Fallot Tetralogy Imaging Findings with MDCT, pre and postoperative findings

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Learning objectives

1. To describe the protocol of acquisition and MDCT technique in pediatric population.

2. To describe temporary/palliative surgery and complete intracardiac repair techniques.

3. To illustrate the main MDCT Angiography pre and post-surgical findings in Tetralogy of Fallot.
Background

1. Tetralogy of Fallot Definition:

Tetralogy of Fallot is the most common cyanotic congenital heart defect, one of the five known as the "Terrible Ts": Transposition of the Great Arteries, Tricuspid Atresia, Total Anomalous Venous Return, and Truncus Arteriosus.

The classic findings consists in four basic lesions: (Fig 1-4)

| 1. Large ventricular septal defect |
| 2. Right Ventricular outflow obstruction (from pulmonary stenosis) |
| 3. Overriding aorta |
| 4. Right ventricular hypertrophy |

Four basic lesions

Timing of presentation and degree of cyanosis correlates precisely with the amount of right ventricular outflow tract obstruction. Patients with severe outflow tract obstruction present early in the neonatal period. Those patients with adequate pulmonary blood flow at birth gradually develop increasing cyanosis during the first few weeks and months of life.

Two of the lesions will determine the extent of the disease pathophysiology. There must be right ventricular outflow obstruction and the VSD must be large enough to equalize pressures in both of the ventricles.

Depending on the severity of the right ventricular stenosis may occur.

1. Right to left shunt (cyanosis and decreased pulmonary blood flow)

Severe pulmonary stenosis

2. Left to right shunt (acyanotic Tetralogy of Fallot)

Mild pulmonary stenosis
2. Diagnostic

Actually a number of cases can be diagnosed by fetal echocardiography, allowing monitoring the development of the pulmonary valve ring, the truncus and the pulmonary branches. Detect rare and sever cases of Fallot.

However in most cases the diagnosis is postnatal.

Clinic and Diagnostic Methods

Clinic presentation:

Cyanosis, respiratory distress grunting, fussiness and agitation, a systolic thrill at the lower and middle left sternal border. A Single S2, an aortic ejection click.

The ECG will show right axis deviation and right ventricular hypertrophy.

On chest radiography the presence of a "Coeur en sabot" or "boot-shaped" heart, which is caused by the absence of the small, atretic pulmonary arteries.

Doppler echocardiography with color flow is the method of choice, the findings are diagnostic for tetralogy of Fallot may be the only examination required before surgery and can assess:

- Interventricular communication, severity of the overriding aorta and differential diagnosis
- Severity and localization of pulmonary stenosis; size of the ring, trunk and pulmonary arteries
- Associated abnormalities
- Postoperative complications

3. Tetralogy of Fallot Treatment. Types of surgery

3a. Complete intracardiac repair (fig. 8)

Complete surgical repair typically involves patch closure of the ventricular septal defect and relief of RV outflow tract obstruction by means of pulmonary valvotomy, resection
of hypertrophied muscle bundles, or placement of a transannular outflow tract patch (in severe PV hypoplasia), placement of a graft conduit between right ventricle and the pulmonary artery may be necessary if anomalous coronary artery obstructs access to right ventricle infundibulum or complete pulmonary atresia is present.

Until recently, the transventricular repair was habitual in all kinds of Fallot, and correction of pulmonary stenosis was performed with implantation of patches of very large expansion in order to avoid future restenosis. However the patients showed progressively pulmonary valve insufficiency and development of large akinetic regions in the outflow tract of the right ventricle, source of arrhythmias and worsening of the pulmonary valve insufficiency. Therefore, the current trend is to preserve as far as possible the function of the valve, limiting as much as possible transannular patch and the extension of it to the right outflow tract.

Currently most surgical groups establish elective surgery (in asymptomatic children) of Fallot in 3-6 months. There are groups who prefer correct Fallot early, including the neonatal period. Early correction of Fallotfavor the correct development of the peripheral pulmonary vasculature and decrease the risk of developing significant pulmonary valve insufficiency.

