Bronchial Obstruction Due to Pulmonary Artery Anomalies

II. Pulmonary Artery Aneurysm

By Stephen Contro, M.D., Robert A. Miller, M.D., Harvey White, M.D., and Willis J. Potts, M.D.

In a previous publication attention was drawn to the occurrence of pulmonary artery anomalies as conditions causing tracheobronchial obstruction. In the present report 2 infants are presented in whom a large aneurysm of the pulmonary artery pressed upon the walls of the left bronchus, producing severe respiratory symptoms.

During the past decade vascular malformations of the superior mediastinum have been the object of intense clinical interest. However, the occurrence of anomalies of the pulmonary artery as a cause of tracheobronchial obstruction has gone almost unrecognized. We previously reported 3 cases in which significant respiratory obstruction was produced by a left pulmonary artery coursing anomalously above the right bronchus and behind the trachea, thus producing a "vascular sling."1

The present report is concerned with the description of 2 patients in whom disabling respiratory difficulties resulted from pressure on the left bronchus by a large pulmonary artery aneurysm, a hitherto undescribed condition.

Case Reports

Case 1. D. M., a 3-month-old white boy was admitted to Children’s Memorial Hospital because of "congenital heart disease and dextrocardia." He was the product of an uneventful gestation and delivery. At birth an imperforate anus was found with a tiny fistulous tract, terminating just lateral to the median raphe. This was repaired and subsequently dilated with satisfactory resumption of bowel function. Cyanosis, especially on crying, rapid and labored respirations, and a systolic murmur were noted shortly after birth.

On admission the infant appeared well nourished with mild cyanosis of the lips and extremities. The heart tones were best heard over the right chest and the cardiac impulse was palpable in the right anterior axillary line. A rather harsh systolic

murmur was heard over the entire chest. In the second and third left intercostal space was a loud to-and-fro murmur. The left chest was hyperresonant and rales were heard at the left base. The liver edge was felt 1 inch below the right costal margin. The electrocardiogram revealed upright P waves in the standard limb leads with shift of the transition zone to the right in the chest leads. Roentgenogram (fig. 1) and fluoroscopy of the chest showed a marked emphysema of the left lung with shift to the right of the heart and mediastinal structures. In addition, a large pulsating mass was present in the region of the pulmonary artery segment. A venous angiography revealed opacification of the left atrium from

Fig. 1. Case 1. Roentgenogram in the frontal projection showing marked emphysema of the left lung with displacement of the heart and mediastinum to the right and a large mass in the region of the left pulmonary artery.

From the Departments of Cardiology, Surgery, and Radiology, Children’s Memorial Hospital, Chicago, Ill.
the right atrium and of the aorta from the right ventricle, and the presence of a huge aneurysm of
the pulmonary artery (fig. 2).

Because of the severity of the respiratory symptoms, surgical relief was attempted. Through a
left thoracotomy a huge aneurysmal dilatation of the left pulmonary artery and similar but less
pronounced changes of the right pulmonary artery were found. The left bronchus was collapsed and
remained so when the aneurysmal left pulmonary artery was elevated. Although it was realized that
pneumoneetomy in this age group is almost uniformly fatal, it was thought that the severe
mediastinal shift could be corrected in no other way. The left pulmonary artery was ligated and
the emphysematous left lung was removed. The infant died 10 hours later.

At autopsy a tetralogy of Fallot with a widely patent foramen ovale was found. The main pul-
monary artery measured 1.5 cm. in diameter at its origin from the right ventricle; the left pulmo-
nary artery, which was removed at surgery, 3 cm. at about 0.5 cm. from the bifurcation; the right
pulmonary artery was also dilated and became bulbous at the right hilum.

