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Case Reports

Total Correction of Tetralogy of Fallot After Percutaneous Coiling of MAPCA: A Case Report

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Introduction

Major aortopulmonary collateral arteries (MAPCAs) are blood vessels that bring systemic blood flow to the pulmonary arteries. They develop in response to decreased pulmonary blood flow and cyanosis. Tetralogy of Fallot (TOF) with pulmonary stenosis is associated with the development of MAPCAs in less than 5% of cases, although it is seen in about two-thirds of patients having pulmonary atresia. MAPCAs are usually clinically silent, presenting only in late cases with haemoptysis. Their presence complicates the operative management of TOF as excessive return of blood floods the operative field. Postoperatively also, the presence of MAPCAs may make it difficult to wean a patient off the ventilator because of pulmonary congestion, and may provoke congestive cardiac failure by raising pulmonary arterial pressures. Therefore, appropriate management of MAPCAs is necessary for both short and long term outcome. Here, we present a case of TOF (with pulmonary stenosis) and MAPCAs in a 13 year old boy, for whom sequential percutaneous coiling of MAPCAs and total correction under hypothermic cardiopulmonary bypass was done.

Case Report

A thirteen year old boy presented with occasional breathlessness, fever and palpitations since birth. Clinically, he appeared not ill-looking, with mildly retarded growth, pulse 80 beats per minute, sinus rhythm, pressure 90/60 mm of Hg, having a systolic murmur heard maximally in the second left intercostals space, radiating to the axilla and the back. His saturation was 92% on room air. He was taking propanolol (10 mg) 12 hourly per operatively.

Preoperative haemoglobin was 15.4 g/dL and platelet count was 290,000/μL.

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Chest x-ray showed a boot-shaped heart with normal transverse diameter. There was a left aortic arch. Lung fields looked clear. ECG showed sinus rhythm with right ventricular hypertrophy.

By preoperative echo, he was diagnosed as TOF with good PA anatomy and MAPCAs. Situs solitus, AV and VA concordance noted. He had a large, non-restrictive malaligned VSD (20 mm in size) with trabecular extension and right to left shunt, 50% aortic over-ride seen. There was severe infundibular, valvar and supravalvular stenosis with the possibility of a supravalvular band or suprapulmonary membrane in the main pulmonary artery (MPA). Valve cusps were markedly thickened with systolic doming. The combined subvalvular and valvular gradient was 90 mm of Hg. Peak pressure gradient at the supravalvular level beyond the band was 114 mm of Hg. MPA was 16 mm, left pulmonary artery (LPA) 9 mm, and right pulmonary artery (RPA) 7 mm. Post-stenotic dilatation of MPA seen. PAs were confluent and 2 MAPCAs to the right lung were noted. Moderate RVH noted. LVIDd 38 mm, LVIDs 28 mm. EF 61%. Good ventricular function noted. Left aortic arch seen. Great vessels in normal relationship.

Cardiac catheterization was done on 16.08.01. The diagnosis of Tetralogy of Fallot was confirmed. Right ventricular outflow obstruction was confirmed to be at all levels, from infundibular to supravalvular (Figs 1 and 2). The number of branches of the pulmonary arteries supplying the lungs was estimated visually to be seventeen. The study showed normal coronary artery anatomy and two MAPCAs supplying the mid and lower zones of the right lung arising from the middle descending thoracic aorta. RV pressures were same as systemic pressures. The lower MAPCA (probably supplying the right apical basal and medial and posterior segments of the lower lobe) had a stenosed origin. The upper MAPCA was smaller and did not appear to have a significant area of distribution within probably the right apical basal segment.

