

CASE REPORTS

An Unusual Manifestation of Brain Tumor: Development of Delayed Hemiplegia After Cardiopulmonary Bypass

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ABSTRACT Cerebral swelling after cardiopulmonary bypass might trigger a critical cerebral consequence resulting from intracranial space-occupying lesion. We experienced a 75-year-old woman who suffered from a delayed left hemiplegia after mitral valve replacement. Urgent diagnostic imaging revealed the presence of a brain tumor with perifocal cerebral edema. Fluid shifts occurring within a few days after the cardiopulmonary bypass, manifesting the focal cerebral edema, played a key role in this unique clinical course. doi: 10.1111/j.1540-8191.2007.00436.x (*J Card Surg* 2007;22:417-418)

Report of cardiac surgery in patients with a brain tumor is extremely rare.¹⁻³ Cardiopulmonary bypass (CPB) is prone to increase permeability of blood brain barrier and result in cerebral swelling.^{4,5} Brain tumor, even though it is clinically silent, might exacerbate the cerebral swelling due to increased extracellular brain water after CPB. We report a case of patient with meningioma who suffered from a delayed left hemiplegia. Fluid shifts occurring within a few days postoperatively might play a key role in delayed development of perifocal cerebral edema.

CASE REPORT

A 75-year-old woman with shortness of breath on exertion was diagnosed to have mitral and tricuspid regurgitation with severe congestive heart failure. No neurological abnormalities were identified on medical screening. Routine anesthesia and CPB techniques were applied. Mitral valve repair was converted to mitral valve replacement with a mechanical prosthesis. Concomitant procedures included tricuspid annu-

loplasty, left atrial appendage closure, and pulmonary vein isolation. The patient was thrown into low output failure necessitating a large amount of catecholamine. The aortic cross clamp time was 166 minutes and CPB time was 238 minutes. Calculated water retention during CPB was 5267 mL. Minimum value of serum sodium was 123 mequiv/L.

After awaking from anesthesia, her extremities were confirmed to move without paralysis according to our instructions. Then she was put under sedation by continuous administration of propofol until the recovery of myocardial function and correction of water retention. Left upper following lower extremity was gradually paralyzed within two days. On postoperative day 2, when she was weaned from sedation and mechanical ventilatory support, left entire paralysis was observed. A neurological examination identified a normally responded patient with complete left hemiplegia (muscle manual test; 0/5), while her sensory function was normal. An urgent diagnostic imaging revealed a brain mass with perifocal edema in right cerebral hemisphere (Fig. 1). This mass with a diameter of about four centimeter was indicated to be a meningioma. A treatment for the cerebral edema including the administration of glycerin and adrenocortical steroid was started but not sufficient to obtain the satisfactory effect. Then, a surgical treatment was indicated on postoperative day 20. The tumor was totally resected under craniotomy and confirmed to be a transitional meningioma by histological examination.

The left hemiplegia gradually improved and had completely resolved since postoperative month 3. At 28 months after operation, she remains doing well with no sign of recurrence.

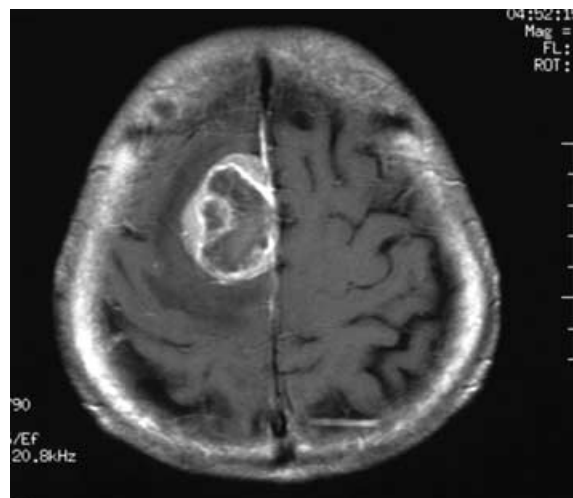


Figure 1. Magnetic resonance imaging of brain shows a mass broadly adjacent to the falx cerebri in the right frontal lobe. Note the severe edema around the mass.

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COMMENT

Experience of cardiac surgery in patients with a brain tumor is extremely rare, reporting on only three cases with meningioma except for pituitary adenoma in the world literatures.¹⁻³ Fortunately, the focal sign or cerebral edema associated with the brain tumor had not been experienced in any case. This is a meaningful report from clinical standpoint that a silent brain tumor induces the delayed hemiplegia related to perifocal cerebral edema after cardiac surgery.

It should be noted that the left hemiplegia appeared after awaking from anesthesia but not during anesthesia and became complete within a few days. This was not attributable to a new onset of cerebrovascular accident, but instead to the perifocal cerebral edema presented by diagnostic imaging. This intriguing clinical course is reflected in the theory that the fluid shifts after CPB occur within the first two to three days postoperatively.¹ This fluid shift might be implicated in the perifocal cerebral edema and result in the development of delayed hemiplegia.

Cerebral swelling after CPB is a well-known complication of cardiac surgery.^{4,5} The mechanism of this phenomenon is supposed to be a decreased plasma osmolarity and increased permeability of blood brain barrier. One of the most important factors to prevent cerebral swelling is the maintenance of normal osmolarity.¹ Animal models with CPB represent that serum sodium level is closely associated with plasma osmotic pressure, which plays an important role in the cerebral edema more than oncotic pressure.^{6,7} Furthermore, maintenance of normal plasma osmolarity by adding hypertonic saline during the CPB is reported to be useful for preventing the cerebral edema.⁶ In this particular case, low sodium level during CPB as well as prolonged CPB time and positive water retention are likely to exacerbate the brain edema.

CONCLUSION

We have reported our experience treating a patient with brain tumor who suffered from the delayed hemiplegia after mitral valve replacement. Fluid shifts occurring within a few days postoperatively might be implicated in the development of perifocal edema and hemiplegia.

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Left Ventricular Rhabdomyoma With Severe Left Ventricular Outflow Tract Obstruction Development of Delayed Hemiplegia After Cardiopulmonary Bypass

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ABSTRACT The incidence of cardiac tumors increased with the improvement of imaging techniques in infants. Rhabdomyomas are the most common tumors in this group of patients. We herein report a 40-day-old male patient with left ventricular rhabdomyoma. The tumor caused syncope attack and supraventricular tachycardia. An emergency operation was planned and the life-threatening lesion was excised via left ventriculotomy. The patient was extubated on postoperative sixth hour and discharged from hospital on the sixth day of the postoperative period without any problem. This successful operation encourages us not to hesitate to perform an operation in newborns with cardiac neoplasms causing hemodynamic instability. doi: 10.1111/j.1540-8191.2007.00437.x (*J Card Surg* 2007;22:418-420)

Cardiac masses called as space-occupying lesions are rare in infancy. Most of these lesions are benign.

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Rhabdomyomas and fibromas are the most common tumors in infants. Myxomas, teratomas, and various forms of sarcoma and angioma have also been reported.¹ Here, we reported an infant suffering from left ventricular rhabdomyoma causing left ventricular outflow tract obstruction.

