Cardiac magnetic resonance imaging in Conotruncal anomalies and complications after their postoperative repair

Abrar Elsallakh (MSc)1 , Emad M. Mashaly (MD)1, Sameh A. Khodair (MD)1, Wesam Elmozy (MD, PhD)2, Rehab Elnagar (MD)1, Rasha Mahmoud Dawoud (MD)1

1 Radiology Department, Faculty of Medicine, Tanta University, Egypt
2 Aswan Heart Center (Magdi Yacoub Foundation) Aswan, Egypt
Radiology Department, Faculty of Medicine, Cairo University, Egypt
Corresponding author: Abrar Elsallakh
abrarelsallakh@gmail.com

Abstract:
Background: Conotruncal anomalies refer to a broad category of complex congenital heart diseases with different surgical management options; cardiac magnetic resonance imaging has expanded its role in diagnosis, and follow-up and markedly integrated in clinical practice. The purpose of this study is to highlight the role of cardiac magnetic resonance imaging (CMR) in the assessment of these anomalies and their post-surgical consequences. **Results:** Cardiac magnetic resonance imaging was performed for 50 patients with conotruncal abnormalities ranging in age from 7 months to 47 years. Tetralogy of Fallot was the most prevalent conotruncal abnormality accounting for 44% of cases. CMR identified variable post-operative complications correlated to each type of contruncal anomaly and its surgical procedure. **Conclusion:** CMR plays an essential role during post-operative follow up of conotruncal anomalies providing precise information on cardiac anatomy, ventricular function, valvular condition, and myocardial fibrosis that would improve the clinical decision; not being hampered by acoustic window of echocardiography or use of ionizing radiation as in CT and catheter angiography. **Key words:** Cardiac magnetic resonance (CMR), Conotruncal anomalies.

Introduction:

Conotruncal anomalies are a broad category of congenital heart diseases involving the ventricular outflow tracts; include tetralogy of Fallot, transposition of the great arteries, double-outlet ventricle, truncus arteriosus, type B interrupted aortic arch, aortopulmonary window, and anatomically corrected transposition of the great arteries [(1)].

Echocardiography and cardiac catheterizations have been the traditional modalities for imaging of CHD including conotruncal anomalies. The last decade had shown the rise of MRI and computed tomography as an accepted imaging modalities for CHD. Cardiac magnetic resonance imaging has been the source for important anatomical and physiological assessment in congenial heart diseases [(2,3)].

Echocardiography is currently used as the initial noninvasive imaging study for almost all patients with known or suspected Conotruncal anomalies. MRI is unlikely to replace echocardiography as the first diagnostic procedure owing to its availability and affordability, yet MRI can demonstrate cardiovascular anatomy without the limitation of acoustic windows or ultrasound penetration of the body [(4,5)].

Furthermore, MRI can provide tomographic images in any imaging plane and with a wide field of view as well as displaying extra-cardiac anatomy, and being the

modality of choice for assessment of size and function of right ventricle in patients with Conotruncal anomalies, providing accurate quantitative data needed to make precise clinical decisions [6,7].
The purpose of this study is to highlight the role of cardiac magnetic resonance imaging (CMR) in assessment of these anomalies and their post-surgical consequences.

Patients and methods:
This prospective study was performed between August 2020 and July 2022 on 50 patients previously diagnosed or suspected by echocardiography to have Conotruncal anomalies.
The patients varied in age from 7 months - 47 years (29 men and 21 women). All of them underwent cardiac MRI examination after fulfilling our research sample and inclusion requirements. Both informed approval and ethical committee permission were granted.
CMR examination was carried on using Siemens Magnetom Sola MRI Scanner, 1.5 Tesla (T).
All patients were examined in supine position, headfirst. The patients were offered ear plugs to reduce repetitive gradient noise.
Children below the age of 4 years or older and unable to standstill or co-operate adequately were examined under general anesthesia with mechanical ventilator (12 patients).
ECG leads were optimally placed. A suitable imaging coils was selected to optimize the signal to noise ratio. A pediatric coil was used in children, adequate coil coverage and placement was confined by reviewing the localizing images.
The pear sensor was used when breath-hold sequences were applied. It was placed over the maximum area of respiratory movement under the coil.
Imaging protocol:
Basic congenital CMR protocol included SSFP localizing images, ECG triggered cine images, 3D whole heart with respiratory navigator and flow images; late gadolinium enhancement sequences were performed in 18 patients ten minutes following administration of contrast, with T1 scout has been used to determine the ideal inversion time for myocardial nulling.
The interpretation was done off-line through available workstation (Philips Medical System)
Flow quantification was done by outlining the ROI throughout the cardiac cycle and then presented as a graph of flow/time, where the backward, forward, net flow and regurgitant fraction were determined.
Calculation of ventricular volumes were done by evaluation of axial slices' stalk at end-systole and end-diastole by contouring their endocardial borders; providing quantitative parameters as end-systolic and end-diastolic volumes as well as ejection fraction and cardiac output.
Visual assessment of late gadolinium enhancement was enough, and no more additional processing was required.
Statistical method: IBM SPSS software version 21.0 (IBM Corp, Armonk, NY) was used for data analysis. Number and percentage were used to describe qualitative data. The normality of the distribution was examined using the Kolmogorov-Smirnov test. The range {minimum and maximum}, mean, standard deviation {SD}, median, and interquartile range {IQR} were used to characterize the quantitative information. Significant findings determined at 5% level.

