal life, requiring early delivery.³

Early detection and intervention in CHDs require a collaborative effort between healthcare professionals, including obstetricians and neonatologists. Prenatal screening has a crucial role in identifying potential cardiac anomalies, allowing for proactive measures and counseling for parents.^{4,5} Advances in imaging techniques, such as fetal echocardiography, enable a more detailed examination of the fetal heart, facilitating the early diagnosis of structural abnormalities.⁵ A study that enrolled 9,782 neonates and infants reported that the introduction of echocardiography as a screening test increased the prenatal diagnosis rate with 44% in case of major CHD, including hypoplastic left heart syndrome, double outlet right ventricle, and severe pulmonary stenosis.⁵ The data strongly indicate that implementing a cardiac center program for prenatal screening has the potential to significantly enhance the early detection of major CHDs. Such a proactive approach not only facilitates the timely identification of critical cardiac anomalies but also equips the medical team with essential insights, enabling them to better prepare for the management of cardiovascular emergencies.⁵

Timely identification during the prenatal period prepares medical teams for potential postnatal complications. Beyond the immediate medical concerns, addressing the long-term implications of CHDs in neonates is equally important. Children who survive critical episodes may face developmental challenges, necessitating a holistic approach to their care.¹

Concerning clinical presentation, any newborn presenting with shock should first undergo septic and hypovolemic shock screening, as these are the two most frequent causes of shock during the first month of life.^{1,6,7} If cardiogenic shock is suspected, the four most frequent causes are congenital heart malformations, arrhythmia, cardiac muscle diseases, or cardiac metabolic disorders, and the baby should be transferred to a pediatric cardiology facility or pediatric intensive care unit.^{6,8}

The aim of this narrative review was to comprehensively explore the critical aspects regarding the diagnosis and management of cardiovascular emergencies in neonate patients. By systematically addressing the complexities of neonatal cardiovascular emergencies, this review could provide valuable insights that can inform medical practices, enhance early recognition, and improve outcomes for newborns facing critical heart conditions.

INITIAL EVALUATION

The initial evaluation starts with the clinical examination, which allows a prompt inclusion into one of the four main categories: cyanosis, shock, arrhythmia, and congestive heart failure.

The 'five Ts' mnemonic provides a framework for facile recall of prevalent cardiac etiologies contributing to cyanosis in the neonatal period.^{9,10} These encompass dextro-Transposition of the great arteries, Tetralogy of Fallot, Truncus arteriosus, Total anomalous pulmonary venous connection, and Tricuspid valve abnormalities. Other less common malformations that can cause cyanosis are hypoplastic left heart syndrome, pulmonary atresia, anomalous systemic venous connection, double outlet right ventricle, or the single ventricle.^{9,10}

The presence of differential cyanosis or reversed differential cyanosis can provide valuable diagnostic insights in a clinical context. Differential cyanosis is characterized by the manifestation of cyanosis in both lower extremities, while the right arm or both arms remain pink.^{11,12} The explanation to this phenomenon is that in patients with pulmonary arterial hypertension and patent ductus arteriosus (PDA), the deoxygenated blood in the pulmonary artery enters the PDA and travels to the aorta and from thereon to the lower half of the body. The color of the left arm depends on where the PDA enters the aorta: if below the left subclavian artery, then the left arm is pink, as it receives blood from the left ventricle, same as the right arm. If the PDA enters the aorta above the left subclavian artery, then the left arm is cyanotic. Differential cyanosis also appears in persistent pulmonary hypertension of the newborn associated with critical aortic stenosis or coarctation, interrupted or hypoplastic aortic arch.^{11,12}

Reverse differential cyanosis is encountered in newborns with transposition of the great arteries associated with high pulmonary vascular resistance, coarctation of the aorta, or interrupted aortic arch. In both cases, the descending aorta receives blood from the pulmonary circulation via a patent PDA, therefore the legs are pink and the arms are cyanotic.^{11,13}