CASE REPORT

A 40-day-old male infant (four kilograms) was admitted to the hospital with diarrhea. He was cyanotic, dyspneic, and tachycardic. During hospitalization, for fluid replacement and antibiotic therapy, paroxysmal supraventricular tachycardia and syncope attack was observed, although routine physical examination of the healthy looking baby showed no significant findings. The chest X-ray revealed normal findings. On transthoracic echocardiographic examination, a cardiac mass obstructing left ventricular outflow tract was detected (Fig. 1A). An urgent operation was planned. Before the operation, a supraventricular tachycardia attack was detected. Following amiodaron (0.1 mg/kg/min) perfusion the cardiac rhythm normalized.

Surgical technique

The patient was taken to the theater for urgent surgery. Cardiopulmonary bypass (CPB) was initiated by cannulation of the ascending aorta and right atrium. Moderate hypothermia (28°C) was used during CPB. Following diastolic cardiac arrest, an oblique aortotomy was carried out just a few millimeters above the coronary ostia. The cardiac tumor was detected, however, it is impossible to resect the mass from this approach and a ventriculotomy (1.5 cm) parallel to the left anterior descending artery (LAD) was performed. A mass infiltrating left ventricular septum was observed. It has nodules on its surface and it has no capsula. The tumor has no relations with mitral and aortic cusps and either with papillary muscles. $1 \times 2 \times 1 \text{ cm}^3$ big tumor, which obstructs the left ventricular space, was excised from the myocardium (Fig. 1B). Ventriculotomy and aortotomy were closed with running sutures. The patient was transferred with moderate dose inotropic perfusion. He was extubated on postoperative sixth hour and discharged from hospital on the sixth day of the postoperative period without any problem.

Microscopic findings

The microscopic examination of the ventricular mass showed large myocardial cells that have cytoplasm distended by glycogen-filled vacuoles. The pathognomic spider cells were observed (Fig. 2).

COMMENT

The incidence of cardiac tumors increased with the improvement of imaging techniques such as sonography, magnetic resonance imaging, and multidimensional echocardiography.¹ Moreover, imaging techniques have contributed significantly to earlier



Figure 1. (A) Echocardiographic view of the mass obstructing the left ventricular outflow tract; (B) Resection of the rhabdomyoma via left ventriculotomy.

treatment and thus improved survival. Primary cardiac tumors are found approximately in 0.08% of infants and children.^{1,2} Clinical symptoms of a left ventricular tumor are related to cavity obstruction and signs of embolism. Because of quick growth of the mass, congestive heart failure has been reported in many of these cases.²

Rhabdomyoma is the most common intracardiac tumor diagnosed in infancy.¹⁻³ The tumors are usually invading the ventricular myocardium but occasionally protruding into the left ventricular space or freely moving mass.³ Ventricular inflow or outflow tract obstruction, arrhythmia, atrioventricular block, pericardial effusion, and even sudden death have been reported as clinical presentation.^{3,4} Spontaneous regression of a rhabdomyoma has also been reported.^{1,3} Therefore, nonsurgical treatment options are available. However, the presence of severe left ventricular obstruction as in our case is an indication for an emergency lifesaving operation.

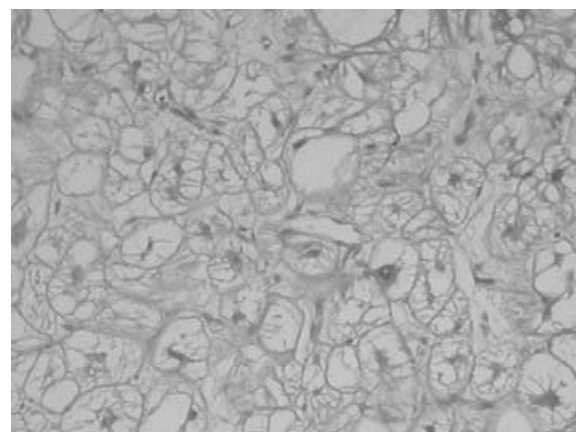


Figure 2. Microscopic evaluation revealed pathognomic spider cells.

On the other hand, rhabdomyomas can be associated with some other congenital diseases. Tuberous sclerosis (TSC) is one of them.³ The association of rhabdomyoma with TSC is well-documented. In our case, the clinical signs are not compatible with TSC. In a meta-analysis of rhabdomyomas in infancy, it is pointed that TSC patients have a higher surgical mortality rate than those without TSC.³ There is a difference in surgical approach between TSC-positive and TSC-negative patients. Nonradical procedures were more often used in patients with clinical signs of TSC, whereas children without TSC symptoms underwent more radical excisions. Tumors with TSC have more tendency of regression.³ In our case, however, macroglossia makes us think about the Beckwith–Wiedemann syndrome characterized with macroglossia, macrosomia, and omphalocele.⁵ In literature review, it has been rarely reported about a combination with this syndrome and rhabdomyoma.¹ Yet, to talk about Beckwith–Wiedemann syndrome, two of above mentioned symptoms are essential.

Rhabdomyomas are frequently multiple, consisting of many tiny nodules less than one millimeter. Our case is unusual because only a single mass was present. The microscopic findings were well-defined, and histological prognostic markers were described completely. Spider cells are pathognomic for rhabdomyoma¹ (Fig. 2).

Isaacs¹ reported that surgical intervention is recommended only for those patients with severe hemodynamic instability or for those with persistent arrhythmias. Neonates with rhabdomyomas but no hemodynamic impairment, or those, in whom only intramural masses can be demonstrated, are not considered surgical candidates. Padolino et al. stated that, for cardiac tumors with extensive invasion, transplantation remains the only therapeutic option.

As a conclusion, in symptomatic infants, surgical excision of the cardiac tumor can be performed with success. For those infants, surgical intervention is a life-saving procedure, although rhabdomyomas have the possibility of regression.

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Anticoagulation After Coronary Artery Surgery in Patients With Polycythemia Vera: Report of Two Cases

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ABSTRACT Polycythemia vera is a myeloproliferative disorder associated with the thromboembolic events. Normalization of the hematocrit and elevated platelet counts is obligatory to reduce the thrombotic risk of patients with PV. Therapeutic strategies include phlebotomy, myelosuppressive agents, and, more recently, interferon- α . In addition, appropriate antiplatelet therapy should be administered to prevent life-threatening complications and reducing the viscosity of the blood. Although aspirin is widely preferred in such patients, this monodrug therapy or combined with clopidogrel as an alternative approach might not be enough, especially after coronary artery surgery. Therefore, warfarin should be added to anticoagulant therapy. This short report describes the use of warfarin, associated with aspirin and clopidogrel as an anticoagulant regimen after coronary artery bypass surgery in two cases with polycythemia vera. We believe that a combination of warfarin with other oral antiplatelet agents may be more effective in preventing the coronary artery bypass graft thrombosis. doi: 10.1111/j.1540-8191.2007.00438.x (*J Card Surg* 2007;22:420-422)

Polycythemia vera (PV) is a myeloproliferative disorder characterized by increased red cell mass in the circulation and frequently associated with the thromboembolic events that more complicate the clinical course of the disorder and are major cause of the mortality and morbidity.¹ Thrombocytosis is present in about 50% of patients with PV.² Thrombotic events are present in 20% to 50% of patients with PV at diagnosis and involve major vessels and the microcirculation.^{1,3} Most often, thrombosis occurs in the cerebral circulation, but it can appear anywhere in the arterial tree, including the heart and coronary arteries.² Thrombotic complications appear to be more often fatal than bleeding complications in these patients and prophylaxis of thromboembolic events remains a key issue

in the management of PV.³ We report two cases of PV who underwent coronary artery bypass grafting (CABG) and warfarin was used as preventing thrombosis in the bypass conduits.

