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INTERVENTRICULAR SEPTAL DEFECT ASSOCIATED WITH ANEURYSMAL DILATATION OF THE AORTIC SINUSES AND AORTIC SINUS-RIGHT VENTRICULAR FISTULA. A CASE TREATED SURGICALLY

HUAN-CHEN CHENG, M. D.

St. Francis Hospital and Sanatorium Roslyn, New York U. S. A

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Though many examples of aortic sinus aneurysms have been described in literature, it is a rare disease witnessed today. The ruptured or unruptured aortic sinus aneurysm associated with interventricular septal defect is especially rare, only a few cases having been reported. The congenital aortic sinus aneurysm associated with interventricular septal defect was thought to be due to the failure of fusion of the proximal bulbar swelling with the upper border of the ventricular septum in the embryo.[1]

The case in this report was diagnosed as congenital heart disease, interventricular septal defect and aortic sinus aneurysm pre-operatively. During an open heart corrective procedure, the patient was found to have interventricular septal defect associated with aneurysmal dilatation of the aortic sinus and aortic sinus-right ventricular fistula. Closure of the interventricular septal defect and aortic sinus-right ventricular septal fistula was performed by utilization of extracorporeal circulation. The patient recovered well.

CASE REPORT

The patient was a six year old male child who was first seen in our consultation clinic on July 9, 1959, with the chief complaints of cardiac murmur, general weakness, shortness of breath and occasional chest pain for a duration of three years. He was then admitted into our hospital on October, 19, 1959 for cardiac diagnostic study and possible corrective cardiac surgery. His mother stated that he was born at full term by a normal spontaneous delivery. He had no significant post natal difficulties. He was noticed to have a cardiac murmur at birth which was attributed to congenital malformation of the heart, but he had been fairly well until three years of age, when he developed frequent episodes of tonsillitis and upper respiratory infection. He also has been complaining of shortness of breath, easy fatigability and occasional precordial pains since then. He appears thinner than his siblings, but there has been no fainting, squatting or visible cyanosis. There is also a history of a car accident in March, 1959, but there was no sudden onset of shortness of breath or chest pain immediately following the accident. The

family history was non-contributory. The mother had no sickness during her pregnancy.

Physical examination revealed a well developed and fairly nourished negro child in no acute distress. The skin appeared to be normal and he was not anemic or cyanotic. The pupils were round, equal and reacted to light normally. Blood pressure was 105/65mm. of Hg. The pulse was regular with a rate of 85 per minute. The respirations were regular with a rate of 24 per minute. Except for hypertrophic tonsils, the head, ears, nose, eyes, and throat were essentially negative. The chest was symmetrical with normal expansion. The lungs were clear on percussion and auscultation anteriorly and posteriorly. The heart revealed that the point of maximal impulse was in the fifth intercostal space just outside of the mid-clavicular line. A systolic thrill was palpated in the third and fourth intercostal space at the left sternal border. On percussion the heart was found to be enlarged bilaterally. On auscultation a Grade III to IV systolic murmur was best heard in the third and fourth intercostal space at the left sternal border. There was also a Grade I diastolic murmur heard in the second intercostal space at the left sternal border. (It should be noted that the presence of the diastolic murmur was questioned by some of our investigators.) The second pulmonic sound was increased. The abdomen and the extremities were essentially negative.

The laboratory data were as follows: Erythrocytes 3.9 mil./mm; hemoglobin 12.3 gms./100 ml.; hematorit 36%; leucocytes 7150 with a normal differential count; Sed rate 8mm per hour; clotting time 3′; bleeding time 1′45″; urinalysis normal. The X-ray examination of the chest showed that the heart was enlarged. All chambers were involved though it was felt that the right side was predominately enlarged. The pulmonary artery was quite prominent. The pulmonary vascular markings were increased (F. 1A). Angiocardiography revealed that there was enlarg-

