Origin of Both Great Arteries from the Right Ventricle and Pulmonary Stenosis

Apropos Case Successfully Corrected*

Morris J. Levy, M.D.,** Richard DeWall, M.D.,*** Larry P. Elliott, M.D.
and Leo Cuello, M.D.

Minneapolis, Minnesota

Congenital cardiac malformations involving the relationship of the two great vessels as they are related to the ventricular chambers are common. A rare condition, however, has been observed in which both great vessels arise from the right ventricle. This malformation was classified by Witham as “double outlet, right ventricle - partial transposition” and he divided the cases into two groups according to whether or not there was associated pulmonary stenosis. Those cases without pulmonary stenosis were designated as of the “Eisenmenger type” and those cases with pulmonary stenosis, as of the “Fallot type.” Edwards, in a later publication, defined this condition as “origin of both great arteries from the right ventricle” and excluded this entity from the cases of persistent truncus arteriosus arising from the right ventricle. Neufeld and associates reviewed five necropsy proved cases with origin of both great vessels from the right ventricle and associated pulmonary stenosis. Although none of the patients previously reported was diagnosed clinically, a suggestion that the clue for proper diagnosis would be in angiocardiographic studies was proposed. More recently, Lucas and associates discussed a case of origin of both great vessels from the right ventricle without pulmonary stenosis. The patient was subjected to operation and anatomic correction was achieved. Unfortunately, death occurred early in the postoperative period, related to massive liver necrosis.

The purpose of this presentation is to report one case with this relatively rare congenital anomaly (origin of both great arteries from the right ventricle with pulmonary stenosis) diagnosed clinically and successfully operated utilizing the cardiopulmonary bypass.

A seven-year-old girl was admitted to the University of Minnesota Hospitals for correction of a congenital cardiac malformation. She was the product of a normal pregnancy and delivery. At the age of 14 months, a cardiac murmur was discovered on a routine examination. At the age of three and a half years, right-sided cardiac catheterization was performed and the diagnosis of a large ventricular septal defect with pulmonic stenosis was made (Table 1). The pressure in the right ventricle was equal to the systemic (100/0 mm. Hg). The pressure in the pulmonary artery was normal (20/10 mm. Hg). A step up in the oxygen content was found at the ventricular

<table>
<thead>
<tr>
<th>Catheter Position</th>
<th>Oxygen cont. (Vol. %)</th>
<th>Oxygen Saturation %</th>
<th>Pressures Mean (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>10.16</td>
<td>64.2</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>10.72</td>
<td>68.5</td>
<td>4/0 2</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>12.08</td>
<td>77.1</td>
<td>100/0</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>13.43</td>
<td>85.8</td>
<td>20/10 15</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>14.53</td>
<td>90.0</td>
<td>100/0</td>
</tr>
<tr>
<td>Aorta (descending)</td>
<td>14.64</td>
<td>9.16</td>
<td>96/60 75</td>
</tr>
</tbody>
</table>

*From the Departments of Surgery and Pediatrics, University of Minnesota.
**Surgeon, on leave of absence, Tel-Hashomer Hospital, Israel.
***Established Investigator, American Heart Association. This study was supported in part by Grants from the United States Public Health Service (No. H-7067), American Heart Association, Minnesota Heart Association, and the Life Insurance Medical Research Fund.
level and an additional increase at the pulmonary artery. The aorta was readily entered during the catheterization. The oxygen saturation of the systemic blood was 90 per cent (left ventricle) and 91.6 per cent (aorta) respectively, indicating an additional right-to-left shunt. Thoracic roentgenograms revealed moderate cardiomegaly, predominantly of the right ventricle. Fluoroscopic examination showed increased pulsations of the ascending aorta, which was noted to be slightly dilated. Until the age of four years, she did well and was asymptomatic when mild cyanosis of the lips after exercise was noted for the first time.

In 1959, at the age of five years, selective aortography was performed to rule out an extracardiac malformation. This study revealed a dilated aorta and bicuspid aortic valve. The ascending aorta was found displaced anteriorly and the aortic valve located unusually high in the horizontal body plane (Fig. 1). By the age of six years, she was hospitalized because of bronchopneumonia. Thereafter, slight cyanosis of the lips at rest was observed, with further increase of exercise intolerance accompanied by cyanotic spells.

