Case Report

Pulsatile Flow in Descending Aorta: Can Coarctation of Aorta be Ruled out by Transesophageal Echocardiography?

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ABSTRACT

A 5-year-old child with L posed great arteries, large subpulmonic ventricular septal defect (VSD), atrial septal defect (ASD), and a large patent ductus arteriosus (PDA) with mild isthmic narrowing was scheduled for surgical correction. Intraoperatively, it was a case of anatomically corrected malposition of great arteries. Due to abnormal positioning of great vessels, the isthmus was ligated instead of the large PDA. The postoperative transesophageal echocardiography showed pulsatile flow in descending aorta as it was being filled by large PDA, and thus iatrogenic coarctation (CoA) was missed. It was detected in the intensive care unit due to the onset of acidosis on blood gas analysis and the presence of gradient between radial and femoral arterial line pressures. The patient was taken for redo surgery, the PDA was then ligated, resection of the isthmic narrowing and repair by end-to-end anastomosis was done.

Keywords: Coarctation of aorta, malposition of great arteries, patent ductus arteriosus

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Submitted: 15-Jan-2019 **Revised:** 25-Aug-2019 **Accepted:** 28-Oct-2019 **Published:** 19-Apr-2021

INTRODUCTION

Anatomically corrected malposition of the great arteries (ACMGA) is a rare congenital cardiac anomaly. In a heart with ACMGA, both great arteries arise from the appropriate ventricles but are abnormally related to each other and to the ventricles.[1] It can be misdiagnosed as transposition of great arteries.[2]

CASE REPORT

A 5-year-old girl was posted for ventricular septal defect (VSD) and atrial septal defect (ASD) closure with patent ductus arteriosus (PDA) ligation; her preoperative echocardiography showed large subpulmonic VSD, small ostium secundum ASD, large PDA with left to right shunt with mild Coarctation of aorta, severe pulmonary arterial hypertension, with L-posed great arteries, and normal biventricular function. The patient had presented with dyspnea on exertion and recurrent respiratory tract infections. On examination, all peripheral pulses were felt with a rate of 98/min and was regular. A pansystolic murmur of grade III was heard at the left sternal border. After induction, right radial artery was cannulated with 22-gauge cannula and right internal jugular vein with 5.5 Fr triple lumen catheter.

Intraoperatively, it was a rare complex cardiac lesion known as anatomically corrected malposition of great arteries with a large subpulmonic VSD, isthmic narrowing, and a large PDA. VSD closure was done with Dacron

Access this article online	
Quick Response Code:	Website: www.annals.in
	DOI: 10.4103/aca.ACA_8_19

How to cite this article: Diwakar A, Chalam KS, Hiremath CS, Dash PK. Pulsatile flow in descending aorta: Can coarctation of aorta be ruled out by transesophageal echocardiography? Ann Card Anaesth 2021;24:253-5.

patch and ASD by direct closure. PDA ligation was done which also addressed the isthmic narrowing. The patient was successfully weaned from cardiopulmonary bypass. Postoperative transesophageal echocardiography showed good biventricular function and the descending aorta showed good pulsatile flow, no turbulence, and no gradient, indicating no Coarctation of aorta. Figure 1.

In the postoperative intensive care unit, there was acidosis on blood gas analysis, and femoral pulses were feeble as compared to radial; hence, the femoral arterial line was placed which showed good pulsatile trace but the pressures were 40/16 mmHg and the radial pressures were 70/36 mmHg. Immediately patient was taken to the operation theatre with suspicion of iatrogenic COA. Due to abnormal positioning of great vessels, the large PDA appeared to be left pulmonary artery and isthmus as PDA which was ligated [Figure 2]. The PDA was then ligated and repair of the isthmic narrowing was done by resection and end-to-end anastomosis by thoracotomy.

DISCUSSION

There are four types of ACMGA: Type 1 (S, D, L) is situs solitus, d-loop ventricles, aorta being left and anterior; Type 2 (S, L, D) is situs solitus, l-loop ventricles, with aorta right and anterior; Type 3 (I, L, D) is situs inversus, l-loop ventricles, aorta is right and anterior; and Type 4 (I, D, L) is situs inversus, d-loop ventricles, with left and anterior aorta. The anomalies associated with ACMGA are VSD, obstruction of right ventricular outflow tract (RVOT), subaortic obstruction, hypoplasia of right ventricle, juxtaposition of atrial appendage, and right aortic arch.[1,3] In patients with situs solitus and AV concordance (S, D, L), the surgical results have been good with 92% survival. The presence of AV discordance or hypoplastic right heart structures, or both conditions makes the outcome after palliative procedure to be poor with only 29% survival.[4]

