

Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery: Successful Direct Reimplantation in a 50-year-old Man

Takemi Kawara, MD, Eiki Tayama, MD, Nobuhiko Hayashida, MD, Masaru Nishimi, MD, and Shigeaki Aoyagi, MD

Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital coronary artery anomaly that is often referred to as Bland White Garland syndrome. Most patients with this anomaly require surgical intervention early in life, and it is extremely rare that patients reach middle age without any symptoms. We present a 50-year-old man with this anomaly, who underwent direct reimplantation of the left main coronary trunk to the ascending aorta. His postoperative course was uneventful, and three and a half years after the operation, he is well and does not require medication. Several surgical procedures can be used to treat this anomaly, but we prefer to use direct reimplantation, whenever technically possible. To our knowledge, this patient is the oldest patient to have undergone a direct reimplantation without any angioplasty. (Ann Thorac Cardiovasc Surg 2003; 9: 197–201)

Key words: anomalous origin of the left coronary artery, Bland White Garland syndrome, coronary artery anomaly, direct reimplantation

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly that is often referred to as Bland White Garland syndrome.¹⁾ Most infants die within the first year of life without treatment.²⁾ Survival beyond infancy depends on the development of adequate collateral circulation from the right coronary artery (RCA) or from an other source to the left coronary artery (LCA). Thus, older patients who survive without surgical intervention are rare. We report the successful surgical treatment of a 50-year-old man with ALCAPA by direct reimplantation of the LCA to the ascending aorta. Details of the intraoperative findings are presented, and we also discuss the optimal operative strategies for adult patients with this anomaly.

From Division of Cardiovascular Surgery, Department of Surgery, Kurume University School of Medicine, Kurume, Japan

Received October 8, 2002; accepted for publication January 31, 2003.

Address reprint requests to Takemi Kawara, MD: Department of Surgery, Kurume University School of Medicine, 67 Asahi-machi, Kurume-city, Fukuoka 830-0011, Japan.

Case Report

A 50-year-old man was referred to our hospital for the evaluation of fatigue and shortness of breath on exertion. He was 169 cm tall and weighed 56 kg. His blood pressure was within the normal range. Physical examination revealed a grade 2/6 regurgitant systolic murmur at the apex and a grade 2/6 continuous murmur at the fourth left sternal border. Chest X-ray revealed the absence of cardiomegaly with a cardiothoracic ratio of 47%. Electrocardiogram (ECG) revealed sinus rhythm and left ventricular hypertrophy, with strain patterns of the ST-T segment in V5 and V6, and Q waves in I, aVL, V5, and V6. A provisional diagnosis of an old anterolateral infarction was subsequently made. Transthoracic echocardiography revealed a dilated left ventricle (end-diastolic diameter 6.4 cm, end-systolic diameter 4.4 cm), a grade 2/4 mitral regurgitation caused by papillary muscle dysfunction, and annular dilatation without prolapsing leaflets. Aneurysmal dilatation of the RCA and a mosaic flow signal in the left anterior descending coronary artery were also found. Exercise stress thallium-201 perfusion scintigraphy revealed reversible atypical patchy hypoperfusion in the



Fig. 1. Preoperative right coronary angiogram. Right coronary artery is dilated and tortuous. Left coronary artery is filled via extensive collaterals and the contrast is seen draining into the main pulmonary artery.

anterior and posterior area of the left ventricle.

