



Fontan completion of a 10-year-old Kawashima patient with extensive arteriovenous malformations: consideration for a lobectomy

Brief Report

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Author for correspondence:

Şima Kozan, School of Medicine, Koç University, Istanbul, Turkey. E-mail: skozan18@ku.edu.tr

Mehmet Biçer¹ , Şima Kozan²  and Metehan Kızılkaya³

¹Department of Pediatric Cardiovascular Surgery, Koç University Hospital, Istanbul, Turkey; ²School of Medicine, Koç University, Istanbul, Turkey and ³Department of Pediatric Cardiology, Koç University Hospital, Istanbul, Turkey

Abstract

A significant contributing factor to the progression of late cyanosis in individuals undergoing Kawashima operation is pulmonary arteriovenous malformations. Following the Fontan procedure, arteriovenous malformations may regress. However, in cases with extensive malformations causing severe cyanosis, lobectomy can also be a possible treatment approach. Thereby, we present our two-step treatment strategy in a late Fontan completion complicated by arteriovenous malformations in a Kawashima patient.

Case presentation

A 10-year-old male patient was referred to our clinic with a history of cyanosis and dyspnoea with a resting saturation of 40–55% and a haematocrit value of 65%. Echocardiographic findings included an anatomy demonstrating an operated Kawashima patient with moderate/severe insufficiency on unbalanced atrioventricular valves with an ejection fraction of 45% and left atrial isomerism with hemiazygos continuation (Video S1 and Video S2). There were frequent ventricular extrasystoles on electrocardiography. At cardiac catheterisation, pulmonary arterial pressure was 14 mmHg, and severe arteriovenous malformations were observed in the left lung (Video S1). Following the balloon occlusion of the left pulmonary artery, saturation increased to 90% and pulmonary artery pressure to 25 mmHg. A two-step surgical approach was planned with a left lower lobectomy prior to the Fontan operation. Following lobectomy, the saturation increased to 68% and the Kawashima pressure to 16 mmHg.

Two weeks later, Fontan completion with intra-extra cardiac conduit and atrioventricular canal repair with polytetrafluoroethylene strip over the bridging leaflets was performed (Video S2). Conduit was anastomosed towards the centre of pulmonary arteries with no fenestration. The patient was weaned from bypass and extubated in the operating room with a saturation of 86%, Fontan pressure of 18 mmHg and mild regurgitation of atrioventricular valves. The patient was on temporary pacemaker support due to complete atrioventricular heart block.

On the postoperative 4th day, an episode of cardiac arrest occurred due to rhythm disturbances and on the 6th day, an implantable cardioverter-defibrillator was implanted. Following the arrest, regurgitation of the atrioventricular valves increased to moderate. Post-operative course was prolonged due to ascites occurrence managed with vasopressin receptor-2 antagonists. Patient was discharged on the post-operative 67th day. In the outpatient follow-up, it was reported that the patient was clinically stable with a persistent saturation of 80–85% which were unchanged from levels recorded at the time of discharge.

Discussion

The approach to late Fontan completions in Kawashima cases complicated by extensive arteriovenous malformations remains unclear. It was previously reported that a gap of more than two years until Fontan completion is an independent risk factor for the emergence of malformations after the Kawashima procedure.¹ Our patient had a 9-year delay after his initial operation. This can explain the extensive malformations present that were causing severe cyanosis. Thus, we agree that following Kawashima operation, Fontan completion should be performed without a delay to avoid progressive cyanosis.

In Fontan patients with arteriovenous malformations, approaches that potentiate a nonfenestrated strategy have been discussed in the literature.² This patient had significant arteriovenous malformations that served as an escape route, favouring a nonfenestrated Fontan completion procedure. Further, we thought the malformations would act to lower the Fontan pressure by lowering the transpulmonary gradient in the early post-operative period undertaking the function of a fenestration. Notably, the possibility of a future angiographic

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intervention to create a fenestration has been acknowledged and considered during the planning and follow-up of this case as a management strategy against Fontan failure.

Fontan completion operation may lead to regression of arteriovenous malformations; nevertheless, the extent of this regression is unclear, and persistent post-Fontan malformations requiring further intervention may occur.^{1–3} Possible treatment options in cases of severe extension of these malformations, particularly with recurrent haemoptysis, include conservative lung resection, local resection, segmentectomy, and lobectomy.^{3,4} Treatment should be individualised and based on the size, number, and location of the lesions as well as the age and general condition of the patient.^{3,4} Our decision to perform an initial lobectomy was made in conjunction with the improvement in saturation with the occlusion of left pulmonary artery during catheterisation. Our intention was for the hibernated cardiac functions to improve by virtue of the increased saturation levels and distribute the operative risk.

Regarding the later valvular insufficiency in the common atrioventricular valve, future surgical intervention is anticipated and will be planned according to the patient's status. Furthermore, given the predicted regression of arteriovenous malformations and the lack of a fenestration, patient is being closely monitored with frequent follow-up for a potential Fontan failure. Overall, the two-step strategy to Fontan completion with an initial lobectomy could be viewed as a feasible approach.

Conclusion

In patients with the Kawashima circulation, the treatment plan for pulmonary arteriovenous malformations is still unclear. Prior to Fontan completion surgery, a surgical treatment approach to arteriovenous malformations by way of a lobectomy can be considered especially in late completions with extensive malformations and cyanosis.

Supplementary material. To view supplementary material for this article, please visit <https://doi.org/10.1017/S1047951123000288>

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Conflicts of interests. None.

Ethical standards. The study was approved by Koç University Committee on Human Research. Ethics approval protocol number: 2022.436.IRB1.162. Written informed consent was obtained from the patient's guardian.

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