Bronchial Obstruction Due to Pulmonary Artery Anomalies

I. Vascular Sling

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

A left pulmonary artery, coursing anomalously above the right bronchus and behind the trachea en route to the left lung, can produce disabling respiratory symptoms. Three cases observed by the authors are presented in detail and 5 similar cases, previously reported, are reviewed. The clinical features are those of tracheobronchial obstruction, occurring early in life, associated with endoscopic evidence of extrinsic pressure on the right bronchus and posterior wall of the trachea. The importance of the clinical recognition of this anomaly is stressed, since it appears to be amenable to surgical correction.

Recent advances in surgical technics have renewed interest in cardiovascular malformations that cause bronchial or esophageal obstruction. Most of this recent work has been concerned with a variety of anomalies produced by a deranged development of the 6 primitive aortic arches. These include double aortic arch, right aortic arch with ductus or ligamentum arteriosum, and anomalous subclavian or carotid arteries. However, the occurrence of pulmonary artery anomalies as a cause of bronchial obstruction has remained almost wholly unrecognized. In a recent publication one of the authors reported a case in which an anomalous left pulmonary artery embraced the right main bronchus and the lower trachea so tightly that it produced marked narrowing of the airways. Two additional patients have since been observed and with the earlier case form the substance of this report.

Case Reports

Case 1. K. B., a white baby girl was seen at Children's Memorial Hospital at the age of 2½ months, because of spells of dyspnea and cyanosis. She was born at full term of an uncomplicated pregnancy. No abnormality was noted at birth nor during the first month of life. Then she began to have brief episodes of dyspnea and cyanosis, especially after crying. She received x-ray therapy to the anterior chest wall after a roentgenogram of the chest was interpreted as revealing an enlarged thymus gland. Her respiratory symptoms increased both in frequency and severity and were associated with periods of unconsciousness. On admission she appeared moderately dyspneic, but not cyanotic at rest. The suprasternal notch retracted with each inspiration and an inspiratory and expiratory wheeze was present.

Chest films (fig. 1) showed narrowing of the lower part of the trachea and a left apical density, interpreted as a thymic shadow; a marked indentation of the anterior wall of the esophagus was seen just below the aortic arch with no posterior displacement (fig. 2). An electrocardiogram and other laboratory tests were within normal limits.

The diagnosis of a vascular anomaly, causing tracheoesophageal compression was considered, and, because of the severity of symptoms and unfavorable prognosis, surgery was undertaken. After a left thoracotomy, cause of extrinsic tracheal compression was not found. With the chest open, bronchoscopy revealed moderate stenosis of the trachea at the level of the carina, involving as well the opening of the left main bronchus. The right main stem bronchus was almost completely collapsed. The bronchoscope could be passed easily into the lumen of the right bronchus, but upon withdrawal of the instrument compression recurred. A dilator was repeatedly passed through the narrowed area without relief of the obstruction. At this point it was deemed that no further surgery could be performed and the chest was closed. The patient's respiratory symptoms grew worse immediately and death occurred in 3 days.

Postmortem examination revealed tracheal stenosis at the carina, partially involving the opening of the left main stem bronchus; ulcerative

From the Departments of Cardiology, Surgery, and Radiology, Children's Memorial Hospital, Chicago, Ill.

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bronchitis; resorption atelectasis of the left lung; and an anomalous left pulmonary artery. While the right pulmonary artery coursed in a normal direction to the right hilum, the left pulmonary artery crossed anterior to the right main bronchus and encircled the trachea, compressing both structures (fig. 3). It then coursed posteriorly between the trachea and esophagus to emerge on the left side, just superior and posterior to the left main stem bronchus and then entered the left lung.

Case 2. C. D., a white baby boy, the product of a full-term pregnancy and uneventful delivery, was first admitted to Children's Memorial Hospital at the age of 9 days because of episodes of dyspnea and cyanosis. These were noticed immediately after birth, were associated with inspiratory retractions, and bore no relation to feeding. Pertinent physical findings on admission consisted of sternal retractions with each breath, audible prolonged expiratory wheeze, decreased breath sounds over the right chest, and displacement of the cardiac dullness to the left.

X-rays (fig. 4) showed marked emphysema of the right lung with displacement of the mediastinal structures to the left; the mediastinum shifted with each expiration; and an esophogram was normal. Bronchoscopy revealed a normal patent larynx with tracheal deviation to the left. The carina was sharp and the left main bronchus was normal, but the right main bronchus immediately above the upper lobe orifice and the lower trachea was compressed to a small slit by pressure on the lateral wall. The bronchoscope was passed into the bronchus without appreciable resistance, but compression recurred when the instrument was removed. Bronchography (fig. 5) also indicated external pressure on the right main stem bronchus and lower trachea. Laboratory tests including an electrocardiogram were within normal limits.