Results:

In our study we enrolled fifty patients, with higher male percentage 58%. The patients' ages varied from 7 months to 47 years, The mean age was 14.5±10.35.

Tetralogy of Fallot, was the most prevalent conotruncal anomaly represents 44%, while the least prevalent were CC-TGA and "conotruncal anomalies associated with single ventricle physiology", each account for 6%, others were D-TGA 22%, truncus arteriosus 14%, DORV 8%.

The most common CMR indication was quantification of ventricular volumes, function and Op/Qs (representing 60%) while the least common was for assessment of LV mass (representing 4%), others include 1st baseline follow up, deterioration of ventricular volume &/or clinical status. **(Table1)**. Eighteen of our patients were planned to undergo contrast administration during their CMR study as being referred for their baseline follow up or for evaluation after deterioration of their clinical condition or ventricular function.

Table (1): Distribution of the studied Conotruncal patients according to CMR indications (n=50):

Indication	No.	Percentage (%)
Assessment of ventricular volumes and function	**30**	60%
Deterioration of clinical status	**7**	14%
1st visit	6	12%
Deterioration of ventricular functions	5	10%
Calculation of LV mass	2	4%

In this study, 14 out of the 18 patients (77.7%) had myocardial enhancement at the LGE sequences; the most common site for myocardial enhancement was VSD and trans-annular patch (85.7%) followed by insertion points (21.4%) and the least common site was left ventricular myocardial enhancement and RV free wall (7.14% for each) with the note that 3 cases had myocardial enhancement at more than one site **(Table 2)**.

Variable post-operative complications were identified by CMR with tricuspid and pulmonary regurgitation were the most prevalent ones with percentage of 90% and 76% respectively, others included right and left ventricular dilation, pulmonary stenosis, aortic regurgitation and residual VSD defects.

Our results were classified according to the post-operative sequel correlated to each type of conotruncal abnormalities and their surgical procedure.

Table (2): Distribution of enrolled 18 Conotruncal patients according to site of myocardial enhancement at late gadolinium enhancement (LGE) sequences:

Site of myocardial enhancement									
No enhancement		**VSD and Transannular patch**		**Insertion points**		**RV free wall**		**LV wall**	
No.	**%**	**No.**	**%**	**No.**	**%**	**No.**	**%**	**No.**	**%**
4	**28.57%**	**12**	**85.7%**	**3**	**21.4%**	**1**	**7.14%**	**1**	**7.14%**

Twenty-two of our patients were diagnosed to have tetralogy of Fallot; eighteen of them underwent trans-annular patch while remaining 4 underwent RV-PA conduit repair depending on their clinical severity and associated findings.

Multiple post-operative complications were shown in (Table3) with a significant statistical correlation between type of surgical repair and post-operative pulmonary and tricuspid regurgitation, occurring in all patients underwent trans-annular patch and 50% of cases underwent RV-PA conduit.

Two out of 22 repaired TOF patients underwent re-intervention in the form of pulmonary valve replacement following primary repair by transannular patch, therefore their results were excluded form tests of significance (Table 3).

Table (3): Distribution of the enrolled repaired TOF patients according to their type of operation and CMR post-operative sequel

Post-operative sequel		Tetralogy of Fallot (n=22)							
		Trans annular patch (TAP) (n=16)		RV-PA conduit (n=4)		X^2	^{MC}P	TAP followed by PVR(n=2)	
		No	%	No.	%			No.	%
RV Volume	Normal	1	6.3%	2	50%	4.804	P^{FE}	1	50%
	Dilated	15	93.8%	2	50%		0.088	1	50%
RV function	Normal	11	68.8%	3	75%	0.060	P^{FE}	0	0%
	Impaired	5	31.3%	1	25%		1.000	2	100%
LV Volume	Normal	10	62.5%	3	75%	0.220	P^{FE}	2	100%
	Dilated	6	37.5%	1	25%		1.000	0	0%
LV function	Normal	15	93.8%	4	100%	0.263	P^{FE}	1	50%
	Impaired	1	6.3%	0	0%		1.000	1	50%
PR	No	0	0%	2	50%	8.889	P^{FE}	1	50%
	Yes	16	100%	2	50%		0.032*	1	50%
PS	No	12	75%	2	50%	0.952	P^{FE}	1	50%
	Yes	4	25%	2	50%		0.549	1	50%
TR	No	0	0%	2	50%	8.889	P^{FE}	0	0%
	Yes.	16	100%	2	50%		0.032*	2	100%
AR	No	14	87.5%	2	50%	2.813	P^{FE}	1	50%
	Yes	2	12.5%	2	50%		0.162	1	50%
VSD	No	16	100%	3	75%	4.211	P^{FE}	2	100%
	Yes.	0	0%	1	25%		0.200	0	0%
RVOT	Stenosis	1	6.2%	0	100%	0.240	P^{FE}	0	0%
	Aneurysm	4	25%	0	0%		1.000	1	50%

Eleven patients were diagnosed to have D-TGA; 5 of them underwent atrial switch operation, 3 underwent Rastelli operation and 1 underwent arterial switch operation based on patient age and associated findings, while the remaining two patients were referred for pre-operative assessment of LV mass “deconditioned LV”; as was planned to undergo staged arterial switch operation by pulmonary artery banding (PAB).