3b. Temporary or Palliative surgery (Fig. 5-7)

In specific cases, when the child's age or the anatomy is unfavorable and can not perform first intention complete intracardiac repair staged correction is indicated (palliative techniques):

- Blalock-Taussig Shunt: between the innominate or subclavian artery and the ipsilateral pulmonary artery GORE-TEX®). It is performed when the pulmonary arteries are small and newborn oxygenation depends ductus. It is also performed in some cases with anomalous coronary.
- Partial correction (right ventricle - pulmonary artery connection leaving open the interventricular communication). Usually in no ductus-dependent infants, but with small pulmonary arteries.
- Balloon dilatation for pulmonary valve
- Valved conduit early implantation (right ventricle - pulmonary trunk). In special anatomies, such as when the pulmonary branches are disconnected from each other and/or one arises from the aorta, in cases of absent pulmonary valve, or if Fallot is associated with complete atrioventricular canal (Down syndrome).
• When coronary artery crossing the pulmonary infundibulum, it is not possible to cross a expansion patch then is necessary the implantation of a conduit (right ventricle - pulmonary artery)
Images for this section:

Fig. 1: Ventricular Septal Defect Male 1 year and 7 months old MDCT Angiography. Four-chamber and short-axis views: Defect in the ventricular septum

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Fig. 2: Right Ventricular outflow obstruction Male 1 year and 7 months old MDCT Angiography. Axial and (A-C) y sagittal (B-D) multiplanar reformatted: Valvular and supravalvular pulmonary stenosis (RV: Right ventricle, PA-arrow: Pulmonary artery, PV: Pulmonary valve)

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Fig. 3: Overriding Aorta Male 1 year and 7 months old MDCT Angiography: the aorta is displaced to the right so that it appears to arise from both ventricles and straddles the ventricular septal defect (AO: Aorta RV: Right Ventricle LV: Left ventricle)

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**Fig. 4:** Right ventricular hypertrophy Male 1 year and 7 months old MDCT Angiography: Four-chamber (A-C) and short-axis (B-D) views. Hypertrophy of the muscular walls of the right ventricle. VD/VI Ratio increased

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Fig. 5: Potts shunt (arrows) Male 6 years old MDCT Angiography: Multiplanar reformatted images: Anastomosis between the descending aorta (AO) and the left pulmonary artery (Lpa). Assessment the integrity and permeability. In this case, the fistula is functional.

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Fig. 6: Blalock-Thomas-Taussig shunt (arrows). Male 7 years old MDCT Angiography: Multiplanar reformatted images: Anastomosis between the right subclavian artery (RSCA) and the right pulmonary artery (RPA). Assessment the integrity and permeability. In this case, the fistula is functional.

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Fig. 7: Blalock-Thomas-Taussig shunt (arrows). Male 1 year 2 months old MDCT Angiography: Multiplanar reformatted images: Anastomosis between the right subclavian artery and the right pulmonary artery (hypoplastic). Assessment the integrity and permeability. This case is a shunt malfunction (ocluded)

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Fig. 8: Post repair surgery status. Male 13 years old, MDCT Angiography: Multiplanar reformatted images: Closure of VSD through Patch (fig. A-B), no evidence of residual shunts, enlargement of the right ventricular outflow tract (RVOT) (fig. C-D)

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Findings and procedure details

1. MDCT Scanning Protocol

MDCT scanners provide images with a high level of anatomical detail. MDCT protocols must be adapted to specific clinical indication and patient characteristics, nevertheless there are some general guidelines regarding MDCT technique.

We generally recommend a biphasic protocol to achieve opacification of both right and left chambers with the following technique:

<table>
<thead>
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<th>ECG-Gated</th>
<th>None</th>
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<tr>
<td></td>
<td>Prospective at 45% of R-R cycle when coronary evaluation is indicated</td>
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<tr>
<td>mAs</td>
<td>1-7 Kg = 10 mAs/Kg</td>
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<tr>
<td></td>
<td>8-12 Kg = +5 mAs/kg</td>
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<tr>
<td>kV</td>
<td>&lt;40 kg = 80 kV</td>
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<tr>
<td></td>
<td>41-70 = 100 kV</td>
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<tr>
<td></td>
<td>&gt;70 kg = 120 kV</td>
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<tr>
<td>FLASH protocol</td>
<td>+ (Pitch 2.8)</td>
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<tr>
<td>Collimation</td>
<td>1 x 128 x 0.6</td>
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<tr>
<td>Iodine contrast</td>
<td>1 ml/kg</td>
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<tr>
<td>Injection rate</td>
<td>1.5-3 ml/seg</td>
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<td>Triggering</td>
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<td>Left chambers = acquisition at the end of injection + 2 sec</td>
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<td>Right chambers = acquisition at 3/4 of the injection + 2 sec</td>
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2. MDCT Diagnostic findings and presurgical assessment

