FETAL TETRALOGY OF FALLOT

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ABSTRACT — Tetralogy of Fallot (ToF) is a common form of congenital heart disease (CHD). Frequency of occurrence is 1/3600 live births and 3,5–7% of infants with CHD. TOF is 5% to 10% of all congenital heart disease. Our database has included 112 cases of ToF in 2013–2016. Definition of fetal ToF was attempted from multiple ultrasound scan planes. Initial fetal echocardiograms were obtained between 12 and 39 weeks of gestation (m = 24.5 weeks). In all cases there was postnatal examination by echocardiography, angiography, surgery or autopsy. Prenatal diagnosis was confirmed in 100%. The termination of pregnance took place in 5 cases.
107 cases have been subjected cardiac surgery in our center and survival rate was 97%.

Tetralogy of Fallot (ToF) is a common form of congenital heart disease (CHD). ToF includes dextraposition of aorta, aortic ventricular septum defect, right ventricular outflow obstruction and hypertrophy of right ventricle in postnatal period. We will not have hypertrophy of right ventricle before birth, fetal right ventricle is larger, left ventricle is normal. Frequency of occurrence is 1/3600 live births and 3,5–7% of infants with CHD. ToF is 5% to 10% of all congenital heart diseases [2].

Classification of ToF

- Pale form (absent or minimum of right ventricular outflow obstruction);
- Classic form (middle and strong pulmonary stenosis);
- Extreme form (Atretic pulmonary valve);
- TF with absent pulmonary valve syndrom.

RVOT obstruction in ToF occur at following levels [1, 2, 3].

- 1. Infundibular stenosis 45%;
- 2. Pulmonary valve stenosis 10%;
- 3. Valvular and infundibular combination in 30%;
- 4. Atretic pulmonary valve in 15%.

Concomitant intracardial pathology:
25% of ToF associated with right aortic arch, 5% of ToF have anomalous coronary arteries, 2% of TOF are associated with atrioventricular septal defect.

Extracardial pathology:
trachea-esophageal fistula, cleft lip, abdominal wall defects, venrticulomegaly, renal anomalies [4,6].

Chromosome complement are 22q11deletion, trisomy 21, trisomy 13, trisomy 18.

Morphology

Anterior craniocaudal or cephalad deviation of the insertion of the muscular outlet septum torgether with hypotrophy of trabeculations on the infundibular free wall constitute the essential features of ToF. Instead of nestling berween the anterior and posterior limbs of the trabecula septomarginalis, the outlet septum is displaced and typically fuses with the anterior limb. This brings the aorta over the ventricular septum so that it has a biventricular origin and accounts for the malalignment ventricular septal defect. The displaced outlet septum, torgether with the trabeculations of the parietal or free wall, produce muscular sub-valvar right ventricular outlet obstruction [2].

HEMODYNAMICS

Due to overriding, aorta receives blood from both right ventricle and left ventricle. Ventricular sep-

tal defect (VSD) is nonrestrictive meaning no gradient across VSD. Pressure difference between in heart's cavieties is 10-15 mm Hg before the birth. There is the general circulation and lungs do not function. ToF is compensated before the birth.

After the birth ToF is cyanotic CHD. RV preferentially ejects into arota because of infundibular stenosis. Hypoxemia took place because took place the infundibular pulmonary stenosis.

MATERIALS AND METHODS

Our database was reviewed for cases with a diagnosis of ToF seen between 2013–2016. There were 112 cases of ToF.

Definition of fetal ToF was attempted from multiple scan planes including basic views: four-chamber view, five-chamber view, long-axis view of left and right ventricle, short view of aortic valve, and, three vessel view. (Fig. 1–8).

Doppler color flow mapping and pulsed Doppler interrogation were used to facilitate identification of great vessel relationship, location and severity of ventricular outflow obstruction and the measurement of maximal pulmonary velocity.

Methods of the echocardiographic identification of ToF were postnatal examination by echocardiography, angiography, surgery or autopsy. Initial fetal echocardiograms were obtained between 12 and 39 weeks of gestation (m = 24.5 weeks).

