Case Reports

A Case of Sinus of Valsalva—Right Ventricular Fistula Successfully Repaired by Surgery

Takeshi KUZUYA, M.D., Hitoshi TANABE, M.D., Hiroshi KURI-
HARA, M.D., and Kizuku KURAMOTO, M.D.

AMONG the congenital anomalies of the heart, aneurysm of sinus of Valsalva has been considered to be a rather rare disease¹⁾ and only 12 cases were found in 1,000 autopsied cases of congenital heart diseases according to Abbott.²⁾ About 20 cases have so far been reported in Japan.^{3)-10), 15)-17)} Since the recent development of the method of temporary cardiopulmonary bypass using pump oxygenator, its surgical treatment has become possible. Although cases of successful operation have been accumulated recently,¹²⁾⁻¹⁴⁾ such success is still regarded as a rare experience in Japan. We are, therefore, reporting a case of successfully repaired sinus of Valsalva-right ventricular fistula.

CASE REPORT

A 24-year-old male student was admitted to Okinaka Clinic on June 19, 1958 with the chief complaints of exertional palpitation and breathlessness. His birth was not unusual. Development of growth in infancy and childhood was uneventful and there were no subjective difficulties until the age of 14. At 14 years of age, occasional palpitation and shortness of breath appeared during exercise and the symptom gradually became worse. At the age of 17, patient was told that he might have "congenital heart disease" by a local physician. At about the same time, a routine X-ray of the chest revealed pulmonary infiltration in the left. In April 1958, patent ductus arteriosus was suspected on routine physical examination because of the continuous murmur, and hospitalization was recommended for a further study. Palpitation, shortness of breath, and arrhythmia during exercise have gradually become severe in past few years, although the patient remained symptom-free at rest. No episode suggesting the rupture of Valsalva sinus aneurysm has been noticed.

Family history: Father, grandmother and a cousin (female) of father's side had "heart disease", the details of which were unknown. All three siblings are in good health.

Physical examination at the time of admission revealed a well nourished young male of medium size without cyanosis. Pulse rate was 70-80/min., slightly bounding, with good filling. Quincke's capillary pulsation was positive. No clubbed finger was present. Respiration was normal. Blood pressure was 120/50 mm.Hg

From the Okinaka Clinic, Department of Internal Medicine, University of Tokyo, Tokyo.

in arms and 140 mm.Hg systolic in legs. Cardiac dullness was extended 2 cm. to the right from the right sternal border and 2 cm. to the left from the left nipple line. The upper border was in the 3rd rib. Apical beat and thrill were felt in the fifth intercostal space 2 cm. outside of the left nipple line. Coarse grade IV continuous machinery murmur was best heard at the 3rd interspace on the left near the left sternal border, radiating from the base to the apex. Systolic phase was louder than diastolic. Chest and abdomen revealed no remarkable changes. Neurological examination was negative.

Laboratory findings: Urine and stool were negative. Hemoglobin 108% Sahli, RBC 497×10^4 , WBC 6,200 with normal differential count. Serological tests for syphilis were negative. Arm to tongue circulation time was 19 seconds with cholere-tin.

An AP chest X-ray showed a slight cardiac enlargement bilaterally, without protrusion of the main pulmonary artery or increase of lung markings. At LAO, left ventricular enlargement was apparent, without forceful hilar vascular pulsation. ECG revealed deep S in V_1 through V_3 , high R in V_4 through V_6 , suggesting left ventricular hypertrophy. Phonocardiograph recorded a continuous murmur in the 2nd to 4th interspace along the left sternal border, having the largest amplitude around the 2nd sound, without definite diastolic accentuation. Near the apex, however, the systolic murmur was larger in amplitude than the diastolic. Right heart catheterization revealed no particular elevation of right ventricular or pulmonary arterial pressure. Right ventricular blood contained definitely more oxygen than right atrial blood, indicating a left to right shunt at the right ventricular

Fig. 1 (Left). Chest X ray, anterior position, showing slight bilateral enlargement of heart.

Fig. 2 (Right). LAO position. Hypertrophy of left ventricle is present.

level. Tentative diagnosis of fistulation to right ventricle of Valsalva sinus aneurysm was made. In order to rule out inter-ventricular septal defect complicated with aortic insufficiency, retrograde aortography was performed. Inserting a catheter from right axillary artery, the catheter was found to enter directly into the right ventricle (pressure 30/0 mm.Hg) from the position immediately above the aortic valve

Fig. 3. RAO position.

