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INTERESTING MEDICAL IMAGE

Mogul in a Baby

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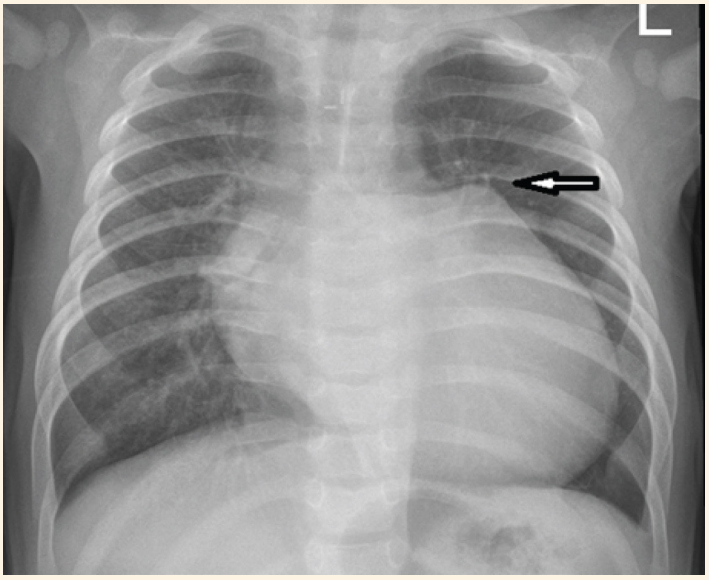

Figure 1: Plain X-ray Chest showing the third mogul on the left heart border (arrow).

The bumps seen on the cardiac silhouette in a plain chest skiagram (the left heart border in particular) have been termed 'moguls' because of the resemblance to mounds of snow formed while skiing.[1] Commonly, four moguls and a variable fifth have been described in adult mitral valve disease referring to the following from above downwards: aortic knob or knuckle, the enlarged main pulmonary artery, the enlarged left atrial appendage (especially in rheumatic mitral valve disease), the ventricular bulge or contour just above the left cardio-phrenic angle (seen with ventricular aneurysm) and the variable fifth mogul at the cardio-phrenic angle caused by pericardial cysts, fat pads or lymphadenopathy.[2]

A 7-month-old female infant (weight = 7.1 kg; height = 75 cm; body surface area = 0.4 m^2) with single ventricle physiology was admitted electively for the second stage operation. She had had a right modified Blalock-Thomas-Taussig (BTT) shunt at the age of 11 days. Echocardiography revealed pulmonary atresia with intact ventricular septum (PA-IVS) with a severely hypoplastic right ventricle (RV) and a hypoplastic but competent tricuspid valve (TV). Dilated myocardial sinusoids were noted. The BTT shunt was patent but narrowed.

Chest radiograph revealed a rather unusual shadow at the left heart border in the third anterior intercostal space [Figure 1].

Pre-Glenn cardiac catheterisation was performed and an aortic root angiogram [Figure 2A] revealed an ectatic left coronary artery that formed the third mogul seen on chest skiagram. Pulmonary angiogram (through the shunt) revealed good pulmonary anatomy and that the left pulmonary artery stopped short of the mogul [Figure 2B]. RV angiogram revealed a severely hypertrophied, small RV with coronary fistulae and opacification of the distal right coronary artery suggesting right ventricle dependent coronary circulation (RVDCC) [Figure 3].

The baby underwent an uneventful redo sternotomy and bidirectional superior cavo-pulmonary (Glenn) shunt with takedown of previous BTT shunt on beating heart cardiopulmonary bypass. Subsequent recovery was uneventful. Parental consent and ethical committee approval were obtained for publication of these images.

