Congenital Heart Disease

Presented by
Society for Cardiovascular Magnetic Resonance

1 Indications

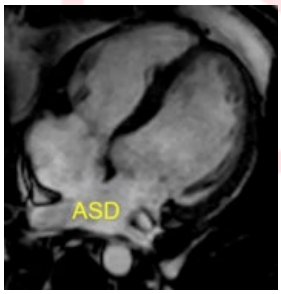
CMR is the gold-standard for assessment of cardiac function and size in pediatrics and adults with congenital heart disease (CHD). Current SCMR guideline and appropriate use criteria (AUC) guideline recommend use of CMR for routine surveillance in patients with repaired CHD of moderate or greater complexity at recommended intervals, those with anatomy and physiology incompletely assessed by echocardiogram, those with change in clinical status and/or new concerning signs or symptoms, and for pre-procedural planning or evaluation.

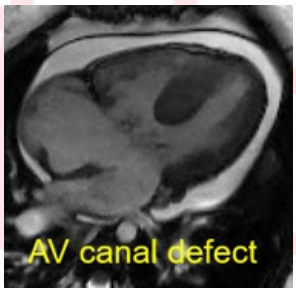
2 Why CMR

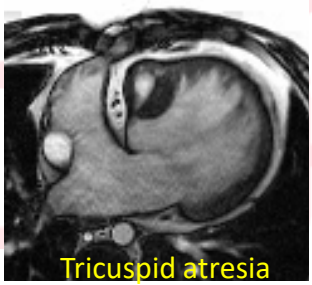
- Improve quality of care for patients with congenital heart disease.
- High accuracy in diagnosis due to excellent resolution of images.
- Excellent image quality independent from body habitus.
- Accurate assessment of morphology, function, strain, myocardial fibrosis, perfusion, valvular morphology and valvular function.
- Noninvasive assessment of intracardiac shunt, residual defects and vascular anomalies, and surveillance.
- Excellent 3D imaging for pre-surgical and pre-procedural planning.
- Advanced technique and assessment with 4D flow imaging.
- Robust prognostic data and no ionizing radiation exposure.

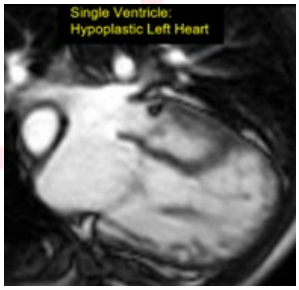
3 Images

Anatomic assessment



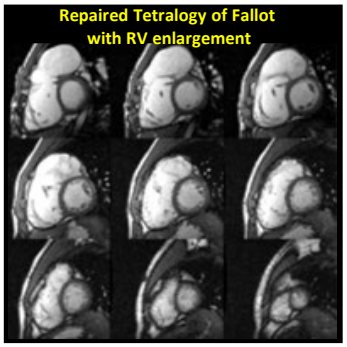




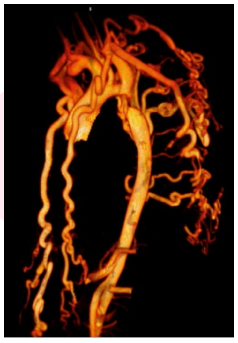


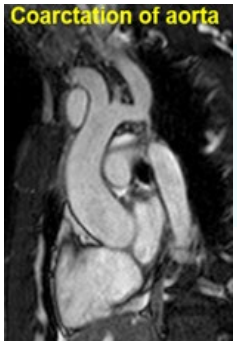
Functional assessment

Volume & EF

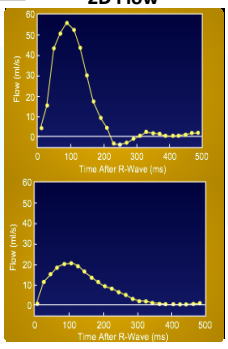


Angiography

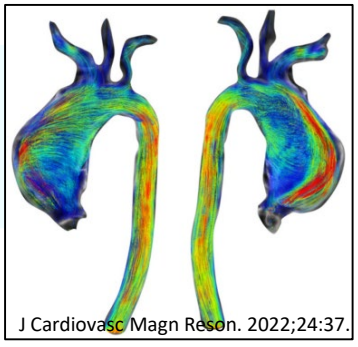




2D Flow



4D Flow



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Images provided courtesy of: *Sihong Huang;* *Helen DeVos Children's Hospital Congenital Heart Center, MI, USA*

4 Appropriate Use Criteria

M= Maybe appropriate, level 4-6; A=Appropriate, level 7-9;

Note: Frequency of test in parenthesis.

