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Late-Presenting Asymptomatic Balanced Double Aortic Arch Associated With Tetralogy of Fallot: Definitive Surgical Correction

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Abstract

Asymptomatic balanced double aortic arch (DAA) is rarely associated with tetralogy of Fallot (TOF). Balanced DAA may present with symptoms caused by tracheal or esophageal compression. However, the lack of consensus on the management of asymptomatic DAA poses a dilemma regarding the need for surgical intervention. We report a case of asymptomatic balanced DAA associated with TOF in an eight-year-old boy who presented with exercise intolerance, fatigue, and increasing cyanosis. Doppler showed severe right ventricular outflow tract obstruction. Intracardiac repair proceeded without ring division. On postoperative day 15, echocardiography showed a peak outflow tract gradient of 28 mmHg and free pulmonary regurgitation.

Categories: Radiology, Cardiology, Cardiac/Thoracic/Vascular Surgery
Keywords: congenital cardiovascular malformation, congenital heart disease, double aortic arch, intra-cardiac repair, tetralogy of fallot, vascular ring

Introduction

Double aortic arch (DAA) is a congenital aortic arch anomaly that forms a complete vascular ring around the trachea and esophagus [1]. Depending on the relative dimensions of the two arches, DAA may be right-dominant, left-dominant, or balanced; right-sided dominance is the most common pattern [1]. Compression of the trachea or esophagus may produce respiratory or feeding-related symptoms [1,2].

Asymptomatic DAA refers to the absence of clinical manifestations attributable to the vascular ring and may be detected incidentally during imaging for another cardiac abnormality [2,3]. DAA may occur in isolation or with associated congenital cardiac defects, including tetralogy of Fallot (TOF) [1,3]. Although surgical treatment is established for symptomatic DAA, evidence guiding the management of asymptomatic vascular rings remains limited, and clinical observation with follow-up has been described [1,2]. Published reports of asymptomatic DAA associated with TOF are uncommon [3]. We describe an eight-year-old boy with asymptomatic balanced DAA and TOF who underwent intracardiac repair of TOF without division of the vascular ring.

Case Presentation

Ethical approval for this study was granted by the Institutional Ethics Committee (IEC-2) of the HM Patel Centre for Medical Care and Education, Anand (Approval No: IEC/BU/2021/Cr.08/273/2026) dated 01/08/2026.

An eight-year-old male patient diagnosed with TOF was referred to our department because of complaints of progressive exercise intolerance, fatigue, and increasing cyanosis. He had a history of dyspnea on exertion (grade IV). Clinical examination showed deep cyanosis with an SpO_2 of 78% and clubbing (grade III). Cardiac auscultation revealed an ejection systolic murmur over the pulmonary area, with no continuous murmurs detected. Imaging revealed bilateral aortic knob indentation, consistent with DAA (Figure *1*).

