Tetralogy of Fallot and its Radiologic Manifestation

Kasia Lipska, Harvard Medical School  Year-III
Gillian Lieberman, MD
What is Tetralogy of Fallot?

The most common cyanotic congenital heart disease characterized by decreased pulmonary blood flow.
Basics of Tetralogy

- It is the most common cyanotic congenital heart defect after infancy and it accounts for 10% of all congenital heart disease cases.
- The degree of obstruction to pulmonary blood flow is the chief determinant of the clinical presentation.
- Associated anomalies: right aortic arch (25%), atrial septal defect (10%), and coronary arterial anomalies (10%)
Basics of Tetralogy

• Clinical presentation:
  Often cyanotic from birth or beginning in the 1st year
  Sudden hypoxic spells in childhood
  Dyspnea and limited exercise tolerance in adulthood
Prognosis and Treatment (in brief)

• Without surgical intervention, most patients die in childhood: 50% die before school age, 25% die before adolescence. Fewer than 10% of cases survive into adulthood.
• Complete surgical correction: closure of VSD and relief of right ventricular outflow obstruction
• Palliative surgery: anastomosis of a systemic artery to a pulmonary artery to increase pulmonary blood flow.
Our patient E.A.

- 55 year old white male
- 3-year history of dyspnea, exertional chest pain, weakness, increased fatigue, and lethargy

Past medical history:
- At age 5, physician noted a “heart murmur” and commented on “a hole in the heart”
- At age 16, E.A. weighed 96 lbs, but had no exercise limitation
- He lead the vigorous life of a fisherman with only mild dyspnea on exertion until age 52
Physical Exam of E.A.

- VS: Pulse 88, BP 130/90
- Chest: clear to auscultation
- Cor: regular rate and rhythm, normal S1/S2, S3/S4, harsh holosystolic ejection murmur at the L sternal border, no right ventricular lift, point of maximum impulse displaced to the L
- Peripheral pulses normal
Menu of tests available for imaging suspected cardiac abnormality

- Plain films of the chest
- Two-dimensional echocardiography
- MR imaging
- Cardiac angiography
  (selective R or L ventriculography)
Plain film

- Characteristic changes in the heart and pulmonary vasculature may be recognizable on a plain chest film
- Fast, inexpensive, non-invasive
- May be useful in the follow-up of patients post-repair
- **Limitations**: findings are often non-specific, plain films do not allow visualization of intracardiac anatomy
Chest X-Rays

• Our patient had 4 plain films of the chest obtained after barium administration to outline the esophagus:
  1. Frontal CXR
  2. Lateral CXR
  3. Left anterior oblique
  4. Right anterior oblique
Frontal Chest X-Ray

Film findings:

- Mild cardiomegaly with narrow cardiac base and rounded elevated apex.
- Right sided aorta
- Concavity in the area of the main pulmonary artery and decreased vascularity

Courtesy of Sven Paulin, M.D., Ph.D. (BIDMC)
Lateral Chest X-Ray

Normal

Patient E.A.

Film findings:
• No definite RV enlargement
• Suggestion of LV enlargement

Courtesy of Sven Paulin, M.D., Ph.D. (BIDMC)
Left Anterior Oblique Chest X-Ray

Normal

Patient E.A.

Film findings:
- Right sided aortic arch

Courtesy of Sven Paulin, M.D., Ph.D. (BIDMC)
Right Anterior Oblique Chest X-Ray

Normal

Patient E.A.

Film findings:
- Right sided aortic arch

Courtesy of Sven Paulin, M.D., Ph.D. (BIDMC)
Differential diagnosis

- **Right aortic arch:**
  - normal variant
  - Tetralogy of Fallot – right sided arch is present in 25-30% of cases, vessels originate from the aorta in exactly opposite order to those arising from a normal aortic arch
  - Persistent truncus arteriosus

- **LVH with clinical signs of L heart failure:**
  - Mild pulmonary stenosis with VSD, i.e. Pink Fallot (L to R shunting)
  - LV failure due to acquired heart disease

- **Other congenital heart defects:**
  -- Patent ductus arteriosus
  -- Transposition of the great arteries
Our patient went on to have
Cardiac Angiography

Report from angiography:

1. On catheterization, the catheter passed from the right to the left ventricle and into the aorta diagnosing a ventricular septal defect.
2. The aorta was right sided.
3. There was demonstrable pulmonary stenosis.
4. Pressure and gas studies were suggestive of left to right shunting.
5. Diagnosis: Pink Tetralogy of Fallot.
Discussion of our patient’s presentation and work-up

Our patient was unusual in:

• presentation – late adult onset and no cyanosis.
• work-up – he was diagnosed in the 1970s when current imaging technology was still unavailable.

Today, cardiac echo and MRI are routine imaging adjuncts to plain film and angiography.

