Valvular Pulmonic Stenosis with Intact Ventricular Septum and Patent Foramen Ovale

Report of Illustrative Cases and Analysis of Clinical Syndrome

By Mary Allen Engle, M.D., and Helen B. Taussig, M.D.

In this malformation the difficulty in sending blood through the stenosed pulmonary valve leads to right heart enlargement and to functional patency of the foramen ovale, through which a venous-arterial shunt occurs. The onset of symptoms and their severity depend on the size of the opening in the pulmonary valve. Characteristic physical findings and results of fluoroscopy, electrocardiogram, circulation time, arterial blood studies, exercise test, angiocardiogram, and cardiac catheterization are presented. Diagnosis is important because these patients develop cardiac failure following a Blalock-Taussig operation, but may be greatly helped by pulmonic valvulotomy.

Valvular pulmonic stenosis with an intact ventricular septum occurs less frequently than pulmonic stenosis associated with a ventricular septal defect and an overriding aorta, as in the tetralogy of Fallot. Greene and co-workers1 after a thorough search of the literature found 68 cases proved by autopsy in which the auricular as well as the ventricular septum was intact. In the remainder of the cases of "pure pulmonic stenosis" the foramen ovale was not completely sealed. This, however, is not necessarily a malformation as the foramen ovale remains patent in a number of "normal" persons.

The first case of valvular pulmonic stenosis and patency of the foramen ovale was reported by Gallois2 in 1809 and is included in Paul's review3 describing three similar cases. Maude Abbott4 in her collection of 1000 cases of congenital malformations of the heart listed 16 additional patients with this malformation. Currens, Kinney, and White,5 from a survey of the autopsy specimens obtained from several Boston hospitals, reported 10 cases with pure pulmonic stenosis, of which 5 had an associated patency of the foramen ovale and an additional case had fenestrations in the septum secundum. Selzer and associates6 have recently reviewed the literature and studied 2 adults who died with this malformation. In addition to those included in Selzer's review, autopsied cases have been reported by Wilson,7 Auerback and Stemmerman,8 Mannheimer,9 Brock,10 Baker and associates,11 and Allanby and Campbell.12

Although the condition has been recognized at the autopsy table for the past 140 years, the correct diagnosis has rarely been made during life. In the past several years we have had the opportunity to study a group of these patients. Although the clinical picture was at first puzzling, it was recognized that the findings in all were quite similar and that all had the same malformation. Subsequent cor-
relation of these findings with postmortem examinations clarified the clinical syndrome. Correct clinical diagnosis is of considerable importance because patients with this type of pulmonary stenosis are not benefited by the Blalock-Taussig operation but are helped by pulmonary valvulotomy. The purpose of this paper is to present the clinical syndrome associated with this malformation. The following cases are selected as representative examples.

![Image](http://circ.ahajournals.org/)

**Fig. 1. Case 1. Anterior-posterior view of chest.** Note cardiac enlargement, prominence of pulmonary conus, large left pulmonary artery, and diminished vascularity of peripheral lung fields.

**Case 1.** W. W. (H. L. H. A 56312), a 24 year old white man, was first examined in July, 1947.

**Past History and Present Illness.** He had had exertional dyspnea for as long as he could remember, and had always tired more readily than other people. The first knowledge of heart trouble dated from 8 years of age, when after a race, he became cyanotic and extremely dyspneic. At that time he was told that he had "leakage of the heart." Thereafter severe exertion produced cyanosis as well as marked dyspnea. He did not, however, squat when tired. His activities were only slightly restricted. He served during World War II in the Merchant Marine.

The development of persistent cyanosis was insidious; the patient was not aware of it until January, 1947, when he was admitted to a hospital because of a left-sided hemiplegia which developed suddenly 16 days after being rendered unconscious in an automobile accident. At that time he was found to be moderately cyanotic and to have clubbing of the fingers. Subsequently the hemiplegia cleared almost completely. However, following this episode, his incapacity, cyanosis, and dyspnea became progressively more severe and he occasionally suffered from swelling of the ankles. In June, 1947, he was found to have a right pneumothorax. After the lung had re-expanded, he was referred to the Cardiac Clinic as a possible candidate for cardiac surgery.

**Physical Examination.** Pulse 102 per minute; respirations 20 per minute; blood pressure 124/94. The patient was a well-developed, well-nourished man with moderate cyanosis and clubbing of the fingers and toes. There was anterior bowing of the left chest. No thrill was palpable over the precordium. The heart was enlarged. The heart sounds were distinct though less loud than normal. The second sound was louder over the pulmonic area than over the aortic area but was not accentuated. There was a harsh but rather soft, blowing systolic murmur best heard in the third and fourth intercostal spaces to the left of the sternum. It was transmitted over the entire precordium and to the left interscapular area posteriorly. There was a presystolic gallop rhythm. The lungs were clear. The liver extended 2 fingerbreadths below the costal margin and was pulsating. The pulsations in the femoral arteries were strong. There was no edema. Except for a residual left facial paralysis the remainder of the physical examination was negative.

