

Case Number 2

Infective Endocarditis in a patient with Tetralogy of Fallot

Gabriella Grech

Reviewed by: Dr. Herbert Felice

Case summary:

Demographic details:

Patient: FG, Male.

Resident in Fgura.

Age: 29 years.

Referred by: Presented at Accident and Emergency.

Mr. FG is a 29-year-old man from Fgura. He is a known case of Tetralogy of Fallot and mitral valve replacement (double-disc prosthetic valve), and presented at Accident and Emergency with a headache and fever. Following history, examination and investigations it was found that the patient was suffering from Subacute Bacterial Endocarditis. He was given antibiotics to treat the infection and due to his high risk of recurrence he continued the antibiotics.

Presenting complaint:

Headache

Fever of 103 degrees

Palpitations

Nausea

These symptoms were of recent onset.

History of presenting complaint:

Three previous episodes of infective endocarditis, the last one being 2 years ago.

Past medical and surgical history:

Past medical history:

- Mild asthma
- Obstructive sleep apnoea

Past surgical history:

- Blalock-Taussig shunt at the age of 9 months
- Mitral valve replacement with a mechanical prosthesis at the age of 7
- Replacement of the prosthetic valve with double-disc prosthesis at the age of 12
- Angiogram – 4 months ago

Drug history:

Drug	Dosage	Frequency	Type	Reason
Amiodarone	200mg	Daily	Anti-arrhythmic	To decrease the risk of developing atrial fibrillation due to multiple heart surgeries.
Warfarin	According to INR	Daily	Anti-coagulant	Due to risk of emboli developing on the mechanical mitral valve.
Fluticasone	100 micrograms	Prn	Inhaled glucocorticoid	Due to mild asthma.

Family history:

The patient has a history of cyanotic heart disease in his family with his uncle suffering from the same condition. His father died of lung cancer at the age of 52.

Social history:

He used to work as a minibus driver but had to stop 4 months ago due to complications of an angiogram. He is a non-smoker and binge drinks heavily - 1 bottle of vodka every weekend. He is active and exercises regularly by playing football and walking.

Systemic inquiry:

- General Health: Looks well in general
- Cardiovascular System: Recent palpitations
- Respiratory System: Some shortness of breath during extreme exercise and sleep apnoea
- Gastrointestinal System: Few vomiting episodes, dysphagia
- Genitourinary System: Nil to note
- Central Nervous System: Sudden headache and fever
- Musculoskeletal System: Nil to note
- Endocrine System: Nil to note

Discussion of results of general and specific examinations:

On examination, the patient had symmetrically warm hands and a pulse of around 60 beats per minute. The JVP was not elevated. There was no sign of anaemia and there was no sign of central cyanosis on examination of the mouth.

On palpation of the precordium a thrill was felt on the left sternal edge.

On auscultation, a pan-systolic murmur (best heard at the apex), high-pitched decrescendo early diastolic murmur (best heard at the upper sternal border) and mid-diastolic murmur (heard best at the apex) were auscultated and the prosthetic click of the valve was heard. Auscultation of the lung bases was normal and there was no sign of sacral oedema.

Differential diagnosis:

- Subacute bacterial endocarditis
- Systemic lupus erythematosus
- Atrial myxoma and other cardiac neoplasms
- Lyme disease
- Antiphospholipid syndrome
- Polymyalgia rheumatica
- Reactive arthritis¹

Diagnostic procedures:

Laboratory exams:

Test: Complete blood count.

Justification for test: To check for sign of infection or anaemia.

Result: Elevated white cell count with an increase in neutrophils.

Conclusion: This supports the idea of subacute bacterial endocarditis.

Test: Urea and Electrolytes.

Justification for test: To assess state of hydration, renal function and electrolyte imbalances.

Result: Normal.

Conclusion: This excludes any renal disease.

Test: Liver Function Tests.

Justification for test: To assess liver function.

Result: Normal.

Conclusion: This excludes any liver disease.

Test: C-Reactive Protein.

Justification for test: Acute phase reactant.

Result: Elevated.

Conclusion: This is an indication of an invasive bacterial condition.

Test: Blood culture.

Justification for test: To detect the presence of actively multiplying bacteria or fungi in the bloodstream, to identify the microorganism present and to guide antimicrobial treatment.

