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Tricuspid Atresia with Polysurgery —A case report—

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Introduction

Since FONTAN[2] reported the successful repair of tricuspid atresia in 1971, various modified surgical techniques have[1,3,4,5,7,8,9] been described. Recently this Fontan-like operation is performed with a wider indication. However, these operations are limited to patients over 5 years old[4]. Thus some patients may undergo polysurgery, palliative and/or radical operations.

We experienced a case of tricuspid atresia Ib in which Glenn operation, left sided Blalock-Taussig operation, Fontan-like operation, closure of a previously undetected ASD and two subsequent operations for hemostasis were performed.

The problems in the surgical procedure following the previous thoracic operations and the postoperative management will be described herein.

Case report

The patient, a 9-year-old boy, was born after a full term pregnancy and normal delivery. A cardiac murmur was detected at the age of 3 months in a hospital in Kyoto City. As the baby could not take in an adequate supply of milk, he was taken to the Department of Pediatrics, Kyoto Prefectural Hospital, where he was diagnosed as ventricular septal defect (VSD) and given medication. Cyanosis increased gradually to the age of 4 months when general cramps developed for periods of 1 to 2 minutes. These anoxic spells increased to a frequency of once in 3 days until the age of 9 months, when he underwent cardiac catheterization and was diagnosed as VSD. Despite continued medication, his general condition became worse and he was referred to the

Key words: Fontan-like operation, Glenn operation, Blalock-Taussig operation, Polysurgery, Surgical procedures.
索引語：Fontan 様手術，Glenn 手術，Blalock-Taussig 手術，頻回手術，手術操作.
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Department of Pediatrics, Kyoto University Hospital at the age of 1 year. The second cardiac catheterization disclosed tricuspid atresia Ib.

On December 6, 1972, when he was 14 months old, he underwent a Glenn operation in the 2nd Department of Surgery, Kyoto University Hospital. On February 12, 1975, subsequent catheterization disclosed a systemic oxygen saturation of 79.2%, a hematocrit value of 51% and a hemoglobin level of 16 gm/dl. A Grade 3/6 systolic murmur was noted in the left sternal border. The cardiac apex was located in the fifth intercostal space at the midclavicular line. Electrocardiogram showed sinus rhythm and QRS axis of −20 degrees, with left ventricular enlargement. Upon chest roentgenogram, cardiac shadow was enlarged with a CTR of 0.51.

However, at the age of 6 years, the child again had dyspnea and a cyanotic crisis. On November 8, 1976, a left sided Blalock-Taussig operation was performed. The postoperative course was uneventful.

Upon discharge, the patient had a Grade 3/6, continuous murmur and mild cyanosis, but no anoxic episodes. A thrill was palpable in the left 2nd intercostal space at the sternal border, and the apical thrust was palpable in the fifth intercostal space at the midclavicular line.

A fourth cardiac catheterization, performed on December 15, 1977, disclosed a systemic oxygen saturation of 82%, a hematocrit value of 49%, and a hemoglobin level of 12 gm/dl. Angiography taken in the left atrium showed a small right ventricle with a well-developed pulmonary artery (Fig. 1). The aortogram revealed a Blalock-Taussig shunt, the blood flow being less than shortly after the operation.

The patient was admitted to our institute on February 26, 1980, in order to undergo functional radical operation, i.e. a Fontan-like operation, because of gradually increasing cyanosis.

Upon admission, he was ten years of age, his weight and height were 127 cm and 22 kg,

