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Coarctation of the Aorta in Infants : A Diagnostic Challenge

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ABSTRACT

Coarctation of the aorta (CoA), usually occurring in the thoracic and rarely in the abdominal aorta, remains an underdiagnosed disease entity in the majority of neonates & infants with a misdiagnosis rate being more than 62%. It is an earnest need to diagnose CoA in them at the earliest possible time to provide appropriate management. We retrospectively analyzed data of 6 infants with CoA at Ibrahim Cardiac Hospital & Research Institute to provide clues for the diagnosis of CoA, describe their echocardiographic and computed tomography aortopulmonary angiogram to match them with the diagnosis of CoA and analyze their prognosis.

ORIGINAL ARTICLE

INTRODUCTION:

Coarctation of the aorta (CoA) is the narrowing of the aorta in the region of the insertion of arterial duct, with or without additional abnormalities of the aortic arch[1]. It usually occurs in the thoracic and rarely in the abdominal aorta. An underdiagnosis rate of over 62% places CoA as the most frequently misdiagnosed critical congenital heart disease[2]. It is a pressing need to pick up CoA as early as possible. Simple pulse oximetry and palpation of peripheral pulses can give us clues that are confirmed by echocardiography and computed tomography aortopulmonary angiogram. Early surgical correction and lifetime echocardiographic follow-up must be performed to improve prognosis. In the present study, we retrospectively analyzed data of patients with CoA at Ibrahim Cardiac Hospital & Research Institute to (i) get the clues for diagnosis of CoA, (ii) describe how the echocardiographic and computed tomography aortopulmonary angiogram matches with the diagnosis of CoA, and (iii) analyze the clinical prognosis.

Case -1

Titly, a 12-month-old female baby was diagnosed with an atrial septal defect (ASD) and visited us for another echocardiogram for confirmation. On clinical examination (prior to echocardiography), her pulses were absent in both dorsalis pedis with radio-femoral delay. Echocardiogram revealed discrete CoA with PG:42 mmHg with bicuspid aortic valve and ASD (Figure 1). Computed tomography (CT) aortopulmonary angiogram revealed severe CoA with hypoplastic aortic arch, ASD, and PAPVC (Partial anomalous pulmonary venous connection) (Figure 2). She underwent balloon dilatation and she is stable now.

Case-2:

Alif, a 3-month-old male baby was referred to us for echocardiography as he has PFO (Patent foramen ovale) and also suffering from renal problems. As we checked his peripheral pulses before the echo, we found absent pulses in both dorsalis pedis. Echocardiogram revealed

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a left-sided aortic arch with juxta-ductal coarctation with a thin antegrade flow and a peak systolic gradient of 23 mmHg across the coarcted segment with parachute MV and moderate MS. The computed tomography aortopulmonary angiogram revealed severe CoA (4.8 mm) from the origin of the left subclavian with a hypoplastic mid/distal aortic arch (Figure 3). He underwent balloon coarctoplasty and now he is 11 months old and stable.

Case-3:

Al-Muttaqin, a 4-month-old male baby having repeated respiratory tract infection came for an OPD consultation. On examination, he had tachycardia with absent peripheral pulses in the lower limbs, tachypneic, bilateral basal crackles, and hepatomegaly (4 cm below the right costal margin). Echocardiography revealed juxta-ducctal CoA with a systolic pressure gradient of 23 mmHg across the narrowed segment (Figure 4). Computed tomography aortopulmonary angiogram revealed juxtaducctal CoA (5.5 mm) from the origin of the left subclavian artery (Figure 5). Medical management was given at that time and advice for coarctoplasty. He is now 12 months old, but coarctoplasty has not been done yet.