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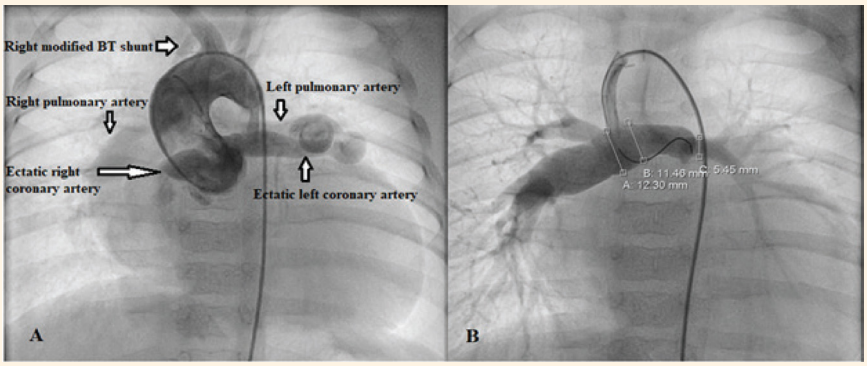

Figure 2: A: Aortic root angiogram showing the ectatic coronary arteries and the mogul. **B:** Pulmonary angiogram through right modified Blalock-Taussig shunt. The left pulmonary artery is not the cause of the third mogul.

Comment

The most common differential diagnosis of a third mogul is an enlarged left atrial appendage. Rarer diagnoses include coronary malformations, an infundibular hump in tetralogy of Fallot, mediastinal mass, cardiac aneurysm, pericardial cyst, pericardial defect and a left pulmonary arteriovenous malformation.[2,3]

Pulmonary atresia with intact ventricular septum comprises several subsets depending on the size of the tricuspid valve (TV) and morphology of the right ventricle (RV) and the possible eventual outcome. Four groups are noted in the spectrum—biventricular, one and a half ventricular, univentricular and the heart transplant group (ongoing myocardial ischaemia).[4]

The formation of coronary fistulae depends on a competent tricuspid valve with pulmonary atresia which leads to supra-systemic RV pressure in systole. This results in opening up of fistulous communications with the coronaries and initially results in bidirectional flow in coronaries, with reversal of flow into the aorta in systole.[5] The resultant to and fro flow leads to eventual occlusion of the communication with the aorta and coronary perfusion becomes dependent on the right ventricle which pumps relatively deoxygenated blood, a state called RVDCC. The more the myocardium that is RV dependent, the higher is the risk of myocardial dysfunction.

Maintenance of good perfusion pressure during surgery is important to avoid myocardial dysfunction. The current patient did not require aortic cross-clamping and cardioplegic arrest, but if such patients do, then it is important to give cardioplegia into the right ventricle as well.[6] Early ligation of the coronary artery fistulae or RV decompression could possibly prevent RVDCC.[7]

The shunt procedure carries the highest risk of mortality because, although it results in better oxygenation, it reduces coronary perfusion and survival depends on a thin line of optimum balance between the two. Thus, an ominous radiological sign is illustrated.

AUTHORS' CONTRIBUTION

PSK conceptualised and designed the work. AMSF performed the cardiac catheterisation and interpreted the images. NAV drafted the manuscript. NAV, CK and AL revised the manuscript. All authors approved the final version of the manuscript.

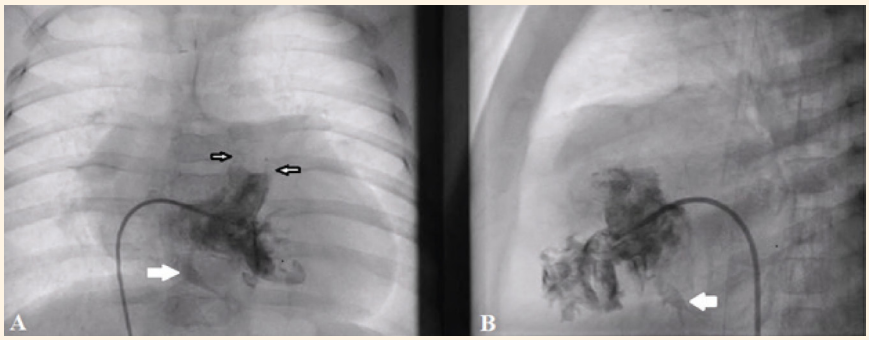

Figure 3: A & B: Biplanar views of the right ventricular angiogram showing a hypoplastic, heavily trabeculated right ventricle with coronary fistulae (small arrows) and opacification of the distal right coronary artery (solid white arrow) suggesting right ventricle dependent coronary circulation.

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