Aberrant Left Pulmonary Artery

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An aberrant left pulmonary artery, a surgically correctable anomaly, is often not thought of in the differential diagnosis of respiratory obstruction in infancy. We have discovered 35 previously reported cases (Lochard, Vert, and Chalnot, 1963; Pontius, 1963; Heinemann, 1964; Neumann et al., 1964; Murphy et al., 1964; Hu Chih-Hsiang, 1964). We report two further cases—one patient diagnosed during life and operated upon, but who died post-operatively, and the other recognized at necropsy.

CASE REPORTS

Case 1. A male Chinese was delivered normally at the Kandang Kerbau Maternity Hospital, Singapore. His birthweight was 3258 g. (7 lb. 4 oz.) and his condition at birth was satisfactory. He was well until 7 weeks old when he was admitted to the paediatric ward with a history of persistent coughs and noisy respiration during the previous 10 days, which had become progressively worse. He had had several cyanotic spells due to choking produced by excessive frothy, mucoid sputum in the throat.

On clinical examination, the child was in severe respiratory distress. He had a loud inspiratory stridor with marked chest recession and slight cyanosis. There was no clubbing of fingers. The pulse rate was 140 a minute, the temperature 37.5°C. (99-5°F.). The throat was injected but no membranes were seen. The heart was clinically not enlarged and the heart sounds were normal. The blood pressure was 80/50 mm. Hg. All peripheral pulses were present and equal. Scattered rhonchi and rales were heard in both lungs. The liver and spleen were not palpable. Other systems were clinically normal. Laboratory findings: Hb 10.1 g./ml., total white cells 19,000, differential count: polymorphs 80, lymphocytes 14, monocytes 3, and eosinophils 3. A throat swab was negative for C. diphtheriae. Chest x-ray film showed a heart of normal size and shape and evidence of bronchitis in the lungs. Lateral x-ray film of the neck showed no obstruction of the upper respiratory tract. An ear, nose, and throat surgeon could not find any abnormality in the larynx. A diagnosis of acute tracheobronchitis was made and the child was treated with chloramphenicol, steroids (for bronchospasms), and oxygen, with improvement in the general condition. The stridor gradually disappeared and the respiratory distress was relieved. The child was then discharged home after 12 days in hospital.

Two weeks later, he was readmitted with a similar history of stridor and cyanotic spells. The clinical findings were the same as in the previous admission. There was no history of dysphagia at any time. Bronchoscopy did not reveal any abnormality. An oesophagogram showed a deep anterior indentation of the oesophagus at about the level of the carina (Fig. 1 and 2). The child's condition deteriorated: he became more and more distressed from respiratory obstruction, had repeated cyanotic attacks, and was referred to one of the authors (N.K.Y.) and operated upon.

A left antero-lateral thoracotomy was done. On dissection of the superior mediastinum, a patent ductus arteriosus was found which was sutured and excised. The aberrant left pulmonary artery, running between the trachea anteriorly and the oesophagus posteriorly and compressing the trachea about 0.5 cm. above the bifurcation, was identified. It was then divided close to its origin and the distal end was brought forwards and anastomosed end-to-side to the intrapericardial portion of the main pulmonary artery (Fig. 3). The vagus, the recurrent laryngeal, and the phrenic nerve were all identified and preserved. The aortic arch and its branches were normal. There was a small left persistent superior vena cava draining into the coronary sinus. The anastomosis was satisfactory and the blood supply to the left lung flowed normally.

The child's immediate post-operative condition was satisfactory, but three hours later he suddenly developed cardiac arrest. Resuscitative measures, including an external cardiac massage and intermittent positive pressure respiration, were instituted. The heart started to beat again and the child was put on a respirator. However, his general condition gradually deteriorated and he died 12 hours after the operation.

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FIG. 1.—Oesophagogram (antero-posterior view) shows characteristic anterior indentation at about the level of the bifurcation of the trachea (arrowed).

FIG. 2.—Oesophagogram (lateral view) shows the anterior indentation of the oesophagus (arrowed).

FIG. 3.—Diagrammatic representation of operation (Case 1). The distal portion of the aberrant left pulmonary artery (LPA) is anastomosed end-to-side to the main pulmonary artery (PA).