PATIENTS

Case 1

A 46-year-old male patient with PV, who has been treated for two years, was admitted to the cardiology clinic for unstable angina. Coronary angiography revealed subsequent atherosclerotic lesions in left anterior descending (LAD) (80% and 90% stenosis subsequently), Circumflex (CX) (70% stenosis), and right coronary artery (RCA) (80% stenosis). The patient was referred to our clinic for coronary artery bypass surgery (CABG). In his blood count, red blood cell (RBC) volume was 38 mL/kg, platelet count was 450,000 and white blood cell (WBC) count was 12,500. His hematocrit levels were 47%. He had been treated by phlebotomy (by removing one pint/week of blood until his hematocrit level drops below 45%) and aspirin (80 mg/day). He was hospitalized and patient was consulted by Department of Hematology. Phlebotomy was applied (as the hematocrit level was 47%) and hematocrit level dropped to 42%. On physical examination, spleen and liver enlargement was detected. Vitamin B 12 levels were found to be slightly increased.

Case 2

A 61-year-old female patient with PV (who has been treated for three years) was admitted to the cardiology clinic due to stable angina pectoris with positive Treadmill test. Coronary angiography revealed atherosclerotic changes on LAD (80% stenosis in a long segment) and right coronary artery (RCA) (90% stenosis). She was referred to our clinic for CABG. In her blood count, RBC volume was 31 mL/kg, platelet count was 375,000, and WBC count was 10,250. Her hematocrit levels were 40%. Bone marrow testing also confirmed her diagnosis of PV. She had been treated by hydroxyurea and aspirin (80 mg/day).

Operative technique

A median sternotomy was performed in all patients. The pericardium was opened, followed by standard heparinization, aortic cannulation, and venous cannulation (using a double-stage atrial cannula). Cardiopulmonary bypass was started. In situ LITA was always the graft of choice for revascularization of the LAD. LITA was harvested with a pedicle and preserved in a sponge with papaverine to avoid spasm. Radial artery was used as the second graft for Cx artery in the young male patient. Saphenous vein graft was used in the female patient for RCA. Cardiac arrest was maintained by crystalloid cardioplegia under moderate hypothermia. All coronary anastomoses were done using a double armed 7-0 polypropylene suture with a continuous suturing technique. Proximal anastomoses were constructed on the ascending aorta with continuous double-armed 6-0 polypropylene sutures using a side clamp during rewarming.

Postoperative course and follow-up

Heparin infusion started on early postoperative period, then warfarin sodium, clopidogrel, and aspirin were added. Postoperative courses were uneventful. Laboratory tests were in normal limits. Both were discharged on postoperative sixth and eighth day with aspirin (100 mg/day) plus warfarin. Up to now, they had no complication (including cardiac complications, infections, and bleeding due to anticoagulant therapy). The international normalized ratio (INR) was maintained between 2.0 and 2.5 in both patients. The fluctuant levels of INR required more frequent follow-up and were arranged by either telephone interviews or in hospital. Both patients continued their treatments for PV. Both patients were invited to hospital for control angiographies one year after discharge. Both the patients gave consent for postoperative angiographic examination after approval of local ethical committee. Postoperative coronary angiograms of the native coronary arteries and grafts were performed by selective catheterizations and Judkin's technique in four planes; anteroposterior, left lateral, right anterior-oblique, and left anterior-oblique positions. Images were commented by two cardiologists and a surgeon. There was neither occlusion nor narrowing in the grafts.

COMMENT

Patients with PV are threatened by increased risk of arterial and venous thromboembolic events.^{1,2} Arterial ischemic complications occur in 24% to 43% of these patients, particularly those with cardiovascular risk factors.¹⁻⁴ Cardiovascular events were responsible for 40% of deaths in PV patients.⁵ Nonatheromatous arterial thrombosis involves all large and medium-sized vessels, particularly cerebral, limb, coronary and digestive arteries. Juvenile myocardial infarction or rapid postangioplasty coronary thrombosis may reveal certain myeloproliferative disorders.³ Also, venous thrombosis is frequent in PV; superficial or deep venous thromboses are seen. The rate of thrombotic events is strongly dependent on some risk factors, such as increased age, elevated platelet count, elevated hematocrit previous thrombosis, and vascular comorbidity.⁵ Control of red cell mass and antithrombotic therapy appears to be effective treatments in limiting the risk of thrombosis.⁵

The exact mechanism responsible for this increased risk is elusive but a hemostatic imbalance has been implicated.^{1,3} Chuncharunee et al.⁶ evaluated patients with extreme thrombocytosis and found that for similar levels of platelet counts patients with myeloproliferative disorders had an increased risk of thrombotic events. Abnormal platelet aggregation response to various agonists, decreased red cell membrane fluidity with decreased deformability, which increases the susceptibility to occlusion, increased prevalence of antiphospholipid antibodies, increased production of thromboxane A2, and elevated levels of plasminogen activator inhibitor-1 have all been implicated in the pathogenesis of these thrombotic events.^{7,8}

Graft patency is dependent on several factors. Blood flow through the graft is an important factor influencing graft patency.⁹ Faulkner et al.⁹ reported that an increased flow rate was associated with less intimal proliferation. The variables used in estimating the flow rate are graft diameter, graft resistance, and the resistance posed by the native coronary vessel. Blood viscosity is also an important factor affecting flow rate and graft patency.^{2,3} Therefore, antiaggregant and anticoagulation treatments have vital roles. The efficacy and safety of low-dose aspirin (100 mg daily) in PV has been formally assessed in a double-blind, placebo-controlled, randomized clinical trial carried out in the setting of the ECLAP project.⁴ It has been shown in this study that aspirin lowered significantly the risk of a primary combined end-point including cardiovascular death, nonfatal myocardial infarction, nonfatal stroke, and major venous thromboembolism, and total and cardiovascular mortality were also reduced by 46% and 59%, respectively. But major bleeding was slightly increased by aspirin. In this study, an antithrombotic preventive strategy with low-dose aspirin is recommended in all PV patients. Other studies confirm this judgment. However, insufficient data are obtained in the English language literature about warfarin treatment in PV patients. In the study of Bachleitner-Hofmann et al.,¹ some promising results were obtained, but the study was retrospective, did not have a control group, and constituted of limited patient size. Warfarin usage in PV patients after CABG remains controversial. In both of our cases, aspirin plus warfarin treatment was effective and sufficient to prevent the graft thrombosis. There is not a certain study about the long-term effects of warfarin treatment in patients with PV. PV is a condition marked by a risk for both thrombosis and hemorrhage. Although the major risk stems from thrombosis, hemorrhage is unpredictable. Therefore, it may seem that warfarin treatment in these patients can bring some risks. However, we believe that a close follow-up of prothrombin and INR levels may prevent warfarin-related bleeding complications.

