

CASE REPORT

ADVANCED

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE

Hemolytic Anemia After an Aortic Dissection Treated With a Valve-Sparing Aortic Root Replacement



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ABSTRACT

A 46-year-old man with a personal history of a repaired aortic dissection was admitted because of hemolytic anemia. The transesophageal echocardiogram displayed an accelerated flow and a residual intimal flap in the proximal descending aorta. A total arch replacement was performed, the flap was removed, and his hemolytic anemia was resolved. **(Level of Difficulty: Advanced.)** (J Am Coll Cardiol Case Rep 2023;18:101921) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

CASE PRESENTATION

A 46-year-old man was presented to the hospital because of asthenia and shortness of breath that had begun 2 weeks prior. Patient denied chest pain and palpitations. His past medical history included a Stanford type A acute aortic dissection (AAAD), requiring a valve-sparing aortic root replacement (David procedure) and hemiarch replacement 6 months ago. No other data were remarkable in his

medical history. His medical treatment was aspirin 100 mg once daily. On arrival, vital signs were as follows: temperature 96.8 °F, heart rate 115 beats/min in sinus rhythm, blood pressure 101/57 mm Hg, and oxygen saturation was 98% on room air. Physical examination demonstrated a systolic murmur in the suprasternal focus. No signs of congestive heart failure were detected. The patient's electrocardiograph showed sinus rhythm at 120 beats/min. His blood tests revealed severe anemia, elevated levels of bilirubin and lactate dehydrogenase, and low levels of haptoglobin, leading to a diagnosis of hemolytic anemia (HA).

LEARNING OBJECTIVES

- To identify hemolytic anemia as a rare but relevant complication after aortic surgery.
- To recognize the presence of a residual intimal flap as a potential etiology of hemolytic anemia owing to an accelerated flow between the true and false lumen.
- To know the different mechanisms that lead to hemolysis after aortic surgery to ensure the best correction of this complication.

QUESTION 1: WHAT IS THE DIFFERENTIAL DIAGNOSIS, AND WHICH INITIAL TEST WOULD YOU RECOMMEND TO RULE OUT A CARDIAC DISORDER AS THE ETIOLOGY OF HA?

ANSWER. This patient with a valve-sparing aortic root replacement (David procedure) and hemiarch replacement 6 months ago, had a symptomatic HA

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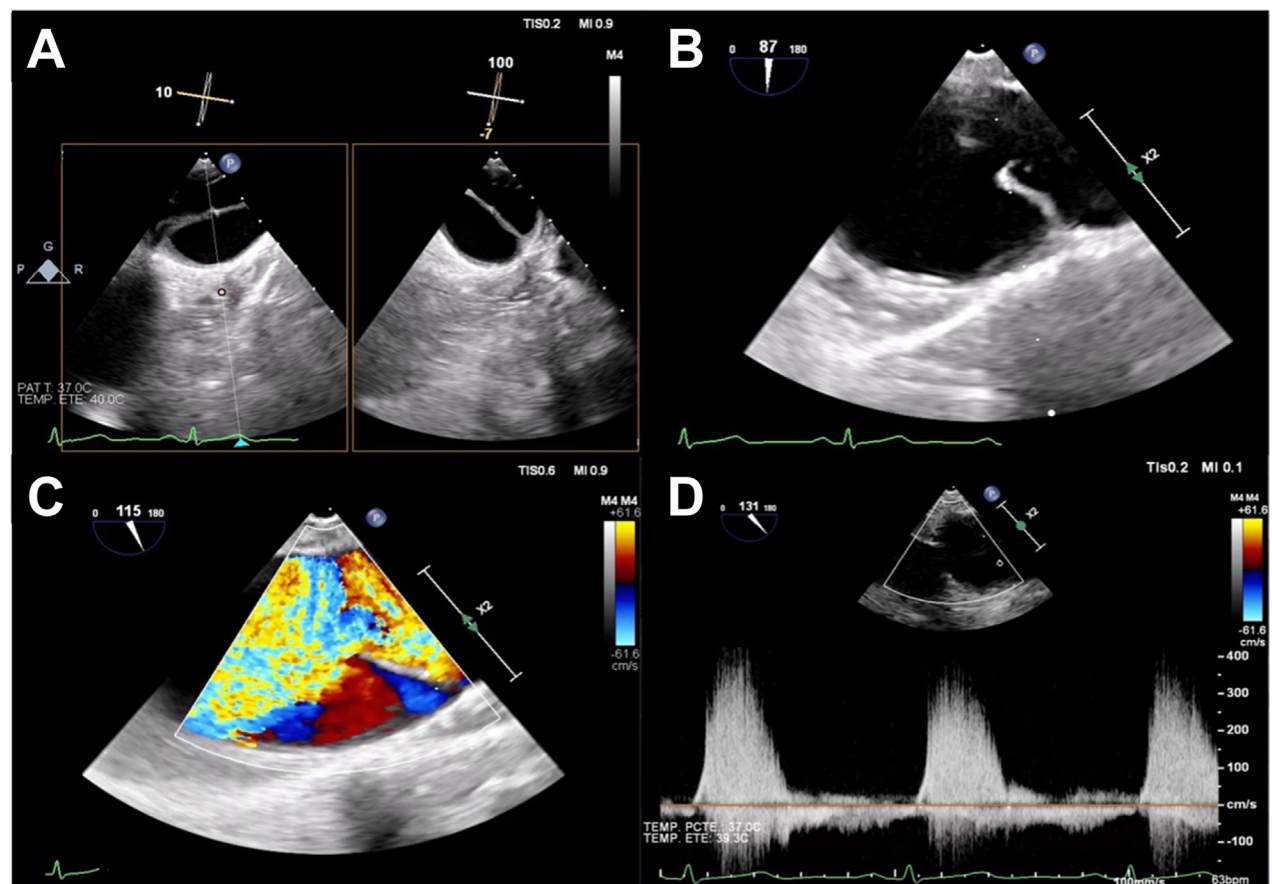
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**ABBREVIATIONS
AND ACRONYMS****AAAD** = Stanford type A acute aortic dissection**CTA** = computed tomography angiography**HA** = hemolytic anemia**TEVAR** = thoracic endovascular aortic repair

