A rare case report: Tetralogy of Fallot, right aortic arch, isolated left subclavian from patent ductus arteriosus, neonatal aortopulmonary window, and hypoplastic right pulmonary artery

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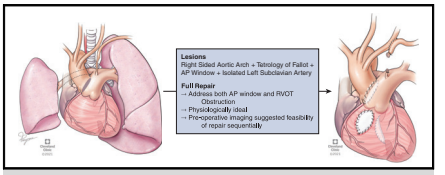


Patient with a complex series of defects underwent complete surgical repair successfully.

CENTRAL MESSAGE

Complete repair of unique conotruncal and aortic arch defect can be achieved by using a virtual image-based platform to facilitate preoperative planning and coordinated multidisciplinary team effort.

See Commentaries on pages 195 and 196.

▶ Video clip is available online.

Tetralogy of Fallot (TOF) accounts for 7% to 10% of congenital heart disease. Right-sided aortic arch (RAA) is frequently seen in TOF, in as many as 25% of cases.[1,2] Aortopulmonary window (APW), defined by a communication between the aorta and the pulmonary artery (PA) with 2 separate semilunar valves, is exceedingly rare, accounting for 0.1% to 0.2% of congenital heart disease.[3] We present a patient with a never-before characterized set of lesions including TOF with RAA, APW, and an isolated left subclavian artery. The case discussed herein was even more complex, as the patient had a hypoplastic right PA and right lung, as well as an imperforate anus, bifid scrotum, and horseshoe kidney consistent with VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) diagnosis. We describe the preoperative planning and surgical management of this uncharacterized set of anomalies (Figure 1). The patient's guardians provided informed written consent for the publication of the study data.

A 3-week-old ex-37-week male (3.4 kg, body surface area 0.2 m^2) presented with increased work of breathing, requiring continuous positive airway pressure to maintain oxygen saturations greater than 90%. Transthoracic echocardiography revealed an anteriorly displaced conal septum, large unrestrictive perimembranous ventricular septal defect with inlet extension, right ventricular hypertrophy, and right ventricular outflow tract obstruction (RVOT) consistent with TOF. The PA z score was –0.49. An 8-mm APW was seen between the aorta and distal main PA, with significant hypoplasia of the right PA and right lung, exacerbating his tenuous respiratory status. Three-dimensional reconstruction of computed tomography imaging provided detailed anatomic definition and spatial relationships of these complex lesions. The RAA was confirmed with an isolated subclavian artery arising from the left PA through a patent ductus arteriosus without evidence of any aortopulmonary collaterals (Figure 2, *A-D*). The right lung hypoplasia shifted the heart posterolaterally, complicating surgical exposure for full repair. However, virtual heart mapping using EchoPixel (EchoPixel) improved understanding of the spatial relationships and topography, particularly regarding accessibility of the right atrium and ventricle, given the extreme rightward displacement of the heart, and the extent of the APW to the arch vessels for

