

Surgical Correction of Right Pulmonary Vein Opening into the Azygos Vein Combined with Atrial Septal Defect

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There are two forms of congenital transposition of the pulmonary vein. All direct communication with the heart is lacking in cases of total transposition, whereas — in partial transposition — only some of the pulmonary veins drain into the right side of the heart either directly or via the systemic veins (V. cava superior, innominate vein, coronary sinus, V. cava inferior, V. cava superior sinistra persistens, left subclavian, portal, gastric and azygos veins, etc.). The anomaly is usually associated with a defect of the atrial septum.

To our knowledge, no more than 11 cases have so far been reported in which the pulmonary vein drained into the azygos vein.

In the 3rd or 4th week of intrauterine life, i.e. at the time of growth of the lung bud, there appears a venous plexus (*plexus intestinus primus*) which is soon communicating with the umbilical veins via the cardinal and the vitello-umbilical veins. Endothelial proliferation from the sinoatrial region of the embryonic heart starts at the same time; it unites with the

said primary pulmonary venous plexus to form the common pulmonary vein. Situated in the midline, this new vessel bifurcates in the course of development, and conveys blood from the (meantime likewise bifurcated) lung bud to the left atrium. After repeated forkings, this vein subsequently invades the lungs, while the left atrium expands. First the trunk of the common pulmonary vein, then its primary and secondary branches are incorporated by the wall of the (meanwhile formed) left atrium so that, finally, blood flows into the left atrium by four permanent pulmonary veins. This process of development goes hand in hand with a degeneration or disappearance of the cardinal and vitello-umbilical veins.

Total or partial transposition is the consequence of an abnormal development of the common pulmonary vein. If, in cases of total transposition, the blood passes through the cardinal and vitello-umbilical veins, these adapt themselves to the increased flow by dilatation. If it is in the left pulmonary trunk that the developmental

anomaly occurs, the blood will flow to the left cardinal vein (via the future V. cava superior persistens sinistra into the coronary sinus and/or through the innominate vein into the V. cava superior and the right atrium). Under suitable conditions, the pulmonary veins of the right side may, in a like manner, open into the right cardinal vein (the V. cava superior).

It also happens that the common pulmonary vein is disturbed in its development while the vitello-umbilical veins fail to degenerate. The pulmonary veins may in such cases drain into the heart (directly into the right atrium, the V. cava inferior, the portal vein, the gastric, the hepatic or the azygos vein) through the preserved vitello-umbilical vessels.

Malformation of the atrial septum, too, may give rise to transposition of the pulmonary veins. Physiologically, the septum primum is formed on the right side of the common pulmonary vein, so that the pulmonary veins issuing from the primary trunk empty into the left atrium. If, however, the atrial septum turns left in the course of development, the orifice of the common pulmonary vein will open into the right atrium.

REPORT OF A CASE

V. D., a female child 11 years of age, was admitted in March, 1964. The patient had been known to have heart disease since the age of 4; at the age of 7 she had had a suspicious X-ray shadow in the thorax. She had frequently had upper respiratory catarrhs.

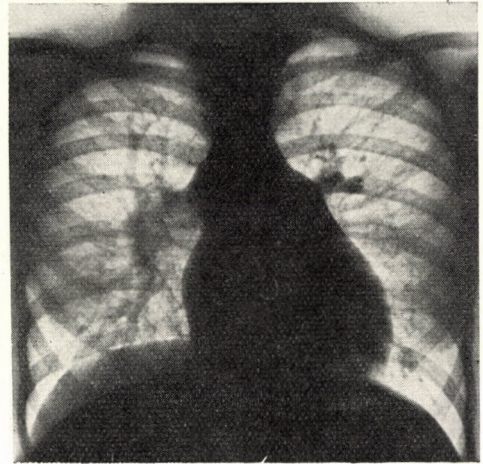


FIG. 1. Preoperative antero-posterior roentgenogram

At admission the child was moderately developed, lean. There was an ejection murmur filling one half to two thirds of the systole, with the punctum maximum in the left parasternal line at the height of the second intercostal space; the second sound was widely reduplicated. Liver and spleen were not palpable. Blood pressure was 120/90 mm Hg; pulse rate, 100/min. Laboratory tests were negative. Thoracic X-rays showed clear perihilar fields, hypervascularization and dilated central vessels in the hilum. The mediastinal shadow was widened at the height of the right hilum whence a shadow, suggestive of an aberrant pulmonary vein, ran upward. The left side of the heart extended to the medio-clavicular line. The right ventricle was considerably dilated, the left was displaced, the retrocardial space free, the aorta normal. The oesophagus showed an impression made by the pulmonary artery (Fig. 1). ECG: limb leads showed a deviation of the electric axis to the right; thoracic leads revealed an incomplete right bundle branch block, right ventricular hypertrophy and tall spiked P-waves.