The physical examination at time of admission showed the patient in no distress, with normal color and good general appearance. Blood pressures in the arms were 100/70 mm. Hg. The remaining vital signs were normal. Cardiovascular examination showed the cardiac impulse to be located at the fifth intercostal space 2 cm. left of the mid-clavicular line. A systolic thrill was palpable over the aortic area and along the second to fourth intercostal space to the left of the sternal border. A grade 4/6 holosystolic murmur was heard loudest along the third and fourth left intercostal spaces along the sternal border. This murmur transmitted well to the apex and to the neck vessels. A grade 2-3/6 diastolic murmur of blowing character was heard at the aortic area. The second pulmonary sound was diminished - obscured by the systolic and diastolic murmurs.

The electrocardiogram and vectorcardiogram (Fig. 2) showed right ventricular hypertrophy of the systolic (pressure) overload type. Vectorcardiographically the QRS sE loop in the frontal (F) and horizontal planes.
FIGURE 3: Plain frontal chest roentgenogram indicating mild cardiomegaly, left aortic arch with slight dilatation of the ascending aorta. The pulmonary vascular markings are decreased. There is a concave outflow tract of the right ventricle. The left pulmonary artery segment seems to be slightly enlarged.

(H) planes was deviated abnormally to the right and anteriorly. This is represented electrocardiographically by right axis deviation (+180°) and an abnormally tall R wave in lead V1. Additional left ventricular hypertrophy is suggested by an accentuated initial anterior force in the horizontal plane (H) represented by a relative deep Q wave in lead V6.

Conventional chest x-ray examination indicated a moderately enlarged right heart, increased cardiac pulsations, and left aortic arch, somewhat enlarged beyond the limits of the normal. There was the impression of a concave outflow tract of the right ventricle. The pulmonary vasculature was diminished (Fig. 3). Right selective ventriculography was performed and showed: the right ventricle appeared to be enlarged; there was mild tricuspid insufficiency as contrast medium opacified also the right atrium; both great vessels were simultaneously visualized; the aorta seemed to be slightly anterior to the pulmonary artery and markedly dilated; the aortic valve was at the projected level of the pulmonary

FIGURE 4: Selective right ventriculography indicating simultaneous filling of the aorta (A) and pulmonary trunk (P.T.). There is no opacification of the left ventricle. Both vessels take their origin from the right ventricle (R.V.). The aorta (A) lies anteriorly and the aortic valve is situated higher than the normally anticipated. At the right ventricular outflow tract (arrow) there is a "dilution effect" corresponding to the location of the ventricular septal defect (arrow). Left: Postero-anterior projection. Right: Lateral projection.
ORIGIN OF BOTH GREAT ARTERIES FROM RIGHT VENTRICLE

valve; the pulmonary trunk was of normal size with fairly good pulsations on it; the left ventricle was not opacified (Fig. 4). Selective aortography was repeated and demonstrated further dilatation of the ascending aorta overlapping the thoracic spine and lying mainly to the right of the midline. The aortic valve was of bicuspid nature and 2+ (on the basis of 1-4) aortic insufficiency was present. A regurgitant stream of opaque medium opacified the right ventricle by retrograde way as injected in the aorta and immediately after, the pulmonary trunk. Only one coronary artery was opacified arising from the posterior sinus of Valsalva (Fig. 5). On the basis of the catheterization data (from previous admission) and the angiocardiographic studies, the diagnosis of double outlet from the right ventricle, infundibular pulmonic stenosis, ventricular septal defect of the "large" variety, and bicuspid aortic valve with aortic insufficiency was made.

On December 19, 1961, through a midline sternotomy, the heart and the great vessels were exposed. The aorta was lying anteriorly, almost parallel to the pulmonary trunk. There was marked enlargement of the aorta, being almost three times the size of the pulmonary artery. A diastolic thrill was felt at the base of the aorta and along the right ventricle. A systolic thrill was felt at the pulmonary valve area and over the right ventricle. The right ventricle was considerably enlarged. Upon further inspection, there seemed to be a single coronary artery. This appeared to be the left which proceeded normally in the sulcus giving off the path of a normal anterior descending branch with the circumflex running completely around the heart to be lost in the right ventricle. Cardiopulmonary bypass was achieved in the usual way—venous drainage by cannulating both venae cavae through the right atrium and arterial return by direct cannulation of the ascending aortas. A helical reservoir bubble oxygenator and heat exchanger were utilized. The extracorporeal circuit was primed with 415

