

Pulmonary artery sling

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ABSTRACT Pulmonary artery sling is a rare cause of respiratory distress the embryological origin of which is not known. Two patients with this abnormality, both of whom are now thriving, are described. Surgery was necessary at the age of 5½ years in one child, who also had an atrial septal defect. The other child, at the age of 2 years, shows improvement of her wheezing, cough, and breathlessness despite having had neither surgical nor medical treatment.

Pulmonary artery sling was first described by Glaevecke and Doeble in 1897.¹ Since then less than 80 other cases have been recorded in published reports.² The first surgical correction was performed by Potts in 1954.³

The aberrant vessel arises from the right pulmonary artery and passes between the trachea and oesophagus to enter low in the left hilum. Thus a sling is formed around the right main bronchus and trachea. The embryological origin is not fully understood.

The treatment of choice for the pulmonary sling is surgery; but both medical and surgical treatment have a high mortality rate. In only one of the two cases we report was surgery necessary, and the other patient improved without any treatment.

Case reports

CASE 1

An 18 month old girl was referred to the outpatient clinic. She was a full term baby with a normal delivery whose birth weight was on the 12th centile. She had been noted to be breathless and wheezy from birth, with a cough that was occasionally productive. The wheezing had been worse over the previous six months and peripheral cyanosis was noticed on two occasions. She had always fed well and gained weight satisfactorily. She had had no syncope or fits. On examination she appeared a healthy child, not cyanosed and with no clubbing; her height was on the 10th centile and her weight on the 50th centile. She had no intercostal indrawing but had both

inspiratory and expiratory rhonchi throughout the chest. Her heart sounds were normal but a systolic bruit was audible in the right axilla. A chest radiograph showed mediastinal shift to the right and was reported as showing agenesis of the right upper lobe; a barium swallow showed an anterior indentation in the middle third of the oesophagus (fig 1).

An initial diagnosis of pulmonary artery sling was made and the child was admitted for cardiac catheterisation. This showed an aberrant left pulmonary artery arising from the right pulmonary artery and forming a sling around the trachea (fig 1).

The child has been seen regularly in the outpatient clinic over the last six months and she continues to thrive with her weight on the same centile; her wheezing, cough, and breathlessness are improving. She is having no medication.

CASE 2

A Pakistani girl, born at full term by a normal delivery, was referred at 8 months. She had a history of two respiratory tract infections, the first at 4 months of age. Both required hospital admission. A chest radiograph taken at the time showed dextrocardia (fig 2).

On examination she was not cyanosed. She had a grade 3/6 systolic murmur audible over her back and a grade 2/6 pulmonary systolic murmur. She was not in heart failure. Cardiac catheterisation showed that she had an atrial septal defect and that the dextrocardia was positional and due to a hypoplastic right lung. She was observed over the next five years. She gradually became short of breath, with some wheezing, and she was reinvestigated at the age of 5½ years. The physical signs suggested a persistent atrial septal defect and the chest radiograph showed an oligoemic right lung field and a plethoric left lung. Cardiac catheterisation on this occasion

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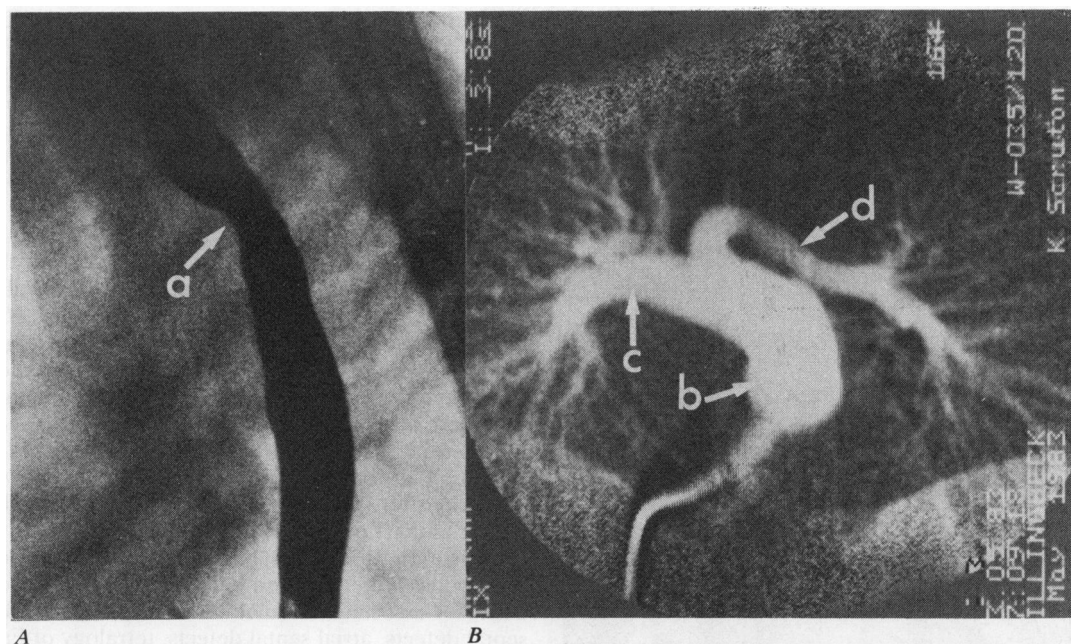