Case 2. I. T., a white baby girl, born at full
term of an uncomplicated pregnancy, was seen at
the Children's Memorial Hospital at the age of
13 days because of rapid shallow respirations and
cyanosis of the lips and fingertips. On admission
the infant appeared well nourished, tachypneic,
with minimal retractions of the suprasternal notch
and mild cyanosis. The heart tones were best
heard over the right precordium and the point of
maximal pulsation was felt in the fourth right
intercostal space and nipple line. A loud to-and-
from murmur with a continuous thrill was present
over the second and third left intercostal space
parasternally. The lungs were clear. The liver
edge was felt just below the right costal margin.
Roentgenogram (fig. 3) and fluoroscopy of the
chest showed the heart and mediastinal structures
markedly shifted to the right with a large pulsat-
ing density at the level of the pulmonary artery
segment. The left lung was emphysematous. A
venous angiogram (fig. 4) revealed a huge
aneurysmal dilatation of the main and left pul-
monary artery with no definite evidence of shunt
in either direction.

The infant’s hospital course was marked by
recurring episodes of dyspnea and cyanosis. Death
occurred during one such attack.

Postmortem examination revealed tetralogy of
Fallot, aneurysm of the main and left pulmonary
arteries, and obstructive emphysema of the left
lung.
Discussion

Aneurysm of the pulmonary artery is one of the rare lesions of the cardiovascular system, occurring at a rate of approximately 1 in 14,000 autopsies. Exceedingly uncommon are those forms of congenital aneurysm that cannot be accounted for by any known cause of pulmonary artery dilatation, viz., atrial septal defect, ventricular septal defect, patent ductus arteriosus, valvular pulmonic stenosis, and primary pulmonary hypertension.

The symptoms and findings of this lesion were comprehensively reviewed by Boyd and McGavack and by Deterling and Clagett. These include dyspnea, cough, chest pain, hemoptysis, and hilar mass with expansile pulsations. However, a thorough search of the literature failed to reveal any case in which a pulmonary artery aneurysm encroached upon the bronchial tree, producing serious obstructive symptoms.

The 2 cases herein presented are strikingly similar. In both a huge aneurysmal dilatation of the pulmonary artery impinged upon the walls of the left bronchus, leading to obstructive emphysema of the left lung and mediastinal displacement to the right. Unlike other forms of tracheobronchial obstruction of vascular nature, this lesion does not appear to be surgically correctible inasmuch as irreversible changes of the bronchial walls necessitate pneumonectomy, which is usually fatal in the first few months of life.

Summary

Two infants with severe respiratory embarrassment secondary to compression of the left bronchus by a large aneurysmal dilatation of the pulmonary artery are presented. Review of the literature failed to reveal any similar case.

Summario in Interlingua

Es presentate le casos de duo infantes con sever embarrasso respiratori secundari a com-
Pressione del bronco sinistre per un grande dilatazione aneurysmica del arteria pulmonar. Le rivista del litteratura non produceva ulle simile caso.

REFERENCES


A simple safe test for the detection of atrial septal defects which is of value in the symptomless patient is described. This test is based upon the rise in the right atrial pressure above the resting level that occurs immediately after performance of the Valsalva maneuver. With an atrial septal defect or a patent foramen ovale the sudden rise in net right atrial pressure during the first few seconds after the end of the Valsalva maneuver causes a right-to-left pressure gradient across the defect with desaturated blood going into the left atrium. The resulting change in arterial oxygen saturation may be detected by the ear oxymeter method. In 32 patients with suspected atrial septal defects study by means of this test proved to be of value. In 12 in whom the diagnosis was proved by cardiac catheterization or thoracotomy, characteristic and reproducible changes in arterial oxygen saturation were found. In 14 further patients with positive oxymeter responses to the Valsalva test, there was catheter evidence of a left-to-right shunt at the atrial level but definite anatomic proof of an atrial septal defect was lacking. Five patients had a negative response. Three subsequently were found not to have atrial septal defects and 2 had congestive cardiac failure associated with atrial septal defects. In these latter 2 patients, Muller's maneuver or elevation of the legs caused a fall in systemic arterial oxygen saturation, enabling the correct diagnosis to be made.

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