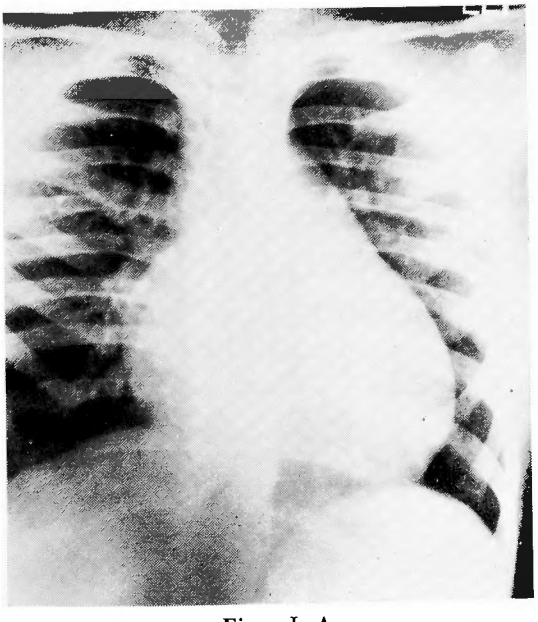

Fig I A

ment of the right ventricle, pulmonary artery and of the left heart. There was re-opacification of the pulmonary system at the time of left heart filling. In addition, there was aneurysmal dilatation of the aortic sinus with dilatation of the ascending aorta (F. 2). The electrocardiography showed that there was no axis

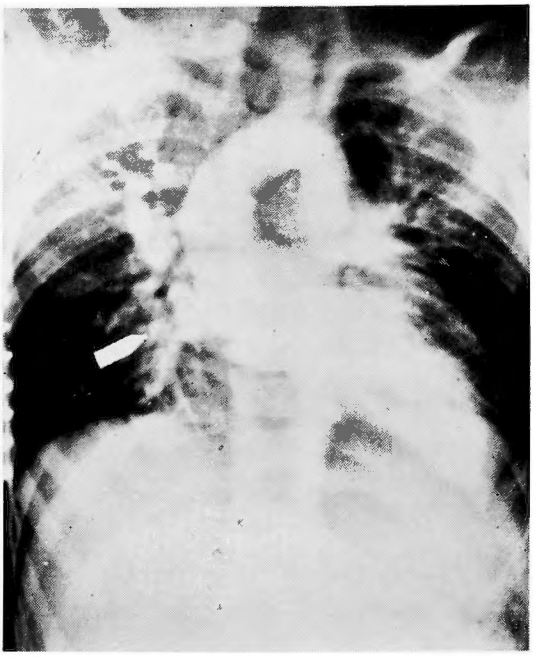

Fig. Ⅱ

deviation and the heart was in semi-vertical position. There was RSR pattern in V3R and V4R. The T waves over the left precordium were peaked and the R waves showed high amplitude (F. 3A). The right heart catheterization revealed that there was hypertension in the right ventricle and the pulmonary artery system

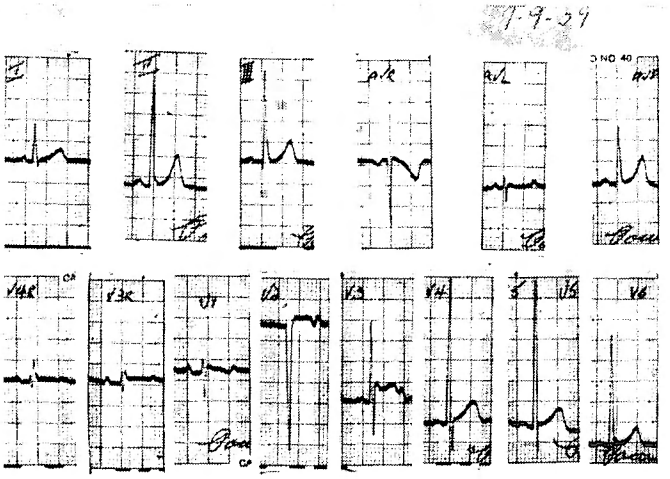

FIG. ⅢA

Table 1 Results of Right Cardiac Catheterization

Site	Blood Oxygen Saturation %	Blood Pressure in mm. of Hg.
Brachial artery	99.4	118/78
Superior vena cava	79.0	
Right atrium	73.5	3.1 (mean)
Right ventricle	84.0	52/1.5
Pulmonary artery	81.0	45/20.5 (30.0mean)
Pulmonary capillary		9.5 (mean)

with a normal pulmonary capillary pressure. There was no evidence of cardiac failure. There was a significant step-up of blood oxygen saturation in the right ventricle. The systemic blood oxygen saturation was normal (T. 1).

