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CASE REPORT

ADVANCED

CLINICAL CASE SERIES

Symptomatic Myocardial Bridging in D-Transposition of the Great Arteries Post-Arterial Switch

Sumeet S. Vaikunth, MD, MED,[a,b] Daniel J. Murphy, MD,[c] Jennifer A. Tremmel, MD, MS,[d] Ingela Schnittger, MD,[d] Robert Scott Mitchell, MD,[e] Katsuhide Maeda, MD, PHD,[f] Ian S. Rogers, MD, MPH[c,d]

ABSTRACT

We present Stanford's experience with patients post-arterial switch operation presenting with chest pain found to have hemodynamically significant myocardial bridging. The evaluation of symptomatic patients post-arterial switch should not only include assessment for coronary ostial patency but also for nonobstructive coronary conditions such as myocardial bridging. **(Level of Difficulty: Advanced.)** (J Am Coll Cardiol Case Rep 2023;8:101730)

Coronary ostial stenoses are frequently invoked as causes of chest pain in dextro-transposition of the great arteries (d-TGA) post-arterial switch operation (ASO).[1] However, many times evaluation reveals no stenoses. Myocardial bridging occurs when there is an intramyocardial segment of an epicardial coronary artery. Although myocardial bridges may be present in approximately 20% to 25% of the general population as benign anatomic variants, there have been reports of clear association with myocardial ischemia, malignant arrhythmias, and sudden cardiac death in patients with and without congenital heart disease. We present Stanford's experience with the diagnosis and management of symptomatic adult arterial switch patients found to have hemodynamically significant myocardial bridges of the left anterior descending artery (LAD).

LEARNING OBJECTIVES

- To evaluate patients' post-arterial switch operation for late coronary insufficiency per American College of Cardiology guidelines.
- To identify nonobstructive coronary artery disease such as myocardial bridging when patients presenting with symptoms have a negative workup for obstructive coronary lesions.
- To recognize that surgical unroofing of the myocardial bridge can relieve symptoms in this population.

CASE SERIES

PATIENT 1. An 18-year-old woman presented with intermittent chest pain for 5 years. A prior exercise

From the [a]Department of Medicine, Division of Cardiovascular Medicine, University of Pennsylvania Perelman School of Medicine, Philadelphia, Pennsylvania, USA; [b]Division of Cardiology, Children's Hospital of Philadelphia, University of Pennsylvania Perelman School of Medicine, Philadelphia, Pennsylvania, USA; [c]Department of Pediatrics, Division of Pediatric Cardiology, Stanford University School of Medicine, Palo Alto, California, USA; [d]Department of Medicine, Division of Cardiovascular Medicine, Stanford University School of Medicine, Palo Alto, California, USA; [e]Department of Cardiothoracic Surgery, Stanford University School of Medicine, Palo Alto, California, USA; and the [f]Division of Cardiothoracic Surgery, Children's Hospital of Philadelphia, University of Pennsylvania Perelman School of Medicine, Philadelphia, Pennsylvania, USA.
The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the Author Center.

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ABBREVIATIONS AND ACRONYMS

ASO = arterial switch operation

CCTA = coronary computed tomography angiography

dFFR = diastolic fractional flow reserve

d-TGA = dextro-transposition of the great arteries

FFR = fractional flow reserve

IVUS = intravascular ultrasound

LAD = left anterior descending artery

SPECT = single-photon emission computed tomography

test demonstrated normal exercise tolerance. Coronary angiogram and cardiac magnetic resonance showed no evidence of obstructive disease or delayed enhancement, respectively. Coronary computed tomography angiography (CCTA) identified an LAD myocardial bridge; repeat cardiac catheterization with intravascular ultrasound (IVUS) and intracoronary hemodynamic evaluation established hemodynamic significance. Her chest pain resolved after myocardial bridge unroofing and she remains asymptomatic.

