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症　　例

Congenital Coronary Arterial Fistula —A Case Report—

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Since Krause[9] reported a case of congenital coronary arterial fistula in 1865, this lesion had been considered as an uncommon lesion.

But with the improvement in facilities for diagnosis and correction of such an anomaly there has been an increase in interest in this lesion and an increase in the number of case reports. Among the various congenital anomalies of the coronary vascular system one of the most important ones is the abnormal communication of the coronary artery with a cardiac chamber or vessel. This anomaly can now be easily and accurately diagnosed by selective coronary angiography preoperatively, and also can be operated on successfully by various surgical procedures such as ligation, division, arteriorrhaphy and aneurysmorrhaphy. But the general lack of sufficient knowledge about the natural history and surgical prognosis can be seen in the lack of uniformity in operative indication and procedures.

A case of a 7-year-old boy with congenital coronary arterial fistula which branched from the left circumflex artery and entered near the sinoatrial node area of the right atrium is presented in this paper, and the surgical problems in this lesion are described.

Case report

N.K., a 7-year-old boy, was referred for evaluation concerning a heart murmur first heard at the age of 2 years and 6 months and thought to be due to a patent ductus arteriosus or congenital coronary arterial fistula.

Key words: Congenital coronary arterial fistula, Surgical procedures, Operative indications, Ultrasonic cardiotomography, Postoperative thrombosis.
索引語：先天性冠動脈瘻，手術々式，手術適応，超音波断層法，術後血栓形成.
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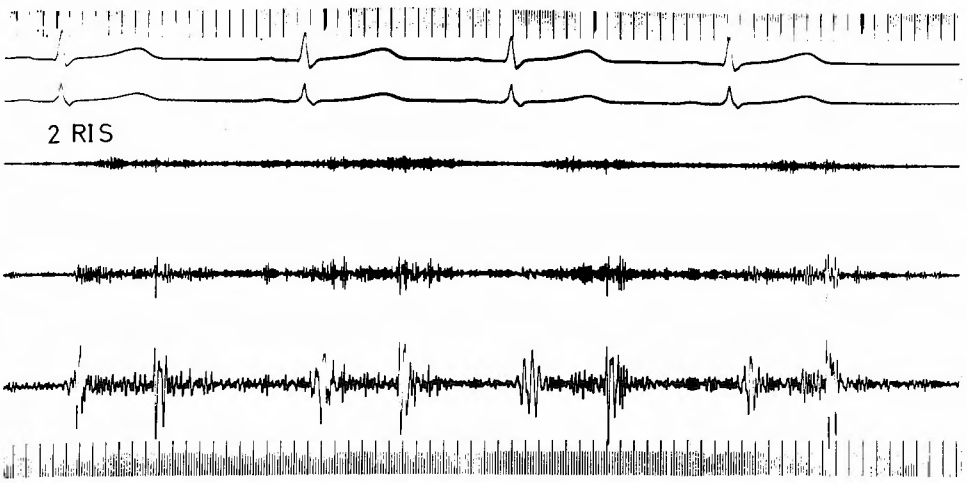
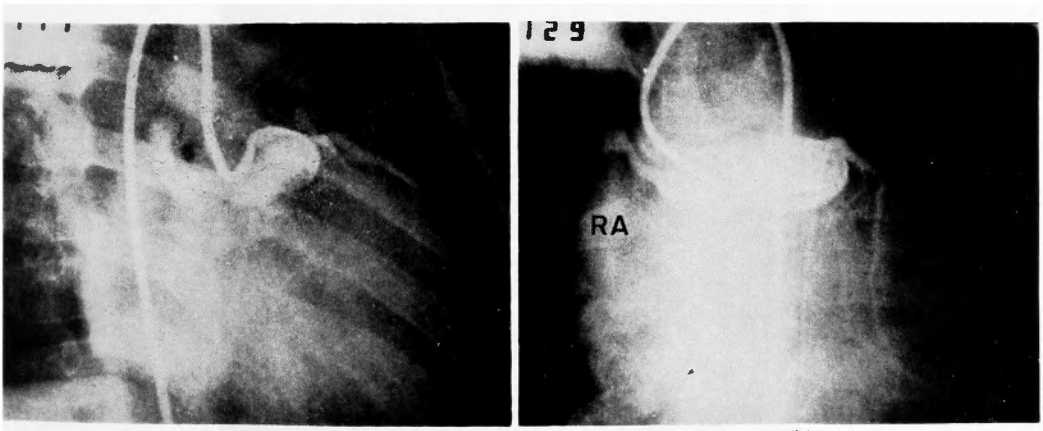


Fig. 1. Phonocardiogram
A grade 3/6 continuous murmur, maximal over the 3rd interspace along the right sternal border, was heard.