Total surgical repair was preceded in 5 patients by urgent palliative percutaneous atrial septostomy (Rashkind) shortly after birth in order to allow adequate mixing of deoxygenated and oxygenated blood.

Many postoperative complications were detected as shown in (Table 4) with pulmonary stenosis was significantly correlated to Rastelli operation as being occurred in all patients.

Table (4): Distribution of the enrolled repaired D-TGA patients according to their type of operation and CMR post-operative sequel :

Post-operative sequel		D-TGA (n=9)						X^2	^{MC}P
		Arterial switch operation (n=1)		Atrial switch operation (n=5)		Rastelli operation (n=3)			
		No	%	No.	%				
RV Volume	Normal	1	100%	4	80%	2	66.7%	0.514	1.000
	Dilated	0	0%	1	20%	1	33.3%		
RV function	Normal	1	100%	2	40%	1	33.3%	1.440	1.000
	Impaired	0	0%	3	60%	2	66.7%		
LV Volume	Normal	0	0%	4	80%	0	0%	5.760	0.083
	Dilated	1	100%	1	20%	3	100%		
LV function	Normal	0	0%	4	80%	1	33.3%	3.060	0.288
	Impaired	1	100%	1	20%	2	66.7%		
PR	No	1	100%	2	40%	0	0%	3.600	0.287
	Yes	0	0%	3	60%	3	100%		
PS	No	1	100%	5	100%	0	0%	9.000	0.013*
	Yes	0	0%	0	0%	3	100%		
TR	No	0	0%	1	20%	0	0%	0.900	1.000
	Yes.	1	100%	4	80%	3	100%		
AR	No	0	0%	4	80%	0	0%	5.760	0.083
	Yes	1	100%	1	20%	3	100%		
Baffles	Leak	0	0%	1	20%	0	0%	--	--
	Stenosis	0	0%	3	60%	0	0%		
Myocarditis	No	1	100%	4	80%	3	100%	0.900	1.000
	Yes	0	0%	1	20%	0	0%		
Implanted coronary artery stenosis	No	0	0%	5	100%	3	100%	9.000	0.114
	Yes	1	100%	0	0%	0	0%		

Four patients were diagnosed to have DORV. Three types of operations were done based on great vessels relationship and VSD location namely, trans-annular patch, RV-PA conduit and Rastelli operation (each was done for 1 patient) while 1 patient was referred for pre-operative assessment of ventricular functions and calculation of Qp/Qs.

Pulmonary regurgitation was a sequel for all repaired DORV patients regardless their type of operation, while pulmonary stenosis was mainly related to post-Rastelli repair (Table 5).

Seven patients were diagnosed to have truncus arteriosus type I and all of them underwent total surgical repair with pulmonary regurgitation was an inevitable post operative sequel with subsequent RV dilation and tricuspid regurgitation.

CC-TGA: Three patients were diagnosed to have CC-TGA, only one of them underwent LV-PA conduit placement while the other two patients were referred for follow up of their systemic right ventricular function.

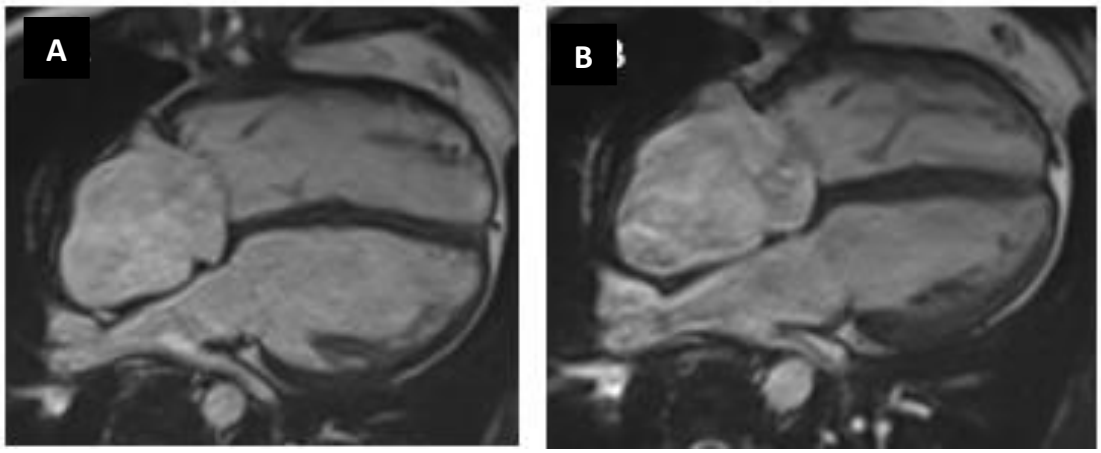
Three conotruncal anomalies (2 DORV and 1 TGA cases) were associated with single ventricle physiology, which includes unbalanced common A-V canal, multiple large non-septetable VSDs and mitral atresia, all of them underwent Glenn

shunt procedure with no veno-venous collaterals were detected as a post-operative sequel.

