Balloon-Expandable Stenting, Percutaneous Coil Embolization, and Amplatzer Septal Occluder; Treatments after Complicated Lateral Tunnel Fontan : A Case Report

WORAKAN PROMPHAN, MD*,
KRITVIKROM DURONGPISITKUL, MD*,
SAMPHAN PORNVILAWAN, MD**,
SOMCHAI SRIYOSCHATI, MD**
JARUPIM SOONGSWANG, MD*,
DUANGMANEE LAOHAPRASITIPORN, MD*,
APICHART NANA, MD*,

Abstract

The authors report a 7-year-old girl with univentricular heart physiology who developed prolonged pleural effusion due to discrete narrowing of the proximal right pulmonary artery, and progressive cyanosis which resulted from leakage of the atrial baffle, multiple veno-venous collaterals after the lateral tunnel Fontan operation. Percutaneous balloon- expandable stent implantation was used to correct the right pulmonary artery stenosis with an excellent result. Cyanosis was improved by coil embolization of the collaterals and occlusion of the baffle leakage with Amplatzer septal occluder. This is the first successful report in Thailand.

Key word : Fontan Operation, Amplatzer Septal Occluder, Stent Implantation, Coil Embolization

PROMPHAN W,
SOONGSWANG J, DURONGPISITKUL K, et al
J Med Assoc Thai 2003; 86: 369-375

The lateral tunnel Fontan operation, which is a modification of the classic Fontan operation, has shown good short[1] and long-term[2] results for patients with univentricular physiology. Complications may occur early as well as late in the postoperative periods. Elevated pulmonary arterial resistance[3] or Fontan pathway obstruction[4] may manifest as fluid retention in the third space. Persistent

* Division of Pediatric Cardiology, Department of Pediatrics,
** Division of Cardiothoracic surgery, Department of Surgery, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand

pleural effusion accounted for 10 per cent of the Fontan patients who died without hemodynamic insufficiency(5). A residual communication or fenestration between systemic and pulmonary venous returns, such as: venous collateral from superior vena cavae to pulmonary vein may cause persistent arterial desaturation with a potential towards paradoxical embolism(6). The authors describe a post lateral tunnel Fontan patient in whom moderate risk preoperative Fontan hemodynamic with narrowing of the proximal right pulmonary artery (RPA), and leakage of the atrial baffle with multiple veno-venous collaterals that was successfully treated with balloon-expandable stent, Amplatzer septal occluder, and percutaneous coils embolization.

CASE REPORT

The patient had an uneventful perinatal history and was delivered normally in 1994. She has had severe cyanosis since birth. Physical examination revealed cutaneous oxygen saturation of 70 per cent, single second heart sound, no heart murmur, and no hepatomegaly. Chest roentgenogram showed no cardiomegaly but decreased pulmonary vasculature. An electrocardiogram demonstrated two forms of P wave (P axis +75 degree and -105 degree), QRS axis +135 degree, poor R progression in the left chest leads, absent Q wave in V5, V6. Echocardiogram revealed situs inversus, isolated levocardia, common atrium, common right ventricular morphologic ventricle, d-transposition of the great vessels, and pulmonary atresia. Cyanosis was improved by 4 mm of right modified Blalock Taussig (BT) shunt on day-2 of life. Cutaneous oxygen saturation was improved from 70 to 90 per cent post-operation. Her growth was around 25 percentile until 2.5 years of age.

Pre-operative right cardiac catheterization was undertaken and showed mean atrial pressure of 5 mmHg, and pulmonary vein wedge pressure of 12 mmHg. Ventricular end diastolic pressure was 7 mmHg. Oxygen saturation in the aorta was 81 per cent. Systemic blood flow and pulmonary blood flow ratio was 1.3. Pulmonary vascular resistance (PVR) was 2.3 Wood unit.m^2 and McGoon ratio was 2 : 1 with mild proximal right pulmonary artery (RPA) stenosis. There was no significant atrioventricular valve regurgitation. She underwent lateral tunnel Fontan (with Gortex^{TM} patch) operation and right modified BT shunt was taken down at the age of 3.5 years. Her acute post-operative cutaneous oxygen saturation was 93 per cent.

