



Evolution of surgical repair of intraseptal anomalous left coronary artery with myocardial ischaemia

Original Article

Cite this article: Kalustian AB, Doan TT, Masand P, Gowda ST, Eilers LF, Reaves-O'Neal DL, Sachdeva S, Qureshi AM, Heinle J, Molossi SM, and Binsalamah ZM (2023) Evolution of surgical repair of intraseptal anomalous left coronary artery with myocardial ischaemia. *Cardiology in the Young* 33: 2342–2349. doi: [10.1017/S1047951123000197](https://doi.org/10.1017/S1047951123000197)

Received: 8 December 2022
Revised: 12 January 2023
Accepted: 13 January 2023
First published online: 2 March 2023

Keywords:

Anomalous aortic origin of coronary artery; anomalous left coronary artery; coronary anomalies; coronary unroofing; intraseptal coronary

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Abstract

Background: Anomalous aortic origin of the left coronary artery with intraseptal course is a rare coronary anomaly associated with an increased risk of myocardial ischaemia. The role and techniques for surgical intervention are evolving, with numerous novel surgical techniques for this challenging anatomy reported in the last 5 years. We report our single-centre experience with surgical repair of intraseptal anomalous left coronary artery in the paediatric population, including clinical presentation, evaluation, and short- to mid-term outcomes. **Methods:** All patients with coronary anomalies presenting to our institution undergo standardised clinical evaluation. Five patients aged 4 to 17 years underwent surgical intervention for intraseptal anomalous aortic origin of the left coronary artery between 2012 and 2022. Surgical techniques included coronary artery bypass grafting (n = 1), direct reimplantation with limited supra-arterial myotomy via right ventriculotomy (n = 1), and transconal supra-arterial myotomy with right ventricular outflow tract patch reconstruction (n = 3). **Results:** All patients had evidence of haemodynamically significant coronary compression, and three had evidence of inducible myocardial ischaemia pre-operatively. There were no deaths or major complications. Median follow-up was 6.1 months (range 3.1–33.4 months). Patients who underwent supra-arterial myotomy (with or without reimplantation) had improved coronary flow and perfusion based on stress imaging and catheterisation data. **Conclusions:** Surgical approaches to intraseptal anomalous left coronary artery with evidence of myocardial ischaemia continue to evolve, with new techniques demonstrating promising improvement in coronary perfusion. Further studies are warranted to determine long-term outcomes and refine indications for repair.

Anomalous aortic origin of a coronary artery is the second leading cause of sudden cardiac death in the young. Although the right coronary artery is more commonly involved, risk of sudden cardiac death is higher when the left coronary artery is affected.¹ Given the risk for myocardial ischaemia and sudden cardiac death, most patients with anomalous aortic origin of a left coronary artery with an intramural or interarterial course are recommended to undergo surgical repair via unroofing or reimplantation to improve coronary patency.^{1,2} In rare cases, an anomalous left coronary originating from a right sinus of Valsalva dives through the ventricular septum inferior to the pulmonary valve in the posterior right ventricular outflow tract prior to emerging at the epicardial surface. This intraseptal coronary segment is subject to compression by the muscular right ventricular outflow tract and is not accessible using the standard surgical approaches for anomalous coronaries. Though previously thought to be a benign variant,¹ recent data have demonstrated an association between intraseptal anomalous left coronary artery and myocardial ischaemia.³ Standardised multimodal evaluation utilising sensitive measures of coronary and myocardial perfusion has provided a framework to begin identifying high-risk features of coronary anomalies, though current data are limited.

In the setting of these challenges, multiple management pathways have emerged for the subset of patients considered at significant risk for ischaemia, balancing the unclear risk of serious adverse cardiac events with the unknown long-term outcomes for novel surgical repair techniques. Close serial monitoring, exercise restriction, and beta-blockade have been shown to reduce symptoms, even in patients with evidence of inducible ischaemia.^{2,3} Coronary artery bypass grafting was initially utilised,^{4–7} but is limited by graft failure secondary to competitive flow,^{1,5} leading surgeons to develop novel techniques to address sources of coronary obstruction.^{8–11}

In this study, we review the literature on the evolution of surgical approaches to this lesion and report our single-centre experience with surgical repair for intraseptal anomalous

left coronary artery with inducible ischaemia in the paediatric population, including clinical presentation, evaluation, and short- to mid-term outcomes.