Fig. 4. ECG. Deep S in V_1 - V_3 , high R in V_4 - V_6 suggesting left ventricular hypertrophy.

(125/60 mm.Hg). Fig. 6 represents the pulling out curve from right ventricle to aorta, clearly indicating the communication between aorta and right ventricle. This procedure was repeated three times with the same results. Opaque media (Sugiuron) was injected when the tip was immediately above the aortic valve to take serial X-ray pictures, which were interpreted as showing the entry of opaque media to the right ventricle.

Under the final clinical diagnosis of fistula of Valsalva sinus aneurysm to right ventricle, the patient was once discharged on July 20, 1958. In October 1958, the patient suffered from acute hepatitis, and was hospitalized again. The recovery was uneventful and all liver function tests became normal.

Fig. 5. Phonocardiogram. Continuous murmur without diastolic accentuation.

Table I. Right Heart Catheterization, Indicating Significant Higher O₂ Content in Right Ventricle than in Right Atrium

	Position	Pressure in mm. Hg	O ₂ content (vol. %)
1	Pulmonary capillary (wedge)	10/0	19.2
2	Right pulmonary artery		16.3
3	Main pulmonary artery		16.5
4	Right ventricle (outflow)	37/0	17.0
5	Right ventricle (inflow)		16.3
6	Right atrium	6/3	14.2
7	Superior vena cava		14.4
8	Inferior vena cava		14.6
9	Femoral artery		18.3

Pulmonary blood flow 12.9 L./min.
Effective pulmonary blood flow 6.3 L./min.
Left to right shunt 6.6 L./min.

On Jan. 15, 1959, he was admitted to Kimoto Clinic, Department of Surgery, University of Tokyo for the operation. On Feb. 11, under the inhalation anesthesia of N₂O-O₂ with ether, thoracotomy was done at the left 4th interspace, with transection of the sternum. On opening the pericardial cavity, thrill was felt immediately below the pulmonic valve from outside. By the finger inserted into the right ventricle, protrusion of Valsalva sinus aneurysm was palpated.

Following the cannulation of left femoral artery, superior and inferior vena cava were also cannulated through the right atrium. After total cardiopulmonary bypass was completed using bubble-type pump oxygenator of De Wall-Lillehei

Fig. 6. Pulling out curve of arterial catheter from right ventricle to aorta, demonstrating direct communication between aorta and right ventricle.

Operation (Feb. 11, 1959)

Fig. 7. Operation.

1. Right ventricle opened, aneurysm of Valsalva sinus protruded through IVSD immediately below pulmonic valve. Through the hole at the top of aneurysm, jet of blood streamed.
2. Aneurysm resected and sewn up.
3. IVSD closed.

type, right ventricle was opened. Directly below the pulmonic valve, high inter-ventricular septal defect of 8 mm. in diameter was noted, through which protruded the aneurysm of sinus of Valsalva 15 mm. in length 8 mm. in diameter into the right ventricle. An opening was seen at the top, through which the jet of blood streamed upon loosening the tape on the aorta. First, this aneurysm was resected and closed with 7 stitches, and the IVSD was closed with 4 stitches. Right ventricle was then sewn, cannulae were pulled out from venae cavae and artery, and the chest was closed leaving bilateral drainages. The time of total cardiopulmonary bypass was 18 minutes and 40 seconds.

After 3 weeks, postoperative pleural effusion cleared up and subsequent course was uneventful. Fifty days after the operation, the patient was discharged with the only complaint of slight palpitation after walking. The coarse continuous murmur disappeared postoperatively. A diastolic murmur, however, was heard