She was more cyanosed at the 1 month post-operative visit and the cutaneous oxygen saturation had dropped to 70 per cent in room air. Echocardiogram showed unobstructed modified Fontan circuit, dehiscence of Gortex^{TM} patch with leakage at the lower part into the pulmonary venous atrium. Cardiac catheterization demonstrated mean pressure in the Fontan circuit was 10 mmHg. Angiogram confirmed the leakage, discrete significant narrowing of the proximal RPA, 2 small collaterals from the right superior vena cavae (SVC) drained into the pulmonary venous atrium and one small collateral from the right subclavian vein drained into the right lower lobe pulmonary vein (Fig. 1). She was taken to the operating room to have the leakage repaired. Her acute post-operative course was complicated by a significant amount of pleural effusion which required prolonged chest tube drainage. Then percutaneous balloon angioplasty and finally PALMAZ stent (P308, Johnson & Johnson Interventional System Co) placement were performed at proximal RPA (Fig. 2) and small collateral vessels were occluded with Gianturco coil embolization (Cook, Inc., Bloomington, IN) at the same admission. Echocardiogram post procedure demonstrated a small communication between the Fontan circuit and the pulmonary venous atrium. Cutaneous oxygen saturation went up to high 80 per cent. The intercostal drainage was successfully removed a week after the intervention. At the follow-up period of 8 months her cutaneous oxygen saturation had gradually dipped to low 80 per cent with an increase in exercise intolerance. The authors decided to re-catheterize her with the plan to close the communication.

Re-catheterization was performed under general anesthesia to determine the possibility of occluding the residual patch leakage by Amplatzer septal occluder (ASO), (AGA Medical Corp, Golden Valley, Minn). The hemodynamic data was obtained during test occlusion which was in the acceptable range for permanent occlusion: Fontan circuit pressure went up from 10 to 14 mmHg, and aortic oxygen saturation rose from 79 per cent to 92 per cent post-occlusion. Then, the authors decided to close the defect with ASO. (Fig. 3) Cefazolin 50 mg/kg was administered 30 minutes before the procedure. Transesophageal echocardiography (TEE) was used to assist in defect sizing and device placement. Sizing of the defect was performed with an occlusion balloon catheter. The balloon size that resulted in total occlusion was 8 mm. Then, the device size was selected to be number 10 (10 mm at waist). A 12 F delivery

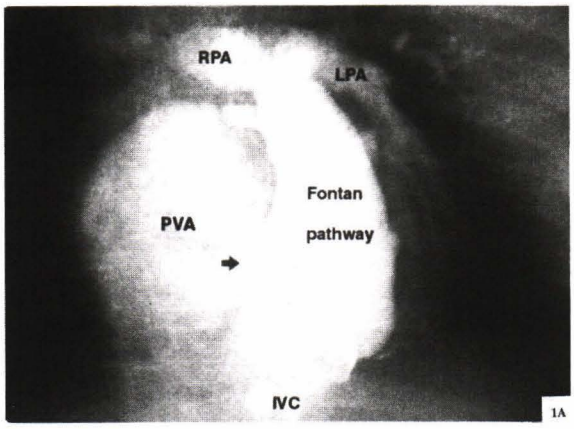

Fig. 1A. Angiography of the Fontan pathway; Demonstrated atrial baffle leakage, 8 mm in diameter (black arrowhead). PVA = pulmonary venous atrium, RPA = right pulmonary artery, LPA = left pulmonary artery, IVC = inferior venacavar.

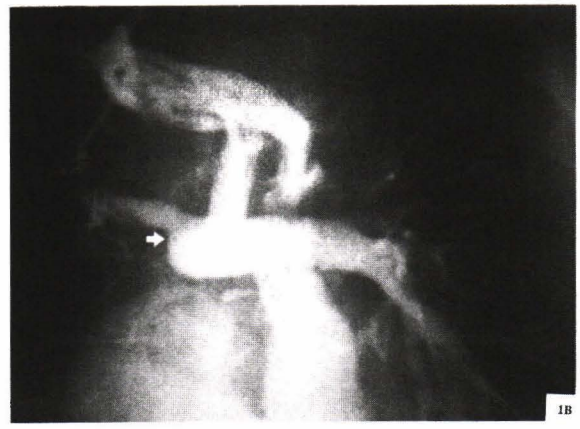

Fig. 1B. Demonstrates stenosis of the proximal RPA (white arrowhead).

