INTRODUCTION

In patients with complex congenital heart disease (e.g., tetralogy of Fallot with pulmonary atresia), major aortopulmonary collateral arteries (MAPCAs) are well described.[1] Isolated MAPCAs without intracardiac defects appear with increased incidence in very low birth infants with chronic lung disease.[2] Isolated MAPCAs with no associated congenital cardiac disease supplying a completely functional lung are a rare entity. In contrast, in pulmonary sequestration, sequestered lung is typically supplied by a systemic artery with drainage to the pulmonary venous system, however, there is a lack of normal connection with the tracheo-bronchial tree.[3] In addition, patients with isolated MAPCAs can present with congestive heart failure, recurrent pulmonary infections, and hemoptysis requiring embolization. We describe a very rare presentation of single aortopulmonary collateral supplying the entire left lower lobe with normal tracheobronchial connection and no pulmonary arterial supply in an otherwise healthy child.

CASE REPORT

An 18-month-old female born at term with no significant past medical history and asymptomatic was referred to us for evaluation of a murmur. The pregnancy was uncomplicated, and she delivers spontaneously. On initial examination, the baby had a Grade 2/6, harsh continuous murmur most predominantly heard in the left infrascapular area. Echocardiogram showed pulmonary flow to the lower lobe, and she was discharged home on postoperative day 2 without complications.
holodiastolic reversal of flow in the descending aorta, and a large collateral artery arising from the descending aorta measuring 6 mm. She was taken for cardiac catheterization to assess hemodynamics and possibly close the collateral. Cardiac catheterization showed normal cardiac index and filling pressures. A large torturous collateral artery was seen arising from the descending aorta and supplying the entire left lower lobe, draining through the left inferior pulmonary vein into the left atrium. There was no pulmonary arterial connection to the left lower lobe [Figure 1]. A chest computed tomography (CT) with virtual bronchoscopy appeared to demonstrate a normal bronchial connection of the lower lobe which was confirmed with bronchoscopy before starting the case. Given the unique findings of an entirely functional lobe being supplied by one MAPCA and evidence of restriction with audible murmur, our multidisciplinary team planned to preserve the left lower lobe by anastomosing the collateral artery to the pulmonary circulation.

On the day of surgery, the patient was positioned for a left-sided, muscle-sparing thoracotomy with cardiopulmonary bypass on stand-by. Once the collateral artery was dissected and its course within the pleural space appreciated, the main left pulmonary artery was isolated. The original intent was direct anastomosis, however intraoperatively, we felt that a direct anastomosis would be under tension and might cause unnatural kink in the arterial flow. The course of the collateral through the parenchyma of the left lower lobe prevented enough mobilization to allow it to be directly connected to the main left pulmonary artery. Therefore, we anastomosed an 8 mm ringed conduit from the left pulmonary artery, coursing anteriorly to the proximal aspect of the collateral artery [Figure 2]. The most proximal aspect of the collateral was then clipped and tied off from the aorta without transecting the artery. The patient tolerated the surgery well. Postoperatively, saturations were >98% suggesting no ventilation-perfusion mismatch and echocardiogram showed unobstructed flow in left pulmonary artery. She was started on aspirin to prevent stenosis/graft thrombosis and discharged home on postoperative day 2 without complications.

**DISCUSSION**

This case presented us with a unique situation of an entire, functional, lobe of the left lung being perfused by a systemic collateral artery. The continuity of the functional lobe with the bronchial tree in a way mirroring MAPCAs suggesting “unifocalization” might be a durable solution. Preoperative CT suggested bronchial continuity with the trachea but was unable to definitively confirm. Hence, in a multidisciplinary meeting, it was felt that the most important feature to preserve the lobe with an anastomosis to the pulmonary arterial circulation was bronchoscopic confirmation of a normal lung ventilation. Once a normal airway was confirmed, the most important decision was between ligating the collateral from the aorta with direct anastomosis to the left pulmonary artery versus a conduit between the two. Intraoperatively, the course of the collateral prevented enough mobilization and decision was made to use a conduit. After selecting the largest reasonable conduit, the final lay appeared satisfactory with adequate perfusion and potential to accommodate future somatic growth. While there are likely several ways in which this congenital anatomy could have been successfully addressed, we feel that our approach treated the patient’s anatomy and volume overload in a manner which will hopefully endure long into adulthood. Follow-up in few months with lung perfusion scan and CT angiogram may be required to access the health and patency of the graft.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
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Conflicts of interest
There are no conflicts of interest.

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