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Brief Report

Concomitant percutaneous treatment of aortic coarctation and associated intercostal aneurysms: pre-procedural recognition is key

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Abstract Intercostal aneurysms are associated with aortic coarctation. Their aetiology is not well-understood but may be related to intrinsic vascular pathology and altered flow dynamics through the intercostal artery. We present the cases of two patients with coarctation and intercostal aneurysms. The aneurysms were recognised on pre-catheterisation imaging studies and were selectively occluded during the same procedure to treat the coarctation. There were no complications; both the patients have no residual coarctation at the most recent follow-up. Intercostal aneurysms associated with coarctation can have significant consequences including late rupture, paralysis, and even death. These aneurysms are common with an incidence of up to 40% with adult-diagnosed coarctation; one treatment plan is to treat both the coarctation and aneurysm during a single catheterisation. Pre-catheterisation CT or MRI may play a role in this strategy.

Keywords: Coarctation of the aorta; collateral arteries; aneurysm; CHD

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Background

Aortic coarctation is a common congenital anomaly, with an incidence of 1 per 1200 in the general population and ~5–7% in patients with CHD.[1,2] Traditional teachings have suggested a bimodal distribution with infantile and adolescent/adult forms; however, more recent evidence suggests a continuum with more severe obstruction – regardless of location – presenting earlier in life.[1] Many patients who present later in life maintain systemic perfusion by developing arterial collaterals that bypass the narrowed segment of the aorta.[3] In these older patients, systemic hypertension is the most common finding leading to diagnosis, but patients also suffer from headaches, left ventricular hypertrophy on electrocardiogram, abnormal chest X-ray findings, and diminished femoral pulses with radial-femoral delay.[4] A few recent case series have discussed the association between intercostal aneurysms and aortic coarctation; although these aneurysms are less commonly discussed, they are a relatively common finding associated with coarctation diagnosed in adults.[5,6] In fact, up to 40% of adult-diagnosed patients have aneurysms, and the vast majority occur in the intercostal arteries.[7] We present the following cases to highlight this association and describe a combined therapeutic approach.

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Cases

We present the cases of two patients diagnosed with aortic coarctation associated with a large intercostal aneurysm. Neither patient had other significant medical issues and both were diagnosed within 2 months of catheterisation.

Patient no. 1 was a 48-year-old Russian nun. She was diagnosed with hypertension during her 2nd decade

