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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, 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 congenital heart disease.

Methods: This study was a retrospective study in which children who had severe respiratory distress with underlying congenital heart disease seen between June 2017 and June 2018 were consecutively recruited from two teaching hospital.

Results: Forty-seven children aged 2 months to 15 years were admitted for severe respiratory distress secondary to Congenital Heart Disease (CHD) at our centre between June 2017 and June 2018. The commonest CHD was isolated Ventricular Septal Defect (VSD) which made up 36.2%, followed by Tetralogy of Fallot (TOF) 23.4%. Identifiable causes of respiratory distress in these children were restrictive airway disease (36.2%) as the most common cause, followed by pulmonary oedema from congestive cardiac failure (27.8%). 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. The commonest Congenital Heart Disease (CHD) was isolated Ventricular Septal Defect (VSD) which made up 17 (36.2%), followed by Tetralogy of Fallot (TOF) which made up of 11 (23.4%) of congenital heart diseases.

Conclusion: Restrictive airway disease was noted as the commonest cause of severe respiratory distress among children with congenital heart disease. Among those with congestive heart failure, infants were more commonly affected.

Keywords: Severe respiratory distress, Restrictive airway disease, Congenital heart disease, Children.

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Introduction

Congenital cardiovascular abnormalities and severe respiratory diseases are two highly linked entities and looks inseparable. It is noted that although severe respiratory distress from whatever cause is common in children, they may be life-threatening especially to infants with congenital heart disease.

Some paediatricians have been faced with the dilemma of encountering a child where both Congenital Heart Disease (CHD) and respiratory disease co-exist. 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 Hyper-Responsiveness (AHR) in children with Congenital Heart Disease (CHD) is not known, some authors have suggested that hyperactive airway disease is commoner in children with Congenital Heart Disease (CHD) than in the general population. For instance, Bode Thomas et al, noted a co-existence of ventricular septal defect and hyperactive airway in two Nigerian children, she managed with bronchodilators and steroids and they responded well. In developing countries like ours, the attending physician may note that the co-existence of these two disease entities may lie mainly in the fact that both could present with similar symptomatology. This could lead to the delayed diagnosis especially when there is a low index of suspicion.

Furthermore, severe respiratory distress in infants with congenital heart disease has also been noted in idiopathic pulmonary arterial hypertension, a rare disorder that is progressive and often lead to right ventricular failure. It may take several months to diagnose with dyspnoea on exertion being the most common presenting complaint. Respiratory distress among children with congenital cardiac anomaly may be caused by broncho-pneumonia. For instance, refractory *Mycoplasma pneumoniae* was isolated by Zhal et al., in infants with congenital heart disease who presented with respiratory

distress and early application of broncho-alveolar lavage shortened the duration of fever and hospitalization, improved laboratory indices and promoted the resolution of wheezing and atelectasis.

In addition, it is also notable that some of the causes of severe respiratory distress in infancy could be due to anomalies of respiratory tract. For instance, Efrati et al., noted that more than 3% of children with cardiac disease also exhibit airway issues, including tracheo-bronchomalacia and airway narrowing, which are known to be latent causes and aggravating factors of persistent and recurrent lower respiratory infections.

Lee et al., also noted these structural defects from the respiratory tract as a common cause of respiratory distress in this age group with Congenital Heart Disease (CHD). Several cases of respiratory distress among infants with congenital heart disease have always been mismanaged as bronchopneumonia or cardiac failure with its attendant morbidity and mortality.

This work is very rare as this is the very first of its kind done in this vicinity. A careful search has shown that the prevalence of severe distress in children with congenital heart disease is rarely discussed.

In this study, we attempt to highlight the commonly observed causes of severe respiratory distress among children with congenital heart disease. 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.

This work is therefore embarked upon to look at observed causes of respiratory distress among infants with congenital heart diseases and associated factors. It also harnessed the prevalence of severe respiratory distress stemming from congenital heart disease. This study will go a long way as an eye opener and to help improve the wellbeing of infants and children with congenital heart disease.

Materials and Methods

This study was a retrospective study in which children with congenital heart diseases seen between June 2017 and June 2018 were consecutively recruited. The hospital receives referral of children with congenital heart disease within the state and beyond.