This is a small case report including only two patients with polycythemia vera undergoing coronary bypass surgery. Larger number of patients with long-term follow-up results will give more definite results. However, we recommend that warfarin should be added to aspirin and clopidogrel therapy after CABG in patients who have polycythemia vera, because combined warfarin with other oral antiplatelets may have a significant preventive effect against coronary artery bypass grafts.

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A Diagnostic and Therapeutic Dilemma: An Unusual Complication of a Coronary Artery Bypass Surgery

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ABSTRACT We report a diagnostic and therapeutic challenge involving an unusual coronary artery bypass complication. This is the first report of extensive systemic-pulmonary shunts involving the left internal mammary artery, lateral thoracic and intercostal arteries following coronary artery bypass surgery. Although internal mammary to pulmonary arterial fistula are uncommon complications of coronary artery bypass surgery, clinicians should keep this in mind in managing patients with unexplained exertional dyspnea, especially with prior history of intrathoracic inflammation, mediastinitis, and disruption of the pleura. We also discuss various therapeutic approaches including surgical and percutaneous interventions. doi: 10.1111/j.1540-8191.2007.00439.x (*J Card Surg* 2007;22:422-424)

A 63-year-old male underwent coronary artery bypass grafting including bypass of the left anterior de-

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scending artery (LAD) with the left internal mammary artery (LIMA) and three saphenous vein grafts. Postoperatively, the patient developed extensive left hemithorax subcutaneous emphysema with a persistent refractory air leak requiring left upper lobectomy. Subsequently, the patient developed a left anterior mediastinal abscess requiring debridement. Unfortunately, the abscess recurred and the patient required another debridement and left pectoralis flap.

During the next three months, the patient experienced increasing exertional dyspnea and angina pectoris. Repeat coronary angiogram demonstrated extensive communication of the LIMA to the pulmonary arterial circulation via a plexus of small vessels (Fig. 1). The fistulous LIMA to pulmonary flow was interrupted by insertion of a covered stent of 16 mm length and 3 mm diameter (JoStent, Abbott Vascular). There was significant improvement in the LIMA flow; however, the LAD flow remained very poor. At the time of the coronary angiogram, a very prominent lateral thoracic artery was visualized and this prompted further imaging. A computed tomography angiogram revealed numerous fistulas between the systemic and pulmonary arterial and venous circulation, originating from branches of the subclavian and lateral thoracic arteries as well as from multiple intercostal arteries (Fig. 2).

Percutaneous embolization of multiple branches of the lateral thoracic, subclavian, thyro-cervical trunk, and multiple intercostals arteries was performed. Shunts to both the pulmonary arterial and pulmonary venous systems were also embolized with larger size embolic agents. Despite this intervention, the patient remained dyspneic with mild exertion. A nuclear shunt study was performed and showed a normal right lung field with early recirculation in the left lung field consistent with residual systemic-pulmonary shunting. Because of the patient's reduced pulmonary reserve secondary to chronic obstructive lung disease, we elected to treat him medically. At five years following the initial coronary artery bypass surgery, the patient is alive but remains incapacitated by moderate exertional dyspnea.

DISCUSSION

Internal mammary artery to pulmonary arterial fistulas are rare. They can be either congenital or acquired. The acquired type had been described in hematologic neoplasia, chronic granulomatous disease,¹ trauma, and chest surgery.²

The first report of acquired internal mammary to pulmonary arterial fistula after CABG was published in 1990 by Johnson et al.³ At least 14 other cases of internal mammary artery to pulmonary arterial fistulas following CABG have been reported.¹⁻¹⁴ However, this is the first report of extensive systemic-pulmonary shunting involving the LIMA, lateral thoracic and multiple intercostals as a CABG complication.

It has been suggested that the actual prevalence of internal mammary to pulmonary artery fistulas is possibly higher than that reported in the literature, since not all of these shunts may have enough hemody-

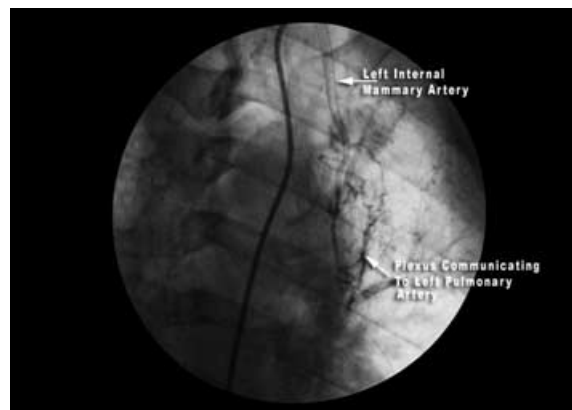


Figure 1. *Fistula of the LIMA to the pulmonary arterial circulation.*

namical impact to cause symptoms.⁴ Most patients presenting with recurrent angina following CABG are thought to have progression of their atherosclerotic coronary disease and are treated medically without undergoing diagnostic coronary angiography.

The etiology of the LIMA-pulmonary artery fistula remains unclear. It has been suggested that disruption of the pulmonary pleura by trauma, repeat sternotomy or metal clips can lead to fistula formation.⁴ Coagulation without clipping side branches of the internal mammary pedicle was also proposed as a potential cause of fistulas.² Kimmelstiel⁶ and Musleh⁷ suggested that intrathoracic inflammation may lead to neovascularization and fistula formation. Najm et al. reported redo CABG in 50% of patients with LIMA-LAD fistulas.⁵ There had been no previous report of extensive systemic-pulmonary shunts involving LIMA, intercostals, and lateral thoracic arteries similar to ours. It is highly possible that the formation of these pulmonary-systemic fistulas in this patient was secondary to the extensive intrathoracic inflammation from the recurrent mediastinitis and the lobectomy.

