A Diagnostic Problem: Noncyanotic Heart Disease with Heart Failure in a Seven Year-Old Boy

A Clinical Pathological Conference held at the University of Kansas Medical Center

The Clinicopathological Conference at this school is somewhat unique. Fourth year students are responsible for the discussion of the case, interpretation of x-ray films, and of electrocardiograms. The comments of the students, in the following presentation, reflect the maturity of the modern-day senior.

CASE PRESENTATION

D R. MAHLON DELP, Moderator: Dr. Weber will you present the history of the case which is to be considered today?

Dr. Weber (Senior Resident in Medicine): This 7 year old boy was admitted to the University of Kansas Medical Center on Nov. 25, 1953 for possible surgery, with the chief complaint of “heart disease”.

Review of the patients medical history disclosed that at the time of his birth, his parents had been informed that he had an enlarged heart. Cyanosis, however, had not been observed to be present either at the time of birth or later. Squatting behavior had never been noticed. Although the parents had observed that the boy had dyspnea on exertion, his physical development had been normal. Until one month before admission to the hospital, the child had attended school and played well with other children. At that time, however, the patient developed an upper respiratory infection. Dyspnea and easy fatigability had been progressive since this illness. At the time of admission he was unable to walk 10 steps without severe dyspnea. Further questioning disclosed that since birth the patient had numerous respiratory infections and that within the three years just prior to his admission to the hospital he had experienced three illnesses diagnosed as pneumonia. Ingual herniorrhaphies had been performed in 1949 and 1951.

The family history was essentially negative. The patient’s mother did not remember any illnesses occurring during pregnancy. There were no siblings.

On physical examination the patient was observed to appear chronically ill and to be in moderate respiratory distress at rest, but he was not cyanotic. A marked pigeon-breast deformity of the chest and slight kyphosis of the dorsal spine were present. The patient’s temperature was 97 F., his pulse rate 124 beats per minute, his respiratory rate 36 per minute, and his blood pressure 90/60. His height was 46 inches and his weight 53 pounds. Veins of the neck were distended and pulsating. The anterior posterior diameter of the chest was increased. The chest was hyperresonant to percussion. The breath sounds were slightly diminished and only an occasional rale was present in the bases. The point of maximal impulse was located in the sixth intercostal space in the midaxillary line. There was a bounding precordial pulsation. A grade II systolic thrill was felt at the apex of the heart. A grade IV systolic murmur was heard along the left sternal border and at the cardiac apex. A grade III diastolic murmur was heard at the pulmonic area. The second pulmonic sound was accentuated. The liver extended 10 cm. below the right costal margin and the splenic tip was palpated at the left costal margin. Pedal edema was not present and pulsations in peripheral arteries were normal.

Urinalysis revealed specific gravity of 1.015; it was otherwise negative. Hemoglobin was 11.1 Gm. per 100 cc. and white blood count was 8,400 with 66 per cent neutrophils. Serology was negative. Nonprotein nitrogen 46.3 mg. per 100 cc., creatinine 1.7 mg. per 100 cc. Serum electrolytes were as follows: Sodium 141 mEq. per liter, potassium 5.7 mEq., chloride 112 mEq., carbon dioxide 27.0 mEq./L. Sedimentation rate was 3 mm. in 1 hour, and the hematocrit 41.5 cc.

In the hospital, the patient received 0.45 mg. of
digitoxin in three divided doses on the first hospital day and thereafter received 0.05 mg. daily. After four days, the child weighed 43 pounds (a 10 pound weight loss) and maintained this weight thereafter. The tachypnea and tachycardia disappeared and the liver-edge receded to a position 2 cm. below the right costal margin.

