

Transposition of the Great Arteries

Template:Infobox - VVV thumb|300px|Traspozice velkých cév, defekt síňového septa a průchozí *ductus arteriosus Botalli* thumb|250px|RTG: transpozice velkých tepen **Transposition of the great vessels** (TGV) is a congenital heart defect, in which the **systemic** and **pulmonary** circulation is **devided into two parallel circuits**. Both main arteries arise from wrong chambers and therefore form two separate circuits: aorta arises from the right ventricle and the main pulmonary artery from the left ventricle.

Deoxygenated venous blood from the body is pumped from the right atrium to the right ventricle and through aorta back into the systemic circulation. Oxygenated blood from lungs goes through the left atrium, left ventricle and main pulmonary artery back into the lungs.

This defect is compatible with life **only in the presence** of atrial septal defect, ventricular septal defect or persistent ductus arteriosus. . In this case mixed (arterial and venous) blood circulates in the systemic circulation. Children are **less cyanotic** when a septal defect is present, but they develop pulmonary hypertension and heart failure may occur sooner. The combination of transposition of great vessels, septal defect and pulmonary stenosis causes cyanosis, but usually not heart failure nor pulmonary hypertension.

Clinical manifestation

This is a critical heart defect with early manifestations of severe hypoxia. Clinical signs include:

- cyanosis,
- tachypnea,
- signs of severe heart failure,
- hyperactive precordium,
- no murmur can usually be heard (unless combination of defects is present).

Diagnosis

- Echocardiography.

Treatment

Maintaining normal body temperature is very important, because hypothermia would worsen the metabolic acidosis caused by hypoxia. prostaglandin E1 is administered prior to the surgery. It improves the oxygenation of systemic blood by keeping ductus arteriosus Botalli open. Atrial septostomy can also be performed. In the first two weeks of life the patient undergoes **corrective surgery**.

- **Anatomical correction** (arterial switch) is the method of choice. The transposed arteries are put into their correct place. This challenging operation includes the transfer of coronary arteries to the future aorta. It is necessary to perform this procedure in the first days of the newborn's life. Functional results are excellent, long-term experiences are not available yet.
- **Rastelli** procedure is indicated when septal defect and pulmonary stenosis are present as well. (This combination is not suitable for anatomical correction). The septal defect is closed so that blood flows from the left ventricle to the displaced aorta and an extracardiac conduit is placed between the right ventricle and the pulmonary arteries (it needs to be changed during patient's life).
- **Functional correction** is nowadays usually not performed. The principle is atrial correction (Mustard or Senning procedure) - the venous inflow into the atria is transferred to opposite chambers using baffle (tunnel). Blood from the superior and inferior vena cava is directed to the mitral valve and then to the left ventricle and main pulmonary artery, blood from the pulmonary veins is directed to the tricuspid valve and then to the right ventricle and aorta. The anatomy of the heart is transposed (aorta arises from the right ventricle and main pulmonary artery from the left). This surgery used to be performed at the age of 3-8 months. The immediate functional result is excellent, but long term results are not. Sometimes there is a loss of sinus rhythm and various arrhythmias occur, these can lead to sudden death. Right ventricular function deteriorates and begins to fail (can't meet the demands of systemic circulation).

Without surgery the prognosis is very poor, most patients die within the first days of their life.

Links

Související články

- Vrozené srdeční vady
- Získané srdeční vady

Externí odkazy

- Transposition of great arteries (YouTube video) (<https://www.youtube.com/watch?v=FGPEua3iBVY>)

Zdroj

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Použitá literatura

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Kategorie:Patologie Kategorie:Vnitřní lékařství Kategorie:Kardiologie Kategorie:Pediatric Kategorie:Chirurgie