

Coarctation of Aorta with Supravalvular Pulmonary Stenosis in an Adult Patient: A Rare Exception of the Fetal Flow Pattern Theory

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Abstract Coarctation of the aorta is a common congenital defect whose overall incidence is 5–8% of all congenital cardiac anomalies. Associated cardiac anomalies have been well described in previous studies examining specific subgroups of coarctation of aorta patients, particularly infants, excluding older children, adolescents, and adults. The association of coarctation with left-sided obstructive lesions (Bicuspid aortic valve, Parachute mitral valve, Mitral atresia, Aortic atresia) were well documented in the literature. The association of coarctation of aorta with right- sided obstructive lesions is rare and a hemodynamic explanation based on fetal flow pattern was given for this. Herein we report a case of coarctation of aorta with right sided obstructive lesion in form of supravalvular pulmonary stenosis in an adult female. To the best of our knowledge this is the first case report where an adult female presented with coarctation of aorta along with supravalvular pulmonary stenosis.

Keywords: *coarctation of aorta, supravalvular pulmonary stenosis, fetal flow pattern theory*

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1. Introduction

Coarctation of aorta was first described by Morgagni in 1760 as a zone of constriction in the descending aorta. [1] Coarctation of the aorta is a common congenital heart defect whose overall incidence is 5–8% of all congenital cardiac anomalies with male dominance [2]. The coarctation of aorta is caused by deformity of the aortic media and intima, resulting in a prominent posterior infolding of the aortic lumen. This deformity typically occurs at or near the junction of the aortic arch and descending thoracic aorta [3]. Associated cardiac anomalies have been well described in previous studies, but these studies tended to examine specific subgroups of patients with coarctation, particularly infants and necropsy specimens [4,5,6,7]. Development of coarctation of aorta in association with various left sided obstructive lesions and not with right sided obstructive lesions was explained by Rudolph, Heymann, and Spitznas (1972) on the basis of fetal flow pattern hypothesis [8]. Here, we report the first case of an adult female presenting with coarctation of aorta and supravalvular pulmonary stenosis, violating this fetal flow pattern hypothesis.

2. Case Report

A 36-year-old female presented to Cardiology Department with left lower limb claudication, fatigue, and exertional dyspnea which were increasing over the last 3-4 years prior to presentation. Her past medical history revealed history of hypertension for which she was taking irregular treatment for the past 15 years. On physical examination, no facial dysmorphism was present, the femoral pulses were palpable bilaterally; however, a radial-femoral pulse delay was noted. The blood pressure was 200/130mmHg in both upper limbs and 140/90mmHg in lower limbs with a systolic pressure gradient of 60 mmHg between upper and lower extremities. Apex beat was in 5th intercostal space in midclavicular line and heaving in character. A grade 1 right parasternal heave was present. Grade 2/6 ejection systolic murmur was heard at the pulmonary area and continuous murmur heard all over the back. On the basis of history and physical examination aortic coarctation was suspected, and further investigations were performed. Results of routine blood chemistry and urine analysis were normal. Twelve-lead electrocardiogram revealed left ventricular hypertrophy and chest x-ray showed rib notching. The two dimensional (2D) transthoracic echocardiography showed a turbulent flow just below the origin of the left subclavian artery, while the continuous wave Doppler revealed a peak flow velocity of 3.2 m/sec in descending aorta, and the peak pressure gradient was estimated at 42 mmHg. Apart from the coarctation of aorta, a significant gradient of 55 mmHg

was also seen in the pulmonary artery. The pulmonary valves were morphologically normal and there was a narrowing seen above the level of the pulmonary valves (Figure 1, Figure 2, Figure 3). The patient then underwent a transesophageal echocardiogram (TEE) that confirmed the findings of coarctation of aorta and a supra valvular pulmonary stenosis. The Computed Tomography angiogram (CT angio) (Figure 4, Figure 5) showed severe coarctation of aorta below the origin of the left subclavian artery, together with poststenotic dilatation. Cathetrization study