Several ductal-dependent lesions should be suspected whenever differential cyanosis is observed, including critical aortic stenosis, critical coarctation of the aorta, or interrupted aortic arch.^{11,12} Conversely, classical cyanosis may indicate numerous ductal-dependent lesions, such as hypoplastic left heart syndrome, tetralogy of Fallot with pulmonary atresia, pulmonary atresia with intact interventricular septum, critical pulmonary stenosis, tricuspid atresia (if there is also severe right ventricular outflow

TABLE 1. The most frequent causes of cyanosis and differential cyanosis and their relationship to the ductus arteriosus

Type of cyanosis	Type of lesion	Ductal-dependent
Cyanosis	Hypoplastic left heart syndrome	Yes
	Tetralogy of Fallot with pulmonary atresia	Yes
	Pulmonary atresia with intact interventricular septum	Yes
	Critical pulmonary stenosis	Yes
	Tricuspid atresia	Yes
	Severe neonatal Ebstein anomaly	Yes
	Transposition of the great arteries	Yes
	Truncus arteriosus	No
	Total anomalous pulmonary venous connection	No
Differential cyanosis	Critical aortic stenosis	Yes
	Critical coarctation of the aorta	Yes
	Interrupted aortic arch	Yes

tract obstruction), severe neonatal Ebstein anomaly (in this case a patent ductus is required to maintain pulmonary blood flow until the pulmonary vascular resistance diminishes), or transposition of the great arteries. Cyanosis could be also observed in various non-ductal-dependent lesions, such as truncus arteriosus or total anomalous pulmonary venous connection (Table 1).¹⁴

All cyanotic CHDs are accompanied by central cyanosis. However, several non-cardiac conditions can also cause cyanosis, such as airway obstruction (laryngotracheomalacia or choanal atresia), hemoglobinopathies, neurologic diseases leading to hypoventilation (intraventricular hemorrhage, apnea of prematurity, cerebral infection, neonatal myasthenia gravis, Wernig–Hoffman disease, or phrenic nerve injury) and pulmonary diseases leading to ventilation/perfusion mismatch (pulmonary hypoplasia or hemorrhage, hyaline membrane diseases, emphysema, atelectasis, pleural effusion, or pneumothorax) or impaired alveolar–arterial diffusion (fibrosis or edema).^{11,15} Therefore, neonates presenting with cyanosis warrant a thorough diagnostic investigation to discern potential major CHDs that necessitate immediate intervention.¹¹

A facile diagnostic assessment with a primary utility in discerning between cardiac and respiratory etiologies of cyanosis is the hyperoxia test. During the test, neonates are subjected to a 10-min administration of 100% oxygen. The anticipated physiological response involves a rise in PaO₂ levels to >300 mmHg. A PaO₂ value persisting below 150 mmHg suggests potential cardiac mixing lesions or persistent pulmonary hypertension. In cases in which the obtained value falls within the range of 150–300 mmHg, the likelihood of methemoglobinemia, central nervous system disorders, or pulmonary disorders is consider-

able.^{10,15} Nevertheless, the hyperoxia test may be falsely positive in hypoplastic left heart syndrome and total anomalous pulmonary venous return, which may respond to oxygenation. The test may be falsely negative in massive intrapulmonary shunts.¹

ADVANCED EVALUATION

Low cardiac output is the hallmark of cardiogenic shock (of any cause). In this particular clinical context, newborns can exhibit pale or cyanotic skin, diminished peripheral pulse, cold extremities, delayed capillary refill time, the presence of the third or fourth heart sound, tachycardia, diminished urinary output, signs of hepatic or pulmonary congestion, irritability, or lethargy.^{6,8,14}

