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Late arrhythmia in patient with repaired tetralogy of Fallot

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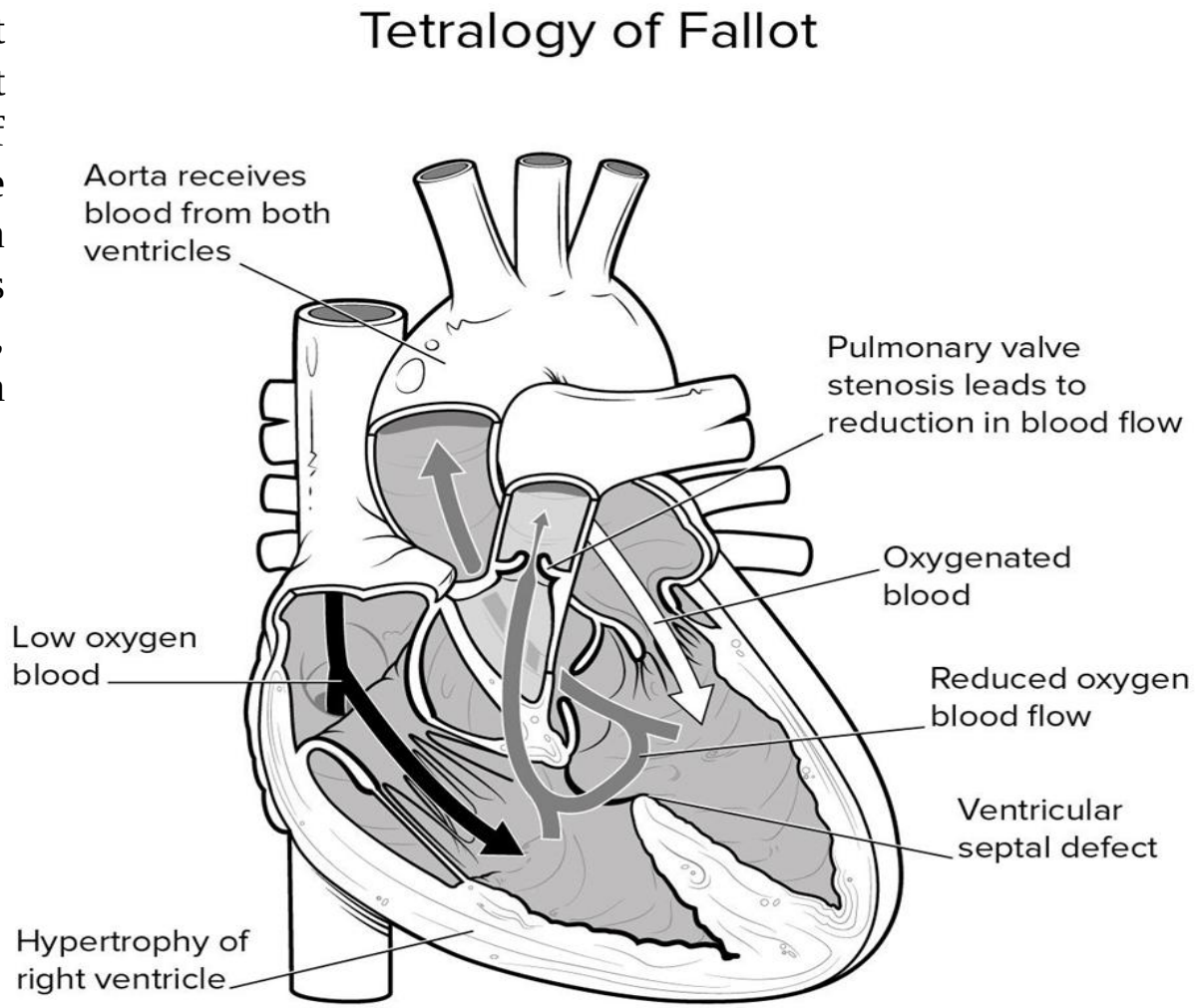
2017

Tetralogy of Fallot (TOF) is one of the most common congenital heart disorders (CHDs). This condition is classified as a cyanotic heart disorder, because tetralogy of Fallot results in an inadequate flow of blood to the lungs for oxygenation (right-to-left shunt) (see the following image). Patients with tetralogy of Fallot initially present with cyanosis shortly after birth. The mortality rate in untreated patients reaches 50% by age 6 years, but in the present era of cardiac surgery, children with simple forms of tetralogy of Fallot enjoy good long-term survival with an excellent quality of life.



