Creation of a dual-coronary system for anomalous origin of the left coronary artery from the pulmonary artery utilizing the trapdoor flap method


Pediatric and Congenital Heart Surgery, Cleveland Clinic Children's Hospital, The Cleveland Clinic Foundation, 9500 Euclid Avenue, Cleveland, OH 44195, USA

Received 18 August 2001; received in revised form 31 May 2002; accepted 26 June 2002

Abstract

Objective: Results of the repair of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) have improved. Direct implantation of the anomalous coronary artery into the ascending aorta establishes a dual-coronary system and is the goal of current surgical approaches. We report the development of our surgical technique for ALCAPA.

Methods: Between September 1993 and December 2000, 13 patients underwent surgery for ALCAPA. There were four males and nine females. Ages ranged from 1 month to 25 years (median = 3.9) and weight ranged from 2.6 to 102 kg (median = 16.8). One patient had previously undergone an operative procedure at an outside institution.

Results: Direct implantation of the anomalous coronary artery into the ascending aorta was feasible in 12 of 13 patients. In situ transfer was performed in one patient with an intramural coronary artery. The first case in the series required an intrapulmonary baffle reconstruction (Takeuchi procedure) because the coronary artery arose remotely from the ascending aorta from the left-anterior sinus of the PA. For coronary transfer, a trapdoor flap was created on the ascending aorta for the implantation of the coronary button and the sinus defect in the main PA was augmented with a pericardial patch. The left ventricular (LV) shortening fraction was improved from a median value of 27% (range 12–36%) preoperatively to 33% (range 24–45%) in the immediate postoperative period (P = 0.004). The LV end-diastolic dimension decreased from a median value of 36 mm (range 22–70 mm) preoperatively to 29 mm (range 19–56 mm) in the immediate postoperative period (P = 0.004). There has been no mortality or reoperation during a median follow-up of 36 months.

Conclusions: Using a standard technique, direct implantation of the anomalous coronary artery into the ascending aorta was achieved in all cases but one. At intermediate follow-up, LV function had improved by echocardiography. No postoperative mechanical circulatory support was required in any of these patients. This operative technique is reproducible and is applicable to the majority of patients with ALCAPA. © 2002 Elsevier Science B.V. All rights reserved.

Keywords: Anomalous origin of the left coronary artery from the pulmonary artery; Direct implantation; Dual-coronary system; Bland-White-Garland syndrome

1. Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly. Past surgical approaches which employed ligation of the anomalous coronary artery were associated with unacceptably high mortality [1,2]. Coronary artery bypass utilizing either the left subclavian or internal mammary artery also yielded unsatisfactory results [3]. Establishment of a dual-coronary artery system has gained widespread acceptance as the current treatment goal [4]. Implantiing the anomalous coronary artery directly into the ascending aorta is ideal but may not be possible for some unusual anatomic patterns in which the anomalous coronary artery arises in a location remote from the ascending aorta or in which the anomalous coronary artery takes an intramural course between the great arteries. In these situations, other surgical options, such as intrapulmonary baffling (Takeuchi procedure), may be required. The evolution of the surgical technique at our center has resulted in increased use of direct implantation of the anomalous coronary artery into the ascending aorta.

* Corresponding author. Tel.: +1-216-445-7833; fax: +1-216-445-3692.
E-mail address: meer@ccf.org (R.B.B. Mee).
2. Materials and methods

2.1. Patients

Between September 1993 and December 2000, 13 patients underwent surgical treatment for ALCAPA. There were four males and nine females. Ages ranged from 1 month to 25 years (median = 3.9) and weight ranged from 2.6 to 102 kg (median = 16.8). Four patients were asymptomatic with cardiomegaly detected incidentally on chest X-ray. Nine patients had symptoms consistent with myocardial ischemia or infarction. Eight demonstrated left ventricular (LV) dyskinesia while one patient demonstrated severe right ventricular (RV) dysfunction. Two patients required mechanical ventilation for respiratory failure due to respiratory syncitial viral pneumonia in one case and sepsis due to necrotizing enterocolitis in the other case. There were 12 primary operations. One patient presented, after a previous Takeuchi procedure, with dehiscence of the PA baffle, which had resulted in a large aortopulmonary window (as discussed subsequently).