On Dec. 3, 1953, cardiac catheterization was performed without difficulty. The oxygen saturation values obtained were as follows: Superior vena cava 88 per cent, inferior vena cava 75.5 per cent, right atrium 94.1 per cent, right ventricle 94.1 per cent, right pulmonary artery 94.1 per cent, left pulmonary artery 95.4 per cent. The oxygen saturation in the femoral artery was 96.7 per cent. The mean wedge pressure was 11 mm. Hg, the pulmonary artery pressure was 42/20, the right ventricular pressure was 44/10, and the mean right atrial pressure was 5 mm. Hg.

On Dec. 10, 1953, surgical treatment was attempted with the patient in a state of hypothermia. The patient was given 25 mg. of Benadryl and 1/200 Gm. scopolamine, preoperatively. Anectine (30 mg.) was given intravenously and an endotracheal tube was inserted. At 8:30 a.m. ice was applied; the patient’s temperature at this time was 37 C. At 9:15 a.m. his temperature having dropped was 30 C. and the ice was removed. At this time cardiac arrest occurred. Thoracotomy was accomplished rapidly and cardiac massage was started. The patient’s body temperature continued to fall and at 10:10 a.m. was 24 C. This was the lowest recorded reading. At 9:45 a.m. a heating pad was applied to the body. An electrocardiogram made at 9:30 a.m. showed ventricular fibrillation. Potassium chloride and later Pronestyl were given into the chamber of the left ventricle without benefit. The electrical defibrillator was used periodically without success. Calcium chloride was later given intracardially, but no improvement in the patient’s condition occurred. After cardiac massage had been carried out for a period in excess of two hours without restotmation of cardiac action, manipulation was discontinued.

Dr. Mahlon Delp, Moderator: Mr. Tucker, will you present your interpretation of the x-ray films?

Mr. Tucker, Fourth Year Student: The plates showed a large heart with right hypertrophy, particularly obvious on lateral view, and a prominent pulmonary outflow tract.

Dr. Delp: Dr. Tice, do you have any additional comments? Perhaps your fluoroscopic findings would be pertinent.

Dr. Galen Tice, Professor of Radiology: Fluoroscopy showed forceful pulsation in the hilar areas on both sides. The combination of a large right heart and forceful hilar pulsation would make me think first of interatrial septal defect. One might, of course, consider patent ductus arteriosus and mitral valvular disease. The latter could be seriously considered if there were displacement of the esophagus. We cannot ignore the excessive hilar activity noted in this patient.

Dr. Delp: Mr. Van Antwerp, may we have your interpretation of the electrocardiogram as well as the catheter pressure readings?

Mr. Van Antwerp, Fourth Year Student: The electrocardiogram showed a heart rate of approximately 125 beats per minute with regular rhythm. The rhythm arose in the sinus node and conduction between atria and ventricles was prolonged (0.2 second). The P waves were large and peaked, suggesting atrial hypertrophy. The QRS complexes were widened (0.12 second) with a delay in depolarization over the right ventricle. I consider this record indicated right ventricular hypertrophy and atrial hypertrophy.

The pressures were recorded by catheterization in the pulmonary capillary (wedge), left and right pulmonary arteries and right ventricle. The following positive interpretations can be made: (1) No pressure gradient existed between the pulmonary artery and the right ventricle; therefore no pulmonary stenosis was present. (2) Slight pulmonary hypertension was present. (3) The “wedge” pressure was not abnormal. (4) The diastolic pressure was elevated in the right ventricle, a finding compatible with right heart failure.

Dr. Delp: Dr. Dimond, do you have any additional comments about these tracings?

Dr. E. Grey Dimond, Professor of Medicine: No.

Dr. Delp: May I suggest these features of the case in a brief summary. This patient had first been recognized at birth as having heart disease. The clearly described murmurs and especially the diastolic murmur deserve careful consideration. Additional information which may be gathered from the electrocardiogram and pressure readings I leave to you. Hypothermia was utilized in preparation for surgery here but this we wish discussed after the differential diagnosis.