Various therapeutic approaches including surgical ligation of the fistula, percutaneous intervention, and

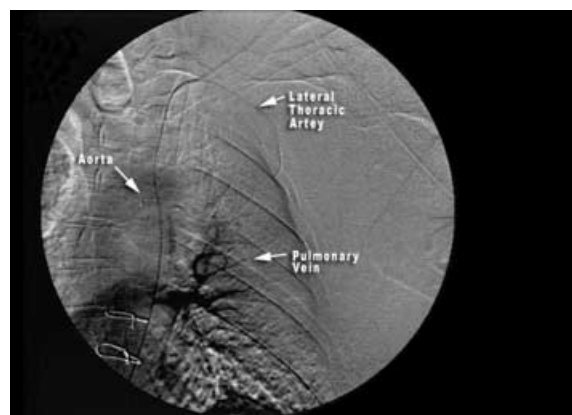


Figure 2. *Angiogram of the left lateral thoracic artery showing fistulas from the lateral thoracic artery to the pulmonary venous system and the aorta.*

conservative medical management for treatment of LIMA-LAD fistulas had been reported. It was initially suggested that all arteriovenous fistulas involving the internal mammary vessels should be surgically closed.³ Blanche et al. and Najm et al. suggested that therapy should be guided by the severity of symptoms, and that conservative management might be adequate in asymptomatic patients with small shunts.^{4,5}

During the last decade, the use of percutaneous embolization has increased.¹²⁻¹⁴ One of the potential complications with percutaneous embolization was the risk of embolizing microparticles into the coronary circulation.¹² Modern microcatheters and the use of newer microcoils have reduced the risks of reflux or protrusion of the coils into the coronary circulation. For our patient, there was a potential risk of embolic agents traversing the pulmonary veins and secondary peripheral embolization. Finally, if the causal stimulus for the shunts was not removed, these shunts may recur despite an apparently successful embolization. This may have happened in this case, since the patient remained dyspneic following the embolizations.

This report emphasized the fact that common symptoms may be the first manifestations of uncommon conditions. Although internal mammary fistula to pulmonary artery is rare, clinicians should keep this in mind when faced with patients with unexplained exertional dyspnea following CABG, especially in patients with prior recurrent history of intrathoracic inflammation or trauma.

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Successful Graft Patency After Factor VIIa Use for Bleeding in Coronary Artery Surgery: An Unusual Complication of a Coronary Artery Bypass Surgery

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ABSTRACT There is a growing literature regarding the use of recombinant activated factor VII (rFVIIa) (NovoSeven, NovoNordisk, Copenhagen, Denmark) to control refractory hemorrhage after cardiopulmonary bypass in cardiac surgery. This supporting evidence is mostly documented as case reports on patients, of which only six involve coronary artery grafting. We present our experience of a patient undergoing coronary artery bypass grafting, who suffered an anaphylactic reaction to protamine preventing safe reversal of systemic heparinization. Subsequent management of postoperative bleeding was successfully achieved only after rFVIIa administration. doi: 10.1111/j.1540-8191.2007.00440.x (*J Card Surg* 2007;22:424-426)

Resternotomy for bleeding following surgical procedures on cardiopulmonary bypass (CPB) carries significant morbidity and mortality. Administration of recombinant activated factor VII (rFVIIa) has been successfully used in several cases to control hemorrhage

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when conventional use of blood products and pharmacological agents were ineffective. We present a patient in whom rFVIIa was used to control bleeding after coronary artery bypass grafts.

A 69-year-old gentleman presented in December 2004 with central chest pain. He had a background of one previous myocardial infarction, insulin dependent diabetes mellitus, and lithotripsy for right renal calculi. His ECG showed left bundle branch block and the troponin peaked at 0.19. A non-ST elevation myocardial infarction was diagnosed. Coronary angiography revealed an occluded right coronary artery, 60% and 90% stenoses of the proximal and distal left circumflex artery, respectively, 80% stenosis of the intermediate branch, and 70% stenosis of the left anterior descending artery. He was referred for coronary artery bypass grafting, which took place electively six weeks later.

The patient had no history of coagulopathy. Aspirin and clopidogrel had been stopped over three weeks before surgery. Preoperative hematological investigations were within normal limits (Hb 13.6, WCC 5.6, Plt 150, INR 0.9, APTTR 1.07, TT 14[†]).

After systemic heparinization with 33,000 units of heparin, CPB was established between the right atrium and ascending aorta. Tranexamic acid 2 g was given. The patient was cooled to 34°C. The radial artery was anastomosed to the distal right coronary, two saphenous vein grafts to the intermediate and first diagonal vessels, and the left internal mammary to the left anterior descending artery.

After rewarming, the patient came off CPB easily, with low filling pressures and no inotropic support. Shortly after administration of 200 mg protamine the patient developed profound hypotension and concomitant elevated airway pressure. An anaphylactic reaction to protamine was suspected. There was no response to adrenaline and metaraminol, so the patient was re-heparinized and CPB was reinstituted. A noradrenaline infusion was commenced and 200 mg hydrocortisone was administered. A left atrial line was inserted. Following 20 minutes of rest on CPB, the patient was taken off CPB with good hemodynamics, and the noradrenaline infusion weaned within 10 minutes. Floseal was applied to all the anastomotic sites and meticulous hemostasis was performed in view of the inability to safely reverse the heparin. Aprotinin was administered at a dose of 2 million units. Four units of fresh frozen plasma (FFP) and two pools of platelets were administered at this stage.

Following transfer to the cardiothoracic intensive therapy unit (ITU), the patient continued to bleed into the drains and the total blood loss for the first two post-operative hours was 1880 mL even though an aprotinin infusion had been running at 50 mL/h and further blood products were administered (4 units of FFP, 10 units of cryoprecipitate, and 3 pools of platelets). He remained coagulopathic with an INR 1.6, APTTR > 5, and TT > 50.

After consultation with the hematologist two doses of 30 µg/kg rFVIIa were given one hour apart. The drainage then rapidly decreased to 180, 20, 60, and 60 mL in the subsequent hours.

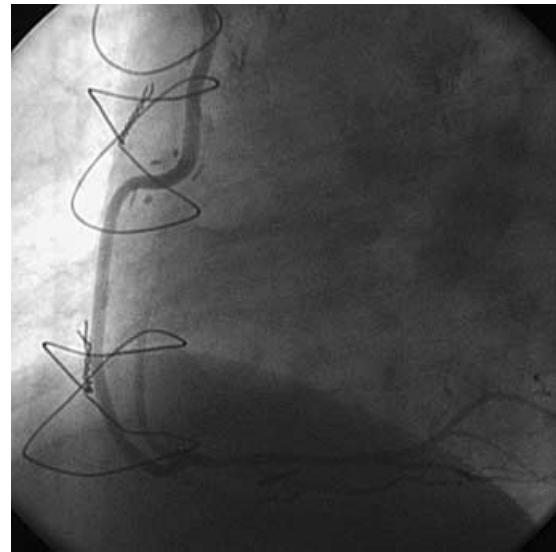


Figure 1. Patent radial artery to right coronary artery graft.

By the next morning (12 hours after rFVIIa), the patient's hemodynamics were stable, drain output minimal and clotting parameters had virtually normalized: INR 0.8, APTTR 1.22, TT 15, Hb 8.0, Plt 126. Only a further four units of FFP were required after the second dose of rFVIIa. The patient was extubated on day 1 and proceeded to have an uneventful remainder of recovery. Subsequent immunological investigations confirmed the allergy to protamine.

A coronary angiogram at three months showed that all the grafts were patent (Figs. 1-3).