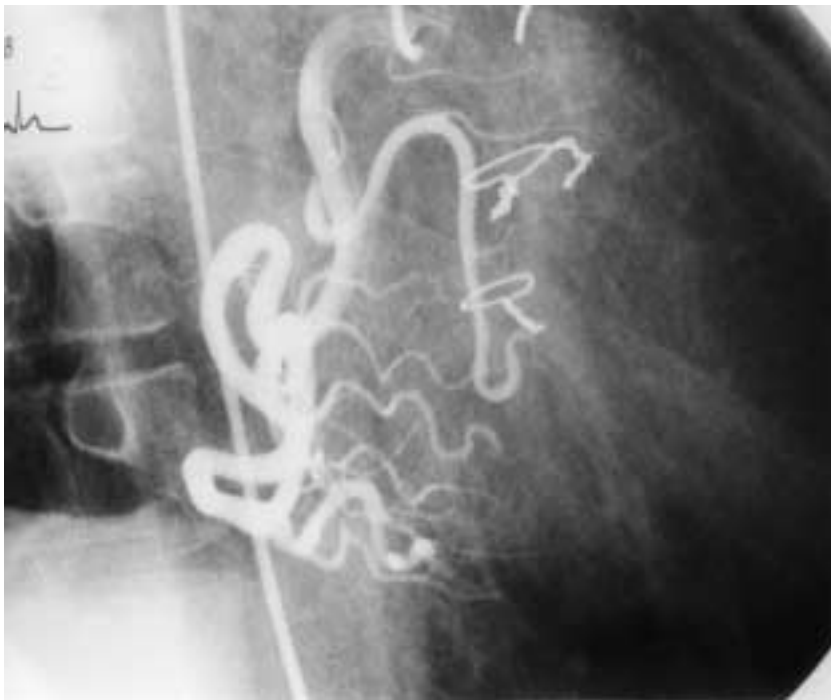
Cardiac catheterization revealed a left-to-right shunt rate of 32%, with a Qp/Qs of 1.48. Coronary angiography (Fig. 1) showed a markedly dilated and tortuous RCA, with collateral filling to the left coronary system, that drained retrogradely into the main pulmonary artery (mPA) via an anomalous left main trunk (LMT) of the LCA. In the view of this, the patient was diagnosed with ALCAPA, and surgical correction was indicated.

Surgical technique

After median sternotomy and pericardiotomy, a congenital pericardial defect (7 cm superioinferiorly \times 5 cm anteroposteriorly) was found on the right side of the superior vena cava. The RCA was markedly dilated and tortuous. The LMT of the LCA on the posterior side of the mPA root was soft and uncalcified. The ascending aorta and the mPA were carefully separated from each other. Standard cardiopulmonary bypass was established with aortic and bicaval cannulation, and the left ventricle was vented. The aorta was cross-clamped and cold blood cardioplegic solution was infused into the aortic root under total cardiopulmonary bypass with moderate hypothermia (at 28°C). At the same time, the pulmonary artery was transected distal to the commissures' level. There was a brisk backflow spurt of arterial blood from the LCA ostium and complete cardiac standstill was not obtained

until the fingertip capped the LCA ostium. Although the heart was cool and ECG once showed flat tracing, electrical activity of the heart returned within a few minutes of antegrade administration of cardioplegic solution. Retrograde continuous cold blood cardioplegia (RCCBC) was subsequently administered to the coronary sinus to obtain continuous cardiac standstill, using a balloon tipped catheter with purse-string sutures placed around the orifice and a tourniquet. However, the RCCBC did not work very well, certainly because of profuse noncoronary collaterals, which caused annoying continuous backflow spring from the LMT ostium even when RCCBC was not being administered. Ventricular fibrillation was often observed and complete cardiac standstill was obtained only after a few minutes of antegrade administration of a cardioplegic solution every 20 minutes.

The ostium of LMT was excised as a large button of the mPA wall and the LMT was clamped to control backflow. The LCA was subsequently dissected and mobilized as far as the origin of the left circumflex artery and the left anterior descending artery. A hole was made in the left posterolateral wall of the ascending aorta to fit the LMT button, using a 4.4-mm puncher several times. The LMT button was subsequently anastomosed to the aorta with a 5-0 polypropylene continuous suture. The right facing pulmonary sinus of Valsalva, which previously hosted the coronary ostium, was then reconstructed



A
B

Fig. 2. A: Postoperative left coronary angiogram: showing widely patent anastomosis of the LMT to the aorta and dilated LMT and left coronary tree. B: Postoperative right coronary angiogram: previously shown collaterals of the LCA are not seen.

using bovine pericardium. The continuity of the anterior pulmonary trunk was reestablished via direct anastomosis. The aortic cross-clamp was released, and the operation was completed in a routine fashion.

The patient's postoperative course was uneventful. Cardiac catheterization and coronary angiography on postoperative day 42 revealed no stenosis at the anasto-

mosis site of the LMT (Fig. 2A) and no collateral filling to the LCA from the RCA. The RCA (Fig. 2B) had become slightly smaller, and the right ventricular branches (former collateral vessels) had become markedly thinner, as assessed by comparison of pre- and postoperative angiograms. Four years after the operation, the patient was found to be symptom-free without medication.

