# Cardiovascular MR Imaging: Exploring the Boundaries
# Case Based Studies in MR - Congenital Heart Disease

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*Goals*

Several cases of adult congenital heart disease (ACHD) as may be encountered in a typical Cardiac MR (CMR) practice will be presented. To properly perform and interpret an ACHD study requires not only an understanding of the pathophysiology of the underlying congenital anomaly, but also familiarity with and ability to recognize the more common corrective surgical procedures as well as a good knowledge of how and when to apply CMR techniques.

This short case-based study presentation will focus on applying the different types of cardiac MR pulse sequences (e.g. CINE, black blood, quantitative flow, viability, MR angiography) in an appropriate manner as dictated by the unique nature of ACHD. As the spectrum of disease (and the accompanying array of MR sequences) is broad, this short case-study can only touch on the topic and serve as an introduction to this rewarding and fascinating facet of MRI.

*Introduction*

Advances in management of pediatric cardiac malformations have enabled most patients to survive well into adulthood. This represents a new population with unique issues never before seen in medicine. Cardiac MR, also advancing at a rapid rate, plays an increasingly significant role in the management of these patients. While echocardiography still plays the dominant role in diagnostic imaging, CMR has proven itself invaluable in many circumstances. Probably most significant are the abilities to a.) accurately assess right and left ventricular volume and function (RV evaluation being typically quite limited with echo, and vital to the evaluation of almost all ACHD), and b.) quantify flow through vessels and conduits, thereby accurately evaluating valvular regurgitation and determining shunt fractions. CMR is also extremely useful for evaluating the complete aorta (e.g. in coarctation), detecting aorto-pulmonary collaterals and anomalous pulmonary veins, characterizing coronary anomalies, and detecting right and left ventricular myocardial fibrosis.

*CMR Pulse Sequences*

Bright blood CINE imaging is the workhorse for most CMR, including ACHD, where it is used for biventricular volume and function measurement, ventricular mass measurement, detecting valvular abnormalities such as regurgitation and stenosis or bicuspid aortic valve, and characterizing ventricular chambers (i.e. right vs. left ventricle or in the case of single ventricles) as well as figuring out anatomical relationships between chambers and natural or surgical conduits. In many cases, ASDs and VSDs, including patent foramen ovale, can be seen with CINE, as can baffle stenoses or leaks in cases of Mustard or Senning repair for transposition.

Quantitative phase contrast, or “q” flow imaging allows for accurate and reproducible measurements of forward and reverse through vessels and conduits, thereby permitting

quantification of valvular regurgitant fractions and shunt volumes, as well as entities such as split pulmonary flow or hemodynamic significance of an aortic coarctation. In addition, peak velocities can be measured to grade stenosis (although echo is often more accurate in this circumstance secondary to difficulty aligning the MR imaging plane exactly with the maximal stenotic jet).

Black blood imaging can be useful for defining the anatomy of cardiac chambers and defects, as well as evaluating the aortic wall for abnormalities such as intramural hemorrhage. It can also be useful for evaluating anomalies such as left ventricular non-compaction.

MR angiography can either be contrast-enhanced, particularly useful for the aorta and pulmonary vessels, or non-contrast, which works nicely for evaluating the coronary origins and aortic root. Both have their respective merits, and are often used in conjunction with one another. Evaluation of the more distal coronary arteries remains problematic and is better suited to CTA, but luckily is seldom required as part of the ACHD workup.

Finally, late gadolinium enhancement (LGE), or "viability" imaging has an emerging role in ACHD, particularly for the evaluation of right ventricular fibrosis in the often abnormally enlarged right ventricles associated with ACHD. In this group, RV LGE is believed to correlate with complications such as fatal arrhythmias. As the ACHD patients age and develop secondary cardiac disease, evaluation of left ventricular LGE and perfusion will be future important considerations.

*Partial Table of ACHD Variants, Treatment, and Primary CMR Evaluation Targets*

| Disease | Common Corrective Procedure | Primary Entities Evaluated by MRI |
|---|---|---|
| **Tetrology of Fallot** | (Prior palliative: Blalock-Taussig Shunt)<br>Definitive: VSD Closure, RVOT outflow repair | RV function and size<br>Pulmonic regurgitation (90% have)<br>PA stenosis, residual VSD<br>RV fibrosis |
| **D-Transposition** | Mustard, Senning (intra-atrial switch)<br>Jatene (arterial switch)<br>Rastelli (RV-PA conduit) | RV function and size<br>Baffle leak (using qflow Qp:Qs)<br>RV fibrosis |
| **L-Transposition** | Senning with arterial switch<br>Senning with Rastelli | RV function and size (esp. if not corrected)<br>Possible PA or aortic stenosis<br>Residual VSD |
| **Single Ventricle** | Palliative: Glenn shunt<br>Definitive: Fontan (cavo-pulmonary conduit) | Cavo-pulmonary evaluation for stenosis etc.<br>Single ventricular function<br>Pulmonary artery/vein evaluation |
| **Aortic Coarctation** | Coarctation repair, e.g. left subclavian flap angioplasty vs. graft | MRA for residual aortic stenosis<br>Qflow to determine hemodynamic sig. of stenosis<br>CINE aortic valve for co-existent bicuspid AOV |
| **Anomalous Pulmonary Veins** | Surgical re-anastamosis | Q-flow for residual shunt<br>MRA for post-surgical pulmonary venous strictures |
| **Marfan's Disease** | Aortic root repair/valve replacement | Aortic root size – longitudinal follow-up<br>Evaluate for aortic dissection<br>Dural ectasia LS spine |
| **Coronary Artery Anomalies** | Coronary re-implantation | Coronary origins<br>LGE left ventricle<br>Other associated cardiac anomalies |