Result: Positive for *Streptococcus mitis*.

Conclusion: This further supports the diagnosis of infective endocarditis.

Instrumental exams:

Test: Electrocardiogram.

Justification for test: To detect any conduction defects.

Result: Right bundle branch block.

Conclusion: This could be a normal variant or due to history of heart defects.

Test: Trans-thoracic Examination.

Justification for test: To check for vegetations or infected tissue.

Result: There is moderate right ventricular outflow tract stenosis with moderate pulmonary regurgitation. There is severe right ventricular dilatation with mildly impaired systolic function. The left ventricle is of

normal size and good systolic function. There is evidence of significant mitral prosthetic stenosis with an estimated valve area of 1.96 cm². There is patient-prosthesis mismatch, however the left atrium is of normal size. The right ventricle is severely dilated with mild right ventricular hypertrophy. There is a left hand sided aortic arch.

Conclusion: The mitral stenosis explains the diastolic murmur, best heard on auscultation at the apex with the patient in the left lateral position. Pulmonary regurgitation explains the high-pitched decrescendo diastolic murmur secondary to pulmonary regurgitation at the upper sternal border and the pansystolic murmur of tricuspid regurgitation was audible in the 4th left intercostal space in the patient as a result of right ventricular dilatation.

Test: Trans-oesophageal Echocardiogram.

Justification for test: To assess for the presence of vegetations or infected tissue.

Result: No vegetations were detected on aortic, tricuspid or mitral valves. The pulmonary valve was not well seen. A shelf-like structure was observed in the left ventricular outflow tract.

Conclusion: This did not confirm the diagnosis of infective endocarditis.

Therapy:

Drugs:

Drug	Dosage	Frequency	Type	Reason
Benzyl penicillin	1.2g	Every 4hrs	Antibiotics	Due to streptococcal infection
Gentamycin	1mg/kg	Every 8hrs	Antibiotics	Due to streptococcal infection

Diagnosis:

Infective endocarditis (IE) is defined as an infection of the endocardial surface of the heart, which may include one or more heart valves, the mural endocardium or a septal defect. Its intracardiac effects include severe valvular insufficiency, which may lead to intractable congestive heart failure and myocardial abscesses. If left untreated, IE is always fatal.

The Duke diagnostic criteria are generally used to make a definitive diagnosis of infective endocarditis. The criteria combine the clinical, microbiologic, pathologic, and echocardiographic characteristics of a specific case²:

Major blood culture criteria for IE include:

- two blood cultures positive for organisms typically found in patients with infective endocarditis,
- blood cultures persistently positive for one of these organisms, from cultures drawn more than 12 hours apart, or
- three or more separate blood cultures drawn at least 1 hour apart.

Major echocardiographic criteria include echocardiogram positive for infective endocarditis, documented by:

- an oscillating intracardiac mass on a valve or on supporting structures, in the path of regurgitant jets, or on implanted material, in the absence of an alternative anatomic explanation,
- myocardial abscess,
- development of partial dehiscence of a prosthetic valve, and
- new-onset valvular regurgitation.

Minor criteria for IE include:

- predisposing heart condition (in this particular case: Tetralogy of Fallot),
- intravenous drug use, fever of 38°C (100.4°F) or higher,
- vascular phenomenon, including major arterial emboli, septic pulmonary infarcts, mycotic aneurysm, intracranial haemorrhage, conjunctival haemorrhage, or Janeway lesions,
- immunologic phenomena such as glomerulonephritis, Osler nodes, Roth spots, and rheumatoid factor,
- positive blood culture results not meeting major criteria or serologic evidence of active infection with an organism consistent with infective endocarditis, and
- echocardiogram results consistent with IE but not meeting major echocardiographic criteria.

A definitive clinical diagnosis can be made based on the presence of:

- 2 major criteria;
- 1 major criterion and 3 minor criteria or;
- 5 minor criteria³.

Congenital heart disease is one of the risk factors of IE. Tetralogy of Fallot represents the largest proportion of IE cases. Repaired Tetralogy of Fallot often has residual aortic regurgitation - a substrate for aortic valve IE. Moreover in this case there was prosthetic valve replacement increasing the predisposition to infection⁴.