From the above findings a conclusion was reached that the patient had interventricular septal defect with a left to right shunt and aortic sinus aneurysm. Open heart corrective surgery was performed on November 10, 1959 by utilization of extracorporeal circulation. The operative findings and the procedure were as follows.

The heart was approached via a trans-sternal fourth intercostal space bipleural incision. The right heart was found to be markedly enlarged. The left ventricle was also enlarged. A Grade II thrill was palpable over the uppermost aspect of the right ventricle, where a slight bulge was detected. The right coronary artery was located considerably more to the left and higher up across the ventricle than usual. This necessitated a smaller incision into the right ventricular cavity and consequently the exposure was not as good as usual.

With the patient on the pump oxygenator at a flow rate of 90cc/kg./min., a right ventriculotomy was performed. The lesions found consisted of an aortic sinus fistula about 8-10mm. in diameter and a separated high interventricular septal defect about 12-14 mm. in diameter. These defects were closed with interrutped #000 atraumatic black silk sutures. These lesions were difficult to correct because of their location, the amount of blood in the field, and the small incision in the ventricle. Closure of these lesions required approximately 46 minutes on the pump oxygenator. During this interval the patient had rather marked tachycardia. The ventriculotomy wound was then closed with interrupted #00 and continous #00 atraumatic black silk sutures. The thrill that was previously present had completely disappeared and a diminution in the size of the right ventricle occurred. The pericardium was then partially closed in the usual manner.

The immediate post-operative course was very smooth and the operative wounds healed well. The sutures were removed on the eighth post-operative day. On the tenth post-operative day, the patient's temperature rose to 101.5°F. then continuously rising to between 102°F and 103°F. in the next five days. During this period, every effort was made to determine the cause of the temperature elevation. Finally the sternal portion of the transverse incision of the chest was suspected of being infected. An exploration was then made and osteomyelitis of the sternum was

discovered. The infection was treated with antibiotics and the patient recovered well and was discharged on December 15, 1959 in apparently good condition.

Residual cardiac murmurs were found after the removal of the wound dressing on the eighth post-operative day. The murmurs were Grade I systolic murmur, best heard in the third and fourth left intercostal space, and a faint diastolic murmur heard in the second left intercostal space. These murmurs persisted until the patient was discharged. During the post-operative period of hospitalization, chest X-rays and electrocardiography were repeated several times, but were not remarkable.

The patient was seen again on April 8, 1960 in our consultation clinic for post-operative follow-up. On physical examination, his general condition was found to be good, not cyanotic or anemic. Blood pressure was 110/75, pulse was 95, regular, and the respirations were 24 per minute. The lungs were clear and the breath sounds were good. The systolic thrill, which was palpable in the third and fourth intercostal space pre-operatively, was no longer present. The second pulmonic sound was found to be only slightly accentuated. However, the diastolic cardiac murmur in the second left intercostal space was found to be increased in intensity and became continous at times. The abdomen and extremities wer eessentially negative. There were no signs of cardiac failure or any other abnormality. On this visit, the patient's mother stated that the patient's appetite has been fairly good and he has been fairly active in the past two or three months. He still occasionally complained of weariness, but there was no chest pain or significant shortness of breath. The patient's mother also recognized that there was improvement in his activity and strength when compared with the pre-operative period. Chest X-rays were taken on this visit and revealed that the heart size was proportoinately smaller and the pulmonary vasculature was diminished (F. 1 B). The electrocardiography was also repeated and suggested an improvement in the left ventricular hypertrophy diastolic overload pattern (F. 3B).

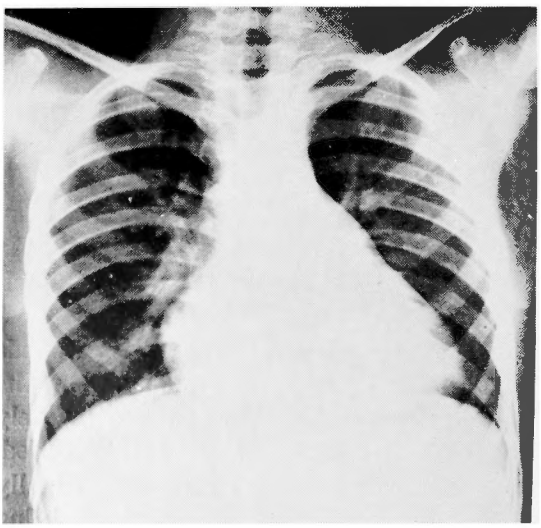

Fig. B I

In comparision of the pre and post-operative findings, we have found that the patient's general condition has improved. The cardiac systolic thrill has disappeared. The second pulmonic sound has diminished. The chest X-rays showed that the pulmonary hypervascularity has diminished to normal appearance. The electrocardiography suggested some improvement in the left ventricular hypertrophy diastolic over-