COMMENT

In our case, the bleeding was refractory to meticulous hemostasis, the administration of blood products (cell saver, FFP, platelets) and aprotinin. The use of

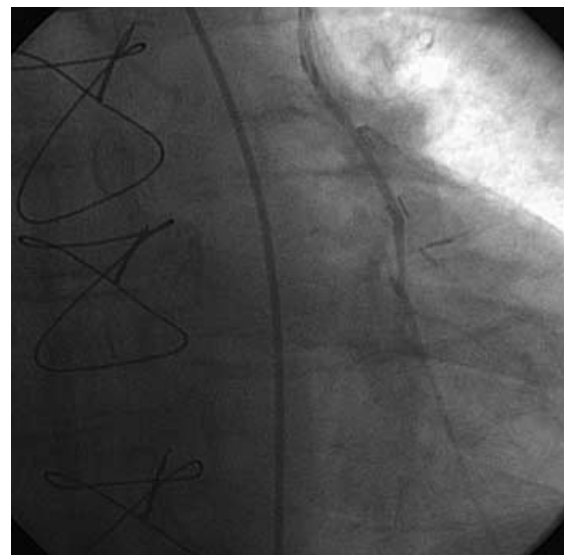


Figure 2. Patent left internal mammary artery to left anterior descending artery graft.

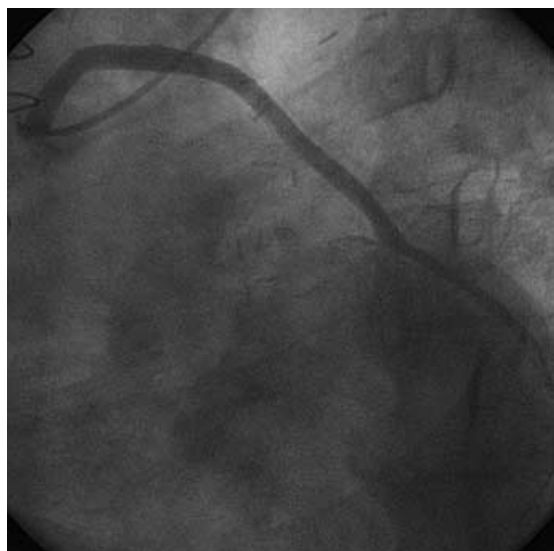


Figure 3. Patent saphenous vein to diagonal branch graft.

rFVIIa minimized the use of further blood products, a protracted ITU stay and the significant morbidity and mortality associated with re sternotomy for bleeding.

Including this report, there are 29 cases documented in the literature regarding the successful use of rFVIIa following postoperative bleeding after procedures on CPB¹⁻⁴ (Herbertson reported a further 17 cases without specific details).⁵ These reports declare that there were no adverse consequences following rFVIIa administration—no reported thromboses, and in particular no coronary thrombosis in the seven reported CABG patients. However, 3 of these 29 cases did die: day 2 post-CABG with multiorgan failure; day 10 post-CABG with decreased cardiac output, multiorgan failure, sepsis; day 34 post-mitral valve replacement with sepsis.

The only case of fatal thrombosis involving surgery on CPB involves a lung transplant patient, who required support by extracorporeal membrane oxygenation as CPB was unsuccessfully terminated.⁶ There rFVIIa was combined with activated prothrombin complex concentrate (APCC). No autopsy was performed, but massive intracardiac and ECMO circuit thrombosis was cited as the cause of death, precipitated by the APCC. The combination of rFVIIa and APCC has been implicated in other cases of thromboembolism not involving CPB including pulmonary embolism in a factor VIII-deficient patient.⁷

However, one cannot dismiss that there are potential thrombotic risks with rFVIIa since acute myocardial infarction has been reported in a patient with hemophilia A given rFVIIa and given the postulated mechanism of action.^{8,9} The rFVIIa is believed to interact with its cofactor tissue factor (TF), expressed at sites of tissue injury. This subsequently stimulates factor Xa generation from factor X, with subsequent thrombin generation. Activated platelets may also play a role in interacting with rFVIIa and provide another pathway for thrombin generation. Furthermore, rFVIIa may stimulate antifibrinolytic activity via activation of in-

hibitors to fibrinolysis. Given the fact that TF will be expressed at coronary graft anastomoses and at unstable atheromatous plaques, graft occlusion is indeed a valid concern.

Our case is invaluable in that it is the first to show that coronary graft patency is maintained following rFVIIa administration. Future cases of CABG patients who received rFVIIa should also be followed up with coronary angiograms to check graft patency, before rFVIIa can be deemed safe to use in CABG patients.

There are not yet any prospective trials nor any standard treatment protocols concerning at what stage and at what dose rFVIIa should be administered. Halkos et al.'s recent retrospective study of nine patients considered rFVIIa use only when 16 U platelets, 4 U plasma, and 10 U cryoprecipitate, whereas other cases opted for rFVIIa at a much earlier stage.¹ The doses used in cardiac surgery elsewhere in the literature vary between 13 and 120 $\mu\text{g/kg}$, most opting for 90 $\mu\text{g/kg}$.

Prospective studies with a standard protocol for administration of more traditional hemostatic agents and blood products before rFVIIa need to be established. The results of Herbertson's randomized controlled trial administering rFVIIa (90 $\mu\text{g/kg}$) or placebo at the end of CPB are eagerly awaited. The efficacy and possible detrimental effects of using higher doses than those we used also need investigation. This is of further importance given the expensive cost of rFVIIa.

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A Case of a Large Intrapericardial Lipoma Occupying Pericardial Space An Unusual Complication of a Coronary Artery Bypass Surgery

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ABSTRACT Although cardiac lipomas are quite rare, they represent the second most common primary benign cardiac tumors. In this case report we present a 71-year-old man with a large intrapericardial lipoma originating from pericardium. In this case, intrapericardial lipoma was diagnosed by echocardiographic methods (transthoracic and transesophageal echocardiography) and imaging techniques such as computed tomography, magnetic resonance imaging. The patient had an uneventful postoperative recovery. She is currently asymptomatic and has no evidence of recurrence at 12 months follow-up. doi: 10.1111/j.1540-8191.2007.00441.x (*J Card Surg* 2007;22:427-429)

Cardiac lipomas are benign tumors and account for approximately 10% of all primary cardiac tumors.^{1,2} Cardiac lipomas can originate from endocardium, myocardium, and pericardium.³⁻⁸ Most of them are found incidentally at the time of autopsy, echocardiography, or cardiac catheterization. Although pericardial lipomas may occasionally result in signs and symptoms of heart diseases, the majority of the cases are asymptomatic. In general, cardiac signs and symptoms depend on the size and location of the tumor. These patients may present with fatigue, malaise, palpitation, chest pain, dyspnea, syncope, and sudden death.⁹⁻¹² Large tumors may affect atrial and ventricular function or compress mediastinal structures due to a mass effect.

We report the unusual case of a patient who presented with a large intrapericardial lipoma occupying pericardial space. The diagnosis of intrapericardial



Figure 1. Transesophageal echocardiogram showing short-axis view of oval-shaped mass (arrows) on the posterolateral wall of the left atrium and left atrial appendage.

lipoma was made by transthoracic (TTE) and transesophageal echocardiography (TEE), as well as by computed tomography (CT), magnetic resonance imaging (MRI), and cardiac catheterization.

