Successful Repair of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) using Pericardial Roll Graft: Case Report with Long-Term Follow-up

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SOUHRN

Kontext: Anomální odstup levé věnčité tepny z plicnice (anomalous origin of the left coronary artery from the pulmonary artery, ALCAPA) je vzácná vrozená srdeční vada spojená se závažnými otázkami z hlediska diagnostiky, operativy i postoperační péče.
Popis případu: Popisujeme případ 16měsíčního pacienta, u kterého byla na našem pracovišti provedena chirurgická korekce ALCAPA. Pacient byl dopraven do nemocnice s těžkými symptomy srdečního selhání, jako jsou dušnost, kašel a neprospívání. Vyšetření prokázalo sníženou funkci levé komory a těžkou nedostatečnost mitrální chlopně v důsledku dilatace levé komory. V tomto případě nebyl nalezen kmen levé věnčité tepny, ale dvě ústí (r. interventricularis anterior a r. circumflexus na plicnici); koronární tepnu se nám podařilo zavřít pomocí 4mm záplaty **z** perikardu (pericardial roll extension). Výkon si nevyžádal použití žádného zařízení pro mechanickou podporu srdeční funkce. Dlouhodobé sledování pacienta (7 let) prokázalo, že pacient výkon přežil a jeho stav byl v Rossově klasifikačním systému srdečního selhání (Ross Heart Failure Classification) hodnocen stupněm I. Kontrolní echokardiografické vyšetření prokázalo zlepšení ejekční frakce levé komory a zmírnění nedostatečnosti mitrální chlopně.
Závěr: Chirurgická korekce anomálního odstupu levé koronární tepny **z** plicnice stále ještě představuje náročný výkon, je však vynikající možností, jak zlepšit pacientovu kvalitu života. Korekce s použitím záplaty (pericardial roll graft) by se mohla stát fakultativním výkonem při řešení této anomálie pro obnovení duálního systému koronárních tepen.

Keywords:
Anomalous origin of the left coronary artery from the pulmonary artery
Case report
Congenital heart disease
Long-term follow-up

ABSTRACT

Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect that has serious challenges from diagnostic, surgical, to postoperative management.
Case description: We report a 16-month-old patient, who underwent ALCAPA repair in our center. This patient came to the hospital with severe heart failure symptoms such as shortness of breath, cough and failure to thrive. Decreased left ventricle function and severe mitral valve regurgitation due to left ventricle dilatation was found. In this case, the left main artery (LMA) was not found and there were two ostial of the left anterior descending artery and circumflex artery on the pulmonary artery; we managed the coronary artery transfer by using 4-mm pericardial roll extension. No mechanical assist device was needed. Long-term follow-up of the patient (7 years) showed that the patient survived and had a class I Ross Heart Failure Classification. Follow-up echocardiography revealed improvement of left ventricular ejection fraction and mitral valve regurgitation.
Conclusion: ALCAPA repair is still a challenging procedure but has an excellent opportunity to improve the quality of life of the patient. Pericardial roll graft could be an optional procedure for ALCAPA repair to restore the dual coronary artery system.

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Introduction

ALCAPA is a rare congenital anomaly that often causes myocardial ischemia and infarction within the first few weeks or months after birth, subsequently leading to left ventricular dysfunction, mitral valve insufficiency, and congestive heart failure.[1] Infant with ALCAPA are subject to acute myocardial ischemia because of a lack of well-developed collaterals and at high risk of death without surgical intervention. In this case report, we report the perioperative management of ALCAPA case with long-term follow-up result.

Case presentation

Clinical case

A 16-month-old patient underwent ALCAPA repair in our center. The body weight was 13 kg. The patient presented with severe heart failure symptoms such as shortness of breath, cough and failure to thrive. Preoperative ROSS Heart Failure Classification was class IV. On echocardiography examination, we found decreased ventricular function (left ventricular ejection fraction was 45%) and severe mitral valve regurgitation.

Surgical management

We used standard procedures, including induction of anesthesia, bi-caval cannulation, antegrade cardioplegia and mild hypothermia (32–34 °C). Cardioplegia was administered through cannulation in the aortic root with the addition of selective perfusion of the left coronary artery using an olive-tipped catheter after opening the pulmonary artery. Interestingly, the left main artery (LMA) was not found and there were two ostial of the left anterior descending artery and circumflex artery on the pulmonary artery. Since the direct implantation was not possible, the reimplantation of the left coronary artery was performed with the addition of a 4-mm pericardial roll as an interposition graft (Figs 1A, 1B). We used 7-0 nonabsorbable monofilament sutures to construct a 15 mm length of pericardial roll over the Hegar dilator. The anastomoses were created using a continuous 7-0 nonabsorbable monofilament suture. The conduits were secured to the epicardium to relieve tension and prevent angulation. The cross-clamp time was 90 minutes and the cardiopulmonary bypass time was 192 minutes. The chest was closed without significant hemodynamic changes and transferred to the pediatric intensive care unit.

We found no major cardiac adverse event (MACE), such as malignant arrhythmia during post-operative care. The patient was extubated 48 hours after surgery. No mechanical-assisted device was used postoperatively. The ICU length of stay was 9 days and the patient was discharged on the day 19 after surgery.