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, 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. 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. It is therefore pertinent to note that instead of calling these group of children as “wheezy”, clinicians opt for the term "reactive airways" to refer to this group of children.

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

Heart failure: Diagnosis of congestive heart failure was made with history of difficulty in breathing, dyspnea on exertion, tachycardia, tender hepatomegaly with displace apex.

Pulmonary edema: Diagnosis of pulmonary edema was made clinically when we got a history of a sudden onset of extreme breathlessness, sweating, 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 confirm the diagnosis.

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

Sample selection

A total number of 200 children with congenital heart disease was diagnosed by means of 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 are those with respiratory distress not stemming from congenital cardiac anomalies, those with severe broncho-pneumonia alone or hypoxaemia not stemming from cardiac disease. Children with congenital heart disease 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 socio demographic characteristics, echocardiographic findings/diagnosis of the congenital heart defect, laboratory estimation of haemoglobin concentration, serum electrolytes, urea and creatinine, chest x ray and blood culture were documented.

Anthropometric indices and oxygen saturation were also elucidated. 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 analysed using Epi Info and SPSS, version 20. 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.

Results

A total number of 200 children with congenital heart disease was diagnosed by means of echocardiography in the study period, of which 47 presented with severe respiratory distress. This gives a prevalence of 23.5% of children with congenital heart disease who presented with severe respiratory distress.

Among children aged 1 month to 15 years, who presented with congenital heart disease between June 2017 and June 2018, forty-seven of them presented with respiratory distress. The children comprised 25 (53.2%) males and 22 (46.8%) females with mean age of 3.9 (4.5) years. Twenty-nine point eight percent 14 (29.8%) of the subjects were infants, 19 (40.4%) were children under five years, 6 (12.8%) were children 6-10 years while 8 (17.0%) were above 10 years. The mean weight and height were 14.4 kg (13.1) and 97.6 cm (31.6) respectively. Thirty-eight point three percent 18 (38.3%) of the patients belong to the middle socioeconomic class, 11 (23.4%) to the lower class, while 13 (27.6%) belong to the upper class (Table 1).

CHD	Frequency	Percent
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
Note: Others: DORV, VSD+ASD, isolated PDA, single ventricle, TAPVR with dextrocardia, TGA, isolated ASD		

Table 1. Specific frequencies of identified congenital heart diseases.

The commonest Congenital Heart Disease (CHD) was isolated Ventricular Septal Defect (VSD) which made up 17 (36.2%), followed by Tetralogy of Fallot (TOF) which made up of 11 (23.4%) of congenital heart diseases. Table 1 shows the frequency of occurrence of Congenital Heart Disease (CHD) in this study. The common presenting features are as in Table 2. The commonest 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%).

Symptom	Frequency	Percent
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 2. Frequency of common symptoms and signs in children with CHD.

Identifiable causes of severe respiratory distress in these children are as shown in Table 3, implicating Reactive Airway Disease (RAD) 17 (36.2%) as the most common cause, followed congestive cardiac failure 13 (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.

Cause of respiratory distress	Frequency	Percent
Atopy	17	36.2
Heart failure (Pul. oedema)	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

Table 3. Identified causes of respiratory distress in children with CHD.

Out of 17 children with isolated Ventriculo Septal Defect (VSD), 4 (35.3%), 6 (23.5%), 4 (23.5%), had congestive heart failure, reactive airway disease, bronchopneumonia as cause of respiratory distress respectively while anaemia, infective endocarditis and metabolic acidosis contributed 1 (2.1%) each. Among children with Teratology of Fallot (TOF), 6 (54.5%) had RAD, 2 (18.2%) were hypoxic while bronchopneumonia, pulmonary oedema 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).

CHD	No	Cause of respiratory distress (%)						
		Atopy	Pulmonary oedema	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	-	50	-	-	-	-
*PDA	2	50	50	-	-	-	-	-
**Others								
Note: *TOF with PDA, In-infective endocarditis, **the frequency of other CHDs is one each								

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

In Table 4, RAD was observed to be the commonest cause of respiratory distress across the different age categories, except in those 6-10 years in whom hypoxia dominated as a cause.

