CASE REPORT

Case Report: ALCAPA syndrome: successful repair with an anatomical and physiological alternative surgical technique [version 1; peer review: 1 approved, 2 approved with reservations]

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Abstract

Anomalous left coronary artery from the pulmonary artery, or ALCAPA syndrome, is a rare congenital cardiac disease that can cause myocardial infarction, heart failure and even death in paediatric patients. Only few untreated patients survive until adult age. Here we present the case of a 33-year-old female patient with paroxysmal tachycardia, syncope and little effort dyspnoea. She was diagnosed with ALCAPA syndrome and underwent surgical correction with an alternative technique of left main coronary artery extension to the aorta.

Keywords

Coronary vessel anomalies , ALCAPA syndrome , Bland-White-Garland syndrome , Adults with ALCAPA , Coronary extension technique

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Approval Status

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Any reports and responses or comments on the article can be found at the end of the article.

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Introduction

ALCAPA syndrome, also known as Bland-White-Garland Syndrome, is a rare congenital heart disease, affecting approximately 300,000 newborns in the USA. This disease has 90% of mortality within the first year of life in untreated patients, due to myocardial ischemia and heart failure. Approximately 18–25% of patients with this congenital heart disease reach adulthood, presenting arrhythmias, heart failure and myocardial ischemia¹. Treatment of the anomalous origin of the left coronary artery from the pulmonary artery includes several surgical techniques, however they are all associated with important morbidity (21%)^{2–5}.

In the adult population, surgical correction is more difficult to resolve, due to the heart dimensions and compensatory disorders in coronary circulation to the left ventricle. We present the case of a 33-year-old female with ALCAPA syndrome and mitral valve severe regurgitation, who underwent successful correction with a physiological and anatomical technique.

Clinical case

A 33-year-old female with medical history of recurrent respiratory infections since childhood, paroxysmal tachycardia in adolescence and some syncope episodes in adulthood accompanied by retrosternal pain during exercise. Physical examination revealed a mitral murmur III/IV. The paraclinical diagnostic methods showed anomalous emergency of left main coronary artery from the pulmonary artery, the right coronary artery dilated, the left ventricle dilated and regurgitant flow in mitral valve (Figure 1).

Surgical technique and postoperative follow-up

Sternotomy and surgical procedure were performed with circulatory support to hypothermia (28°C). The mitral valve was replaced by Mechanical Sorin Carbomedics® valve No 27 to correct valvular dysplasia. The left main coronary artery button was dissected and then connected to a duct constructed with pulmonary wall and bovine pericardium to be anastomosed to the aorta artery. The pulmonary artery was reconstructed with Woven Dacron graft, leaving the previously constructed duct in the back of the Dacron graft. The surgical findings were: right coronary (RCA) dilated and collateral circulation from RCA to left ventricular circulation, LMA arising from the MPA, dysplasia of posterior mitral valve. At 6 months follow-up, the patient remained in functional class I of New York Heart Association and AngioCAT showed patency of the new ductus (Figure 2).

Discussion

ALCAPA congenital anomaly is a rare disease that must be surgically treated in the first year of life. However, between 10–15% of patients reach adulthood and clinically manifest rhythm disorders usually attributed to alterations of the cardiac electrical system, which obscures the underlying pathophysiology of myocardial ischemia^{1–5}.

The blood flow restauration in left main coronary artery from the aorta is the primary objective in the surgical correction of ALCAPA, and there are several surgical options in the paediatric population. Derivation of the left subclavian artery and implementation of an aorto-coronary bypass with saphenous vein or the left internal thoracic artery to the left anterior descending coronary have shown

Figure 1. A – Arrow – RCA dilated arising from the aorta. B – Arrow – LMA arising from lateral aspect of the MPA.

Figure 2. AngioCAT – Arrow – Adequate graft patency (combined pulmonary tissue and bovine pericardium patch).

low short-term effectiveness (60%) and high morbidity with stenosis and thrombosis of bypass graft^{10–14}. The Takeuchi procedure is the most used in the paediatric population, however it has a high incidence (> 21%) of supravulvar stenosis of pulmonary artery¹⁵.

In the first year of life, great arteries are not fully developed and tissues are more “flexible”, which allows a coronary reimplantation. However, child’s growth promotes stenosis in short and medium-term^{7–9}. The major anatomical distances and the less “flexible” tissues in adult patients make the surgical restauration of the left main coronary artery blood flow more difficult. Our surgical team resolved this situation with a duct constructed with pulmonary artery wall (80%) and bovine pericardial patch (20%), leaving this duct in anatomical position behind the Woven dacron graft used for restitution of blood flow in main pulmonary artery (Figure 3). We believe that anatomical position of the new duct permits a physiologic blood flow like in a normal heart. In our case, ischemic symptoms resolved and the patient maintained good functional class at 6 months follow-up and full patency of the graft in AngioCAT.

Figure 3. **A** – LMA taken from the MPA and reconstructed as a tubular structure with bovine pericardium. **B** – LMA anastomosis to the Ao as in a normal position, MPA reconstructed with a pericardial patch. **C** – MPA reconstructed with a Dacron graft.

The ALCAPA physiopathology consists of a relative coronary steal, which promotes low oxygenation in the left myocardial tissue as a consequence of blood flow from pulmonary artery which leads to myocardial ischemia and acute myocardial infarction. The low oxygenation circumstance promotes collateral vessels development and right coronary dilatation, as can be seen in [Figure 4](#). On the other hand, the chronic myocardial ischemia produces papillary muscle and ventricular lateral wall dysfunction, which causes mitral insufficiency. All of this would explain the symptoms presented by the patient.