Complete Intracardiac Repair

Primary correction is the ideal operation for treatment of tetralogy of Fallot (TOF) and is usually performed under cardiopulmonary bypass (CPB). The aims of the surgery are to close the ventricular septal defect (VSD), resect the area of infundibular stenosis, and relieve the right ventricular (RV) outflow tract obstruction (RVOTO).

Temporary or Palliative Surgery

This surgery improved blood flow to the lungs (a shunt between a large artery branching off the aorta and the pulmonary artery. One end of the shunt is sewn to the artery branching off the aorta. The other end is sewn to the pulmonary artery). A complete repair of the four defects was done later in childhood (as in our patient case).

https://emedicine.medscape.com/article/2035949-overview

The four components that make up the "tetralogy" include:

- **a ventricular septal defect (VSD);**
- **pulmonary stenosis (subvalvar, valvar and/or supravalvar);**
- **an overriding aorta;**
- **right ventricular hypertrophy (RV).**

Reasons of late arrhythmia in patient with repaired tetralogy of Fallot

The incidence of atrial arrhythmias after TOF repair is relatively high, about 30%, including atrial fibrillation, flutter, focal or reentrant atrial tachycardia.

- chronic right ventricular systolic pressure overload and increased end-diastolic pressure resulting from pulmonary valve insufficiency lead to increased right atrial pressure, hypertrophy and fibrosis.
- the atriotomy scar provides an anatomical obstacle, which in combination with haemodynamic disturbances, causes atrial arrhythmias, mainly typical atrial flutter or re-entrant tachycardia around the atriotomy scar.
- tricuspid insufficiency was the main risk factor for development of atrial fibrillation or flutter. Also found that risk factor for the development of atrial arrhythmias is the P wave dispersion in different leads.
- intraventricular conduction is typically prolonged in patients after TOF repair, and characterized by the presence of a right bundle branch block, they remain at risk of complete AV block should left bundle branch block develop as a result of degenerative changes.
- Although intra-atrial re-entrant tachycardia is the most common postoperative atrial arrhythmia, other possible causes should not be overlooked. These include accessory pathways and atrioventricular (AV) nodal re-entrant tachycardia, which are relatively easy targets for treatment with radiofrequency ablation.

Postoperative Arrhythmias in Tetralogy of Fallot, Hell J Cardiol 46: 2005

OUR PATIENT

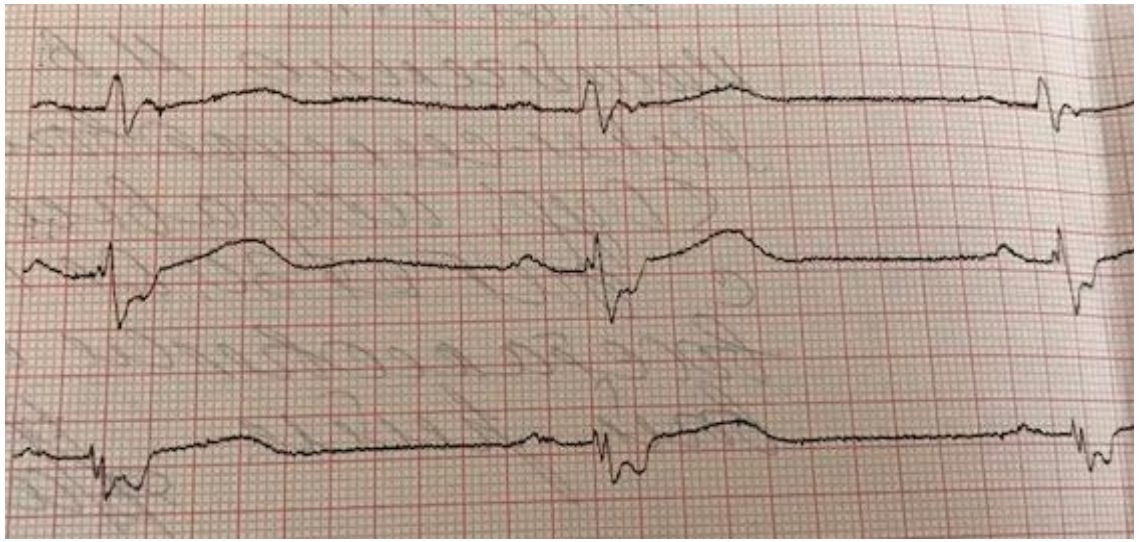
- Patient N.M.V.
- 39 y. old
- IT-specialist
- city resident