CASE REPORT

A 71-year-old man was admitted because of effort dyspnea, chest pain, and palpitations. He had been in good health until one month earlier. His medical history included systemic hypertension and smoking. On examination, his heart rate was 120 beats/min, and blood pressure was 155/95 mmHg. The physical examination, apart from tachycardia, was unremarkable. Heart sounds were normal, no murmur or pericardial rub was heard. Standard 12-lead electrocardiogram revealed sinus tachycardia without any remarkable abnormality. Chest X-ray was normal cardiothoracic ratio, but pulmonary artery trunk was markedly large. TTE demonstrated a large intrapericardial mass compressing the posterior and lateral wall of the left atrium and left ventricle that appear to be noninvasive. The mass compressed the left atrium. Left ventricular systolic function was normal, with ejection fraction estimated at 62%. TEE revealed an echogenic homogenous ovoid mass located along the posterior and lateral surface of the left atrium and left atrial appendage (Fig. 1). This examination also demonstrated that the left atrial and atrial appendage contours were greatly deformed by the mass. In order to confirm and amplify the echocardiographic findings, the patient was performed CT and MRI examination. Thoracic CT showed a mass measuring 3.5 × 10 cm extending posterolateral of the left atrium and left ventricle (Fig. 2). MRI scan demonstrated that the density of the large intrapericardial mass was consistent with lipomatous tissue (Fig. 3). Coronary angiography revealed normal coronary arteries and did not identify any artery feeding the mass. Evaluation of cardiac catheterization showed that intraventricular end diastolic pressure was within normal but pulmonary artery wedge pressure was mildly increased.

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The patient underwent surgery to remove the mass under the diagnosis of pericardial lipoma. During the operation, a large, yellow, soft, encapsulated subepicardial tumor was observed covering up the inferolateral aspect of the heart. The mass was lobulated and pediculated in shape and it had been connected to the posterolateral surface of the left atrium through a pediculum. At the operation, the mass was resected easily. The mass measured 10 cm by 9.5 cm by 3.5 cm. Biopsy specimen consisted of lobulated mature adipose tissue macroscopically, and the histopathology of the mass was of a lipoma with no evidence of malignancy, it was composed of fatty cells. After one year, he was asymptomatic. No signs of recurrence were detected on echocardiography.

DISCUSSION

Lipomas are benign cardiac tumors and have rarely been encountered.^{1,2} They are usually asymptomatic and may occasionally cause symptoms. Most are diagnosed incidentally at autopsy, echocardiography, or cardiac catheterization. They can arise from anywhere in the heart.³⁻⁸ The most commonly involved chambers are the left ventricle and left atrium. The lipomas may originate from in the pericardial space. Cardiac lipomas can affect atrial and ventricular function secondary to the mass effect, and result in symptoms such as fatigue, malaise, palpitation, chest pain, dyspnea, syncope, and sudden death.⁹⁻¹² Lipomas are polypoid or sessile masses macroscopically, and are also encapsulated masses consisting of mature fat cells with varying amounts of fibrous and myxoid tissue.

Echocardiographic methods and imaging techniques such as CT, MRI, and ultrasound allow the diagnosis of cardiac masses.¹³⁻¹⁶ Patients with these diseases are generally difficult to image with conventional transthoracic two-dimensional echocardiography. However, TTE is usually the initial diagnostic method to be performed in patients with a suspected cardiac mass. Tissues characterization of a mass remains difficult to obtain by TTE. In our case, TTE revealed that an echogenic ovoid mass was overlying the



Figure 2. CT scan showing a low-density mass extending posterolateral of the left atrium and ventricle (arrows).

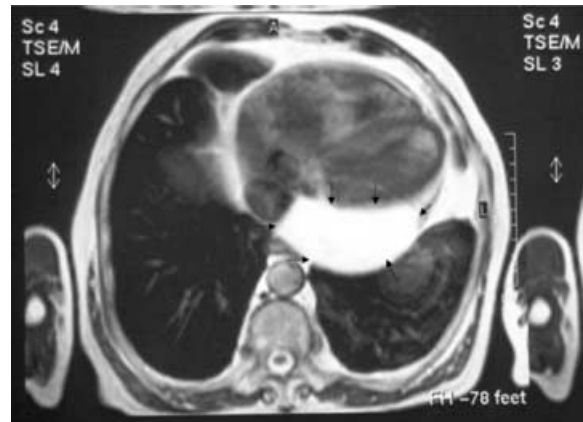


Figure 3. T₁-weighted MR image showing a high signal intensity of the mass (arrows).

posterolateral wall of the left atrium and the left ventricle. TEE can provide high-quality imaging of intracardiac and extracardiac structures in patients who have insufficient acoustic windows. TEE clearly showed an intrapericardial mass on the posterior and lateral surface of the left atrium and left atrial appendage. On TEE examination, the mass did not also appear to be invasive, but the left atrial appendage partly appear compressed. As in the case reported in this article, recent imaging studies including CT and MRI are used to confirm the echocardiographic findings.^{15,16} These imaging techniques are noninvasive means of evaluating cardiac tumors. They provide tissue characterization of the lesion, and are accurate in diagnosis of cardiac lipoma. Especially, MRI examination can demonstrate the lipomatous nature of a mass and can delineate more precisely its spatial extension. Cardiac lipomas are diagnosed by MRI on the basis of their low attenuation and can be distinguished from cardiac lesions, such as myxoma and thrombus, which have higher attenuation. Lipomas are homogeneous and display lower signal intensity than subcutaneous fat. Thorax CT of our patient demonstrated a large homogeneous ovoid mass, covering the lateral wall of the left atrium and left ventricle. MRI revealed that the mass greatly did not deform the left ventricle and left atrial contour and it had low attenuation. Coronary angiography can determine coronary anatomy, may show a vascular network arising from a coronary artery, and also it may guide the surgical approach. In this case, coronary angiography demonstrated that coronary arteries were normal and the left ventricle had normal contraction.

In our patient, complete excision of the tumor was considered possible, because the mass is not impinging on vital structures. At operation, a large anterior intrapericardial mass was found that was not adherent to the parietal pericardium and that was attached by a stalk on the posterolateral wall of the left atrium. The mass was resected successfully. The mass was a large encapsulated yellow lobulated soft-tissue mass that measured 10 × 9.5 × 3.5.0 cm. During 12-month postoperative follow-up period, the patient remained

asymptomatic and is in good condition. No recurrences were described until now.

In conclusion, we present the unusual case with a large intrapericardial lipoma occupying pericardial space. The combination of echocardiographic methods and imaging techniques allowed a precise diagnosis in terms of the localization and tissue characterization of the tumor. CT and MRI provide important information for cases in which a cardiac mass is suspected. These methods describe the appearance of primary and secondary cardiac masses and allow for detailed delineation of intracardiac and extracardiac masses, their extent, and their influence on cardiac function.

