# Successful surgical revascularization of the congential left main coronary artery atresia in an infant

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**ABSTRACT**

Congenital anomaly of the coronary arteries is a rare disease occurring in 1%–2% of all congenital heart diseases. Atresia of the left main coronary artery is one of its least-frequently observed variations, with very few cases presented in the literature (Amaral et al.,2000; Gebauer et al.,2008). The prognosis is unfavorable because it results in myocardial ischemia or infarction, and even sudden cardiac death. Surgical revascularization of the left main coronary artery is recommended in most patients with atresia of the left coronary artery because of unfavorable clinical outcome, of which only a few cases have been reported for successful surgical revascularization (Sunagawa et al., 2005; Gebauer et al., 2008).

**Keywords:** Left main coronary artery atresia, left anterior descending artery, left internal mammary artery, successful revascularization.

**INTRODUCTION**

Coronary and aortic root anomalies represent a small but interesting group of malformations that may occur alone or in association with structural heart disease[1]. Congenital anomalies of the coronary arteries occur in 1-2% of the population[2] of which atresia of the left main coronary artery is the least observed variation[3].In true left main coronary artery atresia as defined by Musiani and colleagues there is no left coronary ostium and no left main stem[4]. Recognition of the coronary artery anomalies requires a high index of suspicion and prognosis of this anomaly is grave.Timely intervention by surgical revascularization of the left main coronary artery is recommended.A very few cases of congenital atresia of left main coronary artery with successful surgical revascularization have been reported[5].

**CASE REPORT**

A 4 months old male infant weighing 5.5 kgs presented with symptoms of hurried breathing and recurrent respiratory tract infections since two months of age.Clinical examination revealed heart rate of 118/min regular, respiratory rate of 40/min with no evidence of cyanosis, edema, pallor, lymphadenopathy. Systemic examination was within normal limit. Chest x-ray showed cardiomegaly with prominent bronchovascular markings (Figure 1). ECG showed equivocal signs of ischemia. 2D ECHO revealed dilated cardiomyopathy, severe LV dysfunction. **(LVEF – 18 %.)** with hypokinesia of septal basal and anterolateral wall of left ventricle. Origin of left main artery was not visualized on angiography.No features of ALCAPA were noted. Single right coronary artery with atresia of LMCA was seen. Faint filling of LAD and circumflex were noted through filling of intercoronary collaterals was observed (Figure 2).Child was admitted to PICU and administered Inj. Dobutamine 5 ugm/kg/min. Syp. Frusemide 2.5 mg bd. Syp.Dixin 0.25 1ml bd. Blood Investigations were normal.Procedure planned was Surgical revascularization of left main coronary artery (left internal mammary to left anterior descending artery graft) with cardiopulmonary bypass.

**Anesthetic Management:**

Pre-operatively, surgical and anesthetic options were discussed and after a thorough pre-anesthetic evaluation it was decided to proceed for on pump beating heart coronary artery bypass graft. Patient was kept NBM for 4 hours prior to surgery. Patient was premedicated Inj. Ketamine 25 mg + Inj. Glycopyrollate 0.05 mg + Inj. Midazolam 0.5 mg i.m. 22 G i.v line was secured on the right hand. Goals were to Maintain stable hemodynamics mean arterial pressure > 45 mm Hg and avoid tachycardia. Patient was preoxygenated with 100% O2 for 3 mintues and induced with Inj. Ketamine 10mg +Inj. Fentanyl 50 ugm +Inj. Midazolam 0.5 mg + Inj. Vecuronium 1mg follwed by Inj Ceftriaxone 150mg+ Inj Amikacin 50 mg and Inj. Methyl Predisolone 150 mg i.v.Nasotracheal intubation with ETT number 4.5 uncuffed was done. Right femoral artery was canulated with 22 G single lumen catheter. Right IJVwas canulated with 4.5 Fn 6 cm triple lumen catheter.

Patient was ventilated on Pressure Controlled mode with FiO2 – 70 % (O2 + Air + Sevoflurane) and respiratory rate of 34/min. Monitors attached were ECG , IBP, CVP, Temperature, and Pulse oximeter. LIMA was harvested. Inj Heparin 20 mg i.v was given. (ACT 570). Aortic and RA cannulation was done .CPB

initiated and temperature was drifted to 320C. LIMA to left anterior descending artery grafting was carried out on pump beating heart with 1 mm shunt without cardioplegia (Figure 3).CPB time was 26 minutes. Priming volume – 426 ml including 250 ml of PCV.Esmolol 0.5mg/kg was used to control heart rate during grafting.Inj. Dobutamine 5 ugm/kg/min and Inj. NTG 1-2 ugm/kg/min was started.Inj. Protamine 25 mg used to reverse heparin (ACT 144s). Patient shifted to intensive care unit and ventilated for 24 hours and later extubated (Table 1). Postoperative period was uneventful.Inotropes were continued till post-operative day 7 and patient was discharged on day 10. Patient was advised digoxin, furosemide, aspirin and enalapril on discharge. Post-operative echo revealed an increase in EF - 30 %.CT confirmed the patency of the graft.