![Figure 5: Selective aortography illustrating dilatation of the ascending aorta (A), which is anteriorly displaced. The aortic valve is located mainly toward right of the midline and also located unusually high. It is of bicuspid nature. Opaque medium opacified also the right ventricle retrograde fashion, through the insufficient bicuspid aortic valve. There is only one coronary artery opacified (arrow) arising from the posterior sinus of Valsalva. Left: Postero-anterior. Right: Lateral projection.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21376/ on 06/26/2017)
ml. of 5 per cent dextrose in distilled water without donor blood as a hemodilution technique was used. Following institution of the cardiopulmonary bypass, the body temperature was quickly reduced to 29° C. per rectum and 16° per esophagus. Upon entering the right ventricle, the aortic valve was clearly identified, taking off from the right ventricle. The valve was bicuspid and insufficient, as a small amount of regurgitant blood was leaking through the valve in the right ventricle. A narrow channel 6-8 mm. orifice size led to a stenotic infundibulum and the pulmonary artery. The crista supraventricularis was markedly hypertrophied and located above the septal defect. Upon excision of muscle and some amount of fibrotic endocardium from the infundibular area, a free access to the pulmonary valve was achieved. The valve was also bicuspid, with mild fusion along both commissures. The infundibular stenosis was of a fibrous muscular type and extended approximately 1½ cm. in length. Beneath the aortic valve, there was a large ventricular septal defect, oval in shape, approximately 2.5 cm. in diameter. The aorta was excluded from a right ventricle by placing patch (3 by 3.5 cm. Teflon felt). This was sutured to the right ventricle beneath the aortic ring in between the both great vessels, as well as to the lower edge of the ventricular septal defect, approximately the half of the entire circumference of the septal defect (Fig. 6). By this, a continuity of the left ventricle to the aorta was achieved, while the patch became a part of the left ventricular outflow tract by the same time separating the both ventricles from direct communications. The ventricular septal defect itself remained essentially wide open to allow the blood to flow from the left ventricle to the aorta. An outflow roof utilizing a tear-drop Teflon patch (2 by 4 cm.) was additionally placed at the right ventricular outflow tract, extending across the pulmonary artery annulus at the valvular commissure. The right outflow tract was thus increased to the size of a #16 Hegar dilator (Fig. 7). The total bypass

Figure 6: (a) Schematic drawing of origin of both great arteries from the right ventricle at the aortic plane. The origin of the pulmonary trunk (P.T.) is not present at this level. L.V.=left ventricle; R. V.=right ventricle; L.A.=left atrium; R.A.=right atrium; A.=aorta. (b) The technique applied for correction of the anomaly. A patch has been sutured between the lower margin of the ventricular septal defect and the right outflow tract beneath the aortic ring. As a result, continuity of the left ventricle with the aorta was achieved (through the septal defect). Also, by separating both ventricles by this method, the intracardiac flow takes normal course.

Figure 7: Diagrams of the malformation in the natural state (a) and diagrammatic representation of the procedures undertaken to correct the infundibular pulmonary stenosis (b and c). The procedure of diverting blood from the left ventricle directly to the aorta is illustrated in Fig. 6. (a) Section at the right ventricular plane (R.V.). The ventricular septal defect lies beneath the aorta (A). The crista supraventricularis (C.S.) lies between the defect and the aorta. The pulmonary outflow tract is obstructed by infundibular muscle and also valvular stenosis. P.T.=pulmonary trunk; R. A.=right atrium. (b) Surgical relief of the pulmonary obstruction by resection of infundibular muscle and pulmonary valvotomy. (c) Additional widening of the right ventricular outflow tract to the pulmonary trunk by placing a "tear-drop" artificial patch.
time was two hours and six minutes, during which period hypothermic cardiac arrest was applied. The aortic pressure at the end of the procedure was 105/70 mm. Hg. A faint diastolic thrill was still palpable at the aortic base.

The specimen resected from the right infundibular area was a single fragment of muscular tissue, measuring 3 by 1 cm. It consisted of irregular portions of myocardium and mass of fibrous connective tissue without evidence of endocardial sclerosis or other pathologic changes.

The patient tolerated the operation well and following an uneventful postoperative recovery, was discharged two weeks later in satisfactory condition.

**Discussion**

Needless to emphasize is the value of accurate preoperative diagnosis in complex cardiac anomalies.

The case under discussion was originally diagnosed as ventricular septal defect and pulmonary stenosis or tetralogy of Fallot. The clinical, electrocardiographic, and right-sided cardiac catheterization findings supported this diagnosis. The presence of systemic pressures in the right ventricle indicated a large ventricular septal defect. A differential oxygen content between the right and left ventricles, respectively, suggested the presence of two ventricles. The systolic gradient across the pulmonary valve indicated the presence of marked pulmonary stenosis.