Table (5): Distribution of the enrolled repaired DORV patients according to their type of operation and CMR post-operative sequel

Post-operative sequel		DORV (n=3)							
		Trans-annular patch (n=1)		RV-PA conduit (n=1)		Rastelli operation (n=1)		X^2	^{MC}P
		No	%	No.	%	No.	%		
RV volume	Normal	0	0%	0	0%	0	0%	--	--
	Dilated	1	100%	1	100%	1	100%		
RV function	Normal	1	100%	0	0%	0	0%	3.000	1.000
	Impaired	0	0%	1	100%	1	100%		
LV volume	Normal	1	100%	1	100%	0	100%	3.000	1.000
	Dilated	0	0%	0	0%	1	0%		
LV function	Normal	0	0%	0	0%	0	0%	--	--
	Impaired	1	100%	1	100%	1	100%		
PR	No	0	0%	0	0%	0	0%	--	--
	Yes	1	100%	1	100%	1	100%		
PS	No	0	0%	1	100%	0	0%	3.000	1.000
	Yes	1	100%	0	0%	1	100%		
TR	No	0	0%	0	0%	0	0%	3.000	1.000
	Yes.	1	100%	1	100%	1	100%		
AR	No	0	0%	1	100%	0	0%	3.000 0	1.000
	Yes	1	100%	0	0%	1	100%		

Case Number 1 (Figure 1)