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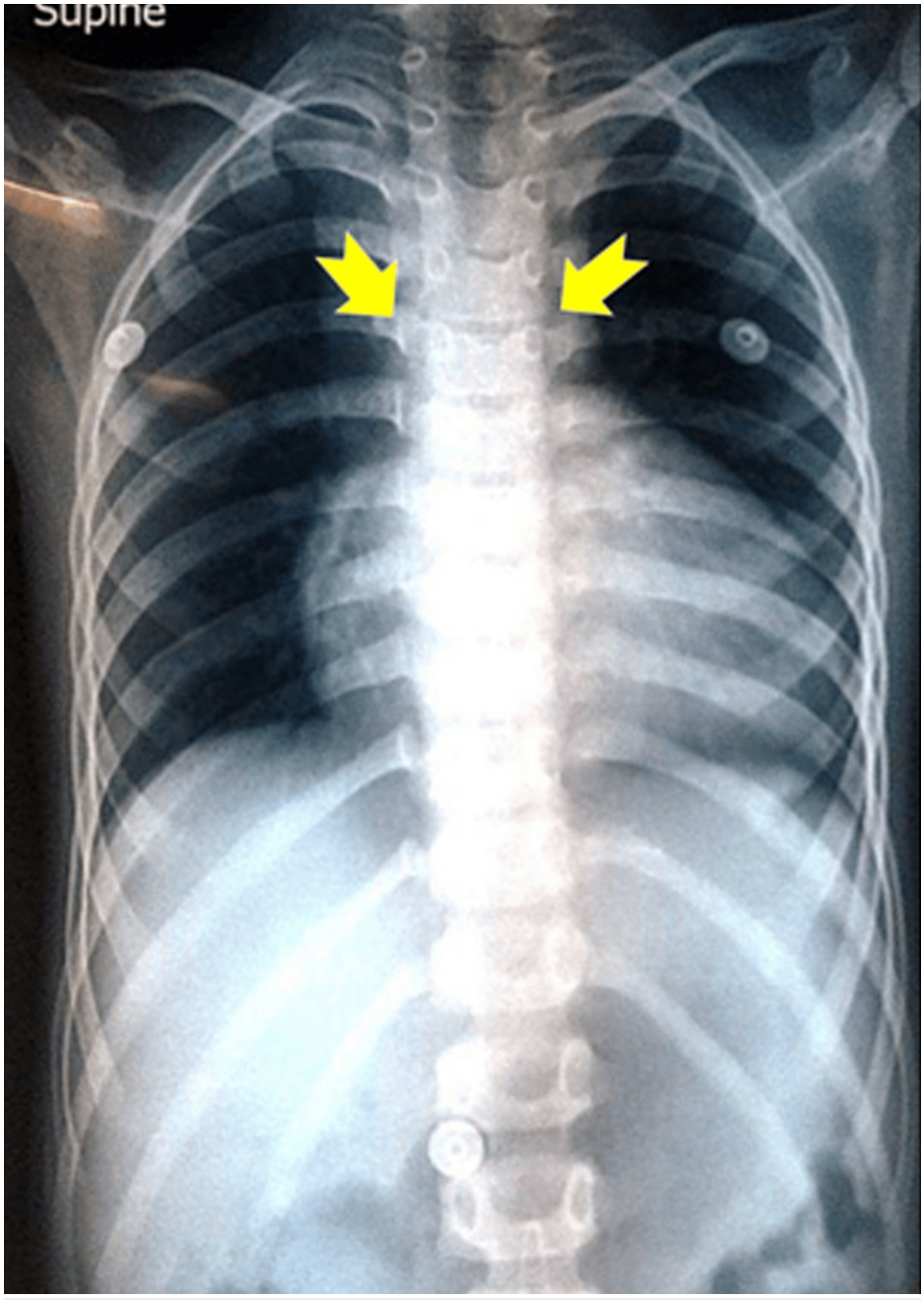


FIGURE 1: Pre-operative plain radiograph of the chest in supine (AP view), showing double aortic knuckles (aortic knobs; yellow arrows)

Image Credit: Dr. Deepakkumar V. Mehta

Preoperative evaluation with 2-D echocardiography showed TOF with a small ventricular septal defect along with supra-systemic right ventricular pressure. The subaortic ventricular septal defect was small and shunting from right to left. Doppler echocardiography indicated severe multilevel right ventricular outflow tract obstruction, with a reported peak gradient of approximately 80 mmHg. The branches of pulmonary arteries were confluent and of adequate size, and the left ventricle was of adequate size. The types and sizes of the arches were not clearly discernible on the 2-D echocardiography.

Cardiac computed tomography angiography showed an overriding aorta, mild-to-moderate subvalvar narrowing (right ventricular outflow tract diameter, 9.6 mm), and mild valvar narrowing (maximum diameter at the valve, 10.4 mm). These CT measurements described the anatomic caliber, whereas Doppler estimated the hemodynamic severity of the multilevel obstruction. Dynamic infundibular narrowing may have contributed to the higher Doppler estimate. A co-dominant/balanced DAA (aortic ring) with a single ascending and descending aorta was identified in the right paramedian position. We also noted fairly large

main right and left pulmonary arteries. The Nakata index was 177.32 mm²/m² [4], and the McGoon ratio was 1.181 [5]. The ascending aorta divided into right and left arches of equal sizes at the T3-T4 intervertebral disc level, which ascended up to the T2 vertebral level and then passed posteriorly quite above the left and right main bronchi, respectively. The right and left aortic arches give rise to their corresponding common carotid and subclavian arteries. Both aortic arches again joined each other at the T3-T4 intervertebral disc level, forming the descending thoracic aorta in the right paramedian position, which slightly turned toward the left side and lay in the midline at the diaphragmatic hiatus level. There were no aortopulmonary collaterals found (Figures *2*, *3*).