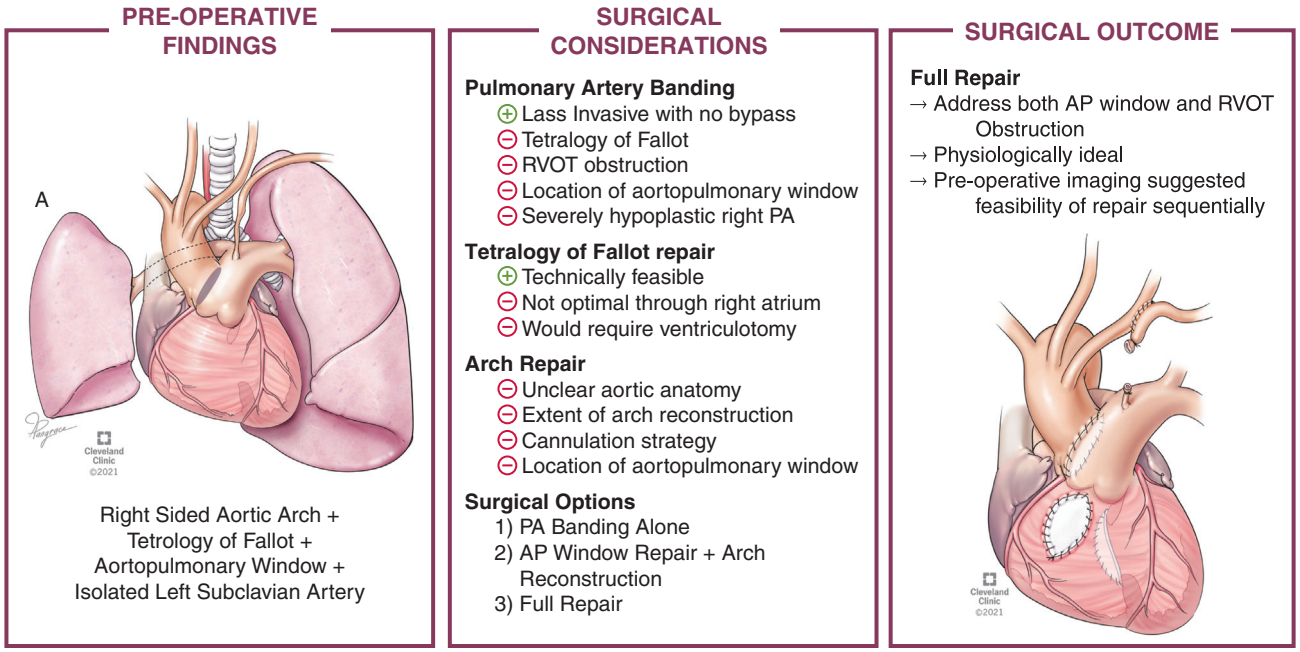


FIGURE 1. Surgical considerations and decision-making. Based on the preoperative anatomy, surgical considerations were discussed during multidisciplinary conference. The benefits and disadvantages of each repair strategy were thoroughly explored before decision. The team consensus was to proceed with full repair as it was physiologically ideal, surgically feasible, and addressed all lesions. *RVOT*, Right ventricular outflow tract obstruction; *PA*, pulmonary artery; *AP*, aortopulmonary. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography ©2022. All rights reserved.

crossclamping and cardioplegia. The surgical team was able to choreograph surgical sequences necessary for complete repair compared to palliation, which prompted multidisciplinary consensus for complete repair.

OPERATIVE PROCEDURE

Median sternotomy and dissection of mediastinal structures were performed on our 3-week-old (3.4-kg) male patient. The patent ductus arteriosus was ligated and divided. The distal ascending aorta was cannulated, bicaval cannulation was performed, and cardiopulmonary bypass was instituted, cooling to 32° C. The right and left PAs were controlled using clips, and a left atrial vent was placed. The APW was identified and dissected. Aortic crossclamp was placed above the APW, and diastolic arrest was achieved using single-dose anterograde del Nido cardioplegia. An aortotomy was made, providing visualization of the APW and branch PAs. A Gore-Tex patch (W. L. Gore & Associates, Inc) was sutured to the posterior margins of the APW and incorporated with the PA and aorta anteriorly so that the patch was sandwiched in between (Figure 3, *A* and *B*). A limited infundibulotomy was performed, and obstructing muscle bundles in the RVOT were sharply dissected. Then, the ventricular septal defect was repaired with a Gore-Tex patch transatrially, and the infundibulotomy was closed with a transannular patch. Crossclamp was then released, allowing coronary reperfusion. Afterwards, the isolated subclavian artery was identified and reimplanted with a side-to-side anastomosis to the left common carotid. By this time, the heart was back in normal sinus rhythm. The patient was weaned off cardiopulmonary bypass with excellent hemodynamics. Postoperative echocardiography demonstrated excellent repair of TOF, good biventricular function, and no intracardiac or extracardiac shunts. Cardiopulmonary bypass and crossclamp time were 87 and 59 minutes, respectively. The patient was extubated on postoperative day 2 and, by postoperative day 7, was tolerating nasogastric feeds and discharged (Video 1).