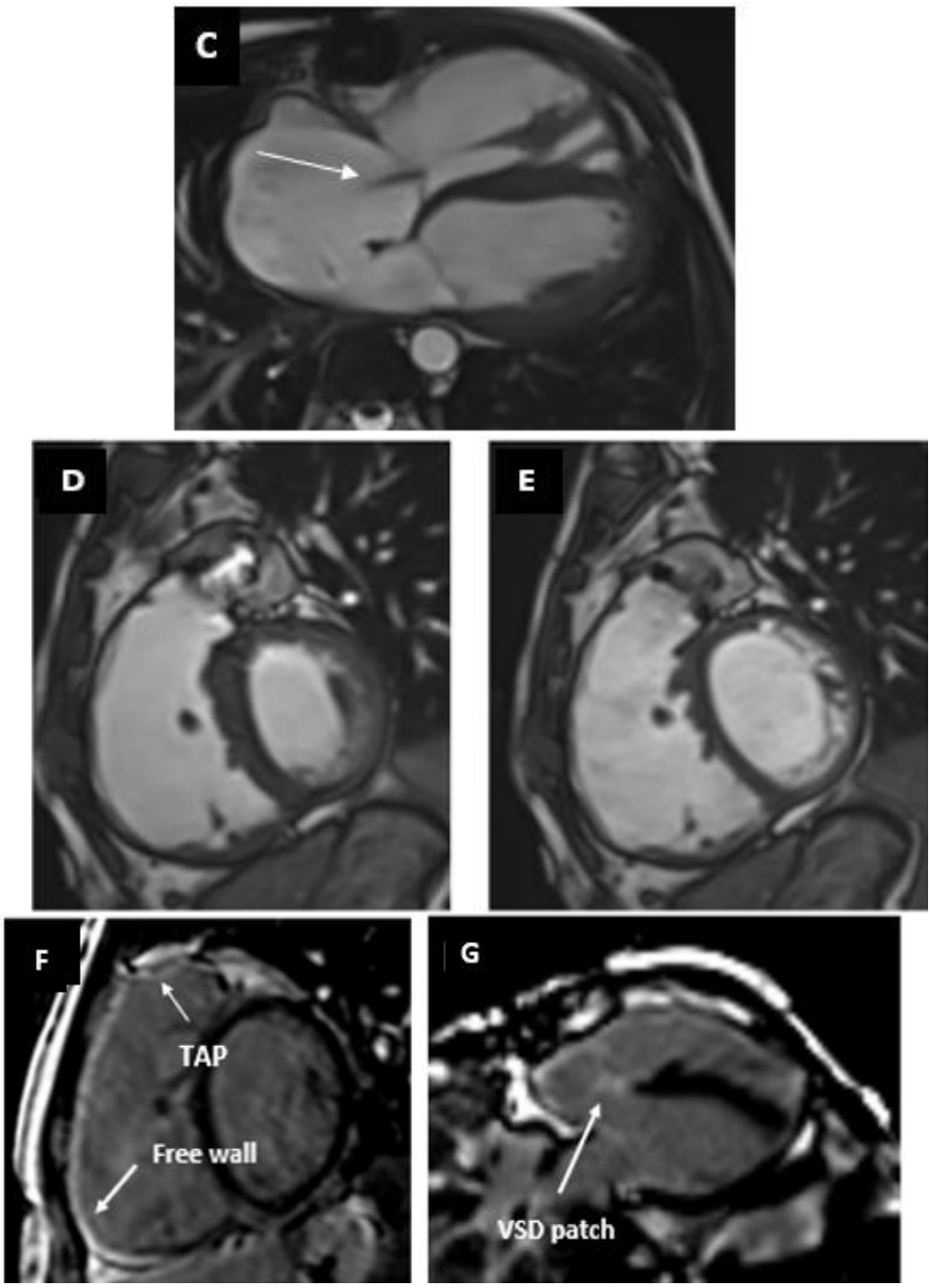


Figure 1: A 23-year-old male patient, had tetralogy of Fallot, underwent Trans-annular patch repair followed by bio-- prosthetic pulmonary valve replacement. 4-chamber cine images at end- diastole (A), at end-systole (B) showing RV dilation with impaired systolic function. Axial cine image (C) during ventricular systole showing dephasing jet of tricuspid regurgitation. RVOT cine images showing stenotic jet of neo-pulmonary valve in systole (D) with intact pulmonary valve at diastole showing no regurgitation in (E). Short axis (F) 3-chamber (G) (LGE) showing enhancement of RV free wall, trans-annular patch (TAP) as well as VSD patch

Case Number 2 (Figure 2)