Fig 1 Case 1: A—Barium swallow showing the indentation in the anterior wall of the oesophagus at the level of the carina caused by the aberrant pulmonary artery (a). B—Pulmonary angiogram showing b—main pulmonary artery; c—right pulmonary artery; d—aberrant left pulmonary artery arising from the right.

showed the atrial septal defect with a 2:1 left to right shunt and an abnormal pulmonary vascular supply (fig 2). The small right pulmonary artery gave rise to a vessel which passed posterior to the trachea and anterior to the oesophagus to reach the left lung. This lung was also supplied by an artery following the normal course. The right lung was hypoplastic. The child was admitted for closure of the atrial septal defect and correction of the pulmonary vascular anatomy.

At operation the pulmonary artery anatomy was identified before bypass was started. The main pulmonary artery divided into two branches, one to the left lower lobe and the other to the right lung. The right pulmonary artery was hypoplastic and gave rise to the aberrant vessel that passed between the trachea and oesophagus to supply only the left upper lobe.

Bypass was started and the ostium secundum defect was closed with interrupted Ethibond sutures. The pulmonary artery was dissected out and a sling placed around the left main bronchus. The aberrant pulmonary artery was divided from the right pulmonary artery and the proximal end was oversewn with continuous 5/0 prolene. The aberrant vessel was pulled from behind the left main bronchus and anastomosed end to side to the main pulmonary artery with 5/0 prolene. Air was removed from the heart, which resumed spontaneous rhythm. Decannulation

was effected without incident. The child was extubated in theatre and returned to the intensive care unit with 50% oxygen administered via a face mask. She returned to the ward the following day and made an uneventful recovery.

The child has been followed up in the outpatient clinic at six monthly intervals for three years. She is thriving and is on the 12th centile for weight and height. She has not suffered any lower respiratory tract infections.

Discussion

Infants with pulmonary artery sling usually present in the first year of life. The average age at onset of symptoms is 2 months and about half have symptoms from birth,⁴ as in our first patient. The anomaly, if not corrected, can be fatal within the first year of life.⁴⁻⁶ Less than 10 cases have been diagnosed in adult life, the anomaly in these cases having been an incidental finding during investigations for unrelated problems.⁵

Patients with pulmonary artery sling present with respiratory symptoms. Respiratory distress and intercostal recession are the commonest features, occurring in 90% of reported cases.⁴ Other symptoms and signs include cyanosis, stridor, inspiratory and expiratory rhonchi, apnoeic episodes, asymmetry of the chest, and difficulty in feeding.^{4,7}

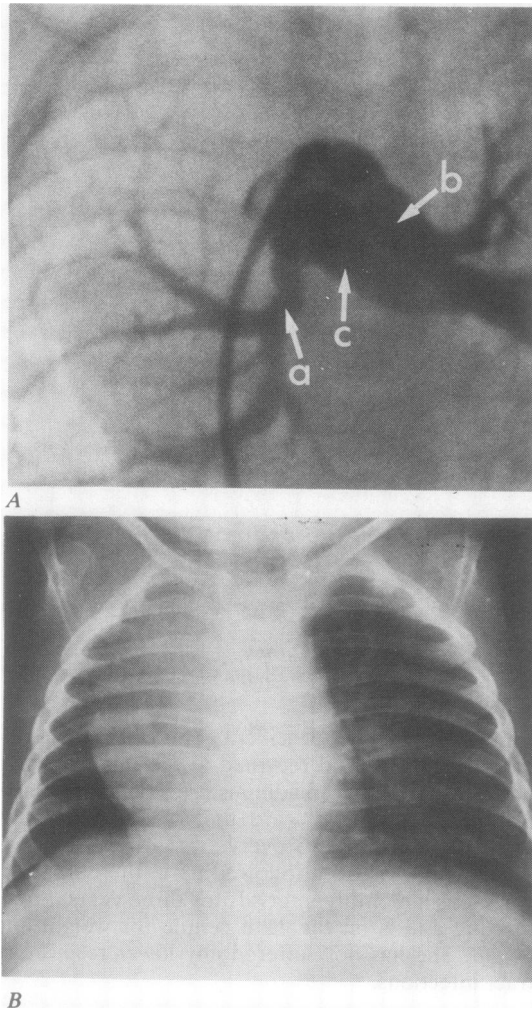


Fig 2 Case 2: A—Pulmonary angiogram showing a—right pulmonary artery; b—aberrant left pulmonary artery supplying the left upper lobe; c—left pulmonary artery arising from the main pulmonary trunk. B—Chest radiograph showing positional dextrocardia and hypoplastic right lung.