**Laboratory Data.** The red blood cell count was 7.68 million per cu. mm.; the hemoglobin level was 19.8 Gm. per 100 cc. and the hematocrit reading was 65. The oxygen content of the arterial blood was 23.4 volumes per cent and the oxygen capacity was 30.1 volumes per cent; hence the saturation was 78.3 per cent. The arm-to-tongue circulation time, determined with Decholin, was prolonged to 18.7 seconds. Electrocardiogram showed high peaked P waves, incomplete A-V block, and right bundle branch block.

**Fluoroscopy** in the anteroposterior view showed that the heart was enlarged to the right and to the left. In the oblique views the cardiac shadow extended abnormally far both anteriorly and posteriorly. These findings were thought to indicate enlargement of the right auricle, the right ventricle, and the left ventricle. The right ventricular pulsations were of exceedingly small amplitude. The pulmonary conus was prominent, and the pulmonary window was obscured. The lung vascular markings, however, appeared reduced, and only very slight pulsations could be seen in the hilar vessels. There was a left aortic arch and no left auricular enlargement. Telerontgenogram showed the cardiothoracic ratio was 57 per cent (fig. 1).

Since the nature of the malformation was not clear, additional studies were undertaken. An exercise test showed a drop in the amount of oxygen consumed per liter of ventilation from 24.7 cc. at rest to 18.6 cc. after exercise. There was a corresponding decrease in carbon dioxide production from 23.1 to 20.9 cc.

**Catheterization of the right heart** was attempted on
two occasions, but on neither occasion could the catheter be introduced into the right ventricle. The right auricular pressure was elevated to 29/15 mm. Hg.

Angiocardiogram showed that the Diodrast entered via a dilated superior vena cava into a large right auricle and a large thick-walled right ventricle. The dye was expelled very slowly from the right side of the heart into a dilated pulmonary artery, where it lingered for an abnormally long time before being distributed to the lungs. Because the right pulmonary artery was hidden behind the enlarged heart, it was at first thought that there was but a single pulmonary artery. No evidence of overriding of the aorta was demonstrable and the left side of the heart was never visualized. The dye still remained in the right heart and pulmonary artery at the end of the series of angiocardiograms. (The last of these x-rays was taken about 11 seconds after the injection of the dye.)

Clinical Impression. It was thought that the patient had pulmonic stenosis and tricuspid insufficiency; the possibility of Ebstein's malformation of the tricuspid valve was considered. A Blalock-Taussig operation was contraindicated because of the enlarged heart, the poor heart action, the pulsating liver, and the failure to demonstrate an overriding aorta.

Course. The patient was given digitalis and a period of bed rest was advised. In spite of therapy he became more dyspneic, cyanotic, and incapacitated. In October, 1947, he was able to walk only four blocks very slowly before becoming exhausted. At this time his arterial oxygen saturation at rest had dropped to 53 per cent. He developed multiple emboli to the right leg, to the brain, and to the lungs. On March 16, 1948 he was admitted to the U. S. Marine Hospital in Seattle, Washington in moribund condition and he died 15 hours later.

Autopsy. (Performed by Dr. George E. Tooley, U. S. Marine Hospital, Seattle, Washington). The heart was markedly enlarged; it weighed 725 Gm. (normal 300 Gm.) (See figure 2.) The great veins entered in the normal fashion. The right auricle was tremendously dilated, and its walls were moderately hypertrophied. There was a gross patency of the foramen ovale which measured 6 by 8 mm. The tricuspid valve measured 12.5 cm. and was normal in appearance. The right ventricle was enormously hypertrophied. The myocardium had an average thickness of 2.4 cm. in contrast to an average thickness of the left ventricle of 1.2 cm. Microscopically the right ventricle showed diffuse myocardial scar-

* We are indebted to Dr. Gordon A. Abbott, Clinical Director of the Marine Hospital, for the photographs of the specimen and the pathology report and to Dr. Stewart Lippincott, Professor of Pathology at the University of Washington Medical School, for permitting us to study the specimen.

ring. There was marked stenosis of the pulmonary valve (fig. 3). The cusps of the pulmonary valve were completely fused into a dome-shaped membrane with only a small central opening measuring 2 by 4 mm. Below the stenotic pulmonary valve was a mural thrombus firmly adherent to the endocardium over an area 2 by 3 cm. The pulmonary arteries appeared dilated. The left auricle, mitral valve, and left ventricle were normal. There was no ventricular septal defect. The aorta arose from

Fig. 2. Case 1. Photograph of heart, showing enlarged right auricle and right ventricle. Patent foramen ovale is indicated by pointer. Hypertrophy of right ventricular musculature is apparent.