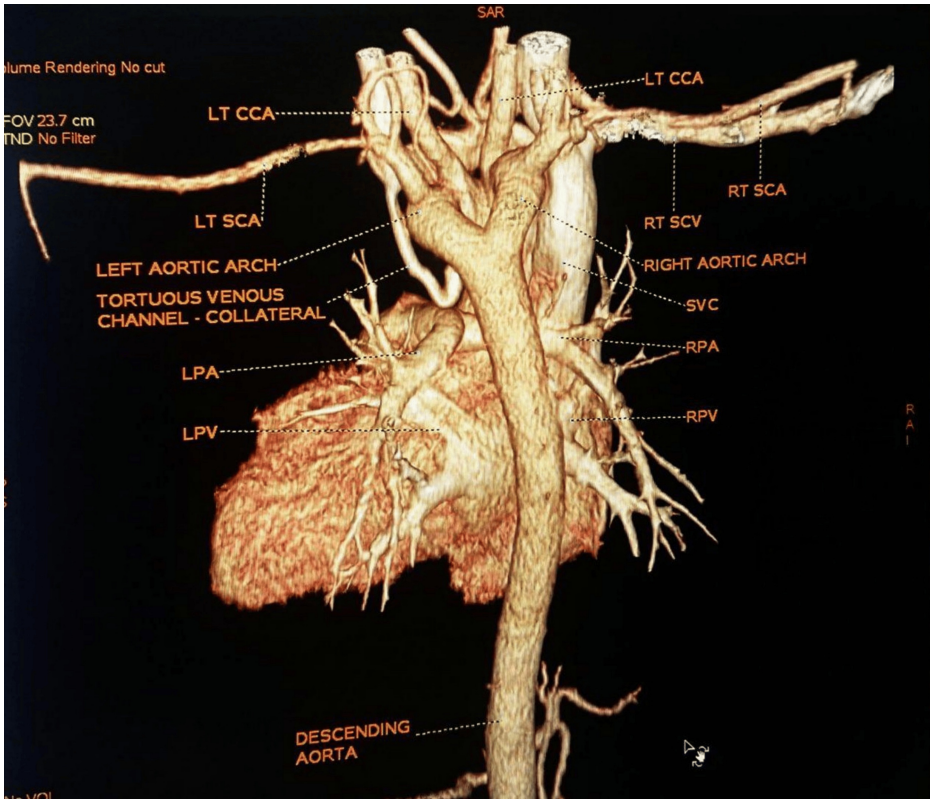


FIGURE 2: Volume rendering (VR) image: posterior view of the heart, showing double aortic arch

CCA, common carotid artery; SCA, subclavian artery; LPA, left pulmonary artery; LPV, left pulmonary vein; RPV, right pulmonary vein; RPA, right pulmonary artery; SVC, superior vena cava; SCV, subclavian vein; Rt., right; Lt., left; SAR, specific absorption rate.

Image Credit: Dr. Deepakkumar V. Mehta

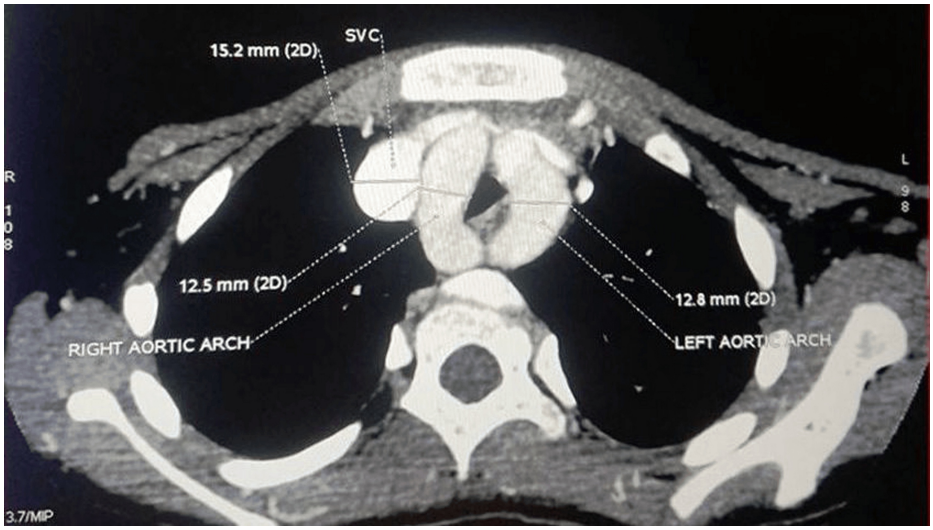


FIGURE 3: Contrast-enhanced axial cardiac CT image demonstrating a double aortic arch