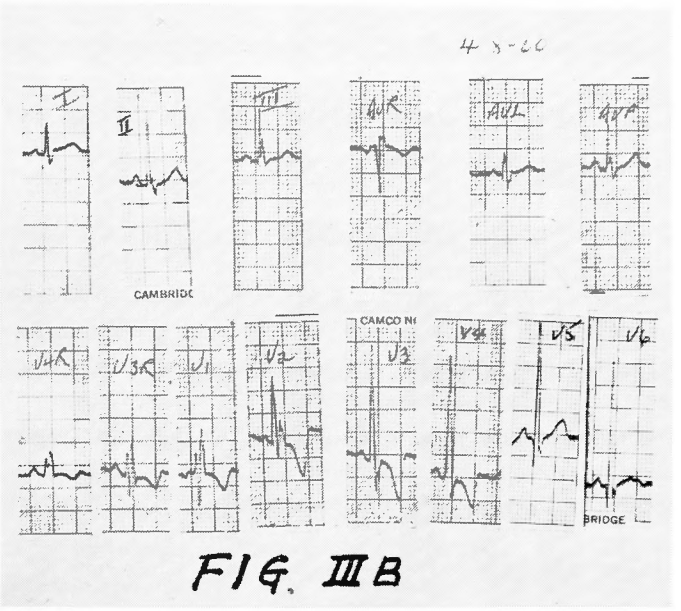

FIG. III B

load pattern. However, the diastolic murmur has increased in intensity and becomes continuous at times. A repeat cardiac catheterization is planned in the near future for further evaluation.

DISCUSSON

1. Jones and Langley have collected forty-seven cases of aortic sinus aneurysm and their conclusion was that patients with unruptured aortic sinus aneurysm were free from symptoms [2]. Steinberg has pointed out that there are no diagnostic features for the recognition of unruptured aortic sinus aneurysm except for a Grade II to III cardiac murmur. Accordingly, a high index of suspicion and angiocardiography are advocated in order to establish the diagnosis during life [3]. In rupture of silent aortic sinus aneurysm described by Oram and East [4], Venning [5], Falholt and his associates [6], at the time of rupture, the patient was usually seized with severe pain in the chest or upper abdomen associated with shortness of breath, general weakness, and often with vomiting. The study of 4,000 cases of aneurysms of the thoracic aorta by Boyd [7], indicated that acquired aortic sinus aneurysm most frequently ruptured into the pericardial sac ; the congenital aortic sinus aneurysm usually ruptured into the right side of the heart. In 1946, Herson and Symons [8] reported one case of ruptured aortic sinus aneurysm associated with interventricular septal defect, and one more was added by Burchell and Edwards [9] in 1951. But both of these cases died of cardiac failure and these complicated lesions were found at autopsy. Until recently in persons suffering from ruptured aneurysm of the sinus of Valsalva, death usually resulted from congestive heart failure or sub-acute bacterial endocarditis. Since extracorporeal circulation was utilized, there were a few cases of rurtured aortic sinus aneurysm treated surgically

by Lillehei [10] and Allison [11].

Rupture of aortic sinus aneurysm can occur at any time during life. It is usually provoked by physical strain. In this case, the patient's family history was non-contributory and the past history revealed that the patient had a cardiac murmur since birth and had been asymptomatic until the age of three years. There was no sudden onset of chest pain or severe shortness of breath and the fistula was found to be of long duration on the operating table. Therefore, it was thought that the fistula was probably congenital in origin. The asymptomatic period, the first three years of his life, was explained on the basis that at that period he was too young to complain. In addition, his activity was rather limited because of his age. Of course, the accident in March, 1959 might be contributory to the rupture of the aortic sinus aneurysm, but there was no exaggeration of symptoms immediately after the accident occurred. As far as his symptoms are concerned, we believe that either one of these complicated lesions could cause the symptoms he had pre-operatively. Post-operatively, especially in the past three months, he has been very active. The chest pain has disappeared and physical exertional tolerance has been much improved.