(Cath study) (Figure 6, Figure 7) revealed coarctation of aorta and supra valvular pulmonary stenosis. Cath data also revealed a 60 mmHg pullback gradient in the main pulmonary artery and a pullback gradient of 70 mmHg across the coarctation of aorta. So patient was diagnosed as having coarctation of aorta with supra valvular pulmonary stenosis. The patient was then referred to cardiothoracic surgery to evaluate her candidacy for surgical therapy. Cardiac surgeon advised for surgery but she refused for surgery.



Figure 1. Parasternal short axis at base showing normal pulmonary valve(thin arrow) and supra valvular obstruction(thick arrow)

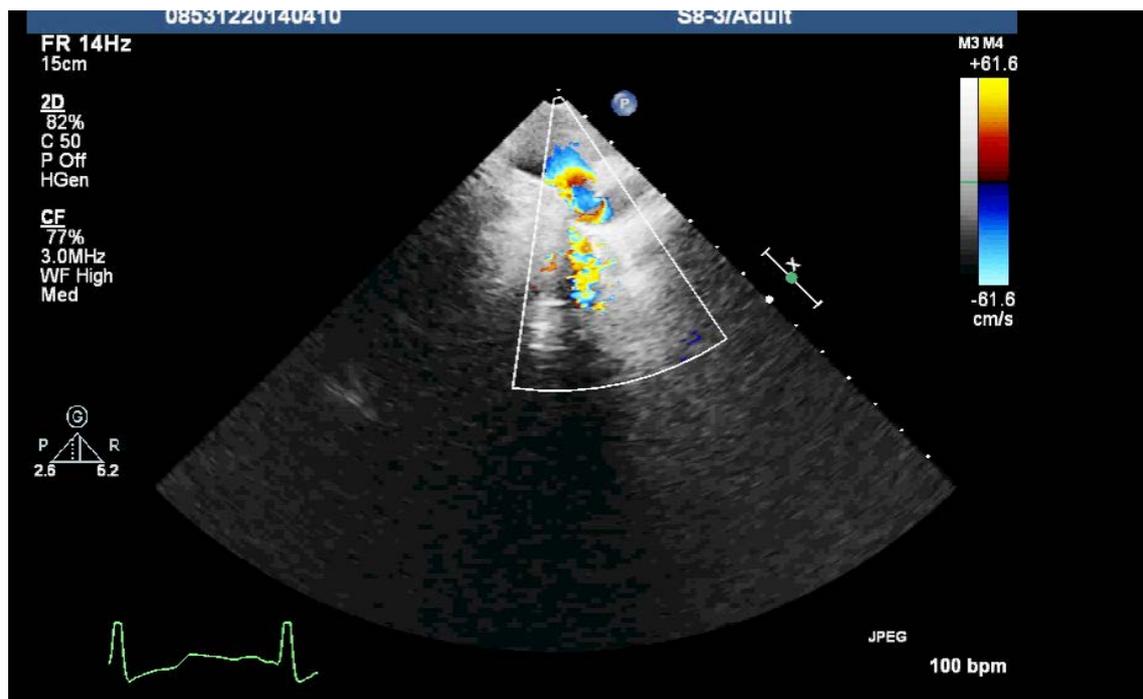


Figure 2. Colour Doppler in parasternal short axis view at base showing turbulence at supra valvular level