Fig. 3. Case 1. Photograph of heart. Note dome-shaped protrusion of stenosed pulmonary valve and small, central perforation in the fused cusps. The dilated pulmonary artery has been laid open.
the left ventricle in the normal manner, and both
the aortic valves and the aorta were normal. There
was generalized visceral congestion. The liver was
enlarged. Both the liver and spleen showed gross
and microscopic evidence of long-standing passive
congestion.

Final Anatomic Diagnosis. Congenital malforma-
tion of heart. Valvular pulmonic stenosis. Post-
stenotic dilatation of pulmonary artery. Patent
foramen ovale. Hypertrophy of right ventricle. Dilat-
tation and hypertrophy of right auricle. Mural
thrombus in right ventricle. Passive congestion of
viscera. Congestive cirrhosis of liver.

Case 2. D. L. (H. L. H. A 62640), a 6 year old
white boy, was first seen in the Cardiac Clinic in
May, 1948.

Past History and Present Illness. There was no
cyanosis at birth or in early infancy, but at about
6 months of age cyanosis and dyspnea appeared on
exertion. At this time a systolic murmur was noted.
At 18 months of age cyanosis became persistent, and thereafter it steadily increased. He easily became dyspneic and fatigued, but he squatted to rest only infrequently. His exercise capacity gradually became more limited. At 6 years of age he was able to walk only three blocks on a good day and was tired after climbing one flight of stairs. There were no episodes of paroxysmal dyspnea, loss of consciousness, or convulsions. The family physician noted progressive cardiac enlargement.

**Physical Examination.** Temperature 37°C; pulse 96 per minute; respiratory rate normal at rest; blood pressure 100/80. He was slender but well developed. There was moderate cyanosis and clubbing of the fingers and toes. There was a precordial bulge. The heart was enlarged but no thrill was palpable. There was a soft, blowing, systolic murmur in the third intercostal space at the left sternal border. The second sound at the base of the heart was weak. The liver edge was 2 fingerbreadths below the right costal margin and was pulsating. The pulsations in the femoral arteries were strong. There was no edema. The remainder of the examination was essentially negative.

**Laboratory Findings.** The red blood cell count was 6.75 million per cu. mm.; the hemoglobin level was 22 Gm. per 100 cc.; the hematocrit reading was 56. The arterial oxygen content was 10.2 volumes per cent; the capacity was 24.4 volumes per cent, giving an arterial oxygen saturation of 78.8 per cent. The arm-to-tongue circulation time with Decholin was 12 seconds. In the unipolar and standard leads, the electrocardiogram showed high, peaked P waves, first degree heart block, and right bundle branch block.

**Fluoroscopy** in the anteroposterior view revealed that the heart was moderately enlarged to the right and to the left (fig. 4). There were strong, asynchronous pulsations of the right and left cardiac borders. There was a convexity in the region of the pulmonary conus, and a large left pulmonary artery was seen to continue from this point into the left lung. The right pulmonary artery appeared to be of normal size. There were no expulsive pulsations visible in either pulmonary artery, and the lung fields were abnormally clear. In the oblique views (fig. 4) there was evidence of great enlargement of the heart both anteriorly and posteriorly. Barium swallow showed a left aortic arch and no enlargement of the left auricle. Telerentgenogram revealed the cardiothoracic ratio to be 69 per cent.

**Clinical Impression.** Valvular pulmonic stenosis with a patent foramen ovale was considered the most probable diagnosis. Ebstein's anomaly of the tricuspid valve was considered a possibility. To differentiate the two conditions and to clarify the diagnosis, additional studies were undertaken.

**Angiocardiogram** (fig. 5) showed that the Diodrast entered the right auricle through the dilated superior vena cava and passed into the large, thick-walled right ventricle. At the end of two seconds, the contrast medium appeared in the main pulmonary artery, which occupied the position of the prominent pulmonary conus seen on fluoroscopy. The contrast solution remained in the right side of the heart and in the pulmonary artery for an abnormally long time. The opacification of the parenchymal pulmonary vascular tree was less than normal. No overriding of the aorta was demonstrated. The left side of the heart was poorly visualized.

**Right heart catheterization** revealed that the oxygen content of the superior vena cava was 9.2 volumes per cent and of the right auricle, 8.4 volumes per cent in one region and 12.2 volumes per cent in another location. The oxygen content of the right ventricle was 12.2 volumes per cent. The pulmonary artery could not be catheterized. The pressure in the right auricle was elevated, and in the right ventricle it was 112/55. Calculation of the blood flows showed a reduced pulmonary artery flow of 1280 cc. and a systemic flow of 1830 cc. per square meter of body surface per minute. Although there was a small shunt of oxygenated blood from the left auricle into the right auricle, as indicated by the higher oxygen content of one of the samples taken from the right auricle, the overall shunt was from right to left and was calculated to be 550 cc. per square meter of body surface per minute. Thus catheterization studies were consistent with the clinical impression of valvular pulmonic stenosis and patency of the foramen ovale.