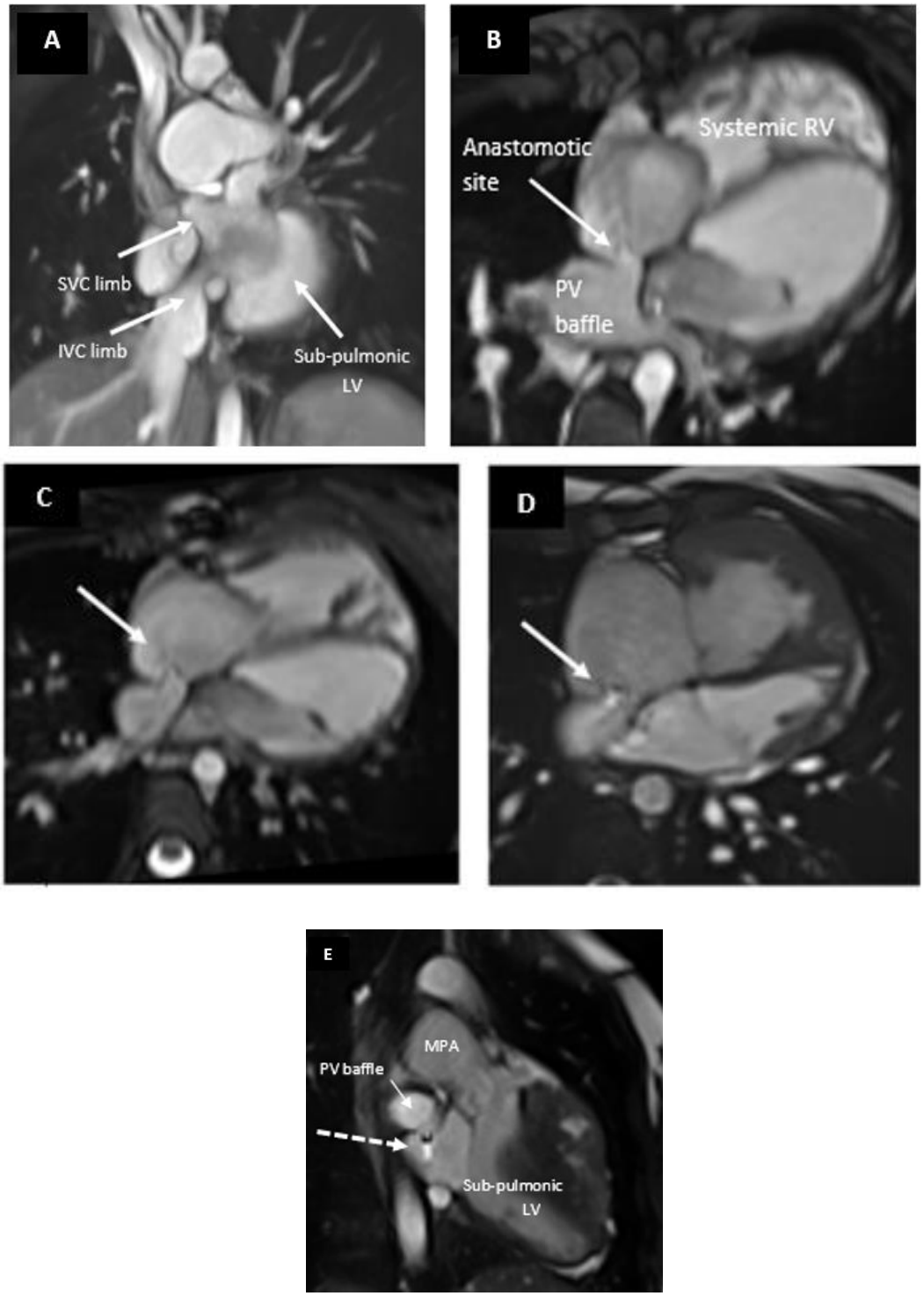
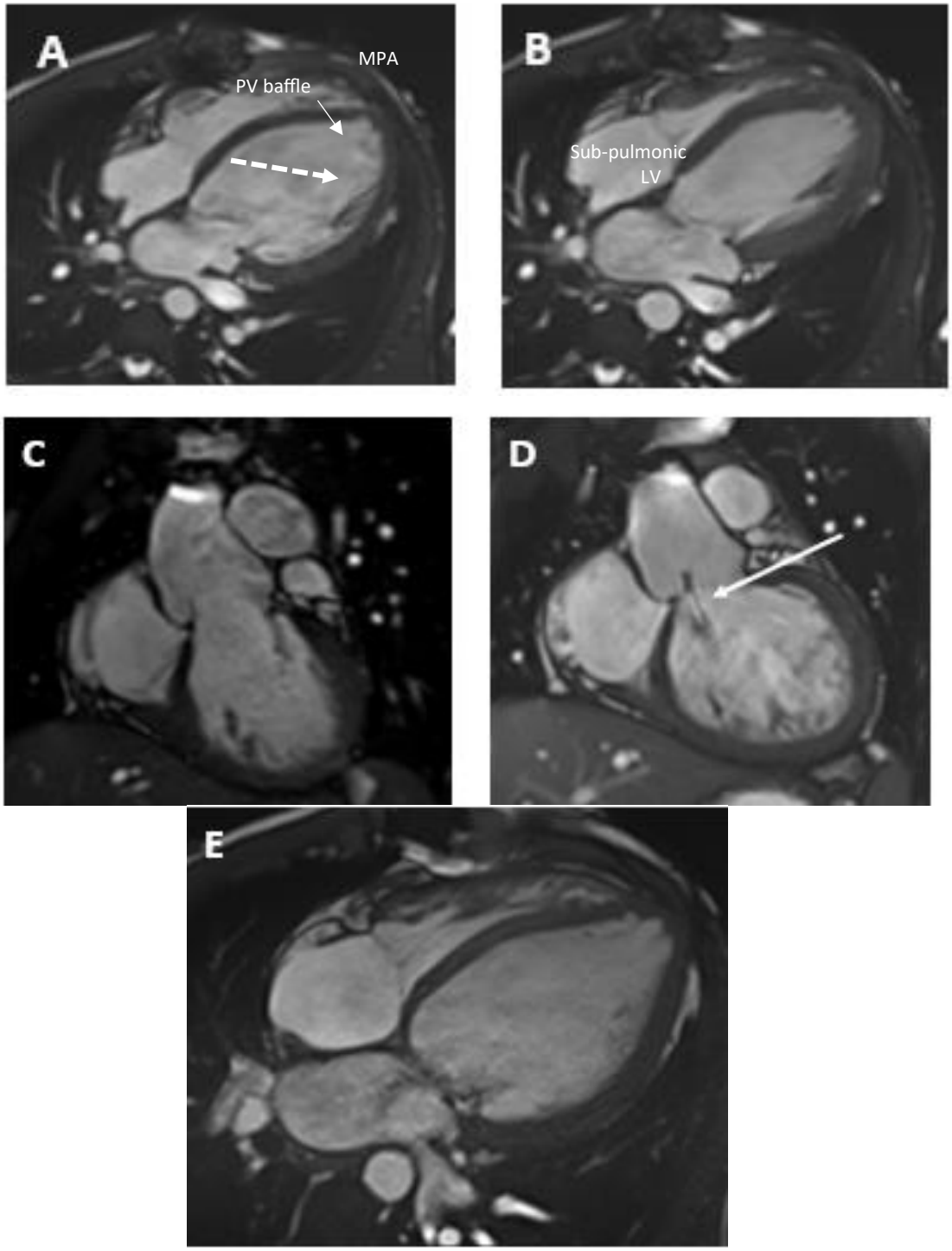
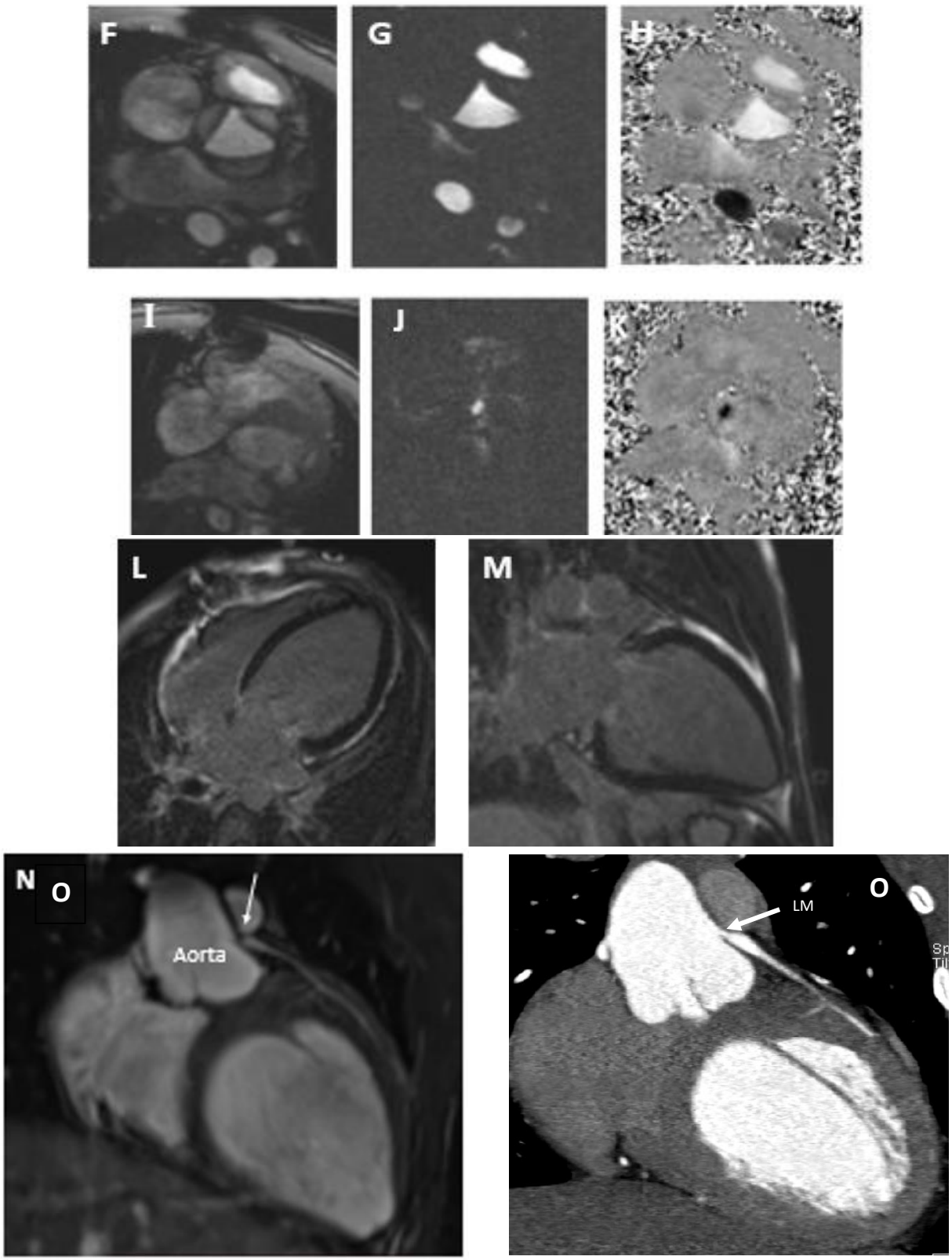