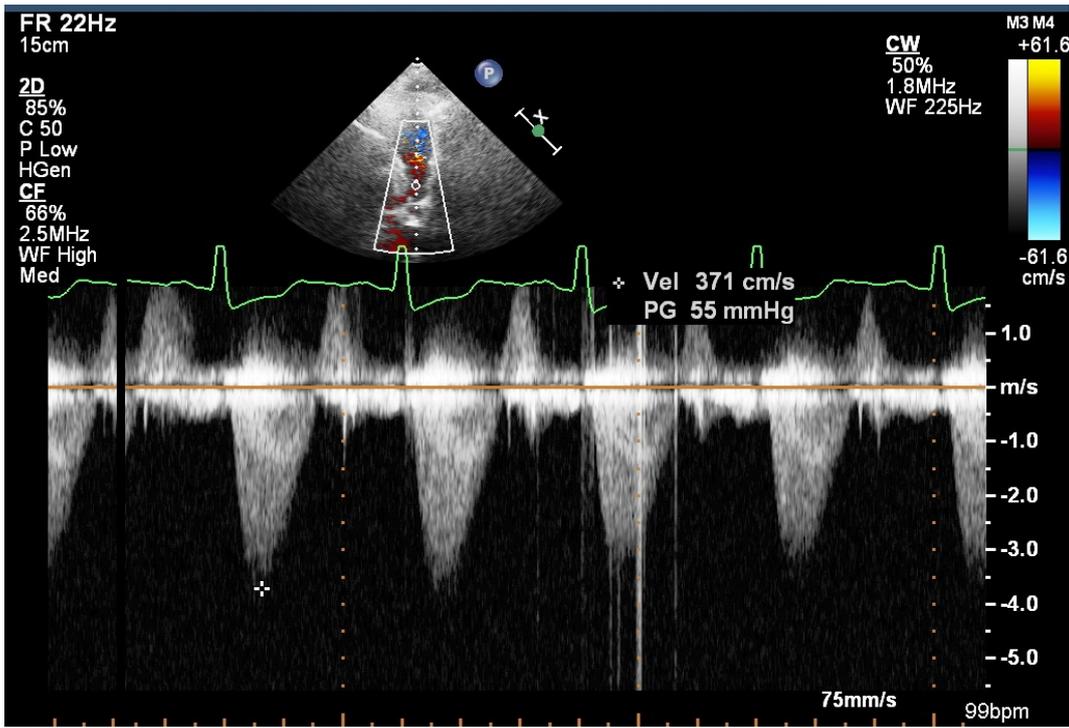


Figure 3. Continuous wave Doppler at supravalvular level in basal short axis view showing gradient of 55mm of hg.