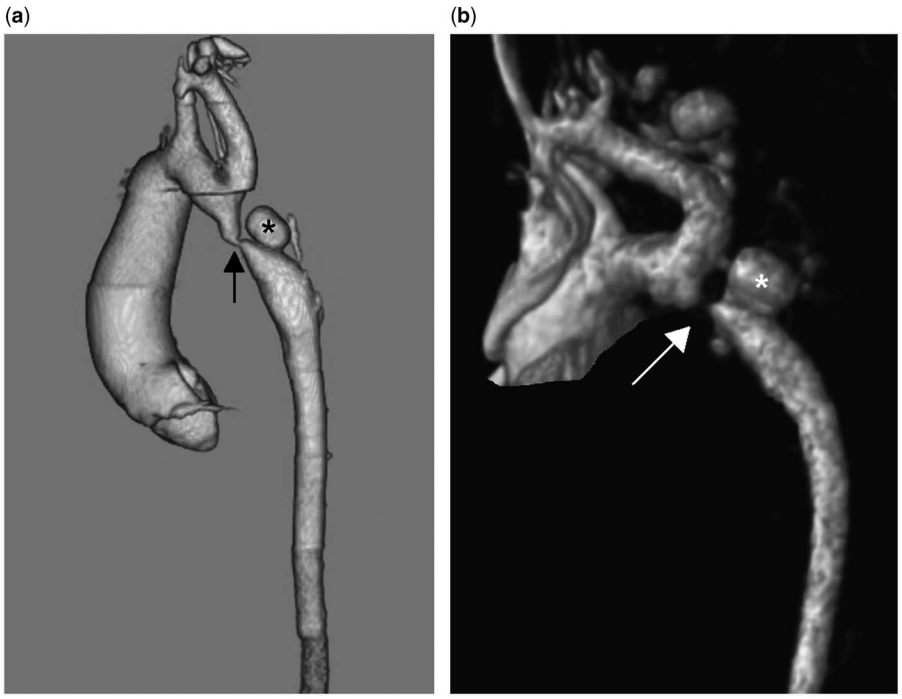

Figure 1.
*Pre-procedural 3D-re-constructed CT (**a**) and MRI (**b**) images of the reported patients. The * denotes the intercostal aneurysm and the arrow points to the coarctation.*

of life and had been medically managed with diuretics and β-blockers. She recently immigrated to the United States and was referred to a cardiologist. Evaluation by our adult congenital cardiology team noted non-palpable femoral pulses with upper extremity blood pressure between 110 and 125 mmHg, no gradient between her arms, and unobtainable pressures in her lower extremities. A contrast-enhanced chest MRI confirmed a discrete coarctation of the aorta (Fig 1a).

Patient no. 2 was a 14-year-old boy who presented with elevated blood pressure. He was asymptomatic and had not received regular medical evaluations. A new physician noted diminished femoral pulses and an upper–lower blood pressure gradient of ~50 mmHg. A contrast-enhanced cardiac CT confirmed aortic coarctation (Fig 1b). Both patients' scans also documented the large intercostal aneurysm (Fig 1). Initial haemodynamic and angiographic assessments in the catheterisation laboratory documented normal cardiac indices. Patient no. 1 had a discrete coarctation measuring 3 mm in minimal diameter, but also had relative hypoplasia of the distal arch and proximal descending aorta extending ~3 cm; the peak systolic gradient from ascending to descending aorta was ~50 mmHg. Patient no. 2 also had a discrete coarctation, minimal diameter of ~3 mm, with only mild and shorter segment hypoplasia of the distal arch, and a peak gradient of ~45 mmHg. The supreme intercostal artery was aneurysmal in both patients: the left in patient no. 1 and the right in patient no. 2 (Fig 2).

In patient no. 1, we placed two Amplatzer Vascular Plug 2^{TM} (St. Jude Medical, St. Paul, Minnesota, United States of America) devices into the feeding intercostal artery just cephalad to – that is, farthest from the aorta – the aneurysm. We then placed three covered stents in her aorta; two were required to treat the coarctation and one was used to cover the aortic ostium of the aneurysm. In patient no. 2, we placed one Amplatzer Vascular Plug-2 device into the intercostal feeding the aneurysm. We were able to treat both the coarctation and cover the aortic aspect of the aneurysm with a single covered stent (Fig 3). There was no gradient from ascending to descending aorta in either patient after intervention with no flow into the excluded aneurysms. Both the patients tolerated the procedure well without complications. Both were doing well, with no evidence of re-coarctation or other complications, at the most recent follow-up.

Discussion

Although recent case reports of arterial aneurysms associated with aortic coarctation are limited, this phenomenon is not rare. In her treatise on coarctation, Abbott describes the presence of numerous aneurysmal vessels associated with coarctation in adults.[8]