Discussion

ALCAPA is a rare anomaly in adults because more than 80% of symptomatic infants with this disease die of heart failure within the first year of life if not treated.²⁾ In adults, collateral blood flow from the RCA to the LCA system and noncoronary collateral blood flow (NCCF) contributes to survival beyond childhood as in our case. However, even in the patients who survive to adulthood, sudden death frequently occurs.²⁾ Therefore, surgical intervention should be performed in all patients, even in asymptomatic adult patients with no objective evidence of ischemia.

The patients who undergo simple ligation of the ALCAPA may survive for varying periods. However, late-onset sudden death in patients undergoing this procedure alone has been reported.^{3,4)} Thus, in the repair of this anomaly, reestablishment of a double-coronary system is desirable. There are several surgical alternatives for the establishment of a double-coronary system, including subclavian-coronary artery anastomosis,⁵⁾ ligation of the left coronary artery combined with coronary artery bypass grafting (CABG) with a saphenous vein graft or an internal thoracic artery,^{6,7)} intrapulmonary tunnel repair,⁸⁻¹⁰⁾ tubular extension of the LMT using pulmonary artery tissue anastomosed to the aorta or other arterial source,^{11,12)} and direct reimplantation of the anomalous left coronary artery to the aorta, as described by Neches et al.¹³⁾ and Grace et al.¹⁴⁾ While no single surgical technique is widely accepted, we prefer the direct reimplantation technique because it is simple and offers physiological coronary flow, possibly with the best patency rate. Although our patient was a 50-year-old man, we were convinced that direct implantation could be performed because of the normal length of the LMT without calcification, and because of the short distance between the course of the LMT at the right facing pulmonary sinus of Valsalva and the ascending aorta. Many reports have confirmed that direct reimplantation can be performed safely in patients of a wide age range.¹⁵⁻¹⁸⁾ Therefore, the left coronary artery should be reimplanted into the aorta, whenever technically possible, even though this procedure requires meticulous dissection of the LMT in adult patients because of the dilated and thin vessel walls. Of previously published studies in which the age of the patient was specified, a 456.6-month-old (about 38-year-old) patient¹⁶⁾ is the oldest patient to have undergone direct reimplantation without LMT reconstruction prior to our patient.

During surgical intervention for this anomaly, well-

managed myocardial protection is essential. During the initial administration of antegrade cardioplegic solution, clamping of the mPA or occlusion of the LMT ostium after the opening of the mPA is required to avoid the steal phenomenon. However, in adult patients with ALCAPA, maintenance of complete cardioplegic arrest is usually markedly difficult, because of the presence of profuse NCCF.¹⁹⁻²¹⁾ NCCF to the heart is supplied from noncoronary routes, such as mediastinal, pericardial, and bronchial collateral channels, through pericardial reflections surrounding pulmonary and systemic veins as well as from the vasa vasorum along major vessels.²²⁾ These collateral vessels appear to function as supplemental feeding arteries when myocardial ischemia occurs.²³⁾ In the case reported by Murata et al.,²⁴⁾ they performed selective bronchial artery angiography after the surgery, in which LMT ostium was closed via mPA resulting in the single coronary system, and demonstrated collaterals from the bronchial artery to the left circumflex artery. However, to our knowledge, there has been no report on this anomaly in which NCCF was angiographically confirmed preoperatively and the fate of those vessels was studied after surgery.

Thus, although it was considered difficult to control NCCF during aortic cross clamping, we employed a combined intermittent antegrade and RCCBC method keeping the LMT clamped, expecting optimal myocardial protection with cardiac standstill. In spite of this, cardiac arrest was obtained for only several minutes after every antegrade administration of cardioplegic solution. Therefore, if ventricular fibrillation often occurs during aortic cross clamping even when cardioplegic solution is being correctly administered, the systemic temperature of the cardiopulmonary bypass should be reduced to less than 28°C, and the left heart should be fully vented to obtain better myocardial protection.^{21,25)}

References

1. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *Am Heart J* 1933; **8**: 787-801.
2. Wesselhoeft H, Fawcett JS, Johnson AL. Anomalous origin of the left coronary artery from the pulmonary trunk. Its clinical spectrum, pathology, and pathophysiology based on a review of 140 cases with seven further cases. *Circulation* 1968; **38**: 403-25.
3. Wilson CL, Dlabal PW, McGuire SA. Surgical treatment of anomalous left coronary artery from pulmonary artery: follow-up in teenagers and adults. *Am Heart*

