Indian Heart Journal xxx (2017) xxx–xxx

Contents lists available at ScienceDirect

Indian Heart Journal

journal homepage: www.elsevier.com/locate/ihj

Original Article

Cardiac MR, myocardial scar and coronary flow pattern in ALCAPA

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ARTICLE INFO

Article history:
Received 26 May 2016
Accepted 10 August 2017
Available online xxx

ABSTRACT

Background: Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a very rare congenital heart defect characterized by myocardial ischemia and ultimately scaring. The scar burden will determine eventual recovery of left ventricular function after corrective surgery.
Material method: All patients with proven diagnosis of ALCAPA and who underwent treatment at present centre were included. Detail echocardiography and cardiac magnetic resonance imaging (CMR) (delayed Gadolinium enhancement) was performed before and after surgery.
Results: There were 4 patients (3 females, age group 3 months to 3 yr, follow up 6months to 20months.) There was no peri operative mortality. All patients had significant improvement in symptom class and LVEF(increase of more than 10%) when evaluated at last follow up. Three patients has pre operative CMR and 3 post operative CMR. All patients had improvement in post operative LVEF, but >50% was observed only in one patient who had less than half thickness delayed gadolinium enhancement. The right coronary flow pattern were unique to disease. The left coronary flow pattern were had significant variation and could predict extent of scared myocardium.
Conclusion: Ischemia in ALCAPA can lead to myocardial scarring even in early infancy. The recovery in left ventricular function is a closely related to scar burden. Coronary flow patterns are unique and give useful insight into disease process and natural history.

1. MNUSCRIPT

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) leads to ongoing myocardial ischemia and scarring. Extent of pre operative myocardial scar will determine recovery in left ventricular function after surgical correction and long term prognosis. Cardiac magnetic resonance imaging with delayed gadolinium enhancement (CMR) is a well established diagnostic modality in adults to evaluate viability of myocardium and scar burden. To best of our knowledge there is a little work done in pediatric substrate in this respect particularly in ALCAPA.[1–9]

Changes in right coronary artery (RCA) pulse wave coronary flow pattern (PWCFP) are diagnostic for ALCAPA.[1–3,10–12] However, there is very little understanding about change in PWCFP of left coronary artery (LCA) as well as its prognostic significance. The present study tries to establish the extent of myocardial scar with CMR and correlate it with PWCFP of LCA.

2. Material and method

This is a single centre observational study. Informed consent was obtained from all patients. It is approved by ethics committee of the hospital. All consecutive patients who underwent corrective surgery for ALCAPA were included. Baseline echocardiography was performed with Philips Epiq 7. Coronaries were assessed in modified short axis, parasternal long axis view and apical 4 chamber view. PWCFP was recorded at lowest Naquest limit with wall filter kept at low. CMR was performed with Philips 3T (Ingenia, Erlangen, Netherlands). Baseline and post Gadolinium enhanced CMR images were obtained. Pre surgical CMR was done on the day of surgery. Post surgical CMRs were done at interval of 7 days to 9 months. Improvement in LVEF more than 10% and functional class by I was defined as successful outcome

Off the 4 patients, 3 (age <1 yr) underwent left subclavian to LAD anastomosis. A 3 yr old girl underwent left internal mammary artery to LAD anastomosis. All surgeries were performed off pump. (without use of heart lung machine). The anastomotic site was fashioned close to left main coronary artery (LMCA) bifurcation so as to allow adequate left circumflex perfusion. LMCA was legated close to its origin from pulmonary artery. Post surgical CMR was

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http://dx.doi.org/10.1016/j.ihj.2017.08.004

performed using same protocol. The study has been approved by ethics committee of hospital.

3. Results

Patients characteristics, baseline and post operative echocardiography CMR findings and surgical details are described in Table 1. Coronary pulse wave Doppler are shown in Image 1. There were no perioperative deaths and none patient required extracorporal membrane oxygenation support. Mean time of ventilation was 45 hrs days (18 day to 90hrsdays). Inotropic support was required for mean 67 hrs (27 hrs to 117 hrs,) There were no significant perioperative complications. None patient required ECMO support. There was improvement in left ventricular ejection fraction, grade of MR and functional class for all the patients. Except one patient all had residual scar on follow up. However, it reduced on follow up. (Image 2,3,4)

4. Discussion

In a new born with ALCAPA, pulmonary artery pressure is the perfusion pressure of LV myocardium. Without any significant intracardiac shunt(ventricular septal defect, aortopulmonary window, patent ductus arteriosus etc) pulmonary artery pressure would drop over a period of weeks as a natural process. The resultant impairment in tissue perfusion leads to ischemia and triggers development of epicardial and endomyocardial collaterals between RCA and LCA. Former collateral are responsible for systolic flow reversal in LCA and characteristic higher systolic flow in RCA. The latter feed left coronary mainly during diastole as they are compressed by contracting myocardium. In spite of extensive collaterals development, communication between LCA and PA decompresses coronaries, worsens tissue perfusion and aggravates ischemia. Long standing ischemia ultimately leads to myocardial infarction and fibrosis. Poor diastolic flow reversal may be result of fibrotic obliteration of intra myocardial collaterals as a result of scarring. In natural history, with progressive scarring, not only baseline LVEF will be low but also diastolic component of LAD flow will be lost. Also, the recovery in myocardial function after surgery will be suboptimal. Thus loss of diastolic component and increase of systolic component suggest scarred myocardium, low baseline LVEF and sub optimal recovery after surgery. [10]