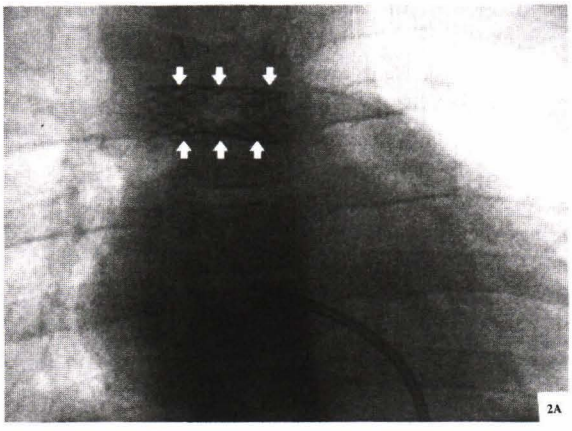

Fig. 2A. Demonstrates post coils embolization of veno-venous collateral vessels and a balloon expandable PALMAZ stent (P308, Johnson & Johnson Interventional System Co) in the RPA (arrowhead).

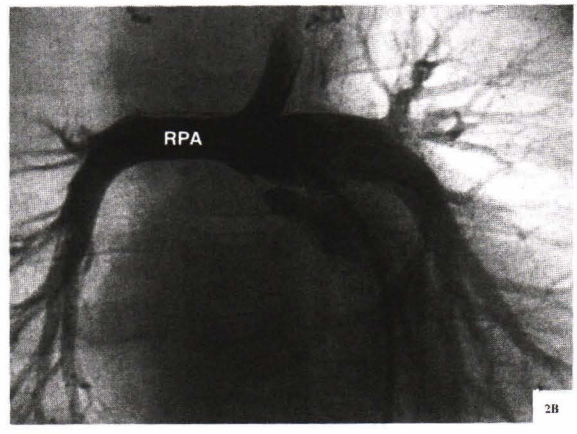

Fig. 2B. Demonstrates angiogram post-stent placement at proximal RPA.

sheath was inserted and advanced across the communication over the guide wire. The wire and dilator were removed. The device was prepared and loaded into the delivery sheath carefully to avoid introduction of air bubbles. The device was advanced through the sheath, and the distal disk was released into the pulmonary venous atrium. With the use of fluoroscopy and TEE, the distal disk was pulled firmly against the baffle; the waist and then the proximal disk were released into the systemic venous side of the baffle. After appropriate placement, a push-pull maneuver and color Doppler mode TEE were performed to confirm the stability. Then, the device was deployed. Serial pressures and oxygen saturations

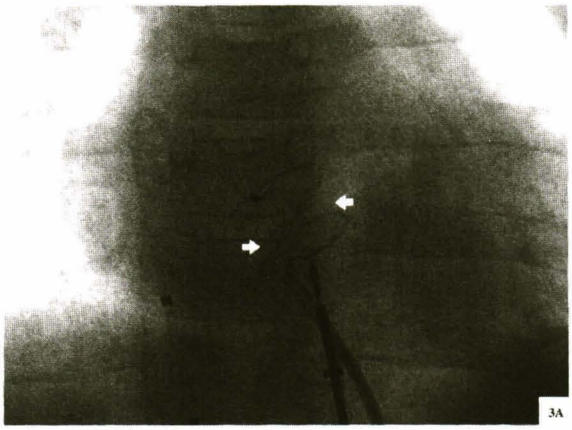

Fig. 3A. Post Amplatzer septal occluder implantation: A) Demonstrates position of device (arrowhead).

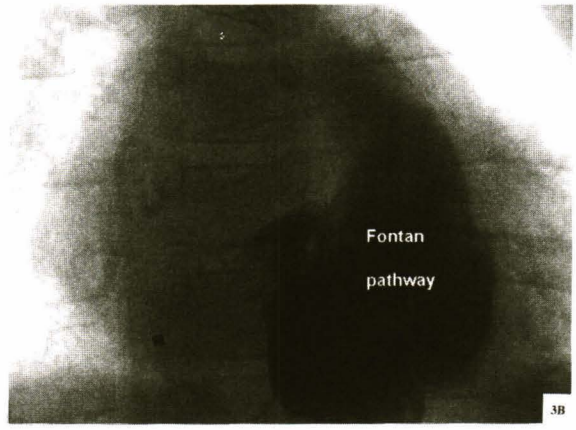

Fig. 3B. Angiography of the Fontan circuit demonstrates nearly complete occlusion of the atrial baffle leakage.

were repeatedly obtained in the baffle and the aorta, and an angiogram was performed with injection in the baffle 10 minutes later.