PATIENT 2. A 25-year-old woman presented with chest pain for 3 years. Nuclear stress testing, CCTA, and coronary angiogram were negative for significant coronary disease. Over the next months, the pain began to occur daily at rest and with exertion. Cardiac catheterization with IVUS and intracoronary hemodynamic evaluation diagnosed an LAD myocardial bridge (identified on retrospective review of CCTA), and she underwent unroofing. She did well postoperatively but began having palpitations and underwent ablation for atrioventricular nodal re-entrant tachycardia. She recently underwent an uncomplicated pregnancy.

PATIENT 3. A 23-year-old woman presented with chest pain and shortness of breath for 6 years. A stress echocardiogram was significant for septal buckling (Videos 1 and 2), and CCTA showed a myocardial bridge (**Figure 1**). Catheterization with IVUS and intracoronary hemodynamic evaluation confirmed a hemodynamically significant LAD myocardial bridge (**Figure 2**). After an unsuccessful trial of nebivolol, she underwent unroofing and continues to teach dance without limitation.

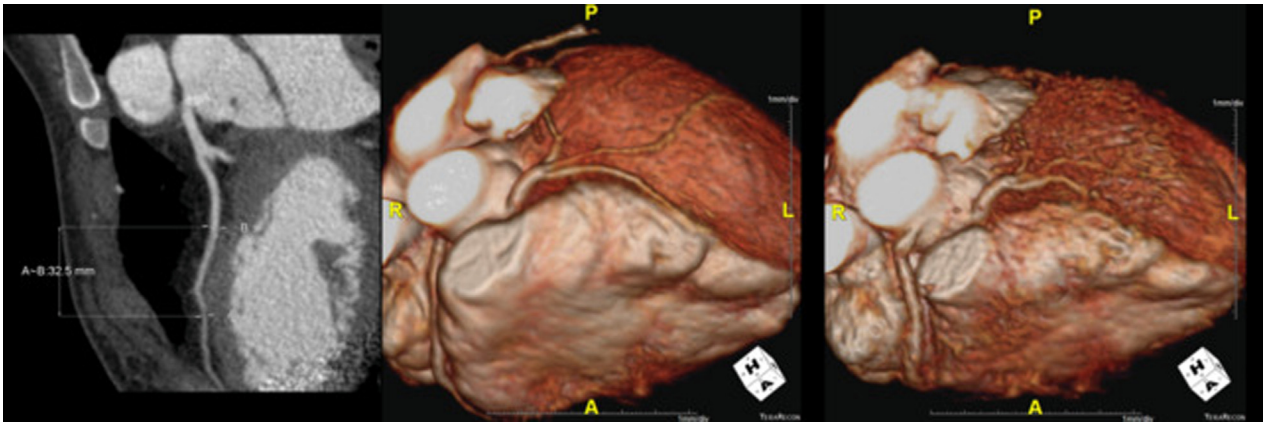
PATIENT 4. A 26-year-old woman presented with daily episodes of chest pain, shortness of breath, and fatigue for 4 years. Cardiac catheterization revealed no evidence of ostial stenosis, but CCTA identified LAD myocardial bridging. Cardiac catheterization with IVUS and intracoronary hemodynamic evaluation confirmed hemodynamically significant myocardial bridging and moderate endothelial dysfunction After an unsuccessful trial of nebivolol and then amlodipine, her chest pain resolved following unroofing and she remains asymptomatic.

PATIENT 5. A 16-year-old male with pectus excavatum presented with syncopal episodes for 3 years. Rhythm monitoring exhibited nonsustained ventricular tachycardia with concern for right coronary artery compression because of its proximity to the sternum. CCTA did not demonstrate compression but revealed an LAD myocardial bridge. Catheterization with IVUS and intracoronary hemodynamic evaluation established hemodynamic significance of the bridged segment and a fully patent right coronary artery. Unroofing along with the Nuss procedure for pectus was performed; he remains asymptomatic.