Simple Congenital Heart Disease

Unrepaired ≥ moderate ASD, VSD, PDA

Routine surveillance (1-2 years)	CMR±MRA (M4)
Change in clinical status, pre-repair planning	CMR±MRA (M5-6)

Post-repair ASD

Change in clinical status and/or new signs or symptoms	CMR±MRA (A7)
Routine surveillance after intervention	
• Surgical ASD repair: asymptomatic or mild symptoms (2-5 years)	CMR±MRA (M4-6)
• Device ASD closure with residual defect, valvular or ventricular dysfunction, arrhythmias, pulmonary hypertension (3-12 months)	CMR±MRA (M5)
• PDA occlusion with LPA stenosis or aortic obstruction (1-2 years) OR after pulmonary stenosis repair with moderate-severe sequelae (1-3 years)	CMR±MRA LPA stenosis (M6) Aortic obstruction (A7) Post PS repair (A7)

Moderate Congenital Heart Disease

Unrepaired

Sinus venosus ASD +/- PAPVC, TAPVC: Pre-repair planning	CMR ± MRA (A7-8)
Ebstein anomaly, tricuspid valve dysplasia: Pre-repair planning	CMR±MRA (A7), stress CMR (M5)
Aortic coarctation or interrupted arch: Routine surveillance (3-5 years)	CMR±MRA (A7-8)
in mild obstruction, new symptoms, pre-repair planning	Stress CMR (M6 for new symptom)
PAPVC involving >1 pulmonary vein: Routine surveillance (3-5 years)	CMR±MRA (M4)
TAPVC: Change in Clinical status, pre-repair planning	CMR±MRA (A7)
AVCD of all types: Change in clinical status, new symptoms, pre-repair planning	CMR±MRA (M5)
TOF: Change in clinical status, new symptoms, pre-repair planning	CMR±MRA (M5)

Post-repair

TOF: change in clinical status, new symptoms, pre-pulmonary valve replacement, routine surveillance with pulmonary regurgitation, ventricular dysfunction, residual RVOT obstruction, branch PA stenosis	CMR±MRA A7-8 (2-3 years) M5 (3-12 months if CHF)
PAPVC, TAPVC, Ebstein, Eisenmenger syndrome, TV dysplasia: change in clinical status, new symptoms, post PS repair with moderate-severe sequelae	CMR/MRA (A7), stress CMR (M5 Ebstein and Eisenmenger)
PAPVC, TAPVC, Ebstein, or TV dysplasia: Routine surveillance (3-5 years)	CMR±MRA (M4-6)
PAPVC, AVCD, or Ebstein with residual defect, pulmonary vein obstruction, LVOT obstruction, valvular or ventricular dysfunction, pulmonary hypertension, ventricular dysfunction, arrhythmias: Routine surveillance (3-12 months)	CMR±MRA (M5)
Aortic coarctation or interrupted arch:	
Routine surveillance (3-5 years), change in clinical status, new symptoms	CMR/MRA (A7-8)
Routine surveillance (3-12 months) in patients with CHF	CMR/MRA (M6)
Routine surveillance within the first year (6-12 months)	CMR/MRA (M5)

Abbreviations

ASD, Atrial Septal Defect; **AVCD,** Atrioventricular Septal Defect; **CHF,** Congestive Heart Failure; **LPA,** Left Pulmonary Artery; **VSD,** Ventricular Septal Defect; **LVOT/RVOT,** left/right ventricular outflow tract; **PAPVC,** Partial Anomalous Pulmonary Venous Connection; **PDA,** Patent Ductus Arteriosus; **PS,** Pulmonary Stenosis; **TAPVC,** Total Anomalous Pulmonary Venous Connection; **TGA,** Transposition of the Great Arteries; **TOF,** Tetralogy of Fallot; **TV,** Tricuspid Valve

4 Appropriate Use Criteria, cont'd

Moderate Congenital Heart Disease, cont'd

Coronary Anomalies

Unrepaired

Change in clinical status, new symptoms	CMR (A7), Stress CMR (A8)
Pre-repair planning	CMR (A7), Stress CMR (M6)
Routine surveillance (1-2 years)	CMR (M5), Stress CMR (M5)

Post-repair

Change in clinical status, new symptoms	CMR (A7), Stress CMR (A8)
Within 30-days post-procedural routine evaluation	CMR (M4)
Ventricular or valvular dysfunction: Routine surveillance (3-6 months)	CMR (M5), Stress CMR (M5)
Asymptomatic or mild sequalae:	CMR (M6), Stress CMR (M4)
Evaluation within 1 year after repair	CMR (M6), Stress CMR (M4)
Routine surveillance (2-5 years)	CMR (M5), Stress CMR (M5)