Figure 2: A 15-year-old female patient, had D-TGA performed atrial switch operation. 3D whole heart (A) coronal oblique MIP image showing SVC and IVC baffle into sub-pulmonic left ventricle via mitral valve , (B) axial oblique image MIP showing pulmonary venous baffle into systemic right ventricle via tricuspid valve. (C) 3D whole heart axial oblique image showing stenosis at the anastomotic site of pulmonary venous baffle, (D) axial cine image showing aliasing and stenotic jet at PV anastomotic site. (E): 3-chamber cine image showing the baffle leak jet (dashed arrow) from the pulmonary venous baffle to systemic baffle.

Case Number 3 (Figure 3)

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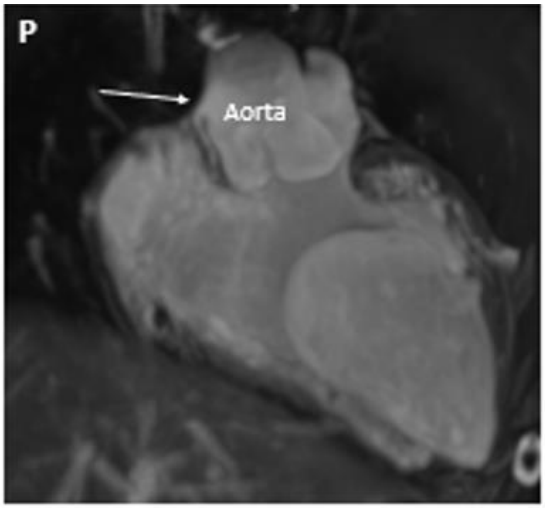

Figure 3: A 20-year-old male patient, had D-TGA , performed arterial switch operation, 4-chamber cine images at end-diastole (A), at end-systole (B) showing dilated left ventricle. LVOT cine images (C) at systole showing no stenotic jet along aorta, (D) at diastole showing dephasing jet of aortic regurgitation. (E): 4-chamber cine image showing dephasing jet of mitral regurgitation during systole. (F,G,H):Through-plane aortic valve flow at systole (Venc=1.5 m/s) showing no alising denoting no aortic stenosis. (I,J,K) Through-plane sub-aortic valve flow at diastole (Venc 1.5 m/s) showing aortic regurgitation. 4- chamber view (L), 3-chamber (M) showed no enhancement in late gadolinium sequences. 3D whole heart (N) showing signal drop off at ostium of re-implanted LM to neo-aortic root that showed LM ostial stenosis by CT (O), while (P) shows normal appearance of RCA anastomotic (arrows).