Pregnancy and delivery were normal; the birth weight was 2900 gm.. He was asymptomatic; his development was smooth. He was not prone to upper respiratory infection and there was no cyanosis, edema, or other symptoms.

Accurate preoperative examination was performed on December 13, 1979, and revealed an abnormal coronary artery which branched from the left circumflex artery and drained into the right atrium.

Physical examination disclosed a well developed and well nourished boy. The pulse was regular, 76 per minute, and the blood pressure was 118/68 mmHg with normal peripheral



(a) (b)

Fig. 2. Preoperative selective coronary angiography
An abnormal coronary artery branched from the left circumflex artery and drained into the right atrium, having an abnormal and tortuous course. The left coronary artery had remakable aneurysmatic dilatation between the root of the aorta and the left circumflex artery.
Fig. 2a was taken in RAO projection. Fig. 2b was taken in A–P projection.

pulses. There was no cyanosis, clubbing, or chest deformity. The first sound was normal, and the second was of normal intensity and normally split. A grade 3/6 continuous murmur, maximal over the third interspace along the right sternal border, was heard and varied markedly with respiration. These findings were confirmed by phonocardiogram (Fig. 1).

Roentgenograms of the chest in frontal and oblique projections revealed slight cardiac enlargement with a cardiothoracic ratio of 54.5%. The pulmonary vascular markings were within normal limits.

The electrocardiogram showed a transiently wandering P-wave but this could not definitely be interpreted as indicating left ventricular hypertrophy. No evidence of myocardial infarction was proved.

Catheterization of the right and left sides of the heart was performed on December 13, 1979. The presence of a left to right shunt at the atrial level was indicated by an increase in blood oxygen content of 0.96 Vol. per cent from the venae cavae to the right atrium. The shunt, 1.08 liters per minute, amounted to 24.5 per cent of the pulmonary blood flow. All pressures were within normal limits. Calculated output and resistances were normal.

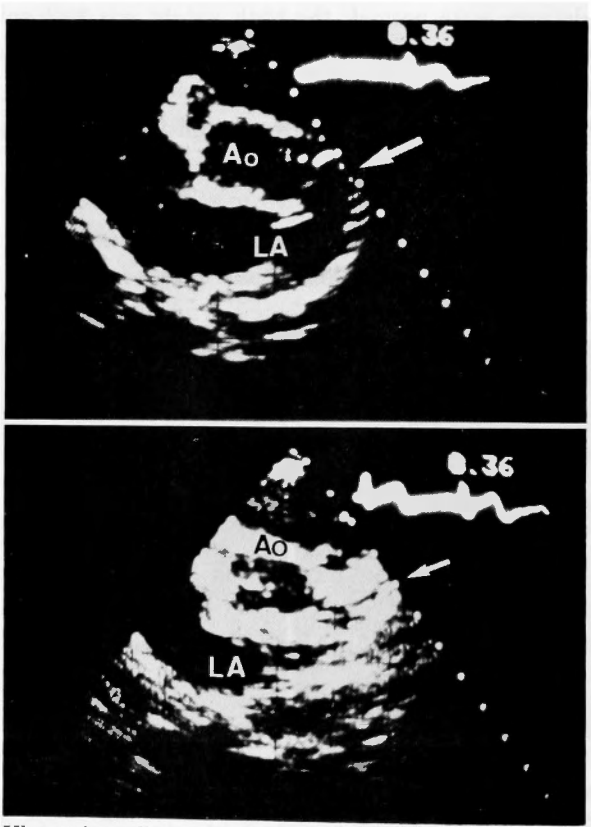


Fig. 3. Ultrasonic cardiotomography.
Arrows point to the aneurysmatic dilatation of the main trunk of the left coronary artery. Fig. 3a was taken in a short axis at a level of 2 cm above the aortic valve. Fig. 3b was taken at the level of the aortic valve.