Any neonatal patient with a cardiovascular emergency must be admitted to a newborn intensive care unit. The monitoring protocol encompasses electrocardiography, temperature assessment, measurement of arterial and central venous pressure, as well as peripheral saturation. Laboratory tests should include lactate levels, troponin, and B-type natriuretic peptide, and other markers of heart failure, myocardial dysfunction, or injury.^{16,17}

A chest X-ray is imperative for comprehensive evaluation, capable of revealing not only data concerning the cardiac silhouette but also indicators of oligemia or pulmonary edema. Echocardiography stands as a proficient modality for the discernment of diverse cardiac malformations. It can provide valuable insights into cardiac anatomy, the ejection and shortening fractions, as well as myocardial performance index. If transthoracic echocardiography images are not satisfactory for any reason, transesophageal echocardiography should be performed (after sedation and

intubation).¹⁸ In patients who underwent surgical correction of CHD, cardiac computed tomography or heart catheterization are required to properly characterize the function of shunts and the integrity of anastomoses.^{19–21}

Venous oximetry can be performed in the superior vena cava, given that many patients usually have a superior vena cava catheter inserted as a central venous access. Central venous oxygen saturation is an important parameter that should be measured in a continuous or intermittent manner, in response to clinical status changes or after interventions.²² Establishing a target threshold of greater than 50% is recommended for effective neonatal management.²³

Cerebral and somatic near-infrared spectroscopy is a relatively new monitoring method that relies on the observation that oxygenated and deoxygenated hemoglobin absorb infrared light differently. It can be used to assess adequate tissue oxygen delivery (class II, level of evidence B recommendation) despite the lack of randomized clinical trials to date.²⁴

Thermodilution cardiac output effectively measures cardiac index, stroke volume, and vascular resistance, and offers data that can be used to calculate oxygen delivery and consumption.²⁵ The method requires a pulmonary artery catheter to be placed, therefore it has limited indications. If the pulmonary artery catheter is inserted, boluses of cold saline are injected during expiration. The drop in temperature is then measured using a thermistor near the catheter tip. Using this method in patients with low cardiac output despite standard therapy is a class IIa, level of evidence C recommendation.²²

If patients have no intracardiac shunting, lithium dilution, transpulmonary thermodilution, and pulse contour analysis are new methods to measure cardiac output. Despite seemingly offering data superior to echocardiography measurements, more data is required before entering standard clinical use.^{22,25}

CARDIAC ARRHYTHMIA

Bradyarrhythmias and tachyarrhythmias represent other neonatal cardiovascular emergencies.^{26,27} Commonly seen arrhythmias include junctional ectopic tachycardia, atrial tachycardias and atrioventricular reentrant nodal tachycardia, sinus bradycardia, ventricular premature contractions, ventricular tachycardia, and fibrillation. Children who underwent heart surgery may develop second- or third-degree AV block, right or, less commonly, left bundle branch block, intra-atrial reentrant tachycardia, or sinus bradycardia.^{26,27}

An important aspect of arrhythmia prevention relies on normal potassium and magnesium levels.²⁸ Normal calcium and sodium levels are also desirable. The pH level is also important, as acidosis promotes ventricular arrhythmia and should be corrected either by administering sodium bicarbonate or increasing minute ventilation.²⁶

The pharmacological treatment of acute arrhythmias depends on the arrhythmogenic substrate and type of arrhythmia. Atrial flutter, atrial fibrillation, as well as atrioventricular nodal reentrant tachycardia may respond to procainamide (10–15 mg/kg loading dose over 30–45 min; infusion 20–40 µg/kg/min). If procainamide proves ineffective, amiodarone should be used (the loading dose of 5 mg/kg i.v. in 1–2 h in order to prevent acute alpha blockade, can be repeated up to three times; infusion 15–20 mg/kg/24 h). Adenosine (50–200 µg/kg rapid bolus) can stop reentrant supraventricular tachycardias but not atrial ectopic tachycardia and junctional ectopic tachycardia. Ectopic atrial foci are better suppressed by esmolol. Verapamil should not be used in newborns.²² Ventricular arrhythmias usually require lidocaine (1–2 mg/kg loading dose followed by 20–50 µg/kg/min infusion), sotalol (1 mg/kg i.v. over 60 min), or amiodarone.²²