DISCUSSION

We describe the management of a patient with a unique constellation of intracardiac and extracardiac defects using virtual-based imaging to inform surgical decision-making. In our patient, extreme rotation of the heart, and hypoplastic right PA further complicated these decisions. Specifically unclear arch anatomy, difficult approach to TOF lesions, and tenuous respiratory status with concern for a reactive pulmonary vascular bed were important considerations. Through the use of virtual software, spatial anatomic relationships became clearly defined, specifically the RAA, RVOT, and isolated subclavian artery, allowing the team to conceptualize several surgical approaches. Comprehensive review demonstrated the challenges of a palliative versus definitive repair strategy. Although PA banding and TOF repair with arch reconstruction were considered, we concluded that a complete repair was technically feasible,

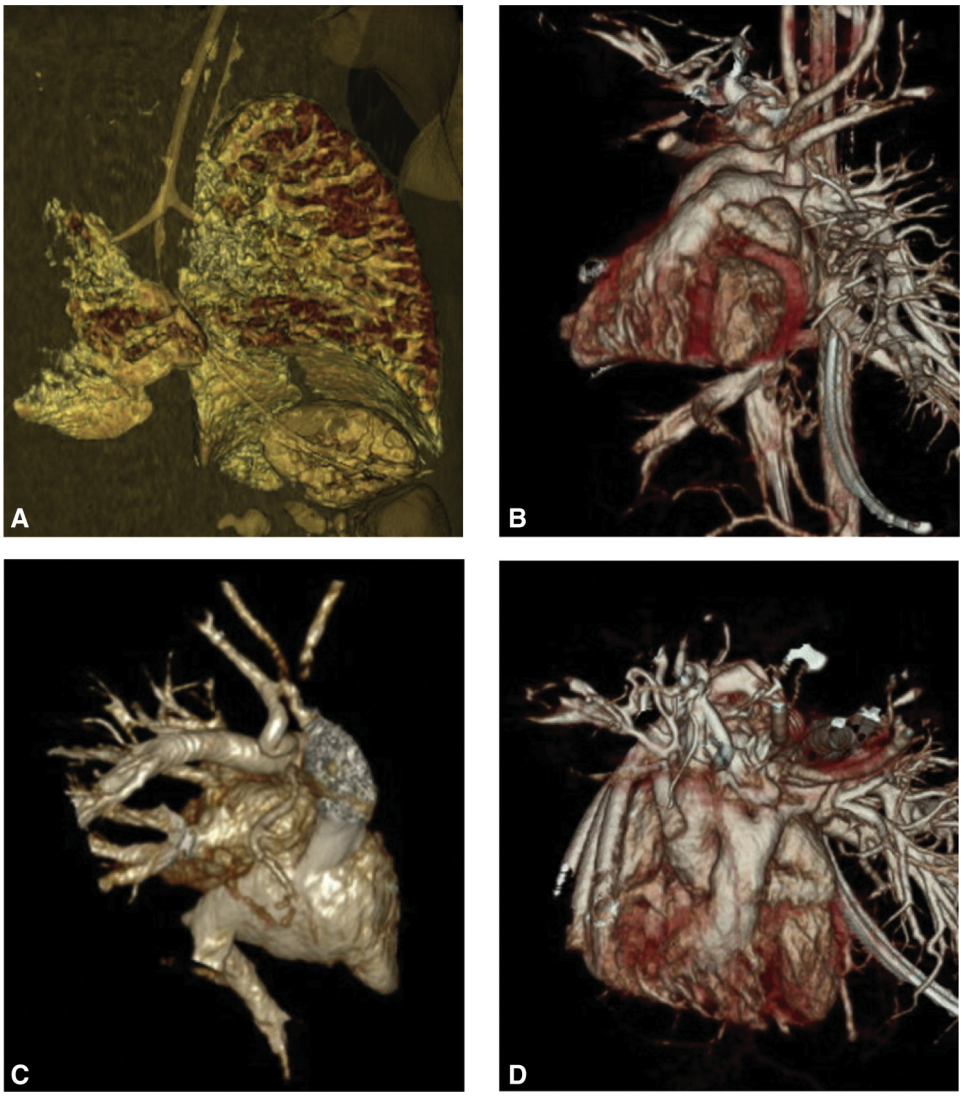


FIGURE 2. Three-dimensional (3D) computed tomography reconstruction. 3D reconstruction of computed tomography provided visualization of (A) hypoplasia of the right lung, as well as (B-D) right-sided aortic arch, isolated left subclavian artery arising from left pulmonary artery, and spatial relationships of surrounding structures.

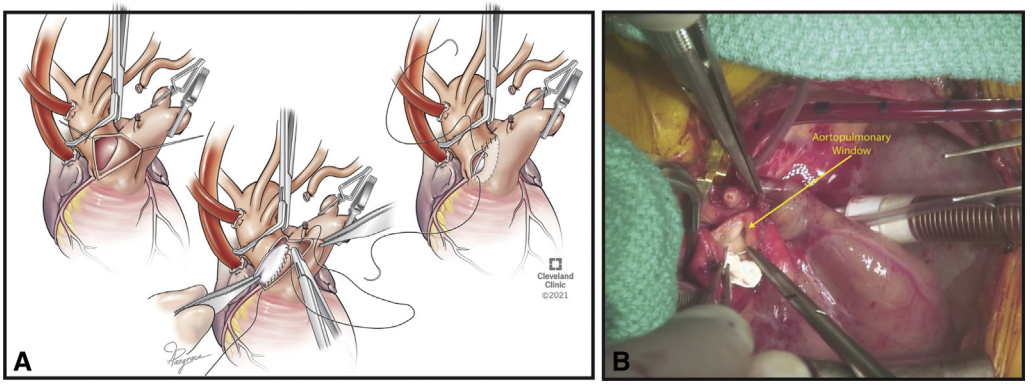
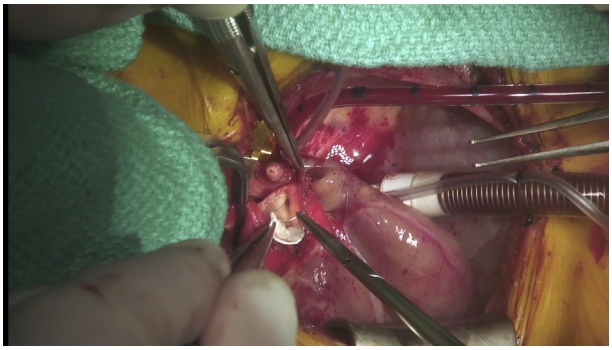


FIGURE 3. Closure of aortopulmonary window. A, Anterior aortotomy was made to provide visualization of the aortopulmonary window. The defect was closed with a Gore-Tex patch using a "sandwich" technique by suturing the margins of the APW posteriorly and incorporating the pulmonary artery and aorta anteriorly. Then the aortotomy was closed. B, Intraoperative view of the aortopulmonary window and patch placement. Reprinted with permission, Cleveland Clinic Center for Medical Art & Photography ©2022. All rights reserved.



VIDEO 1. Case presentation and operative technique. This video abstract details the patient's initial presentation, the associated pathologies, preoperative imaging, surgical considerations, and the operative technique to address the described lesions. Video available at: https://www.jtcvs.org/article/S2666-2507(22)00205-X/fulltext.

and physiologically preferable to partial repair, specifically given the hypoplastic right PA and lung, location of the APW, RVOT, and arch anatomy. The sequence of complete repair was facilitated by virtual rehearsals informed by the preoperative evaluation. Despite extensive comorbidities and uniquely complex anatomy, the patient's successful course can be attributed to this approach—virtual computed tomography imaging technology, surgical planning, and multidisciplinary management.

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