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RESEARCH ARTICLE

A Brazilian Neonatal Unit's Experience with Functional Echocardiography and Diagnostic Findings in Congenital Heart Disease

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ABSTRACT:

Introduction: The diagnosis of congenital heart diseases is a challenge for pediatricians due to the diversity of clinical presentations and symptoms, just like several other diseases prevalent in the neonatal period. Often the diagnosis is established during the gestation period, sometimes the hypothesis is formulated by neonatal screening. However, several cases will require clinical suspicion from the pediatrician. Given the possibility of this diagnosis, the evaluation of the infant cardiologist is essential. Unfortunately, this professional is not always readily available. In the intensive care units, health professionals use heart rate, blood pressure, capillary refill, urinary output and acid-base balance for hemodynamic analysis. These data are significant, but nonspecific. Thus, functional echocardiography can improve hemodynamic evaluation with positive impact on diagnosis. Our objective was to conduct a retrospective analysis of hospitalizations in a Brazilian neonatal unit, with the goal of identifying the diagnoses of congenital heart diseases made by a researcher using functional echocardiography, from July 2015 to September 2017, having as inclusion criteria; clinical manifestations of distress or respiratory failure, characterized by tachypnea, signs of dyspnea, need for oxygen therapy or ventilatory support and/or hemodynamic instability, characterized by tachycardia, cyanosis, cold extremities, prolonged capillary filling time, fine pulses and/or hypotension, low urinary output.

Results: During the 26 months of the study, 362 neonates were hospitalized. Of these, 223 were included in the research for presenting cardiorespiratory manifestations. We observed three cases of obstructive shock related to cardiac tamponade, 18 cases of congenital heart disease, five cases of hypertrophic myocardiopathy, seven cases of myocardial dysfunction not related to structural heart disease, 46 cases of neonatal persistent pulmonary hypertension and 27 cases of patent ductus arteriosus (six with hemodynamics repercussion).

Conclusions: The results showed functional and/or anatomical changes in 105 patients analyzed (46.7%), identified a diversity of structural cardiopathies, improved hemodynamic evaluation with impact on the therapeutic approach. While using functional echocardiography in neonates, clinicians should be aware that undiagnosed critical congenital heart defects can present during this period. They should acknowledge the limitations of skills, and it should not be used as a screening tool for diagnosing or excluding congenital heart defects. A patient with a suspected critical congenital heart defect should be quickly referred to a pediatric cardiologist, even if this implies out-of-hospital patient's transportation. We assume that the systematic implementation of this technology by the medical team can improve diagnostic accuracy, which can positively impact the therapy used.

Keywords: Critical care, neonatology, ultrasonography, echocardiography, point-of-care systems, cardiac tamponade, hypertension pulmonary, heart defectal congenits.

Introduction

The diagnosis of congenital heart diseases (CHD) represents a challenge for pediatricians, considering the various presentations and possible confusion with other diseases prevalent in the neonatal period. Fetal morphological ultrasonography performed by the radiologist in the first trimester of pregnancy seeks to analyze the existence of the four cardiac chambers, as well as analysis of the exits of the ventricles. When risk of CHD is considered, fetal echocardiography is appropriate from 20 weeks of gestational age (preferably between 24 and 28 weeks of gestation) and may direct birth to units specialized in cardiopathies.[1] However, most of the Brazilian population cannot perform this examination and we believe that this situation is like several other countries. A few years ago, pediatric societies suggested neonatal screening by pulse oximetry. [2] But this screening is indicated for over 35 weeks of gestational age and clinically stable and does not prevent diagnostic loss despite good sensitivity. Thus, the suspicion of CHD should be based on clinical data such as murmur, tachycardia and tachypnea, cyanosis, shock. But these presentations may be related to the transition from fetal to neonatal circulation or manifestation of other diseases prevalent in the neonatal period. In the suspicion of CHD, echocardiography societies suggest clinical and imaging evaluation by a specialist with training in congenital cardiopathies. Unfortunately, these professionals are not always readily available. Hemodynamic monitoring in neonatal intensive care unit (NICU) is based on the evaluation of heart rate, blood pressure, urinary output, acid-base balance and capillary filling time. Although these data are important and useful, more detailed hemodynamic information, such as myocardial contractility, valve function and pulmonary blood pressure estimation are limited.[3-6] Thus, the use of functional echocardiography (echo) has gained interest in the NICUs for the analysis of myocardial function, pulmonary and systemic blood flow, intra and extra cardiac shunts and tissue perfusion, measurement of the velocities in the exit pathways and analysis of valve regurgitations.[3-6] Its limitation lies in the fact that it is performed by a general practitioner who may not observe structural defects that justify the clinical manifestation of the patient. There is special concern with cardiopathies such as anomalous pulmonary veins drainage and coarctation of the aorta (CoAo), which pose diagnostic challenge even for the specialist. That is, ideally, echo can improve evaluation and monitoring, but it should only be performed after evaluation of the specialist when CHD is a possibility. As the specialist was not always readily available, we sought to analyze how the identification of CHD occurred in a neonatal intensive care unit with echo performed by an intensive care unit.