The long-term result was evaluated. The ROSS Heart Failure Classification was class I. The patients also had no hospital re-admission history related to cardiac problems. Latest echocardiography data were collected and we found that mitral regurgitation was significantly improved postoperatively with better left ventricular ejection fraction (Fig. 1C).

Discussion

Various surgical methods are performed to increase the survival rate of ALCAPA patients. Earlier, ligation of the anomalous artery originating from the pulmonary artery was performed. However, this method causes complete dependence on the collateral circulation, resulting in a higher initial mortality rate than expected.[2] Currently, the success of ALCAPA repair surgery depends on the success of the dual coronary artery system. Several techniques, such as Takeuchi procedure or direct reimplantation of LCA are the procedure of choice to achieve the best vascularization.[3,4] Several reports have confirmed that the direct reimplantation of the anomalous coronary into the ascending aorta has good early and late results.[5] However, stenosis or total occlusion of the LCA occasionally can be seen after repair. Factors related to this complication are short-length of LCA, the origin of the LCA from the lateral

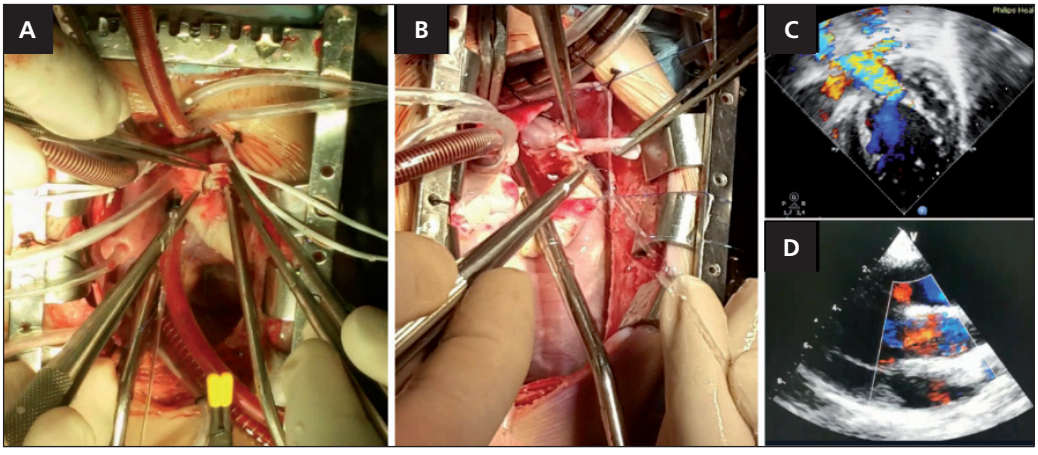

Fig. 1 – Coronary reimplantation to the aortic root using 4-mm pericardial roll (A and B); echocardiography prior to surgery (C) and at 6 years after operation (D) showed improvement of mitral valve regurgitation.

aspect of the PA, and compression by the repaired PA. In addition, the vicious circle of ischemia, LV distension, and increased PA pressure may play a role in stretching the re-implanted coronary artery.[6]

Direct reimplantation of LCA to the aortic root might not be possible and need an additional conduit to minimalize the chance of a kinked artery, such as subclavian artery graft of PTFE graft.[6,7] As we found in our case, the left main artery was unavailable and there were 2 ostia of left anterior descending and left circumflex artery. Mobilization of both the coronary artery to the aortic root was not possible. We used a 4-mm pericardial roll due to the absence of the left main artery. Post operatively, we found no ST-elevation changes and no increment of cardiac marker. We also found that long-term follow-up of the pericardial roll graft showed good flow patency. This method could be an optional procedure for ALCAPA repair.

Normalization of LV function occurs in ALCAPA patients, and some patients experience improvement within 1 month, 50% improvement in less than 1 year.[8] A possible mechanism for restoration of cardiac function after the repair is myocyte hyperplasia in young patients. The potential for complete myocardial recovery after revascularization following chronic myocardial ischemia in this patient can be explained by the hibernation phenomenon.[8] In our study, we found that all the patient who survived had class I ROSS Heart Failure Classification which means that the heart function was significantly improved after the surgery with improvement of heart contractility and regurgitation.

Conclusion

The success in the management of patients ALCAPA is a serious challenge of diagnostic, surgery techniques, cardioplegia techniques and postoperative treatment. The restoration of dual coronary artery system is the best option to achieve the best vascularization of myocardium. After long-term follow-up, all of the patients who survived showed significant improvement of heart function. Therefore, ALCAPA repair is still a challenging procedure but has a good opportunity to improve quality of life of the patient.

Conflict of interest
The authors declare that there is no competing interest regarding the manuscript.

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Ethical statement
All patient parents / legal guardians in this study have understood and agreed to the use of patient personal data related to the writing of scientific articles.

Author contribution
DF was responsible for drafting, literature review, and editing of the manuscript. WA, AH, PB, PR and EM were responsible for literature review and editing. All authors have read and approved the final manuscript.

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