The hospital course was marked by gradual improvement in the respiratory symptoms, so the child was sent home. He took his feedings well, gained weight, and generally did well for approximately 2 months. Then, spells of cyanosis and dyspnea reappeared with increasing frequency associated with a constant expiratory wheeze. Surgery was then considered imperative.

The right hilus, right bronchus, and lower segment of the trachea were adequately exposed through a right thoracotomy. The main pulmonary artery coursed directly toward the right side and divided into its 2 main branches over the hilum of the lung. While the right pulmonary artery went directly to the right lung along normal pathways, the left pulmonary artery circled anteriorly around the right bronchus and lower part of the trachea and then coursed posteriorly and to the left, behind the trachea and in front

Fig. 1 Left. Case 1. Chest film in the frontal projection showing increased density of left upper lung field (lateral projection, which showed a distinct narrowing of the lower portion of the trachea did not lend itself to satisfactory reproduction).

Fig. 2 Middle. Case 1. Barium swallow showing extrinsic pressure defect on the anterior wall of the esophagus at about the level of the carina. Note also the congenital deformity of the upper dorsal spine (hemivertebrae).

Fig. 3 Right. Case 1. Posterior view of the mediastinal structures with probe at site of bifurcation of main pulmonary artery into left pulmonary artery and right pulmonary artery with its apical branch.
of the esophagus and aorta. The right bronchus was reported to be paper thin and transparent at the point of pressure by the artery. The left pulmonary artery was divided at its origin. The distal segment was then moved medial to the right bronchus and trachea and an end-to-end anastomosis was hurriedly performed with the proximal end.

The postoperative course was uneventful with the exception of a tracheobronchitis of short duration. Respiratory symptoms and obstructive emphysema of the right lung disappeared. An x-ray (fig. 6) on the eleventh postoperative day showed equal expansion of the lungs and the heart in normal position. During the ensuing months, the patient suffered occasional attacks of mild stridor, which later disappeared.

Case 3. J. E. S., the fourth child of a healthy mother was admitted to Children’s Memorial Hospital at 15 hours of age because of imperforate anus, cleft lip and palate, and possible congenital heart disease. The infant had been delivered without difficulty and had required no oxygen. Physical examination on admission revealed a well nourished 6-pound white male in good general condition. There was an imperforate anus and a rather severe cleft lip and palate. There was no cyanosis at rest and no cardiac murmur was heard. The heart tones were normal and the femoral arteries were palpable.

X-ray of the chest revealed normal heart and lungs. The hemoglobin was 20.5 Gm. per cent. the white blood cell count was 32,000/mm$^3$, and the urine was normal. The electrocardiogram suggested right heart strain.

At 48 hours of age a primary anorectal anastomosis was done under general anesthesia. The immediate postoperative course was uneventful. About 48 hours after surgery the child had a sudden severe episode of dyspnea, accompanied by gray pallor and cyanosis. The respirations became erratic and the infant died shortly thereafter.

Pertinent postmortem findings were imperforate anus, surgically repaired; cleft lip and palate; persistent left superior vena cava draining into the coronary sinus; common ventricle and atrio-ventricular orifice with 5 valves; dextroposed aorta arising slightly anterior to the pulmonary artery; pulmonary artery originating to the left of the aorta. At a distance of 0.5 cm. from the valve ring the pulmonary artery curved sharply to the right, crossing the midline. As it passed over the right bronchus it divided, one branch going directly to the right lung. The other branch coursed around the trachea near its bifurcation, passed anterior to the esophagus and parallel to the left main bronchus into the left chest and entered the parenchyma of the left lung after dividing into 3 branches.

DISCUSSION

The soft nature of the tracheobronchial cartilages in infancy explains the ease with which extrinsic structures can compress the major airways. Cardiovascular anomalies occupy a prominent place in the etiology of tracheobronchial compression. Although the markedly enlarged heart can produce com-
pression of the bronchial tree, vascular anomalies are more important in the genesis of respiratory tract obstruction. In 1837 Von Siebold first reported a case in which death due to respiratory distress was caused by a double aortic arch. Thenceforth, several similar cases have been reported and much stress has been laid on the common syndrome produced by double aortic arch and constricting ring formed by a right aortic arch with persistent ligamentum arteriosum or anomalous left subclavian artery. The successful surgical treatment of double aortic arch, first reported by Gross in 1945, stimulated new interest in the study of these conditions. Despite the wealth of literature accumulated on this subject, only very rare references are found pertaining to pulmonary artery anomalies, as a cause of respiratory obstruction.