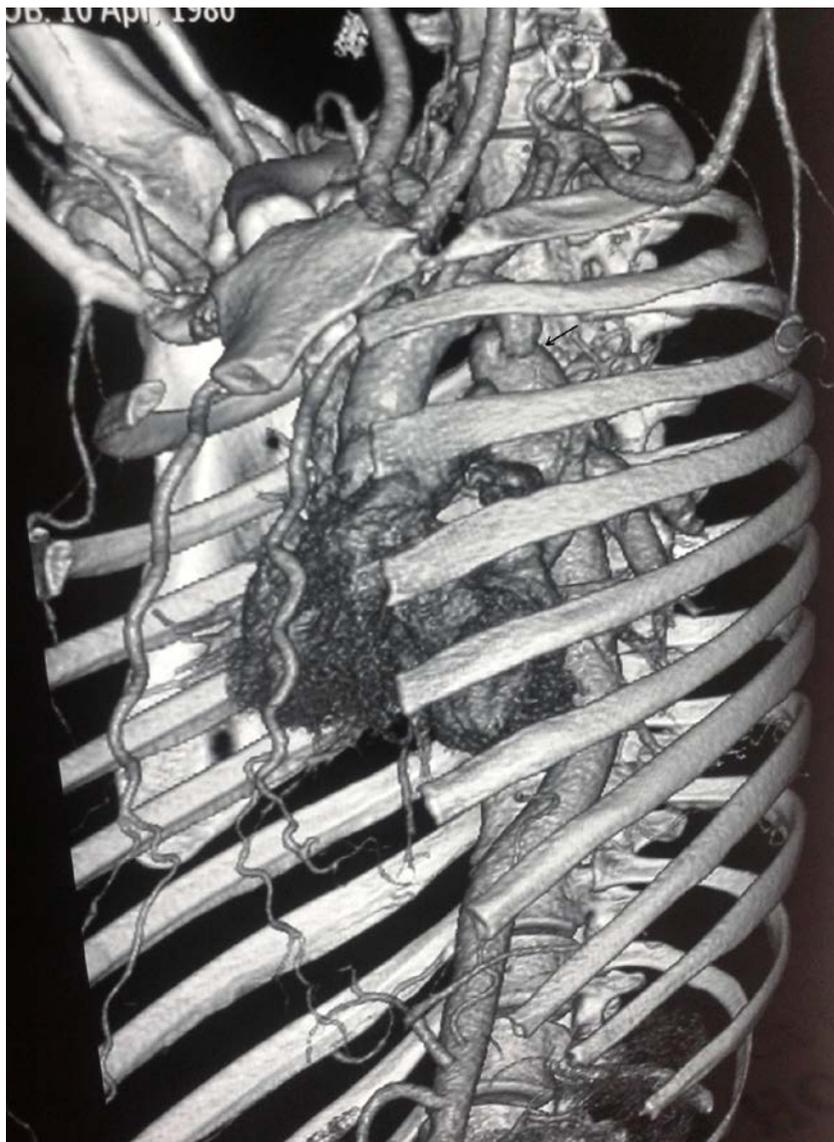


Figure 4. CT angiography showing Coarctation of aorta

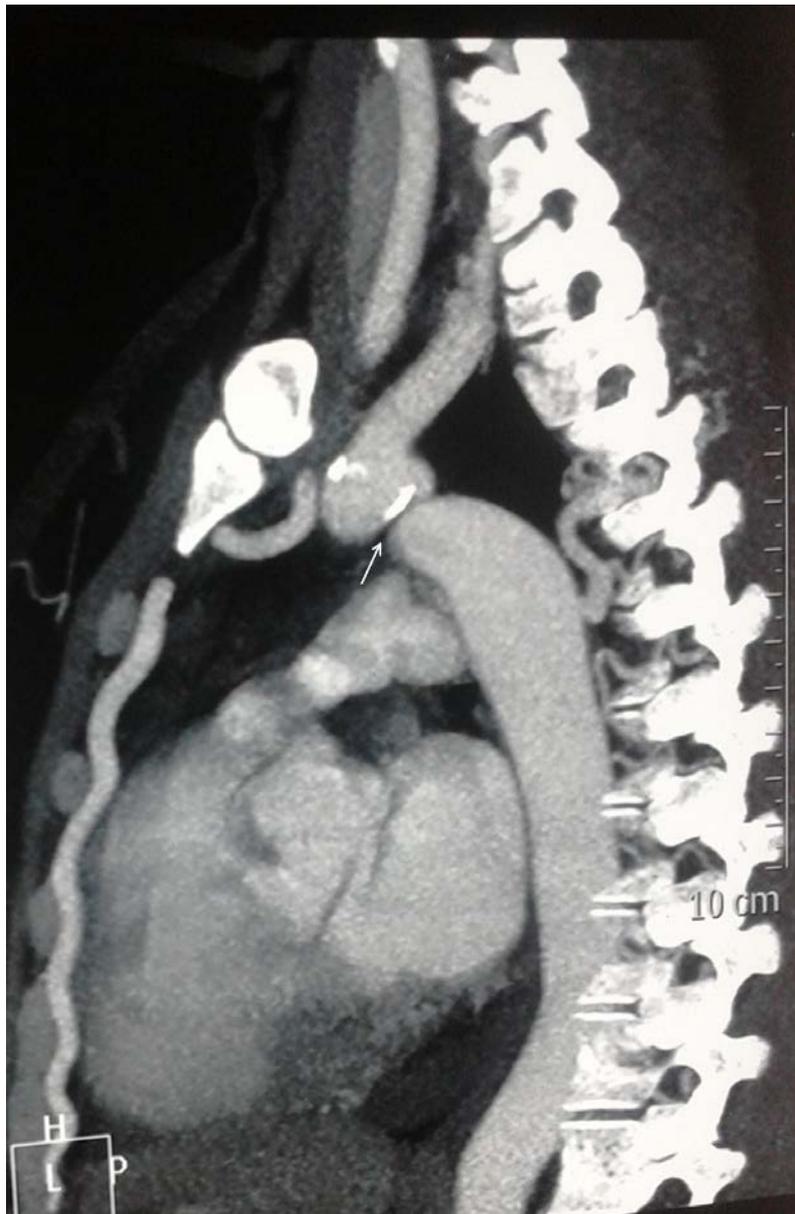


Figure 5. CT angiography showing Coarctation of aorta

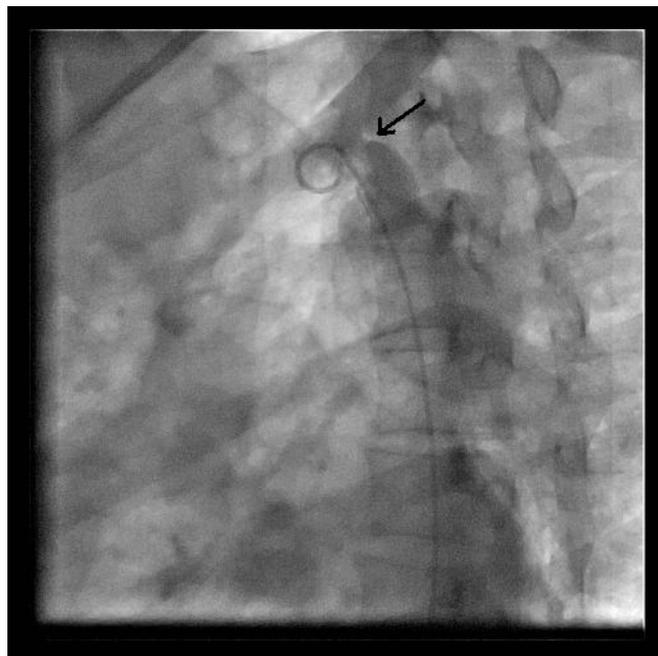