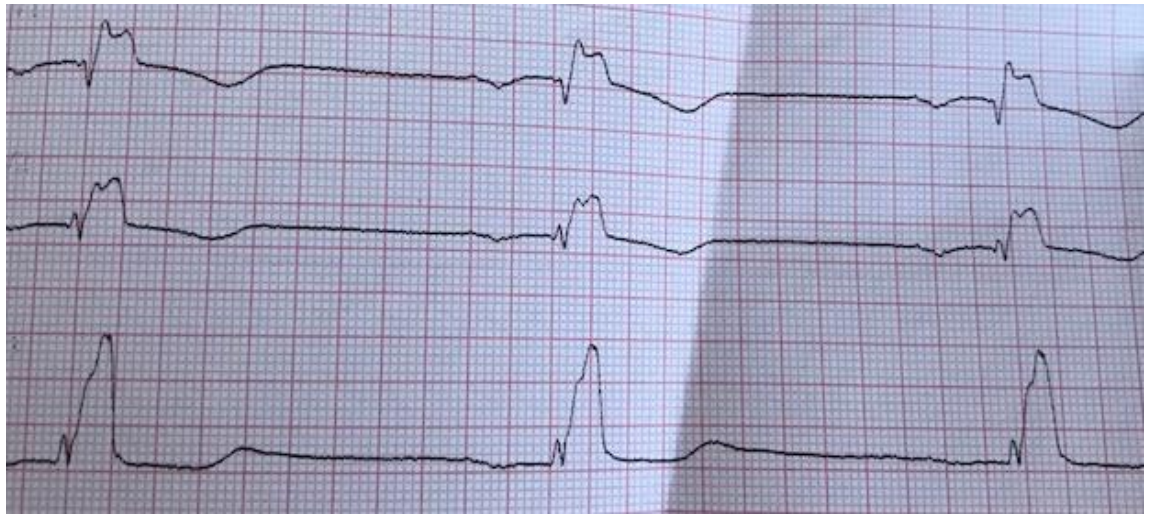
COMPLAINTS

- Periodical palpitation
- Weakness
- Discomfort in the chest

ANAMNESIS MORBI

- Congenital heart defect - tetralogy of Fallot – was surgically treated in 1984, 1990. Patient felt himself well till this year, when after skiing and flight to Georgia in January 2017 he felt bad, first time appeared episode of palpitation but after hospital treatment with amiodarone 31.01.17 sinus rhythm was repaired, received constant therapy with clopidogrel.
- Usual ECG of patient after Fallot tetralogy surgical repairment can be seen below. The same ECG picture patient had before hospitalization in the hospital 06-nov-2017. ECG conclusion: sinus rhythm, complete RBBB, left anterior hemi-block.





ANAMNESIS MORBI

- This time patient felt bad 06-nov-2017 at 4 a.m. in the morning when appeared feeling of high heart rate and after patient's words “he stated feel where the heart is”. At 10.00 a.m. state of the patient worsened: BP was 100/70 mm Hg and heart rate around 125 bts in min. Patient called an ambulance, on ECG was found paroxysm of atrial fibrillation and patient received amiodarone 400mg per oral treatment which had slight effect (heart rate decreased only till 100 bts in min).

- In 2.30 p.m. his state still wasn’t improved, patient visited district cardiologist and after ECG was taken (paroxysm of AF), received again 200mg of amiodarone per oral treatment without effect, by ambulance was delivered to ICU of the hospital.