Case-4:

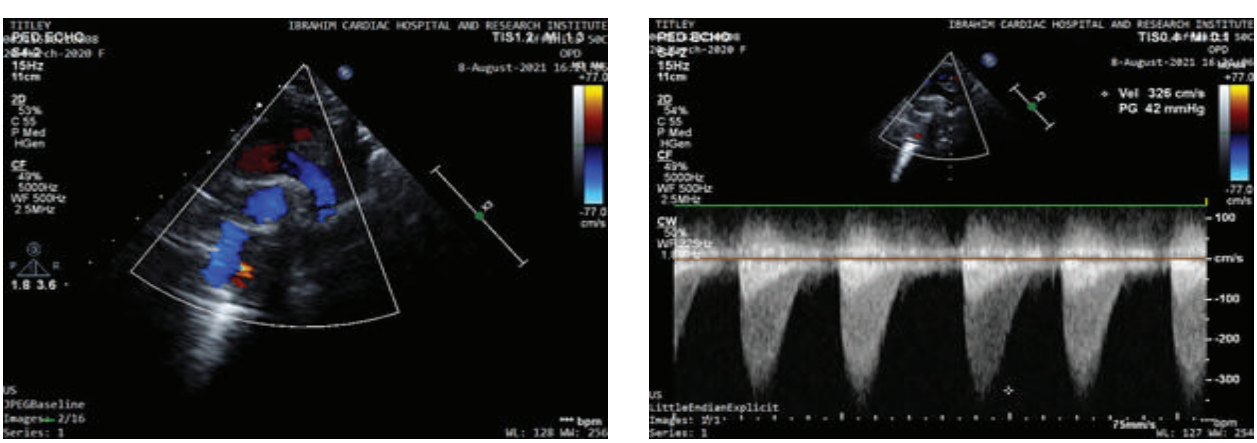
Swad, a male baby underwent balloon coarctoplasty at the age of 3 months. Now at the age of 8 months, he came for follow-up echocardiography. On examination, his peripheral pulses are feeble in his lower limbs at our OPD. Echocardiogram revealed a significant posterior shelf at the level of descending aorta resulting in moderately severe CoA (max PG:30 mmHg). There is post-stenotic dilatation with large ASD and small PDA (Figure 6).

Case-5:

Riad, a 6-month-old male baby had an outside echo which revealed ASD, VSD, and mild PAH. On examination, peripheral pulses were found absent in his lower limbs. A systolic blood pressure difference of more than 40 mmHg was noted in the upper and lower limbs. Echocardiogram revealed there is a post subclavian CoA with a thick membrane with two jets (max PG: 42 mm Hg) with large ASD and subarterial VSD (Figure 7). He was advised for a Computed Tomography Aortopulmonary Angiogram.

Case-6:

Ariyan, a 3-month-old female baby's echocardiogram revealed functionally single ventricle (fSV) (RV systemic ventricle, rudimentary LV), large ASD secundum, large inlet VSD, mild CoA, high RV pressure>>PAH. He was advised for a computed tomography aortopulmonary angiogram. But he lost follow-up.



A B

Figure 1: Transthoracic echocardiographic views of coarctation of aorta. Suprasternal sagittal color Doppler imaging showing absent antegrade color flow across the coarctation site. B) & C) Continuous wave spectral Doppler imaging across the coarctation segment in suprasternal view. Doppler shows increased flow velocity in systole with continuation of flow in diastole (diastolic runoff).