Figure 6. Aortogram during cath study showing coarctation of aorta

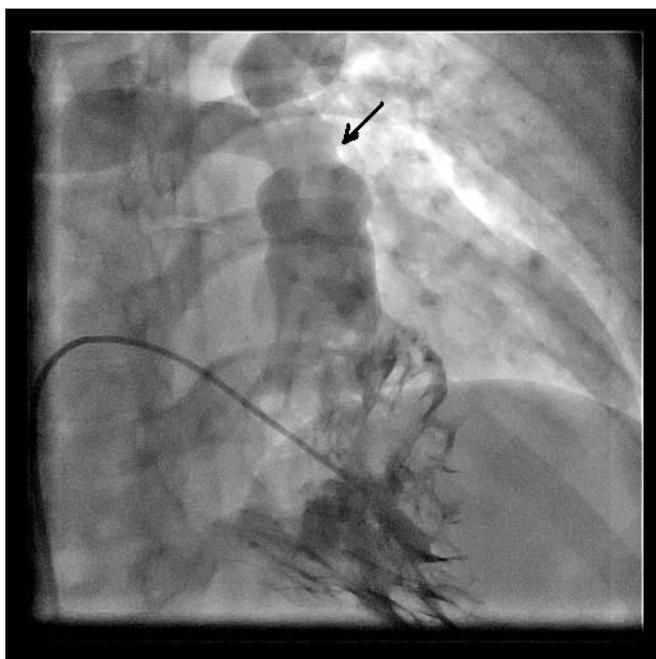


Figure 7. Right ventricle angiogram showing normal pulmonary valve and supravalvular pulmonary stenosis(arrow).