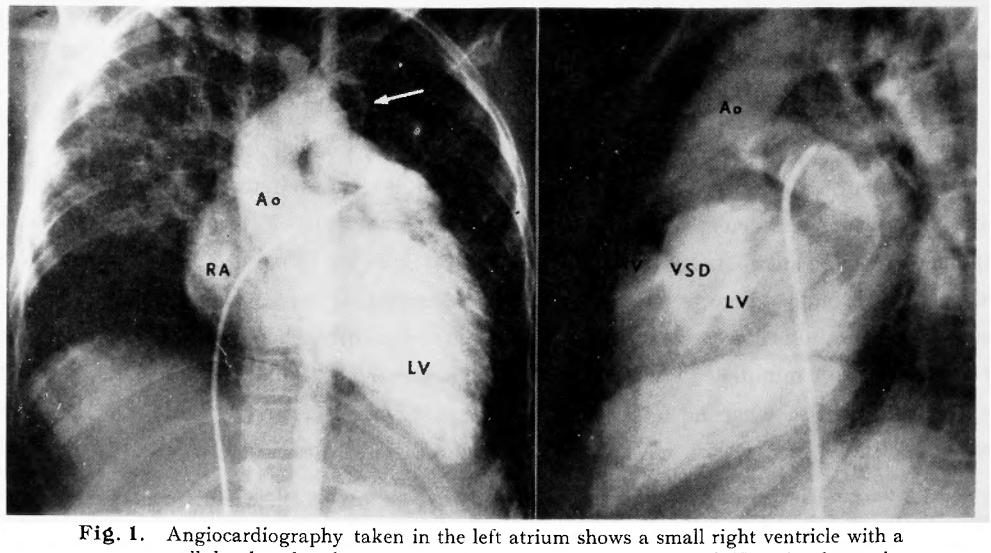

Fig. 1. Angiocardiography taken in the left atrium shows a small right ventricle with a well-developed pulmonary artery. Arrow shows a Blalock-Taussig shunt, the blood flow being less than shortly after the operation.

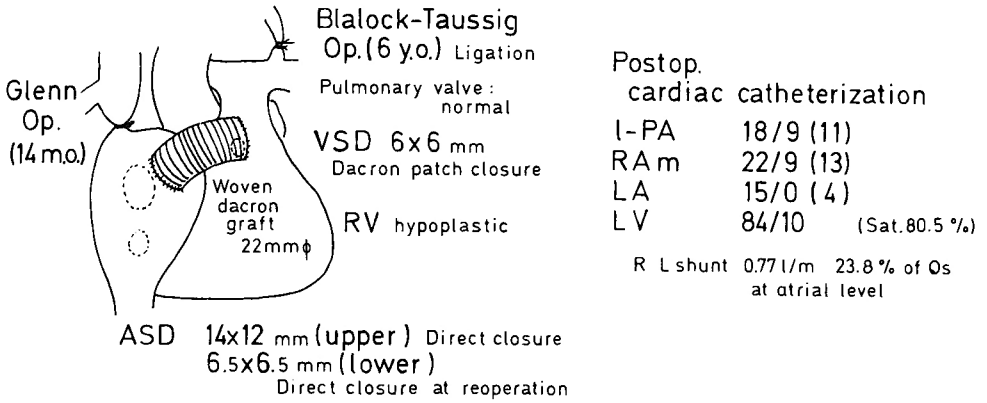

Fig. 2. Surgical procedure and data of the postoperative cardiac catheterization.

respectively. The red cell count was $658 \times 10^4/mm^3$, with a hematocrit value of 52.2%, and a hemoglobin value of 17.7 gm/dl.

On March 3, 1980, a midsternotomy was performed (Fig. 2). There was a palpable thrill over the left pulmonary artery, thus confirming the patency of the previous Blalock-Taussig shunt. Venous drainage was obtained by two cannulas. One cannula was inserted into inferior vena cava (IVC) via the left external iliac vein, and the other was inserted into the superior vena cava (SVC). With the subsequent free access to the right atrial appendage, the Blalock-Taussig shunt was threaded by silk. Moderate hypothermia perfusion by core-cooling was started, followed by cardiac arrest. Cold myocardial cardioplegic solution (250 ml) was infused into the aortic root. In addition, ice slush was placed around the heart. The myocardial temperature was constantly maintained below 15°C, and a left ventricular vent was inserted.

A 3 cm longitudinal incision was made in the atrial appendage. The size of the foramen ovale type atrial septal defect (ASD) was found to be 14×12 mm. The ASD was closed by continuous suture. It was difficult to close the ASD from the short incision of the right appendage, in such a way as to maintain good contraction of the right atrium (RA). The VSD (6×6 mm in size) was closed using a Dacron patch instead of direct closure, in order to maintain right ventricle capacity. Then a woven Dacron vascular graft (22 mm in diameter) was placed between the right atrial appendage and the hypoplastic right ventricle. The well-developed pulmonary valve had three cusps, but no fusion. No additional surgical procedures were necessary as valve function was maintained by using only an autogenous valve in the right heart. The graft was shaped into a circular arc: the external arch was 10 cm long and the internal arch was 3 cm long. The graft was placed just under the midsternotomy. Perfusion ended after 104 minutes, with the patient in a good hemodynamic state.