The preoperative diagnosis was established by two-dimensional echocardiography. We reserve angiography for patients in whom the preoperative diagnosis is in doubt and in all cases with new presentation of dilated cardiomyopathy and no evidence of ALCAPA on echocardiography. Origin of the coronary artery from the main PA was from the posterior sinus [9], from the left-posterior aspect [1], from the left-anterior aspect [1], from the anterior sinus [1] and from the right PA with an intramural aortic course but without connection to the aortic lumen [1].

2.2. Operative technique

Through a median sternotomy, the pericardium was opened to the left of the midline to preserve a pericardial flap for later use. High aortic cannulation at the take-off of the innominate artery and bicaval venous cannulation were used in all cases. High flow, low perfusion pressure cardiopulmonary bypass (CPB) was employed in all cases with the administration of phenoxybenzamine, in patients, sufficiently small so as to require a blood prime. Immediately after commencing CPB, the distal main PA was clamped or the branch PAs were snared to prevent coronary runoff via the innominate artery and bicaval venous cannulation were opened to the left of the midline to preserve a pericardial flap for later use. High aortic cannulation at the take-off of the innominate artery and bicaval venous cannulation were used in all cases. High flow, low perfusion pressure cardiopulmonary bypass (CPB) was employed in all cases with the administration of phenoxybenzamine, in patients, sufficiently small so as to require a blood prime. Immediately after commencing CPB, the distal main PA was clamped or the branch PAs were snared to prevent coronary runoff via the anomalous coronary artery. The aortic cross-clamp was placed high for creation of the trapdoor. Cold, crystalloid cardioplegia solution was infused into the aortic root to induce cardiac arrest with simultaneous occlusion of the outflow of the anomalous coronary artery.

2.3. Direct coronary implantation

The origin of the anomalous coronary artery was carefully inspected and confirmed. After cardioplegic arrest, the main PA was transected above the level of the coronary origin. The coronary orifice was excised with a generous cuff of the pulmonary sinus wall and the proximal portion of the coronary was mobilized with the preservation of branches. A trapdoor incision was made in the ascending aorta, directly opposite the origin of the coronary from the PA (Fig. 1a). The coronary artery was anastomosed to the aorta with a 6-0 or 7-0 prolene suture and after de-airing, the aortic cross-clamp was removed. The defect in the main PA was reconstructed with a generous fresh autologous pericardial patch, which also served to extend the length of the posterior wall of the main PA (Fig. 1b).

2.4. Intramural coronary artery

In this case, the coronary artery arose from the right PA, making a right-angled turn toward the aortic wall (Fig. 2a). The coronary artery then coursed through the aortic wall (intramural segment) without entering the aortic lumen. This was not detected until the origin from the right PA was excised. The aorta and the main PA were then transected and the coronary artery was amputated at its point of entry into its intramural course, proximally (Fig. 2b). The intramural segment of the coronary artery was unroofed internally into the aortic lumen and also externally. A small fresh autologous pericardial patch was placed at the upper end of the unroofed, transected coronary artery externally. The two ends of the transected aorta were then sutured. After removal of the aortic cross-clamp, the proximal right PA and the distal main PA were reconstructed with fresh autologous pericardium (Fig. 2c).

2.5. Reoperation

One patient was referred for repair after an unsuccessful reconstruction with a Takeuchi procedure at another institution. At presentation, the patient had a large aorto-pulmonary window due to dehiscence of the intrapulmonary baffle and partial obstruction of the right PA origin. An RV outflow patch had also been placed at the previous operation, which resulted in significant pulmonary insufficiency and RV failure. At reoperation, the intrapulmonary tunnel was taken down and the aorto-pulmonary window closed with a pericardial patch. The origin of the coronary artery was successfully implanted into the ascending aorta proximal to the aorto-pulmonary window. The outflow patch was removed, the pulmonary valve was reconstructed, and the main PA was patched with fresh autologous pericardium.

