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TRANSTHORACIC 3 DIMENSIONAL REAL TIME ECHOCARDIOGRAPHY FOR THE DIAGNOSIS OF (ALCAPA) ANOMALOUS LEFT CORONARY ARTERY FROM PULMONARY ARTERY

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Contribution

SA conceived the idea and designed the case report. IS, MA collected pictures and helped in final draft. SA did review. All authors contributed equally to the submitted case report.

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ABSTRACT

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare but clinically significant form of congenital heart disease, usually causing myocardial dysfunction and heart failure in infancy. Approximately 90% of patients die within first year of life if left untreated. Cardiac surgery (re implantation of left coronary artery to aorta) is performed immediately after diagnosis. Usually the diagnosis is suspected on transthoracic echocardiography (TTE) and then confirmed by coronary angiogram. We are describing the use of real time 3 dimensional echocardiography (RT3DE) for confirmation of the diagnosis of ALCAPA in our 2 months old patient. The RT3DE images were reviewed and appreciated by the cardiac surgeon. Patient had successful ALCAPA repair without being subjected to invasive procedure of coronary angiography

Key Words: ALCAPA, RT3DE, Echocardiography, Coronary angiography

INTRODUCTION

Coronary arteries are the first branches of aorta and supply oxygen rich blood to the myocardium. ALCAPA is a rare congenital abnormality with an incidence of 1 in 300,000 live births.[1] ALCAPA is also known as Bland-White–Garland syndrome.[2] The left coronary artery (LCA) arises from pulmonary artery, thus carry oxygen poor blood therefore the myocardial oxygen demand is then maintained by acquiring collateral connections from the right coronary artery (RCA). After birth as the pulmonary vascular resistance decreases, the blood from the right coronary artery will get diverted into pulmonary artery via its collateral connections with the LCA. This leads to coronary steal phenomenon, resulting in poor myocardial perfusion, as well as left ventricle volume overload from the left to right shunt.

TTE findings of dilated and dysfunctional left ventricle, shiny hyper echogenic papillary muscles, mitral valve regurgitation along with difficulty in visualization of the origin of LCA from the aortic sinus and dilated RCA would raise the suspicion of ALCAPA. Cardiac catheterization and coronary angiography was the routine for confirmation of the diagnosis before surgery. Current advances in echocardiography devices have improved our capacity to diagnose complex congenital cardiac lesions. We describe the use of RT3DE for confirmation of the diagnosis of ALCAPA in our patient without subjecting to invasive procedure of coronary angiography

CASE REPORT

Two months baby boy presented with 2 weeks history of poor feeding and fast breathing.

No history of febrile illness. Physical examination revealed weight 4 Kg, length 57cm, BP 72 /45 mmHg. Respiratory rate of 46 breaths per minute and heart rate 138 beats per minute. He was pink in room air. Peripheral pulses were palpable. He had gallop rhythm on auscultation. Electro cardio gram showed deep Q wave in lead I, AVL, V4, V5 and V6.

Transthoracic echocardiogram performed using I.E 33 Philips system; Real time three dimensional(RT3DE) echo performed using X-2-7 transducer.

TTE findings: Dilated left ventricle with moderately decreased systolic function, Ejection Fraction of 45 %. Echogenic papillary muscles and Mitral valve chordae with severe mitral valve insufficiency, dilated Left atrium and prominent right coronary artery arising normally from right coronary sinus of aorta. LCA origin could not be confirmed from the Aorta but there was communication between LCA and Main pulmonary artery (Figure 1).

RT3DE full volume as well as full volume color images were obtained.

The origin of Left coronary artery from the main pulmonary artery was clearly demonstrated with 3 dimensional perception as well as the color flow from LCA to MPA on RT3DE. Further cropping of the MPA clearly showed the orifice of the LCA origin situated in the posterior pulmonary sinus of pulmonary valve. (Figure 2)

During hospital course; based on the RT3DE findings the baby was accepted for cardiac surgery without being subjected to cardiac catheterization. The above findings were confirmed in the operating room. Patient had successful ALCAPA repair and discharged in a week time.

Figure 1: Showing 2Dimensional echoA-Apical 4 Chamber; Dilated Left Atrium and Left Ventricle, Echogenic Chordae of Mitral Valve. B- Apical 4 Chamber Color Map ,Moderate to Severe Mitral Insufficiency. C- Parasternal Short Axis Color Map Showing Prominent Right Coronary Artery. D- Parasternal Short Axis Color Map Showing Left Coronary Artery Connected to Main Pulmonary Artery Marked with Astrix. (Ao –Aorta, PA-Pulmonary Artery, RCA–Right Coronary Artery ,LAD-Left Anterior Descending Artery , LCX-Left Circumflex Coronary Artery.

