Original Article

Observed Causes of Severe Respiratory Distress among Children with Congenital Heart Disease

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Abstract

Background: Severe respiratory distress is a clinical feature commonly observed among children with congenital heart disease (CHD), but the underlying cause is often misdiagnosed. **Objectives:** This study is aimed at determining the common causes of severe respiratory distress observed among children with CHD. **Methods:** This study was a retrospective study on children who had severe respiratory distress with underlying CHD seen between June 2017 and June 2018, and were consecutively recruited from two teaching hospitals. **Results:** Forty-seven children aged 2 months to 15 years were admitted for severe respiratory distress secondary to CHD at our center between June 2017 and June 2018. The most common CHD was isolated ventricular septal defect (VSD), which made up 36.2%, followed by tetralogy of Fallot with 23.4%. Identifiable causes of respiratory distress in these children were restrictive airway disease (36.2%) as the most common cause, followed by pulmonary edema from congestive cardiac failure (27.7%). Among children with heart failure, 57.1%, 47.4%, 0%, and 50% of infants, children aged 1–5, 6–10, and above 10 years, respectively, were affected. **Conclusion:** Restrictive airway disease was noted as the most common cause of severe respiratory distress among children with CHD.

Keywords: Children, congenital heart disease, restrictive airway disease, severe respiratory distress

Introduction

Some pediatricians have been faced with the dilemma of encountering a child where both CHD and respiratory disease coexist. It has even been postulated that congenital cardiovascular anomalies are significantly associated with congenital and acquired respiratory disorders. Although the prevalence of asthma and/or airway hyperresponsiveness in children with CHD is not known, some authors have suggested that hyperactive airway disease is more common in children with CHD than in the general population.[2,3] For instance, Bode-Thomas *et al*.[4] noted a coexistence of ventricular septal defect and hyperactive airway in two Nigerian children, they managed with bronchodilators and steroids, and the children responded well. In developing countries like ours, the attending physician may note that the coexistence of these two disease entities may lie mainly in the fact that both could present with similar symptomatology. This could lead to a delayed diagnosis, especially when there is a low index of suspicion.[5-8]

A careful search has shown that the prevalence of severe distress in children with CHD is rarely discussed.

In this study, we attempt to highlight the commonly observed causes of severe respiratory distress among children with CHD. Early identification of the exact cause of respiratory distress in this age group will help avert the numerous complications, morbidity, and mortality that follow it.

Methods

This study was a retrospective study in which children with CHDs seen between June 2017 and June 2018, were consecutively recruited. The hospitals receive referrals of children with CHD from within the state and beyond.

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Study area
The study was carried out at the University of Nigeria Teaching Hospital and Enugu State University Teaching Hospital.

Diagnostic criteria
Severe respiratory distress
This was diagnosed with a history of dyspnea, flaring of ala-nasi, subcostal and intercostal recession, and tracheal tug with oxygen saturation below 94%.

Reactive airway disease
Reactive airway disease (RAD) was diagnosed as clinical features of wheeze or bronchial spasm, without any prior history of asthma.[9] It is important to note that wheeze is quite common in infants, and only a minute fraction of infants who wheeze ever suffer bronchial asthma.[9] It is therefore pertinent to note that instead of calling these groups of children as "wheezy," clinicians opt for the term "reactive airways" to refer to this group of children.[9]

Pneumonia
Pneumonia is an inflammation of the lung parenchyma with variable etiologies.[10] We made this diagnosis from a history of fast breathing, cough, and clinical signs of chest retractions and crepitation. It was finally proven with a chest X-ray showing patchy opacities.[10]

Heart failure
Diagnosis of congestive heart failure was made with a history of difficulty in breathing, dyspnea on exertion, tachycardia, and tender hepatomegaly with a displaced apex.[11]

Pulmonary edema
Diagnosis of pulmonary edema was made clinically when we got a history of a sudden onset of extreme breathlessness, sweating, and cough. Chest auscultation reveals fine, crepitant rales, and occasional rhonchi or wheezes, especially inspiratory. Chest X-ray evidence of Kelly B lines or fluffy opacities confirms the diagnosis.[11]

Metabolic acidosis
We made a diagnosis of metabolic acidosis with a history of deep breathlessness, plasma bicarbonate concentration below 20 mmol/L, and decreased pH equal to or below 7.20.[12]

Sample selection
A total number of 200 children with CHD were diagnosed using echocardiography in the study period, of which 47 presented with severe respiratory distress.