Sinus rhythm was resumed, and the arterial pressure was 110/80 mmHg, and the venous pressure was 16–20 cm of water. The patient required assisted ventilation with a Servo Respirator 900B for 4 days. Unfortunately, cyanosis did not decreased. As expected, oxygen saturation was 82% in FiO_2 of 0.8. The liver was palpable 2 cm below the costal margin. Respiration with moving of the ala nasi and the mandible and also slight cyanosis on the lips were

seen postoperatively.

On the 4th postoperative day (p.o.d.), intermittent mandatory ventilation (IMV) using a Servo Respirator was started; occasionally intermittent positive pressure brething (IPPB) mixture was used. Three days later the patient was placed in an oxygen tent.

On the 8th p.o.d., oral feeding was started. The arterial pressure was 110/70 mmHg, venous pressure 18 cm of water, Po_2 46.2 (mmHg), Pco_2 46.7 and saturation 84.7%. The respiration with moving of the ala nasi continued for 4 months postoperatively. The oxygen tent was removed on the 16th p.o.d..

On the 21st p.o.d., ^{99m}TC-macroaggregated albumin (MAA) was injected into the cubitus vein, and taken up by the lungs, liver and kidneys. These findings demonstrated the existence of right to left shunt as expected immediately following the previous operation. Also the residual ASD was disclosed by angiocardiography injected in IVC (Fig. 3). Reoperation was considered necessary in order to decrease cyanosis and permit increased physical activity.

But serum hepatitis developed on April 18, 1980, which required 4 months treatment. Values of GOT and GTP were within normal limits on August 20, 1980.

Reoperation of the residual right to left shunt at the atrial level was performed on October 6, 1980. The esophageal temperature of the patient was maintained at 30°C by surface cooling, using a mat and blankets. An arterial line was inserted into the right external iliac artery, and a venous line was inserted into IVC via the right external iliac vein.

Prior to midsternotomy, arterial and venous cannulation was performed in order to control any unexpected bleeding; as anticipated, it was very difficult to prepare the graft just under the sternum. While separating the SVC from the surrounding tissue, massive bleeding, with air leakage, occurred. Perhaps the adhered lung surface or a vessel near the hilus of the lung was damaged. At this time extracorporeal circulation was started, and hemostasis from the midsternotomy approach was performed. Cannulation into SVC could not be performed due to severe adhesion around the SVC. Instead of performing SVC-cannulation, a venous cannulation (NO.

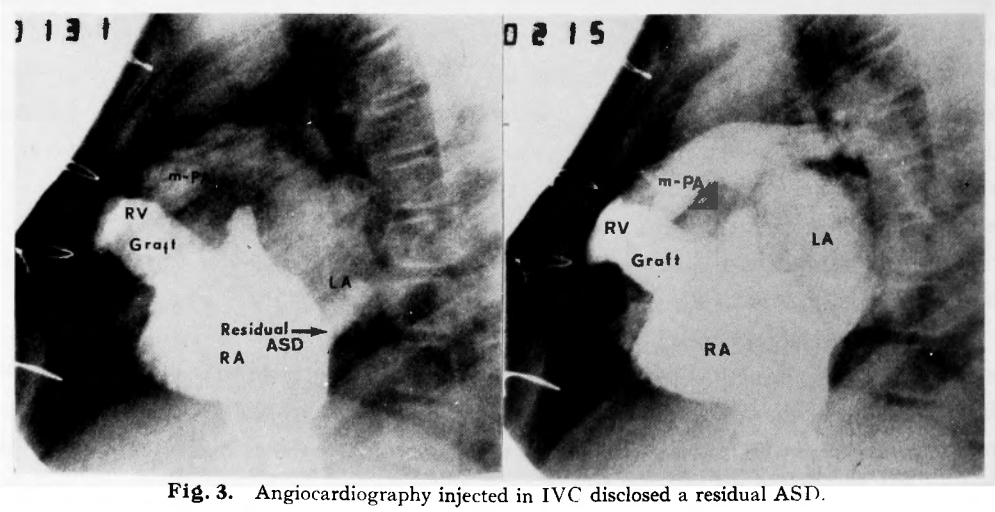

Fig. 3. Angiocardiography injected in IVC disclosed a residual ASD.