Mr. Miller, we want you to discuss the
Differential diagnosis based upon the history, physical findings, and any additional information you gather from pertinent laboratory values. Dr. Dimond will continue the diagnostic discussion. We wish Dr. Reissmann to confine his remarks to hypothermia, its indications, how it is done, and the dangers involved. Dr. Hardin will present his proposed plan of surgery in this case. First, Mr. Miller.

Differential Diagnosis

Mr. Miller, Fourth Year Student: The case today is that of a seven year-old boy who, at birth, was found to have an enlarged heart. This finding raises the possibility of congenital heart disease.

The subsequent developments of dyspnea on exertion, frequent upper respiratory infection and three episodes of illness diagnosed as pneumonia serve only to strengthen this suspicion. The possibility of rheumatic heart disease cannot be ruled out by history, but subsequent developments would tend to point towards congenital heart disease as this patient's main problem. The lack of development of cyanosis through these various infections, plus the clinical picture on admission establish that this child had acyanotic or, at most, potentially cyanotic heart disease.

Displacement of the apical beat to the left mid-axillary line plus x-ray and electrocardiographic findings indicate right atrial and ventricular enlargement. Tachycardia, tachypnea, venous distention, hepatomegaly and cardiomegaly, plus the subsidence of these findings on digitoxin therapy, indicate that this patient was in right heart failure.

The auscultatory finding of systolic murmur actually does not localize the lesion; however, the existence of increased pulmonic second sound, plus fluoroscopic evidence of increased bronchovascular markings and prominent pulmonary conus indicate the existence of pulmonary hypertension.

Thus far, I am able to limit this lesion to one which may cause right heart enlargement and failure without producing these findings on the left side. Common lesions which could give the picture presented are: (1) patent ductus arteriosus, (2) pulmonary stenosis, (3) interventricular septal defect, (4) interatrial septal defect, and (5) Lutembacher's syndrome.

Cardiac catheterization was utilized in this patient, and normal oxygen saturation values were obtained in the superior and inferior vena cava. However, on entering the right atrium, there was found a substantial increase in this value. Thus, in this compartment, there must be mixing of arterial blood with that from the venae cavae. The most common condition which may produce this finding is atrial septal defect.

Another condition which may cause increased oxygen saturation values is anomalous pulmonary venous drainage into the right atrium. This condition can give the exact findings of atrial septal defect, but can only be proved by passing the catheter into this vein.

Other exceedingly rare conditions which are all eliminated as possibilities because of physical and catheterization findings are: (1) persistent atroventricular canal, (2) atroventricular fistula between a coronary artery and coronary sinus, (3) tricuspid insufficiency with interventricular septal defect and (4) patent ductus with pulmonary and tricuspid insufficiency.

Atrial septal defect does cause pulmonary hypertension, right heart enlargement, right heart failure, and can give all of the clinical as well as the catheterization findings. Thus, I feel that this patient did have interatrial septal defect.

This condition is notorious for having associated with it other lesions, congenital or acquired. Abbott describes 373 cases of interatrial septal defect, and out of this group, only 73 were uncomplicated. Interventricular septal defect may accompany this lesion, but, in the case being considered here, there was no increase in oxygen saturation value in the right ventricle over that of the right atrium. The existence of pulmonary stenosis is also disproved in this case due to the finding of pulmonary hypertension. While there is an increased oxygen saturation of 1.3 per cent in the left pulmonary artery over that of the right ventricle, which would bring up the possibility of a patent ductus, I believe this difference is probably due to laboratory error. The lack of
NONCYANOTIC HEART DISEASE WITH HEART FAILURE 127

typical machinery-like murmur and widened systemic pulse pressure also makes this unlikely.