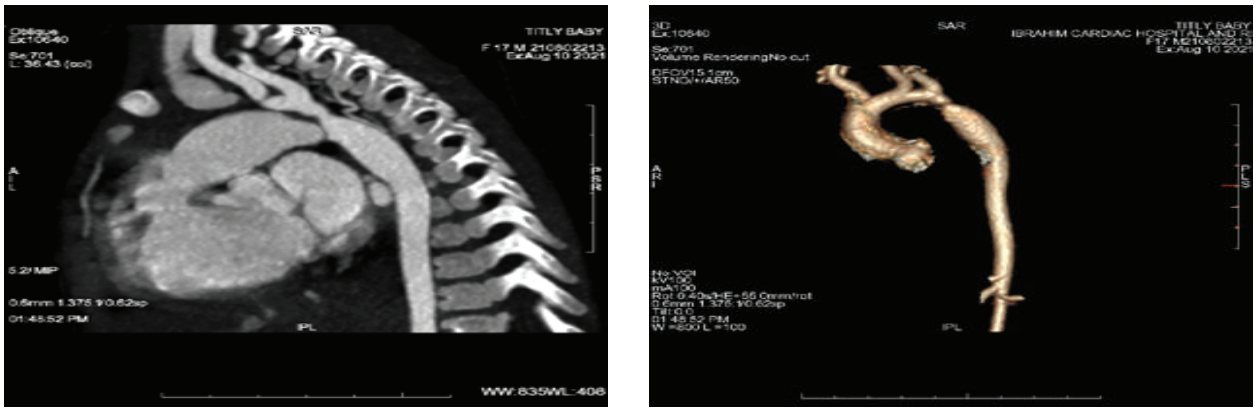


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Figure 2: Computed tomography scan imaging of coarctation of the aorta. A) Two- dimensional sagittal reconstruction showing the site of discrete narrowing at the level of aortic isthmus. B) Three-dimensional reconstructions of computed tomographic angiography showing the site of discrete narrowing in the same patient.

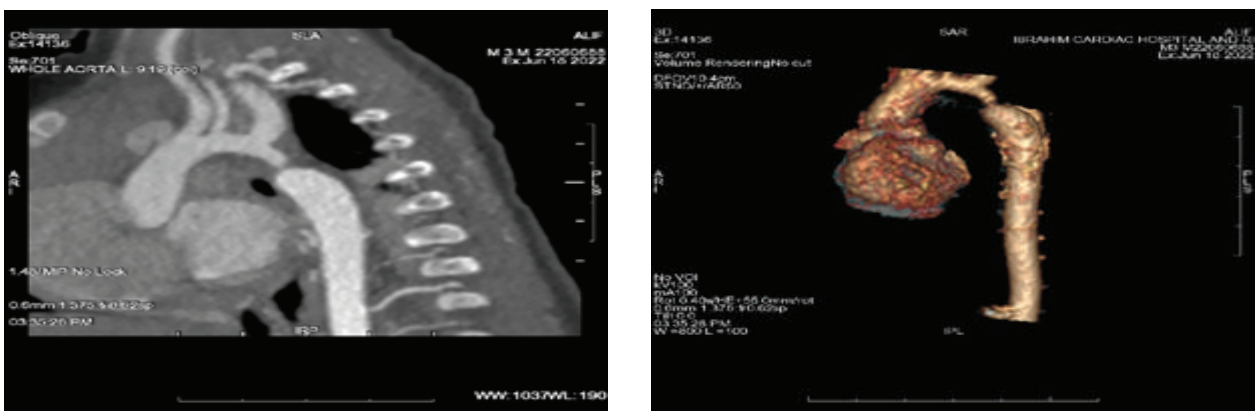

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Figure 3: Computed tomography scan imaging of coarctation of the aorta. A) Two-dimensional sagittal reconstruction showing the site of discrete narrowing at the level of the aortic isthmus. B. Three-dimensional reconstruction of computed tomographic angiography showing the site of discrete narrowing in the same patient.