2.6. Mitral valve repair

Concomitant mitral valve repair was performed in two patients who had more than moderate mitral regurgitation (MR) due to prolapse of the anterior leaflet of the mitral valve resulting from ischemic damage to the papillary muscles. One patient had a mitral annuloplasty performed at both commissures. The other had a ring annuloplasty performed using a Cosgrove-Edwards® annuloplasty ring (Baxter Healthcare Corporation, Santa Ana, CA, USA).
2.7. Follow-up studies

At follow-up, the degree of postoperative MR was assessed by color Doppler echocardiogram for all patients using a scale of 0 to 3. LV function was assessed by shortening fraction (LVSF) which was calculated as end-systolic LV dimension divided by end-diastolic LV dimension (LVEDD). Preoperative and postoperative values were analyzed using a two-way Student’s paired t-test.

3. Results

Implantation of the ALCAPA to the ascending aorta was feasible in all but one patient. In this patient, the ALCAPA was arising remotely from the ascending aorta – from the anterior and leftward aspect of the main PA. In this case, an intrapulmonary baffle (Takeuchi) repair was utilized.

The degree of preoperative MR was 0 in two, 1 in four, 2 in five, and 3 in two patients. CPB time ranged from 74 to 150 (median = 100.5) min and aortic cross-clamp time ranged from 29 to 63 (median = 45.5) min. There was no 30-day or hospital mortality and mechanical circulatory support was not required in any case. The left atrial pressure after discontinuation of CPB ranged from 4 to 9 (median = 7) mmHg. All patients were managed with low-dose infusions of a single inotrope. Except for the two patients who required preoperative intubation for respiratory failure, patients were extubated within 3 days of operation (median = 1), were discharged from the intensive care unit within 5 days (median = 1), and were ready for hospital discharge within 9 days (median = 5). Postoperative echocardiography showed improved MR in six patients.

Of nine patients with significant preoperative ventricular dysfunction, seven were considered to have improved function on postoperative echocardiogram. Preoperatively, the median LVEDD was 36 mm (range 22–70 mm) which decreased to a median of 29 mm (range 19–56 mm) (P = 0.004) in the immediate postoperative period (median of 9 days for immediate postoperative values). Median preoperative LVSF was 27% (range 12–36%) which improved to a median value of 33% (range 24–45%) (P = 0.004) in the immediate postoperative period.

There has been no mortality or reoperation during a median follow-up of 36 months (range 0.6–66 months). Complete echocardiographic data are available for 11 of the 13 patients. Over this period of follow-up, the median LVEDD is 34 mm (range 22–62 mm) (P = 0.60 compared to preoperative LVEDD) and the LVSF is 38% (range 26–58%) (P = 0.004 compared to preoperative LVSF). Currently for these 11 patients, MR is 0 in two, 1 in three, 2 in five, and 3 in one. The patient with a patched aortic intramural segment of the anomalous coronary from the right PA demonstrates no dilatation or stenosis of the coronary origin by echocardiography.
4. Discussion

ALCAPA is a rare congenital anomaly, which is usually lethal without surgical intervention. The pathophysiology is characterized by the absence of prograde flow in the left coronary system often with flow reversal, which results in a ‘steal’ syndrome and subsequent myocardial ischemia. Patients may not be symptomatic in the neonatal period due to the physiologic increase in PA pressure, which maintains some degree of prograde flow in the anomalously arising coronary. Patients classically present within the first few months of life when PA pressures fall and left coronary perfusion pressure diminishes. Rarely, a patient may be asymptomatic and may not come to medical attention until later in life.