Materials and methods

Patient population

The Coronary Artery Anomalies Program at our institution prospectively maintains a database of paediatric patients with intraseptal anomalous coronary arteries who presented at ages ≤ 20 years, beginning in December 2012. Study of these patients was approved by the Institutional Review Board under protocol H-30898, and informed consent was obtained. The database was retrospectively queried for patients who underwent surgical intervention and completed their 3-month post-operative workup. Clinical information was abstracted from their medical records.

Pre-operative assessment and indication for repair

All patients with coronary anomalies presenting to our institution undergo thorough standardised evaluation and multidisciplinary discussion with the coronary artery anomalies team (algorithms previously published).^{2,3} Following initial diagnosis with echocardiogram, CT with coronary angiography further delineates coronary anatomy for pre-operative planning, including length of intraseptal segment, pattern of distal branches, and ostial configuration. Exercise stress testing evaluates the patient's functional baseline, presence of inducible symptoms, and electrocardiographic evidence of inducible ischaemia. Dobutamine stress cardiac MRI identifies baseline or stress-induced perfusion defects, wall-motion abnormalities, or delayed gadolinium enhancement suggestive of myocardial scarring. Finally, cardiac catheterisation – supplemented by coronary angiography as well as coronary functional evaluation with fractional flow reserve, instantaneous wave-free ratio, and intravascular ultrasound – determines the degree of compression exerted on the intraseptal segment corresponding to the risk of ischaemia. Provocative pharmacologic stress with adenosine and dobutamine infusion during invasive cardiac catheterisation testing is performed with a goal heart rate of 75% exercise-predicted maximum. Coronary flow compromise is considered to be haemodynamically significant if the patient has a resting/baseline instantaneous wave-free ratio < 0.89 , instantaneous wave-free ratio during dobutamine infusion < 0.85 , or diastolic fractional flow reserve during dobutamine infusion < 0.8 .

In the presence of concerning symptoms (such as exertional syncope/near-syncope or angina-like chest pain) associated with inducible or baseline ischaemia in the left coronary artery territory and/or significant compression of the anomalous coronary artery with compromised flow, surgical intervention is recommended if the benefits are felt to exceed risks. Shared decision-making with patient and family involves an extensive discussion regarding risks associated with these lesions, implications of the patient's anatomic and functional test results, options for surgical intervention (including disclosure of the lack of long-term experience with novel techniques), and alternative management strategies, including close observation with exercise restriction and possible beta blockade. Ultimately, the selection of operative technique was at the discretion of the consulting surgeon, considering patient anatomic factors as well as contemporarily available published data on the risks, benefits, and outcomes of novel techniques.

Post-operative assessment and follow-up

All patients have electrocardiogram and echocardiogram prior to hospital discharge and 30 days post-operatively. Pre-operative studies are repeated 3 months post-operatively, with particular attention to coronary patency, signs of right ventricular outflow tract obstruction, ischaemic ST changes, reversal of any pre-operatively discovered inducible or baseline ischaemia, and change in coronary compression. Based on expert consensus guidelines, we recommend exercise restriction post-operatively, which may be lifted after 3 months if there is no evidence of inducible ischaemia on post-operative functional testing and the patient has no history of aborted sudden cardiac death^{1,2}. Patients are followed for life with episodic surveillance imaging.

Surgical technique: transconal supra-arterial myotomy and right ventricular outflow tract patch reconstruction

As described by Najm and Ahmad,⁹ the intraseptal coronary was accessed via a right ventriculotomy just inferior to the pulmonary valve annulus. Limited aortotomy was also performed to evaluate the coronary ostia and probe the intraseptal coronary to identify its course. The intraseptal coronary was released by supra-arterial septal myotomy, mobilised, and excluded from the posterior right ventricular outflow tract. Our approach favours a combination of blunt dissection and electrocautery to aid visualisation and avoid coronary injury. The right ventricular outflow tract was elongated and reconstructed using a fresh autologous pericardial patch sutured circumferentially posteriorly and tapered anteriorly for direct re-approximation of the native anterior wall.

Surgical technique: direct reimplantation and limited supra-arterial myotomy

This technique and patient were described in a previous publication⁸. With the aim to relieve interarterial compression and correct an acute angle takeoff, direct reimplantation was accomplished by transecting the proximal left main coronary artery from a single right coronary and directly anastomosing it to a neo-ostium created in the right coronary sinus of Valsalva. Via a right ventriculotomy, and with the assistance of a coronary probe, a supra-arterial myotomy was performed to release the majority of the intraseptal

segment into the right ventricular cavity while maintaining ventricular wall integrity at the entry and exit sites of the coronary artery.