By selective coronary angiography in the both oblique projections, an abnormal coronary artery which branched from the left coronary artery and drained into the sinoatrial (S-A) node of the right atrium was found. The left coronary artery had remarkable aneurysmatic dilatation between the root of the aorta and the left circumflex artery, and the course of the abnormal coronary artery resembled S-A node artery branched from the left circumflex artery and was tortuous (Fig. 2).

Ultrasonic cardiotomography revealed the aneurysmatic dilatation of the main trunk of the left coronary artery (Fig. 3) and these findings were comparable to the findings of the selective coronary angiography of the left coronary artery.

Surgical correction was performed under extracorporeal circulation on January 21, 1980. The right atrium was slightly enlarged and a small swelling was seen just in the area of the S-A node over which a strong thrill was felt continuously (Fig. 4). The right coronary artery was normal in course and diameter. The left coronary artery had aneurysmatic dilatation between the root of the aorta and the bifurcation of the left circumflex artery (Fig. 5). After the bifurcation of the left circumflex artery, the abnormal coronary artery ran horizontally over the posterior wall of the left atrium, and turned up and away from the cardiac wall and soon turned abruptly down draining into the area of the S–A node of the right atrium. This abnormal coronary artery was regarded as S–A node artery, a branch of the left circumflex artery, considered anatomically by its course. Because the abnormal S-A node artery had an orifice in the area of the S–A node, there was the possibility that the direct closure of the abnormal coronary artery from the inside of the right atrium might lead to arrhythmia. So the S-A node

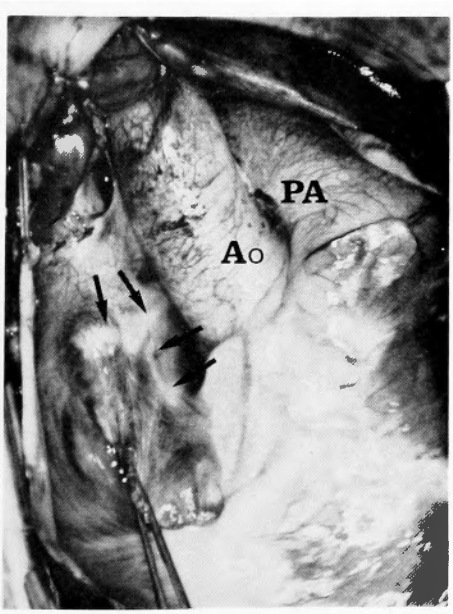


Fig. 4. An abnormal coronary artery (arrows) drained into the right atrium, the area of the S–A node.
Ao: aorta PA: pulmonary artery

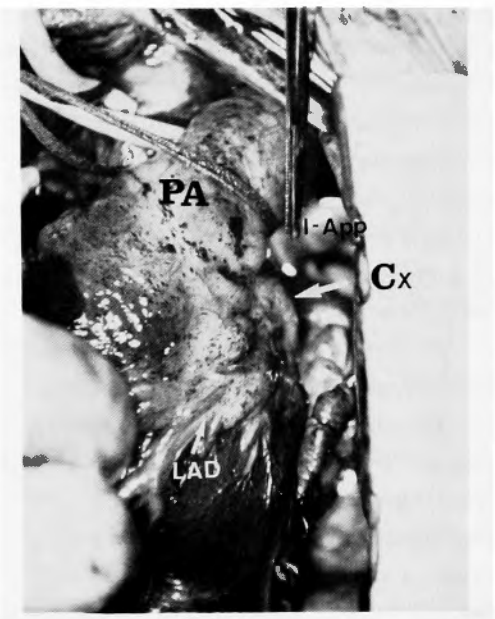


Fig. 5. The left coronary artery had aneurysmatic dilatation between the root of the aorta and the bifurcation of the left circumflex artery.
PA: pulmonary artery, Cx: circumflex tree, 1-App: left atrial appendage, LAD: left anterior descending artery.