**Course.** The catheterization procedure was a short one, and the patient returned to the ward in good condition. Shortly afterwards, he appeared restless. He was examined by several doctors and given a small dose of morphine. His condition was thought to be satisfactory. Suddenly, about three hours after completion of the catheterization, he gasped, became deeply cyanotic, and the respirations and heart action ceased. Resuscitative measures were of no avail.

**Autopsy.** (No. 21233 performed by Dr. Jack M. Burnett.) The chief interest centered about the enlarged heart. The enlargement was limited to the right auricle and the right ventricle, which formed the anterior surface of the heart. Venous return was normal. The right auricle was hypertrophied and was about twice the size of the left auricle. Although in part protected by a thickened valve, the foramen ovale was patent for an area of 2 by 8 mm. The tricuspid valve was bicuspid but competent; its edges were slightly thickened. The right ventricle was greatly hypertrophied. Its wall in some areas measured 2.5 cm. in thickness, whereas the thickness of the left ventricle was 1.2 cm. Near the apex of the right ventricle there was patchy scarring of the myocardium. The endocardium was

*We are indebted to Dr. Richard Bing and his associates for these studies.
uniformly slightly thickened. The ventricular septum was intact. The leaflets of the pulmonic valve were completely fused, producing a conical protrusion into the pulmonary artery. There was a small opening 2 mm. in diameter at the apex of the cone. The main pulmonary artery distal to the stenosed valve was about twice the usual diameter, and its uniform walls were thin. The left auricle and left ventricle, the mitral and aortic valves, the aorta, and the coronary arteries were normal. The ductus arteriosus was obliterated. The bronchial arteries were slightly enlarged; their lumina each measured 1 mm. in diameter. The remainder of the postmortem examination was not remarkable.


Case 3. C. N. S. (H. L. H. A 65191), a 2 year old white girl, referred to the Cardiac Clinic in August, 1948.

Past History and Present Illness. Cyanosis was noted at birth, and it progressively deepened. Dyspnea appeared after moderate exertion, but there were no attacks of paroxysmal dyspnea, and she did not assume a knee-chest position when fatigued. She was unable to walk. From the age of 8 months she gained weight poorly. At the time of her examination in the clinic, the mother felt that the child's condition was gradually deteriorating.

Physical Examination. Temperature 37 C.; pulse 140 per minute; respirations 30 per minute; blood
pressure 114/70 in the arm and 118/78 in the leg. She was a small, underdeveloped child who weighed only 24½ pounds at the age of 2 years. There was slight clubbing of the fingers and toes, and even at rest there was cyanosis. With crying she became dyspneic and markedly cyanotic. Examination of the chest showed no precordial bulge; no thrill was felt. The heart was enlarged; the heart sounds were of good quality. In the third intercostal space to the left of the sternum was a loud, harsh, systolic murmur which was propagated over the precordium and upward to the left shoulder; over the pulmonic area it obscured the second heart sound. The liver was palpable one fingerbreadth below the costal margin and was pulsating. There was no edema. The remainder of the physical examination was negative.

**Laboratory Findings.** The red blood cell count was 9.55 million per cu. mm., the hemoglobin level was 21.5 Gm. per 100 cc. and the hematocrit reading 76. A sample of femoral arterial blood, obtained when the child was crying, revealed an arterial oxygen saturation of only 14 per cent. The oxygen content was 3.1 volumes per cent, the oxygen capacity 21.2 volumes per cent, and the carbon dioxide content 36.8 volumes per cent. The electrocardiogram showed marked right axis deviation. There were high, peaked P waves, first degree heart block, and evidence in the unipolar precordial leads of right bundle branch conduction disturbance. T₂ and T₁ and the T waves in the precordial leads were deeply inverted.

**Fluoroscopy** revealed a heart that was tremendously enlarged both to the right and left in the anteroposterior view, and both anteriorly and posteriorly in the left anterior-oblique position. The pulmonary conus region was full; there were, however, no pulsations in the pulmonary arteries, and there was diminished vascularity of the lung fields. There was a left aortic arch. No enlargement of the left auricle was demonstrated after barium swallow. The fluoroscopic findings were interpreted as indicative of diminished pulmonary circulation and great enlargement of the right auricle and right ventricle and questionable enlargement of the left ventricle. Teleroentgenogram showed the cardio-thoracic ratio was 81 per cent (fig. 6).