The 3 cases herein reported are strikingly similar and represent instances of a "vascular sling" formed by an aberrant left pulmonary artery that encircled the right main stem bronchus and the trachea. As a consequence the right bronchus and posterior wall of the trachea were compressed, causing respiratory embarrassment. A thorough search of the literature revealed 5 similar cases. The clinical features of this anomaly consist of symptoms and signs of tracheobronchial obstruction, occurring shortly after birth. Although other malformations (such as double aortic arch and related developmental anomalies of the fourth aortic arch, tracheomalacia, tracheal stenosis, tracheal web, tracheo-esophageal fistula, etc.) can result in similar respiratory derangement, the radiologic and endoscopic findings are sufficiently distinctive in most of the cases to permit a correct diagnosis. While bronchoscopic examination indicates the presence of extrinsic compression on the right bronchus and lower trachea, the esophogram does not show a posterior indentation and thus excludes a true vascular ring. Compression of the anterior wall of the esophagus, as noted in case 1 and in the first case reported by Wittenborg et al., further suggests that the cause of obstruction must lie anterior to the

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*In reference to this type of anomaly, the term "vascular sling" rather than "vascular ring" should be adopted to avoid confusion with more common systemic arterial malformations causing constriction of the trachea and esophagus.
esophagus. Obstructive emphysema of the right lung, as noted in case 2 and in the case reported by Morse and Gladding,7 lends additional support to the diagnosis of this malformation.

The embryologic origin of such a vascular anomaly is of some interest. Early in embryonal life (4 mm. to 40 mm. of embryonic length) each pulmonary artery, forming from the primitive pulmonary arteries and elements of the sixth arch, seeks a vascular channel in the sixth arch, accompanying the newly sprouted lung bud (fig. 7A). Normally, the left pulmonary artery finds its connection in the hilar area of the left lung bud and, as the bronchial tree progresses in its development, the usual relationship of the left pulmonary artery to the left bronchus results. If, however, the timing is upset during the formation of these structures, an anomaly may result. For example, if the growth of the left pulmonary artery is delayed or that of the primitive left lung bud is accelerated, they cannot meet normally. The normal path of the left pulmonary artery to the left lung bud would be obstructed, so to speak, by the overgrown left primitive bronchus. In order to establish its mesenchymal connection, the primitive left pulmonary artery now would have to pass anterior to the right bronchus, posterior to the trachea, then to the left into the tissue surrounding the left lung bud (fig. 7B). Similar embryologic speculation on the origin of this anomaly is offered by Scheid.5

It appears that major developmental anomalies are often associated with the "vascular sling" formed by an aberrant left pulmonary artery. They may dominate the clinical picture. However, the occurrence of this lesion as an isolated anomaly, which is surgically correctible, seems to be almost as frequent (our case 2, Morse's case,7 and Welsh's case6). Therefore the clinical recognition of this condition is not only of academic interest but also of practical concern.

Whenever suspicion arises of an anomalous left pulmonary artery, thoracotomy should be performed on the right side.

CONTRO, MILLER, WHITE, AND POTTS

Summary

Three patients are reported in whom the left pulmonary artery coursed anomalously anterior to the right bronchus and behind the trachea with consequent compression of these structures. A review of the literature revealed 5 similar case reports. The clinical picture associated with this congenital lesion is dominated by symptoms and findings of bronchial obstruction, when not obscured by other major associated anomalies. Bronchoscopic findings indicative of extrinsic pressure and the absence of indentation of the posterior wall of the esophagus assist in the delineation of this lesion. Because this anomaly is surgically correctible, importance of its clinical recognition is stressed.

SUMMARIO IN INTERLINGUA

Es reportate le casos de tres patientes in qui le arteria sinistro-pulmonar sequeva un anormal curso anterior al broncho dextere e in retro del trachea, con le effetto de compression del mentionate structuras. Un revista del litteratura revelava reportos de 5 simile casos. Le tableau clinic associate con iste congenite lesion es dominate per symptomas e constatationes de obstruction bronchial, providite que illos non es cedate per altere major anomalias associate. Constatazione bronchoscopiche que indica le presentia de un pression extrinsee e le absentia de indentation del pariete posterior del esophago es de adjuta in le delineation de iste lesion. Proque le anomalia pote esser corrigithe per medios chirurgic, le importantia de su recognition clinic es sublineate.

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STEPHEN CONTRO, ROBERT A. MILLER, HARVEY WHITE and WILLIS J. POTTS

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