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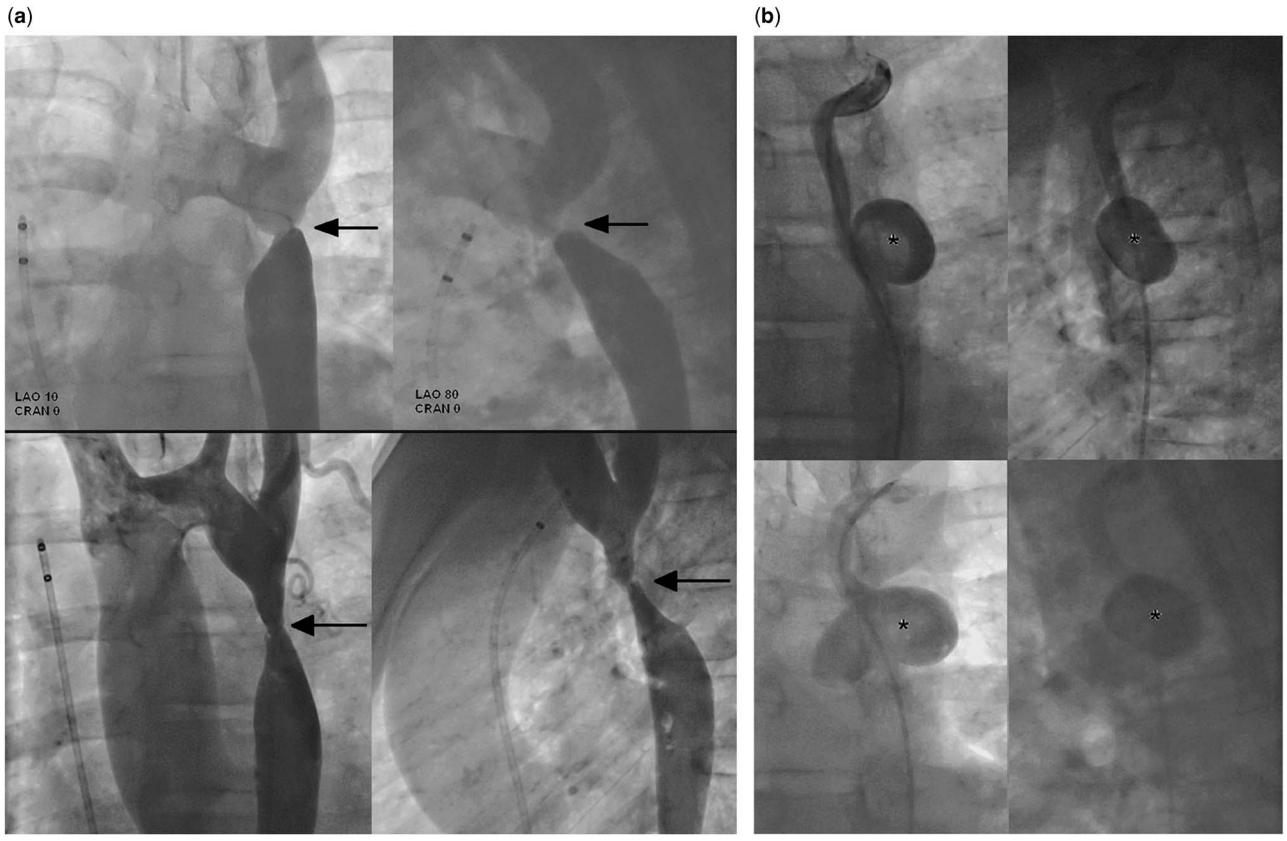

Figure 2.
*Descending aortic and selective intercostal angiograms demonstrating the coarctation (**a**) and intercostal aneurysm (**b**) in both patients.*

She refers to what is likely the first case of a patient with coarctation who died from a ruptured intercostal aneurysm, first published in 1858 by Leudet.[8] A later review by Braimbridge documented the presence of intercostal aneurysms in ~40% of adult-diagnosed, unrepaired coarctation patients.[7] He noted that the incidence of aneurysms increased with patient age. He also found that intercostal artery aneurysms were most common followed by aneurysms of the descending aorta distal to the coarctation; aneurysms of the ascending aorta or brachiocephalic vessels were exceedingly rare.[7]

The precise mechanisms leading to aneurysm formation remain unclear. The following two factors are likely implicated: altered vessel histology and flow dynamics. The abnormal histology has been noted since at least Dr Abbott's time, who described many vessels in coarctation patients as "thin-walled and atheromatous".[9] Subsequent authors have labelled this phenomenon as "cystic medial necrosis", but more recent evidence demonstrated that the underlying arterial histopathology is characterised by depletion and disarray of elastin fibres in the vessel wall.[10,11] Aneurysm formation is possibly related, at least in part, to this loss of structural integrity.