SVC, superior vena cava; 2D, 2 dimensional

Image Credit: Dr. Deepakkumar V. Mehta

Surgical procedure

Intra-cardiac repair and right ventricular outflow tract reconstruction were performed using a 0.1-mm polytetrafluoroethylene (PTFE) patch with a PTFE monocusp valve and a trans-annular SYNKROSCAFF bovine pericardial patch (SynkroMax Biotech Pvt. Ltd., Chennai, India). The ventricular and atrial septal defects were closed with glutaraldehyde-treated pericardial patches.

Intracardiac repair was prioritized because of progressive cyanosis and severe right ventricular outflow tract obstruction. The history included exertional dyspnea but did not identify symptoms specifically attributable to tracheoesophageal compression. After induction of anesthesia, flexible bronchoscopy showed no stenosis or bronchomalacia in either main bronchus. CT demonstrated two patent arches of similar caliber (right, 12.5 mm; left, 12.8 mm). Given the clinical history and bronchoscopic findings, the left arch was mobilized and marked, and the vascular ring was left undivided.

The patient received mechanical ventilation for 72 hours and was discharged after a 16-day hospital stay. Echocardiography on postoperative day 15 showed no residual ventricular or atrial septal defect, a peak right ventricular outflow tract gradient of 28 mmHg, free pulmonary regurgitation, mild-to-moderate tricuspid regurgitation, and preserved biventricular systolic function as described in the report. The available records do not establish a reliable postoperative oxygen saturation or provide a structured assessment of respiratory or gastrointestinal symptoms after surgery. No later outpatient assessment was available for this report (Table *1*).

Sr. No.	Parameters	Patient Values
01	Patient's age at intra-cardiac repair	8 years
02	Patient's weight at intra-cardiac repair	17 kg
03	Length of hospital stay	16 days
04	Length of stay in the cardiac surgical intensive care unit (CSICU)	10 days
05	Mechanical ventilatory support time	72 h
06	CPB time	233 min
07	ACC time	173 min

TABLE 1: Intraoperative and Postoperative Demographic and Clinical Data

ACC, aortic cross-clamp time; CPB, cardiopulmonary bypass

Discussion

A balanced DAA forms a complete vascular ring that may compress the trachea or esophagus. Moreover, TOF as a cyanotic CHD is occasionally accompanied by symptomatic DAA. When symptomatic DAAs coexist with intracardiac defects, the two defects can be fixed in a single stage at the same time [6] or managed in stages. Talwar et al. had previously described the repair of the DAA with a posterolateral thoracotomy and cardiac malformation in a similar scenario [6]. It has been reported that TOF with DAA can be repaired in stages using a median sternotomy followed by posterolateral thoracotomy later as a follow-up procedure [7].

TOF rarely coexists with asymptomatic DAA [3]. On the other hand, the scarcity of data in the literature about the symptomatic and asymptomatic vascular rings raises concerns about the ideal management for these patients.

Tracheal or esophageal compression causing symptoms is an established indication for ring division [1]. Observation has been described in patients with asymptomatic vascular rings [2], and some patients may develop symptoms later [8]. In this case, the available records describe the early inpatient course and leave the longer-term respiratory and swallowing outcomes uncertain.

Bhende et al. described computed tomography-based assessment of pulmonary vascular indices and tailored PTFE monocusp preparation in TOF repair [9]. These considerations are relevant to the preoperative anatomical assessment and right ventricular outflow tract reconstruction in our patient; however, that report does not directly address the management of associated asymptomatic DAA.

Conclusions

This case describes the early inpatient course after intracardiac repair of TOF with right ventricular outflow tract reconstruction in a child whose balanced DAA was left undivided. With follow-up limited to the index admission, the long-term course of the unrepaired vascular ring and the applicability of this approach to other patients remain uncertain.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

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Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. Institutional Ethics Committee (IEC-2), HM Patel Centre for Medical Care and Education, Anand issued approval IEC/BU/2021/Cr.08/273/2026 dated August 01, 2026. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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