Anesthesia was reversed, and routine post-catheterization precautions were observed. She remained in the hospital for one night, and daily aspirin (3-5 mg/kg) was initiated. Before hospital discharge, the transthoracic echocardiography demonstrated complete device occlusion. She was discharged on digitalis, enalapril, and aspirin.

At her most recent examination, 2 months after the cardiac catheterization, her cutaneous oxygen saturation was 94 per cent. She was well and in functional class II, and able to attend school regularly. Mild hepatomegaly was also observed.

DISCUSSION

Early survival after Fontan operation improved from 75 per cent to 83 per cent in the 1970s and to more than 90 per cent in the current era, despite application of the operation in patients with higher risk factors[7-9]. Elevated pulmonary arterial pressure, high pulmonary vascular resistance, and pulmonary artery distortion were widely accepted as risk factors for early failure of the Fontan operation[10-13]. The present patient, had prolonged right pleural effusion soon after surgery that could be explained by rather high PVR and some degree of discrete proximal right pulmonary artery stenosis. However, she had leakage of atrial baffle that preserved her cardiac output and limited the extent of increased central venous pressure. To date, studies of balloon-expandable stent in congenital heart disease have suggested feasibility of implantation and showed gratifying immediate and intermediate results especially in late childhood[14-16]. Compared with percutaneous balloon angioplasty of discrete pulmonary artery narrowing, stent placement resulted in better outcome regarding diameter and pressure gradient[17]. As in the presented patient, after the stent had been placed, pleural effusion was considerably reduced within 1 week. There was no recurrence of stenosis during the intermediate-term follow-up. In the future, if restenosis occurs, this implantable stent could be re-expanded to a larger diameter by balloon catheter.

After completion of the Fontan circuit, most of the patients would have percutaneous oxygen saturation at low 90 per cent due to some of the venous blood flow, which drained from the coronary sinus to the pulmonary venous atrium. The presented patient experienced persistent cyanosis, which resulted from significant leakage of the atrial baffle and multiple veno-venous collateral vessels. This could have resulted from high PVR and high pressure in the Fontan circuit. In some institutions, routine fenestration is performed in the Fontan operation to improve the immediate post-operative outcome[18]. However, this can affect a significant degree of aortic deoxygenation, and may increase the risk of embolic complication and acquired closure later on. A recent study [19] also demonstrated improvement of oxygenation, reduction of the need for anticongestive medication,

and improvement of somatic growth at 10 years follow-up after fenestration closure in Fontan patients. For these reasons, test occlusion was performed. Marked increases in right-sided pressure or falls in cardiac output at test occlusion are taken as signs to defer closure[19]. The presented patient had a slight increase in systemic venous pressure during the test occlusion, and aortic saturation rose to 95 per cent.

Transcatheter closure of fenestration was first described by Bridges et al[20] in 1990 with the "clamshell" double umbrella device. Subsequent reports described experiences with the Gianturco coil [21,22] and Amplatzer septal occluder (ASO) [23-25], which are made from a self expanding nitinol mesh. The ASO has several advantages over the clamshell device in closing fenestration: the smaller delivery sheath, the specific size for different fenestration diameters, and the retrieval and repositioning capability[24]. In Thailand, this is the first case report on closing of the atrial baffle leakage of lateral tunnel Fontan circuit by ASO with an excellent immediate result after the procedure.

In conclusion, successful percutaneous coil embolization of veno-venous collaterals, balloon-expandable stent implantation of the discrete right pulmonary artery stenosis, and occlusion of the atrial baffle leakage with the Amplatzer septal occluder in a child with univentricular heart physiology were reported. Presently, interventional cardiac catheterization is a very useful adjunctive treatment for surgery in congenital heart diseases.