Mitral insufficiency treatment is still under discussion; some authors prefer valvular reconstruction, considering that the failure is due to papillary muscle dysfunction; nevertheless, an important proportion of insufficiency recurrence still exists. In a

sense other authors prefer to replace the mitral valve with a valvular prosthesis. In the case that we presented, the surgical team observed a valve dysplasia which prevented valvular reconstruction, so it was decided to replace the mitral valve with a mechanical prosthesis.

To summarise, the ALCAPA or Bland-White-Garland syndrome treatment is a real surgical challenge in the adult population. However, we believe that the alternative procedure presented in this article consisting of pulmonary artery wall and bovine pericardial construction of a new duct, which connects the left main coronary artery re-establishing a normal anatomical situation and permitting a physiological blood flow to left ventricle, are a viable and probably successful surgical alternative in adult patients without risk of pulmonary stenosis.

Figure 4. ALCAPA syndrome pathophysiology.

Consent

Written informed consent for publication of their clinical details was obtained from the patient.

Author contributions

Vilá Mollinedo Luis Gustavo (Research, article description and development), Andrés Jaime Uribe (Master Supervision), Jose Luis Aceves Chimal (Article development, Master supervision), Rest

(Research, obtaining consent, photography). All authors agreed to the final content of the article.

Competing interests

No competing interests were disclosed.

Grant information

The author(s) declared that no grants were involved in supporting this work.

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Roxanne E. Kirsch

Division of Cardiac Critical Care, Department of Critical Care and Anesthesiology, and Department of Bioethics, The Hospital for Sick Children, University of Toronto, Toronto, ON, Canada

Thank you for this concise description of the adult presentation of ALCAPA and offer of an alternate surgical procedure for restoration of coronary blood flow from aorta to LCA.

This presents another surgical option for the armamentarium. While you make biologically plausible suggestions as to why this procedure may prove superior, the actual superiority cannot yet be known as only the passage of time and experience with the technique will determine that. This should be more explicitly stated, so as not to be misleading. In that bent, I might revise the final sentence to: "...permitting a physiological blood flow to the left ventricle is viable and may prove to diminish the risk of pulmonary stenosis post repair of ALCAPA compared with other techniques."

As a non-surgeon, and a pediatrician, I cannot comment on the likelihood of improved coronary patency personally.

Also, it might be helpful for the reader to know the degree of residual mitral regurgitation at the six month follow up, and if you intervened on the valve during the surgical repair (I do not believe you did from my review of this case presentation), and whether this patient's presenting symptoms had abated at the time of 6 month follow up.

My additional minor suggestions are:

1. the sentence: ...the paraclinical diagnostic methods showed anomalous emergency of left main coronary artery.... should read: "emergence of the left main coronary artery"
2. I would consider an alternate term for the use of "duct". This initially seemed like it was referring to a ductus arteriosus before I rapidly realized this was referring to the "tube graft" you had created.

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Reviewer Report 26 July 2016

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Stephan Achenbach

Department of Internal Medicine II (Cardiology), University of Erlangen, Erlangen, Germany

There is nothing wrong with the description of a surgical technique for ALCAPA repair in the form of a case report. This, however is no proof of superiority over other approaches.

The main concern is presentation:

"ALCAPA syndrome, also known as Bland-White-Garland Syndrome, is a rare congenital heart disease, affecting approximately 300,000 newborns in the USA." Affecting 300,000 newborns? In which time period?

"This disease has 90% of mortality"; "important morbidity"; "surgical correction is more difficult to resolve"; "The paraclinical diagnostic methods showed anomalous emergency of left main coronary artery from the pulmonary artery"; "physiopathology" - examples of incorrect use of the English language.

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard, however I have significant reservations, as outlined above.

Author Response 26 Jul 2016

Luis Gustavo Vilá Mollinedo, National Medical Centre 20 de Noviembre, ISSSTE, Mexico City, Mexico

First of all, thank you for your time and recommendations.

Addressing all this:

- We're presenting this approach as an alternative, also we think it's a bit superior

having a coronary extension than a baffle procedure, due to the kinking or obstruction probability of these, but also just displaying what we did.

- I have changed the English use of all the terms and sentences you have proposed, although our English isn't great, we hope to have improved the readability.
- Finally it was our mistake, the sentence about the incidence should read "Affecting 1 of 300,000 newborns".

Thanks for your support, any other outlining would be appreciated.

Competing Interests: No competing interests were disclosed.

Reviewer Report 25 July 2016

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Aiden Abidov

Department of Medicine and Radiology, University of Arizona College of Medicine, Tucson, AZ, USA

A few minor things:

- Abstract: I recommend using a term "mild exertional dyspnea" instead of " little ...dyspnea".
- Figure 3: I would rather remove the Legend from the Figure (makes it less crowded).
- Figure 1 - would show a 3rd plate with a longitudinal view of the LM artery (if available) for a better understanding of the anatomy. Just an origin view is not that impressive.

Competing Interests: No competing interests were disclosed.

I confirm that I have read this submission and believe that I have an appropriate level of expertise to confirm that it is of an acceptable scientific standard.

Author Response 26 Jul 2016

Luis Gustavo Vilá Mollinedo, National Medical Centre 20 de Noviembre, ISSSTE, Mexico City, Mexico

First of all, thank you for your evaluation, really helpful, I changed the term of dyspnea for the one you suggested. As for the Figures, we have removed the legend, and as for the CT scan I do not have the one with the longitudinal view.

Thanks for your help again.

Competing Interests: No competing interests were disclosed.

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