MRI Angiography in the preoperative evaluation of patients with Tetralogy of Fallot

Poster No.: C-0536
Congress: ECR 2013
Type: Educational Exhibit
Authors: V. N. Bakare¹, K. Taori², A. Disawal¹, J. Rathod¹, P. S. Wavare³;
¹Nagpur, MAHARASHTRA/IN, ²Nagpur, Ma/IN, ³Nagpur/IN
Keywords: Arteries / Aorta, Cardiovascular system, Vascular, MR-Angiography, MR, Diagnostic procedure, Computer Applications-3D, Haemodynamics / Flow dynamics
DOI: 10.1594/ecr2013/C-0536

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method ist strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

1. To describe the spectrum of Anatomical alterations in patients with Tetralogy of Fallot (TOF).
2. To preoperatively evaluate patients suspected of having Tetralogy of Fallot with help of MR Angiography under three categories---
   
a) Morphological Assessment
b) Functional Assessment
c) Contrast enhanced MR Angiography

and describe the spectrum of findings including pulmonary artery anomalies, aorto-pulmonary collaterals(Fig.6), right sided aortic arch (Fig.1,8,9), atrial and ventricular septal defects, persistent left superior vena cava(Fig.7), coronary artery abnormalities, aberrant right subclavian artery, and others.

3. To describe suitable Indications, Advantages and limitations of MR Angiography as compared to other imaging modalities.

4. To correlate MR Angiography findings with intraoperative findings and improve the diagnostic skills by retrospective study.

In this poster we will review the contribution of MR Angiography with special focus on Gadolinium enhanced MR Angiography and three dimensional reconstructed images to diagnose the vascular abnormalities in patients with Tetralogy of Fallot.

The Aim is to improve the diagnostic accuracy of imaging and provide the vascular surgeon all the anatomical alterations present in every individual case so as to allow safer and successful surgery and its final outcome.
Images for this section:

**Fig. 1:** Coronal MR Angiography image showing right sided aortic arch with Mirror image type of branching pattern. (Left brachiocephalic trunk, Right CCA and Right subclavian artery)

© RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Background

Tetralogy of Fallot is one of the most common congenital cyanotic heart diseases in children, which accounts for approximately 4-8% of all congenital cyanotic heart diseases and presents with cyanotic spells, repeated pulmonary infections, failure to thrive and at times with consequences of paradoxical embolism.

The causative factor is misalignment of the crista supraventricularis with associated underdevelopment of the infundibulum. The four features included are overriding of the aorta (Fig.3) with subaortic ventricular septal defect and right ventricular outflow tract obstruction with consequent right ventricular hypertrophy (Fig.4). As a consequence of right ventricular outflow tract obstruction, there is development of bronchial and aortopulmonary collateral vessels (Fig.6) which supply a variable amount of blood to the lungs. Tetralogy of Fallot is often associated with other anomalies like pulmonary artery atresia of various degrees of severity (Fig.2), right sided aortic arch (Fig.1,8,9), atrial and ventricular septal defect, persistent left superior vena cava (Fig.7), coronary artery abnormalities, and aberrant right subclavian artery, which are necessary to be detected before surgical correction of this condition. Contrast enhanced MRI angiography plays an important role in the assessment of the deep anatomic structures which are difficult to assess with echocardiogram and selective angiography. Further, cine MRI images gives additional and detailed information about the cardiac function, valve patency, and the hemodynamic significance of the vascular stenosis.

Chest X ray shows right ventricular hypertrophy in the form of enlargement of the cardiac silhouette with lateral and upturned apex. There is pulmonary oligemia and absent or rudimentary pulmonary nuchal which gives concavity to the right heart border which is classically described as "Boot shaped heart" in literature.

Echocardiography was used traditionally as one of the tool to diagnose most of the components of TOF. Beekman et al concluded that MR imaging was superior to echocardiography for the evaluation of right ventricular hypertrophy and overriding aorta. In a study by Greenberg et al in which echocardiography and MR imaging were directly compared in the evaluation of pulmonary abnormalities postoperatively in children with tetralogy of Fallot, MR imaging was confirmed to have greater sensitivity. Echocardiography is often of limited use because of poor acoustic window for proper evaluation. The difficulties encountered are improper visualization of stenosis and aneurysmal dilatation, and the patency of the hypoplastic pulmonary arteries.