Surgical technique: coronary artery bypass graft

In the setting of complex coronary anatomy with an intraseptal collateral from the right coronary supplying a diminutive (approximately 1.25 mm diameter) distal left anterior descending artery, coronary artery bypass grafting was felt to be the only surgical option to improve perfusion. On cardiopulmonary bypass and cardioplegia, the left internal thoracic artery was anastomosed to the small distal left anterior descending artery. This patient was also mentioned in a previous publication³.

Statistical analysis

Descriptive statistics were performed using STATA 17.0 software (StataCorp LLC, College Station, TX). Categorical data are presented as absolute and relative frequencies. Continuous variables are summarised by median and ranges.

Table 1. Patient demographics and operative information

Patient	Symptoms	Age (yrs)	Weight (kg)	Year	Repair Type	CPB (min)	CXT (min)	LOS (days)
1	Syncope*, dyspnea*	4	60.5	2017	CABG	149	100	3
2	Pre-syncope*, chest pain*, dyspnea*	16	80.9	2019	Direct reimplantation, limited supra-arterial myotomy	179	135	4
3	Syncope†, palpitations†	17	84	2022	Transconal supra-arterial myotomy, RVOT patch	173	137	4
4	Syncope†, palpitations†, chest pain†, dyspnea*	15	30.4	2022	Transconal supra-arterial myotomy, RVOT patch	174	137	8
5	Pre-syncope*, fatigue*	10	34.1	2022	Transconal supra-arterial myotomy, RVOT patch	172	101	4
<i>Median</i>		<i>15.2</i>	<i>60.5</i>			<i>173</i>	<i>101</i>	<i>4</i>

* Symptom occurring with exertion only, † symptom occurring at rest and exertion.

CABG = coronary artery bypass graft, CPB = cardiopulmonary bypass time, CXT = aortic cross-clamp time, LOS = post-operative length of hospital stay, min = minutes, yrs = years, RVOT = right ventricular outflow tract.

Results

Patient presentation and coronary anatomy

From 2012 to 2022, five patients aged 4–17 years underwent surgical intervention for intraseptal anomalous left coronary artery at our institution. Repair techniques were coronary artery bypass graft (patient 1), direct reimplantation with limited supra-arterial myotomy (patient 2), and transconal supra-arterial myotomy with right ventricular outflow tract reconstruction (patients 3–5).

The patients' presenting symptoms and operative information are summarised in Table 1. Three patients were female (60%). None had any cardiac comorbidities, arrhythmias, or baseline ST segment changes on electrocardiogram. Four patients (80%) had a family history of early heart disease or sudden death. All were symptomatic pre-operatively. The youngest (patient 1) presented at age 4 with frequent exertional syncope since the age of 2. Three patients (60%) had at least one syncopal episode, and the remaining 2 (40%) had near-syncopal events with dizziness and vision changes. Other common symptoms were exertional dyspnoea ($n = 3$, 60%), chest pain ($n = 2$, 40%), and palpitations ($n = 2$, 40%). Pertinent findings on pre-operative workup are reported in Table 2. No patient had wall motion abnormalities or ventricular dysfunction on echocardiography, and there were no true ischaemic ECG changes on exercise stress testing (though two patients [40%] experienced exercise-induced symptoms). Three patients (1, 2, 5; 60%) had evidence of inducible subendocardial ischaemia on dobutamine stress cardiac MRI pre-operatively, of whom one (patient 2) also had delayed myocardial hyperenhancement and inducible septal hypokinesis consistent with prior myocardial infarction. All patients had evidence of haemodynamically significant coronary compression on pre-operative cardiac catheterisation, evidenced by at least one abnormal parameter (baseline/dobutamine instantaneous wave-free ratio or fractional flow reserve; Table 3). Axial imaging (all patients) and intravascular ultrasound (performed on patients 2–5) also showed decreased calibre of the intraseptal coronary segment in all except patient 5.