RL, right lung; LL, left lung; RPA, right pulmonary artery; A, aorta; RV, right ventricle; LV, left ventricle; T, trachea.

**Necropsy.** The findings at thoracotomy were confirmed. The end-to-side anastomosis of the aberrant left pulmonary artery to the main pulmonary artery was intact. The whole length of the trachea and major bronchi was congested and narrowed, especially at a point about 0.5 cm. above the carina. An anomalous bronchus, supplying the upper lobe of the right lung, was found about 1 cm. from the bifurcation. Other systems were grossly normal.

**Case 2.** A 2494 g. (5 lb. 8 oz.) full-term, male Chinese baby was delivered normally. The baby was feeble and blue at birth. Crepitations were heard in both lungs. A provisional diagnosis of aspiration pneumonia was made, the baby’s mouth and pharynx were cleared by suction, and oxygen by hood was administered with good results. However, it was noticed that the baby began to turn blue the moment he was taken off the oxygen.

Two hours after birth, he was seen by a paediatrician and was found to have some difficulty in breathing. Auscultation of the lungs revealed an obstruction to air entry, with decreased breath sounds at the beginning of every inspiratory phase, followed by crepitations all over
both lung fields. All other systems were clinically normal.

The following day, the baby was noticed to be pink when at rest, but when stimulated to cry, developed respiratory distress and turned blue. Before any investigations could be carried out to determine the cause of the respiratory obstruction, the baby died, about 34 hours after birth.

**Necropsy.** The main pulmonary artery was found to arise normally from the right ventricle and to run upwards and to the right. Just below and to the right of the bifurcation, it branched into the right and left pulmonary arteries. The former entered the right lung normally; the latter, however, coursed above the right main bronchus, close to its origin, and then ran behind the trachea, just above the bifurcation, and anterior to the oesophagus to enter the left lung normally (Fig. 4 and 5). At the point where the aberrant left pulmonary artery was pressing on it, the trachea was narrowed.

The heart showed a high ventricular septal defect. The foramen ovale was anatomically patent. The valves and coronary blood supply were normal. The ductus arteriosus was absent. The aorta and the great vessels were normal. The liver appeared grossly normal. The gall-bladder was absent and the extrahepatic bile-ducts were atretic. All other systems were grossly normal.

**Discussion**

The first case of an aberrant left pulmonary artery was described by Glaevecke and Doehle in 1897. Scheid in 1938 reported the second case. Since then, more cases have been described.

In 1959, Sherman recognized the aberrant left pulmonary artery as a distinct anatomical anomaly with the following characteristics.

1. An elongated pulmonary trunk arises normally from the right ventricle and runs upwards and to the right of the midline following the course of the right pulmonary artery.

2. The left pulmonary artery arose extrapericardially from this trunk just in front and below the origin of the right main bronchus. It courses anterior and superior to the right main bronchus and then runs between the trachea anteriorly and the oesophagus posteriorly.

3. The aberrant left pulmonary artery is in contact with these air passages along its course and the point of maximum contact and compression is variable.
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(4) It invariably compresses the oesophagus anteriorly.

In most of the 35 previously reported cases, the right main bronchus bore the brunt of the compressing force. In some, as in both our cases, the trachea was compressed just above the bifurcation. In a small number of cases, the compression involved both the right main bronchus and the trachea. At operation or at necropsy, the point of compression of either the trachea or the right main bronchus can be recognized as a stenotic segment with the aberrant left pulmonary artery overlying it.

In a number of the previously reported cases, no mention of the race was made. However, gauging from the countries these reports came from, it could be assumed that they were all Caucasians. If this was so, then only 3 of the 35 cases were non-Caucasians—two being Negroes (Sherman, 1959; Jacobson et al., 1960) and the other Chinese (Hu Chih-Hsiaung, 1964). Both our cases were Chinese.

Both sexes may be affected. Of the 35 previously reported cases, 19 were male and 14 were female. In two cases, no mention of the sex was made. Both our cases were male.

Clinically, an aberrant left pulmonary artery presents as respiratory distress. The patient is usually dyspnoic and cyanosed with marked chest recession. Sometimes there may be a stridor or a wheeze. The expiratory phase is often prolonged. These signs and symptoms are usually present at birth. Sometimes, however, they appear in the neonatal period and only rarely in the paediatric age-group. Antemortem diagnosis is possible if only the anomaly is thought of. It can be confirmed by the following.