Complex Congenital Heart Disease

D-TGA

Unrepaired: Change in clinical status, pre-repair planning	CMR (M4-6)
Repaired (arterial switch operation, Rastelli, atrial switch operation)	
Change in clinical status, new symptoms	CMR (A7) , Stress CMR (M6)
CHF – Routine surveillance (3-12 months)	CMR +/- MRA (M4-6)
Asymptomatic – Routine surveillance (3-5 years)	CMR (A7), Stress CMR (M6 for arterial switch)
≥ Moderate systemic ventricular or valvular dysfunction, LVOT or RVOT obstruction, branch PA stenosis, presence of an RV-RA conduit, arrhythmias: Routine surveillance (3-12 months)	*Arterial Switch* – CMR (M6), Stress CMR (M5) *Rast*elli – CMR (M5) *Atrial switch* – CMR (A7), Stress CMR (M5)
Progressing neo-aortic root dilation, or neo-aortic regurgitation: Routine surveillance (1-2 years)	*Arterial switch* – CMR/MRA (A8)
Coronary evaluation in asymptomatic patients	*Arterial switch* – CMR (M8), Stress CMR (A7)

Single-ventricle

Unrepaired: Evaluation prior to planned surgical palliation	CMR (A7)
Change in clinical status, new symptoms	CMR (M6)
Postoperative:	
Stage 1 Palliation – Change in clinical status, new symptoms, evaluation prior to planned stage 2 palliation	CMR/MRA (A7)
Stage 2 Palliation – Routine surveillance (1-2 years), Change in clinical status, new symptoms, evaluation prior to planned stage 3 palliation	CMR/MRA (A7-8)
Stage 3 Palliation –	
Change in clinical status, new symptom	CMR/MRA (A7), stress CMR (M5)
CHF: Routine surveillance (3-12 months)	CMR/MRA (M6)
Valvular or ventricular dysfunction, arrhythmia, or other cardiac complications: Routine surveillance (3-12 months)	CMR/MRA (M6), stress CMR (M4)
Asymptomatic: Routine surveillance (3-5 years)	CMR/MRA (A8), stress CMR (M5)

4 Appropriate Use Criteria, cont'd

Complex Congenital Heart Disease, cont'd

Congenitally Corrected Transposition of the Great Arteries (CC-TGA)

Unrepaired	
Change in clinical status, new symptoms	CMR±MRA (A7), Stress CMR (M6)
Pre-repair planning	CMR±MRA (A8)
≥ moderate systemic AV valve regurgitation	CMR±MRA (M6)
– Routine surveillance (6-12 months)	CMR±MRA (M5)
CHF – Routine surveillance (3-12 months)	CMR±MRA (A7)
Asymptomatic – Routine surveillance (3-5 years)	
Repaired (anatomic or physiologic repair)	
Change in clinical status, new symptom	CMR±MRA (A7), Stress CMR (M6)
CHF – Routine surveillance (3-12 months)	CMR±MRA (M6)
Asymptomatic – Routine surveillance (3-5 years)	CMR±MRA (A7), Stress CMR (M5)
Repaired (anatomic repair)	
Valvular or ventricular dysfunction, RVOT or LVOT obstruction, or presence of a RV-PA conduit – Routine surveillance (6-12 months)	CMR±MRA (A7), Stress CMR (M5)
Routine surveillance (1-2 years) after the 1st year following repair with no or mild sequelae	CMR±MRA (M5), Stress CMR (M5)
Repaired (physiologic repair)	
≥ moderate systemic AV valve regurgitation, systemic RV dysfunction, and/or LV-to-PA conduit stenosis – Routine surveillance (3-12 months)	CMR±MRA (A7), Stress CMR (M5)
CHF symptoms – Routine surveillance (3-12 months)	CMR±MRA (M6)

Truncus Arteriosus

Unrepaired	
Change in clinical status, new symptoms, pre-repair planning	CMR±MRA (A7)
Postoperative	
Change in clinical status, new symptoms	CMR±MRA (A7), Stress CMR (M5)
Asymptomatic child or adult with ≥moderate truncal stenosis and/or regurgitation	
– Routine surveillance (3-6 months)	CMR±MRA (M5), Stress CMR (M4)
– Routine surveillance (1-2 years)	CMR±MRA (A7), Stress CMR (M5)
Asymptomatic child or adult – Routine surveillance (3-5 years)	CMR±MRA (M6), Stress CMR (M5)
Residual VSD, presence of an RV-to-PA conduit, or branch PA obstruction, or in patients with heart failure symptoms	
– Routine surveillance (3-12 months)	CMR±MRA (M6)
CHF symptoms – Routine surveillance (3-12 months)	CMR±MRA (M6)

For complete list and detail, please consult references below.

1. Sachdeva R, et al. ACC/AHA/ASE/HRS/ISACHD/SCAI/SCCT/SCMR/SOPE 2020 Appropriate Use Criteria for Multimodality Imaging During the Follow-Up Care of Patients With Congenital Heart Disease. *J Am Coll Cardiol. 2020;75:657-703.*

2. Society for Cardiovascular Magnetic Resonance/European Society of Cardiovascular Imaging/American Society of Echocardiography/Society for Pediatric Radiology/North American Society for Cardiovascular Imaging Guidelines for the use of cardiovascular magnetic resonance in pediatric congenital and acquired heart disease : Endorsed by The American Heart Association. J Cardiovasc Magn Reson. 2022;24:37.