

**CASE REPORT**

# Translunar access for coronary intervention in a 23-year-old with accelerated coronary atherosclerosis due to congenital aortic hypoplasia

Jonathan D. Dawkins<sup>1</sup> | Ari M. Cedars<sup>1,2</sup> | Jeffrey M. Schussler<sup>1,2,3</sup>  <sup>1</sup>Texas A&M College of Medicine, Bryan, Texas<sup>2</sup>Division of Cardiology, Department of Internal Medicine, Baylor University Medical Center, Dallas, Texas<sup>3</sup>The Baylor Scott & White Heart and Vascular Hospital, Dallas, Texas**Correspondence**

Jeffrey M. Schussler, The Baylor Scott &amp; White Heart and Vascular Hospital, Dallas, TX.

Email: Jeffrey.Schussler@Baylorhealth.edu

**Key Clinical Message**

The use of alternative arterial access and advanced imaging is not only applicable to mainstream adult cardiology, but helpful and sometimes necessary in taking care of adult patients with complex congenital heart disease.

**KEYWORDS**

aortic hypoplasia, atherosclerosis, congenital, translunar, transradial

## 1 | INTRODUCTION

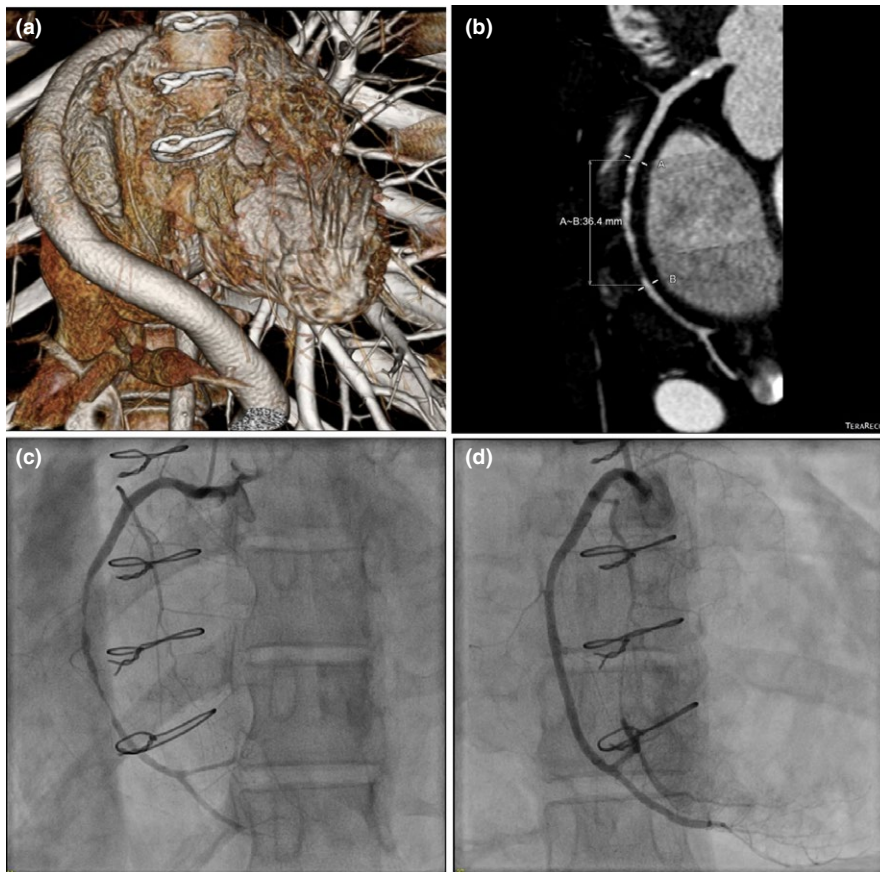
A 23-year-old woman with a history of diffuse aortic hypoplasia, right-sided branch pulmonary artery stenosis, and long-standing hypertension presented with accelerated atherosclerosis and required percutaneous coronary intervention. Due to her size and access limitations, this was performed through a 5Fr, right ulnar approach. This case illustrates several features and techniques which are germane to the care of adults with congenital cardiac disease. It also highlights the use of coronary computed tomography and translunar access which allowed for the diagnosis and treatment of this unusual case.

Congenital heart disease (CHD) is becoming increasingly more prevalent among adults in the Western world.<sup>1</sup> The rate of survival to adulthood among children born with CHD has steadily increased with advances in diagnostic and surgical techniques. By 2010, more than 90% of CHD patients were expected to survive to adulthood, and two-thirds of the living CHD population were adults.<sup>1</sup> As prevalence increases, more adult cardiologists will begin to see adults with congenital heart disease (ACHD) in their practices. These patients present frequently with unique and complicated problems and require specialized care.

## 2 | CASE REPORT

A 23-year-old woman presented to her local (general) cardiologist with complaints of progressive dyspnea with exertion and chest pain beginning within the past 8-12 months. The patient had a history of complex congenital heart disease including diffuse aortic hypoplasia with an interposition graft between her ascending thoracic aorta and her abdominal aorta at the level of her renal arteries (Figure 1—panel A), right branch pulmonary artery stenosis with right lung hypoplasia, mild pulmonary valvular stenosis, and a patent ductus arteriosus which had been ligated at the time of her aortic surgery. She was followed regularly by a pediatric cardiologist until the age of 18, but then had been lost to follow-up until the time of this presentation.