Material and Method:

1) STUDY LOCATION:

Hospital Santa Isabel de Ubá-MG, philanthropic institution that serves users of the Unified Health System, agreements and individuals, reference for high-risk pregnant women in the micro-region, totaling approximately 500,000 inhabitants. It has 168 beds, six neonatal intensive care beds and four pediatric intensive care beds.

2) ETHICAL ASPECTS:

The work was approved by the Human Research Ethics Committee of the Federal University of Viçosa and registered at Plataforma Brasil under number 64217116.8.0000.5153.

3) EXPERIMENTAL DESIGN:

We retrospectively analyzed patients hospitalized in the NICU of the Santa Isabel Hospital from June 2015 to September 2017. Inclusion criteria: patients aged zero to 28 days, admitted or who had developed manifestations of distress/respiratory failure (tachypnea, dyspnea, need for oxygen supplementation to maintain saturation above 90%) and/or hemodynamic instability (prolonged capillary filling time, fine pulses, tachycardia, hypotension) that also underwent evaluation of the researcher in the period of said instability, with the realization of the echo. Those patients who underwent echocardiography with an imaging specialist had a report of their examination filed for comparison with the analysis performed by the researcher.

4) CARDIAC ULTRASONOGRAPHY OR FUNCTIONAL ECHOCARDIOGRAPHY:

Patients, after clinical evaluation of the researcher, were submitted to echo, using a portable GE Vivid device with the use of a sectorial transducer. The examination was performed with the patient in supine position or left lateral decubitus if clinical conditions allowed. The purpose of the echo in the NICU was to seek indicative signs of dysfunction, without having to exhaust resources related to the anatomical definition of the organ. It sought punctual responses such as the presence or absence of pericardial effusion, systolic and diastolic myocardial dysfunction, in addition to evaluating blood flow through the chambers, perform objective and subjective analysis of the ejection fraction of the left ventricle, and shortening fraction of the left ventricle, calculate cardiac output of the left ventricle, pulmonary and aortic flow velocity, to analyze the existence of intra and extra cardiac

shunt, to investigate signs indicative of neonatal persistent pulmonary hypertension and to diagnose patent ductus arteriosus (PDA). The acoustic windows used were the same as the specialist uses: subcostal, apical, four chambers, longitudinal long and short axis, suprasternal. In the subcostal and apical windows, the objective was to visualize the four chambers, observe size, muscle mass, contractility, atrioventricular and ventricular-arterial agreement, pericardial effusion research. In longitudinal windows, evaluate pericardial effusion, left ventricular contractility, ejection fraction and left ventricular shortening. In the longitudinal window short axis, to evaluate trunk and bifurcation of the pulmonary artery, research of patent arterial channel and analysis of the left ventricle. In the suprasternal window, visualize aortic arch and descending aorta with analysis of flow velocity in the descending aorta.

5) STATISTICAL ANALYSIS:

The descriptive analysis was presented in absolute numbers, relative frequencies and measures of central tendency (median and interquartile range) after using the Kolgomorov-Smirnov test.

6) RESULTS:

During the 26 months of the study, 362 neonates were hospitalized. Of these, 223 were eligible for the research because they presented respiratory and/or hemodynamic manifestations, fulfilling the inclusion criteria. As for the age of admission to the research, this varied from the first hour of life to 25 days of life, 33 with a median of one day. The other demographic characteristics of these patients are displayed in Table 1.

Table 1. Characteristics of the sample of patients admitted to the NICU, Ubá-MG, from June 2015 to September 2017.

