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Retroaortic Innominate Vein in Association With Pulmonary Atresia/Major Aortopulmonary Collaterals

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ABSTRACT

BACKGROUND Retroaortic innominate vein (RAIV) is an extremely rare anomaly of systemic venous return. The prevalence of RAIV has been estimated to be 0.02% in individuals without congenital heart disease and 0.5% in those with congenital heart disease. Previous studies have demonstrated an association between RAIV and both conotruncal abnormalities and right aortic arch. Our center specializes in surgical procedures for patients with pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs), and we have frequently observed the presence of RAIV in these patients. The purpose of this study was to evaluate the prevalence and anatomic characteristics of RAIV.

METHODS This was a single-center retrospective review of medical records to identify patients with an RAIV. The study period was 2002 through 2022, with approximately 20,000 patients evaluated during this time frame.

RESULTS A total of 99 patients were identified with an RAIV. Of the 99 patients, 64 (65%) had PA/VSD/MAPCAs. Other diagnoses included tetralogy of Fallot (n = 16), tetralogy of Fallot with pulmonary atresia (n = 3), double outlet right ventricle (n = 4), peripheral pulmonary artery stenosis (n = 3), and other (n = 9). In the cohort of patients with PA/VSD/MAPCAs, 69% had a right aortic arch and 30% had completely absent central pulmonary arteries.

CONCLUSIONS The overwhelming majority (90%) of patients with RAIV had conotruncal abnormalities, with PA/VSD/MAPCAs accounting for the preponderance of cases in our center.

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Retroaortic innominate vein (RAIV) is an extremely rare anomaly of systemic venous return.[1] The prevalence of RAIV has been estimated to be 0.02% in individuals without congenital heart disease (CHD) and 0.5% in those with CHD.[2,3] More than 80% of patients with an RAIV have obstruction of the right ventricular outflow tract, with tetralogy of Fallot (with or without pulmonary atresia) being the most common diagnosis. Patients with RAIV have also been noted to have a high prevalence of right aortic arch.[4]

Our surgical center specializes in surgical procedures for patients with the diagnosis of pulmonary atresia with

IN SHORT

- In a retrospective review, we identified 99 patients with a retroaortic innominate vein. Of the 99 patients, 64 had the diagnosis of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs).
- In the 64 patients with PA/VSD/MAPCAs, the prevalence of absent central pulmonary arteries was 30% or approximately 88% higher than anticipated in this group.
- This report describes an association between retroaortic innominate vein and PA/VSD/MAPCAs.

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ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCAs). PA/VSD/MAPCAs is a relatively rare form of CHD with an incidence of 1 in 70,000 births. The intracardiac anatomy of PA/VSD/MAPCAs is characterized by a malalignment-type VSD with an overriding aorta. However, because of the presence of pulmonary atresia and the absence of a ductus arteriosus, the only possible source of blood flow to the pulmonary arteries is through interconnections with the MAPCAs. Patients with PA/VSD/MAPCAs have either diminutive or completely absent central pulmonary arteries.

We have noted a significant prevalence of RAIV in our patients undergoing surgical repair of PA/VSD/MAPCAs. The purpose of this study was to evaluate the prevalence and anatomic characteristics of RAIV in patients with PA/VSD/MAPCAs.

PATIENTS AND METHODS

This was a single-center retrospective study of patients identified with an RAIV. Most patients were identified through the syngo Dynamics echocardiographic database. Additional patients were identified through a search of The Society of Thoracic Surgeons Congenital Heart Surgery Database. The study was from 2002 through 2022. This study was approved by the Stanford University institutional review board (IRB protocol ID 33924 approved April 14, 2015 and renewed March 31, 2021). The need for written consent was waived by the institutional review board.

During the 20-year study period, it is estimated that approximately 20,000 unique patients have undergone diagnostic evaluation or surgical treatment. The diagnosis of RAIV can be reliably made by echocardiography (Figure 1) and computed tomography angiography (Figures 2, 3).

RESULTS

A total of 99 patients were identified with an RAIV. All 99 patients with an RAIV had echocardiography, and 94 of the 99 (95%) had the recognition of RAIV made by this modality. Eighteen patients with RAIV underwent computed tomography angiography, and 17 of 18 (94%) had the recognition of RAIV made by this modality. There were 5 patients who had the diagnosis of RAIV made at the time of the surgical procedure. A summary of the diagnoses is shown in the Table; 89 (90%) patients had conotruncal defects, and 69 of the 99 (70%) patients had a right aortic arch. Subsequently, 98 of the 99 patients with RAIV underwent surgical treatment.

