Respiratory Distress due to Bronchial Compression in Persistent Truncus Arteriosus

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Clinical and pathologic findings are described in a seven-month-old boy who suffered from persistent truncus arteriosus and suddenly developed signs of respiratory distress. Bronchial compression in patients with persistent truncus arteriosus has only rarely been reported.

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Respiratory distress in patients suffering from persistent truncus arteriosus is usually due to congestive heart failure. In rare instances, emphysema or atelectasis have been reported, but only two of these reports1,2 specify the cause of the lung abnormalities. Once stenosis of the left main bronchus was observed at autopsy, but without clinical correlation.3 As far as we are aware, only Calder and associates4 have documented a patient with persistent truncus arteriosus and respiratory distress caused by bronchial compression. In this report we document the clinico-pathologic correlation in such a patient.

CASE REPORT

A full-term boy, born after an uneventful pregnancy and weighing 4,000 grams, developed dyspnea and cyanosis at the age of six days. A cardiac murmur was noticed and heart failure developed in a few days. Physical examination revealed slight dyspnea and cyanosis at rest. Auscultation revealed a grade 2/6 ejection murmur with its maximum at the second left intercostal space. Chest roentgenogram demonstrated an enlarged heart with a small vascular pedicle and a left-sided aortic arch. There were prominent vascular markings in both lung fields, which otherwise were normal. The cardiac catheterization data are shown in Table 1; angiography demonstrated the presence of a persistent truncus arteriosus.

Initially, the child improved with a medical regimen of digitalis and diuretics. However, at the age of six months he again became severely dyspneic and rapidly developed signs of heart failure. A chest roentgenogram at that time showed hyperinflation of the left upper lobe with a slight shift of the mediastinum to the right (Fig 1A). Bronchography revealed an obstruction of the left main bronchus at the site of origin of the upper lobe bronchus (Fig 1B). Before further attempts could be made to evaluate this condition, the child deteriorated and died of heart failure at the age of seven months.

Autopsy confirmed the existence of a classic type 1 persistent truncus arteriosus.5 From the truncus a left-sided aortic arch originated which was tightly connected to the main left pulmonary artery by an obliterated ductus arteriosus (Fig 2). The left upper lobe bronchus originated at the usual distance from the carina ruling out the possibility of an eparterial

Focus 1. Anteroposterior chest roentgenogram at age six months. A (left), shows hyperinflation of the left upper lobe. B (right), Bronchographic display of bronchial obstruction at the level of the origin of the left upper lobe bronchus (arrow). Note the situs solitus anatomy of the tracheobronchial bifurcation.
bronchus on the left, as can be seen in situ inversus or the "asplenia syndrome" (see Fig 1B). However, the left main bronchus and the origin of the upper lobe bronchus both displayed a slit-like lumen because the bronchi were squeezed between the aortic arch and the left pulmonary artery (Fig 2).

**Discussion**

In patients with congenital heart defects, in particular those with a left-to-right shunt, bronchial compression by dilated "hypertensive" pulmonary arteries is well documented. It is of interest, therefore, that bronchial compression in patients with persistent truncus arteriosus has hardly been reported. Calder and associates have described such a patient, in whom the development of bronchiectasis required pneumonectomy at the age of nine years. Autopsy revealed a right aortic arch and compression of the right main bronchus. This was the only case of bronchial compression among a series of 100 cases of persistent truncus which they reviewed. A few other reports mention obstructed bronchi, but none of these discusses the mode of origin. Bischoff and associates described a patient with persistent truncus in whom they were unable to advance a bronchoscope through the left main bronchus. It is interesting that the autopsy revealed no gross abnormalities at the site of the anticipated obstruction. One wonders whether external compression by arteries could have been the underlying mechanism, a condition easily missed at autopsy. Rowe and Vlad also reported a patient in whom at autopsy a slightly narrowed left main bronchus was demonstrated. In their case, the bronchus took a more horizontal course than normal, which could be due to dilatation of the left atrium. Victorica and associates, from a survey of 14 patients with persistent truncus, described one patient in whom a stenosis of the left main bronchus was detected at autopsy. Chondromalacia was present at the affected site, but the authors do not discuss the cause of this abnormality.