RESULTS

There were 112 cases of ToF, including : ToF with: middle pulnomary stenosis — 75, strong stenosis of PA — 27, extreme form — ToF and atresia of pulnonary artery – 1, ToF with absent pulmonary valve syndrom — 9.

The level of pulmonary obstruction was determined according to the next formula (supplement 1).

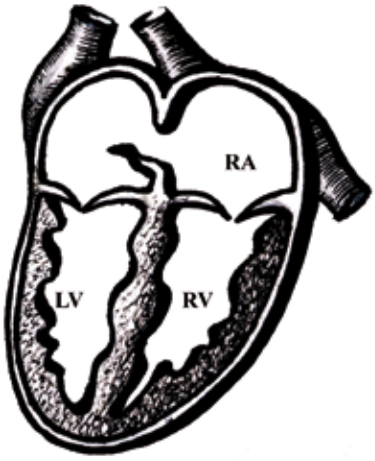


Fig.1A. *four chamber view*

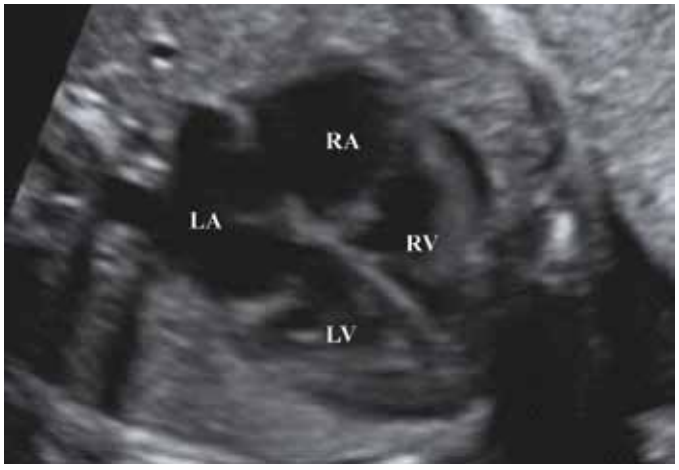


Fig. 1B. *22 week of gestation*

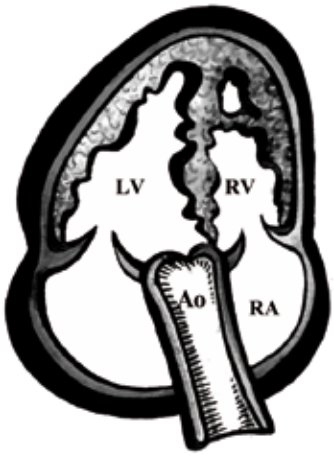


Fig. 2A. *Five chamber view*

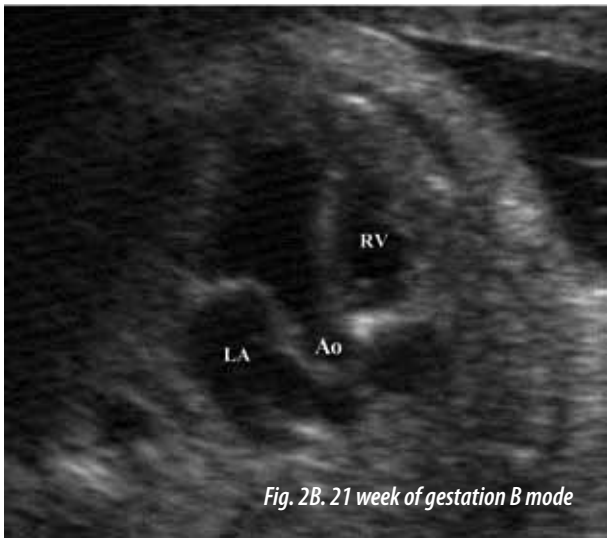


Fig. 2B. 21 week of gestation B mode

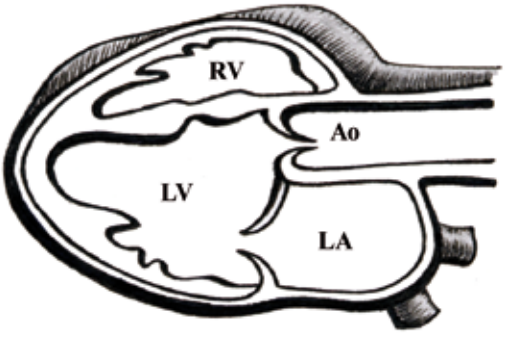


Fig. 3A. *LV outlet tract*

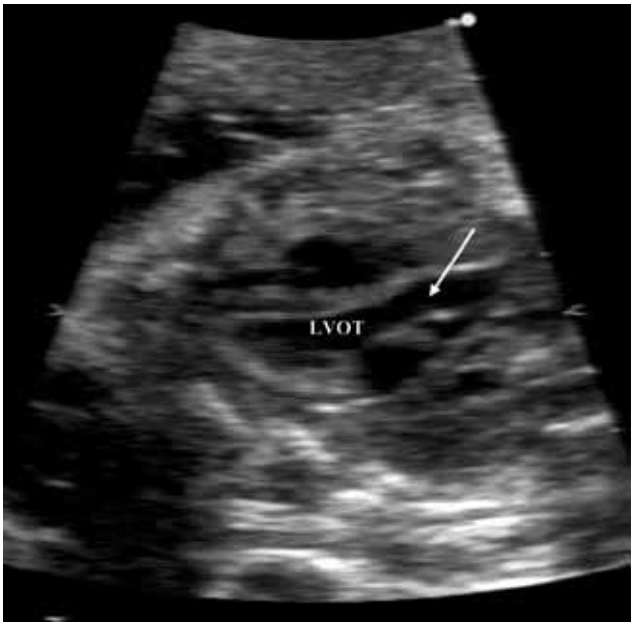


Fig. 3B. *20 week of gestation, B mode*

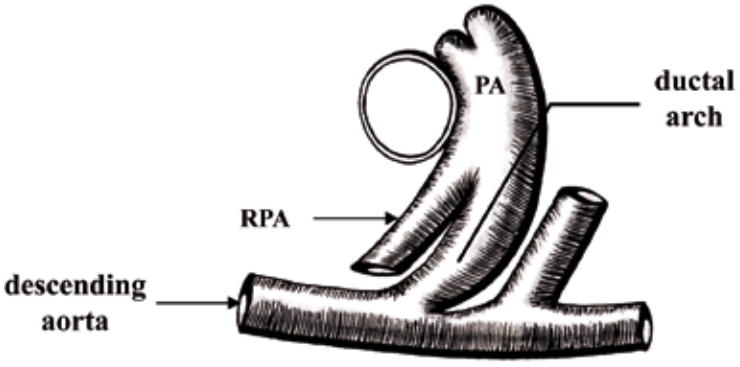


Fig. 4A. *Ductal arch*

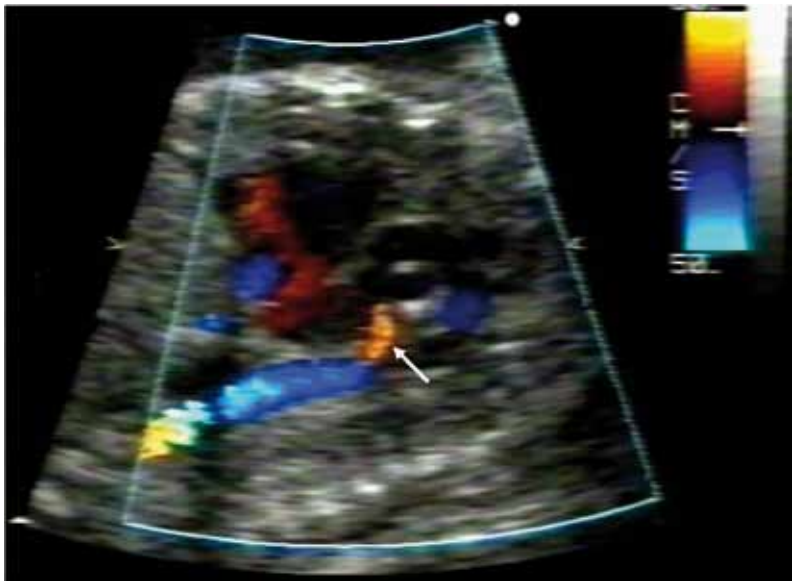


Fig. 4bB. *Color Doppler mapping. Modified aortic arch projection showing the aorta throughout. The arrow indicates the open arterial duct, which connects with the descending aorta*

Regress of pulmonary artery growth from middle pulmonary stenosis to strong pulmonary stenosis took place in 10 cases between 26–34 weeks of gestation. There were 6 cases of ToF with middle stenosis pulmonary artery and atrioventricular septal defect, 5 cases of ToF with severe pulmonary stenosis and face's, abdominal's and renal anomalies . There was chromocome abberation (Down syndrome) in one case.

Postnatal verification took place in all cases. Prenatal diagnosis was confirmed in 100%.

All patients with ToF were offered the cardiosurgerist, genetic's specialist. The delivery of all babies with this CHD was in specialized maternity hospitals. The method of delivery was determined by odstetric and somatic status of pregnant women.

The termination of pregnance took place in 5 cases. There were cases of ToF with severe pulmonary stenosis and extracardial pathology.

