Cardiology in the Young

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Vascular ring and sling repair – addressing the root of the problem

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Guest Editorial

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Chiu and colleagues present a case report of a 9 yea old with a vascular sling (left aortic arch, aberrant right subclavian artery) and Duchenne's muscular dystrophy with symptoms of dysphagia, who underwent a palliative management strategy consisting of a subclavian-carotid bypass and plug at the base of the subclavian leaving the retroesophageal course of the subclavian artery.[1] Unfortunately, post-intervention cross-sectional imaging was not included, such post-intervention cross-sectional imaging would have demonstrated the space-occupying lesion still in place behind the oesophagus.

An aberrant right subclavian artery is one of the most common vascular anomalies, occurring in 0.5% of the population,[2] and does not form a vascular ring but can cause oesophageal compression by a "sling" and the associated space-occupying effect of the aberrant vessel against the posterior wall of the oesophagus, especially when the thoracic inlet is anatomically diminutive. Most of these patients with left aortic arch and aberrant right subclavian do not experience symptoms secondary to this anomaly; however, a minority demonstrate significant dysphagia secondary to the retroesophageal course of the aberrant right subclavian artery, while more commonly patients may report a slight globus sensation without functional compromise. It is critical that these patients undergo a thorough evaluation to rule out any intrinsic oesophageal disease or other reasons for their symptoms.

For patients with dysphagia secondary to the anomalous vessel, the space-occupying aberrant artery must be removed, otherwise the sling effect will persist even when the vessel has been "defunctionalised". The removal of this aberrant artery is accomplished by transferring the subclavian to another artery, commonly either:

1. as a single-stage operation within the thorax with transfer of the subclavian, or
2. as a two-stage approach with transfer of the subclavian to the right common carotid via a small supraclavicular incision, and, at the second stage, removal of the residual retroesophageal stump of the aberrant vessel via a thoracotomy.

We advocate completely avoiding graft material such as Dacron or polytetrafluoroethylene, both of which have poor patency rates, especially in the very young. Such definitive surgical repair can be performed safely with minimal morbidity in experienced hands, both at our institution,[3] and elsewhere.[4,5] Our ongoing analysis of this population at Mayo Clinic includes 35 patients, with four (11%) of these 35 patients presenting with impending rupture or aortic dissection. Among those who underwent elective, staged revascularisation of the subclavian and stump take down (13 patients with a median age of 23 years):

- there were no deaths,
- the median length of stay was 6 days,
- two patients (15%) had chylothorax managed non operatively, and
- one required vocal cord injection for temporary neuropraxia of the recurrent laryngeal nerve.

All 13 patients had either complete resolution or improvement in their symptoms.

While the case report of Chiu and colleagues[1] describes a palliative technique that may resemble the surgical repair while avoiding the thoracotomy, it importantly does not remove the aberrant artery from behind the oesophagus. We have first-hand experience with several patients who underwent "hybrid" techniques for addressing vascular rings or slings without resolution of symptoms and then developed complications as well, leading to very complex, high risk open surgical repairs. Two such cases exemplify these issues:

- One patient was a 28-year-old man with a right aortic arch and aberrant left subclavian artery who underwent left carotid artery to subclavian artery bypass, plug of the aberrant vessel, and stent graft placement of the distal arch and descending aorta. Unfortunately, this procedure was complicated by disruption of the femoral artery and stenosis of the left carotid artery to subclavian artery bypass leading to arm ischaemia. His dysphagia had also worsened after these procedures. He then presented to our institution for revision of his left carotid artery to subclavian artery bypass, excision of the aberrant vessel, and reconstruction of the arch to proximal descending thoracic aorta with a Dacron graft.

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This procedure required hypothermic arrest with retrograde cerebral perfusion. The patient did quite well post-operatively, was extubated the afternoon of surgery, and stayed in the intensive care unit one day. He was discharged to home with no complications.

- A second patient was a 29-year-old woman with Marfan syndrome who had undergone numerous cardiac operations starting as a child with an aortic valve repair, followed by homograft aortic valve replacement, followed by pectus repair, followed by reoperative replacement of the aortic root and hemi-arch replacement. She had a left aortic arch with aberrant right subclavian artery with a Kommerell' s diverticulum that had dilated over time. At an outside institution, she underwent an endovascular palliation because of concern about the number of previous interventions she had undergone. Unfortunately, they were unable to completely exclude the aberrant right subclavian artery, and the aneurysmal right subclavian artery continued to grow, and the stent in the aorta caused a dissection. These complications prompted referral to Mayo Clinic where she underwent staged open repair: first reoperative replacement of the aortic root and aortic arch replacement with an elephant trunk (the root had become too small for her), followed by reconstruction of her descending aorta. She did well after both operations.

We are also aware of other such patients who have undergone similar "minimally invasive" palliative and/or hybrid strategies without resolution of their symptoms. Therefore, we would warn other practitioners in the field to exercise caution regarding "minimally invasive" strategies such as these that do not address the root of the problem.

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