<table>
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<th>Cardiac Catheterization Data</th>
<th>Oxygen saturation (%)</th>
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<td>Superior caval vein</td>
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<td>Left ventricle</td>
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<td>Truncus arteriosus</td>
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<td>Left pulmonary artery</td>
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<td>60/25</td>
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Figure 2. Photographs of heart specimen exhibiting the abnormal relationship between the great arteries and the left-sided bronchi. A (left), shows left anterior oblique view of the heart-vessel specimen in relation to the bronchial bifurcation. The upper lobe bronchus (LUB) is compressed between the aortic arch (Ao) and the left main pulmonary artery (LPA); the two are tightly joined by an obliterated ductus arteriosus (DA). PTA = persistent truncus arteriosus; LV = left ventricle. B (right) shows a posterior view after cephalic displacement of the aorta (Ao). The left main bronchus at the site of origin of the left upper lobe bronchus (LUB) is markedly compressed (arrows). Note the situs solitus anatomy of the tracheobronchial bifurcation, with a regularly positioned eparterial bronchus (EB) on the right. LA = left atrium.
In our patient, the baby developed respiratory problems, which were shown to be due to bronchial compression. The post-mortem studies revealed that abnormal topography of the pulmonary artery and the main bronchial bifurcation on the left had caused this complication. The normal topography dictates that the left main pulmonary artery will course over the left main bronchus before it sweeps posteriorly, using the upper bronchus as a "hinge." In persistent truncus arteriosus we have always encountered the usual relationship. In the present case, however, the left main pulmonary artery passed anterior to the upper lobe bronchus, a situation normally encountered on the right side. The possibility of bilateral right-sidedness, as in the "asplenia syndrome," or situs inversus were considered, but neither one of these possibilities could be substantiated. The postmortem studies made it absolutely clear that the abnormal topography had played a major role in causing bronchial compression. At the time the patient was clinically evaluated we were not aware of any report of bronchial compression in persistent truncus arteriosus and indeed considered this a less likely phenomenon, particularly since a relatively high origin of the left main pulmonary artery in persistent truncus arteriosus is an established feature of this condition. However, our pathologic studies indicated that the presence of a ligamentum arteriosum may have had a major impact, since it tightly connected both aortic arch and left main pulmonary artery, thereby compressing the upper lobe the main left bronchi (Fig 2). Awareness of the potential of bronchial compression as a cause of respiratory distress in persistent truncus arteriosus might have led us to advocate a more aggressive therapeutic approach.

REFERENCES


Clorial Hydrate Overdose and Cardiac Arrhythmias*

Karen Bowyer, M.D.;** and Stephen P. Glasser, M.D.†

Two cases of clorial hydrate overdose were associated with multiform ventricular tachycardia. Both were refractory to a number of antiarrhythmics, but responded to propranolol administration. Two of nine other cases reported in the literature also responded to beta-blocking agents. Mechanisms for the arrhythmia and its apparent response to beta blockade are discussed.

Eleven cases have now been reported of cardiac arrhythmias (mainly multiform ventricular tachycardia) occurring in subjects without evidence of underlying heart disease following clorial hydrate overdose. Two cases reported herein are such instances, and the literature is reviewed. Although a larger series is necessary to establish the appropriate treatment regimen for ventricular arrhythmias occurring in the setting of clorial hydrate overdose, our review suggests that beta-blocking agents might be the treatment of choice.

CASE REPORTS

CASE 1

A 17-year-old white man ingested 28 clorial hydrate (Noctec) capsules four hours prior to admission. On arrival to the emergency room, he was comatose and an ECG was obtained. Figure 1, panel A, is a rhythm strip taken at this time demonstrating multiform ventricular tachycardia. An endotracheal tube was placed for ventilatory support, and gastric lavage was performed. Lidocaine (75 mg intravenous bolus) followed by an intravenous drip of 4 mg/min resulted in little improvement (Fig 1, panel B). Propranolol (1 mg intravenously), however, resulted in junctional rhythm (Fig 1, panel C). Subsequent chest x-ray film demonstrated pulmonary edema without associated cardiomegaly. Arterial blood gas levels were obtained and the pH was 7.29; Pco₂, 52 mm Hg and the Pco₂, was 34 mm Hg. A flow directed balloon catheter was placed and revealed a pulmonary wedge pressure of 5 mm Hg and pulmonary artery pressure of 25/10 mm Hg. A diagnosis of “noncardiac pulmonary edema” was made, and the patient was treated with general ventilatory support. Within 24 hours, he was awake, alert, and in sinus rhythm. He was discharged to psychiatric care three days later.

CASE 2

A 67-year-old white woman with no known heart disease ingested 60 clorial hydrate tablets (500 mg each, total dose 30 gm) several hours prior to admission. She was found by

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