Discussion:

Conotruncal anomalies are a broad category of congenital heart diseases involving the ventricular outflow tracts; include tetralogy of Fallot, transposition of the great arteries, double-outlet ventricle, truncus arteriosus, type B interrupted aortic arch, aortopulmonary window, and anatomically corrected transposition of the great arteries [(1)].

Conotruncal anomalies refer to a group of congenital heart defects involving the outflow tracts of the heart and the great vessels. Developmental abnormalities of the embryonic conus arteriosus (infundibulum) and the truncus arteriosus may result in abnormal ventriculo-arterial alignments and connections, outlet septation defects, or outlet hypoplasia, stenosis, or atresia. The outflow tract of the embryonic univentricular heart begins with a common outlet that undergoes a complex, highly choreographed sequence of events that result in complete separation of the left and right ventricular outflow tracts, which lead into the aorta and main pulmonary artery. Numerous genes tightly control this process and migration of mesenchymal cells from the embryonic neural crest is critical to the development of these areas. Genetic errors and faulty migration of neural crest cells are likely responsible for many of the defects [(8,9)].

Cardiac MRI has evolved into a widely accepted clinical tool in congenital heart disease evaluation as it gives anatomical and functional data that surpasses catheterization and echocardiography provide accurate information regarding vascular and valvular flow as well as shunts quantifications, it also has the capability of evaluating extra-cardiac structures with high spatial resolution[(2)]. Furthermore,

cardiac MRI provides data reconstruction in different planes allowing visualization of complex cardiac heart diseases, not hampered by use of ionizing radiation [10,11].
This study conducted upon fifty patients, with higher male percentage 58%. The patients' ages varied from 7 months to 47 years. Tetralogy of Fallot was the most prevalent conotruncal anomaly represents 44%, while the least prevalent were CC-TGA and "conotruncal anomalies associated with single ventricle physiology", each account for 6%.
Compared to Samira Saraya et al, [12] who studied 42 patients of Conotruncal anomalies including 25 females and 17 males and their ages ranges between 6 months to 18 years and showed agreement as tetralogy of Fallot was the most common anomaly (represented 45%) and DORV was the least frequent (represented 7 %).
In this study, 22 patients had tetralogy of Fallot with trans-annular patch done for majority of cases (81.81%) while reminder of cases underwent RV-PA conduit (18.18%). This shows agreement with Davlouros et al, [13] who found that trans-annular patch was the most common used type of repair (52.8%) among the studied 36 patients of repaired TOF with while the second common repair was RVOT repair (44.5%) with conduit repair being the least common used type of repair (2.8%). The most common post-operative sequel in repaired TOF patients in our study, was pulmonary and tricuspid regurgitation (100% of all cases underwent TAP and 50% of cases of RV-PA conduit) with subsequent RV dilation (93.8% for cases underwent TAP and 50% of cases underwent RV-PA conduit), while pulmonary stenosis occurred in 25% of cases underwent TAP and 50% of cases underwent RV-PA conduit.
Regarding the post-operative sequel for repaired D-TGA patients in this study, post-operative pulmonary stenosis was significantly related to Rastelli operation, this was the same discussed by Mark Gerard et al, [14], who reported that re-operation for RVOT obstruction was most frequently performed for patients pos- Rastelli repair; while coronary artery stenosis was reported in our study as a post operative sequel for arterial switch operation, that was in agreement with Qing-Yu Wu et al, [15], who stated that coronary ostial stenosis after arterial switch operation is a potentially surgical life threatening consequence.
In DORV patients, surgical options were determined based on their pathophysiological sub-types, (transposition of the great arteries {TGAs} type, tetralogy of Fallot {TOF} type, ventricular septal defect {VSD} type or remote VSD type). In our study, two DORV cases were "TOF like" and they underwent trans-annular patch and RV-PA conduit while one patient was "TGA-like" and underwent Rastelli operation with their post-operative outcome were the same as previously discussed for TOF and TGA repair. Similarly, Adam Dorfman et al, [8] stated that repair for "TOF-like DORV" patients suffer from long-term consequences similar to those following Fallot repair. Likewise, in patients with Taussig-Bing type DORV, postoperative complications are comparable to those detected after TGA repair.
Our study enrolled 7 patients with truncus arteriosus, all of them underwent complete surgical repair and referred for post-operative assessment with pulmonary regurgitation and RV dilation being an inevitable post-operative complication. Adam Dorfman et al, [8] stated that CMR has an increasing role in evaluation of pot-operative truncus arteriosus for ventricular functions assessment as well as pulmonary and neo-aortic valve regurgitation fraction calculation and exclusion of residual shunts.

Limitation of the study:
This study has a limitation of smaller sample size as they were classified into multiple groups and sub-groups.

Conclusion:
In conclusion, CMR plays a crucial part during post-operative follow-up of conotruncal anomalies providing precise information on cardiac anatomy, ventricular function, valvular condition and myocardial fibrosis that would improve the clinical decision; not being hampered by acoustic window of echocardiography or use of ionizing radiation as in CT and catheter angiography.

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