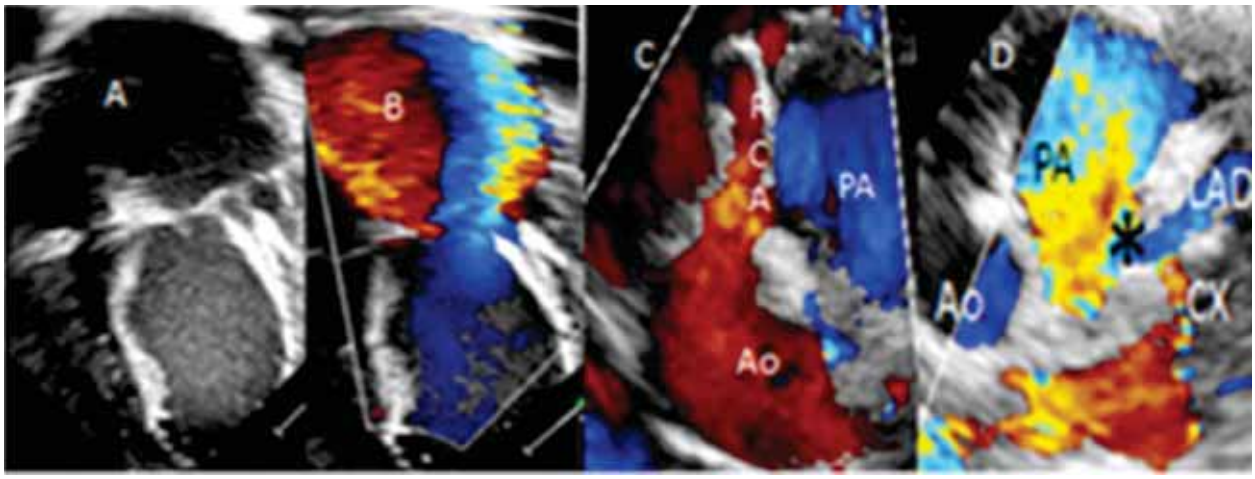
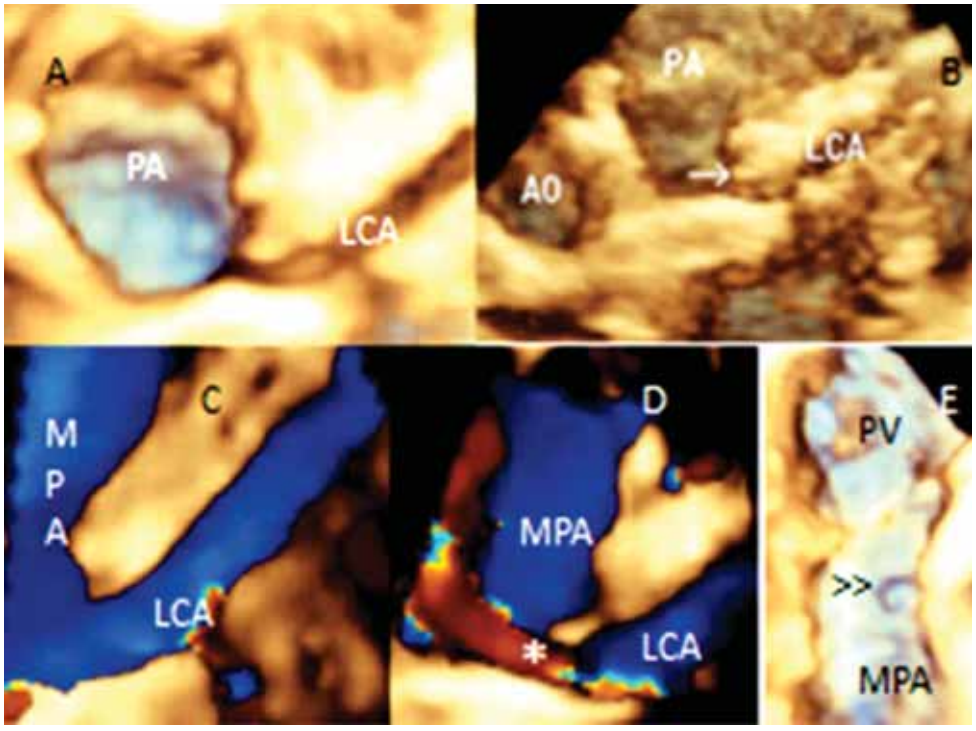


Figure 2: Showing RT3DE . A and B : 3DRTEImages Taken from Parasternal Short Axis Views Showing Origin of LCA from MPA. C and D: 3DRTE with Color Showing Retrograde Flow from LCA into MPA ,marked *. E: 3DRT Image Sagittal Section of MPA Showing the Orifice of LCA Marked Double Open Arrow Head. PV (Pulmonary Valve).



DISCUSSION

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare but clinically significant form of congenital heart disease, usually causing myocardial dysfunction and heart failure in infancy. This anomaly occurs in about 1 in 300,000 children (0.5% of those with congenital heart disease),but remains one of the more common causes of pediatric myocardial dysfunction. Correct diagnosis of ALCAPA in a timely manner is important for patient's management, regardless of the age at presentation.

Cardiac catheterization with angiography has traditionally been the imaging modality used for the diagnosis. However, as echocardiographic techniques have improved, echocardiography can provide diagnostic imaging without the need for more invasive imaging in ALCAPA. Although age-related differences in echocardiographic markers for ALCAPA have been described, but use of 3D echocardiography for the confirmation of ALCAPA in infants has not been reported.[3]

CONCLUSION

Transthoracic RT3DE is feasible and reliable noninvasive imaging modality for diagnosis of ALCAPA in infants.

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