without angina or heart failure. The differential diagnosis included autoimmune HA, mechanical HA, and HA owing to hematological disorders. Therefore, our first thought would be to rule out the noncardiac etiologies of HA. The first step to rule out a cardiac disorder as the etiology of HA is a transthoracic echocardiogram. Transesophageal echocardiography may help to assess aortic graft complications, if there is suspicion of such complications as the etiology of HA after performing transthoracic echocardiogram.

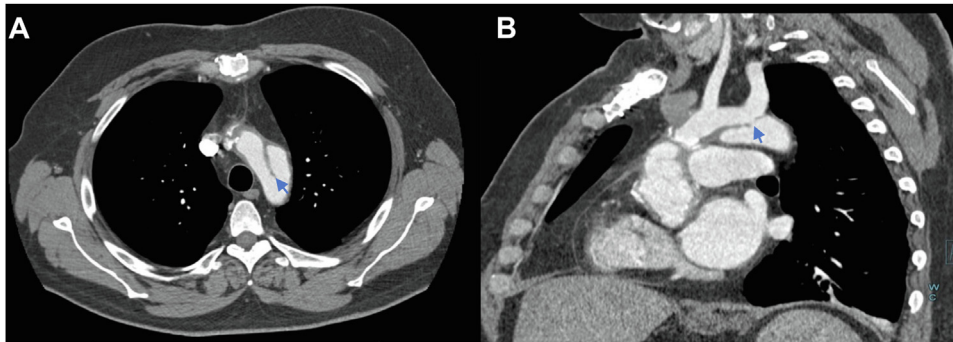
Pharmacological treatment was revised and noncardiac etiologies of HA were ruled out (including

a negative Coombs test and normal plasma ADAMTS13 antigen levels). The transthoracic echocardiogram showed an accelerated flow distal to the hemiarch graft in the suprasternal view. Biventricular size and function were preserved. A transesophageal echocardiogram was performed. It displayed a residual aortic dissection in the proximal descending aorta (**Figure 1A, Video 1**) and an intimal flap with a patent communication between the true and false lumen (**Figure 1B, Video 1**). In addition, an accelerated flow at that point was detected (**Figures 1C and 1D, Video 1**) with a pathological flow velocity (**Figure 1D**). Moderate regurgitation of the native aortic was detected (**Video 1**). No intracardiac shunts or other accelerated flows were detected.

FIGURE 1 Transesophageal Echocardiogram

(A) Transesophageal echocardiogram from mid-esophageal biplane view showing a residual aortic dissection in the proximal descending aorta. **(B)** Transesophageal echocardiogram from upper esophageal aortic view at 87° demonstrating an intimal flap distal to hemiarch graft. **(C)** Transesophageal echocardiogram from upper esophageal aortic view at 115° showing an accelerated flow through the communication between true and false lumen. **(D)** Pulse wave Doppler exhibiting an increased velocity of aortic flow at this point.

FIGURE 2 Computed Tomography Angiography of the Thoracic Aorta



(A) Axial view of computed tomography angiography showing a residual intimal flap (blue arrow) in the aortic arch. (B) Sagittal view of computed tomography angiography exhibiting a residual intimal flap (blue arrow) in the aortic arch that was extended to the origin of the left subclavian artery.