Sixty-four (65%) patients had the diagnosis of PA/VSD/MAPCAs; 56 of these patients had bilateral MAPCAs. There were 8 patients who had unilateral MAPCAs, including 4 patients with a left-sided ductus arteriosus, 2 patients with the left pulmonary arising from the ascending aorta (left hemitruncus), and 2 patients with

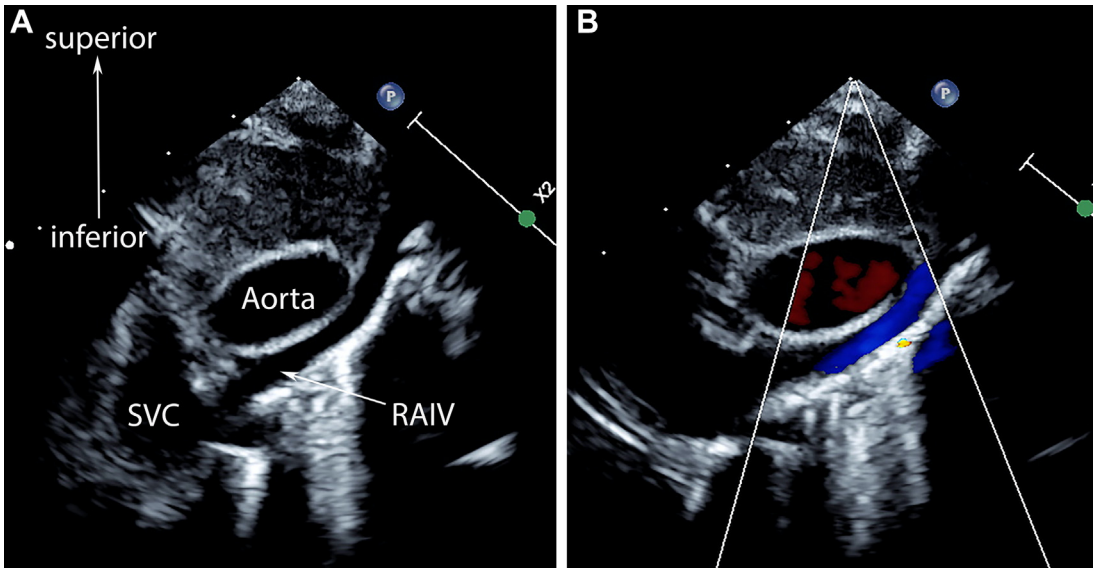

FIGURE 1 (A) Echocardiogram demonstrating the presence of a retroaortic innominate vein (RAIV). The innominate vein is visualized posterior to the aorta and then entering into the right superior vena cava (SVC). (B) Color Doppler demonstrates an inferior trajectory of the venous flow with the retroaortic innominate vein (blue) opposite that of the aorta (red).

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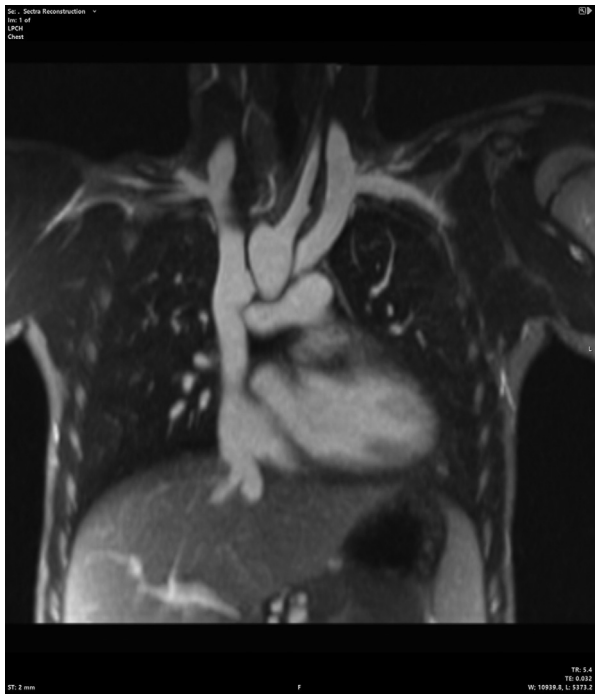

FIGURE 2 Computed tomography angiography demonstrating the presence of a retroaortic innominate vein.

the right pulmonary arising from the ascending aorta (right hemitruncus).

Eighteen (28%) of the 64 patients with PA/VSD/MAPCAs had 22q11 microdeletion. There were 2 patients with Alagille syndrome.