DISCUSSION

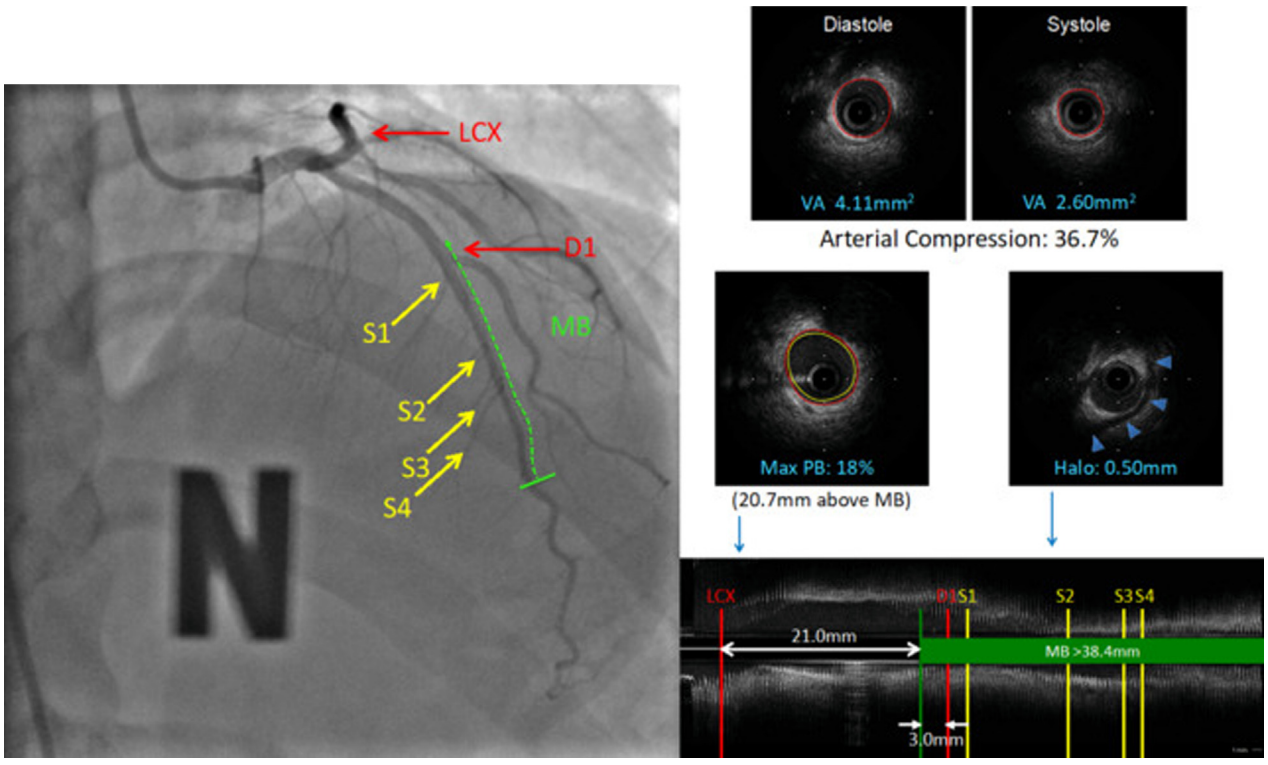
We present Stanford's experience with 5 symptomatic arterial switch patients found to have LAD myocardial bridges. See **Tables 1 and 2** for median and individual characteristics, respectively. All underwent surgical

FIGURE 1 Coronary Computed Tomography Angiography for Patient 3



Coronary computed tomography angiography shows segment of left anterior descending artery with myocardial bridging **(left and middle)** with compression during systole **(right)**.

FIGURE 2 **IVUS Map for Patient 3**



Coronary angiography of the left anterior descending artery with a myocardial bridge (MB) as assessed by intravascular ultrasound (IVUS) in patient 3. The MB is identified by the **green dotted line**. S1 to S4 are septal branches; D1 is a diagonal branch. The **image on the left**, an IVUS "map", represents anatomic data from IVUS superimposed upon a coronary angiogram. The percentage of systolic compression (36.7 %) is shown. The **blue arrows** outline the echolucent half-moon sign ("halo") indicative of an MB with maximum thickness of 0.50 mm. LCX = left circumflex artery; Max PB = maximum plaque burden; VA = vessel area.