Neither of our patients showed intercostal recession as a sign of obstruction, although they were breathless and had coughing and wheezing. A soft systolic murmur at the upper left sternal edge has been described when there is no other congenital cardiovascular abnormality.⁴

The diagnosis of pulmonary vascular sling rests on the investigations. Plain chest radiography may show abnormalities of aeration of the right lung, such as obstructive emphysema, or, if the bronchus is completely obstructed, atelectasis. The carina may be positioned abnormally low with a wide angle

between the right and left main bronchi.⁸ The left hilum is low.^{7,9} A mediastinal mass may be evident at the level of the carina and seen between the trachea and oesophagus on lateral views.⁹ Both our children had abnormal lung fields. Patient 1 had agenesis of the right upper lobe, while patient 2 had such a greatly reduced right lung field that there was positional dextrocardia (fig 2).

A barium swallow shows there is indentation of the oesophagus anteriorly, at the level of the carina^{2,4,9} (fig 1). This sign was anticipated by Welsh and Munro in 1954,¹⁰ and first demonstrated by Wittenberg *et al* in 1956.¹¹ The pulmonary artery sling is the only vascular anomaly which causes indentation of the anterior oesophagus. Dysphagia and regurgitation are rarely a problem, although they have been reported in adults.⁵

Computed tomography has recently been shown to demonstrate clearly the anomalous origin of the left pulmonary artery^{2,5} and is a useful non-invasive investigation. It does not, however, obviate the need for cardiac catheterisation because of the high incidence of associated cardiac anomalies. Ventricular septal defects, atrial septal defects, tetralogy of Fallot, and coarctation of the aorta have all been reported in association with the aberrant left pulmonary artery.^{2,5} To demonstrate the aberrant vessel by cardiac catheterisation the contrast must be injected when the catheter is in the pulmonary tree because other abnormalities may obscure the left pulmonary vascular system, as occurred in the first cardiac catheter study in case 2. Congenital cardiac abnormalities may occur in as many as half the cases of pulmonary artery sling.¹³

There has been only one other published case with both an aberrant and a normal left pulmonary artery.¹³ In this case the anomalous vessel arose from the right pulmonary artery and passed between the oesophagus and trachea, supplying only the posterior part of the left lung. The other vessel arose from the main pulmonary trunk.

Abnormalities of the tracheobronchial tree are common—for example, a tracheal origin of the right upper lobe bronchus and, of more significance, tracheal and bronchial stenosis. The stenosis is due to complete cartilaginous rings and absence of the pars membranacea. Long segments of the tracheobronchial tree may be affected. About half of the infants with pulmonary artery sling have abnormal bronchial cartilage and this may cause persistent respiratory distress despite surgical relief of the sling.^{2,6,7,12,14} Other congenital abnormalities, affecting the gastrointestinal tract and genitourinary and endocrine systems, have also been reported.¹²

The explanation for this abnormality in embryological terms has not emerged. Although

various theories have been suggested none can be substantiated.^{4,6,8} Our two cases support the idea that the aberrant left pulmonary artery is originally a branch of the right pulmonary artery, which migrates during development.

Potts in 1954 was the first to perform an operation to correct the pulmonary artery sling.³ The left pulmonary artery is divided between clamps behind the trachea and the distal segment passed in front and reanastomosed anterior to the trachea by an end to side anastomosis to the main pulmonary artery. Various approaches to the pulmonary arteries have been attempted—for example, unilateral or bilateral thoracotomy or, if there is another cardiac anomaly to correct, median sternotomy. Particular consideration should be given to the possibility that vessels arise from the anomalous pulmonary artery and supply the right lung. Interrupted sutures are used for the vascular anastomosis in these small children to allow some growth.¹³

Other operations described include division of the right main bronchus to release the sling and its reanastomosis.⁴ The ductus arteriosus or ligamentum arteriosum may be divided, which allows the pulmonary artery to move to the right and so release the sling.^{4,10} Of 26 patients, reviewed by Grover *et al.*,⁴ who had the pulmonary artery divided and reanastomosed, only 19 survived the operation. The intraoperative and postoperative morbidity and mortality are greatly increased by the use of a right rather than a left thoracotomy.⁷ The major cause of morbidity is cardiac arrhythmia. Patient 2 had a median sternotomy as the atrial septal defect was closed at the time of operation, and this approach provided excellent exposure. The prognosis is adversely affected by associated abnormalities, particularly those of the tracheobronchial cartilage. Where the cartilaginous rings are complete respiratory distress persists in the postoperative period.^{2,4,13}

Sade⁹ reviewed 65 patients with pulmonary artery sling. There were 17 who survived operation; 10 of these had follow up catheterisation and only one showed a patent left pulmonary artery. A collateral

circulation for the left lung does develop but does not prevent the development of pulmonary hypertension. Pulmonary artery sling has high morbidity and mortality rates whether treated conservatively or by operation.

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