The authors should be congratulated in reporting such an interesting and rare anomaly.

My only question would be for the authors to elaborate further on the reasoning for choosing saphenous vein graft as a conduit of choice in this young 48-year-old patient and not the left internal thoracic artery (LITA).

1.1 Comment: I do not understand their explanation. Since this individual does not have evidence of coronary artery disease, the utilization of LITA, would not have created inadequate flow in the left anterior descending coronary artery.

Answer: We would like to thank the reviewer, this is probably the best question and challenge about this particular case, and we strongly agree this would need further discussion. Indeed, one the best proposed treatments to treat this pathology is to revascularize the myocardium by providing dual coronary perfusion by either re-implanting the left main coronary into the aorta whenever possible, or buy using a graft to bypass the left myocardial territory after ligating the left main. The few case reports and literature suggest, in the young patients with ALCAPA, to use LITA to revascularize the left anterior descending artery (LAD) when appropriate.

Intra-operatively, we were surprised by the fragility of the tissue, and the enormous size of the LAD. Of note, this case presents one of the oldest patients ever operated for this pathology, and issue with the enlarged vessels and collaterals posed to us serious challenge during the surgery. We believed that a large saphenous vein harvested in the calf, compared to a smaller LITA, would better fit the size and the required flow to bypass this enormous LAD vessel. Supporting our decision, a significant back flow was found when the LAD was opened to create the anastomosis on the beating heart, suggesting important collateral blood flow to the LAD territory. Moreover, we were unable to achieve complete cardioplegic arrest, and were forced to use circulatory bypass and the beating heart to revascularize the myocardium, suggesting significant bronchial artery collateral blood flow.

In order to match the size of the large LAD, allow unrestricted flow and avoid competitive flow between the graft and the collaterals from the RCA, the largest possible conduit was used. The choice of a large saphenous vein conduit over an arterial graft (such as the internal thoracic or the radial artery) was in response to this situation. General guidelines of coronary revascularization to treat coronary artery disease (CAD) strongly recommend the use of LITA to bypass the LAD due to its long term reported permeability. In the other hand, there is paucity of data comparing the long term patency of LITA versus saphenous vein to bypass the LAD in ALCAPA patient. In a series of 6 adult patients who underwent saphenous vein bypassgrafting and direct ALCAPA closure from inside the PA, Moodie and associates reported a graft patency rate of 80% at a mean follow-up of 5.8 years. (Moodie DS, Fyfe D, Gill CC, et al. Anomalous origin of the left coronary artery from the pulmonary artery (Bland- White-Garland syndrome) in
adult patients: long-term follow-up after surgery. Am Heart J 1983;106:381–8.)
Moreover, 10-year patency of a SVG to a LAD larger than 2.0 mm is 88% at 10 years,
as reported by Goldman and al. (Long-Term Patency of Saphenous Vein and Left Internal Mammary Artery Grafts After Coronary Artery Bypass Surgery, JACC 2004).

1.2 Comment: Also they should explain why they did not consider an interposition arterial graft with end to end anastomosis between the aorta and the left main artery.

Answer: Intra-operatively, we found an enormous LAD, with several collaterals smaller vessels. The initial step was to encircle the left main coronary to have proper control to perform a safe ligation. However, we were surprised by the fragility of the tissue and the active bleeding coming from behind the coronary due to the small collaterals. Having performed double ligation of the left main without complication, we decided not to perform an end to end anastomosis to the left main coronary because of the frailty of the surrounding tissues and the inability to mobilize the bypassed vessel sufficiently to achieve an hemodynamically favorable end-to-end anastomosis.

Reviewer #2: Jean-Francois Legare

Well-written manuscript.
Interesting case but it was unclear to this reviewer how the management or the case fits in what is already available from the literature.
Suggested major revisions:

2.1 Comment: Comment and expand on how the management of this patient differs from the several case reports seen in the literature

Answer: There is a paucity of report on adult patients operated for ALCAPA syndrome. Usual management on youngest patients is more straightforward and involves the re-implantation of the left main coronary to the aorta, the simple ligation of the left main, or the bypass of the coronaries. Because this patient was treated in his adult life, and the coronary arteries enlarged with time, we were faced with enormous coronary vessels and significant collateral blood flow. Of note, compared with younger patients who are treated in their first years of life with significant coronary blood flow steal due to left to right shunt, this older patient demonstrated little myocardial ischemia. In this particular case we are reporting, it is possible that the ligation of the left main would suffice to terminate the right to left shunt and coronary steal. We believe this case should be reported because of the late presentation, and the few symptoms related to myocardial ischemia reported by the patient.

