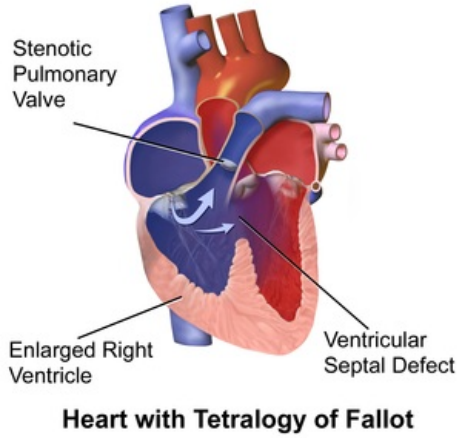
Tetralogy of Fallot

Tetralogy of Fallot is, along with transposition of the great arteries, the most common cyanotic/critical congenital heart defect. It is characterized by a combination of 4 anomalies:

1. Ventricular septal defect;
2. dextroposition aorta, sits above the septal defect;
3. pulmonary stenosis – infundibular or valvular, hypoplasia of the pulmonary trunk and branches may also be present;
4. hypertrophy of the right ventricle (RV).

If a atrial septal defect is also present, we speak of **pentalology of Fallot**.



Tetralogy of Fallot

This combination causes decreased pulmonary flow and cyanosis. Cyanosis is more severe the tighter the pulmonary stenosis. Blood from the right ventricle (deoxygenated) is thus ejected into the descending aorta (right-left shunt). Pulmonary stenosis and right ventricular hypertrophy usually progress, and hypoxemia deepens at the same time. Sometimes secondary pulmonary atresia can occur with severe cyanosis and the development of multiple aorto-pulmonary collaterals. On the contrary, with mild pulmonary stenosis, the cyanosis does not have to be severe and the patient can live to adulthood even without surgery (the so-called pink Fallot). [1]

Obstruction of blood flow through the lungs can sometimes lead to a complete blockage, resulting in the clinical picture of **hypoxic seizures** (so-called *spelling*). The child suddenly turns gray/blue (skin, nails, lips) after crying or feeding or when restless. These seizures are most common in infants around 2 to 4 months of age. First aid consists of laying on the side and pulling the knees to the chest (older children instinctively squat in these situations) [2]

Surgical solution depends on the age and size of the child, the anatomy of the pulmonary vascular bed, the size of the pulmonary valve annulus, the degree of right ventricular hypertrophy, the anatomy of the coronary arteries and the presence of associated heart defects. The optimal primary correction between 2 and 12 months of age is transatrial correction, in which the infundibular stenosis is resected and the VSD is closed via the right atrium and pulmonary artery. After surgical correction, the risk of late complications remains, such as pulmonary stenosis, pulmonary valve insufficiency, arrhythmia and right-sided heart failure. [3]

Clinical picture

- Cyanosis,
- heart failure,
- not thriving,
- exertional dyspnea with relief when squatting,
- paroxysmal hypercyanotic seizures (or hypoxic seizures); they last minutes to hours.

Systolic murmur is caused by blood flow through the narrowed infundibulum, it disappears during a seizure (!).

Apart from very severe forms of tetralogy, we only see cyanosis during times of increased physical activity in the second half of the first year. If the cyanosis lasts long, clubbed fingers will develop.

A hypoxic attack is caused by a transient excessive contraction of the muscles of the outflow tract of the right ventricle (infundibula), which accentuates the stenosis of the truncus pulmonalis, reduces the flow of blood through the pulmonary stream, increases the right-left shunt through the ventricular defect, and reaches the aorta greater proportion of desaturated blood. Small children (newborns) have deepened and accelerated breathing, pronounced cyanosis, cry and may even lose consciousness during a hypoxic attack. In toddlers and older children, seizures are not as dramatic, forcing them into a squat; squatting increases venous return and increases resistance in the peripheral arterial bed, thereby increasing pulmonary flow. [4]

The natural course is always unfavorable, cyanosis progresses, embolizations to the brain occur.

Diagnosis

- Echocardiography,
- EKG; right ventricular hypertrophy,
- systolic murmur above the lung,
- complicated forms need to be examined angiographically (mainly the pulmonary canal).

The precordium is quiet, the second sound is quiet, the blood flow through the stenotic infundibulum produces a systolic murmur (it can be heard along the left edge of the sternum).

Therapy

Hypoxic seizures can be temporarily controlled with beta-blockers, newborns with critical hypoxemia are given prostaglandin E1 to maintain airway patency. Some conditions are suitable for emergency balloon valvuloplasty.