3. Discussion

Coarctation is a narrowing in the descending aorta, at the insertion site of the ductus arteriosus, adjacent to the origin of the left subclavian artery. However, obstruction could also occur in the transverse aortic arch, or abdominal aorta. It can be discrete or tubular, and is associated with various cardiac and noncardiac abnormalities in up to 50 % of patients. [9,10,11] The haemodynamic explanation given by Rudolph et al. (1972) for juxtaductal coarctation is that it represents an exaggeration of the normal aortic anatomy in the newborn. The neonatal aorta is characterized by a disparity in cross-sectional area between the aortic isthmus, the segment of aorta between left subclavian artery and ductus arteriosus, and the descending aorta (Sinha et al., 1969) and sometimes mistakenly referred to as tubular coarctation of the aorta, but it must be stressed that this is the normal appearance in the neonate [12]. The haemodynamic explanation offered for this state of affairs is derived from studies on fetal lambs (Dawes, Mott, and Widdicombe, 1954) [13] and the preivable human fetus (Rudolph et al., 1971) [14] where dimensions of the great vessels appear to reflect flow through them in utero. Each ventricle ejects approximately 50 per cent of the combined output to its respective great vessel. As blood passes from ascending aorta, the innominate, left common carotid, and left subclavian arteries draw their normal 20 to 25 per cent of the output, so aortic dimensions decrease and as a result the isthmus is 30 per cent narrower than the ascending aorta. Because of the low arterial Po₂ (Cook et al., 1963) [15] and lack of mechanical expansion of the lungs (Lauer et al., 1965) [16] the fetal pulmonary vascular resistance is high and only a small proportion of the right ventricular output passes to the lungs. Most of the flow to the main pulmonary artery passes through the widely patent ductus arteriosus to descending aorta. Thus the latter receives both the majority of the right ventricular output together

with flow from the left ventricle that has passed through the aortic isthmus. As a result the descending aorta is considerably wider than the aortic isthmus and the junction of isthmus and descending aorta is characterized in the normal neonate by an indentation on the posterior wall of the aorta opposite the ductus. It is an exaggeration of this normal posterior indentation that results in isolated juxtaductal coarctation. The explanation offered for the lack of association of coarctation with right ventricular outflow tract obstructive lesions or dominant right-to-left shunts is based on postulated flow patterns in the fetus. As less blood is ejected from the right ventricle directly to the pulmonary artery, most of the combined fetal ventricular output leaves the heart via the ascending aorta which is a proportionally larger structure than normal area to the descending aorta. In all congenital cardiac anomalies where there is a dominant right-to-left shunt at birth with oligoemic lung fields, i.e. pulmonary atresia ± ventricular septal defect, tetralogy of Fallot with infundibular stenosis the aortic isthmus will be wider than the descending aorta. When this is so, coarctation caused by a narrowed isthmus will not be expected. Furthermore, as there is no disparity in size between isthmus and descending aorta, no indentation on the posterior aortic wall is present, and hence juxtaductal coarctation also is not seen, thus suggesting that diminution of pulmonary blood flow or right ventricular outflow tract obstruction appears to protect against development of coarctation. This contrasts strongly with the left sided obstructive lesions where diversion of blood away from the aorta will tend to exaggerate disparity in size between isthmus and descending aorta causing more chances for development of coarctation of aorta. So, the co-occurrence of coarctation of aorta with supra valvular pulmonary stenosis is just a coincidental finding or some developmental factors responsible for this, which might be studied in future. To the best of our knowledge this is the first case report wherein an adult female patient presented with coarctation of aorta and supravalvular pulmonary stenosis, violating the hypothesis of fetal flow pattern theory.

Statement of Competing Interests

Authors have no competing interests.

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