21) into innominate vein was performed. The esophageal temperature of the patient was brought down to 20°C by extracorporeal circulation. During circulatory arrest, the closure of the residual ASD was attempted, as total perfusion could not be performed without performing SVC taping. An incision, 4 cm long, was made longitudinally on the previously implanted graft near the ostium of the atrial side, with no damage to the atrial wall. A second ASD, 6.5×6.5 mm, was present 2 cm below the previously closed ASD. This ASD was also a secundum type. Because of the position of the previous incision, it was impossible to discover this second ASD. This ASD was completely closed by direct suture. The incision on the graft was dilated with a Dacron patch.

Postoperatively, the blood pressure was 95/60 mmHg and venous pressure was 24 cm of water. Bleeding from the drainage continued at a rate of 150 to 200 ml/hr. After 6 hours, remidsternotomy was performed in order to stop the hemorrhaging. The bleeding point was not found because adhesion blocked the operative view. Thus the posterior side of the hilus of the lung was covered with a large amount of Oxycel® which was not removed until the patient's condition improved. The midsternotomy was closed after stopping the bleeding from the lung surface and air leakage with many felt mattress sutures.

The following day the blood pressure was 90/50 mmHg, venous pressure 23 cm of water, pH 7.515, Po_2 56.1, Pco_2 36.7 and saturation 91.0% in FiO_2 of 0.90. Urination was adequate due to diuretics. The liver was palpable 6 cm below the costal margin.

On the 3rd p.o.d., blood gas analysis showed pH 7.506, Po_2 47.4, Pco_2 36.7 and saturation 86.6% in FiO_2 of 0.80. Chest roentgenogram showed that neither side of the lung field was clear indicating that the pleural effusion had not decreased. The patient's blood pressure was higher in the left lateral position than in the right lateral position. In the left lateral position the blood could easily flow from IVC into the left lung across the jumping graft. The patient was placed in the Fowler's position to faciliate the return of the blood into the right lung. Morphine hydrochloride was administered in order to decrease pulmonary vascular resistance and sedate the patient.

On the 5th p.o.d., the pleural cavity was opened and the previously packed Oxycel® was removed. Upon opening the right pleural cavity in the fourth intercostal space, the lung and the parietal pleura could not be separated because of adhesion and thus it was not possible to view the hilus of the lung. At this time, the condition of the patient was better than during the previous operation. Another incision in the sixth intercostal space was made to view the hilus of the lung. But air leakage of the lung surface developed at many points. A massive clot and the remaining Oxycel® were removed by the careful stripping off of the large masses and by suction. As an additional clot in the left pleural cavity was suspected from chest roentgenogram, a left thoracotomy was performed in the 4th intercostal space. However, only a little effusion was found. Postoperatively, the respiratory sound was improved and coughing decreased in frequency. In blood gas analysis, Po_2 was 48.8, Pco_2 46.0, saturation 85.4% in FiO_2 of 0.90. Systemic blood pressure was 110/55 mmHg, and venous pressure was 22 cm of water. A massive air leakage from the right lung was found. The liver was palpable 4 cm below the costal margin.

On the 6th p.o.d., subcutaneous emphysema occurred. Respiratory sound was improved, but a little crepitation was continuously audible. As artificial respiration was continued, sedation by morphine hydrochloride was necessary, but occasionally, Myoblock® and Droleptan® were used to inhibit any autogenous respiratory movement which might interfere with the artificial respiration. Air leakage from the surface of the lung continued. Subcutaneous emphysema developed due to obstruction of drainage by fibrin around the internal ostium of the intrathoracic drain. This air leakage may have been caused by the high intratracheal respiratory pressure of the artificial ventilation.