Table 2 also summarises the abnormal origin and course of each patient's major coronary branches. Patients 1, 2, and 4 had a single coronary ostium from the right coronary sinus with anomalous branches supplying the left coronary circulation. Patient 1 had complex anatomy with a right coronary artery supplying the left coronary system via a collateral with a long intraseptal course and branching. The collateral vessel supplied a ramus intermedius

artery, very small left anterior descending artery (which was either interrupted or had myocardial bridging), and circumflex. Intravascular ultrasound was not performed for patient 1 due to the small calibre of the coronary vessels; however, significant systolic compression of the intraseptal coronary was observed. Patient 2's left main coronary artery arose from the proximal single right coronary artery with acute angulation prior to an interarterial and intraseptal course. Patient 4 had a single coronary trunk with trifurcation into the right coronary, retroaortic left circumflex, and left anterior descending artery with intraseptal course. Patients 3 and 5 had anomalous intraseptal left coronary arteries originating from separate ostia within the right coronary sinus, with bifurcation into the left anterior descending and left circumflex branches distal to the intraseptal segment's emergence from the myocardium. Representative pre- and post-operative imaging for patient 3 is shown in Figure 1.

Post-operative course and follow-up

Median post-operative length of stay was 4 days (range 3 to 8 days). Prior to discharge, patient 1 had intermittent accelerated ventricular rhythm and sinus tachycardia (maintained on atenolol). Patient 5 had ventricular fibrillation while weaning from cardiopulmonary bypass, which resolved after defibrillation without recurrent arrhythmias. Patient 4 had asymptomatic moderate pleural effusions (managed with diuresis). Patient 4 had asymptomatic frequent premature ventricular contractions comprising up to 10% of beats on Holter monitoring 1 month post-operatively that resolved on repeat 3-month study. In the setting of her ectopy and abnormal dobutamine stress cardiac MRI, she was started on low-dose beta blockade; however, it was poorly tolerated (episode of near-syncope and hypotension at 4 months post-operation) and discontinued within several weeks. While this episode is attributed to the beta blockade, she is undergoing cardiac event monitoring at the time of this report. In the first 6 months post-operatively, 2 patients (40%) visited the emergency department with symptoms but were discharged with routine follow-up after stable electrocardiograms and normal troponins: patient 2 had chest pain, dizziness, and dyspnoea and patient 3 had intermittent palpitations and mild dyspnoea. Three patients (60%) reported symptoms at some point in the post-operative period, although none had correlating abnormalities on further workup. Patient 1 was admitted to the hospital after a syncopal episode 1 year

Table 2. Summary of multimodal evaluation and follow-up status

	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5
Coronary Anatomy Type*	A3 LCA from RCA, intraseptal bifurcation	A1 LCA from RCA, epicardial bifurcation	A2 LCA from separate origin in right sinus, distal bifurcation	B1 Intraseptal LAD from RCA, retroaortic LCx from RCA	A2 LCA from separate origin in right sinus, distal bifurcation
Intraseptal Segment Length (cm)	Unknown	3	3.3	4.2	2.5
Pre-op Echo EF (%)	68	62	67	Qualitatively normal systolic function	62
Post-op Echo EF (%)	56	57	66	67	68
Pre-op EST					
Symptoms	Not done	Chest pain, dizziness	None	Dyspnea	None
Ischaemic Changes	Not done	None	Transient nonspecific upsloping inferolateral ST segment	None	None
Post-op EST					
Symptoms	None	None	None	None	None
Ischaemic Changes	None	None	None	None	Transient nonspecific inferolateral ST-T changes
Pre-op DSCMR					
Wall-motion abnormality	None	Inducible	None	None	None
Perfusion defect	Inducible	Inducible	None	None	Inducible
Delayed hyperenhancement	None	Subendocardial	None	None	None
Post-op DSCMR					
Wall-motion abnormality	None	Baseline	None	None	None
Perfusion defect	Inducible	None – resolved	None	None	None – resolved
Delayed hyperenhancement		Transmural	None	Subendocardial	None
Follow-up time	2.8 years	1.2 years	6.1 months	4.7 months	3.1 months
Follow-up status	Single post-operative syncopal event, currently asymptomatic, tolerating exercise	Asymptomatic, tolerating exercise	Mild intermittent palpitations, tolerating exercise.	Event monitor ongoing for PVCs and near-syncope/hypotension while on beta blocker, otherwise asymptomatic. Exercise restriction.	Asymptomatic, cleared for exercise.