The peculiar right coronary flow pattern is diagnostic of ALCAPA in adults. [1–3] To the best of our knowledge it has been demonstrated in early infancy for the first time. (Fig. 1d,). It definitely adds to diagnostic utility of same and should be specifically sought for in every patient with depressed left ventricular systolic dysfunction irrespective of age and even in early infancy. It will of interest to know when this pattern develops after birth.

Though of similar age group, the flow patterns, CMR and improvement in LVEF of 3 vs 2/4 were strikingly different. Late diastolic peaking of flow reversal in LAD may be result of delayed relaxation of ischemic but viable myocardium (Patient 3). While inverted 'U' may suggest component of fixed obstruction along with dynamic. It may also suggest that, intra myocardial collaterals develop earlier than epicardial. Presence of significant flow from epicardial collaterals may also be an indicator of diminished flow in myocardial collaterals and scarred tissue. Very minimal diastolic flow in Pt 1, was result of extensive scar because she presented late. The probable reasons for early onset of scarring would be 1) run off from LMCA or 2) lesser collateral development or 3) altered tissue response to ischemia. Lange et al. has also reported that post operative outcome has been influenced by baseline LVEF, age, height and weight at presentation.[12,14]

Secinaro et.al., found that in post ALCAPA repair patients, presence of late gadolinium enhancement was suggestive of coronary occlusion. Post operative LVEF of all of the study patient was more than 60%.[8] While in a pre and postoperative comparative CMR study by Latus wt.al, only 2 of 8 patients had baseline scar (10

Table 1
Pre and post operative details.

No	Age sex	Pre operative Echocardiography/Cardiac MR	Coronary flow on pulse wave	Operative details (off pump)	Post operative
1	3 Yr F	LCA from facing sinus, no collaterals in IVS, LVEF 11%, moderate mitral regurgitation, Positive longitudinal strain of apical segments LDESV/LVEDV:173/194 ml/m^2	Predominant systolic flow reversal and mild diastolic flow. (Fig. 1a)[8]	Left internal mammary artery to LAD anastomosis and LMCA ligation	CMR (9 month) LVEF: 31%, trans mural enhancement of entire apex, anterior segments and mid and distal lateral wall. Mild MR (Fig. 4) LDESV/LVEDV: 96/139 ml/m^2
2	4 mth F	LCA from non facing sinus. collaterals in IVS,Moderate MR, Cardiac MR: LVEF 16%, >50% contrast enhancement of apex, anterior and lateral wall LDESV/LVEDV:150/179 ml/m^2	Prominent diastolic (inverted U) and mild systolic flow reversal (Fig. 1b)	LMCA ligation and left subclavian artery to LAD anastomosis	Cardiac MR (4 weeks): LVEF 39%, more than 50% contrast enhancement of entire apex, mid anterior and apical lateral wall. No contrast enhancement of Basal and mid lateral segments and improvement in contractility. Mild MR ECHO (15 months): LVEF 43%, apical segments hypokinetic, mild MR LDESV/LVEDV: 74/129 ml/m^2
3	4 mth M	LCA from facing sinus., collaterals in IVS, severe MR, LVEF 28% LDESV/LVEDV:131/182 ml/m^2	Prominent diastolic flow reversal (late diastolic peak). Insignificant systolic component (Fig. 1c)	LMCA ligation and Left Subclavian artery to LAD anastomosis	Cardiac MR: LVEF: 51%, no regional wall motion abnormality, moderate MR, subendocardial enhancement of apex and apical lateral wall. (1 week), (Fig. 3) ECHO(16 month): LVEF 62%, good contractility of all segments, no MR LDESV/LVEDV: 61/123 ml/m^2
4	3 mth M	LCA form facing sinus, Collaterals in IVS, severe MR, LVEF 13% Cardiac MR: >50% contrast enhancement of apical segments, mid anterior and lateral wall LDESV/LVEDV: 171/197 ml/m^2	Prominent diastolic flow reversal and significant systolic flow reversal (findings similar to Pt 2)	LMCA ligation and Left Subclavian artery to LAD anastomosis	ECHO (18 months): LVEF 43%, mid anterior anteroseptal and aplical segments severely hypokinetic. No MR (Fig. 2) LDEDV/LVESV: 58/109 ml/m^2

Please cite this article in press as: P. Bhalgat, et al., Cardiac MR, myocardial scar and coronary flow pattern in ALCAPA, Indian Heart J (2017), http://dx.doi.org/10.1016/j.ihj.2017.08.004