On the 10th p.o.d., pus-like exudation flowed out of the intrathoracic drainage. The patient's temperature was 38–39°C which continued until the 16th p.o.d. As pyothorax developed, washing of the right thorax by saline solution with antibiotics was immediately performed; pus culture was negative. Spontaneous respiration continued, with a Pco_2 between 50–60 mmHg. Rales were present on both lungs. Because of the high frequency of ventilation, the MA-1 Bennet Respirator was replaced by a Servo Respirator 900B.

A massive dose of antibiotics was administered in order to prevent infection of the graft (Cefamezin® 4 gm/day, Panimycin® 200 gm/day, Viccillin S® 6 gm/day and Shiomarin® (Latamoxef, 6059-S 2 gm/day). On the 13th p.o.d., exudation from intrathoracic drain became clear, however, abdominal bulging developed; which may have been due to an increased level of ascites following increased pulmonary resistance. Peritoneal drainage was immediately performed and ascites flowed out at a rate of 50 ml/hr for 3 days.

On the 14th p.o.d., pneumothorax developed.

On the 15th p.o.d., blood gas analysis revealed Po_2 58.7, Pco_2 31.0, saturation 91.9% in FiO_2 of 0.65; air leakage continued. The patient's temperature was from 37 to 38°C; urination was normal and melena was excreted. Fresh blood and medication were given; Tragirol® and Transamin® were effective for hemostasis.

On the 16th p.o.d., isosthenuria (80–90 ml/hr, s.g. 1020–1022) and air leakage continued without change. Drainage from the peritoneal cavity was removed. Intra-lipid® was intravenously infused. Meteorism occurred markedly. Blood gas analysis revealed Po_2 56.3, Pco_2 34.0 and saturation 91.5% in FiO_2 of 0.40. Meteorism gradually disappeared and bowel movement became normal and active.

On the 19th p.o.d., an elementary diet (Elental® 160 gm/day, 600 Kcal) was given daily through a transnasal tube.

On the 22nd p.o.d., IMV decreased to one-half (flow 8 l/min, rate 25/min) and Elental® was increased to 320 g/day (1200 Kcal).

On the 23th p.o.d., the intrathoracic drainage was clamped; as pneumothorax did not occur for 24 hours, the drain was removed.

On the 26th p.o.d., the patient was placed in an oxygen tent (Po_2 55.0, Pco_2 45.0, pH 7.46 and saturation 90.0%) and the intratracheal tube was removed.

On the 27th p.o.d., the patient's mental state became unstable. He wanted to leave his bed and he became irritable. Watching TV was very effective in stabilize his mental state. His

physical and mental condition improved daily. The patient was discharged on March 18, 1981 (5 months postoperatively).

Cyanosis was seen only in the nail bed; this may have been due to the poor ventilation following his previous operation. He is doing well, though being still controlled by diuretics.

Discussion

In 1971, FONTAN[2] reported successful cases of tricuspid atresia in which a complex procedure combining the implantation of two homograft valves with cavo-pulmonary anastomosis. Since then, this procedure has been modified and simplified. BJÖRK[1], MURRAY[7] and GALE[3] reported the simplest method, and recently these methods are called a Fontan-like operation. With the simplification of surgical procedures, this Fontan-like operation is performed in the following congenital heart diseases which have low pulmonary vascular resistance:

1) type Ib tricuspid atresia
2) other types of tricuspid atresia having low pulmonary resistance
3) hypoplastic right heart syndrome
4) certain forms of common ventricle[3]
5) the tricuspid stenosis dominant form of Ebstain's anomaly[6]

The key to success in the Fontan-like operation is the presence of low pulmonary vascular resistance. Therefore, it may be difficult to achieve low mortality when this procedure is performed in infants and young children less than 5 years old. Thus, this group temporarily needs some palliative operations; increasing pulmonary blood flow in order to take preventive action against frequent anoxic episodes and subsequent complications. Some of these patients may undergo several operations; some palliative and some radical. In our case, Glenn operation and the left sided Blalock-Taussig operation were performed in order to alleviate cyanosis at the age of 1 and 6 years, respectively.