Cardiac catheterization gave the following results.

Pressure (mm Hg)		Blood oxygen, per cent
V. cava superior	—	68
V. cava superior at opening into right atrium	—	95
V. cava inferior	—	78
Right atrium	8/3	91 86.5, 89, 83
Right ventricle	40/2	85, 84
Pulmonary artery, trunk	40/12	83.5
right branch	30/10	85
left branch	40/12	85
Pulmonary capillary	18/10	—
Left pulmonary vein, wedge overall	18/10 18/6	94 90
Left atrium	8/2	91.5
Pulmonary vein draining into V. cava superior	—	93.5

The catheter passed from the right to the left atrium and the left pulmonary vein. The atrial defect was at the lower part of the septum. There was a free passage from the V. cava superior to the right pulmonary vein.

The patient was operated upon on June 16, 1964, in hypothermia of 30°C. From a longitudinal median sternotomy the pericardium was opened along the right phrenic nerve. The right atrium and ventricle were considerably enlarged. The pulmonary artery was thicker than the aorta, and a systolic thrill was palpable above it. A 4 cm segment of the V. cava superior, before its opening into the right atrium, had a width of about 2.5 cm. The right pleural cavity was opened and the overlapping part of the pericardium was detached from the V. cava superior. The right lung was composed of two lobes separated by an about 2 cm thick and 3 to 5 cm wide septum which extended to the hilar area. Since it impeded orientation, it was divided along its whole length. The hilar structures

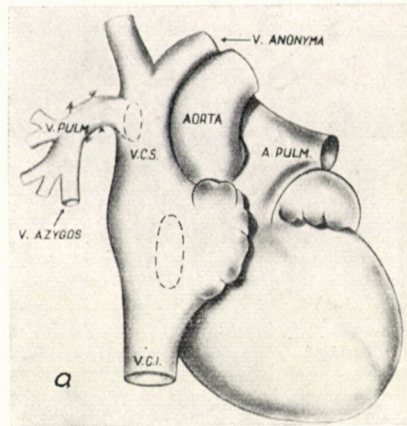


FIG. 2. Schematic illustration of the operation

FIG. 2a. Preoperative condition. Dotted line indicates septal defect and opening of the azygos vein. Side branches of the latter are ligated

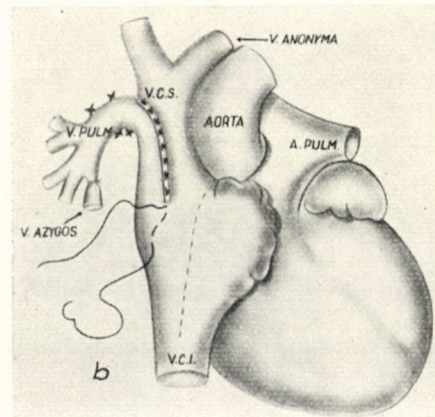


FIG. 2b. The V. cava superior is longitudinally divided by knot suture passed through a Teflon band. Half purse string suture is applied to the outer surface of the right atrium. The azygos vein is ligated

were dissected free of the neighbouring tissues. The veins of the upper and the lower lobe were well distinguishable. The two pulmonary veins opened into the azygos vein which had there a diameter of 2.2 cm. This vein was only 1 cm in dia-