Conventional angiography was used to complement the finding the echocardiography. Geva et al compared MR imaging with conventional angiography in the evaluation
of pulmonary arteries and collateral aortopulmonary vessels in 23 patients and found complete agreement between the features depicted on conventional angiograms and those depicted on MR angiograms with regard to diagnosis of hypoplasia or stenosis of a pulmonary artery branch. Choe et al evaluated whether MR imaging could depict pulmonary arterial anatomy in greater detail than routine angiography in patients with congenital or acquired occlusion of the left pulmonary artery or with pulmonary atresia.

Recent advances in MR imaging are superior to traditional echocardiography and alternative to conventional angiography because it provides both anatomic and functional information about cardiovascular anomalies. There is good correlation between the MRI angiography and conventional angiography findings with respect to the right ventricular output tract obstruction, in patients of TOF. Holmqvist et al reported 93% agreement with regard to a finding of main pulmonary artery stenosis, 79% agreement with regard to that of right pulmonary artery stenosis, and 86% agreement with regard to that of left pulmonary artery stenosis (n = 14). Agreement with regard to the finding of right ventricular enlargement was 50%; differences in assessment, in this instance, were classified as "mild" instead of "moderate" change, or as "normal" instead of "mild" change. Assessments with regard to the presence of overriding aorta and the patency and appearance of a Blalock-Taussig shunt were in agreement in 79% and 100% of cases. Right ventricular outflow measurement with the use of velocity-encoded MR images, was found superior to echocardiography in a study reported by Rebergen et al. MR imaging also provides a practical method of quantifying pulmonary regurgitation, as reported in the studies by Helbing and De Roos and by Rebergen and colleagues. Three dimensional Contrast enhanced MRI angiography with Maximum Intensity Projection (MIP) reconstruction is particularly useful for assessment of the deep anatomic structures such as pulmonary arteries, the location and caliber of aortopulmonary collaterals, the arch of aorta and its branching pattern. It also provides information about the dimensions of the right and left ventricular outflow tracts. Cine MRI images taken in various planes provide detailed information about the cardiac function, valve patency and hemodynamic significance of the vascular stenosis. A balanced turbo field-echo (BTFE) sequence in axial, coronal and sagittal plain is useful for morphological assessment of the cardiac structures and thoracic vessels.

TOF is usually associated with pulmonary artery atresia of various degree of severity (from mild hypoplasia to complete atresia) (Fig.2). Other common associated anomalies include right sided aortic arch (Fig.1,8,9) in about 25% cases and atrial septal defect in 5% cases. Other associations include persistent left superior vena cava (Fig.7), coronary artery abnormalities and aberrant right subclavian artery.
**Fig. 2:** Coronal MR Angiography image showing the hypoplastic main pulmonary trunk.

**References:** RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN

MRI imaging is non-invasive imaging of choice with high sensitivity and specificity to evaluate Tetralogy of Fallot and associated abnormalities in pre-operative evaluation of the patient for obtaining information about the hemodynamic status of the patient and further surgical planning. MRI can also be used as an important tool in assessment of the post-operative patient with surgical correction.
Imaging findings OR Procedure details

Patients suspected of having Tetralogy of Fallot based on clinical background and on primary imaging modalities like X ray and echocardiography were subjected to further MR study.

Our Population:

8 patients suspected of having Tetralogy of Fallot on echocardiogram were selected for further MRI evaluation. The age group of patients included was between 0 to 30 years with prior proper well informed consent. There was no any operative history in any patient.

Protocols

Machine Philips Achieva 1.5 Tesla MRI machine

Patient position : supine

sedation was given in non-cooperative patients.