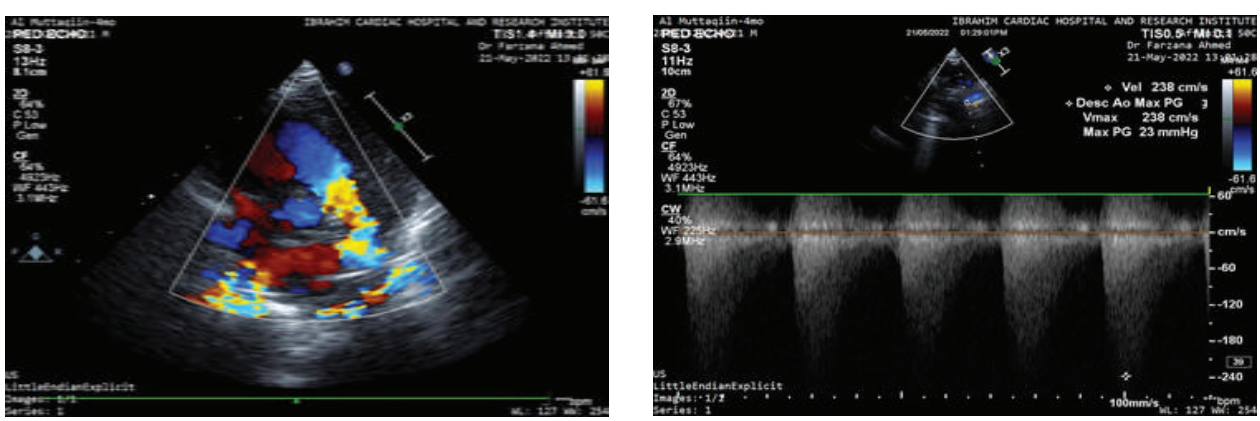

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Figure 4: Continuous wave spectral Doppler imaging across the coarctation segment in suprasternal view. Doppler shows increased flow velocity in systole with continuation of flow in diastole (diastolic runoff).

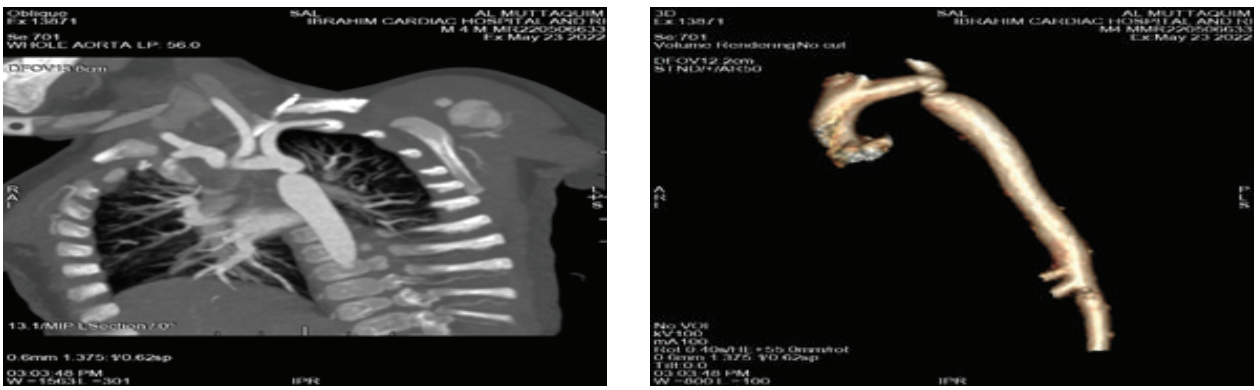

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Figure 5: Computed tomography scan imaging of coarctation of the aorta. A) Two-dimensional sagittal reconstruction showing the site of discrete narrowing at the level of origin of the left subclavian artery. B. Three-dimensional reconstruction of computed tomographic angiography showing the site of discrete narrowing in the same patient.