unroofing with symptom improvement. The incidence of myocardial bridging in patients with transposition of the great arteries is unknown. Although a prior case of myocardial bridging in a patient with d-TGA post-atrial switch has been reported, ours is the first of significant myocardial bridging in patients post-arterial switch.[2]

The ASO involves switching the great arteries to re-establish the appropriate ventriculoarterial relationships. In so doing, the coronary arteries are harvested and transferred to the neo-aorta. This coronary transfer is the most challenging aspect of the operation; any kinking during this process can lead to coronary stenoses. The prevalence of coronary events is 7% in patients post-ASO, with 89% of these events occurring in the first 3 months postsurgery.[1] Single-photon emission computed tomography myocardial perfusion imaging is abnormal in 12% of ASO patients, whereas late coronary stenosis or occlusion is observed in 11%.[3] Thus, anatomical and physiologic screening to evaluate coronary patency and myocardial ischemia are Class IIa recommendations of the 2018 American College of Cardiology adult congenital heart disease guidelines.[4]

Myocardial bridging can be a benign variant, but in patients with refractory chest pain despite normal anatomical and physiologic testing, further investigation needs to be performed. We have devised a protocol to evaluate these patients to determine the significance of the myocardial bridge. The tunneled segment of the coronary artery, which lies under the bridge of the overlying myocardium, can be compressed during systole but also during diastole (especially in tachycardic states in which diastolic filling time decreases), leading to a decrease in flow and flow reserve. The dynamic nature of chest pain and ischemia from myocardial bridging is not reliably captured on single-photon emission computed tomography myocardial perfusion imaging because the ischemia occurs only in late systole to early diastole

TABLE 1 Median Values of Series of Patients

Age, y	23 (16-26)
Bridge length, mm	41.47 (31.9-64.0)
Halo thickness, mm	0.64 (0.50-0.99)
Systolic compression, %	27.8 (19.8-36.7)
Diastolic FFR	0.69 (0.53-0.70)
Length of stay, d	5 (3-6)
Follow-up since surgery, y	5.6 (0.8-8.1)

Values are median (range).
FFR = fractional flow reserve.

and affects a small distribution of the myocardium within the bridged segment.[5] Our center favors stress echocardiography as first line in diagnosis because we have found that patients with myocardial bridging have focal end-systolic to early-diastolic septal buckling with apical sparing on this imaging modality (90% sensitivity and 83% specificity) (Videos 1 and 2).[6] CCTA is ordered to evaluate coronary arteries for potential obstructive disease but also allows for the visualization of myocardial bridges. Invasive assessment with IVUS also can be used to evaluate the bridge length as well as arterial compression, thickness of the myocardial bridge (halo), and plaque burden. This IVUS "map" can be used for surgical planning, and diastolic fractional flow reserve (dFFR) is calculated at rest and with dobutamine stress. A dFFR <0.76 is significant.[7] dFFR differs from the typical mean fractional flow reserve (FFR) because myocardial bridges cause diastolic pressure gradients but normal or negative systolic pressure gradients; therefore, the mean FFR may underestimate the hemodynamic significance. Furthermore, the length and depth of bridging on computed tomography correlate with IVUS measurements, and a myocardial bridge muscle mass index, the product of length and depth of the bridge, of 31 or greater correlates with abnormal dFFR.[7,8] Furthermore, dobutamine stress is necessary given the dynamic nature of ischemia from bridges. Finally, we have observed that at least 50% of adult patients symptomatic enough to undergo invasive assessment also demonstrate coronary endothelial dysfunction (ie, epicardial coronary spasm) with intracoronary acetylcholine administration, which might serve as a potential source of chest pain even after successful myocardial bridge unroofing.[8]