On physical examination, the cardiologist heard a carotid bruit, and given her history, he referred the patient for a computed tomographic angiogram of the pelvis, abdomen, thorax head, and neck. This revealed evidence of the above-described abnormalities as well as diffuse vascular disease including occlusion of the celiac axis, 50% stenosis of the left subclavian artery, and near occlusion of the left internal carotid artery with



**FIGURE 1** A 23-year-old patient with congenital aortic hypoplasia: surgical aortic bypass demonstrated by computed tomography (panel A). A high-grade stenosis is seen of the right coronary artery (RCA) by computed tomographic coronary angiography (panel B) as well as invasive coronary angiography (panel C). Invasive coronary angiography of the RCA post stent placement (panel D)

a suspected 6–7 cm bilobed aneurysm of the basilar artery. As a result, the patient was referred for cerebral angiography with possible coil occlusion of the aneurysm.

This procedure was approached via the femoral artery and resulted in a large retroperitoneal hematoma and hemorrhagic shock requiring an ICU stay. Her course was complicated by respiratory failure due to pulmonary edema, acute liver injury, acute kidney injury, and acute pancreatitis. During this hospitalization, the patient complained of chest pain and ECG showed dynamic inferior ST and T wave changes. A troponin was checked and was found to be elevated with a peak of 76 ng/mL. Given the patient's tenuous clinical status, and known vascular access difficulties, cardiac catheterization was deferred. A subsequent transthoracic echo was found to show *massive left ventricular hypertrophy* without focal wall motion abnormalities. A conservative approach was taken and only medical therapy was used. The patient was placed on aspirin at the time of discharge but refused beta blockade. Further plans were made to assess any ischemic area at risk with future stress testing.

After she was stabilized, an ischemic workup was ordered, and a nuclear stress test indicated inferior ischemia. Given her diffuse vascular disease, and concern that the ischemia might be due to diffuse vascular narrowing not amenable to percutaneous coronary intervention, CT coronary angiography (CTCA) was performed. The CTCA showed mid-right coronary stenosis (Figure 1—panel B).

With her history of complications from the femoral approach, and small caliber distal aorta, she was set up for a transradial catheterization. Due to the caliber of the radial artery, this was not feasible, and she was catheterized via a transulnar approach (a known and accepted alternative). Coronary angiography demonstrated hyperdynamic left ventricular function with a normal left main, LAD, and circumflex arteries. The right coronary had a >95% stenosis in the mid-portion of the artery (Figure 1—Panel C). Using a 5Fr, guiding catheter (RBU, Medtronic guide sheath, Fridley, MN, USA, 5 Fr, 10 cm) with a 2 mm balloon (Apex Balloon, Boston Scientific Corporation, Marlborough, MA, USA, 2 × 20 mm) and stented with a 2.25 × 38 mm SYNERGY (Synergy DES Stent, Boston Scientific Corporation, 2.5 × 38 mm) stent, postdilated at 18 atmospheres with a 2.5 mm balloon (NC Quantum Apex Balloon, Boston Scientific Corporation, 2.5 × 20 mm; Figure 1—Panel D). She was placed on a 12-month dual antiplatelet therapy of clopidogrel 75 mg and aspirin 81 mg daily, and has done well in follow-up.

### 3 | DISCUSSION

Coronary artery disease (CAD) is rare in young women, but there are some unusual structural or genetic causes that may accelerate this process. Structural causes are less common, but tend to offer a more complicated prognosis.<sup>2</sup> Congenital

aortic hypoplasia is a rare disorder characterized by diffuse tubular narrowing of the entire aorta and its branch vessels. Given the rarity of the condition, there are little available data to guide prognostic counseling or management. Nevertheless, from the standpoint of risk factors for accelerated atherosclerosis, the condition might be considered etiologically similar to traditional focal coarctation of the aorta.

Typical coarctation of the aorta is characterized by a discrete narrowing in the aorta at the level of the aortic isthmus, typically recognized as the region between the left subclavian artery and the ligamentum arteriosum. While many more mild cases of coarctation are asymptomatic, they have long-term insidious side-effects.<sup>3</sup> Relative hypoperfusion of the renal parenchyma leads to the activation of the renin-angiotensin-aldosterone system which can result in severe systemic hypertension.<sup>4</sup> In addition, as a result of chronic exposure to elevated afterload, patients with this condition are predisposed to accelerated coronary and cerebral atherosclerosis, as well as left ventricular hypertrophy (LVH) similar to that seen in the present patient.<sup>5,6</sup>

## 4 | CONCLUSION

Cardiovascular sequelae of and treatment for patients with distal aortic hypoplasia has not been extensively documented in the adult cardiovascular literature. This case demonstrates possible complications and highlights the importance of a collaborative approach between ACHD specialists, interventional cardiologists, and the use of noninvasive cardiovascular imaging as well as modern vascular access techniques in order to individualize and optimize a treatment strategy for a patient with a unique subset of clinical issues.

## CONFLICT OF INTEREST

None declared.

## AUTHOR CONTRIBUTION

JMS: involved in secondary and final edits; JDD: involved in initial and secondary edits; AMC: involved in initial edits.

## ORCID

Jeffrey M. Schussler  <https://orcid.org/0000-0002-3446-2129>

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