Intervention was staged. The selective catheterization of MAPCAs was performed by means of televised fluoroscopy, with reference to the thoracic aortogram

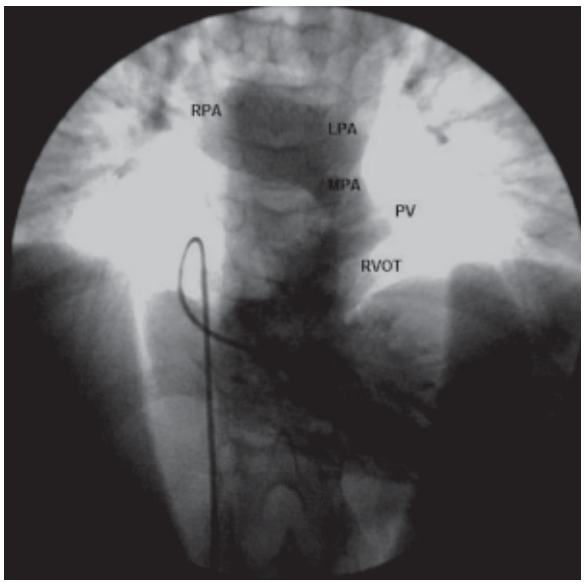

Fig.-1: *Image from cardiac catheterization study: anteroposterior view*

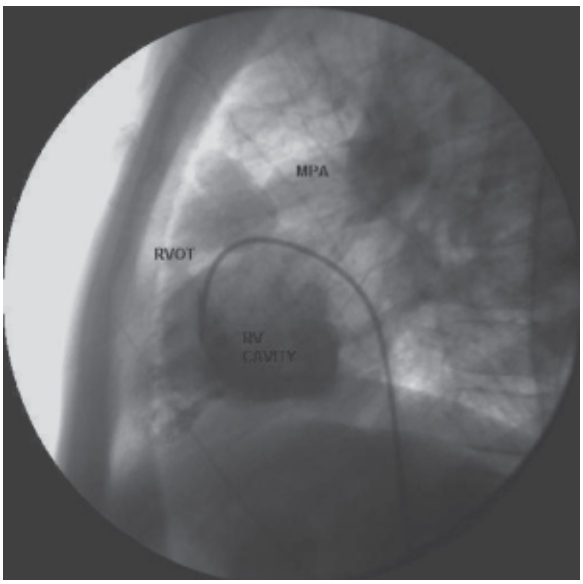

Fig-2: *Image from cardiac catheterization study: lateral view*

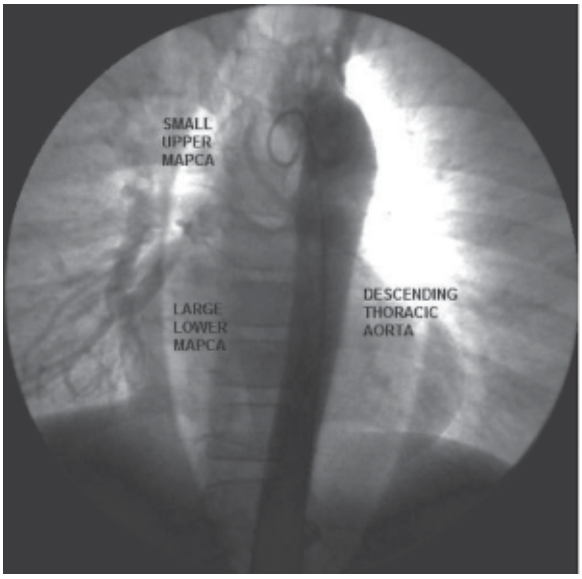

Fig.-3: *Descending thoracic aortogram showing MAPCAs*

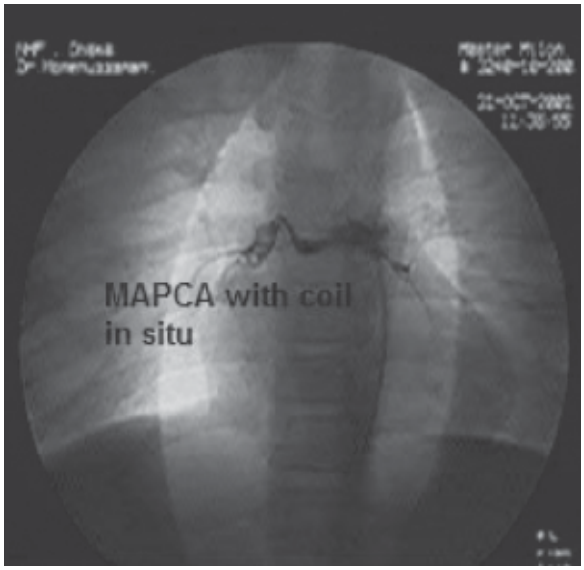

Fig.-4: *Descending thoracic aortogram showing occluded MAPCA after coiling*

that had been taken at the time of cardiac catheterization. Initial coil embolization of right lung lower lobe MAPCA was done using 4 Cook coils, passed into the proximal part of the vessel through a 6 Fr catheter, resulting in complete interruption of flow, as demonstrated by angiogram. The procedure was uneventful and the results excellent (Figs 3 and 4).

This was followed by total correction of TOF under moderate hypothermic cardiopulmonary bypass with cold blood cardioplegic arrest with topical cooling. Lowest rectal temperature was 26ºC.

Peroperatively, he was found to have a large VSD (3.0 x 3.5 cm) with 75% aortic overriding. There was severe supravalvular and infundibular pulmonary stenosis (PS) with mild valvlular PS. Pulmonary arteries were good sized. GORETEX® (polytetra-

fluoroethylene) patch closure of VSD was done. Infundibular resection with pericardial patch enlargement of right ventricular outflow tract (RVOT), percicardial patch enlargement of MPA and pulmonary valvotomy was done. There was no neurological complication. Length of hospital stay was 7 days. He was discharged on digoxin and diuretics. One month after discharge, he was readmitted for congestive cardiac failure, which responded well to increased dose of diuretics.