All patients in this series had a "usual" ostial coronary pattern with no other coronary anomalies and demonstrated reduced dFFR and underwent surgical unroofing. The technique for surgical unroofing has been outlined in detail in prior works.[9,10] In this specific series, because of previous surgeries, scar formation and adhesions on the heart surface are usually observed. It is important to dissect the entire coronary artery for good visualization. Then, complete unroofing should be performed by cutting the myocardium on top and both sides of the coronary artery with a beaver knife and/or scissors. Two patients initially opted for medical management for 5 months and 1 year, respectively, which did not sufficiently reduce symptoms. We preferentially use a particular beta blocker (ie, nebivolol), which stimulates beta-3 receptors, leading to the release of nitric oxide, as first line for medical management. Calcium-channel blocker therapy, such as with diltiazem, can also be helpful. Percutaneous coronary intervention with stent placement is associated with stent fractures and target lesion revascularization after in-stent

TABLE 2 Individual Characteristics

	Age/Sex	Surgical History	Medical History	Symptom	Distance From LAD Origin (CT), mm	Length (IVUS), mm	Halo Thickness, mm	Systolic Compression, %	dFFR	LOS, d	Current Medications	Follow-Up, y
Patient 1 DOS: 3/9/13	18 y/Female	ASO and coarctation repair at 7 days	None	Chest pain	38	Not available	Not available	19.8	0.66	5	None	8.1
Patient 2 DOS: 1/17/14	25 y/Female	ASO at 6 days	AVNRT HTN Pre-eclampsia	Chest pain	5	32	0.56	27.8	0.59	6[a]	Nifedipine[b]	7.3
Patient 3 DOS: 8/21/15	23 y/Female	ASO at 3 days	None	Chest pain	27	38	0.5	36.7	0.70	5	None	5.6
Patient 4 DOS: 11/16/18	26 y/Female	R mBTT shunt/MPA band/VSD closure ASO at 2 months	Prior alcohol use	Chest pain	25	64	0.99	36.6	0.53	3	Labetalol[c]	2.4
Patient 5 DOS: 6/1/20	16 y/Male	ASO at 5 days	Pectus	Chest pain	34	45	0.72	23.5	0.69	4	Metoprolol, aspirin	0.8

[a]Pleural effusion requiring chest tube placement. [b]For blood pressure control because of recent pre-eclampsia. [c]For HTN and aortic dilation given the pregnancy risk with prior losartan.
ASO = arterial switch operation; AVNRT = atrioventricular nodal re-entrant tachycardia; CT = computed tomography; dFFR = diastolic fractional flow reserve; DOS = date of surgery; HTN = hypertension; IVUS = intravascular ultrasound; LAD = left anterior descending; LOS = length of stay; MPA = main pulmonary artery; R mBTT = right modified Blalock-Taussig-Thomas; VSD = ventricular septal defect.

restenosis.[11] Coronary artery bypass grafting can lead to graft occlusion given the risk of competitive flow because of the dynamic nature of myocardial ischemia.[5] Thus, we recommend against stent placement or coronary artery bypass grafting and instead prefer surgical unroofing of such symptomatic myocardial bridges.

Patients with myocardial bridging are at an increased risk for atherosclerosis, which occurs in the segment proximal to the bridge, as detected by IVUS.[8] Routine follow-up is necessary in this young patient cohort, not only for the common complications associated with the arterial switch itself such as aortic root dilatation, aortic regurgitation, and branch pulmonary stenosis but also for the development of acquired atherosclerotic disease, which is prevalent in the aging population of patients with congenital heart disease.[12]

CONCLUSIONS

Coronary ostial patency post-coronary reimplantation is frequently investigated in those with chest pain post-arterial switch. Other causes of chest pain including myocardial bridging and endothelial dysfunction should also be considered. This case series demonstrates LAD myocardial bridging in 5 symptomatic young adult arterial switch patients. Unroofing of the myocardial bridge led to symptom relief in all patients.

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ADDRESS FOR CORRESPONDENCE: Dr Sumeet S. Vaikunth, Perelman Center for Advanced Medicine, 11th Floor, South Pavilion, 3400 Civic Center Boulevard, Philadelphia, Pennsylvania 19104-5127, USA. E-mail: sumeet.vaikunth@pennmedicine.upenn.edu.

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KEY WORDS chest pain, coronary vessel anomaly, transposition of the great arteries

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