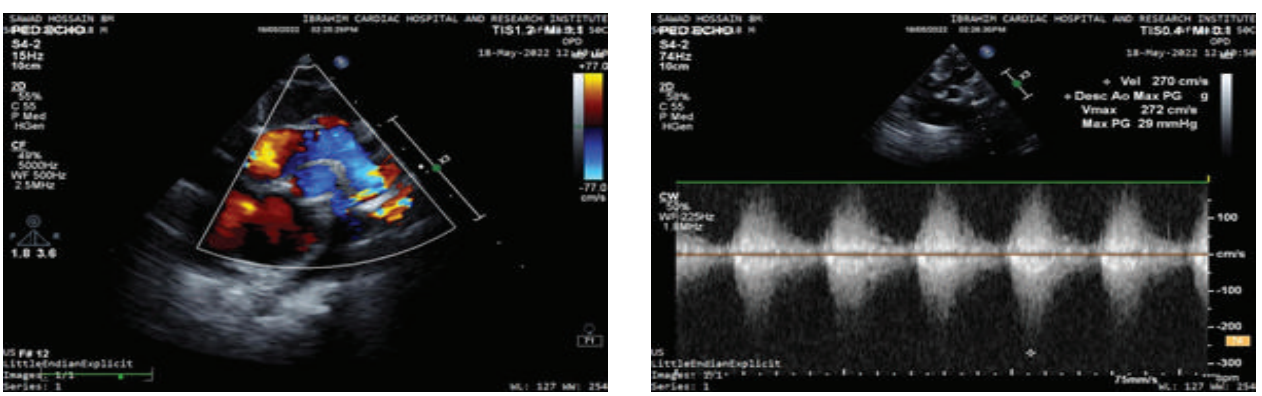

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Figure 6: Transthoracic echocardiographic views of coarctation of the aorta. Suprasternal sagittal color Doppler imaging showing absent antegrade color flow across the coarctation site. B) Continuous wave spectral Doppler imaging across the coarctation segment in suprasternal view. Doppler shows increased flow velocity in systole with continuation of flow in diastole (diastolic runoff).

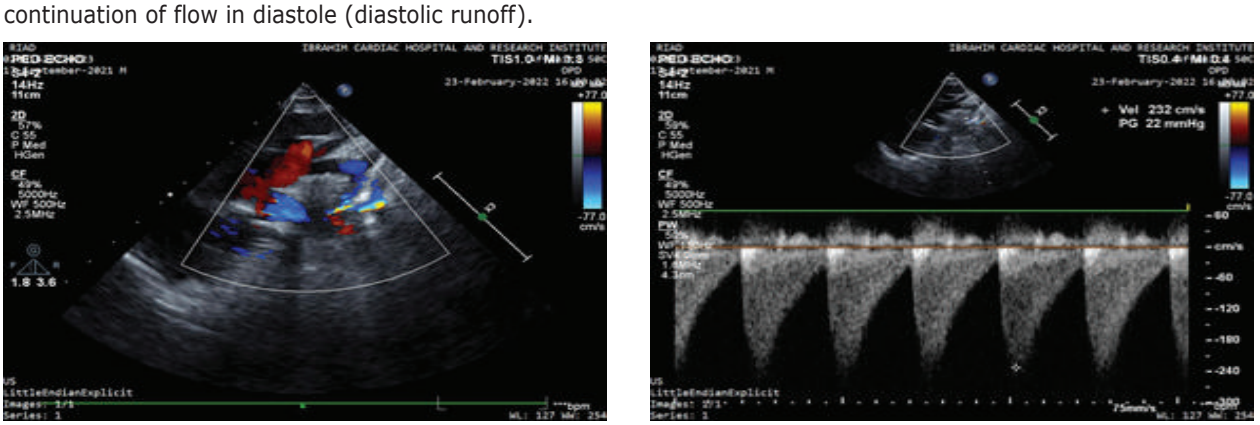

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Figure 7: Transthoracic echocardiographic views of coarctation of the aorta. A) Suprasternal sagittal color Doppler imaging showing absent antegrade color flow across the coarctation site. B) Continuous wave spectral Doppler imaging across the coarctation segment in suprasternal view. Doppler shows increased flow velocity in systole with continuation of flow in diastole (diastolic runoff).

RESULTS:

We identified 6 infants who attended our OPD between Jan 2021 to Dec 2022. The mean age was 6 ± 6 months (range: 3-12 months). Out of 6 patients, 4(66.7%) were male and 2(33.3%) were female. At diagnosis, 5(83.3%) patients had no palpable pulse in the lower limbs and 1(16.7%) had a feeble palpable pulse. Electrocardiographic tests were performed in all 6 cases (100%). Fifty percent of the patients underwent computed tomography angiography. The mean peak descending aortic gradient (DAG) was 29.8 mmHg. Other common echocardiographic findings were bicuspid aortic valve (BAV) (16%), atrial septal defect (66.6%), ventricular septal defect (33.3%), patent ductus arteriosus (16%) and parachute mitral valve insufficiency (16%). It would be worthwhile to note that pulmonary artery hypertension occurred in 50% of the cases. Three (50%) patients had done computed tomography aortopulmonary angiogram and all (100%) revealed CoA which matches our Echo findings. Two (33.3%) babies had CoA with hypoplastic aortic arch. Three (50%) patients underwent balloon coarctoplasty and all of them are stable now.