**Clinical Impression.** Valvular pulmonic stenosis with a patent foramen ovale or an auricular septal defect.

**Course. Angiocardiography** was undertaken to confirm the diagnosis. The patient showed no reaction to the test dose of Diodrast; so after a cutdown in the right arm, 9 cc. of the Diodrast was injected and a series of anteroposterior films taken. The contrast medium entered through a dilated superior vena cava into a very large right auricle and then into the large, thick-walled right ventricle. Some of the Diodrast filled the main pulmonary artery, which occupied the region of the full pulmonary conus, and a small amount of contrast medium entered the larger branches of the pulmonary circulation. The right side of the heart emptied very slowly. The dye was still present in the right ventricle and main pulmonary artery at the conclusion of the series of films seven seconds after start of injection. Neither the aorta nor the left side of the heart was visualized by this time.

The patient seemed well immediately following the procedure, but five minutes later she suddenly stopped breathing, and the heart sounds could not be heard. Resuscitative efforts were futile.

**FIG. 6. Case 3. Anterior-posterior view of chest.** Note tremendous cardiac enlargement and the fullness in region of pulmonic conus.

**Autopsy.** (No. 21480 performed by Dr. Lloyd Shannon). The heart was greatly enlarged, its tremendous size being due entirely to the large right auricle and right ventricle, which formed the anterior aspect of the heart. The heart weighed 140 Gm. (normal 50 Gm.). Venous return was normal. The right auricle was extremely dilated and its wall moderately hypertrophied. The foramen ovale was widely patent, measuring 2 cm. in diameter. The tricuspid valve was bicuspid. The right ventricle was markedly hypertrophied. Its wall measured 1.8 cm. in thickness, whereas the wall of the left ventricle measured 1 cm. The ventricular septum was intact. The pulmonary valve was extremely stenosed. The valve cusps were fused to such a degree that they formed a cone with only a pinpoint opening about 1 mm. in diameter at the apex of the cone. The pulmonary artery distal to the valve was dilated and was of approximately the same size as the aorta. The left side of the heart and the aorta were normal. There was probe patency of the small ductus arteriosus. Two of the bronchial arteries were slightly enlarged. The spleen was enlarged, but the liver was normal.
No cause for the sudden death was found. The lungs were well aerated. There were no evidences of air emboli. The trachea and bronchi were free from obstruction. Examination of the cranial cavity revealed no abnormalities.


**DISCUSSION**

These three cases illustrate the clinical syndrome, which is readily explicable on the basis of the pathologic anatomy. The stenosed pulmonary valve places a tremendous load on the right side of the heart and causes difficulty in the expulsion of blood into the pulmonary artery. This leads to hypertrophy of the right ventricle and to progressive increase in the systolic pressure. Gradually the right ventricle becomes unable to empty itself completely, and the diastolic pressure within the ventricle rises. This in turn increases the work of the right auricle; consequently, the right auricle hypertrophies, and the pressure within that chamber rises.

When the pressure in the right auricle exceeds that in the left auricle, if the foramen ovale is not completely sealed, it is forced open, and venous blood is shunted from the right auricle into the left auricle. When the volume of unoxygenated blood so shunted into the systemic circulation is sufficiently large, the level of visible cyanosis is reached. The cyanosis is at first transitory, then becomes persistent, and deepens as the patient grows older. As the volume of the venous-arterial shunt increases, polycythemia appears. The increase in the height of the hemoglobin level renders the cyanosis more intense.

At birth there probably is less tendency for the foramen ovale to close than in a normal heart. However, so long as the pulmonary orifice is relatively adequate in size and the pressure in the right auricle is lower than in the left auricle, the valve of the foramen ovale is functionally closed.

The patenty of the foramen ovale is the only means by which an intracardiac venous-arterial shunt can occur in this malformation. The evidence is strong that without such a shunt, there is no decrease in arterial oxygen saturation. It has been well demonstrated that a diminution in pulmonary circulation *per se* will not produce arterial unsaturation. In those patients with pulmonic stenosis as part of the tetralogy of Fallot in whom Bing was able to catheterize the pulmonary vein, he found the oxygen saturation of the pulmonary vein to be normal (95 to 96 per cent). Indeed, in one of our patients with valvular pulmonic stenosis and a patent foramen ovale to be included in a subsequent report, the pulmonary vein was catheterized, and its oxygen saturation was found to be 97 per cent.

Analysis of the findings in our patients and those in the literature corroborates the belief that in this condition the cyanosis due to arterial oxygen unsaturation is related to the patency of the foramen ovale. The foramen ovale was patent in 41 cases previously reported. Of these patients on whom clinical data is available, 35 showed cyanosis, and only 1 did not. The last mentioned was a 4 ½ month old baby who died of an unrelated cause (subdural hematoma). In this instance, although there was probe patency of the foramen ovale, it appeared to be functionally closed.