Summary of pertinent workup findings pre-operatively and post-operatively, with most-recent post-operative findings reported when multiple studies of that type were performed. Follow-up information is given as the time from operation to most recent patient contact at our institution and most recent clinical information.

* Categories of pre-operative coronary anatomy origin and course as previously described by Doan et al.³ in Figure 1; summary descriptions of the patients' coronary anatomy are provided in the table.

DSCMR = dobutamine stress cardiac magnetic resonance imaging, Echo = echocardiogram, EF = left ventricular ejection fraction, EST = exercise stress testing, LCA = left coronary artery, LCx = left circumflex coronary artery, Pre-op = pre-operative, Post-op = post-operative, RCA = right coronary artery.

Table 3. Haemodynamic coronary assessment

		Patient 1		Patient 2		Patient 3		Patient 4		Patient 5		
		Pre	Post	Pre	Post	Pre	Post	Pre	Post	Pre	Post	
FFR	Rest	0.91	0.94	0.79	0.88	0.94	0.88	0.99	0.97	1	1	0.99
	Dobutamine	0.75	0.95	0.84	0.70	0.96	0.76	0.95	0.87	0.94	0.96	0.95
	Dobutamine diastolic	0.67		0.79	0.69	0.89	0.77	0.95	0.81	0.89		0.91
iFR	Rest	0.86	0.87	0.73	0.85	0.94	0.86	0.95	0.98	1	1	0.96
	Dobutamine			0.77	0.65	0.95	0.73	0.95	0.80	0.90	0.82	0.91
IVUS (%)					42	26	46.5	16	51	14.8	3.3	5.5

Haemodynamic coronary data from pre- and post-operative cardiac catheterisation for each patient. Red indicates abnormal values.

FFR = fractional flow reserve (normal > 0.8), iFR = instantaneous wave-free ratio (normal rest > 0.89, dobutamine > 0.85), IVUS = intravascular ultrasound (% compression).

post-operatively (first post-operative recurrence). A long-term cardiac event monitor was implanted, which has since demonstrated no arrhythmias.

At a median follow-up of 6.1 months (range 3.1 to 33.4 months), there were no deaths, reoperations, or post-operative catheter-based interventions. At the time of most recent follow-up, patient 3 reported mild intermittent palpitations, and the remaining patients reported complete symptom resolution. All patients with the exception of patient 4 were cleared of their exercise restrictions and tolerating activity without symptoms.

Pertinent findings on post-operative studies are summarised in Table 2. Patient 1's initial post-operative dobutamine stress cardiac MRI showed resolution of her stress perfusion defect, but follow-up scan 3 years post-operatively demonstrated recurrence at the basal anterior septum despite patent graft on angiography. Patient 2's post-operative study showed no inducible perfusion defects, though the regional wall motion abnormality was now present at rest and associated with myocardial thinning and transmural delayed hyperenhancement, consistent with the pre-operative myocardial infarction. Patient 4 developed a new area of delayed hyperenhancement along the right ventricular outflow tract and basal anteroseptal and inferoseptal wall, corresponding to the region of surgical incision. Patient 5 had a subendocardial inducible perfusion defect in the mid-septum pre-operatively, which resolved post-operatively.

All patients had pre- and post-operative cardiac catheterisation with fractional flow reserve and instantaneous wave-free ratio, and all but patient 1 had intravascular ultrasound (Table 3). Patient 1 (coronary artery bypass graft) had two post-operative catheterisations at 1 and 3 years. At first, her fractional flow reserve and instantaneous wave-free ratios increased from pre-operative values, but by repeat catheterisation, they had decreased, some to below pre-operative values. However, the interpretation of these measures is limited in the setting of competitive flow to the graft, which was confirmed patent by angiography. The remaining patients' post-operative catheterisations were performed at a median of 3.5 months following surgery (range 2.9 to 4.5 months). The patients who underwent supra-arterial myotomy (patients 2–5) had normal coronary perfusion measures post-operatively with normalisation of all pre-operative abnormal values. Patients who had significant coronary compression on intravascular ultrasound (patients 2–4) had improvements on post-operative catheterisation, with CT angiography and dobutamine stress cardiac MRI also demonstrating coronary patency with improved calibre of the previously intraseptal segment.