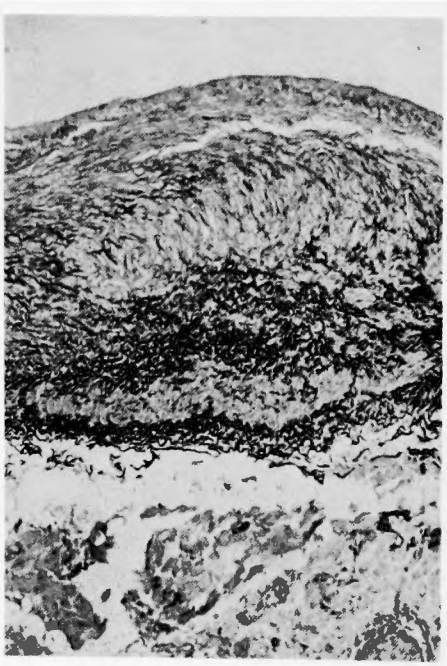


Fig. 6. Histological study revealed that the wall of aneurysmatic coronary artery has three layers without any inflammatory findings but the elastic fibers were heavily ruptured. (Van Gieson's connective fiber stain)

artery was ligated and divided just before the drainage into right atrium in the distal portion.

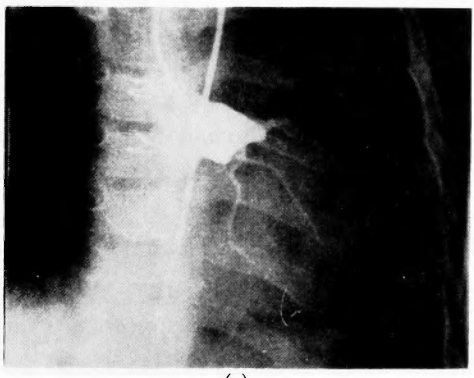
A longitudinal incision was made over the aneurysmatic main trunk of left coronary artery and the orifice was closed with direct suture from the inside and the edges of incision were sutured together gradually decreasing in diameter.

Postoperative course was uneventful; no change in electrocardiogram was found and cardiac murmur was not heard at all.

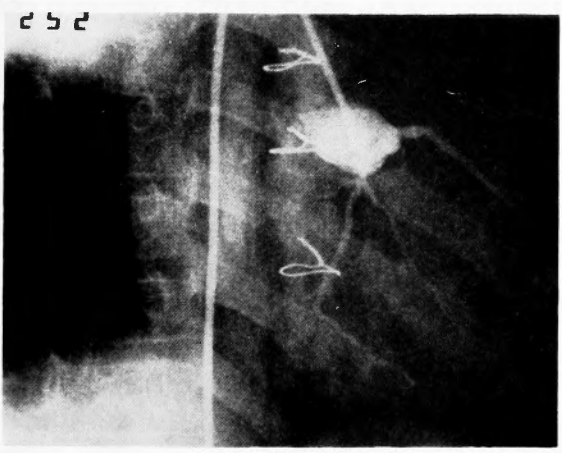
Anti-coagulant therapy by Warfarin® was administered to prevent the development of a thrombus in the coronary artery for a period of 3 months postoperatively.

A small specimen of the wall of the aneurysmatic coronary artery was removed for histological study. Histological study revealed that the wall of aneurysmatic coronary artery has three layers without any inflammatory findings but the elastic fibers were heavily ruptured (Fig. 6).

Postoperative cardiac catheterization and selective coronary angiography were performed



(a)



(b)

Fig. 7a, b. Postoperative selective coronary angiography
The aneurysmatic coronary artery was almost the same as in the preoperative state. Fig. 7a was taken in A-P projection. Fig. 7b was taken in RAO projection.

on May 21, 1980. Selective coronary angiography showed that the aneurysmatic coronary artery was almost the same as in the preoperative state (Fig. 7).

Discussion

Congenital coronary arterial fistula is an uncommon lesion. The improvement in facilities for diagnosis, especially selective coronary angiography and ultrasonic cardiotomography has stimulated interest in this lesion and increased the number of case reports.