- J* 1979; **98**: 440–6.
4. Backer CL, Stout MJ, Zales VR, et al. Anomalous origin of the left coronary: a twenty-year review of surgical management. *J Thorac Cardiovasc Surg* 1992; **103**: 1049–58.
 5. Meyer BW, Stefanik G, Stiles QR, Lindesmith GG, Jones JC. A method of definitive surgical treatment of anomalous origin of left coronary artery. *J Thorac Cardiovasc Surg* 1968; **56**: 104–7.
 6. el-Said GM, Ruzyllo W, Williams RL, et al. Early and late result of saphenous vein graft for anomalous origin of left coronary artery from pulmonary artery. *Circulation* 1973; **48** (Suppl): III2–6.
 7. Kitamura S, Kawachi K, Nishii T, et al. Internal thoracic artery grafting for congenital coronary malformations. *Ann Thorac Surg* 1992; **53**: 513–6.
 8. Hamilton DI, Ghosh PK, Donnelly RJ. An operation for anomalous origin of left coronary artery. *Br Heart J* 1979; **41**: 121–4.
 9. Takeuchi S, Imamura H, Katsumoto K, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg* 1979; **78**: 7–11.
 10. Midgley FM, Watson DC Jr, Scott LP III, et al. Repair of anomalous origin of the left coronary artery in the infant and small child. *J Am Coll Cardiol* 1984; **4**: 1231–4.
 11. Vigneswaran WT, Campbell DN, Pappas G, Wiggins JW, Wolfe RW, Clarke DR. Evolution of the management of anomalous left coronary artery: a new surgical approach. *Ann Thorac Surg* 1989; **48**: 560–4.
 12. Tashiro T, Todo K, Haruta Y, Yasunaga H, Nagata M, Nakamura M. Anomalous origin of the left coronary artery from the pulmonary artery. *J Thorac Cardiovasc Surg* 1993; **106**: 718–22.
 13. Neches WH, Mathews RA, Park SC, et al. Anomalous origin of the left coronary artery from the pulmonary artery. A new method of surgical repair. *Circulation* 1974; **50**: 582–7.
 14. Grace RR, Algolini P, Cooley DA. Aortic implantation of anomalous left coronary artery arising from pulmonary artery. *Am J Cardiol* 1977; **39**: 608–13.
 15. Fernandes ED, Kadivar H, Hallman GL, Reul GJ, Ott DA, Cooley DA. Congenital malformation of the coronary arteries: the Texas Heart Institute experience. *Ann Thorac Surg* 1992; **54**: 732–40.
 16. Lambert V, Touchot A, Losay J, et al. Midterm results after surgical repair of the anomalous origin of the coronary artery. *Circulation* 1996; **94** (Suppl): II38–43.
 17. Moraes F, Lincoln C. Anomalous origin of left coronary artery. Evolution of surgical treatment. *Eur J Cardiothorac Surg* 1996; **10**: 603–8.
 18. Cochrane AD, Coleman DM, Davis AM, Brizard CP, Wolfe R, Karl TR. Excellent long-term functional outcome after an operation for anomalous origin of the left coronary artery from the pulmonary artery. *J Thorac Cardiovasc Surg* 1999; **117**: 332–42.
 19. Chan RKM, Hare DL, Buxton BF. Anomalous left main coronary artery arising from the pulmonary artery in an adult: treatment by internal mammary artery grafting. *J Thorac Cardiovasc Surg* 1995; **109**: 393–4.
 20. Sivasubramanian S, Krishnamurthy SMRG, Thirumalai P, et al. Anomalous origin of the left coronary artery from the pulmonary artery: is reconstruction of the double coronary artery always necessary? *J Thorac Cardiovasc Surg* 1996; **111**: 901–3.
 21. Arsan S, Naseri E, Keser N. Adult case of Bland White Garland syndrome with huge right coronary aneurysm. *Ann Thorac Surg* 1999; **68**: 1832–3.
 22. Bloor CM, Liebow AA. Coronary collateral circulation. *Am J Cardiol* 1965; **16**: 238–52.
 23. Bjork L. Angiographic demonstration of extracardial anastomosis to the coronary arteries. *Radiology* 1996; **87**: 274–7.
 24. Murata N, Ohta H, Suzuki K. An adult case of anomalous origin of the left coronary artery from the pulmonary artery with the coronary artery-bronchial artery anastomosis. *Nippon Kyobu Geka Gakkai Zasshi* 1992; **40**: 2113–9. (in Japanese)
 25. Akins CW. Hypothermic fibrillatory arrest for coronary artery bypass grafting. *J Card Surg* 1992; **7**: 342–7.