2. Edwards and his associates [12] described a case of congenital aortic sinus aneurysm which ruptured into the right ventricle. The patient had a continuous murmur maximum in the second left intercostal space with a systemic blood pressure of 180/0-60mm of Hg.. Herson and Symons [8] also found a high pulse pressure and a continuous murmur in their patient who had an aortic sinus-right auricular fistula. In the report of Brown and his associates [11], in a patient with an aortic sinus-right ventricular fistula, a loud precordial systolic murmur was best heard in the fourth left intercostal space at the parasternal line, which was widely conducted throughout the chest; and an early, high-pitched diastolic murmur, maximum in the fourth left intercostal space. At times the murmur appeared to be continuous in the fourth left intercostal space, and the systemic blood pressure was 140/0mm of Hg.. But Macleod only found systolic and diastolic murmurs in a patient with cardioaortic fistula opening into the right auricle [13].

In our patient, the systolic thrill in the fourth left intercostal space which we believe was caused by the ventricular septal defect, and the Grade III to IV systolic murmur in the third and fourth left intercostal space was from the same source. The Grade I diastolic, blowing murmur in the second intercostal space at the left sternal border was thought to be caused by the aneurysmal dilatation of the aortic sinus. Even though the angiocardiography revealed peculiar re-opacification inferior and somewhat anterior to the sinus of Valsalva of the aorta (F. 2), the diagnosis of cardioaortic fistula could not be made since the patient's systemic bloop pressure was 105/60mm of Hg. on physical examination and was 118/78mm of Hg. on cardiac catheterization at the same time that the right ventricular diastolic pressure was normal (T. 1).

Post-operatively, the systolic thrill in the third and fourth left intercostal space had disappeared and the systolic murmur at the same area had markedly diminis-

hed. However, the diastolic murmur which was located in the second intercostal space at the left sternal border became a continuous one and the intensity has increased. This we believe was caused either by the aneurysmal dilatation of the aortic sinus or a small residual fistula, since the wall of the aneurysm and the rim of the fistula were very thin and it is quite possible to have the closed fistula partially reopened. On his last visit to our clinic in April, 1960, his general condition was good. The blood pressure, respirations and pulse were within the normal limits. The chest was clear. The cardiac findings were the same as described above.

3. The pre-operative electrocardiography revealed RSR pattern in V3R and V4R. The T-waves over the left precordium were peaked and the R-waves showed high amplitude. Bi-ventricular hypertrophy with left ventricular diastolic overload pattern was suggested as is often noted in left to right ventricular shunts. Either one of the patient's combined lesions could give these findings. The post-operative electrocardiography revealed right ventricular hypertrophy and prolonged interventricular conduction. This is in part due to conduction and myocardial changes associated with the operative procedure. The amplitude of the T and the R-waves over the left precordium has decreased, suggesting an improvement in the left ventricular hypertrophy diastolic overload pattern (F. 3A, B).

The pre-operative chest x-rays showed cardiac enlargement, the pulmonary vascular markings were increased, and the pulmonary segment was prominent. These findings suggested an intracardiac left to right shunt, but there was no localized swelling to indicate an aneurysm. The post-operative chest X-rays revealed that the pulmonary vascular markings were reduced to near normal and the size of the heart was proportionately smaller (F. 1B).

On angiocardiographic examination, five minutes after the dye injection there was opacification of the left atrium and the left ventricle. These chambers were both enlarged. There was dilatation of the ascending aorta and also marked dilatation of the sinus of Valsalva of the aorta. At the time of left heart opacification, there was re-opacification of the pulmonary vasculature suggesting a left to right shunt. The level of the shunt was of course not definite. However, in the lateral view, one noted re-opacification inferior and somewhat anterior to the sinus of Valsalva of the aorta. Within this region there was rather dense collection of the dye which suggested that there might be a communication between the base of the aorta and the right ventricle. There was also certainly a communication to allow a left to right shunt either at the atrial or ventricular level (F. 2).

The pre-operative right cardiac catheterization data revealed that there was hypertension in the right ventricle and the pulmonary artery system with a normal pulmonary capillary wedge pressure. There was no evidence of cardiac failure. The systemic blood oxygen saturation was normal, but there was a significant step-up of blood oxygen saturation in the right ventricle without further step-up in the pulmonary artery. Therefore, the conclusion was made that there was a left to right shunt at the ventricular level with hypertension in the right ventricle and

the pulmonary artery on this study (T. 1).