Male gender	136(61%)
Female Gender	86 (36,6%)
Gestational Age (weeks)	
23-28 weeks and 6 days	34(15,2%)
29-31 weeks and 6 days	27(12,1%)
32-36 weeks and 6 days	94(42,2%)
37 weeks onwards	68 (30,5%)
Weight (grams)	
Less than 1,000 grams	28(12,6%)
1,000 to 1,499 grams	42(18,8%)
1,500 to 2,499 grams	75 (33,6%)
Above 2,499 grams	78 (35%)

Variables expressed in absolute and relative frequencies.

The results showed functional and/or anatomical changes in 106 patients (46.7%). Of these 106 patients, 39 had changes in the therapeutic approach as can be seen in table 2. There were three cases of obstructive shock secondary to cardiac tamponade related to the use of central peripheral insertion catheter (PICC) and the echo is decisive in the immediate diagnosis and therapeutic indication. The echo identified 18 cases of CHD (two complete atrioventricular canal defects, four partial atrioventricular canal defects, three of interventricular communication, three of pulmonary stenosis, two of transposition of large vessels, one of tetralogy of Fallot, two of hypoplastic left heart syndrome and one of coarctation of the aorta. Five cases of hypertrophic myocardiopathy related to hyperinsulinism and seven clinical situations with systolic dysfunction unrelated to structural cardiopathies were observed. There was one case of pulmonary stenosis and one case of CoAo diagnosed by the specialist echocardiographer and not described by the researcher. There were also two other cases of CoAo not diagnosed by the specialist echocardiographer or the researcher, with conclusion performed by computed tomography angiography. We identified 46 cases of persistent pulmonary hypertension of the newborn (PPHN): in 14 cases (30%), the medical team did not consider this hypothesis as a cause of cardiorespiratory manifestations. We identified 27 cases of PDA, six of which showed signs of hemodynamic repercussion.

Table 2: Findings using functional echocardiography and therapeutic changes.

	Diagnosis	Procedure or therapeutic change in the diagnostic group
Cardiac Tamponade	3(1,3%)	3 (100%)
PPHN	46 (20,4%)	20 (43,4%)
CHD	18 (8%)	5 (27,7%)
Hypertrofic cardiomyopathy	5 (2,2%)	2 (40%)
Myocardial dysfunction without stuctural defect	7 (3,1%)	3 (42,8%)
PDA	27 (12%)	6 (22,2%)
Total	106 (47,1%)	39 (36,8%)

Source: researcher. PPHN: Persistent Pulmonary Hypertension of the Newborn, CHD: Congenital Heart Disease, PDA: Patent Ductus Arteriosus.

Considering the 106 patients with altered echocardiogram, 55.2% had exams performed by the researcher and the specialist echocardiographer. Cases of myocardial dysfunction, channel dependent cardiopathies and PPHN started treatment before the specialist's evaluation since it was not always readily available. Thus, the main purpose of the examination by the specialist was to rule out CHD not identified by the researcher (a case of pulmonary stenosis was diagnosed in this context). Patients identified as having CHD by echo were evaluated by the specialist, with good diagnostic agreement. However, the specialist observed unidentifiable anomalies in the echo in five cases, which did not alter the initial conduct.

Discussion: Anatomical or functional cardiovascular disorders are frequent occurrences in NICU, requiring high index of clinical suspicion and adequate evaluation. Diagnosis is made based on clinical history, physical examination, chest radiography, electrocardiogram, and often echocardiography.[3-7] In this way, the availability of imaging specialist cardiologists is considered essential for complete analysis. Unfortunately, most hospitals do not have a full-time specialist available. In addition, functional disorders may require frequent reassessments for the analysis of the evolutionary picture and response to drugs.[4] Thus, the acquisition of ability in echocardiographic examination has been gaining interest among intensivists.[3-7] In several European countries the form of certification of echo is discussed. The main medical societies consider important the distinction between basic and advanced training.[8] Thus, it is expected that the professional with basic training can recognize pericardial effusion, subjectively ventricular dysfunction, regurgitation, and valvular dysfunction. The professional with advanced training can assess subjectively and objectively the ejection fraction, measure cavity dimensions, measure flow velocity and cardiac output, analyze transmittal blood flow, analyze blood flow through cavities, distinguish the presence of anatomical abnormalities, but always recognizing the limitations of the technology in use.[9] In our study, we aimed to retrospectively describe the findings of echo by an intensive, non-specialist imaging, as a complement to clinical examination and to analyze the functional diagnoses and findings of CHD. We observed three cases of cardiac tamponade related to the use of PICC, and in two cases there was no clinical suspicion. In all three cases, the echocardiographic diagnosis was fundamental for deciding on the need for pericardiocentesis. PPHN was the hemodynamic disease with the highest series. PPHN is characterized by maintenance of high pulmonary vascular resistance, right-to-left shunting of blood through fetal circulatory pathways and hypoxemia. It is often associated with systemic hypotension and low cardiac output due to increased right ventricular afterload and myocardial dysfunction.[10] 46 cases of PPHN were identified. It was possible to establish this diagnosis based on the estimation of pulmonary blood pressure from the analysis of tricuspid regurgitation speed (when the hypothesis of obstruction in the right ventricular outflow route was ruled out) or through indirect signs, as deviation of the interventricular septum to the left, right-left or bidirectional deviation through the patent of the ductus arteriosus. [3,6,10] We identified 27 cases of PDA, six with signs of hemodynamic repercussion, characterized by increased left chambers, pseudo normal flow pattern by the mitral valve, high flow velocity through the arterial channel and reduction of diastolic pressure in the descending aorta with visualization of diastolic retrograde flow. Works performed by Seghal et al. and El-Khuffash et al. show that a score obtained from echocardiographic data selects patients in whom the patent canal closure has an impact on the outcome.[11,12] In the future, double-blind randomization may clarify whether canal closure in selected cases has an