Another potential factor relates to the altered flow through the intercostal artery, which acts as a collateral. Bramwell documented this reversal of flow in a post-mortem study, elegantly delineating the numerous vessels, which potentially act as collaterals[12] (Table 1). A large amount of published data associates flow and arterial branching angles to the development of intracranial aneurysms.[13–16] Given the reversed flow associated with coarctation, abnormal shear forces may develop along the arterial walls – especially at branching points – that promote the formation of aneurysms through processes previously described.[17–19] More data are needed to truly understand the factors leading to aneurysm formation in patients with coarctation.

Aneurysms have numerous, potentially significant deleterious effects. The most obvious is the risk of rupture; the patient described by Leudet was a 37-year-old woman in whom the intercostal eroded into her left main bronchus leading to severe haemoptysis and death.[9] Rupture has also been reported after treatment for coarctation, many years after surgery in one case.[20,21] In addition to rupture, aneurysms may calcify and enlarge.[22] Moreover, cases of paralysis have been reported secondary to compression of the anterior spinal artery by the pulsatile aneurysm.[23]

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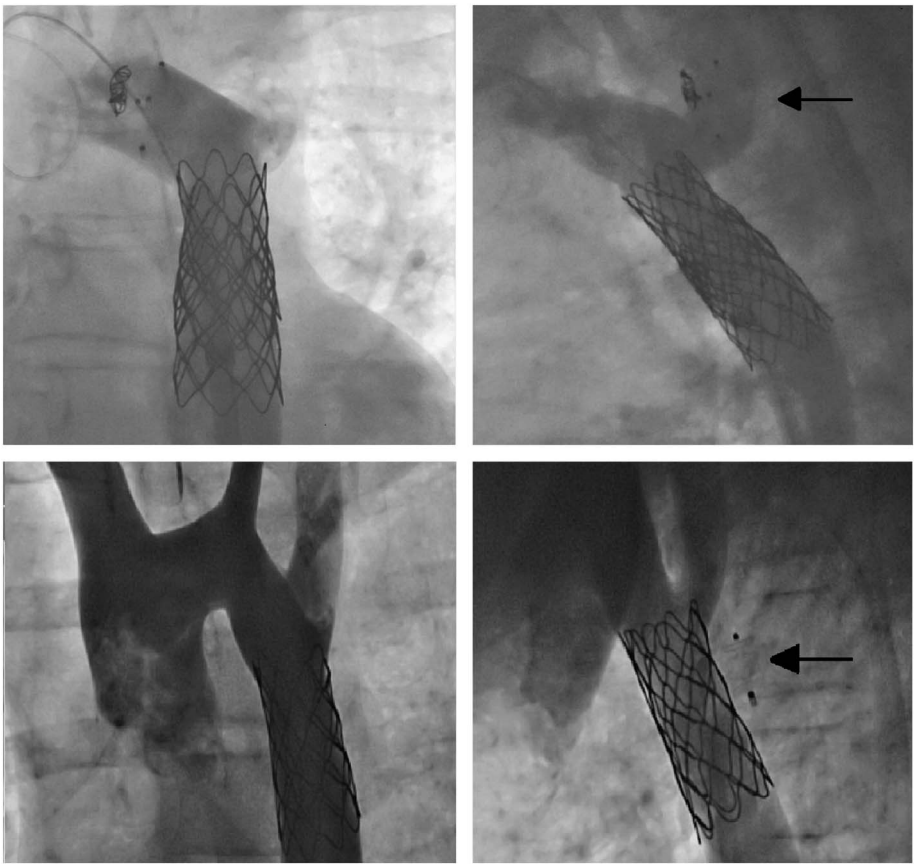

Figure 3.
Descending aortic angiograms after device placement and deployment of covered stents in both patients. Note that the aneurysms do not fill either retrograde or antegrade.