Discussion

As understanding of the natural history of rare variants of anomalous coronary artery anatomy improves, configurations previously considered benign, including intraseptal anomalous aortic origin of the left coronary artery, are being linked to myocardial ischaemia.³ As treatment algorithms evolve to identify and mitigate these risks, new surgical techniques for improving coronary perfusion have emerged. Previously, surgical intervention on such lesions was reserved for patients with existing ischaemia. However, with increasing utilisation of multimodal evaluation to detect high-risk lesions and promising early results from novel techniques, there is a shift towards offering earlier surgical repair to patients presenting without baseline ischaemia. The risk–benefit ratio for surgical intervention on the intraseptal coronary artery is changing rapidly in the setting of recent data demonstrating short- to mid-term safety and efficacy of new surgical options, increased experience with these techniques in the paediatric population, and improved understanding of high-risk features of intraseptal anomalous coronaries. Surgery is becoming an appealing option with the potential to lower the risk of ischaemia and sudden cardiac death while allowing for resumption of an active lifestyle. Patients with high-risk features who were previously not offered intervention may now be candidates based on this evolving paradigm.

Ultimately, more information regarding outcomes for patients with surgical intervention, medical management, or observation will be required to strike a balance in patient selection for operative management. This can only be achieved with regimented approaches to workup and surveillance, utilising multiple modalities for both functional and anatomic coronary assessment. Paediatric patients with these lesions face significant uncertainty, with many years of future exposure to risks associated with their anatomy or to the effects of a surgical intervention. The need for detailed and multidisciplinary discussion regarding the choice of management in this population cannot be overstated. In our experience, the development of a dedicated coronary artery anomalies team is integral to fulfilling these goals and standardising practice.

Overall goals for surgical intervention on anomalous coronaries are to relieve extrinsic compression while minimising turbulent flow and disruption to surrounding structures. Techniques such as direct reimplantation for interarterial coronaries, aortic unroofing for intramural coronaries, and ostioplasty for proximal stenosis are becoming more widely accepted for coronary arteries with anomalous origin and/or course. The challenging intraseptal



Figure 1. Representative imaging for patient 3, who underwent transconal supra-arterial myotomy and right ventricular outflow tract patch reconstruction. (a) Pre-operative three-dimensional CT reconstruction demonstrating intraseptal course of the anomalous left coronary artery (orange) through the posterior right ventricular outflow tract (red); (b) representative angiography images demonstrating improved patency of the intraseptal coronary (red arrow) post-operatively (right) compared to pre-operatively (left); (c) representative coronal CT images demonstrating improved separation of the coronary from the right ventricular outflow tract.

segment was previously only addressed with coronary artery bypass grafting,⁴⁻⁷ which is fraught with disadvantages such as competitive graft flow and graft failure⁵, and is now generally avoided if other options are feasible. Patient 1 in our series developed recurrent perfusion defects after several years and had recurrent syncope after coronary artery bypass grafting. In the paediatric population, graft durability and growth potential are of particular

concern. Newer strategies seek to address the underlying anatomic sources of compression to improve perfusion. Common themes in these surgical approaches to the intraseptal coronary include supra-arterial “unroofing” myotomy,⁸⁻¹² though means of access differ (right ventriculotomy,⁸⁻¹⁰ pulmonary root harvest^{11,12}). Some methods include patch reconstruction of portions of the posterior right ventricular outflow tract to create space for the

formerly intra-septal segment.^{9–12} Such techniques may also be combined with existing methods of addressing any proximal compression.^{8,11}

Our institution previously described a case combining direct reimplantation of the proximal left coronary artery with the release of the intraseptal segment via right ventriculotomy and limited supra-arterial myotomy.⁸ This patient (2) has thus far had a positive outcome with improvement in indices of coronary perfusion and stable cardiac function after a pre-operative myocardial infarction. However, he is left with two areas with potential for compression where the coronary was not released at its entry and exit sites from the ventricular septum. Najm and colleagues first published their experience with transconal supra-arterial myotomy and patch reconstruction of the right ventricular outflow tract in 2019.⁹ Mid-term outcomes in their series of 14 consecutive patients (median operative age 49 years) were favourable with resolution of pre-operative symptoms, patent left coronary artery, improvement in fractional flow reserve, and no deaths or major complications after a median follow-up of 11 months.¹⁰ Gaillard et al. also published a series including five cases in which they accessed the intraseptal segment by harvesting the pulmonary root similar to the Ross procedure, performed septal myotomy, and then reconstructed the right ventricular outflow tract with an augmenting posterior patch.¹¹ One of their patients who had concurrent coronary reimplantation required immediate post-operative angioplasty with stent placement, but the remainder were asymptomatic and alive at follow-up.¹¹ A similar technique is described in a case report by Agati et al.¹² in Italy for an 11-year-old asymptomatic boy with intraseptal anomalous aortic origin of the left coronary artery and inducible perfusion defect on nuclear myocardial stress perfusion test that normalised on repeat study 6 months post-operatively. We have not performed this technique as there is no evidence that a complete pulmonary root mobilisation provides superior results to septal myotomy, and it may pose a greater risk of distorting outflow tract geometry.