Table II. The Cases of Valsalva Sinus Aneurysm in Japan

Authors	Age Sex	History	Heart sound	Blood pressure	X-ray findings of heart
1. Koba- yashi <i>et</i> <i>al.</i> 1944	22 M	"Heart disease" since age 3-4. Exertional dyspnea.	Continuous mur- mur. DM*>SM*. Max. at L. 3-4 ICS.	128/30-0	Enlarged bilaterally. RVH
2. Sugie 1955	23 M	Congenital heart disease at 4 years. Decompensation since age 15. SBE at age 19.	CM* max. at 4. ICS. L. f.b.* out- side of L. sternal border. P ₂ accent. DM accentuated.	152/0	Enlarged bi- laterally. L. 2nd arc protruded Pulm. conges- tion. Hilar dance (##)
3. Ishihara 1955	39 M	Exertional dyspnea since 3 years. De- compensation since 1 year. Ascites pre- sent.	CM max. at 3. ICS 1 f.b. out- side of L. sternal border SM>DM. No P ₂ accentuation.	156/0	Marked bi- lateral enlarge- ment. Pulmonary congestion.
4. Sugiura 1956	28 M	"Valvular disease" during childhood, later congestive failure. SBE since last year.	CM max. at 3. ICS. DM accentuated.	114/40-0	
5. ,,	19 M	At age 17, attack of loss of consciousness with agonizing feel- ing in the chest. At age 18 similar attack.	SM max at L. 3. ICS. DM slight to the left.		Enlarged bilaterally.
6. Oguro <i>et</i> <i>al.</i> 1957	37 M	"Heart disease" since infancy. Ex- ertional dyspnea.	CM max. at L. 3. ICS. DM>SM.	170/0	Enlarged bi- laterally. L. 2nd arc promi- nent. Pulmonary congestion.
7. Tasaka <i>et al.</i> 1959	52 M	2M ago, acute on- set of dyspnea and epigastric pain after running, thereafter exertional dyspnea.	High-pitched DM at the base.	158/0	L. 1st and 4th arc prominent.
8. Shibuya 1959	33 M	Since boyhood "congenital heart disease". 1 year ago, precordial pressure feeling, dyspnea.	CM, L. 2-3 ICS. DM>SM.	156/0	Enlarged bi- laterally. L. 2nd arc prominent. Pulmonary congestion.
9. our case	21 M	Since age 14, exer- tional dyspnea, palpitation. At 17 "heart disease".	CM max. at L. 3. ICS. SM>DM.	120/50	Bilaterally en- larged.

* DM=diastolic murmur

SM=systolic murmur

CM=continuous murmur

ICS=intercostal space

f.b.=fingerbreadth

(Only those with adequate description are cited)

ECG	Cardiac catheterization	Clinical diagnosis	Course	Final diagnosis
LVH, RVH. incomplete RBBB.	+	Fistula of Valsalva aneurysm (congenital)	unknown	
Left type.	+	Aortic septal defect + PI? or Valsalva sinus fistula?	Died shortly after operation	Rupture of RCS** (3 holes) to RV** + IVSD
Right type. ST depression.	—	?	Died 14 days after admission due to congestive failure.	Rupture of RCS to RV
	—	Rupture of sinus of Valsalva to RV** (congenital)?	Died after 4 months due to decompensation.	Rupture of RCS to RV
	+	high IVSD	Operated.	Aneurysm of sinus of Valsalva + IVSD
	+	Rupture of Valsalva sinus to RV?	Died 1 Y. 8 M. after appearance of symptoms due to congestive failure.	Fistula of right Valsalva sinus + IVSD
LVH. Slight myocardial damage.	—	Aortic insufficiency	Died 9 M. after onset.	RCS and LCS** ruptured into LV**
LVH. RVH.	+	Ruptured aneurysm of sinus of Valsalva to RV?	Died 10 M. after onset.	Rupture of RCS to RV + IVSD + Thickening of R. cusp of aortic valve
Suggestive of LVH.	+	Fistula of Valsalva sinus aneurysm to RV.	Operated and cured.	Fistula of aneurysm of Valsalva sinus to RV + IVSD

** RV=right ventricle

LV=left ventricle

RCS=right coronary sinus

LCS=left coronary Sinus

in the 2nd-3rd interspace along the left parasternal line, which probably represents pulmonary insufficiency resulting from involvement of pulmonic valve during the suture of IVSD.

DISCUSSION

Nearly 20 cases of Valsalva sinus aneurysm have been already reported in Japan, including 12 operated and 7 autopsied cases.^{3)-10), 15-17)} Table II summarizes those cases provided with adequate description.

Cardiac murmur of this disease resembles that of patent ductus arteriosus, IVSD with aortic insufficiency, aortic septal defect, etc. In the present case, retrograde aortography gave a confirmatory evidence for the diagnosis of Valsalva sinus aneurysm as the tip of the catheter went into the right ventricle. Although Morrow and his associates³³⁾ reported the passage of catheter through the fistula, the present case was the first case in Japan in which the diagnosis was confirmed by the method mentioned.