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. We could not compare this high incidence with any study as careful search showed none. Children with congenital heart disease 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 infant with congenital heart disease is not well appreciated, making even management difficult.

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, prolonged expiratory distress seen in infants with congenital heart disease has been erroneously blackmailed as cardiac asthma. Snashaall et al. pointed out the fact that small and large airway narrowing in infants with congenital heart disease could be precipitated by acute elevation of pulmonary or bronchial vascular pressures. This could be due to reflex bronchoconstriction triggered by C-fibers with their endings in the lung parenchyma, bronchi and pulmonary blood vessels. He also pointed out that bronchial responsiveness to bronchoconstriction drugs is increased in infants with left ventricular failure partly due to reflex mechanisms.

Again, it has been reported that mucosa inflammation of the bronchus may also contribute to this respiratory distress and bronchodilator drugs has been useful. Furthermore, it was noted by Tsubata et al. that bronchial obstruction or hyperactive air way disease triggered by this congenital heart disease can cause retention of fluids and indeed create a nidus for even infections leading to bronchopneumonia. This increased narrowing and retention of secretion and eventual infections can also cause respiratory distress among infants with congenital heart disease.

Matsuoka et al. also confirmed RAD as the commonest cause of respiratory distress in his study. He noted the effects of pulmonary congestion on the development of atopic asthma in 31 infants with congenital heart disease. They noted that respiratory distress did not resolve after surgery of the underlying congenital heart disease in seven patients, six of whom had a family history of allergy. 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 a cause of respiratory distress in children with congenital heart disease has been postulated by Kussman et al. He noted that extrinsic airway compression must be considered in the presence of respiratory insufficiency. He noted that airway compression is very 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 oedema leading to congestive cardiac failure as the second commonest cause of respiratory distress in this study. It has been opined in some studies. They noted that the major pathological pathway of respiratory distress in infancy with congenital heart disease mainly stems from pulmonary oedema and metabolic acidosis. It is important to note that respiratory distress induced by pulmonary oedema can arise from left ventricular failure causing volume overload form a left to right shunt like in Ventricular Septal Defect (VSD) and Patent Ductus Arteriosus (PDA). It could also arise from pressure impact from obstruction of left ventricular outflow such as aortic stenosis, coarctation of the aorta etc. Impairment of pulmonary venous return to the left ventricle like in total anomalous pulmonary venous drainage could also result in pulmonary oedema. It is pertinent to note that recognition of pulmonary oedema 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 paediatrician that frequent episodes of respiratory distress in infants with congenital cardiac defects are been treated as bronchopneumonia or even as bronchial asthma without necessarily knowing that it is actually from left ventricular failure with attendant pulmonary oedema.

It is rather surprising to note that majority of children with respiratory distress has been erroneously managed as respiratory pathology without giving credence to the cardiac disease. Granted that cardiac disease are easily diagnosed when there is murmur and such respiratory distress in them will be duly attended to by decongesting the pulmonary oedema, yet there is gamut of congenital heart lesion which may never give you a murmur such as Transposition of Great Artery (TGA) with intact septum, a very large ventricular-septal defect, cortriatum sinistrum and yet may present with respiratory distress. This situation may mimic respiratory disease and thus pulmonary oedema may be missed and mismanaged.

We also reported few cases of bronchopneumonia as causes of respiratory distress among children with congenital heart disease. Most previous reports identified Congenital Heart Disease (CHD) as an underlying cause of recurrent pneumonia *i.e.,* when there are two or more pneumonia episodes in a year. Children with large sized ventricular septal defect and patent ductus arteriosus tend to present early and have more severe disease including pneumonia. This is because left to right shunt lesion usually cause over flooding of the lungs and thus create 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. The elaboration of hydrogen ions and severe hypoxaemia from metabolic acidosis stimulates the respiratory centre and triggering distress. In children with right to left shunts, there is marked decrease in systemic venous blood to the lungs such that this metabolic acidosis remains uncompensated. Some clinicians therefore tend to misdiagnose this as heart failure and manage with digoxin and thus causing worsening acidosis. The best thing to do here is the use of oxygen and correction of acidosis. It is important to note that cardiac and pulmonary correlates of respiratory distress among infants with congenital heart disease are intertwined. Yamagish et al. also noted that this similar pathophysiology makes management of patients with Congenital Heart Disease (CHD) all the more complex. He noted that respiratory distress in children with congenital heart disease could be structural as a result of compression causing laryngo-malacia.