FURUSHIMA et al.[4] reported one case and reviewed 106 cases in Japan. OLDHAM et al.[15] reviewed 200 cases in Europe and America.

In most cases of congenital arterial fistula, aneurysmatic dilatation of the associated coronary artery and its tortuous course are usually seen. In our case remarkable aneurysmatic dilatation was seen in the main trunk of left coronary artery and a tortuous course was seen in the distal portion.

Originating coronary artery is usually to the right side and occurs at about the same rate of 55 to 57 per cent in Japan, Europe and America.

The receiving chambers are, in order of frequency, as follows: right ventricle, right atrium, pulmonary artery and left ventricle. In FURUSHIMA's[4] review the number of left coronary artery to right atrium fistulas as compared to right coronary artery to right atrium fistulas were 16 and 12 respectively. The other types, found in greater frequency, are as follows: right coronary artery to right ventricle (30 cases), left coronary artery to right atrium (16 cases), right coronary artery to right atrium (12 cases) and right coronary artery to left ventricle (10 cases).

SAKAKIBARA[17] classified the types of drainage into cardiac chamber into 3 groups; type 1) the distal end of the branch of the main coronary artery, type 2) the side of the main coronary artery, type 3) the distal end of the main coronary artery. The cases in which the associated coronary artery drains into the right atrium, left atrium or pulmonary artery belong to type 1. The cases in which the associated coronary artery was connected to the right ventricle belong to type 1, type 2 or type 3, in about the same frequency. The cases of the associated coronary artery draining into the left ventricle usually belong to type 3. In types 1 and 3, there may not be any problems in surgical treatment. However, in type 2 a careful choice of surgical procedures is needed on closing the fistulas. In most cases selective ligation of the fistula may be safely done because blood flow can be maintained by coronary artery on the other side.

In the congenital coronary arterial fistula clinical features depend on the extent of the shunt. A large shunt may cause right heart failure.

The continuous murmur of this lesion has a striking characteristic and resembles those of patent ductus arteriosus, ruptured aneurysma of the sinus of Valsalva, ventricular septal defect combined with aortic regurgitation and so on.

It is possible to differentiate the coronary arterial fistula from other lesions with similar heart murmur if the abnormal position of the murmur and the shallowness of the murmur are noticed. The maximal area of the heart murmur is related to the position of drainage into the cardiac

chamber, for example, if the entrance is in the upper portion of the right atrium, the cardiac murmur is heard loudest over the 3rd interspace along the right sternal border as seen in the case reported here.

On studying operative indication accurate knowledge of the natural history and surgical prognosis are greatly needed. Whether the diameter of the coronary artery having aneurysmatic dilatation decreases or remains the same as in the preoperative state varies from case to case. Cases have been reported in which the fistula was closed naturally or in which no symptoms were present. Therefore, there may not always be operative indications in all cases with congenital coronary arterial fistula. But the causes of death and complications of this disease are, as many authors[3,5,6,7,11,12,14] have pointed out, rupture of the aneurysmatic coronary artery, congestive heart failure or pulmonary hypertension following large left to right shunts, subacute bacterial endocarditis[20], and angina or/and myocardial infarction following the development of thrombus and occlusion of the coronary artery.

KONNO[8] said that if the following conditions are present, operation on the congenital coronary arterial fistula may be indicated:

1. The shunt is more than 30 per cent even though no symptoms are present.
2. Ischemic changes or strain patterns in electrocardiogram
3. Progressive pulmonary hypertension
4. Progressive congestive heart failure
5. Past history of bacterial endocarditis
6. Great possibility of rupture of aneurysmatic coronary artery
7. The loud murmur becomes a disadvantage in getting a job.

None of the above conditions applied in our case. But we decided to operate in order to protect against myocardial damage following thrombosis and prevent bacterial endocarditis.

Some of the various surgical procedures for this lesion are, as follows:

1. Ligation and/or division

This procedure is simple and does not require extracorporeal circulation. If the trees of the fistula support nutrition, myocardial ischemia will subsequently occur. Many authors have recommended that prior to ligation and/or division it is necessary to perform a temporary test occlusion of the coronary artery for 15 to 20 minutes in order to comfirm by electrocardiogram that no ischemic pattern or arrhythmia will develop. But there have been cases in which fatal myocardial damage occurred even though there were no changes in the electrocardiogram during the test period. There is the possibility that progressive retrograde development of thrombus may have occurred. Therefore it seems advisable, if the fistula does not support nutrition, to close both the proximal and distal portions.