Aneurysm of sinus of Valsalva may be congenital or acquired. According to Jones & Langley,¹¹⁾ various associated anomalies often accompany the congenital type; such as anomalies of aortic cusps (15/23), or IVSD (10/23) etc. Negative serological tests for syphilis and the presence of high inter-ventricular septal defect as well as the history seem to make our case most likely that of congenital. Since no episode suggesting the rupture of the aneurysm was known in this case, the fistula might have been present from the birth.

Although successful operations for this defect have been gradually increased in number recently, only 5 cases of survivals¹⁵⁾⁻¹⁷⁾ are recorded among 12 operated cases in Japan. Postoperative recurrence of fistula was reported in a case treated only with the suture of IVSD.¹⁶⁾

With the recent advances in intra-cardiac surgery under direct vision, a correct diagnosis has become a matter of not only academic interest but practical importance.

SUMMARY

A case of successfully repaired sinus of Valsalva-right ventricular fistula in a young male is presented. Exertional palpitation and dyspnea, slight enlargement of heart, and the continuous machinery murmur maximally heard at the left 3rd intercostal space, were the chief clinical manifestations and signs. Diagnosis was suspected by the right heart catheterization and confirmed with the direct entrance of arterial catheter into the right ventricle from immediately above the aortic valve in retrograde aortography.

Under the total cardiopulmonary bypass, using the pump oxygenator, aneurysm of Valsalva sinus was resected and sutured, and associated high IVSD was also closed. Postoperative course was uneventful.

Brief summary of cases of this disease in Japan is also tabulated.

ACKNOWLEDGMENTS

We should like to express our deepest gratitude to Prof. Okinaka, Dr. Ikeda and the staffs of cardiac group of Okinaka Clinic, for their advice and encouragement. Our appreciation is also due to Prof. Kimoto, Dr. Furuta and the staffs of Kimoto Clinic, Department of Surgery, University of Tokyo, for their help in clinical study and the operation.

REFERENCES

1. Taussig, H. B.: Congenital Malformations of Heart, The Common Wealth Fund, New York, 1958.
2. Sano, T., Hellerstein, H. K., and Keating, D. R.: Clinical Aspects of Congenital Heart Disease, Igaku-Shoin, Tokyo, 1956 (In Japanese).
3. Kobayashi, T. and Shiba, M.: Respiration and Circulation **2**: 298, 1954 (In Japanese).
4. Sugie, S., et al.: Respiration and Circulation **3**: 270, 1955 (In Japanese).
5. Ishihara, T.: J. Jap. Soc. Int. Med. **44**: 319, 1955 (In Japanese).
6. Sugiura, M., et al.: J. Jap. Soc. Int. Med. **45**: 653, 1956 (In Japanese).
7. Oguro, C., et al.: Respiration and Circulation **5**: 443, 1957 (In Japanese).
8. Oguro, C.: Jap. Circulation J. **20**: 589, 1957 (In Japanese).
9. Tasaka, S., Koide, K., Ogata, E., and Nakamura, K.: J. Jap. Soc. Int. Med. **48**: 1306, 1959 (In Japanese).
10. Shibuya, M., Ishii, S., Yamaguchi, S., and Kanai, M.: Respiration and Circulation **7**: 509, 1959 (In Japanese).
11. Jones, A. M. and Langley, F. A.: Brit. Heart J. **11**: 325, 1949.
12. Morrow, A. G., Baker, R. R., Hanson, H. E., and Mattingly, T. W.: Circulation **16**: 533-538, 1957.
13. Lillehei, C. W., Stanley, P., and Varco R. L.: Ann. Surg. **146**: 456, 1957.
14. Kay, J. H., Anderson, R. M., Lewis, R. R., and Remberg, M.: Circulation **20**: 427, 1957.
15. Ozawa, Y.: Proc. of the 15th General Assembly of the Japan Medical Congress, April 1959, Tokyo, IV: 495, 1959 (In Japanese).
16. Kimoto, S.: *ibid.* IV: 503, 1959 (In Japanese).
17. Sakakibara, S.: *ibid.* IV: 522, 1959 (In Japanese).