Forty-four (69%) patients with PA/VSD/MAPCAs and an RAIV had a right aortic arch; 31% had a left aortic arch. Central pulmonary arteries were present in 45 (70%) and were absent in 19 (30%). Retroesophageal MAPCAs were present in 54% of the patients. Coronary artery anomalies were identified in 15% of the patients.

COMMENT

In this single-institution study, we identified 99 patients with an RAIV for an estimated prevalence of 0.5% of patients undergoing screening for CHD. Ninety percent of the patients had a conotruncal defect, and 70% had a right aortic arch. Two-thirds of the entire cohort had the diagnosis of PA/VSD/MAPCAs, no doubt a reflection of the large number of referrals that the surgical center receives because of our interest and expertise. We estimate that our entire experience with PA/VSD/MAPCAs is approximately 800 patients, meaning that the prevalence of RAIV in this entity is 8%. This report indicates a relationship between RAIV and patients with MAPCAs.

By definition, an RAIV passes through the space between the ascending and descending aorta and thus shares (or possibly competes with) the same space normally reserved for the branch pulmonary arteries. It is thus conceivable that the presence of an RAIV could result in an underdevelopment of the pulmonary arteries from an embryologic standpoint. All patients with MAPCA-dependent pulmonary circulation have diminutive branch pulmonary arteries, and between 16% and 20% have completely absent central pulmonary arteries. In this study focusing on patients with RAIV and

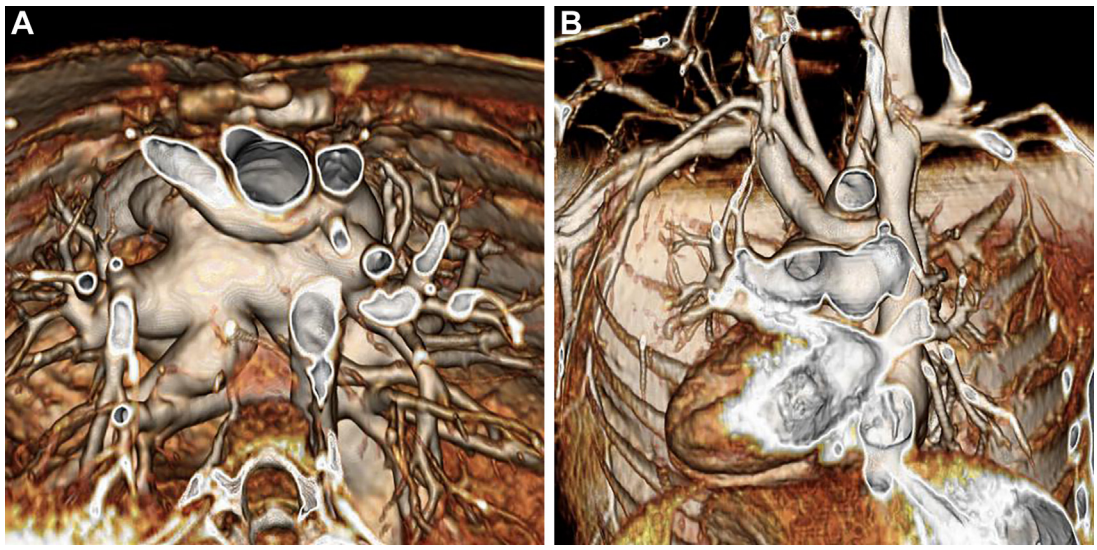

FIGURE 3 (A) Three-dimensional reconstruction of a retroaortic innominate vein shown from a cephalad view. (B) Three-dimensional reconstruction of a retroaortic innominate vein shown from a posterior view.

TABLE Summary of Patients With a Retroaortic Innominate Vein

Diagnosis	No.	%
PA/VSD/MAPCAs	64	65
Tetralogy of Fallot	16	16
Tetralogy of Fallot/pulmonary atresia	3	3
Double outlet right ventricle	4	4
Peripheral pulmonary artery stenosis	3	3
Tetralogy of Fallot/absent pulmonary valve	1	1
Truncus arteriosus	1	1
Coarctation aorta	1	1
Ventricular septal defect	1	1
Loews-Dietz syndrome	1	1
Double aortic arch	1	1
Hypoplastic aortic arch	1	1
Atrial septal defect	1	1
Murmur evaluation	1	1

PA/VSD/MAPCAs, pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries.

MAPCAs, we identified 30% of the cohort with completely absent central pulmonary arteries or approximately 50% to 88% higher than expected. Whether this finding is based on an RAIV competing for the aortopulmonary window space or is intertwined in the genetic coding is currently unknown.