Of the 71 patients with pulmonic stenosis and no defect in the auricular or ventricular septa, cyanosis of slight degree was commented upon in 29. As there was no opportunity for a venous-arterial shunt in these patients, the cyanosis was in all probability not due to arterial oxygen unsaturation. On the contrary, it seems quite likely that cyanosis was caused by venous stasis. That such may be the cause is well illustrated by Arnett’s patient, who when decompensated showed splotchy cyanosis of the ear lobes, the tip of the nose, and the cheeks. His arterial oxygen saturation, however, was 94.6 per cent. At autopsy the foramen ovale, though not sealed, was closed and was guarded on both sides by large folds.

The elevated pressure in the right side of the heart in patients with pulmonic stenosis with an intact ventricular septum eventually causes venous congestion and enlargement of the liver, and ultimately, if the right ventricle fails, may
lead to peripheral edema and ascites. The patency of the foramen ovale acts for a time as an “escape valve” and lessens the load on the right side of the heart by the shunt of blood from the right auricle into the left. Even while the tricuspid valve remains competent, the forceful contractions of the right auricle may cause pulsations of the jugular veins and the liver.

It seems probable that the size of the opening in the fused pulmonary leaflets is fixed at birth and does not increase in size as rapidly as does a normal valve. What may be a relatively adequate pulmonic opening for a small, inactive infant becomes progressively less adequate as the patient grows and his activity increases. As the stenosis thus becomes increasingly more severe, the heart progressively enlarges, and symptoms appear and steadily become more pronounced. The smaller the orifice of the stenotic valve, the earlier is the onset of cardiac difficulties. If the stenosis is extreme at birth, the pulmonary orifice may be inadequate even for a baby, and the patient may die in infancy or early childhood. This concept explains the variations in longevity and in severity of symptomatology in these patients. The average age at death of those reported in the literature was 19 years, but the range was from 4 months to 57 years.

The primary symptom is dyspnea on exertion; this is usually present from early life. It generally precedes the onset of cyanosis and is out of proportion to the cyanosis. It is due to the difficulty in the direction of blood to the lungs. As the shunt of blood through the foramen ovale from the right auricle to the left becomes established and the pulmonary flow is thereby diminished, exertional dyspnea becomes more pronounced. The easy fatigability is also related to the slow delivery of blood to the lungs for oxygenation. The pulmonary stenosis renders it impossible for the individual to increase the pulmonary blood flow in the normal manner with exercise.

The physical findings are readily explicable on the basis of the pulmonary stenosis. The presence of a precordial bulge is due to the hypertrophy of the right ventricle during early childhood. The systolic murmur, maximal in the pulmonic area, is caused by the ejection of blood through the stenosed pulmonary orifice. The stenosis of the pulmonic valve causes a reduction in the intensity of the second sound over the pulmonary area. Enlargement and pulsations of the liver are manifestations of the hypertrophy of the right side of the heart and the high pressure in the right auricle.

The electrocardiographic findings of marked right axis deviation and right ventricular hypertrophy and the frequent finding of right bundle branch block are related to the increased thickness of the right ventricular wall and possibly to the diffuse scarring of the right ventricular myocardium. The high, peaked P waves probably reflect the enlargement of the right auricle. Arterial oxygen unsaturation is present after the shunt from right to left through the foramen ovale has become established. This in turn leads to compensatory polycythemia. The prolonged circulation time is related to the difficulty in the expulsion of blood from the right ventricle. It has been our clinical experience that although the foramen ovale is patent, rarely is enough of the test material shunted from the right auricle through the left auricle and thence to the left ventricle and the systemic circulation to give a short circulation time.

The x-ray and fluoroscopic findings are also explicable on the basis of the pulmonic stenosis. There is progressive enlargement of the right ventricle and the right auricle. On rotation of the patient into the left anterior-oblique position the left ventricle is often displaced so far posteriorly by the large right ventricle that it far overlaps the spine and gives the appearance of enlargement of both the right and left ventricles (see figure 4). The force of contraction in the right auricle and right ventricle seen on fluoroscopy is worthy of comment. Often a “rocking boat” effect is seen in the anteroposterior view as first the right auricle, forming the right heart border, contracts forcefully, and then the hypertrophied right ventricle, forming the left heart border, contracts. The amplitude of the pulsations has been conspicuous in all but 2 of our patients (case 1 and another not included in this report). Moreover, in these 2 the pulsations of the right
auricle were strong, whereas the contractions of the right ventricle were so weak that pulsations were scarcely visible. This finding was a late manifestation, as indicated by the fact that both patients died within a few months after such pulsations were seen. This observation suggests that when the hypertrophied right ventricular musculature begins to fail, the force of its contractions becomes much reduced. Consequently, such a finding is of grave prognostic importance.