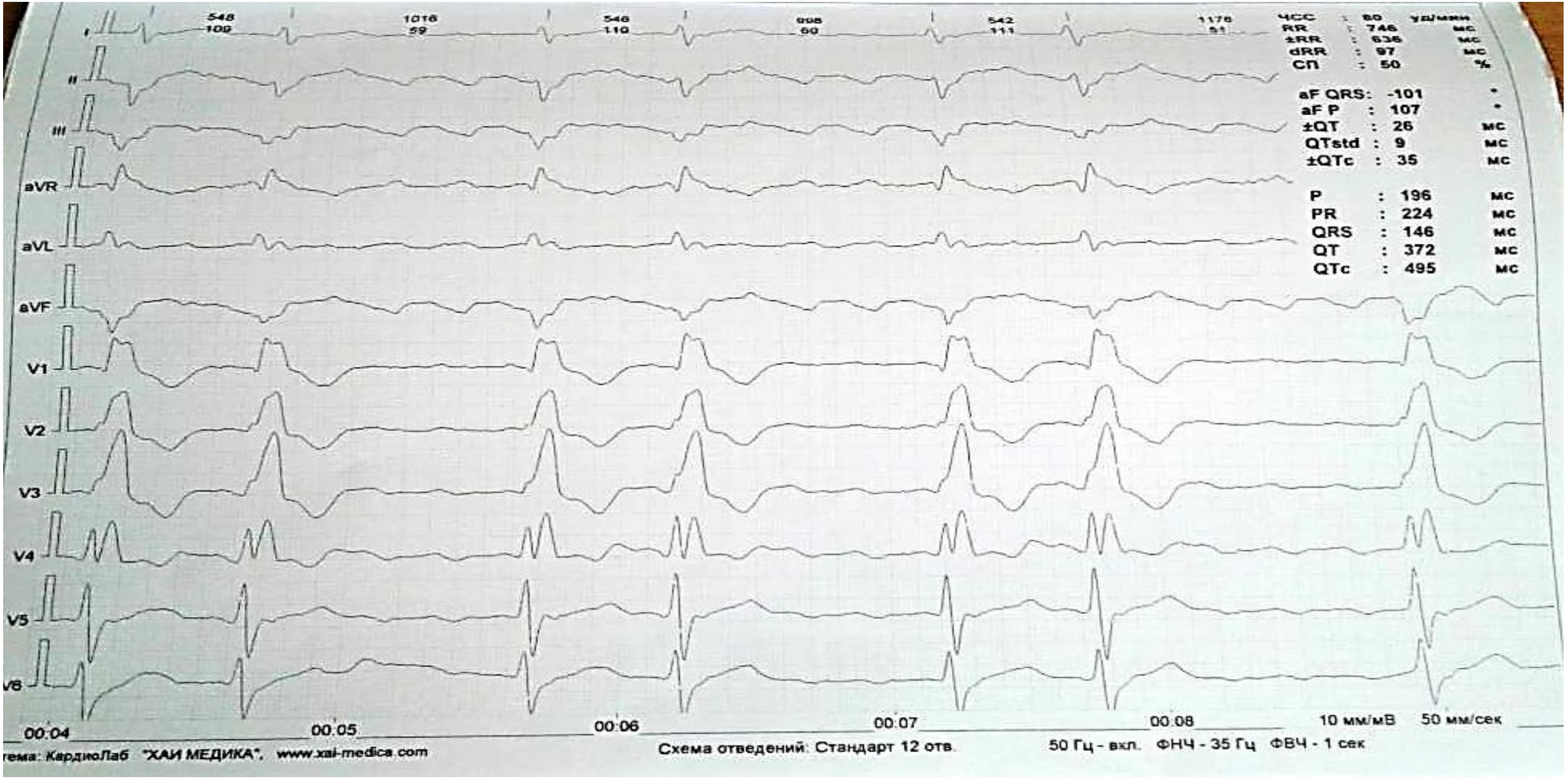
ANAMNESIS VITAE

- Hereditary diseases are not identified
- Allergic history is not burdened
- Childhood infections - chickenpox
- Sexually transmitted diseases were denied
- Smoker - no, do not abuse alcohol
- CV disease family history: nothing