Contrast Agent : Gadolinium

Assessment Categories :

Morphological assessment  - A balanced turbo field-echo (BTFE) sequence in axial, coronal and sagittal plain, which is a balanced steady-state free precession sequence, was used for morphological assessment of the cardiac structures and thoracic vessels. With this fast imaging sequence, it is also possible to acquire a stack of single-shot anatomic images with short breath-hold times that provides a detailed anatomic and topologic assessment of the heart and great vessels.

Functional assessment  - ECG gated cine MRI pulse sequences were used to obtain information about cardiac wall motion, blood flow patterns across the cardiac valves and deformed large vessels.

Contrast enhanced MR Angiography  - 3D Contrast enhanced MRI angiography of the aorta with maximum intensity projection (MIP) formatting was later done to for visualization of the great vessels of the thorax and the abdomen. Adequate post processing was done as per requirement.

Observations :
The four basic components of Tetralogy of Fallot were present in every case i.e. Over-riding of the aorta(Fig.3) with sub-aortic ventricular septal defect and right ventricular outflow tract obstruction with right ventricular hypertrophy (Fig.4). The sub-aortic VSD was in range of 8 to 18 mm with associated overriding of the aorta over the ventricular septum. There was right ventricular hypertrophy (Fig.4) in every patient and consequent right atrial dilatation (Fig.5) in 6 patients. The right ventricular outflow tract obstruction was in the form of infundibular pulmonary stenosis. The veno-atrial and atrio-ventricular connections were normal in all cases.

There was hypoplasia of the main pulmonary trunk (Fig.2) and its branches in all of the cases with post-stenotic dilatation in 3 of them. 5 patients had valvular pulmonary stenosis due to hypoplastic valves. The high velocity blood flow across the stenotic segments of the pulmonary artery was seen as regions of signal void from dephased spins. There was pulmonary oligemia in 6 cases. One of the patients had multiple dilated Aorto-Pulmonary collateral anastomotic channals (MAPCAs) (Fig.6) connecting the descending aorta with the terminal part of the left and right pulmonary arteries with their resultant aneurysmal dilatation. There was incidental finding of double superior vena cavae noted in the same patient with right SVC opening in right atrium and left SVC opening in the coronary sinus (Fig.7). There was right sided aortic arch(Fig.1,8,9) in two patients of which one had Mirror image type of branching pattern (Fig.1) and the other had aberrant origin of left subclavian artery(Fig.9). Patent ductus arteriosus was seen in one of the patient(Fig.10). There was incidental finding of origin of left vertebral artery directly from the arch of aorta in one case.

<table>
<thead>
<tr>
<th>PATIENT NO.</th>
<th>AGE (yrs)</th>
<th>RIGHT VENTRICULAR HYPERTROPHY (Fig.4)</th>
<th>OVERRIDING OF AORTA (Fig.3)</th>
<th>RIGHT VENTRICULAR OUTFLOW OBSTRUCTION</th>
<th>SUBAORTIC VENTRICULAR SEPTAL DEFECT (mm)</th>
<th>OTHER ASSOCIATED ABNORMALITIES</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>8</td>
<td>PATENT DUCTUS ARTERIOSUS.</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>7</td>
<td>----</td>
</tr>
<tr>
<td>3</td>
<td>9</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>12</td>
<td>----</td>
</tr>
<tr>
<td>4</td>
<td>10</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>14</td>
<td>----</td>
</tr>
<tr>
<td>5</td>
<td>11</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>15</td>
<td>MAPCAs and DOUBLE SVC</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>---</td>
<td>---</td>
<td>---</td>
<td>---</td>
<td>---</td>
<td>---</td>
<td>------------------------------------------------------------------</td>
</tr>
<tr>
<td>6</td>
<td>22</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>16</td>
<td>RIGHT SIDED AORTIC ARCH WITH MIRROR IMAGE TYPE OF BRANCHING PATTERN</td>
</tr>
<tr>
<td>7</td>
<td>27</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>12</td>
<td>----</td>
</tr>
<tr>
<td>8</td>
<td>28</td>
<td>Y</td>
<td>Y</td>
<td>Y</td>
<td>18</td>
<td>RIGHT SIDED AORTIC ARCH WITH ABERRANT LEFT SUBCLAVIAN ARTERY</td>
</tr>
</tbody>
</table>

(KEYS - Y- YES,
MAPCA-Multiple Aorto-Pulmonary Collateral Anastomotic Channels,
SVC-superior vena cava).
Fig. 3: Axial BTFE (balanced turbo field-echo sequence) image showing overriding of the aorta across the ventricular septum.