impact on the outcome of bronchopulmonary dysplasia and neonatal mortality. Several CHD were identified, but there was no statistically significant number to analyze the diagnostic agreement between researcher and specialist for each of them. The studies suggest that performing functional echocardiography does not rule out the evaluation of the specialist, who is the qualified professional for diagnosis of congenital heart diseases. [3,6,8] However, considering the local difficulties regarding the availability of the specialist, something common to several other services in Brazil, it is attractive to imagine the aid of this technology by intensivist. Although the number of cases is limited, we had good agreement, including the correct diagnosis of transposition of large vessels, tetralogy of Fallot, pulmonary stenosis, atrioventricular septal defect (partial form and complete form), interventricular communication, hypoplastic left heart syndrome. The researcher's examination did not perfectly complete all the anomalies but allowed functional analysis and appropriate therapeutic approach. Five cases of hypertrophic myocardiopathy were identified: four were children of diabetic mothers and one was a carrier of persistent neonatal hypoglycemia. There was diagnostic failure in one case of pulmonary stenosis, and researcher performed diagnosis of PPHN. In both situations, there is tricuspid regurgitation, but the diagnosis of pulmonary hypertension can only be considered when ruling out obstruction in the right ventricular outflow tract. In this case, the researcher did not measure the speed of pulmonary blood flow, which was elevated and compatible with obstruction in the right ventricular outflow tract. As for CoAo, the final diagnosis was established in four cases, two suggested by the echocardiographer and two diagnosed by computed tomography angiography based on clinical suspicion. In one of the two cases diagnosed by the specialist, the researcher suggested the existence of the disease. Considering the availability of internet, it is possible to send an image for discussion with a specialist, which is particularly attractive for units that do not have an echocardiographer 24 hours a day. It is essential that the emergency physician recognize its limitations and, considering the suspicion of congenital heart disease, request evaluation of an echocardiographer.

Recent cardiac echo guidelines have emphasized the importance of close collaboration among neonatologists, pediatric cardiologists, and radiologists.[8,13] A rapid bedside cardiac ultrasound can provide critical information in neonatal emergencies while waiting for comprehensive echocardiography.[13] We recognized as a major limitation of this work the fact of being retrospective. In addition, the researcher had access to the clinical data of the patients. Although this is a limiting fact, it is important to remember that we consider imaging as an extension of physical examination and not its substitute.

Conclusion: The echo allowed the diagnosis of obstructive shock by cardiac tamponade, indicating pericardiocentesis in a timely manner, identified a diversity of structural cardiopathies, improved hemodynamic evaluation with impact on the therapeutic approach. It is important for clinicians to be aware that undiagnosed critical congenital heart defects can arise just during this time when using echo in neonates. Acknowledging their limitations is important, and it should not be used as a screening tool for diagnosing or excluding congenital heart defects. It is crucial to quickly refer a patient with a suspected critical congenital heart defect to a pediatric cardiologist, even if it requires transportation from an out-of-hospital patient. Our assumption is that the medical team's systematic performance of this technology can enhance diagnostic accuracy, which can have a positive impact on the therapy used.

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