(Received for publication on August 16, 2002)

REFERENCES

1. Gentles TL, Gauvreau K, Mayer JE Jr, et al. Functional outcome after the Fontan operation: Factors influencing late morbidity. J Thorac Cardiovasc Surg 1997; 114: 392-403.
2. Stamm C, Friehs I, Mayer JE Jr, et al. Long-term results of the lateral tunnel Fontan operation. J Thorac Cardiovasc Surg 2001; 121: 28-41.
3. Chussat A, Fontan I, Besse P, et al. Selection criteria for Fontan's procedure. In: Anderson RH, Shineburne EA eds. Pediatric Cardiology. Edinburgh : Churchill Livingstone; 1977: 559-66.
4. Mayer JE Jr, Bridges ND, Lock JE, et al. Factors associated marked reduction in mortality for Fontan operation in patients with single ventricle. J Thorac Cardiovasc Surg 1992; 103: 444-52.
5. Kirklin JW, Barratt-Boyes BG. Tricuspid atresia and the Fontan operation. In: Kirklin JW, Barratt-Boyes BG. eds. Cardiac surgery: Morphology, diagnostic criteria, natural history, techniques, results and indications. 2nd ed. Vol II. New York : Churchill Livingstone; 1993: 1081-8.
6. Wilson DG, Wisheart JD, Stuart AG. Systemic thromboembolism leading to myocardial infarction and stroke after fenestrated total cavopulmonary connection. Br Heart J 1995; 73: 483-5.
7. Mair DD, Hagler DJ, Julsrud PR, et al. Early and late results of the modified Fontan procedure for double-inlet left ventricle: the Mayo Clinic experience. J Am Coll Cardiol 1991; 18: 722-32.
8. Sanders SP, Weight GB, Keane JF, et al. Clinical and hemodynamic results of the Fontan operation for tricuspid atresia. Am J Cardiol 1982: 49; 1733-40.
9. Gentles TL, Mayer JE, Gauvreau K, et al. Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome. J Thorac Cardiovasc Surg 1997; 114: 376-91.
10. DeLeon SY, Ilbawi MN, Idriss FS, et al. Fontan type operation for complex lesions: Surgical considerations to improve survival. J Thorac Cardiovasc Surg 1986; 92: 1029-37.
11. Mayer JE Jr, Helgason H, Jonas RA, et al. Extending the limits for modified Fontan procedures. J Thorac Cardiovasc Surg 1986; 92: 1021-28.
12. Bartmus DA, Driscoll DJ, Offord KP, et al. The modified Fontan operation in children less than 4 years old. J Am Coll Cardiol 1990;15:429-35.
13. Kirklin JK, Blackstone EH, Kirklin JW, et al. The Fontan operation: Ventricular hypertrophy, age and date of operation at risk factors. J Thorac Cardiovasc Surg 1986; 92: 1049-64.
14. O'laughlin MP, Slack MC, Grifka RG, et al. Implantation and intermediate-term follow-up of stents in congenital heart disease. Circulation 1993; 88: 605-14.
15. O'laughlin MP, Perry SB, Lock JE, et al. Use of endovascular stents in congenital heart disease. Circulation 1991; 83: 1923-39.
16. Hosking MC, Benson LN, Nakanishi T, et al. Intravascular stent prosthesis for right ventricular outflow obstruction. J Am Coll Cardiol 1992; 20: 373-80.
17. Rothman A, Perry SB, Keane JF, et al. Early results and follow-up of balloon angioplasty for branch pulmonary artery stenosis. J Am Coll Cardiol 1990; 15: 1109-17.
18. Lemler MS, Scott WA, Leonard SR, et al. Fenestration improve clinical outcome of fontan procedure: a prospective, randomized study. Circulation;105: 207-12.
19. Goff DA, Blume ED, Gauvreau K, et al. Clinical outcome of fenestrated Fontan patients after closure, the first 10 years. Circulation 2000; 102: 2094-9.
20. Bridges ND, Lock JE, Castaneda AR. Baffle fenestration with subsequent transcatheter closure: Modification of the Fontan operation for patients at increased risk. Circulation 1990; 82: 1681-9.
21. Sommer RJ, Recto M, Golinko RJ, et al. Transcatheter coil occlusion of surgical fenestration after Fontan operation. Circulation 1996; 94: 249-52.
22. Sanatani S, Sett SS, Human DG, et al. Extracardiac Fontan operation with tube fenestration allowing transcatheter coil occlusion. Ann Thorac Surg 1998; 66: 933-4.
23. Tofeig M, Walsh KP, Chan C, et al. Occlusion of Fontan fenestration using the Amplatzer septal occluder. Heart 1998; 79: 368-70.
24. Tofeig M, Walsh KP, Arnold R. Transcatheter occlusion of a post-Fontan residual hepatic vein to pulmonary venous atrium communication using the Amplatzer septal occluder. Heart 1998; 79: 624-6.
25. Moore JW, Murdison KA, Baffa GM, et al. Transcatheter closure of fenestrations and excluded hepatic veins after Fontan: Versatility of the Amplatzer device. Am Heart J 2000; 140: 534-40.