Due to concomitant myocardial damage, surgical treatment of ALCAPA remains challenging and carries significant mortality and morbidity even in the current era [5]. Historically, ligation of the anomalous coronary was performed as an early surgical treatment to eliminate steal from the coronary into the PA. This resulted in high mortality in symptomatic infants [6]. Improvement in outcomes was achieved with bypass grafting to the coronary artery using either subclavian artery, mammary artery, or saphenous vein grafts [2]. However, recovery of myocardial function was often poor in these patients and late graft occlusion was common. More recently, the concept of establishing a dual-coronary system has been advocated and has been widely accepted as the procedure of choice. This is ideally achieved with direct implantation of the anomalous coronary into the

Fig. 2. Surgical technique for intramural coronary artery. (a) Origin of the anomalous coronary artery from the right PA with an intramural segment in the aortic wall. (b) The origin of the anomalous coronary was excised from the PA and the intramural segment was unroofed. (c) The upper portion of the coronary artery was augmented with a pericardial patch. The defect on the right PA was filled with a pericardial patch.
ascending aorta, to establish a normal anatomical configuration [7]. However, the origin of the coronary may be on the leftward side of the main PA or in the anterior sinus at some distance from the aorta. In this situation, direct implantation may not be feasible. Takeuchi et al. [8] described the technique of intrapulmonary baffle redirection of the anomalous coronary to the ascending aorta, which may be useful in these cases. This procedure, however, is associated with late complications including baffle obstruction, baffle leak, and PA obstruction [9]. In an effort to avoid the use of intrapulmonary baffle, some have used a flap of the main PA wall to extend the length of the LCA with an anterior sinus origin [7,10,11]. Turley et al. [10] reported that this technique was successfully employed in four of 11 patients.

During this study period, continued effort has been made to achieve direct implantation of the anomalous coronary artery. The evolution of this technique has been derived from experience with coronary transfer for the arterial switch operation. By making a trapdoor flap on the aortic wall, direct implantation of the coronary artery is almost always possible without stretching or kinking (Fig. 3). A generous pericardial patch, used for the reconstruction of the PA, extends the posterior wall and reduces tension against the transposed coronary (Fig. 4). Direct implantation was feasible in the 12 most recent consecutive patients in this series, including the single reoperation and the most recent case with origin of the anomalous coronary from the anterior sinus of the PA.

Mitral valve repair was performed in two patients who had significant prolapse of the anterior leaflet resulting from...
ischemic papillary muscle dysfunction. Mitral insufficiency, resulting from annular dilatation without structural deformity, should improve postoperatively without accompanying mitral valve procedures, due to the establishment of normal perfusion in the left coronary system [12]. Schwartz et al. [13] reported that MR with mild to moderate degree tends to improve without mitral valve repair. In our cohort, improvement of MR was observed in six patients at the time of discharge, despite the fact that mitral valve repair was performed in only two patients.

The recovery of the myocardium resulting from the establishment of a dual-coronary system, appears promising according to recent reports [12,14,15]. In our cohort, seven of nine patients who had preoperative ventricular dysfunction demonstrated improvement in ventricular function postoperatively. Normalization of ventricular function has been reported to occur within 2–7 months after dual-coronary repair in ALCAPA which is corroborated in the present study [13,14]. Interestingly, the tendency for recovery from ventricular dysfunction is observed in adults as well as in children. A 20-year-old patient with severe dyskinesia of the RV free wall demonstrated significant recovery postoperatively.

Implantation of the coronary artery into the ascending aorta should be attempted to avoid the late failures associated with either the intrapulmonary tunnel or a bypass graft. Our study suggests that the trapdoor technique may be applicable for the majority of the cases with ALCAPA including those patients in whom the anomalous coronary artery arises from the leftward aspect of the main PA or from the anterior sinus. The technique is simple, reproducible and allows a margin of error in relocating the ALCAPA thereby providing a safe method for the establishment of a dual-coronary system.

References