Final treatment and follow up:

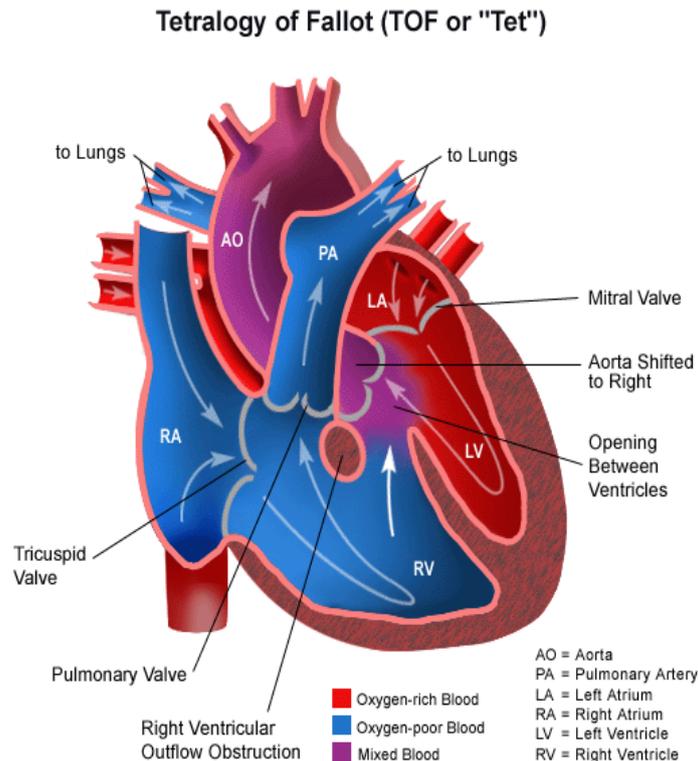
He was given a 2 week course of intravenous benzyl penicillin and gentamicin and was advised to continue benzyl penicillin for 2 weeks prophylaxis. The patient was given an appointment to be followed up at 'Grown-ups with congenital heart defects' clinic.

Fact Box 2:

Alexia Grech

Name of Condition: Tetralogy of Fallot

Tetralogy of Fallot is a congenital heart defect⁵ which involves four abnormalities of the heart. These include ventricular septal defect (VSD), pulmonary stenosis, misplaced aorta (often described as an aorta over riding the septum) and right ventricular hypertrophy. They usually result in an insufficient amount of oxygenated blood reaching the body⁶.



Risk factors:

The exact cause of Tetralogy of Fallot is not yet known but there are several factors and conditions that may increase the risk of having a child with this heart defect, which are associated with the mother carrying the child during pregnancy. These include: viral illness, alcoholism, poor nutrition and age (older than 40)⁸. The condition is also seen more in babies with Down syndrome and DiGeorge syndrome⁹.

Symptoms and Signs:

- Peripheral & central cyanosis.
- Shortness of breath and difficulty in breathing especially during exertion.
- Fainting¹⁰.
- Clubbing of fingers and toes¹¹.
- Growth and development in children are slower¹⁰.
- Cervical lymphadenopathy (> 15 mm diameter, usually unilateral, single, non-purulent and painful).

Investigation to confirm diagnosis:

- Heart murmurs upon auscultation.
- Electrocardiogram: shows the four abnormalities of the heart which are ventricular septal defect (VSD), pulmonary stenosis and misplaced aorta (often described as an aorta over riding the septum) and right ventricular hypertrophy.
- Chest X Ray: shows cardiomegaly and pulmonary oedema¹².
- Treatment: Tetralogy of Fallot is repaired with open-heart surgery, either soon after birth or later in infancy. The surgery aims to repair the defects: the ventricular septal defect, pulmonary stenosis, and misplaced aorta so the heart can work as normally as possible¹³.

Long term complications:

- Incompetent heart valves
- Arrhythmias
- Pulmonary artery branch stenosis
- Right ventricular aneurysms
- Residual ventricular septal defects
- Coronary artery disease

Prognosis:

The outlook for a child born with Tetralogy of Fallot is much better today than it was in the past. Advances in testing and treatment mean that most children who have this congenital heart defect survive into adulthood. However, they require long-term care provided by specialists in order to remain as healthy as possible¹⁴.

References:

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