QUESTION 2: WHAT ARE THE MOST COMMON CAUSES OF HA AFTER AORTIC SURGERY AND WHICH ARE THE BEST TESTS TO EXCLUDE THEM?

ANSWER. An accelerated flow provoked by an inverted Teflon felt strip¹ or a kinked prosthetic graft are the main causes of HA after aortic surgery.² Computed tomography angiography (CTA) is helpful in identifying these complications. Additionally, more advanced imaging techniques such as 4-dimensional flow magnetic resonance imaging have demonstrated a highly accelerated vortex flow in a kinked graft as the cause of hemolysis.^{2,3} To prevent these complications, the internal Teflon felt strip should be as narrow as possible, with its most proximal portion sutured to prevent it from curling.¹ Moreover, the length of the graft must be measured exactly and if, despite this precaution, severe kinking occurs, the anastomosis should be repeated. In cases with moderate kinking, symmetrical U-shaped Prolene stitches applied on both sides, have been described to decrease the graft length.⁴

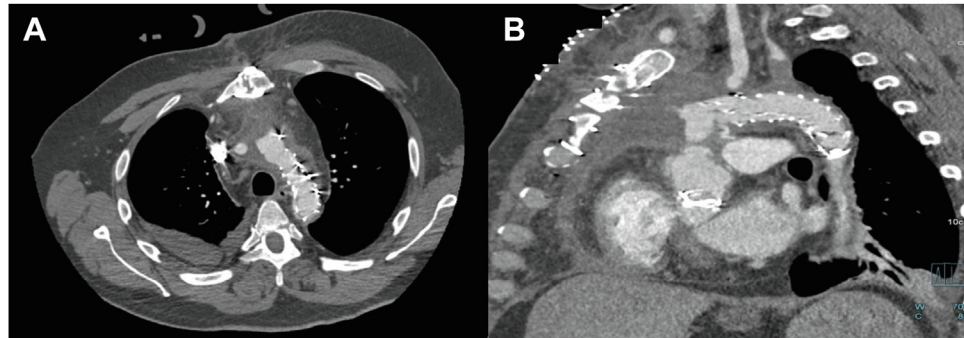
A CTA of the thoracic aorta was carried out. CTA confirmed the presence of a residual aortic dissection in the proximal descending aorta. An intimal flap was detected adjacent to the left subclavian artery (Figures 2A and 2B). No other signs of new aortic dissection in other aortic segments were detected. There were no signs of graft stenosis/kinking either in transesophageal echocardiography or CTA.

QUESTION 3: WITH PERSISTENT SYMPTOMATIC ANEMIA, WHAT IS THE NEXT APPROPRIATE MANAGEMENT STRATEGY?

ANSWER. Malperfusion syndrome, refractory pain, and uncontrollable hypertension are some of the potential post-AAAD surgery long-term complications associated with residual chronic aortic dissection. HA after aortic dissection repair owing to a residual intimal flap is a rare situation.⁵ Known causes of hemolysis after cardiac surgery include perivalvular leakage after prosthetic valve replacement, structural deterioration of prosthesis, severe patient-prosthesis mismatch, and endoleaks. However, only a handful of cases of hemolysis related to aortic surgery have been reported in literature.¹⁻³

The accelerated flow created between the true and false lumen at the residual intimal flap point was presumed to be the etiology of the HA. A multidisciplinary meeting was warranted for medical decision-making of the management. The heart team meeting included cardiology (general and interventional), cardiothoracic surgery, vascular surgery, cardiac anesthesia, nursing, and medical ethics. Eventually, a surgical approach was chosen.

An aortic arch replacement procedure was performed using a frozen elephant trunk procedure, carrying out a resection of the intimal flap, which was believed to be the cause of flow acceleration. Additionally, the aortic valve was replaced owing to moderate central aortic regurgitation. There were no relevant complications in the postoperative period

FIGURE 3 Computed Tomography Angiography of the Thoracic Aorta After Frozen Elephant Trunk Procedure

(A) Axial view of computed tomography angiography after frozen elephant trunk procedure. (B) Sagittal view of computed tomography angiography displaying the exclusion of the false lumen and intimal flap after frozen elephant trunk procedure.

and the patient was discharged. The patient remained asymptomatic in the next visits to the clinic and the blood tests did not show anemia. A CTA of the thoracic aorta was carried out 3 months after the surgical intervention (Figures 3A and 3B).

QUESTION 4: WOULD IT BE ADVISABLE TO IMPLEMENT A TEAR-FOCUSED STRATEGY THAT INCLUDES EXCLUSION OR RESECTION OF AN ENTRY TEAR IN THE ASCENDING AORTA AND ARCH DURING AORTIC REPAIR PROCEDURES FOR PATIENTS WITH ACUTE TYPE A AORTIC DISSECTION?