OBJECTIVE STATUS

- Conciseness - clear, state – moderate severe, body position - active
- Patient can orientate himself in place, time, his personality
- Pale skin and mucosae
- Thyroid: mild hyperplasia of thyroid, soft, homogenous
- Musculoskeletal system – left pectoralis major muscle is bigger than right one due to postsurgical scar present. Scar in the center of sternum
- BR – 16-18 /min
- Lung percussion: no clinically significant changes
- Lung auscultation: hard breathing
- Borders of the heart: left border – outside of midclavicular left line on 2 cm
- Heart auscultation: arrhythmic, eusystolia, heart tones – muffled
- Pulse – rhythmic, 76 bts/min
- BP 105 / 80 mm Hg
- Abdomen: normal size, symmetric, unpainful
- Liver: normal size, no pain during palpation in right hypochondrium
- Spleen: normal
- Pasternatsky symptom – negative from both sides
- Edemas: absent

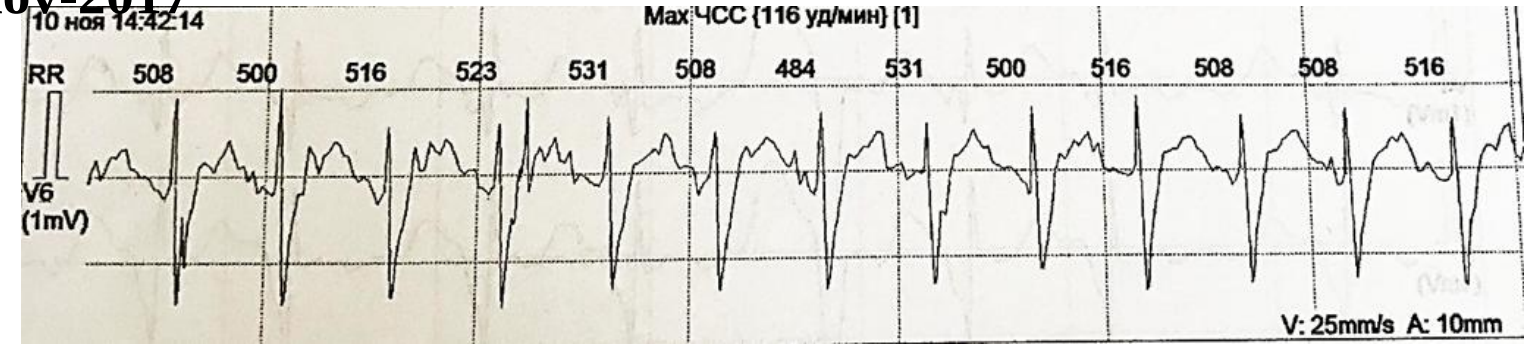
ECG during admission 06-nov-2017

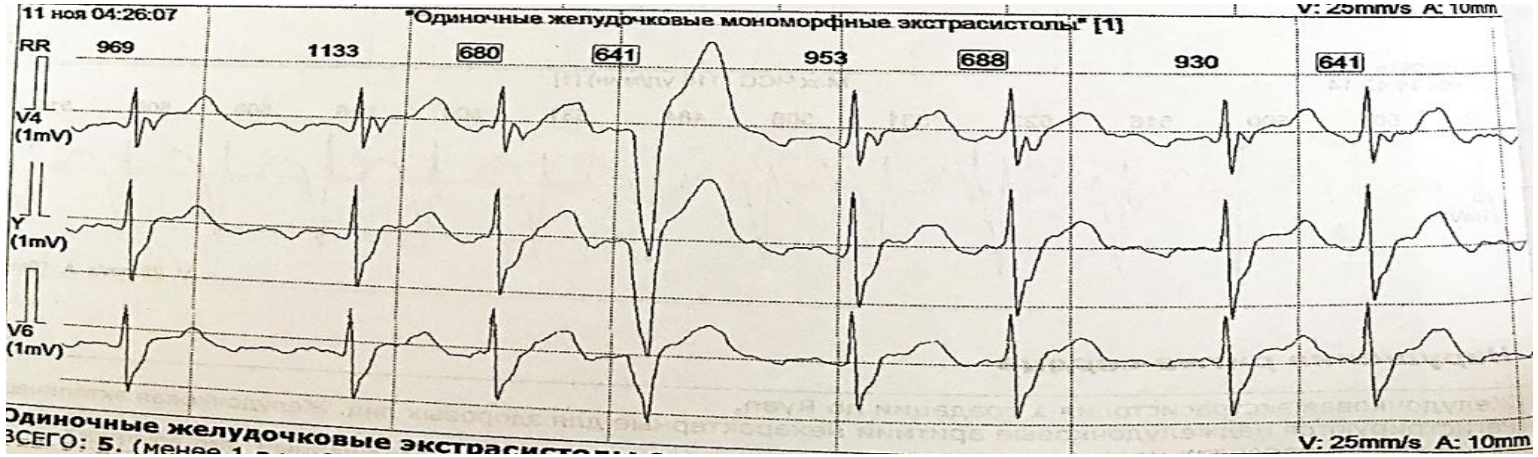


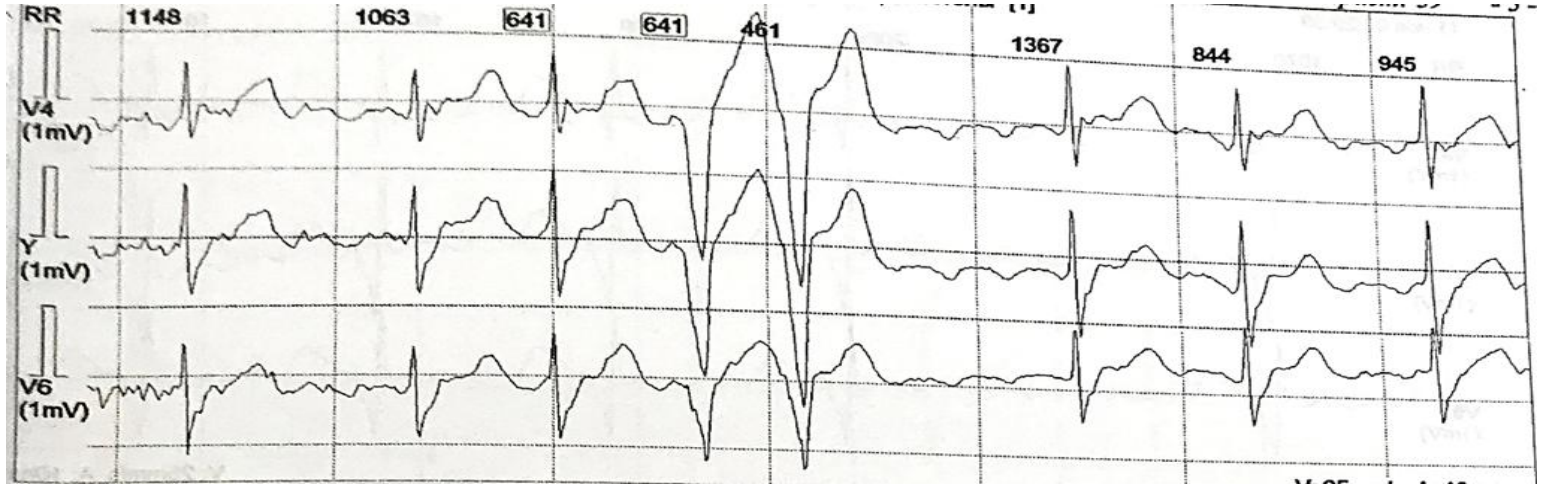
Conclusion: AF (HR-80 bts in min), eusystolia. RBBB. Left anterior LBB hemi - block

HOLTER ECG from 10-nov-2017

Conclusion:
Average HR in day time – 76, average HR in night time – 56 bts in min. Circadian index of HR – 135% (normal). Max HR -116 bts in min. Min HR – 50 bts in min.
Alternation of correct and non-correct atrial fibrillation with complete RBBB. Segregated ventricular premature contractions: 1 –in day time, 4-night time.
Paired ventricular premature contractions at night – 1.







