Prevalence and Anatomy of Retroesophageal Major Aortopulmonary Collateral Arteries

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Background. **Major aortopulmonary collateral arteries (MAPCAs) are the sole source of pulmonary blood flow in patients with pulmonary atresia and absent ductus arteriosus. The anatomy of MAPCAs can be highly variable, both in the number of MAPCAs supplying each lung and the anatomic origin and course of the MAPCAs. This study evaluated the prevalence and anatomy of retroesophageal MAPCAs in patients undergoing repair of pulmonary atresia/ventricular septal defect/MAPCAs.**

Methods. **This was a concurrent analysis of 68 consecutive patients (March 2013 through October 2015) undergoing a primary surgical procedure for pulmonary atresia/ventricular septal defect/MAPCAs. A detailed analysis of the MAPCA anatomy was made intraoperatively for each patient, including the total number of MAPCAs to each lung and the presence or absence of a retroesophageal course. These data were correlated with the preoperative cardiac catheterization images.**

Results. **A retroesophageal MAPCA was identified during the operation in 45 of the 68 patients (67%), all of which were located on the side opposite the arch. For the 36 patients with a left aortic arch, 77% had a retroesophageal MAPCA compared with 53% of patients with a right arch. Forty-six percent of retroesophageal MAPCAs coursed within the muscular fibers of the esophagus (intraesophageal) and were more common to the left lung than the right (72% vs 32%). A midsegment stenosis was present in 84% of the retroesophageal MAPCAs, and this was more severe when the MAPCAs were intraesophageal than when they were not (80% vs 42%).**

Conclusions. **These data demonstrate that two-thirds of patients had a retroesophageal MAPCA and that there were significant differences in prevalence and anatomy depending on the side of the aortic arch. These data provide important insights into the origin and course of retroesophageal MAPCAs.**

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Major aortopulmonary collateral arteries (MAPCAs) are the sole source of pulmonary blood flow in patients with pulmonary atresia (PA) and an absent ductus arteriosus [1, 2]. The anatomy of MAPCAs can be highly variable, both in the number of MAPCAs to each lung and the anatomic origin and course of the MAPCAs [3–5]. This anatomic variability becomes clinically relevant when MAPCAs are used in the reconstruction of a pulmonary vascular bed [6, 7]. Many groups have adopted the "unifocalization" of MAPCAs for the treatment of patients with pulmonary atresia with ventricular septal defect (VSD) and MAPCAs (PA/VSD/MAPCAs) [8–14].

Our surgical group has gained an extensive experience with the identification, dissection, and unifocalization of MAPCAs. We observed that some MAPCAs coursed behind the esophagus before entering the lung parenchyma. These "retroesophageal" MAPCAs always course to the lung contralateral to the side of the aortic arch, as dictated by the anatomic relationship between the descending aorta and esophagus (Fig 1). We also observed that retroesophageal MAPCAs originate from the lateral side of the aorta facing the esophagus before taking a posterior route. This lateral origin and posterior course contrast to most MAPCAs, which originate from the anterior surface of the aorta and take an anterior route to reach the lung. As a consequence, some knowledge is required of retroesophageal MAPCAs anatomy to facilitate identification and surgical technique for dissection of these MAPCAs for unifocalization.

Although we were aware of the presence of retroesophageal MAPCAs for many years, it was also evident that very little was known about the prevalence or anatomy of retroesophageal MAPCAs. The purpose of this study was to evaluate the prevalence and anatomy of retroesophageal MAPCAs.

Material and Methods

The Stanford University Institutional Review Board approved this study. Patients eligible for inclusion in the study were those undergoing a primary surgical procedure for PA/VSD/MAPCAs. The study summarizes our experience with 68 consecutive patients undergoing PA/VSD/MAPCA operations from March 2013 through October 2015.

