Pulmonary artery banding as adjunct therapy for ventricular recovery after ALCAPA-repair: a case report

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Case Report

Keywords: ALCAPA, pulmonary artery banding, dilated cardiomyopathy

Posted Date: March 25th, 2022

DOI: https://doi.org/10.21203/rs.3.rs-1482454/v1

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Abstract

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly commonly associated with severe but reversible left ventricular (LV) dysfunction.

We present an ALCAPA case of persisting left ventricular failure with inability to wean off the ventilator and inotropes after successful coronary reimplantation, in whom pulmonary artery banding enabled bridging to myocardial recovery. Pulmonary artery banding may be considered as a bridge-to-recovery in selected cases of severe left ventricular dysfunction after ALCAPA-repair.

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary anomaly with an estimated incidence varying from 1 in 4000 to 1 in 300,000 births [1–3]. Due to low pressure perfusion with de-oxygenated blood (or even coronary steal phenomenon), chronic myocardial ischemia can lead to myocardial infarction and progressive dilated cardiomyopathy [4]. Depending on the severity of ventricular dysfunction, moderate to severe mitral regurgitation is seen at the time of diagnosis [5]. If left untreated, ALCAPA is associated with a high mortality, especially when presentation is at a young age [4]. Currently, surgical correction aims to restore antegrade coronary perfusion by direct coronary reimplantation or intrapulmonary tunnel-type repair after which spontaneous and progressive myocardial recovery is observed in most cases [1, 6]. However in some patients temporary post-operative mechanical circulatory support (MCS) – either by extracorporeal membrane oxygenation (ECMO) or left ventricle assist devices (LVAD) – is required to allow myocardial recovery (CPB) [7]. A recent publication from the European Congenital Heart Surgeons Association Database reports on an in-hospital mortality of 6% (out of 907 patients undergoing ALCAPA repair between 1999–2019) [7]. In this cohort MCS was required in 7.3% of patients, almost exclusively in infants, resulting in a significantly higher mortality [7].

Whereas treatment/bridging options for dilated LV cardiomyopathy (LV-DCM) now include pulmonary artery banding (PAB)[8, 9], this approach was previously not considered for the failing and dilated LV after ALCAPA-repair. We present a case of persistent circulatory failure, despite coronary reimplantation, where PAB proved a successful bridge to ventricular recovery.

Case Presentation

A 10-month old infant was admitted at our Paediatric Intensive Care Unit because of failure to thrive (weight Z-score −2.8 and length Z-score −1.2), tachypnea and excessive sweating during feeding. Echocardiography showed severe left ventricular dilatation (LV EDD 50mm; Z-score +11) (Fig. 1), grade II mitral regurgitation and poor systolic function (EF 23–28%). The diagnosis of ALCAPA was made by transthoracic echocardiography (Fig. 2). Preoperative stabilization was initiated by administration of loop...
diuretics and surgery was scheduled. Coronary reimplantation was performed successfully and weaning of CPB was achieved with epinephrine and levosimendan.

Two attempts at ventilatory weaning on post-operative day (POD) 6 and 16 proved unsuccessful with need for reintubation. On POD 21 there were still remnant clinical signs of circulatory failure (prolonged capillary refill time, cold extremities, hepatomegaly and feeding intolerance), a high inotropic need (combination of milrinone, epinephrine and norepinephrine) and a persistently severely dilated left ventricle (LV EDD 53mm; Z-score +12.3) with poor LV function on cardiac ultrasound.

Considering the predominant LV dysfunction and preserved RV function, pulmonary artery banding (PAB) was forwarded as a valid option to aid weaning from inotropic and ventilatory support. On POD 27, PAB was performed after pre-operative circulatory optimization with levosimendan. The tightness of the banding was guided by transoesophageal echocardiographic evaluation of ventricular dimensions and the position of the interventricular septum (IVS). At an RV pressure of 55mmHg (for a systemic pressure of 80mmHg) a mild shift of the IVS was obtained without compromising the circulation. Recovery from the PAB was uneventful, inotropics could be completely stopped within 4 days. Finally, respiratory support was withdrawn at day 15 after PAB (36 days after initial repair).

During follow-up progressive recovery of systolic function and decrease of left ventricular dimensions ensued (Fig. 3), accompanied by gradual clinical improvement. After 1 year the child was doing well, showing complete recovery of LV function (LV EDD z-score −1.6 and EF 73%) on echocardiography. Since the gradient across the banding had increased to 80mmHg, partial de-bandaging was performed by balloon dilatation in the cathlab with a Powerflex balloon, alleviating RV pressure-overload and normalizing septal position while preserving normal LV function (Fig. 1).

**Discussion**

After ALCAPA-repair, recovery of systolic LV function typically takes several months, in some cases even up to 2 years [5, 10, 11]. Although temporary mechanical circulatory support (by ECMO or LVAD) is indicated in patients with ALCAPA who can't be weaned off bypass [5, 10]; in the child presented here three weeks had already passed since corrective surgery and initial successful weaning off CPB but at the cost of persisting clinical heart failure and need for inotropic and ventilatory support. Thus, a bridging-solution enhancing recovery was needed, taking into account that a longer-term MCS in such small-size infants is associated with a considerably high complication rate as infection, thrombosis, or bleeding [9]. After successful use as bridge-to-recovery in LV dilated cardiomyopathy, the use of PAB has recently been proposed as an interesting bridge-to-weaning adjunct in LV-DCM after cardiac surgery [8].

Since recovery of LV function was expected with time and RV function preserved, our patient proved a potential candidate for PAB. Although little change in echocardiographic measurements was recorded during the first days after the procedure, benefit of this mechanical improvement was clear by gradual weaning of inotropes and ventilatory support in the following days. The combination of pressure-loading of the RV and improving the LV function efficacy through restoring the LV geometry related to the septal
shift, enabled to surpass the critical phase of inotropic-dependent heart failure. Then gradual but slow LV myocardial recovery characteristic to deep myocardial stunning resulted in complete normalization of echocardiographic measures of the left ventricle after one year. To our knowledge, this is only the second time PAB was used successfully as a bridge-to-recovery after ALCAPA-repair [8].

Conclusion

In a case of persistent circulatory failure after corrective surgery for ALCAPA due to LV failure in the presence of normal right heart function, PAB may be considered as a helpful adjunct to enhance ventilatory and inotropic weaning. Gradual myocardial recovery resulted in further resolution of clinical symptoms, with echocardiographic measures of left ventricle function normalizing after one year.

Statements And Declarations

The authors have no competing interests to declare that are relevant to the content of this article. The authors did not receive support from any organization for the submitted work. Informed consent was obtained from legal guardians.

References


Figures

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Figure 1

Echocardiographic evolution on 4-chamber and short axis views
Figure 2

Echocardiographic diagnosis of ALCAPA

Images show Left Coronary Artery originating from the pulmonary artery with reversed flow indicating coronary steal.

Figure 3
Evolution of LV end-diastolic dimension (as Z-score)

LV EDD: left ventricle end-diastolic diameter; POD: post-operative day(s); LV: left ventricle