Mitral stenosis may accompany an interatrial septal defect, this condition being called Lutembacher's syndrome. By history, this patient had an enlarged heart at birth, a finding uncommon in uncomplicated atrial septal defect. With Lutembacher's syndrome cardiac enlargement is the usual finding. The murmur of mitral stenosis although commonly present, has not been observed in a large number of these cases; in the case under discussion it was not heard. Left atrial enlargement is not always present in this condition, and again in this case evidence of such enlargement was absent. There is one finding which does not agree with this diagnosis, and this is the lack of evidence of a greatly enlarged pulmonary artery; however, the roentgenographic findings indicated some enlargement of that artery, and I feel that the diagnosis of Lutembacher's syndrome best explains this patient's illness.

Dr. E. Grey Dimond, Professor of Medicine: Our preoperative diagnosis was interatrial septal defect with or without anomalous pulmonary veins. We arrived at this diagnosis in the following manner:

From history alone we could classify the problem as one of congenital heart disease, acyanotic type. The very loud pulmonary second sound, the diastolic blowing murmur of pulmonary insufficiency, the heavy motile pulmonary artery shadows by fluoroscopy, the electrocardiographic evidence of atrial and right ventricular hypertrophy, all were points indicating this to be a lesion overloading the right heart and pulmonary circulation. This narrowed the field to atrial septal defect, anomalous pulmonary veins, Lutembacher's syndrome, perhaps patent ductus arteriosus or ventricular septal defect.

Realizing that a patent ductus but rarely may be present without the typical murmur, we felt justified in eliminating this as a possibility.

We could not eliminate ventricular septal defect completely without catheterization, although the usual course of a patient with a ventricular septal defect is certainly not one of right heart failure at the age of seven. We have, in the past, seen a two year-old boy with similar findings in whom a large, high, ventricular septal defect was found. However, this is rare and we rather confidently disregarded that possibility in the present case.

Lutembacher's syndrome (interatrial septal defect and mitral stenosis) could not be completely discounted, but we were unable to hear any apical diastolic murmur. Actually, even if we had heard an apical diastolic murmur, I believe I would have simply assumed that the murmur was due to altered flow through the left heart and not due to a mitral stenosis. Repeatedly, our group as well as others, has made the diagnosis of Lutembacher's syndrome, based on the usual findings of atrial septal defect plus an apical diastolic murmur, only to be embarrassed to find a normal mitral valve at surgery or autopsy.

We have previously seen a young child with actively pulsating pulmonary vessels, right ventricular hypertrophy, and right heart failure, who was found at autopsy not to have congenital heart disease, but severe, primary pulmonary endarteritis. The right heart catheterization was done in the present patient primarily to eliminate this condition.

The data obtained at catheterization effectively proved a left to right shunt at the atrial level. We were not successful in threading the catheter into an anomalous pulmonary vein, so we sent the child to surgery with the diagnosis of interatrial septal defect with or without anomalous pulmonary veins.

Although seven is a somewhat older age for subendocardial fibroelastosis, we have seen heart failure in a child of nine with this diagnosis and, therefore, further qualified our preoperative diagnosis by adding to the above: with or without subendocardial fibroelastosis.

Dr. Kurt Reissmann, Associate Professor of Medicine: If one wants to open the right heart in order to repair a septal defect, one has to block the blood flow through the chambers of the heart by occluding the venous inflow. Two methods have been proposed to achieve such a bloodless heart and yet to prevent anoxic tissue damage:

(1) An extra corporeal heart-lung device
that permits the by-pass of the patient’s heart and lungs and maintains systemic flow. This can be realized either as a mechanical pump and oxygenator or as a human donor whose circulation is connected with that of the patient.

(2) Hypothermia. The application of hypothermia is based upon the reduction of tissue metabolism and oxygen requirements at lowered body temperature. At a body temperature of 27 C, for example, one can completely occlude the cerebral circulation in a dog for 15 minutes and no demonstrable brain damage will result.