ANSWER. A tear-oriented approach, entailing exclusion or resection of the entry tear in the ascending aorta and arch, is recommended in patients undergoing aortic repair to mitigate dissection-related complications. This recommendation is supported by studies published in the medical literature.^{6,7} If entry tears are found in the distal arch or proximal descending aorta, implantation of a frozen elephant trunk may be considered as the treatment of choice, as suggested by current guidelines.⁸

In our case, the entry tear of the previous AAAD was located in the ascending aorta with extension to the proximal arch. As a result, the surgical team did not perform a frozen elephant trunk procedure, opting instead for a David procedure plus hemiarch replacement. During the surgery, the distal anastomosis was performed without cross clamp in the aortic arch, and there was no tear in the mid-distal arch. Our hypothesis is that the distal anastomosis, owing to the inflammatory tissue of the acute aortic

dissection, provoked an intimal tear, which subsequently led to the observed gradients and hemolysis in this particular case.

QUESTION 5: WOULD A PERCUTANEOUS APPROACH EMPLOYING A THORACIC ENDOVASCULAR AORTIC REPAIR BE DEEMED A VIABLE ALTERNATIVE TO REOPERATION IN THIS PARTICULAR CASE?

ANSWER. Thoracic endovascular aortic repair (TEVAR) may be considered as an alternative to reoperation for entry tear coverage and optimization of true lumen perfusion.⁹ However, owing to the zone in which the flap was located in our case, the vascular surgeons at our center did not have an optimal landing zone for TEVAR. If we had decided to perform a TEVAR in this case, a surgical debranching would have been required. To date, surgery remains the gold standard for aortic arch pathology, rather than TEVAR.

There is increasing evidence that a significant correlation exists between the volume of cases performed and the outcomes in aortic medicine.⁹ Implementing strategic workflow processes and systems can significantly increase the volume of aortic cases for both elective and emergent situations. Hence, it is recommended to establish aortic centers as they represent the only effective way to comprehensively understand the natural course of the disease, offer a complete spectrum of treatment options within a single framework, and address potential complications.⁹

CONCLUSIONS

HA secondary to an accelerated flow at a residual intimal flap point is a rare complication. This case highlights the importance of conducting a meticulous imaging evaluation of the thoracic aorta and aortic valve in individuals with HA following aortic surgery. The implementation of a multidisciplinary approach in an aortic center, supported by workflow processes

and systems that facilitate prompt diagnosis, treatment, and appropriate surgical management, can effectively minimize morbidity and mortality in complex aortic procedures.

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REFERENCES

1. Sakaguchi M, Takano T. Hemolytic anemia caused by aortic flap and inversion of felt strip after ascending aorta replacement. *J Cardiothorac Surg*. 2016;11:117.
2. Takei Y, Shibasaki I, Suzuki K, et al. Hemolytic anemia caused by an excessively kinked prosthetic graft after total arch replacement detected by 4-dimensional flow magnetic resonance imaging: a case report. *Medicine*. 2022;101:e29617.
3. Uchida W, Tokuda Y, Takehara Y, Usui A. Mechanical haemolytic anaemia assessed with four-dimensional flow cardiac magnetic resonance. *Eur J Cardiothorac Surg*. 2019;56:813-814.
4. Nezcic D, Milicic M, Boricic M, Micovic S. Modification of an old technique to correct kinking of tubular graft interposed to reconstruct ascending aorta. *Heart Lung Circ*. 2021;30:e139-e141.
5. Tadros R, Tang G, Barnes H, et al. Optimal treatment of uncomplicated type B aortic dissection. *J Am Coll Cardiol*. 2019;74:1494-1504.
6. Inoue Y, Minatoya K, Oda T, et al. Surgical outcomes for acute type A aortic dissection with aggressive primary entry resection. *Eur J Cardiothorac Surg*. 2016;50:567-573.
7. Heo W, Song SW, Lee KH, et al. Residual arch tears and major adverse events after acute DeBakey type I aortic dissection repair. *Ann Thorac Surg*. 2018;106:1079-1086.
8. Inoue Y, Matsuda H, Matsuo J, et al. Efficacy of entry exclusion strategy for DeBakey type III retrograde Stanford type A acute aortic dissection. *Eur J Cardiothorac Surg*. 2021;59:481-489.
9. Czerny M, Schmidli J, Adler S, et al. Current options and recommendations for the treatment of thoracic aortic pathologies involving the aortic arch: an expert consensus document of the European Association for Cardio-Thoracic surgery (EACTS) and the European Society for Vascular Surgery (ESVS). *Eur J Cardiothorac Surg*. 2019;55:133-162.

KEY WORDS aorta, computed tomography, dissection, echocardiography, hemolytic anemia

APPENDIX For a supplemental video, please see the online version of this paper.