At our institution, we currently offer surgical intervention to patients with an intraseptal coronary artery with evidence of myocardial ischaemia (based on the presence of significant symptoms and/or functional testing) or compromised coronary flow (based on catheter-based measures) when the benefits of the proposed intervention are felt to exceed its risks. Our methods for evaluation and management algorithms are detailed in prior publications^{2,3}, though an increasing amount of data and experience with these promising new techniques has led to more frequent operative intervention on high-risk patients in the past few years. Our favoured approach to the intraseptal segment at this time is that pioneered by Najm and colleagues⁹ as it provides a complete release of the intramuscular segment and improved coronary perfusion with normalisation of fractional flow reserve and instantaneous wave-free ratio on follow-up catheterisation. The wide, long right ventricular outflow tract patch allows ample space for the coronary artery and the native wall anterior anastomosis permits somatic growth. In our limited experience, we have not observed any evidence of outflow tract obstruction on echocardiography or other modalities. Theoretically, this technique could be combined with techniques such as ostioplasty or reimplantation for relief of concomitant proximal coronary obstruction, though we have not encountered this situation to date. We have performed this intervention on patients as young as 10 years of age with good short-term results, but more experience is needed to determine the optimal age of intervention. At this time, we recommend individualised risk assessment and informed discussion of all options for

paediatric patients, with the strongest preference for surgical intervention in the presence of severe ischaemic symptoms, aborted sudden cardiac death, or evidence of myocardial infarction.

Our short- to mid-term outcomes with operative intervention for paediatric patients with intraseptal coronaries are generally promising, with no mortalities or coronary reinterventions. Patients had improved symptoms and coronary perfusion following supra-arterial myotomy procedures. However, such procedures are not without significant potential perioperative risk, and the long-term outcomes remain unknown. One patient developed a new area of delayed myocardial enhancement on post-operative MRI, potentially related to the right ventriculotomy site, though with preserved cardiac function. Most patients had electrocardiographic changes of unclear significance in the ST segment and T-waves post-operatively, consistent with previously reported findings¹³ following the repair of anomalous aortic origin of coronary arteries that were not associated with adverse midterm outcomes. One patient had frequent premature ventricular contractions post-operatively, though asymptomatic and since resolved. Although no malignant arrhythmias have been observed in our population to date, the long-term incidence and significance of arrhythmias remain to be seen.

Limitations

Major limitations of this report include the small number of patients given the rarity of the anatomy and short duration of follow-up given the recent experience with novel procedures. Small sample sizes limit the use of comparative statistical methods to detect differences among populations. It also limits generalisability and precludes drawing conclusions regarding the efficacy and safety of this procedure in the long term.

Conclusions

Novel techniques to relieve anatomic compression of anomalous intraseptal coronary arteries demonstrate positive short-term outcomes with improved coronary perfusion and symptoms. Rigorous multimodal pre- and post-operative evaluation of cardiac anatomy and function is paramount for the care of these patients. Surgery should only be considered when there is a significant risk for myocardial ischaemia and objective evidence of impaired coronary flow. Continued follow-up is necessary to determine long-term outcomes for these procedures and delineate which patients are most likely to benefit from surgical intervention.

Acknowledgements. We thank the Texas Children's Hospital Coronary Artery Anomalies Program (CAAP) for their dedication to advancing evidence-based patient care.

Financial support. No dedicated or external funding were utilised for the completion of this project.

Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (Determining The Natural and "Unnatural" History of Anomalous Aortic Origin of a Coronary Artery With an Interarterial Course (AAOCA): Establishing A Multi-Institutional Registry, Protocol H-30898) and with the Helsinki Declaration of 1975, as revised in 2008, and has been approved by the institutional committees (Baylor College of Medicine).

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