Confirmed tetralogy of Fallot is always indicated for surgery:

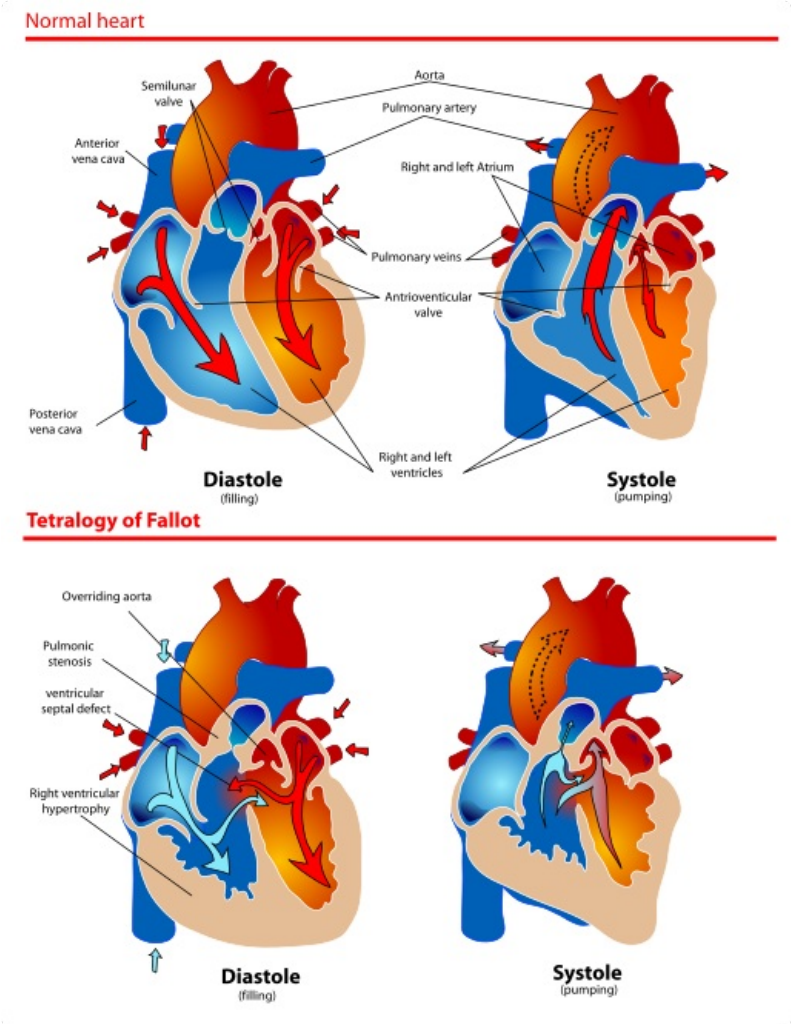
- **corrective surgery:** we indicate it as planned in toddlerhood - removal of the stenosis and expansion of the outflow part of the right ventricle by infundibulectomy, closure of the ventricular septal defect with a patch and plastic surgery of the lung.[5] The functional result is usually excellent and permanent. Regurgitation (lower performance of patients) can occur due to obstruction of the stenotic valve.
- **palliative surgery:** coupling between the pulmonary artery and a. subclavia – modified coupling according to Blalock-Taussig or coupling with Gore-Tex® prosthesis (PTFE 5-6 mm).

[5]

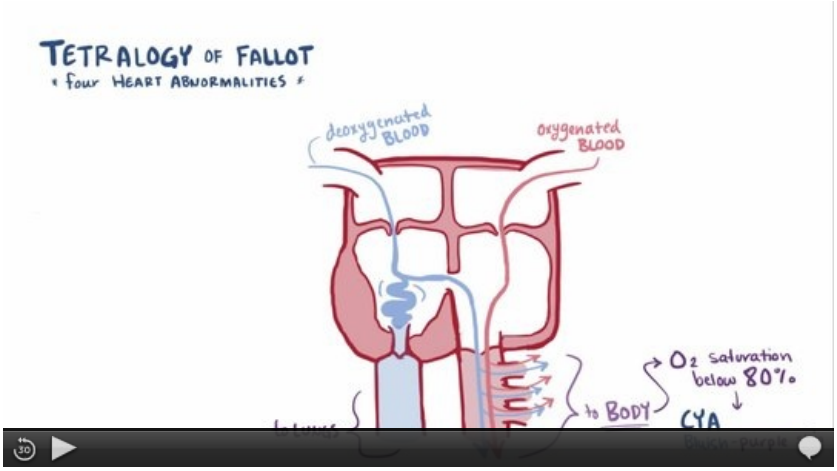
Prognosis

Untreated tetralogy of Fallot is accompanied by a right-to-left shunt, chronic cyanosis, and polycythemia. It is a predisposition to ischemic stroke, brain abscess, bacterial endocarditis and congestive heart failure.

Diagram of diastole and systole



Summary video



Definition, pathogenesis, symptoms, complications, treatment.

Links

Related article

- Congenital heart defects
- Acquired heart defects
- Central cyanosis

References

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2. https://www.mayoclinic.org/diseases-conditions/tetralogy-of-fallot/symptoms-causes/syc-20353477
3. TLÁSKAL, T. *KOREKCE FALLOTOVY TETRALOGIE – ČASOVÁNÍ A CHIRURGICKÉ TECHNIKY* [online]. [cit. 2013]. <http://www.cksonline.cz/21-vyrocni-sjezd-cks/sjezd.php?p=read_abstrakt_program&idabstrakta=511>.
4. – PROVAZNÍK, Kamil – HEJCMANOVÁ, Ludmila. *Preklinická pediatrie.* 2. edition. 2007. pp. 114. ISBN 978-80-7262-438-6.
5. VANĚK, Ivan. *Kardiovaskulární chirurgie.* 1. edition. 2003. 236 pp. ISBN 8024605236.

External links

- Fallotova tetralogie (česká wikipedie)
- Tetralogy of Fallot (anglická wikipedie)
- Tetralogy of Fallot (video na youtube.com) (https://www.youtube.com/watch?v=uHv2RHnTpJg)

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