Study population
The subjects studied included children between the ages of 1 month and 15 years who were admitted for severe respiratory distress secondary to congenital cardiac anomalies. Subjects excluded, were those with respiratory distress not stemming from congenital cardiac anomalies and those with severe bronchopneumonia alone or hypoxemia not stemming from cardiac disease. Children with CHD who had respiratory distress without fulfilling the defined criteria for severe respiratory distress were also excluded.

Children who fulfilled the inclusion criteria were consecutively recruited into the study.

Information on sociodemographic characteristics, echocardiographic findings/diagnosis of the congenital heart defect, as well as laboratory estimations of hemoglobin concentration, serum electrolytes, urea and creatinine, chest X-ray, and blood culture were documented.

Anthropometric indices and oxygen saturation were also elucidated. A careful history and physical examination were used with the aid of laboratory investigation to delineate the cause of severe respiratory distress.

Data analysis
Data were analyzed using Epi Info and SPSS, version 20 (Chicago). Frequency and percentages were used for categorical data. Mean and standard deviation were used to summarize the details of the data that were normally distributed.

Ethical consideration
Ethical clearance for the study was obtained from the Research and Ethics Committee of the University of Nigeria Teaching Hospital, Enugu. The IRB institution name is University of Nigeria Teaching Hospital, Ituku-Ozalla, Enugu, the approval number is IRB 00002323, and the approval date is February 18, 2019.

Results
A total number of 200 children with CHD were diagnosed using echocardiography in the study period, of which 47 presented with severe respiratory distress. This gives a prevalence of 23.5% of children with CHD who presented with severe respiratory distress.

Forty-seven of them presented with respiratory distress. The children comprised 25 (53.2%) males and 22 (46.8%) females, with a mean age of 3.9 (4.5) years. Fourteen (29.8%) of the children were infants, 19 (40.4%) were children under 5 years, 6 (12.8%) were children aged 6–10 years, while 8 (17.0%) were above 10 years of age. The mean weight and height were 14.4 kg (13.1) and 97.6 cm (31.6), respectively. Among parents who gave information on social class, 18 (38.3%) of them belong to the middle socioeconomic class, 11 (23.4%) to the lower class, while 13 (27.6%) to the upper class.

The most common CHD was isolated ventricular septal defect (VSD) which made up 17 (36.2%), followed by tetralogy of Fallot (TOF) which made up 11 (23.4%) of CHDs. Table 1 shows the frequency of occurrence of CHD in this study. The common presenting features are as shown in Table 2. The most common feature necessitating presentation to hospital was fast breathing which occurred in 41 (87.2%) of the patients while convulsion occurred in only 3 (6.4%) patients.

Identifiable causes of severe respiratory distress in these children are as shown in Table 3, implicating RAD in 17 (36.2%) as the most common cause, followed by congestive

cardiac failure in 13 (27.7%). Among children with heart failure, 57.1%, 47.4%, 0%, and 50% of the infants, children aged 1–5, 6–10, and above 10 years, respectively, were affected.

Of 17 children with isolated ventriculoseptal defect (VSD), 4 (23.5%), 6 (35.2%), and 4 (23.5%) had congestive heart failure, RAD, bronchopneumonia as the cause of respiratory distress, respectively, while anemia, infective endocarditis, and metabolic acidosis contributed 1 (2.1%) each. Among children with TOF, 6 (54.5%) had RAD, 2 (18.2%) were hypoxic, while bronchopneumonia, pulmonary edema, and metabolic acidosis contributed 1 (9.1%) each. Two (66.7%) of the three children with atrioventricular septal defect had RAD as the cause of respiratory distress, and 1 (33.3%) had cardiac failure [Table 4].

Discussion

We noted an overall prevalence of severe respiratory distress in this study as 23.5%. This is very high and depicts the importance of this study. Children with CHD usually present with respiratory distress, but the actual cause of this distress is not well elaborated in many studies. The actual mechanism of respiratory distress among infants with CHD is not well appreciated, making the management difficult.[13] This study showed that RAD is the major cause of severe respiratory distress in more than half of the subjects. It is important to note that bronchoconstriction and prolonged expiratory distress seen in infants with CHD have been erroneously blackmailed as cardiac asthma. Snashall and Chung[14] pointed out the fact that small and large airway narrowing in infants with CHD could be precipitated by acute elevation of pulmonary or bronchial vascular pressure. This could be due to reflex bronchoconstriction triggered by C-fibers with their endings in the lung parenchyma, bronchi, and pulmonary blood vessels. They also pointed out that bronchial responsiveness to bronchoconstriction drugs is increased in infants with left ventricular failure partly due to reflex mechanisms.[14]