Table 1. Collateral channels in coarctation of the aorta.

1. The scapular and cervical anastomoses
   The following arteries form a network around the scapula and in the cervical region:
   (a) Supra-scapular and transversalis colli (from thyroid axis).
   (b) Posterior scapular and superficial cervical (from transversalis colli).
   (c) Long thoracic and sub-scapular with its dorsal is scapulas branch (from axillary artery).
   From this network, descending branches anastomose with the lateral and dorsal branches of the aortic intercostals.

2. The internal mammary anastomoses:
   (a) Superior epigastric → deep epigastric branch of external iliac.
   (b) Musculo-phrente → phrenic branches of the thoracic and abdominal aorta.
   (c) Mediastinal branches → mediastinal branches of the aorta.
   (d) Anterior intercostals → terminal branches of aortic intercostals.

3. The intercostal anastomoses:
   (a) The terminal branches → the intercostal branches of the internal mammary.
   (b) The lateral branches → the sub-scapular and long thoracic.
   (c) The dorsal branches → the posterior scapular.
   (d) The first and second intercostals (arising from the subclavian by the superiorintercostal) → the upper aortic intercostals.
   (e) Each intercostal → those above and below.

4. The spinal anastomoses:
   The vertebral artery, arising from the first part of the subclavian, re-inforces the spinal arteries in which the blood flows downwards to reach the spinal branches of the aortic intercostals. These pass through each intervertebral foramen. There are also branches from the inferior thyroid that pass through the intervertebral foramina in the neck to join the spinal arteries.

Reproduced with permission from Bramwell and Jones[12]

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We decided to exclude the aneurysm from both sides, given the known abnormal histological profile of systemic arteries in patients with coarctation as well as the reported complications even after treatment of the coarctation. We postulate that, in some patients, covering only the aortic end of the aneurysm may allow further aneurysm growth – with potential complication – by the feeding vessel. Others have demonstrated that covered stents can be effective in treating coarctation and aortic aneurysm.[24,25] In addition, the technique of excluding an aneurysm on both sides – with either a coil or a device – has also been reported in a patient after aneurysm rupture.[26,27] We combined both strategies in our approach.

In addition to the medical consequences, unrecognised aneurysms have the potential to mislead operators in the catheterisation laboratory. Unrecognised aneurysms may fill slowly after coarctation stent placement. The slow, incomplete filling of large aneurysms may mimic flow into the mediastinum from a transmural aortic tear, potentially causing concern for post-intervention aortic rupture. This may lead to placement of additional stents to exclude the aortic aspect of the aneurysm where one well-placed stent may have dealt with both lesions if recognised earlier. Furthermore, excluding the vessel feeding the aneurysm is difficult – if not impossible – after placement of an aortic stent.

The potential for aneurysm growth may persist if the feeder is not occluded. Therefore, pre-procedural recognition of the aneurysms is critical. Recent studies have shown that MRI and CT have the capability of demonstrating intercostal and other aneurysms.[28,29] This evidence, coupled with the increasing availability of MRI and CT, suggests that these modalities may be indicated as part of the pre-procedural work-up of adult-diagnosed coarctation.

Conclusions

Intercostal aneurysms are a common association with adult-diagnosed coarctation of the aorta. The consequences of these aneurysms are varied and may be significant, including massive haemorrhage and death. We lack a complete understanding of the factors that lead to the development of these aneurysms, but the abnormal histopathological features of the vasculature and reversed flow in the collateral arteries may play a role. Therefore, one strategy to address these lesions is concomitant treatment of the coarctation with complete exclusion of the aneurysm, thereby reducing the potential for ongoing aneurysm growth with complications.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant US national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008. The Children's Hospital of Philadelphia Institutional Review Board's approval was sought, but was not required given that this report involved retrospective results from only two patients.

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