4. In this case, the nature and the location of the murmurs, the normal systemic blood pressure, the palpable systolic thrill in the third and fourth left intercostal space, the chest X-rays, and the electrocardiographic findings suggested an interventricular septal defect possibly associated with another lesion of the heart. Right cardiac catheterization findings supported this diagnosis. On angiocardiography, in addition to the re-opacification of the pulmonary vasculature at the time of left heart opacification, the marked dilatation of the sinus of Valsalva of the aorta was also found. The diagnosis of interventricular septal defect associated with aneurysmal dilatation of the sinus of Valsalva of the aorta was therefore established, pre-operatively. In this patient, who had cardio-aortic fistula (aortic sinus-right ventricular fistula), the systemic blood pressure was normal and the right ventricular diastolic pressure was only 1.5mm of Hg.. The only explanation for these findings was that the fistula was too small to affect the systemic pressure.

5. In the past six months of the post-operative period, the patient has been doing well. He has been very active and the patient's mother has noted marked symptomatic improvement. Chest X-rays showed that the pulmonary vascular markings are reduced to near normal. The electrocardiography indicated some improvement in the left ventricular hypertrophy overload pattern. We consider that this operation has been successful. However, in view of the continuous cardiac murmur discovered on his last visit, a right cardiac catheterization will be repeated in the near future for further evaluation.

SUMMARY

1. A description is given of a six year old negro, male child with congenital cardio-aortic fistula associated with interventricular septal defect.

2. The patient was born at full-term and has been developing fairly well. A cardiac murmur was discovered shortly after birth, but there was no history of cyanosis, sub-acute bacterial endocarditis, cardiac failure, acute onset of dyspnea, or chest pain.

3. The patient was diagnosed as congenital heart disease, interventricular septal defect associated with aneurysmal dilatation of the sinus of Valsalva of the aorta, pre-operatively. Open heart corrective surgery was performed by utilization of extracorporeal circulation. During the operation, the patient was found to have interventricular septal defect associated with dilatation of the sinus of Valsalva of the aorta and aortic sinus-right ventricular fistula. Both of the lesions were repaired and the operative technique described briefly. The patient recovered from the operation well and improvement was demonstrated symptomatically as well as clinically.

4. Ruptured aneurysm of the sinus of Valsalva of the aorta into the cardiac chambers can be surgically repaired with no greater risk than any other open cardiac surgery.

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和 文 抄 録

心室中隔欠損と Valsalva 動脈瘤破裂の合併症例に対する直視下心臓内手術の経験

聖フランス病院 （ニューヨーク）

鄭 煐 陳

私は心室中隔欠損と Valsalva 動脈瘤破裂の合併症例に対し，これを，人工心肺による直視下手術により治癒せしめ得たので報告する．

症例は6才の男子，心雑音，呼吸促迫を主訴として来院，聴診上第3乃至第4肋間，左胸骨縁に第Ⅲ～第Ⅳ度の収縮期雑音，第2肋間左胸骨縁に第Ⅰ度の拡張期雑音を聴取し，Angiocardiography により，左右両心室の拡大を認め，左心室造影時に再び，肺動脈の造影を認め，更にValsalva 動脈瘤の拡大が認められた，

心カテ所見では，右心室圧，肺動脈圧の軽度上昇と右心室及び肺動脈の酸素飽和度の上昇が認められた．動脈血酸素飽和度は正常値を示した．その他一般検査では異常を認めなかつた．

以上の所見から，心室中隔欠損兼 Valsalva 動脈瘤との診断で，人工心肺使用下に直視下手術を行なつた．

手術時所見では，右冠状動脈の位置異常があり，右心室壁の前上方を横切つていたため心室切開創が小さく，手術は甚だ困難で46分に及び遮断時間を必要としたが，8～10mmに及ぶ Valsalva 動脈瘤の破裂と，12～14mmの心室中隔欠損を認め，これらを縫合閉鎖した術後9ヵ月の所見では，術前の収縮期雑音は著しく減じたが，第2肋間左胸骨縁の拡張期雑音は増加した．然し，臨床的には著明な改善が認められた．