Let’s now review the imaging of some typical patients with classic presentations of Tetralogy of Fallot.
Plain film of Tetralogy of Fallot

(Patient 2: 4-year old boy)

Film findings:

- Heart size is normal
- Cardiac apex is elevated above the diaphragm
- Pulmonary vessels are decreased

http://skiagram.com (SouthBank University, London)
Plain film of Tetralogy of Fallot

- The classic "coeur en sabot," or boot-shaped, cardiac silhouette is caused by the elevation of the right ventricle due to right ventricular hypertrophy, with a concavity in the area of the main pulmonary artery.
- Summary: A cyanotic baby with decreased pulmonary vascularity and a boot-shaped heart is highly suggestive of Tetralogy of Fallot.

Courtesy of Sven Paulin, M.D., Ph.D. (BIDMC)
Two-dimensional Echo

- **Technique**: Multiple ultrasonic beams are transmitted through a wide arc
- Easily accessible, relatively inexpensive
- May be the only imaging required to accurately diagnose TOF and associated lesions
- Detects and displays anatomic anomalies, wall and valve motion, abnormal shunts, intracardiac masses (vegetations, thrombi, tumors)
- Transesophageal echo is especially useful in identifying aortic and atrial abnormalities
Two-dimensional Echo

In Tetralogy, 2-D Echo is used to:
• establish the presence of associated abnormalities,
• assess the severity of right outflow tract obstruction,
• assess the size of the main pulmonary artery and its branches,
• determine the number and size of VSDs.

Limitations: narrow field of view
Two-dimensional transthoracic echo
(Patient 4: a pediatric patient, pre-op)

Echo Findings:
1. VSD
2. Overriding Aorta

University of Kansas Medical Center
http://www.kumc.edu/kumcpeds/cardiology/cardiology.html
Cardiac angiography

In Tetralogy, angiography may be used to:

- confirm the diagnosis
- obtain additional anatomical data – appearance of the right ventricular outflow tract, of the main pulmonary artery, and of its branches, as well as the origin and course of the coronary arteries
- obtain hemodynamic data – severity of R → L shunting, ventricular and arterial pressures

Limitations: Invasive procedure requiring ionizing radiation and intravascular contrast administration, relatively expensive
Right ventricular angiography
(Patient 5: a child with uncorrected Tetralogy)

Film findings:

1. Hypertrophied, trabeculated right ventricle
2. Stenosis at the MPA bifurcation

http://www.neosoft.com/~rlpierce/tof.htm
(Pediatric Cardiology Almanac)
Right ventricular angiography 
(Patient 5: a child with uncorrected Tetralogy)

Film findings:

During systole, there is R → L shunting through a VSD which accounts for the contrast in the aorta.

http://www.neosoft.com/~rlpierce/tof.htm
(Pediatric Cardiology Almanac)
MR Imaging

• **Technique**: spin echo (SE) versus gradient reversal echo (GRE) pulse sequences
• Provides anatomic and functional imaging in arbitrary anatomic section

• SE provides highest contrast resolution, but limited temporal resolution
• GRE has limited contrast resolution but can be used to reconstruct images obtained from many short intervals of the cardiac cycle
MR imaging

• **Limitations**: less accessible than an ultrasound, contraindicated in patients with metallic hardware or claustrophobia

• More expensive than ultrasound, but cheaper than angiography
MR imaging
(Patient 6: 40-year old woman with severe Tetralogy variant - pulmonary atresia and aorto-pulmonary collaterals)

Patient’s MRI:
- Coronal
- Collaterals
- Atretic pulmonary vessels

Line drawing of patient’s anatomy:
- Collaterals from aorta to pulmonary bed

Courtesy of Tal Geva, M.D. (Children’s Hospital)
MR imaging of Tetralogy
(Patient 7: 22-year old man with uncorrected Tetralogy)

Film findings:
1. Right ventricular hypertrophy
2. Pulmonary stenosis
3. Post-stenotic dilatation

Courtesy of Tal Geva, M.D.
(Children’s Hospital)
Summary

• We have reviewed typical and atypical presentations of tetralogy of fallot on plain films, echo, angiography and MR via 7 different patients. Remember TOF in a cyanotic baby with decreased pulmonary vascularity and boot-shaped heart.
References


• Lilly: Pathophysiology of Heart Disease, Second edition, Chapter 3 “Diagnostic imaging and catheterization techniques” pp. 39-65. 1998

Acknowledgements

Many thanks to Dr. Tal Geva, Dr. Gillian Lieberman, Dr. Linda Miles, and Dr. Wayne Monsky. Thanks to Catherine Crosland for her camaraderie during our late hours at the Computer Lab.

Big thanks to Beverlee Turner for her assistance with software, hardware, and with averting impending disasters.

A special thanks to Dr. Sven Paulin for his wonderful guidance and resources.

Special thanks to Larry Barbaras and Ben Crandall our WebMasters