Hypothermia is induced by placing the anesthetised patient in crushed ice or cold water, or by means of a special blanket through which a cooling solution circulates. Anesthesia should be sufficiently deep to prevent shivering which is accompanied by an increase in oxygen consumption and delays the lowering of the body temperature. The desired temperature depends on the type of surgery to be performed. In the case of an open repair of a septal defect, a body temperature of 28 C is said to give the surgeon up to 10 minutes time to perform his task. The complicating and limiting factor in the application of hypothermia in cardiac surgery is the behaviour of the heart and its tendency to undergo ventricular fibrillation or cardiac arrest.

Our group has studied recently close to 100 dogs in hypothermia, and we were impressed by the unpredictability of the temperature level at which cardiac complications occurred. Some animals fibrillated at 26 C.; others, under the same conditions, tolerated 18 C. The tendency to fibrillate was greatly enhanced when the heart was manipulated. In our experience, electrical defibrillation was much less successful in the hypothermic animal than in the normothermic, which makes fibrillation in hypothermia a particularly serious complication.

The failure of the heart in hypothermia is probably not a simple matter of hypoxia or myocardial ischemia, because no oxygen debt could be demonstrated by measurement of the coronary arteriovenous oxygen difference and lactate or pyruvate level. The energy yielding enzymatic processes are slowed in hypothermia, but they are still adequate for tissues upon which no demands for work are placed during the cold state. The heart, however, continues to work. Even the idling heart, that is, the beating heart that ejects no blood, has a considerable energy expenditure. The characteristic lengthening of the active phase, which at 25 C. occupies as much as 70 per cent of the cardiac cycle, indicates that in the cold state, the energy for systolic contraction is not available instantaneously but only within a prolonged period of time. When the heart’s temperature is lowered further, the energy yielding reactions are slowed to such an extent that sufficient energy is no longer available for a coordinated cardiac contraction and the heart will fail in spite of adequate blood and oxygen supply.

From the foregoing, it is understandable that, so far, all attempts to prevent cardiac arrhythmia in hypothermia by means of drugs or various electrolyte solutions have met with little success. In the patient under discussion, ventricular fibrillation occurred at the relative high body temperature of 29 C. I think it likely that the considerable dilatation and hypertrophy of the heart in this case contributed to the early occurrence of fibrillation.

Dr. Creighton Hardin, Assistant Professor of Surgery: Although this discussion is limited to our plan of repair of an atrial septal defect under hypothermia, I think brief mention should be made of methods used of which has led to a successful repair of atrial defect. One may divide them into indirect or relatively blind procedures and the direct, dry-field intracardiac methods.

Gross has utilized a rubber or plastic “well” sutured to the side of the dilated right atrium. An incision is made within the portion of the atrium contained within the core of the “well.” Cohn, Murray, and Bailey use a method of repair of atrioseptectomy. The atrial wall is invaginated and sutured to the atrial septum.

The surgeon is desirous of having an open, dry field in which to work so that any septal repair may be done under direct vision. The pump-oxygenator of Gibbon and the use of
hypothermia are current methods by which this latter procedure may be undertaken.

In using hypothermia the surgeon is relying on general body cooling of the vital organ systems to prevent irreversible anoxia. Occlusion of the venae cavae provides a dry field for the surgeon. Even so the surgeon works against the clock but he has the advantage of visualizing directly the operative field.

The repair of the actual defect, if small and centrally located, can be readily done by simple direct suture. Larger defects precluding the above repair necessitate the use of homologous or autogenous tissue or a plastic prosthesis, atrial appendage, either a free or pedicle graft, vein, or pericardium which will heal over in about 10 weeks. Polyethylene, nylon, polyvinyl sponge are rapidly endothelialized by the sixth week.

The surgical advantages of open cardiac surgery for complete visualization, accurate placement of the sutures and placement of tissue or plastic for the larger defects are obvious.

Contraindications for the surgical repair of atrial septal defects are high pulmonary vascular resistance and beginning reversal of blood flow through the septal defect.