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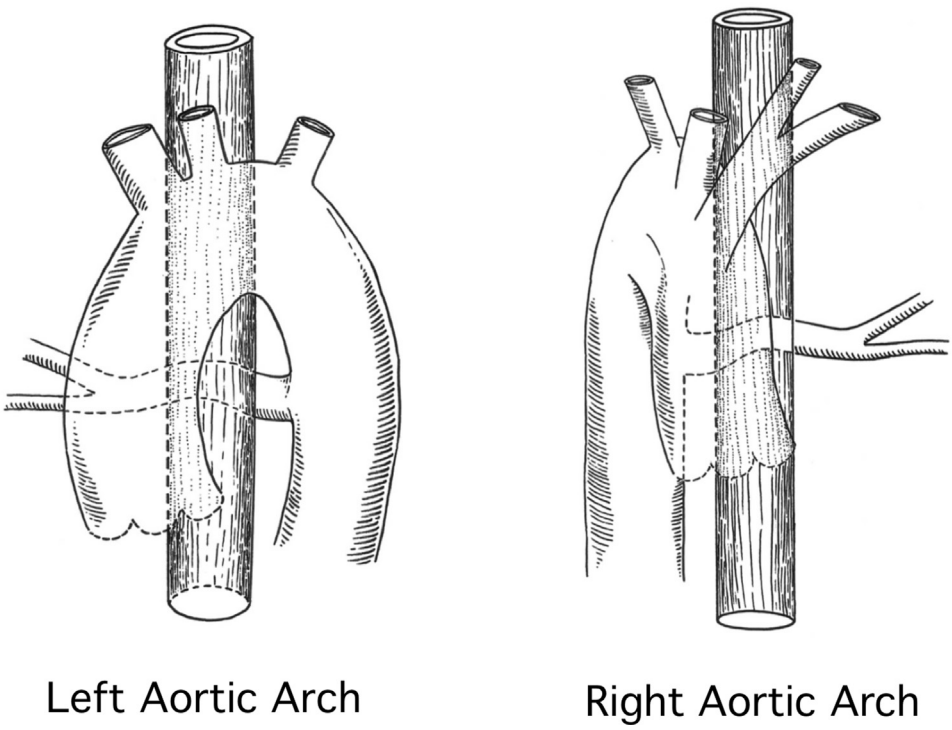

Fig 1. Artist's illustration shows the anatomic relationships between the side of the aortic arch and the position of the esophagus. This anatomy dictates that retroesophageal major aortopulmonary collateral arteries (MAPCAs) occur only to the lung contralateral to the side of the arch. In the current study, 36 patients had a left aortic arch and 32 had a right aortic arch. The origin of retroesophageal MAPCAs is on the lateral side of the aorta facing the esophagus.

We perform the entire MAPCA dissection before cardiopulmonary bypass is initiated. This dissection is performed between the ascending aorta and superior vena cava and inferior to the tracheal carina. The descending aorta is located, and the origins and course of the MAPCAs are identified. For patients with a left-sided aortic arch, the right lateral side of the aorta is also inspected for the presence of retroesophageal MAPCAs (Fig 2A). Conversely, patients with a right-sided aortic arch undergo inspection of the left lateral surface of the aorta (Fig 2B).

When a retroesophageal MAPCA is identified, the MAPCA is dissected on both sides of the esophagus until it is completely mobilized. Removing the transesophageal echocardiography probe during the dissection of the MAPCAs is essential. The retroesophageal MAPCAs in some patients traverse through some of the esophageal muscle fibers (intraesophageal), and in this circumstance, the fibers are divided to free the retroesophageal MAPCA.

Many of the retroesophageal MAPCAs had evidence of stenoses at the point where the MAPCA crossed behind the esophagus. This observation can be visualized on the preoperative angiogram (Fig 3) and subsequently confirmed during the operation when the MAPCAs are probed and surgically opened for inspection.

A detailed analysis of the MAPCA anatomy was performed intraoperatively for all patients. This included (1) the total number of MAPCAs to each lung, (2) the presence or absence of a retroesophageal course, (3) the presence or absence of an intraesophageal course, (4) the degree of stenosis for the retroesophageal MAPCAs, and (5) segmental distribution of blood flow derived from retroesophageal MAPCAs. This information was documented concurrently and was subsequently correlated with the preoperative cardiac catheterization images.

There were 39 boys and 29 girls, and 25 patients had deletion of chromosome 22q11. At the time of the operation, the patients were a median age of 4.4 months (range, 1 to 62 months) and a median weight of 5.1 kg (range, 2.3 to 21.8 kg).

Fifty-two patients (77%) underwent a single-stage complete repair, including unifocalization of MAPCAs, closure of the VSD, and placement of a conduit from the right ventricle to reconstructed pulmonary arteries. Unifocalization of MAPCAs and a central shunt was performed in 12 patients (16%), and an aortopulmonary window was created in 4 patients (7%) [15].

Results are reported as the mean ± standard error. Statistical comparison of groups was performed with analysis of variance.

Results

A retroesophageal MAPCA was diagnosed in 45 of the 68 patients (67%) intraoperatively. However, the prevalence of retroesophageal MAPCAs differed significantly depending on whether there was a left or right aortic arch (Fig 4). The prevalence of retroesophageal MAPCAs was 77% for the 36 patients who had a left aortic arch vs 53% in the 32 patients who had a right aortic arch ($p < 0.05$).

Of the 45 patients with retroesophageal MAPCAs, 41 (91%) had 1 retroesophageal MAPCA, and 4 had 2 retroesophageal MAPCAs (Fig 5). Of the patients with