2. Selective ligation of the fistula

This procedure may be done safely, without risking myocardial ischemia, when the associated coronary artery has an orifice in the side wall; not when there are multiple orifices scattered over a wide area[17].

3. Tangential arteriorrhaphy

As shown in COOLEY's report[2], in the case of multiple orifices scattered over a wide area, many overlapping U-mattress sutures were performed on the longitudinal axis along the border of the associated coronary artery and the associated coronary artery in the functional state was separated from the cardiac chamber. However, cases of regression following this procedure have been reported[6,13].

4. Closure from the inside of the chamber

This procedure may be suitable when the fistula terminates in the right atrium and atrial septal defect is complicated. It is difficult to close the orifices which are between the trabeculae. Also the possibility of postoperative hypofunction of the cardiac chamber, right and left ventricle, may be increased by the incision. This procedure is not recommended when the orifice of the fistula is located in the left ventricle.

5. SYMBAS' internal arteriorrhaphy[19]

In this method the associated coronary artery is incised longitudinally and the orifice is closed with sutures from the inside. It is possible to decrease the diameter of the aneurysmatic coronary artery by aneurysmorrhaphy. This procedure is not only applicable to when the orifice is located at the end of the fistula or on the side of the fistula, but also permits the blood flow to remain normal. However LIOTTA[10] and SYMBAS[19] have pointed out that with this method there is the possibility that a thrombus may develop at any time from an early stage to a long postoperative period. They recommended the administration of anti-coagulant drugs. The development of a thrombus in the aneurysmatic coronary artery does not depend on the existence of the fistula. The main factor in the development of a thrombus is thought to be congestion following the closure of the fistula. It is reasonable to perform aneurysmorrhaphy in the aneurysmatic fistula.

ARAYA et al.[1] reported a case with rupture of the aneurysmatic fistula following surgery in which closure of proximal side was not performed. SUNADA et al.[13] reported a case in which ligation was performed on both sides of the aneurysmatic portion. However, there is the possibility that a thrombus may develop in the remaining blind portion of the fistula.

6. Ligation of the associated coronary artery and aorto-coronary bypass[10]

This method is restricted to adult cases. It is impossible to perform on children because of patency of graft, technical problems and growth.

There have been many reports[13] that residual shunts occurred following ligation and tangential arteriorrhaphy.

NOGUCHI et al.[13] reported that the main causes of occurring residual shunts following operation are: the remaining fistula, recanalization of closed fistula and the characteristic structure of coronary artery. Therefore closure by more than triple ligation and division of the fistula are desirable together with the suture-ligation from the inside of the fistula in order to prevent recurrence.

Summary

The case of a 7-year-old boy with left coronary artery to right atrium fistula was reported here. The fistula was closed with ligation and division at the distal portion just before entrance into right atrium, and was closed with sutures from the inside at the proximal portion. Additional aneurysmorrhaphy was performed at the proximal portion. Anti-coagulant drug was administered for three months postoperatively. The postoperative course was uneventful. Ultrasonic cardiotomography was useful in detecting and observing the aneurysmatic dilatation of the aortic root of the fistula during diagnosis and follow-up study.

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和文抄録

先天性冠動脈瘻の1手術治験例

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症例は7才男児，左冠状動脈—右房瘻の1例で，人工心肺使用下に閉鎖手術を行った．瘻は，回旋枝より分枝し，右房洞結節部に流入していた．又，左冠動脈主幹部は瘤状に拡大していた．閉鎖に当り，末梢側は，洞結節の損傷を避けるために，その流入直前で，外部から瘻を結紮，切断した．又，中枢側は，冠動脈を縦切開し，直視下に瘻起始部を内側より閉鎖し，更に，一部冠動脈瘤の縫縮を行った．術後3ケ月間，血栓形成防止の目的で，抗凝固療法を行った．

なお，本症の診断及び術後観察には超音波断層法が有力と考える．