As the clinical course of the patient deteriorated with a shift from acyanotic to cyanotic, surgical correction was considered. Since the information obtained from the cardiac catheterization did not exclude the presence of infundibular stenosis, selective right ventriculography was performed. The study yielded the diagnosis of origin of both great arteries from the right ventricle. The position of both semilunar valves in the same horizontal body plane, the displacement of the ascending aortic segment anteriorly, and the aortic insufficiency (on the basis of bicuspid aortic valve) in the right ventricle during repeated left-sided angiocardiology, indicated the diagnosis of origin of both great vessels from the right ventricle.

The surgical approach utilized in this patient was similar to that described by C. W. Lillehei in the case reported by Lucas[4] with the difference that in the case under discussion, associated pulmonary stenosis was also present and relieved by valvotomy and infundibuloplasty.

It is obvious that this entity not properly recognized may give the false impression of ventricular septal defect and pulmonary stenosis. The hemodynamic manifestations are essentially those observed in tetralogy of Fallot, in that the clinical profile depends upon the degree of pulmonary stenosis. Yet, not recognized during operation, “correction” of this condition by closure of the ventricular septal defect in the usual way, would be disastrous.

To the best of our knowledge, this is the first case with origin of both great arteries from the right ventricle with pulmonary stenosis, clinically diagnosed and successfully corrected.

**Summary**

A case with origin of both great arteries from the right ventricle with pulmonary stenosis has been discussed. Preoperative diagnosis was obtained by angiocardiographic studies and confirmed at operation. Successful correction was achieved by redirecting the outflow to the aorta in such a way that the left ventricle would empty properly in the aorta by the use of cardiopulmonary bypass technique. Special care to relieve the associated pulmonary stenosis as well as interruption of the pre-existing intracardiac shunt was also undertaken.

**Resumen**

Se discute un caso en el que ambas arterias mayores emergían del ventrículo derecho, con estenosis pulmonar. El diagnóstico preoperatorio se obtuvo por la angiocardiografía y se confirmó en la operación. Se logró la corrección satisfactoria por medio de rectificación de la corriente aórtica, de tal manera que el ventrículo izquierdo se vaciaría adecuadamente en la aorta por el uso de
la técnica de la desviación cardiopulmonar. Se puso especial cuidado en mejorar la estenosis pulmonar coexistente, así como en interrumpir la intercomunicación intracardíaca pre-existente.

**Resumen**

Discusión d'un cas associant la néissance des deux grosses artères du ventricule droit et d'une sténose pulmonaire. Le diagnostic pré-opératoire a été fait par des études angiocardio-graphics et confirmé à l'opération. La correction chirurgicale a été faite avec succès en redirigeant le flux sanguin dans l'aorte de telle manière que le ventricule gauche s'évade correctement dans l'aorte par l'utilisation d'une technique de Bypass cardiopulmonaire. Un soin particulier a été apporté à réduire la sténose pulmonaire associée, et l'interruption du shunt intra-cardiaque pré-existant a été également faite.

**Zusammenfassung**


**References**


**Mucoviscidosis in Pathogenesis of the Association: Emphysema-Digestive Ulcer**

Having discussed the data in the literature related to the frequency of the association of emphysema to peptic ulcer, the authors report a study of 115 emphysematous patients submitted to the sweat test. A case of ulcer has been noted among 79 emphysematous subjects with negative tests, i.e., 7.6 per cent, while eight ulcer have been found in 56 emphysematous subjects with mucoviscidosis, i.e., 22 per cent. It appears, therefore, that the great frequency of gastro-duodenal ulcers in emphysematous subjects is to be attributed to the presence of this genetic defect.


**Roentgenologic Appraisal of Solitary Pulmonary Nodules**

The reported incidence of malignancy in solitary pulmonary nodules varies from 17 to 28 per cent. At the Mayo Clinic, 44 per cent of 340 cases selected for thoracotomy and 25 per cent of 705 cases in which a nodule was found on a routine roentgenogram of the chest were proved to be malignant. The true incidence probably lies somewhere between these extremes but it is sufficiently great to justify surgical exploration in any individual case unless overwhelming evidence can be elicited to indicate that the lesion is benign.

Evidence of malignancy is obtained most commonly by careful evaluation of roentgenologic evidence. On recent roentgenograms, only those features relating to homogeneity and density supply evidence which is reliable in predicting that a given lesion is benign. If calcium is evident within the nodule, the latter is probably benign. In only one instance in 200 nodular carcinomas was calcification demonstrable in preoperative roentgenograms. The risk of a calcified mass being malignant is less than the risk of thoracotomy. If comparison of previous with recent roentgenograms reveals that a nodule has failed to grow for at least two years, the chances are excellent that it is benign.


Downloaded From: http://journal.publications.chestnet.org/pdaccess.ashx?url=/data/journals/chest/21376/ on 06/26/2017