There was no significant difference in SpO_2 between the males and the female children with congenital heart disease though the mean saturation is 89% in room air. This simply means that hypoxia also play a role as a cause of respiratory distress among children with congenital heart disease especially the right to left shunt lesion. The pathway of respiratory distress from hypoxaemia is the same as that from metabolic acidosis. In addition, hypoxia due to congenital heart disease has also being linked to abnormal neurogenesis and impaired cortical growth. Hypoxaemia also have a link with pulmonary oedema. This they do by impairing the airway epithelial Na+ transport, which is important in reabsorption of lung fluids.

It is very noteworthy to point out that anaemia is not one of the causes of respiratory distress as seen in this study. The mean

haemoglobin concentration was 12.4 G/L. Contrary to our findings, Konstantino et al. noted a prevalence of anaemia to be 13.1% and was common in patients with congenitally corrected transposition of great arteries and Ebstein anomaly of the tricuspid valve. The commonest pathogenesis of anaemia in their study was iron deficiency and use of diuretic.

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

Conclusion

Reactive Airway Disease (RAD) is noted as the commonest cause of respiratory distress among children with congenital heart disease across all age groups. Among those with congestive heart failure, infants are commonly affected.

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, approval number is IRB 00002323 and approval date is 18/02/2019.

Author Contributions

JMC and EO conceived and designed the study. VO, NU conducted data gathering. BC performed statistical analyses. JMC wrote the article.

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Declaration of Patient Consent

The authors certify that they have obtained all appropriate patient consent forms.

Conflicts of Interest

None.

References

1. Chen T, Qiu L, Zhong L, et al. Flexible bronchoscopy in pulmonary diseases in children with congenital cardiovascular abnormalities. Exp Ther Med 2018; 15: 5481-5486.
2. Qun J, Zhou J, Yu Y. An official Chinese thoracic society clinical pratice guideline: The etiologic assessment of bronchoalveolar lavage in infectious lung disease. Chin J Tuberc Respir Dis 2017; 40: 578-583.
3. Bode-Thomas F, Hyacinth IH, Yilgwan C. Co-existence of ventricular septal defect and bronchial asthma in two Nigerian children. Clin Med Insights Case Rep 2010; 3: 5-8.
4. Hayes Jr D. Idiopathic pulmonary arterial hypertension misdiagnosed as asthma. J Asthma 2007; 44: 19-22.
5. Zhang Y, Chen Y, Chen Z, et al. Effects of bronchoalveolar lavage on refractory *Mycoplasma pneumoniae* pneumonia. Respir Care 2014; 59: 1433-1439.
6. Efrati O, Gonik U, Modan-Moses D, et al. The role of flexible fibreoptic bronchoscopy in evaluation of pulmonary diseases in children with congenital cardiac disease. Cardiol Young 2007; 17: 140-144.
7. Guillemaud JP, El-Hakim H, Richards S, et al. Airway pathologic abnormalities in symptomatic children with congenital cardiac and vascular disease. Arch Otolaryngol Head Neck Surg 2007; 133: 672-676.
8. Lee SL, Cheung YF, Leung MP, et al. Airway obstruction in children with congenital heart disease: Assessment by flexible bronchoscopy. Pediatr Pulmonol 2002; 34: 304-311.
9. Gheorghiade M, Follath F, Ponikowski P, et al. Assessing and grading congestion in acute heart failure: A scientific statement from the acute heart failure committee of the heart failure association of the European Society of Cardiology and endorsed by the European Society of Intensive Care Medicine. Eur J Heart Fail 2010; 12: 423-433.
10. Jung B, Martinez M, Claessens YE, et al. Diagnosis and management of metabolic acidosis: Guidelines from a French expert panel. Ann Intensive Care 2019; 9: 1-7.

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