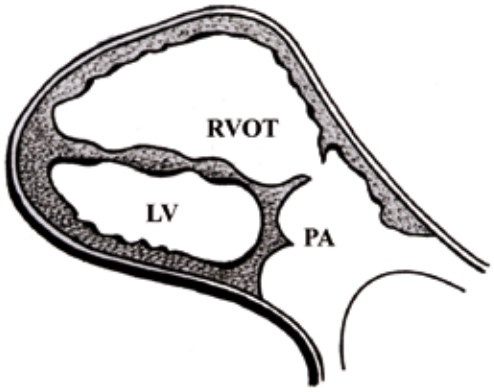


Fig. 5A. *Long axis view of the right ventricle*

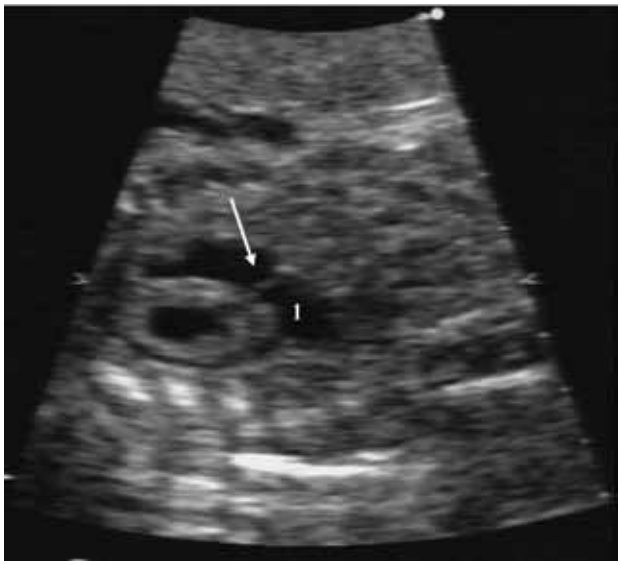


Fig. 5B. *19 week of gestation, B mode*

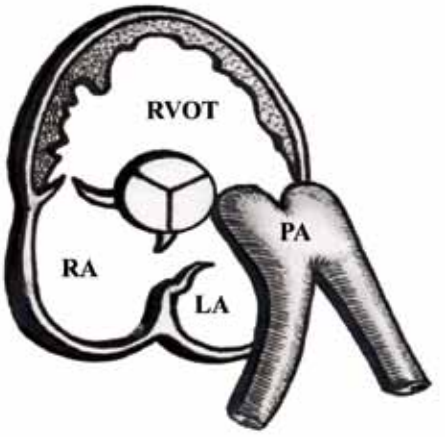


Fig. 6A. *Short axis view of the aortic valve*

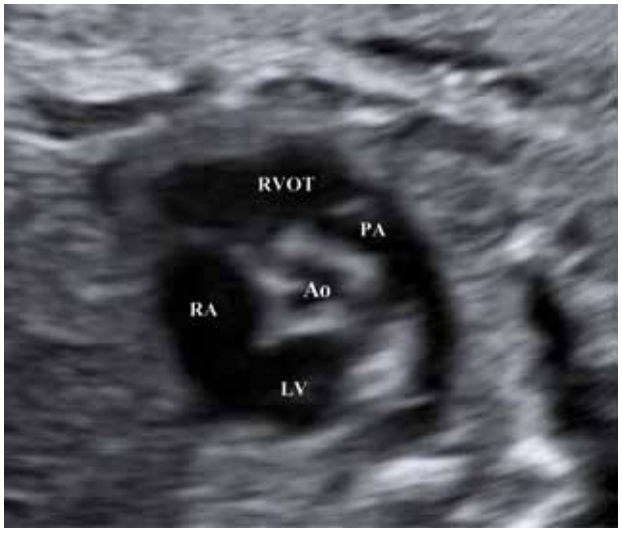


Fig. 6B. *20 week of gestation, wB mode*