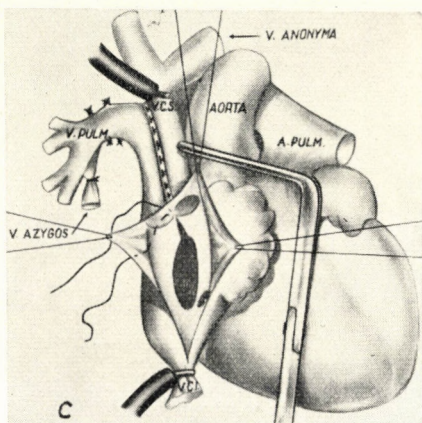


FIG. 2c. The V. cava superior and inferior, the aorta and pulmonary artery are clamped; the right atrium has been opened. The atrial septum is split open toward the V. cava superior

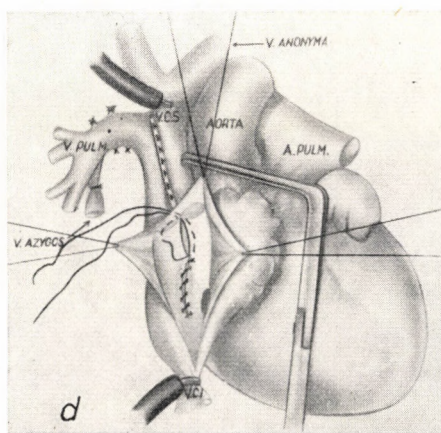


FIG. 2d. The original defect is closed by running suture. The lower, left and upper border of the septal defect are taken up by a purse string suture beginning at the outer surface of the atrium, and then passed again to the outer surface

meter along its normal course below the level of the hilum, and assumed the said width only at the orifice of the pulmonary veins. The azygos vein was ligated before the pulmonary orifices. The intercostal veins, 2 to 3 mm in diameter, opening

likewise into the azygos vein, were also ligated. The azygos vein and the superior V. cava were exposed. The opening of the azygos vein into the V. cava superior was about 4 cm before the latter's atrial orifice. The V. cava superior along its entire length was divided into two tubes by means of densely applied U-sutures. A teflon band, each, was placed above and under the V. cava superior, and the stitches were passed through these bands. In this way a separate channel was made for the blood coming from the pulmonary veins, and a separate one for the blood of the V. cava superior. The last stitch was made to form a half purse-string suture along the wall of the right atrium (Figs 2a–b). The venae cavae, the aorta and the pulmonary artery were clamped, and the right atrium was widely opened. A septal defect of the fossa ovalis type, measuring about 1.5 cm by 2.5 cm appeared at a great distance from the orifice of the channel made for transporting the blood of the pulmonary veins. Therefore the septum was split toward the V. cava superior (Fig. 2c). After closing the original septal defect with running suture, the purse-string suture was passed from the outer wall of the atrium into its cavity, embracing the lower left and upper

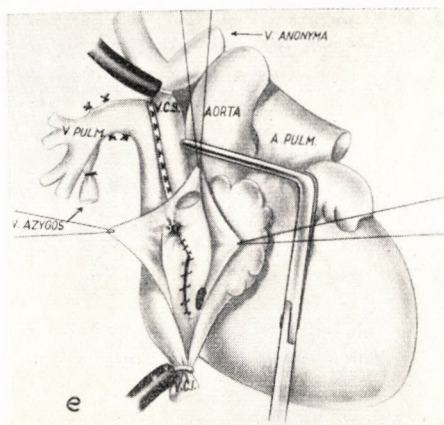


FIG. 2e. The septal defect is repaired, the blood flows from the pulmonary vein into the left atrium

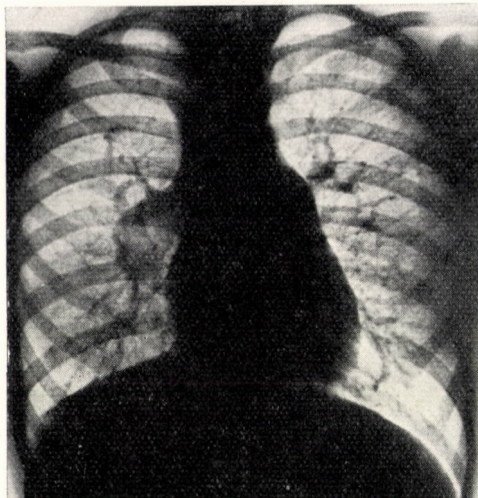


FIG. 3. Postoperative antero-posterior roentgenogram

borders of the split septum and passing the thread on the right side of the channel made for the blood of the V. cava superior to the outer surface of the atrium, to the point where the purse-string had started (Fig. 2d). On knotting the purse-string suture, the atrial septum was pulled to the wall of the atrium. The tube made for the blood of the pulmonary veins curved thus into the left atrium, while the blood of the V. cava superior was free to flow into the right atrium (Fig. 2e). The wound of the right atrium was closed and the clamps and ligatures were released. Tonicity of the heart seemed satisfactory, and its action was rhythmical. The pericardium was reunited, a drain was placed into the right half of the thorax, the sternum was united by wire strands and closed.