Again, it has been reported that mucosa inflammation of the bronchus may also contribute to this respiratory distress and bronchodilator drugs have been useful.[8,14,15]

Furthermore, it was noted by Morgan *et al*.[15] that bronchial obstruction or hyperactive airway disease triggered by this CHD can cause retention of fluids and indeed create a nidus for infections, leading to bronchopneumonia. This increased narrowing and retention of secretion and eventual infections can also cause respiratory distress among infants with CHD.[8,14,15]

Matsuoka *et al*.[16] also confirmed RAD as the most common cause of respiratory distress in their study. They noted the effects of pulmonary congestion on the development of atopic asthma in 31 infants with CHD. They noted that respiratory distress did not resolve after surgery of the underlying CHD in seven patients, six of whom had a family history of allergy.

Table 1: Specific frequencies of identified congenital heart diseases

CHD	Frequency (%)
VSD	17 (36.2)
TOF	11 (23.4)
AVCD	3 (6.4)
RHD	2 (4.3)
TOF+PDA	2 (4.3)
Others	12 (25.5)
Total	47 (100)

Others: DORV, VSD+ASD, Isolated PDA, Single ventricle, TAPVR with dextrocardia, TGA, Isolated ASD. VSD: Ventricular septal defect, ASD: Atrial septal defect, PDS: Personal digital assistant, TGA: Transposition of great artery, TOF: Tetralogy of Fallot, PDA: Patent ductus arteriosus, CHD: Congenital heart disease DORV: Double outlet right ventricle, AVCD: Atrioventricular canal defect, RHD: Rheumatic heart disease, TAPVR: Total anomalous pulmonary venous drainage

Table 2: Frequency of common symptoms and signs in children with congenital heart disease

Symptom	Frequency (%)
Fast breathing	41 (87.2)
Poor weight gain	24 (51.1)
Breathlessness	23 (48.9)
Cough	21 (44.7)
Easy fatigability	17 (36.2)
Fever	14 (29.8)
Cyanosis	9 (19.1)
Generalized weakness	6 (12.8)
Bilateral leg swelling	4 (8.5)
Convulsion	3 (6.4)

Table 3: Identified causes of respiratory distress in children with congenital heart disease

Cause of respiratory distress	Frequency (%)
Atopy	17 (36.2)
Heart failure (Pulmonary edema)	13 (27.7)
Pneumonia	5 (10.6)
Hypoxia	5 (10.6)
Atopy with hypoxia	2 (4.3)
Metabolic acidosis	2 (4.3)
Anemia	1 (2.1)
Atopy+infective endocarditis	1 (2.1)
Unidentified cause	1 (2.1)
Total	47 (100)

It is therefore pertinent to note that pulmonary congestion in infancy may increase the risk of RAD in genetically predisposed children.

Airway hyperactivity and obstruction as the causes of respiratory distress in children with CHD have been postulated by Kussman *et al*.[17] They noted that extrinsic airway compression must be considered in the presence of respiratory insufficiency.[17] They noted that airway compression is very

Table 4: Specific congenital heart diseases and identified causes of respiratory distress

CHD	No	Cause of respiratory						
		Atopy	Pulmonary edema	Pneumonia	Acidosis	Hypoxia	Anemia	In
VSD	17	23.5	35.2	23.5	2.1	-	2.1	2.1
TOF	11	54.5	9.1	9.1	9.1	18.5	-	-
AVCD	3	66.7	33.3	-	-	-	-	-
RHD	2	50.0	-	50.0	-	-	-	-
PDA*	2	50.0	50.0	-	-	-	-	-
Others**								

*TOF with PDA, In- infective endocarditis, **The frequency of other CHDs is one each. CHD: Congenital heart disease, PDA: Patent ductus arteriosus, TOF: Tetralogy of Fallot, VSD: Ventricular septal defect, AVCD: Atrioventricular canal defect, RHD: Rheumatic heart disease

common with some underlying cardiac anomalies, such as those that cause left-to-right shunt which in turn leads to dilatation of the pulmonary arteries.