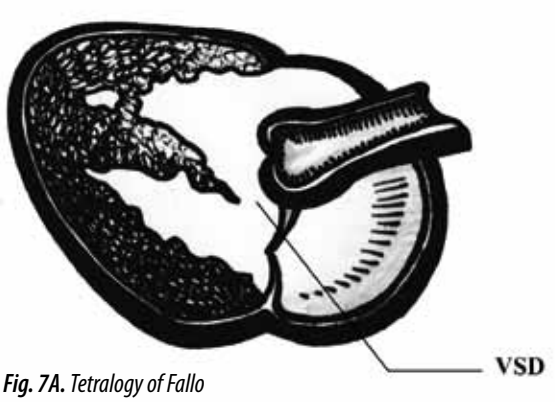


Fig. 7A. *Tetralogy of Fallo*

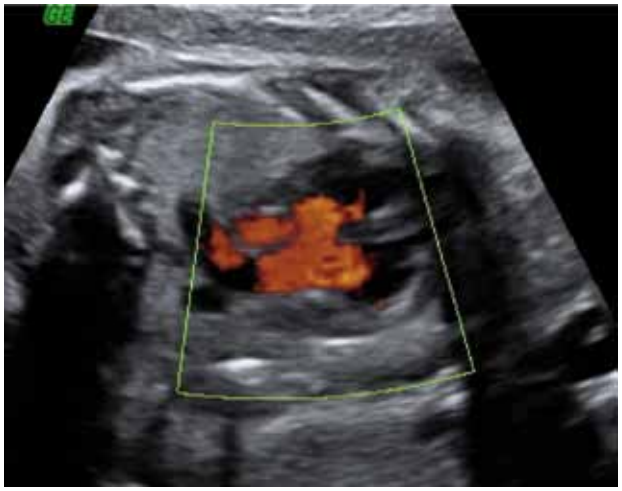


Fig. 7B. *31 week of gestation, Power Doppler Mode*

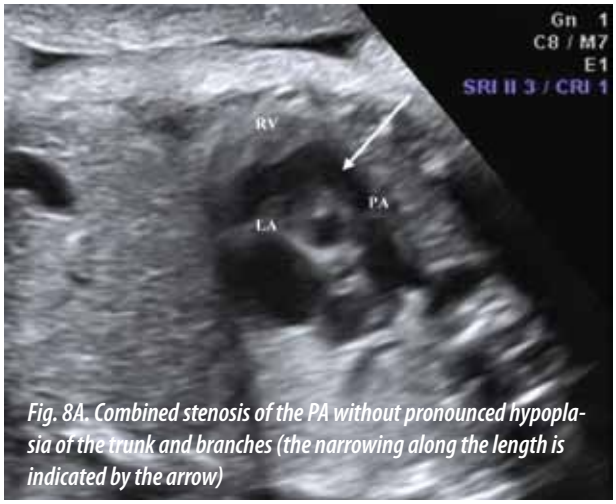


Fig. 8A. Combined stenosis of the PA without pronounced hypoplasia of the trunk and branches (the narrowing along the length is indicated by the arrow)