2.2 Comment: highlight the novelty of the present case report when compared to previous reports.
**Answer:** This case report is of interest because this is one of the oldest ALCAPA patient ever operated. Please refer to comment #1. This patient presented little myocardial ischemia compared to younger patients who usually suffer from myocardial infarct and heart failure at time of diagnostic or first symptoms. Timing of surgery and management in this adult patient with almost no ischemic symptoms warrant report and discussion, especially on the necessity of bypassing the left myocardial territory after the ligation of the left main coronary.

**2.3 Comment:** given the choice of SVG for revascularization some additional data would greatly help the present manuscript

**2.3.1 Flow data on the graft done and discussion on the potential impact of extensive collaterals**

**Answer:** We decided to use a large saphenous vein graft to bypass this enormous LAD in order to match the size and blood flow. The extent of adaptative collateral flow could have caused competitive flow with the graft, but that was not the case as we measured an excellent flow. SVG flow was measured at 117ml/min with the MediStim VeriQ Flowmeter at the time of surgery which is a lot more than we usually measure in LITA grafts. We believe this larger graft will provide adequate flow and long-term patency to overcome extensive collaterals.

**2.3.2 Follow-up stress test or imaging:**

**Answer:** Unfortunately there is no follow-up stress test or imaging performed to date. However, patient's NYHA class is 1, and patient did not report any chest pain on exertion. Of note, patient is very active and initiated an exercise program to lose weight.

**2.4 Comment:** discuss how this patient may have survived into his 40's without significant myocardial damage.

**Answer:** We thank the reviewer for this excellent question, and this question supports the importance to report this clinical case. ALCAPA is usually found in young patients in their first years because of significant tissue ischemia resulting in myocardial infarct and heart failure. This 48-year-old patient did not present significant clinical signs of myocardial ischemia, probably due to adequate and compensated collateral blood flow from the right to the left coronary arteries. However, this patient reported dyspnea on exertion and possibly mild chest pain. It is possible that, in this particular case, this patient survived into his 40's because of adaptive collateral blood flow in the myocardium, but symptoms were related to left to right shunt and volume overload.

**2.5 Comment:** discuss potential sacrifice on longevity of using SVG.

**Answer:** In a series of 6 adult patients who underwent saphenous vein bypass grafting and direct ALCAPA closure from inside the PA, Moodie and associates reported a graft patency rate of 80% at a mean follow-up of 5.8 years. (Moodie DS, Fyfe D, Gill CC, et

Moreover, a SVG flow of 120ml/min was measured at the time of surgery which is significantly higher than what we routinely measure in LITA pedicles. We believe that this high blood flow in the SVG, along with dual antiplatelet treatment (AAS and Clopidogrel) will result in excellent long term patency.

In addition, 10-year patency of a SVG to a LAD larger than 2.0 mm is 88% at 10 years, as reported by Goldman and al. (Long-Term Patency of Saphenous Vein and Left Internal Mammary Artery Grafts After Coronary Artery Bypass Surgery, JACC 2004).

Up to now, no difference in mortality or left ventricular function has been demonstrated in the literature between techniques providing a dual coronary perfusion system (saphenous vein graft, left internal thoracic artery graft, left subclavian anastomosis and direct aortic implantation). (Dodge-Khatami A, Mavroudis C and Backer CL: Anomalous origin of the left coronary artery from the pulmonary artery: Collective review of surgical therapy. Ann Thorac Surg 2002; 74:946-955.)

There is however a difference in long-term mortality between the aforementioned techniques and an isolated LMCA ligation providing a simple coronary perfusion system.

Reviewer #3: Qingyu Wu

3.1 Comment: Left coronary artery from the pulmonary artery must affect the myocardial blood supply in diastolic period even though there is collateral flow from the right coronary artery, and operations of patients with symptom should be done.

Answer: We agree with the reviewer.

3.2 Comment: Up to now, to establish the coronary circulation system is the best choice for correcting this abnormality, which is not much difficult even for neonatal or infant.

Answer: We agree with the reviewer, this is the best treatment whenever anatomically possible and safe.

3.3 Comment: It needs to use the fresh autograft pulmonary artery wall to extend the left main coronary artery, and then implant to the aortic-root afterwards.

Answer: We agree, this is a very good surgical strategy, we included this alternative to the manuscript. (Wu QY, Xu ZH.Surgical treatment of anomalous origin of coronary artery from the pulmonary artery. Chinese Medical Journal 2008; 121(8):721-724)

3.4 Comment: The patent rate of using right saphenous vein graft is not satisfactory, which is only 45%-55% in ten years, so the procedure reported in this paper is not the best technique.

Answer: Please refer to comment 1.1, 2.2 and 2.5.