We noted pulmonary edema leading to congestive cardiac failure as the second most common cause of respiratory distress in this study. It has been opined in some studies.[8,18,9] They noted that the major pathological pathway of respiratory distress in infancy with CHD mainly stems from pulmonary edema and metabolic acidosis.[19] It is important to note that respiratory distress induced by pulmonary edema can arise from left ventricular failure causing volume overload from a left-to-right shunt as in the ventricular septal defect (VSD) and patent ductus arteriosus (PDA).[19] It could also arise from pressure impact from obstruction of left ventricular outflow such as aortic stenosis and coarctation of the aorta. Impairment of pulmonary venous return to the left ventricle as in total anomalous pulmonary venous drainage could also result in pulmonary edema.[19]

It is pertinent to note that recognition of pulmonary edema from congestive heart failure due to left-to-right shunts in infants is crucial though may be difficult, especially in neonates. This is because the onset could initially be insidious. It could just present with feeding difficulties and fatigability. However, the constant finding among them is tachypnea. Among the signs usually observed, the presence of enlargement of the liver and displaced apex was the reliable sign for the diagnosis of heart failure even in patients with the primary lesions in the left heart.

Indeed, it remains a puzzle for the paediatrician, that frequent episodes of respiratory distress in infants with congenital cardiac defects are being treated as bronchopneumonia or even as bronchial asthma, without necessarily knowing that it is actually from left ventricular failure with attendant pulmonary edema.

It is rather surprising to note that majority of children with respiratory distress have been erroneously managed as respiratory pathology without giving credence to the cardiac disease. Granted that cardiac diseases are easily diagnosed when there is a murmur, yet there is some congenital heart lesion that may never produce a murmur. Children with such congenital heart disease without clinical murmurs may still present with respiratory distress from possible pulmonary oedema. This situation may mimic respiratory disease, and thus, pulmonary edema may be missed and mismanaged.

We also reported a few cases of bronchopneumonia as the causes of respiratory distress among children with CHD.[20] Most previous reports identified CHD as an underlying cause of recurrent pneumonia, i.e., when there are two or more pneumonia episodes in a year.[20,21] Children with a large-sized ventricular septal defect and PDA tend to present early and have more severe disease, including pneumonia. This is because left-to-right shunt lesion usually causes over flooding of the lungs and thus creates a niche for bacterial infections.

Metabolic acidosis is another cause of severe respiratory distress reported in our study. Reduction in systemic arterial oxygen tension results in marked anaerobic respiration and release of organic acids, especially lactic acids.[22] The elaboration of hydrogen ions and severe hypoxemia from metabolic acidosis stimulates the respiratory center and triggers distress. In children with a right-to-left shunts, there is marked decrease in systemic venous blood to the lungs such that metabolic acidosis remains uncompensated. To do here is the use of oxygen and correction of acidosis.[22] It is important to note that cardiac and pulmonary correlates of respiratory distress among infants with CHD are intertwined. A study[23] has also noted that this similar pathophysiology makes the management of patients with CHD all the more complex. They noted that respiratory distress in children with CHD could be structural as a result of compression causing laryngomalacia.

There was no significant difference in SpO_2 between the male and the female children with CHD though the mean saturation is 89% in room air. This simply means that hypoxia also plays a role as a cause of respiratory distress among children with CHD, especially the right-to-left shunt lesion. The pathway of respiratory distress from hypoxemia is the same as that from metabolic acidosis.[24] In addition, hypoxia due to CHD has also been linked to abnormal neurogenesis and impaired cortical growth. Hypoxemia also has a link with pulmonary edema. Hypoxemia impairs airway epithelial Na+ transport, which is important in the reabsorption of lung fluids.

It is very noteworthy to point out that anemia is not one of the causes of respiratory distress as seen in this study. The mean hemoglobin concentration was 12.4 g/L (3.7). Contrary to our findings, Dimopoulos *et al*.[25] noted a prevalence of anemia to be 13.1% and was common in patients with congenitally corrected transposition of great arteries and Ebstein anomaly of

the tricuspid valve. The most common pathogeneses of anemia in their study were iron deficiency and the use of diuretics.[25]

When we separate the causes of respiratory distress into age groups, we still noted atopy to be common across all age groups, except 6–10 years of age. In addition, on breaking the cause of respiratory distress into types of lesion, atopy is more common in TOF while pulmonary edema from heart failure is more common in a ventricular septal defect. This can simply be explained from the fact that left-to-right shunt lesion is more predisposed to lung over flooding, heart failure, and pulmonary edema while those with a right-to-left shunt have their lungs spared from over flooding but may present with bronchospasm due to polycythemia and hypoxia.

Conclusion

RAD is noted as the most common cause of respiratory distress among children with CHD across all age groups. Among those with congestive heart failure, infants are commonly affected.

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Conflicts of interest

There are no conflicts of interest.

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