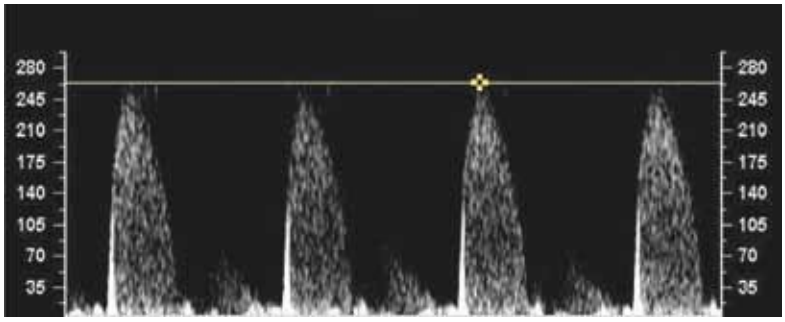
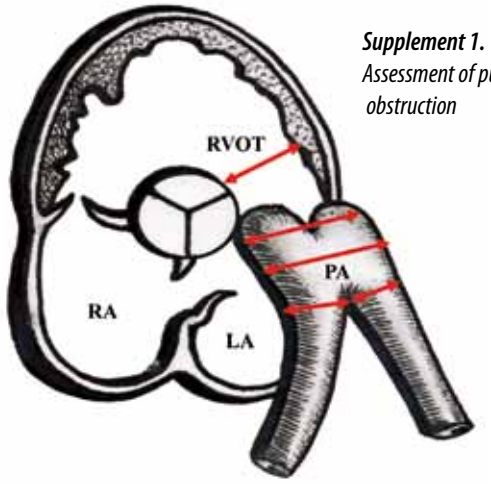


Fig. 8B. *34 week of gestation, Pulsed-wave dopplerography*



Supplement 1.
Assessment of pulmonary obstruction

Short-axis view of the aortic valve. Measurements of pulmonary levels: RV outlet tract, pulmonary valve, mean pulmonary artery, right, left pulmonary arteries for comparison with the rate of gestation
*[D PA measurement. (mm) / D PA coome. AG (mm)] * 100%*

Middle obstruction < 75% vs normal parameters AG ;
Strong obstruction 75–50% vs normal parameters AG
Severe obstruction 50–25%; vs normal parameters AG
Critical obstruction >25% vs normal parameters AG

107 cases have been subjected cardiac surgery in our center and survival rate was 97% . There were isolated form of ToF.

DISCUSSION

Tetralogy of Fallot is a common prenatal diagnosis. Prenatal detectability of this CHD was excellent. But very important to know anatomical details of ToF in every case.

Postnatal prognosis was determined by level and power of pulmonary's stenosis and presence of a combined pathology. The most cases in our cohort had middle stenosis of PA (66,9%) in the same time all cases of atrioventricular septal defect took place in this group. The severe form of TF were isolated heart pathology. Respectively , it was no simple to evaluate the postnatal prognosis in every case.

Our data showed , when the diagnosis of ToF was made in utero, karyotype , detailed anatomical survey and detailed general ultrasound scrinning should be offered in every case. It was important to do the monitoring pulmonary artery growth in fetal life and look for associated extracardiac anomalies throughout pregnance.

Expert review of anatomy of ToF, elimination of combined pathology was positive background for successful surgical repair. Our Center had good experience of ToF's surgical repair. Survival rate was 98% in 10 years after surgical repair during 1 year after birth [1].

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