Although there is a prominence of the pulmonic conus region and the pulmonary arteries are normal in size or enlarged, pulsations in the pulmonary arteries are absent or minimal, and the peripheral lung vascular markings are diminished. These findings are in keeping with the low pressure in the pulmonary artery. The prominence of the conus region was demonstrated both by angiogram during life and by postmortem examination to be due to the dilatation of the pulmonary artery distal to the valvular stenosis. This poststenotic dilatation may be the result of the ejection of a small volume of blood with each ventricular contraction into a pulmonary artery that was originally normal in size. The force of the ejection is almost completely broken by the pulmonary stenosis so that only a thin jet of blood enters the pulmonary artery through the small central perforation. As there is little force to distribute the blood into the pulmonary circulation, the blood pools in the main pulmonary artery before it sluggishly circulates through the lungs.

The finding on angiography of delay of the dye in the right side of the heart and in the pulmonary arteries reflects the difficulty in expulsion of blood from the right ventricle forward into the pulmonary circulation. Early visualization of the aorta does not occur until some of the contrast solution has passed from the right auricle through the defect in the auricular septum to the left. The volume so shunted is relatively small, and the aorta is often visualized in only one of the two series of films taken in the anteroposterior and in the lateral positions. Although there may be faint opacification of the aorta, there is not the dense, early filling which is seen when the aorta overrides the right ventricle.

Because of the pulmonic stenosis and the inability to increase the pulmonary blood flow with exercise, the exercise test usually shows a drop in the oxygen consumption per liter of ventilation with exercise compared with that at rest. The measurement on cardiac catheterization of an elevated pressure in the right auricle and particularly in the right ventricle and of a low pressure in the pulmonary artery is in keeping with the hemodynamic changes proximal and distal to an obstruction at the pulmonary valve.

This malformation requires differentiation from other malformations of the heart which are associated with cyanosis and inadequate pulmonary circulation. The two chief malformations with which it is likely to be confused are the tetralogy of Fallot and Ebstein's malformation of the tricuspid valve.

The differentiation from the tetralogy of Fallot is important because, while the latter malformation is dramatically helped by the Blalock-Taussig operation, each of the 3 patients with valvular pulmonic stenosis who were operated upon because of an erroneous diagnosis of tetralogy of Fallot failed to do well after operation. Although the dyspnea and cyanosis were improved after the circulation to the lungs was increased, the increased volume of blood returned to the left auricle tended to close the valve of the foramen ovale, and thereby increased the strain on the right side of the heart. In each of the above-mentioned patients operation caused tremendous cardiac enlargement and cardiac failure with a huge pulsating liver.

These patients have since been reoperated upon, 2 by Mr. Brock and 1 by Dr. Blalock. The pulmonic stenosis was approached directly through the right ventricle, and the pulmonic

* Because of the death of one patient (case 3) immediately following angiography, we feel this procedure is dangerous in patients with extreme pulmonary stenosis and impending heart failure, as indicated by a very low arterial oxygen saturation, great cardiac enlargement, or poor amplitude of pulsations in the right ventricle. Under such circumstances the sudden injection of contrast material under pressure may add too great a load to the already overtaxed right heart and may deprive the patient of oxygen.
valve was cut; subsequently, the subclavian-pulmonary artery anastomosis was closed. The first patient was in severe heart failure and had recently developed a hemiplegia. He died immediately after the operation. The diagnosis of valvular pulmonic stenosis with patency of the foramen ovale was confirmed at autopsy. The other 2 patients were greatly improved. In each instance the liver promptly receded and ceased to pulsate, and the patient regained compensation. The arterial oxygen saturation rose to nearly normal levels, the symptoms were alleviated, and the patients have done well. Pulmonic valvulotomy has since been performed with gratifying results on other patients with this malformation. These will be reported at a later date.14

The clinical picture presented by the tetralogy of Fallot is quite distinct from that produced by valvular pulmonic stenosis with a patent foramen ovale. The absence of marked cardiac enlargement and the concave curve in the pulmonary area, the absence of pulsations of the liver, and the short circulation time differentiate the tetralogy of Fallot from "pure" pulmonic stenosis. The following features are also helpful. In the tetralogy cyanosis and dyspnea are proportionate to each other and usually appear in early infancy. Attacks of paroxysmal dyspnea are common. The patient typically assumes a squatting position to rest. The electrocardiogram shows right axis deviation and right ventricular hypertrophy; however, there is usually no prolongation of auriculoventricular or of intraventricular conduction time. In doubtful cases either angiocardiology or cardiac catheterization aid in the differentiation of the two malformations.