MAPCAs originating from the descending aorta may also occupy the space anterior to the bronchi and thus could potentially be affected by the competition for space. Most patients with PA/VSD/MAPCAs have between 2 and 4 MAPCAs originating from the descending aorta and coursing anteriorly and then laterally to reach the lung parenchyma. An alternative anatomic pathway is for a MAPCA to come off the lateral aspect of the aorta and go behind the esophagus (ie, retroesophageal MAPCAs).[5] Previous studies have demonstrated that retroesophageal MAPCAs are more likely to lack interconnections with the pulmonary arteries and to have a higher prevalence of absent pulmonary arteries.[6] In this study, the prevalence of retroesophageal MAPCAs in patients with an RAIV was 54% and thus not different from patients with predominantly right arches.

Another form of CHD that typically has diminutive branch pulmonary arteries is peripheral pulmonary artery stenosis. This entity is often linked to Williams syndrome, Alagille syndrome, elastin arteriopathy, and some conotruncal abnormalities. In our series of 145 patients who underwent surgical repair of peripheral pulmonary artery stenosis,[7] we found just 3 patients with an RAIV. All 3 of these patients had Alagille syndrome (2 of the 3 had a right aortic arch). There were 21 patients who had tetralogy of Fallot with peripheral pulmonary artery stenosis in this series, and none had an RAIV. Although this is a relatively small sample size, it suggests there is no association between tetralogy of Fallot/peripheral pulmonary artery stenosis and RAIV.

In the process of searching for patients with RAIV, we found 16 who did have the diagnosis of tetralogy of Fallot with pulmonary stenosis and confluent pulmonary arteries. RAIV is relatively uncommon in this entity, with the literature reporting a 1% to 2% prevalence. Our experience is consistent with these numbers as we estimate the prevalence at 2% to 3%. In contrast, left superior vena cava with anomalous drainage to the coronary sinus is common in tetralogy with pulmonary stenosis. This anomaly of systemic venous drainage is seen in 10% of patients with tetralogy of Fallot and pulmonary stenosis, whereas it is exceedingly rare in PA/VSD/MAPCAs.[8] These differences in the prevalence of systemic venous anomalies between the "standard" form of tetralogy of Fallot and PA/VSD/MAPCAs probably hold some clues about the embryology of these entities but have yet to be elucidated.

A left superior vena cava with anomalous drainage to the coronary sinus is observed in 0.3% of patients without CHD and 5% of patients with CHD.[9] Thus, a left superior vena cava is approximately 10 times more common than RAIV. Left superior venae cavae are relatively common in tetralogy of Fallot, double outlet right ventricle, and truncus arteriosus. However, there is increasing interest in and evidence to support the theory that left superior venae cavae can be the cause of left ventricular inflow obstruction secondary to compression of the left atrium by a dilated coronary sinus.[10] The net result during embryogenesis of the heart is underdevelopment of left-sided heart structures ranging from borderline left ventricle all the way to hypoplastic left heart syndrome. Recent efforts at rehabilitating borderline left ventricles when there is a left superior vena cava with drainage to the coronary sinus include redirecting flow so that it can no longer impede left ventricular inflow—usually by dividing the cava at its junction with the heart and anastomosing the divided end to the right superior vena cava. The results of this strategy have been favorable to date.

Given the relationship between a left superior vena cava with drainage to the coronary sinus and left ventricular inflow obstruction, the obvious question is whether there is an analogous situation with RAIV. In our surgical practice, although RAIVs do take up a portion of the space in the aortopulmonary window, we have found that there is still adequate space for the reconstructed pulmonary arteries and unifocalized MAPCAs. Therefore, we have not attempted to move the innominate vein anterior to the aorta. One practical consideration why translocation is not all that practical is that the ascending aorta is typically large in PA/VSD/

MAPCAs because it carries 2 or 3 cardiac outputs. Whereas the RAIV may conceivably play a role in the underdevelopment of the branch pulmonary arteries during embryogenesis, it is unlikely (in our opinion) to play a physiologic role in constraining pulmonary artery growth after birth.

In summary, this study was performed to assess the association between RAIV and PA/VSD/MAPCAs. We were able to identify 64 patients with this combination and therefore estimate the prevalence of RAIV in PA/VSD/MAPCAs at 8%, or perhaps 4-fold higher than that seen in tetralogy of Fallot with pulmonary stenosis. Patients with RAIV and PA/VSD/MAPCAs had a considerably higher than expected prevalence of absent central pulmonary arteries. This finding provides circumstantial evidence that RAIV can create a competition for space and affect the embryogenesis of pulmonary artery development.

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DISCLOSURES

The authors have no conflicts of interest to disclose.

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