The suspension of circulation lasted $6\frac{1}{2}$ minutes.

While dissecting out the hilar structures, the thoracic duct may have been injured because a mixture of blood and chyle was withdrawn by thoracic puncture on the 5th postoperative day. After repeated aspirations of fluid, the chylothorax subsided without further intervention by the end of the 4th week.

At a follow-up in May, 1965, the child had gained 6 kg in weight and 13 cm in length. X-rays showed the right half of the chest to be clear, the pleural shadow to have been absorbed, the vascularization of the lungs to have decreased and the cardiac shadow to be diminished (Fig. 3).

DISCUSSION

A survey of the hitherto published 11 cases in which the pulmonary vein drained into the vena azygos reveals that in 4 cases one or two lobar veins [2, 5, 7, 11], in another 4 cases all veins of the right lung [12, (2 cases), 1, 9], in one case all veins of the left lung [6] and in 2 cases all veins of both lungs [3, 8] emptied into the azygos vein.

Cardiac and vascular anomalies such as atrial septal defect [1, 6, 8]; V. cava superior opening into the left atrium [8]; bilocular heart [3]; lack of V. cava inferior [7]; anomalous arteries [8, 9]; patent ductus arteriosus [8]; infundibular pulmonary stenosis [3], were associated in four cases with congenital pulmonary malformations such as agenesis of the left lung with monolobular right lung [2, 6, 12] or agenesis of the left lung with a right lung of two lobes [12].

In 8 cases it was only *post mortem* that these anomalies were detected [2, 3, 5, 6, 8, 9, 12 (2 cases)], while diagnosis was made *in vivo* in 3 cases [1, 7, 11]. Surgical intervention was performed in a single case only [1]. The patient underwent two operations, both with the aid of extracorporeal circulation. The first operation was merely exploratory. In the second, a channel for the blood of the pulmo-

nary vein was formed in the lumen of the V. cava superior by a longitudinally divided Dacron prosthesis. The systemic veins draining into the azygos vein were carefully ligated, while no mention is made of the ligation of the azygos vein itself before the orifices of the pulmonary veins.

In the present case, all veins of the right lung opened into the azygos vein, and this anomaly was associated with a pulmonary malformation (right lung consisting of 2 lobes and a pleural membrane separating the two lobes) as also with an atrial septal defect.

When correcting a left-to-right shunt caused by the anomalous pulmonary vein, the undesired formation of a right-to-left shunt should be prevented by ligating not only the systemic veins draining into the azygos vein, but also the latter vein distally to the orifice of the pulmonary veins.

Ligation of the azygos vein has no haemodynamic effects. When it is eliminated, a number of collateral veins become operative, multiple anastomoses are formed at the height between the second and fifth thoracic vertebrae [10, 4], so that venous drainage is secured toward the V. cava inferior and superior via the paravertebral, intervertebral, spinal and even the lumbar network of veins.

Operation in hypothermia made it possible in the present case to dispense with extracorporeal circulation; the employed surgical technique (a single purse string suture which began at the surface of the atrium, took up the borders of the defect in the cavity, passed to and knotted on the surface where it had started) allowed to place into the left atrium the orifice of the tube made for the blood of the pulmonary veins and to repair the septal defect in a single session within the time limit determined by the hypothermia.

SUMMARY

After a brief survey of the embryonic development of aberrant pulmonary veins, a case is reported in which all veins of the right lung (consisting of two lobes) emptied into the azygos vein, this anomaly having been accompanied by a defect of the atrial septum. Successful surgical repair has been performed in hypothermia. Of 11 cases of this kind, hitherto reported in literature, only a single one was surgically dealt with, and the technique applied in that case has been compared with the modified procedure used in the present one.

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