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 4: Axial BTFE image showing Right Ventricular hypertrophy with deviation of the interventricular septum to left side.

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 5: Axial BTFE image showing right atrial dilatation.

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 6: Coronal MR angiography image showing dilated collateral from the descending aorta upto the terminal part of right pulmonary artery resulting in its aneurysmal dilatation.

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
**Fig. 7**: Coronal MR Angiography image showing double SVC

**References**: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 8: Axial BTFE image showing Right sided aortic arch

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 9: Coronal MR angiography image showing right sided aortic arch with aberrant origin of left subclavian artery.

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN
Fig. 10: Coronal MR Angiography image showing the Patent Ductus Arteriosus

References: RADIOLOGY, GOVT. MEDICAL COLLEGE, NAGPUR, GOVT. MEDICAL COLLEGE, NAGPUR - Nagpur/IN

Most of these patients were then subjected to corrective surgeries as per the need. The intra-operative findings were noted in each of the case. Then retrospectively again the MR Angiography findings and Surgical findings were compared in every respective case to improve the diagnostic accuracy. Fortunately, there was no any major significant difference between the imaging and surgical findings.
Conclusion

Imaging Techniques, Comparison

1. **X Ray**: Primary Screening modality. Typically shows the "Boot shaped heart". Widely available and relatively cheaper, but radiation exposure present. No further detail information available.

2. **Echocardiography**: second line investigation, no radiation exposure. It demonstrates right ventricular hypertrophy, overriding aorta, septal defects, and pulmonary arterial abnormalities to some extent. Echocardiography is often of limited use because of poor acoustic window for proper evaluation. The difficulties encountered are improper visualization of stenosis and aneurysmal dilatation, and the patency of the hypoplastic pulmonary arteries.

3. **Conventional Angiography**: Invasive procedure, with exposure to radiation and iodinated contrast agents. It demonstrates the vascular anatomy, however surrounding structural anomalies not visualised. Therapeutic procedures possible.

4. **MR Angiography**: Recent advancement, Morphological and functional assessment possible along with post contrast (Gadolinium) angiography. Multiplanar image reformatting possible which improves diagnostic accuracy. No radiation exposure.

MRI imaging is non-invasive method of choice to evaluate the complex abnormalities and get detailed anatomic and functional information in addition without risk of radiation and contrast reactions to iodinated agents. Echocardiography is recommended for screening, initial diagnosis and follow-up of patients with congenital heart diseases. Conventional angiography should be used for interventional procedures such as embolization of collateral vessels. All these modalities are to be considered complementary in pre and post-operative evaluation of the patient.
References

- **Beekman RP, Beek FJ, Meijboom EJ.** Usefulness of MRI for the pre-operative evaluation of the pulmonary arteries in the tetralogy of Fallot. Magn Reson Imaging1997; 15(9): 1005-1015.


Personal Information

Dr. Vishal Nandkishor Bakare
Resident Doctor
Dept. of Radiology, Govt. Medical College, Nagpur, Maharashtra, India
email: vishalnb1154@gmail.com, vishalnb@yahoo.com

Dr. Kishor Taori
Professor and Head,
Dept. Of Radiology, Govt. Medical College, Nagpur, Maharashtra, India
email: kishortaori@gmail.com

Dr. Amit Disawal
Senior Assistant Professor
Dept. Of Radiology, Govt. Medical College, Nagpur, Maharashtra, India

Dr. Jawahar Rathod
Senior Assistant Professor
Dept. Of Radiology, Govt. Medical College, Nagpur, Maharashtra, India

Dr. Prasad Wavare, Dr. Rakhi P Puria, Dr. Mansi Jain
Resident Doctors
Dept. of Radiology, Govt. Medical College, Nagpur, Maharashtra, India