To assess the basic components Fallot Tetralogy
To assess associated injuries:

- A patent foramen ovale or atrial septal defect (Pentalogy of Fallot) (fig. 9)
- Right Aortic Arch (fig. 10)
- Pulmonary artery atresia (fig. 12)
- Anomalous coronary artery (that can hinder a surgical correction) (fig. 11)
- Agenesis or hypoplasia of the pulmonary valve with pulmonary insufficiency
- Patent ductus arteriosus (fig. 13)
- Aortic valve Insufficiency
- Anomalous pulmonary venous drainage
- Persistent left superior vena cava (fig. 14)
- Aberrant subclavian artery (fig. 13)
- Collateral vessels in aorto pulmonary window (fig. 15)
- Rarely, tracheoesophageal fistula, rib anomalies and scoliosis.

3. MDCT Post-operative findings

Most patients who undergo complete cardiac repair have an uncomplicated postoperative recovery.

Over 85% of children who undergo early surgical correction will survive into adulthood. (may remain asymptomatic for 10-30 years)

Late complications:

- Eventually lead to progressive exercise intolerance, right heart failure, symptomatic arrhythmias, or sudden cardiac death.

Morbidity and mortality of TOF repair are usually associated with:

- Abnormal RV hemodynamics. Pulmonary regurgitation (PR) and stenosis are common complications and eventually lead to chronic RV volume or pressure overload and subsequent RV dilatation and failure

Two stages:

In the early (compensated) stage, there is an asymptomatic increase in end-diastolic volume and hypertrophy of the RV.

In the later (decompensated) stage, there is a decrease in the RV mass-to-volume ratio, an increase in RV end-systolic volume, compromised LV filling due to marked RV dilatation, and markedly reduced RV and LV ejection fractions
4. Multidetector CT of Late Postoperative Complications. PR and RV Dilatation (fig. 16 - 19)

- **Pulmonary Regurgitation** is one of the most commonly encountered complications following TOF repair.
- **Right Ventricle hypertrophy:** Pulmonary regurgitation eventually progresses to Right ventricle dilatation and right heart failure, may lead to life-threatening arrhythmias.
- **Pulmonary Ventricule incompetence** seen at long-term follow-up of these patients is partially related to the method of RVOT reconstruction used during initial repair.
- **VR multidetector CT** is excellent in depicting the morphology of the RVOT and cardiac abnormalities related to PR. Enlarged ventricular volumes are major criteria for PV replacement.
- **RV size and function** (fig. 8), for functional and volumetric assessment of the RV and LV, in the end-systolic and end-diastolic phases of the cardiac cycle, permitting derivation of stroke volume and ejection fraction.
- **Wall motion abnormalities**, which often result from altered ventricular mechanics due to volume overload.
- **Residual Pulmonary Artery Stenosis** occurs approximately 10%-15%. Stenosis may occur in the central or peripheral Pulmonary Arteries and can be a complication of prior palliative Blalock-Taussig shunt placement.
- **RVOT Aneurysm** (dyskinesia) outward movement during systole of part of the ventricular wall or its repaired outflow tract (related in part to RVOT or transannular patching during initial repair), extreme myectomy of the infundibulum or ischemic insult may contribute to its formation.
- **Recurrent Ventricular septal defect** finding that may require intervention (fig. 17).
- **Vascular Stents** endovascular stents and stent-mounted valves is an important indication for cardiac multidetector CT and must evaluate both the position and the patency of these stents. (fig. 18)
- **Implanted Devices** Patients with pacemakers, ICD (Implantable cardioverter defibrillator), coils, or other devices (fig. 19).
- **Anomalous Coronary Arteries** Common abnormalities include the left anterior descending artery arising from the right CA, the left anterior descending artery arising separately from the right sinus of Valsalva, and the right CA originating from the left CA or left anterior descending artery, some patients may have only one CA. (fig. 11)
Images for this section:

**Fig. 9:** A patent foramen ovale or atrial septal defect (fig. E arrow). Female 1 year and 8 month, MDCT Angiography: Multiplanar reformatted images: Ventricular septal defect (Fig. A), Right Ventricular outflow obstruction (Fig. B), Overrinding aorta (Fig. C), Right ventricular hypertrophy (Fig. D). Interatrial communication (Fig. E). Entity known as PENTALOGY OF FALLOT.

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**Fig. 10:** Right Aortic Arch (fig. A - B). Female with diagnosis of Fallot, MDCT Angiography: Multiplanar reformatted images: demonstrating right aortic arch. TR (trachea)

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**Fig. 11:** Unique coronary artery Male 10 years old with diagnosis of Fallot (fig. A-B), MDCT Angiography: Multiplanar reformatted images unique coronary artery with Circumflex branch extending to the right atrioventricular sulcus.
Fig. 12: Tetralogy of Fallot with severe pulmonary stenosis: Female 16 years old with diagnosis of Fallot (fig. A-B circle), MDCT Angiography: Multiplanar reformatted images.
**Fig. 13:** Patent ductus arteriosus (fig. B) and aberrant right subclavian artery and (fig. C - D). Male with diagnosis of Fallot, MDCT Angiography: Multiplanar reformatted images: Right subclavian artery (RSC) crosses the posterior mediastinum behind the esophagus on its way to the upper extremity. Patent ductus arteriosus (PDA) communication between the descending thoracic aorta and the left pulmonary artery.

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**Fig. 14:** Post repair surgery status. Female 6 years old, MDCT Angiography: Multiplanar reformatted images: persistent left superior vena cava (asterisk fig. A-B).

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**Fig. 15:** Collateral arteries: Female 16 years old with diagnosis of Fallot (fig. A-B), MDCT Angiography: Multiplanar reformatted images: Multiple collateral arteries (fig. A-C) from right mammary artery, Left subclavian artery and aortopulmonary window.
**Fig. 8:** Post repair surgery status. Male 13 years old, MDCT Angiography: Multiplanar reformatted images: Closure of VSD through Patch (fig. A-B), no evidence of residual shunts, enlargement of the right ventricular outflow tract (RVOT) (fig. C-D)

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**Fig. 16:** Post repair surgery status. Male 13 years old, MDCT Angiography: Multiplanar reformatted images: enlargement of the right ventricular outflow tract (RVOT) (fig. A). Dilated right ventricle with hypertrophic wall (fig. B). Dilated and calcified conus arteriosus (fig. C-D)

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Fig. 17: Post repair surgery status. Male 5 years old, MDCT Angiography: Multiplanar reformatted images: Multiple muscular ventricular septal defects (circle fig. A-B). Closure VSD Patch with residual shunt (circle fig. C) and a polypoid image (arrow fig. D) probably in relation to vegetation.

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**Fig. 18:** Post repair surgery status. Male 13 years old, MDCT Angiography: Multiplanar reformatted images: Stents in the confluent pulmonary artery branches (fig. A-B), no evidence of thrombus, stenosis or fracture. Incomplete stent apposition (fig. C-D) in the left pulmonary artery (Lpa).

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Fig. 19: Post repair surgery status. Male 5 years old, MDCT Angiography: Multiplanar reformatted images: Apical Amplatzer muscular ventricular septal defect occluder in proper position (fig. A-D)

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Conclusion

1. MDCT is a suitable method for pre-surgical and post-operative follow-up evaluation in patients with Tetralogy of Fallot.
2. Due to new low-dose protocols, extremely low acquisition time and no need for sedation, MDCT is a useful alternative in pediatric patients.
3. MDCT is a suitable alternative in patients in whom the magnetic resonance is contraindicated (implanted devices).
4. A proper understanding of diagnostic findings and potential postsurgical complications is mandatory.
References