There are many problems of hypothermia which require further study. They have been presented by Dr. Reissmann. Until certain of these problems have been satisfactorily answered through further laboratory studies now in progress, we are not using hypothermia again clinically.

We should mention, however, that successful repairs of atrial septal defects have been reported by Lewis and Taufic, and by Swan and his co-workers.

Dr. Delp: Dr. Skelton will give the pathologist’s findings.

Pathology Report

Dr. Floyd R. Skelton, Instructor in Pathology: The role of the pathologist in the discussion of such a case as we have today seems rather straightforward. Consequently, the following report will be presented in three sections: First, the autopsy findings: second, a description of the embryologic development of the cardiac malformation; and third, a brief clinical-pathologic correlation.

At autopsy the body was that of a slightly undernourished but fairly well developed 7 year old boy with no evident cyanosis, but with a distinct pigeon-breast deformity. Since the gross examination of the organs showed that the significant lesions were confined essentially to the heart, this organ will be considered in some detail. A large interatrial septal defect was found measuring 6 by 4 cm. and located in inferior posterior portion of the septum, in such a manner that the attached margins of the septal leaflets of the mitral and tricuspid valves were approximated to form the base of the septal defect. The foramen ovale was present above the defect and, while anatomically open, was functionally closed. The great dilatation and hypertrophy of the wall of the right atrium was readily apparent and contrasted sharply with the small size and thinness of the wall of the left atrium. The pulmonary veins entered the left atrium in a normal fashion. As is usual in cases such as this, the left ventricle was small relative to the right. The leaflets of the mitral valve formed the base of the large interatrial septal defect, and careful examination of this valve failed to reveal significant morphologic changes. The valvular ring measured 8.2 cm. in circumference. The hypertrophy and dilatation of the right ventricle was also striking and was associated with dilatation of the pulmonary ring as evidenced by a circumference of 7.8 cm. In contrast, the circumference of the aortic valve was 5.2 cm. It is of interest that the pulmonary valve cusps measured 2 cm. in width, while the aortic valve cusps measured only 1.2 cm. The tricuspid valve was normal in appearance and measured 13 cm., in circumference. The ascending thoracic aorta was small, measuring 8.3 cm. in circumference above the pulmonary valve. This dilatation extended into the main branches so that the right main pulmonary artery measured 4.7 cm. in circumference and the left main pulmonary artery measured 2.7 cm. in circumference. The only other gross observation of interest was the evident hemosiderosis of the lungs and the manner in which the small branches of the
pulmonary arteries projected above the cut surface in the peripheral regions of the lungs.

An explanation for the projection of pulmonary artery radicles above the cut surface of the lung was readily apparent on microscopic examination. The walls of the arteries and arterioles were considerably thicker than normal, apparently as a result of medial muscular hypertrophy since the media showed the most marked thickening. This medial hypertrophy extended into even the smallest arterioles of the pulmonary vascular tree. When sections of pulmonary tissue were subjected to Gomori aldehyde-fuchsins stain which imparts to the elastic tissue a deep purple color, the internal elastic lamella in the pulmonary arteries and arterioles was observed to be thickened and reduplicated.

Sections of pulmonary tissue stained with hematoxylin and eosin disclosed alveoli containing many large pigment-laden macrophages. That this pigment was hemosiderin was indicated by its blue color in Perls' stain. The presence of these cells substantiated the gross morphologic and the clinical evidence of congestive cardiac failure. The bronchi contained amorphous material, and were lined by tall, columnar, mucous-containing cells resting on a greatly thickened basement membrane. The origin of these latter changes is somewhat obscure, but may be related to the repeated bouts of upper respiratory infection and pneumonia which this patient experienced over the few years of his life.

Of course, as a consequence of the protracted period of cardiac massage, some evidence of trauma would be expected in the heart and this was present as focal areas of hemorrhage in the epicardium and in fibrous connective tissue within the myocardium.