การรักษาผู้ป่วยหลังผ่าตัดฟอนแทน ด้วยการอุดหลอดเลือดด้วยขดลวด การใส่โครง-ลวดถ่างคาหลอดเลือด และ Amplatzer septal occluder : รายงานผู้ป่วย

วรการ พรหมพันธุ์, พบ, จารุพิมพ์ สูงสว่าง, พบ*,*
กฤตวิกรม ดุรงค์พิสิษฏ์กุล, พบ, ดวงมณี เลาหประสิทธิพร, พบ**
*สัมพันธ์ พรวิลาวัลย์, พบ** อภิชาติ นานา, พบ*, สมชาย ศรียศชาติ, พบ***

รายงานผู้ป่วยเด็กหญิงไทยอายุ 7 ปี ซึ่งได้รับการวินิจฉัยว่ามีหัวใจอยู่ด้านขวา มีเวนตริเคิล เพียงห้องเดียว ร่วมกับลิ้นหัวใจพัลโมนารีตันและได้รับการผ่าตัด lateral tunnel Fontan เมื่ออายุ 3.5 ปี หลังผ่าตัดมีน้ำในช่องเยื่อหุ้มปอดติดต่อกันเป็นเวลานาน ร่วมกับมีการรั่วของท่อลัดเส้นเลือดดำ (atrial baffle) บริเวณเอเตรียมและมีการเชื่อมต่อที่ผิดปกติ (collaterals) ระหว่างเส้นเลือดดำ superior vena cavae กับเส้นเลือดดำพัลโมนารี (pulmonary veins) ส่งผลให้ผู้ป่วยยังมีอาการเขียวเหมือนก่อนผ่าตัด ผู้ป่วยได้รับการขยายหลอดเลือดแดงพัลโมนารีด้านขวา และใส่โครงลวดถ่างคาหลอดเลือด (balloon expandable stent implantation) ไว้ ร่วมกับการอุดหลอดเลือด collaterals ด้วยขดลวด (coil embolization) นอกจากนี้ผู้ป่วยได้รับการปิดท่อลัดเส้นเลือดที่มีรูรั่วด้วย Amplatzer septal occluder ได้สำเร็จ ทำให้ผู้ป่วยหายเขียวและมีคุณภาพชีวิตที่ดีขึ้นเป็นรายแรกในประเทศไทย

คำสำคัญ : การผ่าตัดฟอนแทน, การอุดหลอดเลือดด้วยขดลวด, การใส่โครงลวดถ่างคาหลอดเลือด, อุปกรณ์ปิดรูรั่วผนังกั้นหัวใจห้องบนชนิดแอมพลาสเซอร์

วรการ พรหมพันธุ์, จารุพิมพ์ สูงสว่าง, กฤตวิกรม ดุรงค์พิสิษฏ์กุล, และคณะ
จดหมายเหตุทางแพทย์ ฯ 2546; 86: 369–375

* หน่วยกุมารเวชศาสตร์โรคหัวใจ, ภาควิชากุมารเวชศาสตร์
** หน่วยศัลยศาสตร์หัวใจและทรวงอก, ภาควิชาศัลยศาสตร์, คณะแพทยศาสตร์ศิริราชพยาบาล, มหาวิทยาลัยมหิดล, กรุงเทพ ฯ 10700