At follow-up on 09.12.01, RVOT was found to be of good caliber, with mild pulmonary valvular stenosis (gradient across PV 36 mm of Hg). In 2004, the pulmonary gradient had come down to 6 mm of Hg. No residual VSD was seen. Good LV systolic function was noted. Heart rate was 90 beats per minute. He is on combination of frusemide (10 mg) and spironolactone (25 mg) as a single daily dose.

Discussion

The protocol followed by Cooley et al[1] for the management of Tetralogy of Fallot takes into account patient size, systemic arterial saturations and anatomy. Modified Blalock-Taussig shunt (MBTS) is done in symptomatic babies (hypercyanotic spells, ductal dependent pulmonary circulation), weighing less than 4 kg who then undergo complete repair at 6 to 12 months. Asymptomatic babies weighing less than 4 kg in whom pulmonary artery isolation is threatened, also undergo MBTS with repair at 6 to 12 months. All other patients undergo complete repair after 6 months[1]. In Bangladesh, Tetralogy of Fallot is corrected from around three years of age, with a weight of at least about 10 kg. However, presence of major aortopulmonary collateral arteries (MAPCAs) necessitates modification of the procedure and constitutes a risk factor for the development of postoperative morbidity and mortality.

MAPCAs are vessels communicating between the systemic arteries and the pulmonary vessels. They are persistent elements of the splanchnic vascular plexus, which develop in response to persistent hypoxia and insufficient pulmonary blood flow. It is debated whether they are actually bronchial arteries or persistence of the earliest pulmonary circulation[2]. The term “major” distinguishes them from acquired collaterals and does not indicate any particular size. They are highly variable in their number (usually between one to six), course, origin, arborization and histopathological make-up[3]. When they communicate directly with a pulmonary artery, they are called “large” MAPCAs and are usually greater than 2 mm diameter. Arteries with an indirect connection are termed “small”[4]. A segment of lung may be supplied solely from the true pulmonary arteries, solely from the aortopulmonary collaterals, or from both[3,5].