1. An oesophagogram which shows an anterior indentation of the oesophagus at about the level of the carina. Hiller and Maclean (1957) consider this fairly diagnostic and claim that the failure of diagnosis is due to not screening and filming the patient in the full lateral position. Our first case was diagnosed by an oesophagogram.

2. A bronchoscopy which shows a compressed trachea or right main bronchus which is easily distended by the bronchoscope, but which collapses upon withdrawal of the instrument.

3. A chest x-ray film which will show an obstructive emphysema of the right lung and a left mediastinal shift, if the right main bronchus alone is compressed.

4. A pulmonary angiogram which demonstrates the presence of the anomalous vessel.

Potts, Holinger, and Rosenblum (1954) were the first to perform a successful surgical correction of the anomaly. They divided the aberrant left pulmonary artery and re-anastomosed it in front of the trachea. Most of the other successful operations were based upon the technique of Potts et al. Hiller and Maclean (1957) ligated the aberrant left pulmonary artery at its origin and anastomosed its distal end to the broadside of the main pulmonary artery. They claimed this avoided the tendency to kink at the bifurcation and allowed a larger area of vascular anastomosis. This technique was employed in our first case. Lochard et al. (1963), however, found that in their case, because of the lower position of the aberrant left pulmonary artery in relation to the right main bronchus and trachea, it was only necessary to section the former and to bring forward the anomalous vessel. The right bronchus was then re-anastomosed.

An aberrant left pulmonary artery is usually associated with other congenital anomalies, especially with some other vascular anomalies and/or a congenital heart (Wittenborg, Tantiwongse, and Rosenberg, 1956; Contro et al., 1958; Schutt and Robb, 1959; Jacobson et al., 1960; Niwayama, 1960; Hu Chih-Hsiaung, 1964). Other coexistent anomalies that have been described are complete tracheal rings (Scheid, 1938; Jacobson et al., 1960), stenotic trachea, single-lobed left lung, two-lobed right lung and absence of the left lobe and isthmus of the thyroid (Niwayama, 1960); cleft lip and palate, and imperforate anus (Contro et al., 1958); hypoplasia of the left kidney and ureter, malrotation of the intestines, and fusion of the third and fourth lumbar vertebrae (Wittenborg et al., 1956); diaphragmatic hernia (Heinemann, 1964); Hirschsprung's disease (Murphy et al., 1964); and absence of the gall-bladder (Jacobson et al., 1960). The latter was seen in our second case. Anomalous double right bronchi in our first case and absence of the ductus arteriosus and atresia of the extrahepatic bile-ducts had not been previously described.

In three of the reported cases, the patients were mongols (Wittenborg et al., 1956; Schutt and Robb, 1959; Le Tan-Vinh, Alagille, and Nguyen van Phuoc, 1961), one of whom had a normal twin sib (Schutt and Robb, 1959).

The embryology of the aberrant left pulmonary artery is not quite clear. Scheid (1938) and Contro et al. (1958) suggested that the anomalous vessel was the normal vessel displaced by a timing upset in its formation. During embryonic development, each pulmonary artery, forming from the primitive pulmonary arteries and the sixth arch, joins a vascular channel in the hilar region of the developing lung. As the bronchial tree progresses in its development, the usual relationship of the pulmonary artery to the bronchus results. If, however,
the growth of the left pulmonary artery is delayed or that of the left lung is accelerated, they cannot meet as usual. The left pulmonary artery will then have to take an anomalous course. Quist-Hanssen (1949) felt that the ductus arteriosus present in most cases prevented this displacement of the left pulmonary artery. He therefore postulated that a normal pulmonary artery did not develop and that the aberrant vessel arose from collateral vessels which had developed from the foetal mediastinal plexus connecting the two lung hila.

SUMMARY

Two cases of an aberrant left pulmonary artery in Chinese infants are described. The first patient was diagnosed by an oesophagogram and operated upon, but developed post-operative cardiac arrest and died. The second case was diagnosed at necropsy. The clinical and pathological presentation of an aberrant left pulmonary artery is reviewed. The embryology of the anomalous vessel is discussed.

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