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Successful Cardiac Surgery in a Former Conjoined Twin With Heart Malformation and Pulmonary Hypertension An Unusual Complication of a Coronary Artery Bypass Surgery

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ABSTRACT Pulmonary hypertension was diagnosed in a former conjoined twin presenting with a ventricular septal defect, moderate congestive heart failure, and dyspnea with hypercapnia due to instable bony chest. He underwent successful corrective cardiac surgery and chest wall reconstruction at the age of 12 months. Subsequently, mechanical ventilation was performed for three months to prevent right heart failure and pulmonary hypertension until thoracic stability allowed the tracheostoma to be closed. doi: 10.1111/j.1540-8191.2007.00442.x (*J Card Surg* 2007;22:429-431)

CASE REPORT

Conjoined twins are rare, with an incidence that varies from 1:50,000 to 1:200,000 newborns.¹ Of these rare defects, thoracopagus is the most common variety (40%), and this is mostly lethal since union of the cardiovascular systems occurs in the majority of cases.² No previous reports are available of a former thoracopagus twin undergoing successful cardiac surgery for congenital heart defects weeks or months after successful separation.

As reported in the Chinese Journal of Surgery, thoracopagus twin boys were successfully separated at the age of 45 days by Dr. D. Li at Fujian Medical University, China.³ Both children had shared the pericardium, diaphragm, and liver parenchyma (Fig. 1). The chest junctions were at the sternum and the inferior pairs of the ribs. One boy underwent surgical separation without severe complications and was discharged home some months later. However, the other boy remained in hospital with a multitude of problems; the large chest defect was closed only with a polyester pericardial patch graft and a silicon supporter. With his congenital heart defect, his diaphragmatic dysfunction and the unfixed sternum, extubation, and spontaneous breathing failed,

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Figure 1. The thoracopagus twin boys before surgical separation. Reproduced with kind permission from Dr. D. Li, Department of Pediatric Surgery, Fujian Medical University, Fuzhou, China.

and he underwent tracheotomy. After 11 months of hospitalization, cyanotic crisis occurred, the hemodynamics became impaired, and the child was transferred to the Deutsches Herzzentrum, Berlin.

With a body weight of seven kilograms and an oxygen saturation of 88% (FiO_2 , 0.4), the child had chronic hypercapnia with arterial CO_2 of 80 mmHg. The X-ray showed a dextropositioned enlarged heart and asymmetrical bony chest (Fig. 2). Echocardiography revealed a 20-mm perimembranous VSD, an atrial septal defect, and a mild infundibular stenosis with a gradient of less than 10 mmHg.



Figure 2. The X-ray showing the dextropositioned enlarged heart and asymmetrical bony chest.

Because of evidence of existing pulmonary hypertension, cardiac catheterization was performed. Systolic pressure in the left ventricle was 111 mmHg, and in the right ventricle 105 mmHg, mean pulmonary pressure was 73 mmHg, and mean left atrial pressure was 7 mmHg. Pulmonary hypertension still responded to mild hyperventilation and oxygen supply (Rp:Rs was 1.4:1, with FiO_2 0.4, and decreased to 0.63:1, with FiO_2 1.0).

A spiral CT confirmed the dextroversion cordis combined with the large thoracic wall defect with missing corpus sterni and deformation of the right-sided ribs (Fig. 3). Because of the aberrant rotated and dextroverted heart, an uncommon right lateral incision was chosen. Surgical correction with patch closure of the VSD, infundibulectomy, and reconstruction of the thoracic wall was performed without problems. Within the following five days, pulmonary hypertensive crisis and right heart failure were treated with catecholamines, mild hyperventilation, oxygen supply, and nitric oxide.

The child was retransferred to the hospital in China, where artificial ventilation proceeded for the next few months to prevent PHT. Three months after corrective surgery and chest wall reconstruction, the patient could be weaned from mechanical ventilation and the tracheostoma was closed. At follow-up four years later, the child is living with his twin brother and his parents in Fujian, China. He is breathing spontaneously, without pulmonary hypertension, and is thriving, with psychomotoric development concurrent with his age and only mild thoracic deformation.

COMMENT

Few reports are available in the literature of thoracopagus twins who both survived surgical separation because of the serious cardiac anomalies in about 75%,⁴ which may prevent separation or affect survival. To the best of our knowledge, there is no case of successful corrective cardiac surgery for congenital

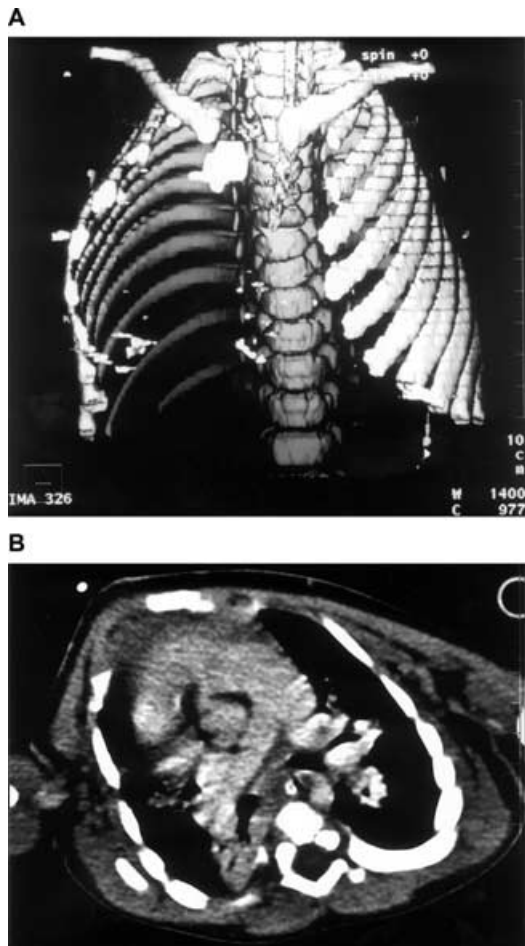


Figure 3. Spiral CT showing the absent corpus of the sternum, the absent processus xiphoideus, and the right-sided rib deformations as a 3D reconstruction (A) and in an axial view (B).

heart disease among surviving former conjoined twins undertaken weeks or months after separation.

In our case, a 1 year-old former thoracopagus twin with VSD, pulmonary hypertension, and only mild infundibular stenosis underwent successful corrective cardiac surgery, although severe PHT was triggered by left-to-right shunt via a large VSD and chronic hypoventilation with hypercapnia due to an instable bony chest.

Within the past 20 years, conjoined twins have almost vanished from our neonatal units because they

are usually diagnosed in the first trimester and such pregnancies are normally terminated before 20 weeks of gestation. A higher prevalence of congenital malformations among twins than among single births has been reported.⁵ Nazer found that in stillbirths there was a significant difference between single and twin stillbirths (7.1% vs. 35.2%), and natural selection seems to work against malformed twins.⁶ Despite the unusual nature of our case, the high standards in surgery and intensive care and our experience in treating VSD and PHT resulted in a favorable outcome. Accurate prenatal diagnosis of complex anatomic connections and associated anomalies has only become possible recently with the use of modern ultrasonography, echocardiography, and fetal magnetic resonance imaging. These advances in prenatal diagnosis allow detailed, accurate evaluations of conjoined twins.⁷ Careful prenatal studies and optimal postnatal management involving different specialties may uncover cases in which separation is life-saving for both twins.

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