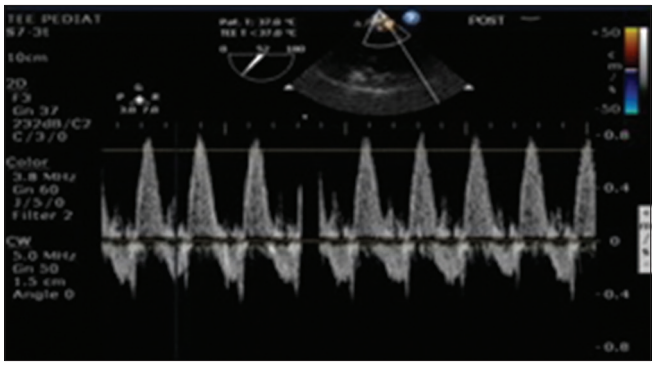

Figure 1: TEE Image showing pulsatile laminar flow in descending aorta

As seen in the Figure 2, the aorta is parallel and lateral to the pulmonary artery making visualization of the arch difficult through the midline sternal opening. The large PDA which continued as the descending aorta was mistaken for left pulmonary artery and the isthmic narrowing appeared to be a PDA and was, thus, inadvertently ligated.

CoA examination by Transesophageal echocardiography is difficult as examination of the distal ascending aorta and proximal aortic arch is limited by the anterior position of the air-filled trachea relative to the oesophagus. However turbulent, eccentric jets or flow acceleration in the descending aorta by colour-flow Doppler can be detected. Reliable images are obtained by transthoracic echocardiography (TTE).[5] Two-dimensional TTE can precisely locate the area of narrowing. Color Doppler will show turbulent flow and continuous-wave Doppler study will help to assess the gradient and shows the classical diastolic tail sign. Patients with significant CoA have gradients between the proximal and distal aorta during ventricular systole, and in severe CoA, it persists during ventricular diastole. These patients have continuous flow in the abdominal aorta instead of pulsatile flow.[6]

In this case, the isthmus was ligated; however, on transesophageal echocardiography, the descending aorta was distally producing a pulsatile laminar flow as the PDA was feeding the descending aorta. It could have been detected if there was a femoral arterial pressure monitoring which was not placed earlier as the femoral pulses were palpable making coarctation of aorta unlikely. Ligation of isthmus did not cause increase in aortic line pressures on cardiopulmonary bypass or it was managed by vasodilators

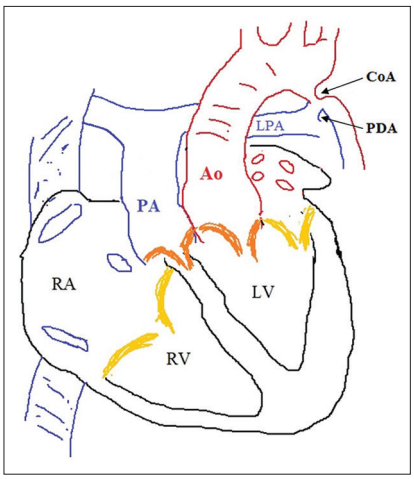

Figure 2: Schematic diagram of anatomically corrected malposition of great arteries. RA–Right atrium, RV–Right ventricle, LA–Left atrium, LV–Left ventricle, PA–Pulmonary artery, Ao– Ascending aorta, DA–Descending aorta, SVC–Superior vena cava, IVC–Inferior vena cava, PDA–Patent ductus arteriosus

and as the descending aorta was being filled by PDA, there was no acidosis or decrease in urine output during the intraoperative period.

It was diagnosed due to the onset of acidosis in the immediate postoperative period, gradient between the radial and femoral pressures, and the concern from surgical side to rule out iatrogenic CoA during postoperative transesophageal echocardiography.

We emphasize the need to carefully assess the anatomy and plan repair intraoperatively in rare congenital anomalies. There is possibility of PDA continuing as descending thoracic aorta in ACLMA which has not been reported so far. Femoral artery canulation is mandatory in cases with large PDA. A high index of suspicion for coarctation of aorta with onset of acidosis on pump or the immediate postoperative period is needed. Pulsatile laminar flow with no gradient on echocardiography in the descending aorta does not always rule out coarctation of aorta. Other associated anomalies have to be considered.

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.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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