

Fallot's tetralogy

VV Fallot's tetralogy, along with large artery transposition, is the most common cyanotic / critical congenital heart defect. It is characterized by a combination of 4 anomalies:

1. *Ventricular septal defect;*
2. *dextroposition of the aorta, abuts the septal defect;*
3. *pulmonary stenosis* - infundibular or valvular, hypoplasia of the trunk and branches of the lung may also be present;
4. *right ventricular hypertrophy (PK).*

If an atrial septal defect is also present, we speak of Fallot's pentalogy.

This combination causes decreased lung flow and cyanosis. The tighter the cyanosis, the tighter the pulmonary stenosis. Blood from the right ventricle (deoxygenated) is thus expelled into the adjacent aorta (right-left short circuit). Pulmonary stenosis and right ventricular hypertrophy usually progress and at the same time hypoxemia worsens. Sometimes secondary pulmonary atresia with severe cyanosis and the development of multiple aorto-pulmonary collaterals can occur. On the contrary, with mild pulmonary stenosis, cyanosis may not be severe and the patient may live to adulthood without surgery (so-called pink fallot).

Obstruction of blood flow through the lungs can sometimes lead to complete occlusion, creating a clinical picture of hypoxic attacks (so-called spelling). The baby suddenly turns gray / bluish (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 placing your knees on your side and pulling your knees to your chest (older children instinctively sit in a squat in these situations).

The **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 primary correction is between the 2nd and 12th month of age, namely the transatrial correction, in which the infundibular stenosis is resected and the VSD closes through the right atrium and lungs. After surgical correction, there is a risk of late complications such as pulmonary stenosis, pulmonary valve insufficiency, arrhythmias and right heart failure.

Clinical picture

- cyanosis
- heart failure
- failing,
- exertional dyspnoea with relief while sitting squatting,
- paroxysmal hypercyanotic seizures (or hypoxic seizures); they take minutes to hours.

The systolic murmur is given by the blood flow through the narrowed infundibule, it disappears during a seizure (!).

In addition to many severe forms of tetralogy, we see cyanosis only at a time of increased physical activity in the second half of the first year. If cyanosis lasts a long time, clubbed fingers develop.

The hypoxic attack is caused by a transient excessive contraction of the right ventricular outflow tract muscle, which accentuates the stenosis of the truncus pulmonalis, reduces blood flow through the pulmonary circulation, increases the right-left short circuit through the ventricular defect and a larger proportion of desaturated blood enters the aorta. Young children (newborns, infants) have deep and rapid breathing, severe cyanosis, crying and may even lose consciousness during a hypoxic attack. For toddlers and older children, the seizures are not so dramatic, they force them to squat; squatting increases venous return and increases resistance in the peripheral arterial bed, increasing lung flow.

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

Diagnosis

- Echocardiography
- ECG; right ventricular hypertrophy,
- systolic murmur above the lungs,
- complicated forms need to be examined angiographically (especially the pulmonary circulation).

The precordium is calm, the second sound is quiet, 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 affected by beta-blockers, newborns with critical hypoxemia are given prostaglandin E1 to maintain ductal patency. Some conditions are suitable for urgent balloon valvuloplasty.

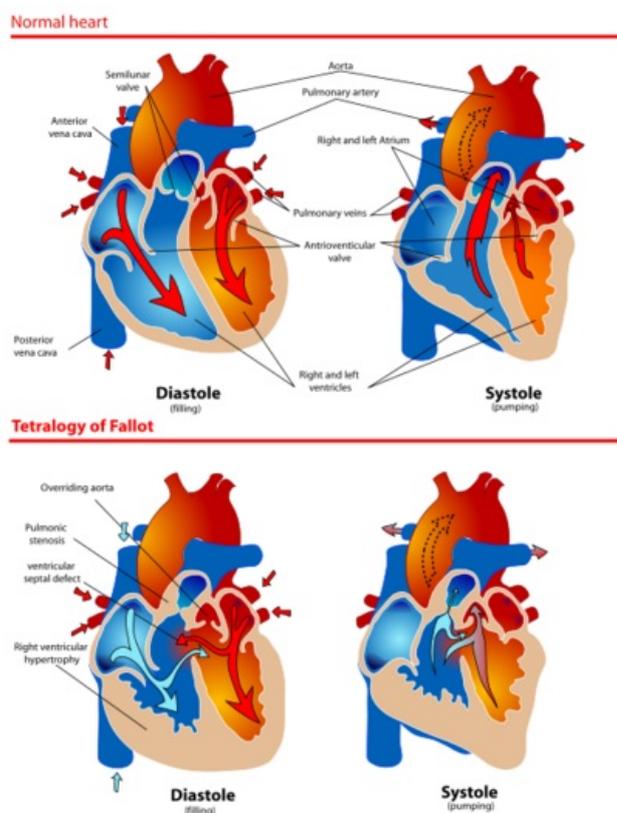
The confirmed Fallot tetralogy is always indicated for the operation:

- **corrective surgery:** we indicate it planned in infancy - removal of stenosis and dilation of the outflow part of the right ventricle by infundibulectomy, closure of the ventricular septal defect with a patch and pulmonary plastic surgery. The functional result is usually excellent and lasting. Regurgitation (lower patient performance) may occur due to the stenotic valve.
- **palliative surgery:** joint between the lungs and the subclavian artery - modified Blalock-Taussig joint or Gore-Tex® prosthesis joint (PTFE 5-6 mm).

Prognosis

Untreated Fallot's tetralogy is accompanied by right-left shunting, chronic cyanosis, and polycythemia. It predisposes to ischemic stroke, brain abscess, bacterial endocarditis and congestive heart failure.

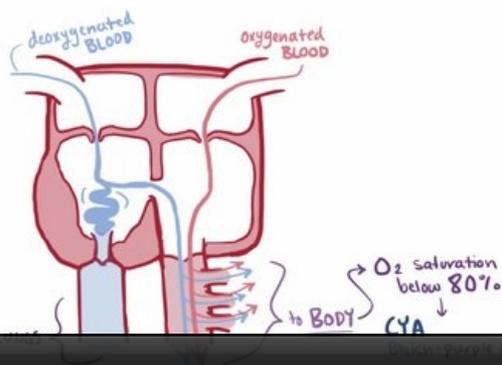
Scheme of diastole and systole



Summary video

TETRALOGY OF FALLOT

Four HEART ABNORMALITIES



Video in English, definition, pathogenesis, symptoms, complications, treatment.

Links

related articles

- Congenital heart defects
- Acquired heart defects
- Central cyanosis

Reference

1. <https://www.prolekare.cz/casopisy/vnitri-lekarstvi/2015-12/fallotova-tetralogie-drive-a-dnes-editorial-57206>
2. <https://www.mayoclinic.org/diseases-conditions/tetralogy-of-fallot/symptoms-causes/syc-20353477>
3. TLÁSKAL, T. CORRECTION OF FALLOT TETRALOGY - TIMING AND SURGICAL TECHNIQUES. Paper presented at the XXI Conference. annual congress of the Czech Society of Cardiology. Brno. 4-5.5.2013 Also available from <http://www.cksonline.cz/21-vyrocní-sjezd-cks/sjezd.php?p=read_abstrakt_program&idabstrakta=511>.
4. LEBL, Jan, Kamil PROVAZNÍK and Ludmila HEJCMANOVÁ, et al. Preclinical pediatrics. 2nd edition. Prague: Galén, 2007. pp. 114. ISBN 978-80-7262-438-6.
5. VANEK, Ivan, et al. Cardiovascular surgery. 1st edition. Prague: Karolinum, 2003. 236 pp. ISBN 8024605236.

External links

- Fallot's tetralogy (Czech wikipedia)
- Tetralogy of Fallot (English wikipedia)
- Tetralogy of Fallot (video on youtube.com) (<https://www.youtube.com/watch?v=uHv2RHnTpJg>)

Reference

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- TASKER, Robert C., Robert J. MCCLURE and Carlo L. ACERINI. Oxford Handbook by Pediatrics. 1st edition. Oxford University Press, 2008. pp. 248. ISBN 978-0-19-856573-4.