Cardioversion is also feasible in this age group. Currents of 0.5–1 J/kg are used after adequate analgesia and sedation. The second shock, if required, should have a double intensity. Defibrillation for ventricular fibrillation or pulseless ventricular tachycardia requires higher energies, 2–5 J/kg. Intractable arrhythmias may benefit from extracorporeal membrane oxygenation, ventricular assist device support, or ablation.^{22,26}

HEMODYNAMIC MANAGEMENT OF CRITICAL CONGENITAL HEART DISEASE PATIENTS

Therapeutic goals in critical patients include improving central and peripheral perfusion, and oxygen delivery and consumption. The following therapeutic classes are available for pediatric use (drugs and doses are presented in Table 2): inotropes (dopamine, dobutamine, epinephrine), vasodilators (nitroprusside, nitroglycerin, inhaled nitric oxide, prostaglandins, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, sildenafil), inodilators (milrinone, levosimendan), chronotropes (isoproterenol), vasoconstrictors (norepinephrine, phenylephrine, vasopressin), and β-Adrenergic antagonists (esmolol).²² The vasoactive inotrope score and index have been used in children who underwent congenital heart disease surgery and the index predicted poor outcomes if above 3. The index takes into consideration the duration of

TABLE 2. Pediatric doses of frequently used hemodynamic drugs

Drug	Dose
Dopamine	2–28 µg/kg/min
Dobutamine	2–20 µg/kg/min
Epinephrine	0.02–0.2 µg/kg/min
Nitroprusside	0.2–5 µg/kg/min
Nitroglycerin	0.2–10 µg/kg/min
Prostaglandin E1	0.01–0.2 µg/kg/min
Enalapril	0.1 mg/kg/day qd/bid
Captopril	0.1–0.5 mg/kg/day q8h
Lisinopril	0.07 mg/kg/day qd
Losartan	0.1 mg/kg/day
Sildenafil	0.5–2 mg/kg
Milrinone	Loading 25–100 µg/kg Infusion 0.27–0.75 µg/kg/min
Levosimendan	Loading 6–12 µg/kg Infusion 0.05–0.1 µg/kg/min
Isoproterenol	0.01–0.2 µg/kg/min
Norepinephrine	0.02–0.2 µg/kg/min
Phenylephrine	0.02–0.3 µg/kg/min
Vasopressin	0.01–0.05 U/kg/h
Esmolol	100–700 µg/kg/min

the required inotrope support as well.²²

Nutrition is another critical issue for acutely ill newborns. Adequate caloric intake is required to promote protein catabolism.²⁷ In cases in which gastrointestinal integrity is preserved, the preferred nutritional options include breast milk, fortified breast milk, or appropriate formulas. Conversely, if gastrointestinal function is compromised, central parenteral nutrition, incorporating dextrose, amino acids, and intralipid constitute alternative solutions. Enteral nutrition (nasogastric or nasoduodenal feeds or even feeds via a gastrostoma) is preferred whenever possible because even small amounts prevent intestinal villi involution.²⁸

CONCLUSIONS

The evaluation and management of CHDs in neonates represent crucial aspects of pediatric care, given the significant prevalence of this condition. Timely recognition and intervention are imperative, as a substantial number of affected neonates become critically ill soon after birth. This review underscores the importance of a multidisciplinary approach, involving neonatologists, pediatricians, and obstetricians, in addressing the diagnostic and therapeutic challenges posed by cardiac emergencies in newborns. The insights presented herein aim to guide medical practices, enhance early recognition, and ultimately im-

prove outcomes for newborns facing critical heart conditions. Continued research and clinical advancements remain essential to refine strategies and optimize care for this vulnerable patient population.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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