Co-existence of Ventricular Septal Defect and Bronchial Asthma in Two Nigerian Children

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Abstract: Congenital heart diseases (CHD) often present with recurrent or chronic breathing difficulties, as do chronic airway diseases such as asthma. Both are relatively common, and may sometimes co-exist. However, there is a paucity of literature from developing countries to that effect. We present two children diagnosed with ventricular septal defect, later also found to have clinical features consistent with co-existing asthma. We highlight the diagnostic challenges we encountered as well as the crucial role of a careful family respiratory history in children with congenital heart disease.

Keywords: Congenital heart disease, CHD, asthma, ventricular septal defect
Introduction
Clinicians are sometimes faced with patients in whom both Congenital Heart Disease (CHD) and airway disease co-exist. Indeed, congenital cardiovascular anomalies are believed to be significantly associated with congenital and acquired airway disorders. Although the prevalence of asthma and/or airway hyper-responsiveness (AHR) in children with CHD is not known, some authors have suggested that asthma or AHR is more common in children with CHD than in the general population. Tsubata and co-workers reported that 6 out of 10 of their patients with congestive heart failure (CHF) secondary to CHD had AHR elicited by histamine challenge, while Ackerman et al identified an extremely strong association between a particular CHD (pulmonary atresia with VSD) and persistent AHR. Matsuoka et al have suggested that pulmonary congestion in infancy may increase the risk of atopic asthma in genetically predisposed children. Overall, there is a paucity of literature, particularly from developing countries, concerning the co-existence of airway disease including asthma and AHR, with CHD.

For clinicians practicing in a developing country, the clinical significance of the co-existence of these two conditions may lie mainly in the fact that both could present in a similar manner, since many CHD as well as asthma, present chiefly with respiratory distress. This could lead to the delayed diagnosis of one or the other—especially when there is a low index of suspicion. In this report, we attempt to highlight these diagnostic problems by presenting our experience with two cases of VSD who were later diagnosed as having co-existing bronchial asthma.

Case Reports
Case 1
J.I. was first admitted with bronchopneumonia and congestive heart failure at the age of one month. He had had breathing difficulties since birth, but developed cough, catarrh and fever one week before presentation. A loud harsh pan-systolic murmur at the left lower sternal edge, typical of a VSD was also present. J.I was treated with antibiotics and diuretics, and discharged home. The VSD was confirmed by echocardiography at the age of 5 months. He had several other episodes of bronchopneumonia throughout early childhood, which were usually treated with oral antibiotics on outpatient basis. His mother learnt to recognize the symptoms (breathing difficulties with or without fever) early and admitted that she often treated him at home with antibiotics even before the next clinic visit. At the age of 7 years, J.I underwent successful closure of his VSD. A few weeks later, his younger sister was diagnosed with childhood asthma when she presented with cough, breathlessness and wheezing. Family history revealed that their mother and another older brother were also asthmatic. Approximately four months after J.I’s surgery, he presented at follow-up clinic with cough and difficulty in breathing similar to some of the previous episodes he had had before the surgery. There was no associated fever. On examination, he was dyspnoeic, tachypnoeic and had widespread rhonchi in both lung fields. He responded dramatically to inhaled salbutamol (delivered via a metered dose inhaler fitted with a spacer device). Both J.I and his sister are doing well on PRN inhaled salbutamol. His mother reported that J.I only needed to use the inhaler about once a month.

Case 2
M.Y. also suffered during infancy, from recurrent bronchopneumonia and congestive cardiac failure (CCF) secondary to a large ventricular septal defect (VSD), which necessitated several hospital admissions. The large VSD was confirmed by echocardiography at the age of 5 months. At about 2 years of age, M.Y. was again admitted, supposedly for CCF secondary to the VSD. He was extremely dyspnoeic and was placed on intranasal oxygen. Closer examination revealed that M.Y was cyanosed and had a silent chest, despite the extreme dyspnoea. This prompted a diagnosis of acute severe asthma in a child with pre-existing VSD. He died before any further management could be instituted. Subsequently, his mother delivered another male child who had no heart disease but was diagnosed in infancy with asthma and admitted frequently for acute severe asthma. His mother later also admitted to being an asthmatic patient.

Discussion
The first case reported above clearly demonstrates the reversal of symptoms of asthma (respiratory distress and wheezing) after the use of bronchodilator
in a child whose VSD had been surgically corrected. The second patient however died soon after he was noticed to have typical features of acute severe asthma. In both patients, it appears that the loud VSD murmur may have earlier in the course of management, diverted attention away from the possibility of a co-existing asthma. It is possible that many more similar cases have been so missed, and that even when rhonchi are auscultated, they could be ignored or attributed to pulmonary congestion or ‘cardiac asthma’ and cyanosis to severe bronchopneumonia and/or CCF. This is coupled with the fact that the family history in children with CHD is often focused on family history of heart disease, parental age and possible predisposing maternal illnesses in pregnancy, but not particularly on family or maternal history of atopy or chronic respiratory illnesses such as asthma.

Both cases highlight the need for a high index of suspicion of asthma in children with CHD presenting with recurrent respiratory difficulties. Despite their early presentation with obvious clinical features of large VSD confirmed by echo in early infancy, they somewhere along the line (unnoticed by the clinicians), probably developed features of bronchial asthma as well, with the symptoms continuing to be attributed to the VSD. It is possible that some of such ‘chest infections’ that are common in young children with left to right shunt lesions could actually be asthma episodes as these two cases have shown.

The relationship between CHD and pulmonary function is not fully understood. It has been suggested that children with CHD could have several fundamental physiologic mechanisms operating, whereby airflow limitations could occur and result in a higher propensity for developing airway hyper responsiveness than the general population. Matsuoka et al\textsuperscript{1,6,7} postulated that pulmonary congestion in infancy may increase the risk of atopic asthma in genetically predisposed children. Both of our patients were genetically predisposed, with strong family history of asthma and had pulmonary congestion in infancy from large VSD. Other workers have highlighted the role of pulmonary hypertension in some of the patients presenting with asthma-like symptoms. Some cases of pulmonary arterial hypertension (PAH) have been misdiagnosed as asthma, and some of the reports of CHD co-existing with asthma were in patients who also had pulmonary hypertension.\textsuperscript{4,8,10} VSD and several other CHD are often associated with PAH, and our 2 patients are also likely to have had PAH since they both had large VSD. However, in the 2 cases reported by Rothman et al the PAH could not be adequately explained by the cardiac lesion nor by respiratory mechanical factors.\textsuperscript{4} In one report, extrinsic pulmonary airway obstruction by dilated pulmonary arteries was actually demonstrated, via fibreoptic bronchoscopy and computed tomography.\textsuperscript{9} Yet other authors\textsuperscript{10} have suggested that bronchomalacia secondary to external vascular compression, may be the direct cause of the respiratory distress that some children with CHD experience. Nevertheless, the consensus would appear to be that the relationship between CHD and pulmonary function is not yet fully understood and that the prevalence of asthma in children with CHD is still unknown.\textsuperscript{6}

The most obvious conclusion from this report, which is consistent with the experiences of earlier workers,\textsuperscript{5,8} is the necessity of a detailed family history in all children with respiratory problems including those with CHD. Early attention to this detail in case number two might have led to an earlier consideration of asthma as an additional problem, the institution of appropriate management and could possibly have prevented his demise pending the surgical correction of his VSD.

However, further work is required, not only to determine the true prevalence of asthma or AHR in children with CHD, but also the mechanism of the apparently higher propensity for AHR in some CHD.

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**Disclosures**

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