MAPCAs are debatably an unreliable source of blood to the lungs[2,6]. About 60% have stenotic segments. Those which do not have stenoses may cause hypertensive vascular disease in the supplied segments, which may present clinically as haemoptysis at a later stage[7,8]. As they are clinically inapparent, their presence may be inferred if there is long standing cyanosis in a patient with severe pulmonary stenosis or atresia.

In this case, the lower MAPCA was morphologically similar to the large variety, with a narrowed take-off from the descending thoracic aorta. The lung segments supplied by the MAPCAs appeared to have dual supply, but predominantly from the MAPCA. An estimated three bronchopulmonary segments were supplied by the lower MAPCA, while probably the remaining seventeen were supplied from the pulmonary arteries. The upper MAPCA was small.

Percutaneous coil embolization of large MAPCAs under fluoroscopic control is a useful technique. The Cook coil is introduced through an angiographic catheter which has been passed into the aorta by the Seldinger method, and positioned accurately in the orifice of the collateral vessel. When the coil is placed in the vessel, it assumes a helical configuration and stimulates thrombosis, leading to rapid occlusion[9]. As the procedure is done under local anaesthesia, the patient’s status can be accurately monitored[3]. Prior to attempted percutaneous occlusion, it is very essential to delineate all the MAPCAs upto the level of the diaphragm[5].

Small MAPCAs probably do not require intervention. Occlusion of the MAPCAs before opern heart surgery is important because otherwise there is excessive return to the left heart when the aorta is cross clamped on cardiopulmonary bypass, flooding the operative field. Although this can be dealt with by placing a large LV vent, postoperatively the patient may be difficult to wean off the ventilator because of excessive pulmonary flow. Also, postoperative cardiac failure

might be caused by a large left-to-right shunt via the persistent MAPCA[9,10]. Again, dissection and occlusion of MAPCAs at the time of cardiac surgery invites profuse bleeding from the raw tissues after heparinization, and so makes the surgery difficult.

It should be remembered that as the blood supplied by the MAPCA to the lungs may be an essential component of pulmonary blood flow, preoperative occlusion may cause arterial desaturation, and measures for instituting cardiopulmonary bypass. Again, pulmonary embolism due to migration of the coil into the pulmonary artery may occur[10].

Coiling of the MAPCAs may also be done after surgery to allow better growth of native pulmonary arteries[3]. It must be emphasized that coil embolization of MAPCAs and cardiac surgery is very much a team effort[9].

The aim in the case of tetralogy of Fallot with highly developed MAPCAs is to correct the cardiac anomaly and at the same time establish a single source of blood supply to the lungs from the pulmonary arteries, and to promote the growth of the native pulmonary arteries[4]. When 15 or more pulmonary segments are connected to the central pulmonary circulation, the addition of a unifocalization procedure to incorporate additional pulmonary segments adds little to the overall reduction in pulmonary artery resistance[3].

In this case, as the MAPCAs supplied part of the lung with dual supply, and as an estimated 17 bronchopulmonary segments were supplied by the pulmonary arteries, it was not felt necessary to centralize these vessels, but simply to make the surgery easier (by decreasing pulmonary venous return on cardiopulmonary bypass) and to prevent future pulmonary vaso-occlusive disease, the large lower MAPCA was occluded by coil embolization. Total corrective surgery was uneventful, and the patient has done well on follow-up except for an episode of congestive cardiac failure one month after surgery.

Major aortopulmonary collateral arteries occur in less than 5% of cases of tetralogy of Fallot with pulmonary stenosis. However, MAPCAs may constitute an important source of blood supply to the lungs in this situation. Although, clinically they usually do not give rise to problems preoperatively, when present, they need to be properly evaluated, and managed, usually before total correction is attempted. Percutaneous occlusion by coiling is a good option in this regard.

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