Anatomic Diagnosis

Primary. Congenital malformation of the heart consisting of an extensive defect of the interatrial septum.

Hypertrophy and dilatation of the right atrium and ventricle with dilatation of the pulmonary artery, advanced; weight of heart and lungs 770 Gm., normal 350 Gm. Normal mitral valve.

Medial hypertrophy of small and medium sized radicles of the pulmonary artery in both lungs.

Chronic passive congestion of the lungs, moderate.

Multiple subepicardial and myocardial hemorrhages (history of cardiac arrest and massage for two hours before death).

Focal atelectasis and hemorrhages in all lobes of both lungs.

Acute passive congestion of the liver, adrenals, and kidneys, slight.

It will be recalled that it is within the first few weeks of fetal life that the primitive two-chambered heart is transformed into a four-chambered organ. This is accomplished by the downward and forward growth of the septum primum from the roof of the single atrium toward the atrioventricular canal and the upward growth of the interventricular septum from the lower portion of the single ventricle. In the region of the atrioventricular canal, endocardial cushions develop, which normally unite with the septum primum and the interventricular septum, thus dividing the single atrium and ventricle into right and left chambers. The ostium primum, which is between the lower edge of the septum primum and the endocardial cushions, normally is obliterated when the septum primum and endocardial cushions fuse. Occasionally fusion does not occur. The resultant persistent ostium primum gives rise to some of the largest interatrial septal defects, such as that in the present case.

A persistent ostium primum has no effect on the heart during fetal development when the right atrial pressure is greater than the left, and when the flow of blood is normally from the right to the left atrium. However, following birth, the development of greater pressure in the left atrium than in the right leads to reversal of this flow so that the blood passes from the left to the right atrium. Coincident with the reversed flow of blood, there is greatly increased pressure within the chambers of the right heart and within the pulmonary system, so that hypertrophy and dilatation of both the right atrium and ventricle and enlargement of the pulmonary artery occur. If the
dilatation of this artery is sufficiently great, actual insufficiency of the pulmonary valve may ensue. In the pulmonary artery radicles within the parenchyma of the lungs, the medial hypertrophy and the thickening and reduplication of the internal elastic lamella is a reaction to this increased pressure in the pulmonary circulation. Of course, the blood passing from the left into the right atrium is oxygenated and, in consequence, the oxygen tension of the blood present within the right atrium, right ventricle, and pulmonary arteries is abnormally high. All of these consequences of the interatrial septal defect were manifest in the clinical examination of this patient.

Despite the increased flow of blood through the pulmonary circulation, and consequent increased return to the left atrium, this chamber does not hypertrophy because the blood readily passes into the right atrium through the large interatrial septal defect. It has been suggested\(^1\) that the passage of blood from the left to the right atrium is not entirely due to the increased return of blood to the former chamber but that it is augmented by the fact that this chamber has a greater hydrostatic pressure by virtue of its anatomic location above the level of the right atrium.

The relatively small size of the left ventricle is commensurate with the decreased volume of blood which it is called upon to handle. The decreased output of the left ventricle, which eventuates in oxygen starvation of the systemic circulation, explains the usual underdevelopment of patients with this cardiac malformation. The decreased output is also reflected in a low systemic blood pressure and in a narrow pulse pressure, both of which were present in this case.

One is tempted to explain the murmurs in this case entirely on the basis of the cardiac malformations. Taussig\(^2\) relates the systolic murmur and thrill either to the interatrial septal defect or to the eddy currents set up in the greatly dilated pulmonary arteries. The diastolic murmur in this patient may well have been the result of dilatation and insufficiency of the pulmonary ring, although little evidence of valvular insufficiency was visible on gross morphologic examination.

The accentuated second pulmonic sound can be explained on the basis of increased pressure within the pulmonary circulation.

No morphologic findings relative to the period of hypothermia were found, not a surprising circumstance.

REFERENCES