This clinical syndrome even more closely resembles that due to Ebstein's anomaly of the tricuspid valve with patency of the foramen ovale.20 This malformation is not amenable to surgery. It is differentiated by the following features. In Ebstein's anomaly cyanosis is more marked than the dyspnea. The heart sounds usually are of poor quality, often there is a gallop rhythm, and there may be a diastolic murmur in addition to the systolic murmur. The liver may be enlarged, but unless the patient is in heart failure, there are no pulsations palpable at its margin. On fluoroscopy there is an absence of fullness in the region of the pulmonic conus, and the pulsations of the right auricle and the right ventricle are much reduced in amplitude. Angiocardiograms, as in valvular pulmonic stenosis, show delayed emptying of a large right auricle and ventricle, but the contrast solution can usually be seen to extend almost to the margin of the cardiac silhouette, indicating that the right auricle and right ventricle are thin-walled. Although cardiac catheterization is not recommended if the diagnosis of Ebstein's anomaly is suspected, in 1 patient in whom it was performed, the findings differed from those of valvular pulmonic stenosis in that the pressures in the right auricle and right ventricle were not greatly elevated.20

Cor pulmonale with patency of the foramen ovale21 may be confused with this malformation. The quality of the second sound at the base may aid in the differentiation of these two malformations. In patients with pulmonic stenosis, the second sound at the base is weak, whereas in cor pulmonale it is usually accentuated. In doubtful instances cardiac catheterization and measurement of the pressure in the pulmonary artery may be necessary to differentiate the two conditions.

The Eisenmenger complex may be suggested by the delay in onset of cyanosis and the full pulmonary conus. The chief differential points in this malformation are the absence of a pulsating liver, the frequent finding of a blowing, diastolic murmur, and the fluoroscopic visualization of enlarged and pulsating pulmonary arteries and congested lung vascular markings. Due to the overriding aorta there is a short circulation time. The exercise test usually shows a rise in oxygen consumption per liter of ventilation after exercise. Right heart catheterization shows a high pressure in the pulmonary arteries as well as in the right ventricle.22 It may be possible to catheterize the aorta from the right ventricle. On the angiocardiogram there is prompt emptying of the right side of the heart, the pulmonary circulation is increased, and there is early visualization of the overriding aorta.
Summary

Three illustrative cases of valvular pulmonic stenosis with patency of the foramen ovale proved by autopsy have been presented. Although the malformation has been recognized at postmortem examination for the past 140 years, the correct diagnosis heretofore was rarely made before death. Nevertheless, correlation of the clinical and pathologic findings in the group of patients collected from the literature and of those which we have studied reveals that the clinical picture presented by this malformation is so characteristic as to permit its accurate diagnosis during life.

The salient features of the clinical syndrome are: delayed onset of cyanosis, dyspnea which is usually greater in severity and earlier in appearance than is the cyanosis, absence of squatting to rest after easy fatigue, a precordial bulge, an enlarged heart with a pulmonic systolic murmur and a weak second sound, and an enlarged and pulsating liver. Fluoroscopy shows vigorous pulsations in the enlarged right auricle and, unless it is failing, in the large right ventricle. The pulmonic conus is prominent, and there are large pulmonary arteries with minimal or absent expansile pulsations. The peripheral lung fields are abnormally clear. The electrocardiogram shows evidence of either right bundle branch block or right axis deviation and right ventricular hypertrophy. There is often a first degree A-V heart block and the P waves are high and peaked. The circulation time is usually prolonged. There is arterial oxygen unsaturation and compensatory polycythemia. Angiocardiogram shows delayed emptying of the hypertrophied right auricle and right ventricle and the dilated pulmonary artery; visualization of the aorta early in the series of films occurs only after some of the contrast solution has passed from the right auricle into the left auricle and left ventricle. Right heart catheterization shows an elevated pressure in the right auricle and right ventricle and a low pressure in the pulmonary artery.

The malformation must be differentiated from the tetralogy of Fallot, from Ebstein’s anomaly of the tricuspid valve, from cor pulmonale, and also from the Eisenmenger complex. The differential diagnosis is discussed. It is important to make the correct diagnosis, for patients with this malformation may be helped by valvulotomy.

REFERENCES


2 GALLOIS: Bull de la Faculté de méd. de Paris 133, 1809. Quoted by Paul.


14 TO BE REPORTED BY THE DEPARTMENT OF SURGERY, Johns Hopkins Hospital, Baltimore, Maryland.

15 ROSSMAN, J. L.: Congenital atresia and stenosis of


21 Cor pulmonale with patency of the foramen ovale to be reported from the Cardiac Clinic of the Harriet Lane Home, Johns Hopkins Hospital.

MARY ALLEN ENGLE and HELEN B. TAUSSIG

Circulation. 1950;2:481-493
doi: 10.1161/01.CIR.2.4.481

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1950 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/2/4/481

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/