Fontan-like operation itself has the following problems:

1) Development of supraventricular arrhythmia and chronic abdominal congestion resulting from postoperative overload of the right atrium.
2) An increase of pulmonary vascular resistance resulting from nonpulsatile flow
3) Development of thrombosis in the conduit
4) Infection, especially when a graft is used
5) Postoperative changes in distribution of pulmonary and bronchial arteries.

However, these problems have not been resolved because of the absence of long-term results from a sufficient number of cases.

Regarding the surgical techniques, there are a few points that deserve consideration. The cannulation of SVC should be performed in such a manner that the contracting parts of the RA are avoided. The cannulation of IVC should be performed at the site of the external iliac vein. When it is difficult to cannulate into SVC, because of severe surrounding adhesion, it may be advisable to cannulate into an innominate vein. But during the closing of ASD, cardiac arrest may be necessary because of the impossibility of total perfusion which results from not taping the

SVC. In reoperation of Fontan-like operation, i.e., a reclosure for residual ASD, a technique of profound hypothermia combining a core-cooling by H-L bypass and surface cooling is advisable. The incision in the RA appendage should be placed near its apex. This principle should be adopted even on reoperation.

Some authors[1,4,7] have recommended that the closure of the ASD, which should always be made with a patch, is somewhat difficult due to the limited incision, but this can be facilitated by a short period of circulatory arrest.

BJÖRK[1] emphasized that in patients with functioning Glenn anastomosis the left side of the heart must be vented adequately after the closure of the ASD, as otherwise the blood from the upper part of the body will directly enter the pulmonary vein through the Glenn anastomosis and overload the left ventricle. In addition, there were some reports of residual ASD after a Fontan-like operation. The majority of these cases were found upon the reoperating of ASD, though the cases in which an other ASD was found were rare. When right to left shunt through the ASD remains after a Fontan-like operation, postoperative management is easy with good systemic pressure, but cyanosis can not be alleviated.

Polysurgery of the chest may result in respiratory disturbance due to ventillation failure, and severe adhesion around the RA may result in disability of contraction. In these cases, good results can be not achieved. In the procedures reported by BJÖRK[1] and GALE[3], no artificial valve or graft were used. Therefore, it is hoped that these methods will eliminate the need for palliative procedure and later reoperation. When a conduit is used, it is advisable not to place it under the sternum, because as the sternum presses the conduit the blood flow is disturbed. Also it is difficult to safely prepare the conduit upon reoperation.

The purpose of the Fontan-like operation is to provide a simple surgical technique, without the use of artificial material, that can be performed for infants and young children and which does not pose any problems as the heart grows.

Finally, it should be emphasized that when prolonged intensive care is necessary, consideration must be given to the patient's mental condition.

Summary

A case of tricuspid atresia Ib, in which Glenn operation (at 14 m.o.), left sided Blalock-Taussig operation (6 y.o.), Fontan-like operation (9 y.o.), closure of a previously undetected ASD (10 y.o.) and two subsequent operations for hemostasis were performed, is described herein. Profound hypothermia combined with surface cooling and core-cooling by H-L bypass is useful and advisable for polysurgery of the heart, especially when cannulation into SVC is impossible due to severe adhesion.

After a Fontan-like operation, if an ASD is still present, systemic blood pressure may be good, but it is difficult to alleviate cyanosis. It should be emphasized that when prolonged intensive care is necessary, consideration must be given to the patient's mental condition.

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和文抄録

6回手術をうけた三尖弁閉鎖症の1例

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三尖弁閉鎖症 Ib 型で1才2ヶ月の時 Glenn 手術，6才の時 Blalock 手術，9才の時 Fontan 様手術，見落した ASD 閉鎖のための再開心術，それに続く2回の止血と多量の Oxycel 除去のための再開胸と計6回手術を行った一例を報告し，その術式と術後管理について言及した．

Fontan 様手術の術後再開心術は，表面冷却法を併用した体外循環下超低体温法で行うと有利であった．又 SVC 脱血は無名静脈から行うと安全であった．

Fontan 様手術々後，遺残 ASD が存在すれば，チアノーゼの改善は認められなかったが，血圧は容易に得られた．ICU 入室期間が長びくと精神療法も必要となる．