DISCUSSION:

CoA is not uncommon congenital heart disease (CHD) and merits early detection and treatment. It comprises about 5-8% of all CHD.[3] This relatively common defect may occur as an isolated defect or in association with various other lesions, most commonly bicuspid aortic valve and ventricular septal defect (VSD).[4] Mild coarctation may not be diagnosed until adulthood. Babies with severe coarctation of the aorta may show symptoms shortly after birth. Imaging plays a crucial role in the diagnosis, therapeutic planning, and follow-up of patients with CoA.[5] In this case series, we evaluated data from 6 infants with coarctation of the aorta. Our main objectives were to identify the most frequent clinical findings that could benefit the early diagnosis and to describe the course of echocardiographic measures and confirmation by CT aortopulmonary angiogram as cardiac MRI is not available in our center. Our main findings were impalpable peripheral pulse in most patients. Echocardiographic findings completely matched the CT scan. Balloon coarctoplasty was the treatment in our cases. Infants had a mean pressure gradient of more than 20 mmHg. In a study, Begum[3] reported 50 patients with CoA who underwent coarctoplasty during a period of 2 years (Jan, 2007-Dec, 2009). Among them 22 were female and 19 were neonates and 03 were infants. The neonates presented with heart failure while infants had a pressure gradient of more than 20 mmHg. In another retrospective study, Begum et al[3] found 20(7.87%) infants with coarctoplasty out of 254 infants with CHD over a period of 5 years-(from 2014-2019).

Although surgical repair has been the primary treatment for CoA at most centers, some centers use balloon angioplasty as a palliative strategy in neonates too sick for major surgical procedures. Balloon angioplasty appears to be associated with a higher rate of coarctation than surgical repair and the rate of complications (including femoral artery injury) is high during infancy. Some centers use low-profile endovascular stents in very sick infants, which has the advantage of not producing aneurysms. Primary coronary stent implantation is a feasible bridging therapy to surgery in very low birth weight infants with critical aortic coarctation and may be preferably postponed until 3 kg with PGEs.[7]

Another study demonstrated that the degree of relief of aortic obstruction and the frequency with which re-intervention is required are similar between surgical and balloon therapy groups, but the morbidity and complication rates were higher with the former strategy. Balloon angioplasty may be an effective alternative to surgery for the relief of CoA.[8] In our study, all cases were stable with balloon angioplasty at the time of diagnosis. Most cases of balloon angioplasty were done between 3-6 months of age except for one case whose balloon angioplasty was done at the age of 12 months. The management guidelines by the American Heart Association 2011, for adult and

pediatric patients with CoA, recommend correction of coarctation in early childhood to prevent the development of systemic hypertension. If coarctation escapes early detection, the repair should be performed at the time of diagnosis.[9]

Conclusion: The study concluded that the majority of infants with coarctation of the aorta presented with impalpable peripheral pulse or feeble pulse in arteria dorsalis pedis which were later confirmed by echocardiography and computed tomography aortopulmonary angiogram. So simple pulse oximetry and palpation of peripheral pulses can give us clues that could be confirmed by echocardiography and computed tomography aortopulmonary angiogram. Early surgical correction and lifetime echocardiographic follow-up must be performed to improve prognosis. However, the coarctation treatment strategy has changed over time. Nowadays coarctectomy is performed on the pump with aortic arch enlargement with the concomitant repair of associated lesions, whereas in patients with simple coarctation, balloon dilatation, and stenting is the preferred approach.

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