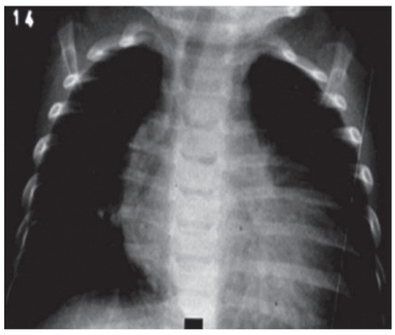

**Figure 1: Chest x-ray**

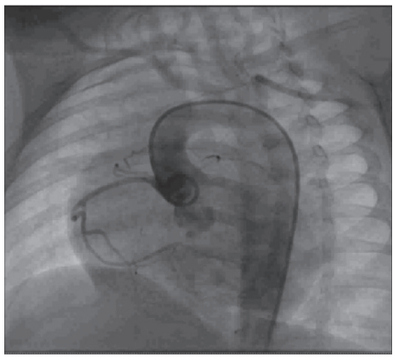

**Figure 2: Angiography.**

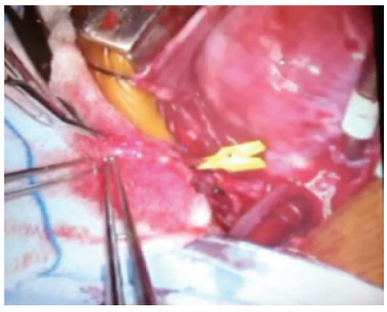

**Figure 3: Left internal mammary harvesting.**

## DISCUSSION

True LMCA atresia is indeed a rare condition, with only 64 reported cases that we know of.The incidence of all coronary anomalies is low in the general population, ranging between 0.46 and 1.55%; Yamanaka and Coll[6] identified 1,686 anomalous cases in a review of 1,26,595 coronary angiograms; 56 of these patients had a single coronary artery, but no cases of true LMCA atresia were found. Infants mostly present with failure to thrive and myocardial infarction, while children and adolescents often have syncope as a presenting symptom.Usually LMCA occurs as an solitary lesion but it can be associated with biscuspid aortic valve supravalvular aortic stenosis[7,8], pulmonary stenosis and should be particularly differentiated from ALCAPA[9]. As LMCA atresia carries an unfavorable prognosis and medical therapy does not seem to help, we think that surgical revascularization is the procedure of choice.Two methods have been described in children CABG using IMA/ SVG or reconstruction of the LMCA with aortic wall / pericardial patch.In pediatric patients the long-term results of CABG can be questioned, especially if saphenous vein grafts are used, although good results are reported with simple IMA grafts to the LAD in children.[10,11,12].In our case successful revascularization was done of the LIMA graft to the LAD artery on pump (beating heart) which according to pubmed is the first case of its kind from India.

## CONCLUSION

Congenital atresia of the left main coronary artery is a rare disease. It should be considered when diagnoses of dilated cardiomyopathy or anomalous origin of the left coronary artery from the pulmonary trunk in infants or in adults suspected of having coronary insufficiency are made. Myocardial revascularization surgery with grafting of the internal thoracic artery seems presently to be the method of choice; however, the possibility of surgical angioplasty should be considered. Our case according to pubmed search is the first reported case of successful revascularization of left main coronary artery atresia in an infant from India.

Given the high risk related to the presence of LMCA atresia and in view of the uniformly good results obtained by CABG, we recommend the above mentioned anesthetic technique for safe surgical revascularization in pediatric patients with LMCA atresia.

**Table 1- Intraoperative Events**

| TIME | EVENTS | CVP | ABP (mmHg) | PR | SPO2% | INPUT-OUTPUT |
|---|---|---|---|---|---|---|
| 8:15 am | Premedication | - | - | 110 | 99.9 | Kidrolyte 2ml/kg/hr. |
| 8:50 am | Heparin administered | 8 | 76/40 | 123 | 99 | 10 ml |
| 9:15 am | On CPB | 8 | 81/46 | 115 | 99 | 10 ml |
| 9:43 am | Off CPB | 5 | 69/43 | 118 | 99 | 30 ml |
| 9:50 am | Protamine administered | 7 | 82/58 | 110 | 99 | 30 ml |
| 10:30 | Shifted to PITU | 9 | 81/53 | 116 | 99 | 50 ml |

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