BLOOD COUNT

	06/11/17	Normal Range
Hemoglobin, g/l	156	130 – 160
Red blood cells, 1012	4.81	4.0 – 5.0
Ht	47%	40-48%
White blood cells, 109	9,1	4 - 9
ESR, mm/h	3	1 -10
Bands	4%	1.06 – 6%
Segments	46%	47 – 72%
Eosinophils	1%	0.5 – 5%
Monocytes	6%	0.1 – 3%
Lymphocytes	43%	19 – 37 %
Platelets	264,6	180-320

Conclusion: lymphocytosis with minimal segments level decreasing

BIOCHEMISTRY TEST

DATA	Patient's ranges,	N
Glucose, mmol/l	4.9	4.22 – 5.5
Total bilirubin, mkmol/l	9,0	1,7 – 21
AST, U/l	28,8	< 37
ALT, U/l	48,0	< 41
ALT (from 11/01/17), U/l	51,0	< 41
Total protein, g/l	76.3	66 - 87
Creatinine, mkmol/l	102	82 – 106
Urea, mmol/l	6,6	3,0 – 9,2
Ca, mmol/l	2.37	2.02 – 2.6
K, mmol/l	4.59	3.6 – 5.5
Mg, mmol/l	0.75	0.66 – 1.07

Conclusion: increased ALT concentrations in the serum as clopidogrel day by day therapy side-effect

CARDIAC ENZYMES TEST DATA

06-nov-2017	Patient's ranges,	N
13:00		
CFK-NAC, u/l	88	< 171
CFK-MB, u/l	12,9	0 - 24

07-nov-2017	Patient's ranges,	N
7:00		
CFK-NAC, u/l	62	< 171
CFK-MB, u/l	10.4	0 - 24

Conclusion: all is normal ranges

LIPID PROFILE

	Patient's ranges,	N
Total cholesterol, mmol/l	5.03	< 5.2
VLDL, mmol/l	0.34	< 1.0
LDL, mmol/l	3,19	< 3.5
TAG, mmol/l	0,77	< 2.3
HDL, mmol/l	1,49	≥ 0.9
Index of atherogenicity	2.37	< 3.0

Conclusion: all is normal ranges

HEART ULTRASOUND from 08/11/17

Aorta: dilated, cuspids are thickened. Ascending aorta – d 46 (20-37mm). Aortic regurgitation I stage.
Tricuspid valve – regurgitation II stage. Pulmonary trunk valve – regurgitation I stage, d = 29mm. Pressure in pulmonary trunk is 21,0 mm Hg (< 15). Mitral valve – cuspids are thin, M-shaped movements in different direction, anterior cuspid in left atrium cavity during systole, regurgitation I stage.
EF – 75% (N - 55 – 78%). FS – 44% (N - 28 – 44%).
Left Ventricle:
FDD – 50 mm (N – 35 – 55mm)
FSD – 28 mm (N – 23 – 38 mm)
Posterior wall thickness in systole– 11,2 mm (N – 6 – 13mm).
Intraventricular septum size in diastole– 12,2 mm (6 – 11 mm)
Right Ventricle:
Diameter – 33,1 mm (N – 9 – 26 mm)
Wall thickness – 6,8 mm (N – 3 - 6 mm), thickened.
Left atrium – not enlarged – 34,1 mm in diameter (N – till 38 mm)
Right atrium – dilated – 45,6 mm in diameter (N – 25-37). Interatrial septum – not changed, no defects.

Conclusion: Tetralogy of Fallot surgical repair state. Dilation of ascending part of aorta and aortic root, aortic regurgitation I stage. Dilation and hypertrophy of right ventricle and dilation of right atrium. Pulmonary hypertension I degree. Tricuspid regurgitation II stage. Mitral valve prolapse I degree, regurgitation I stage. No cardiac thrombus.

THYROID GLAND EXAMINATION

	Patient's ranges,	N
Ab to thyroglobulin, mE/ml	14.0	< 100
TSH, mkME/ml	2,1	0.23 – 3.4

Conclusion: all is normal ranges

THYROID ULTRASOUND from 08/11/17

Left lobe – 43*15,5*15,6mm, volume – 7.3 cm3. Right lobe – 50,8*16,9*17,5 mm. Average volume of both lobes – 16.0cm3. Isthmus – 5,0 mm.

Conclusion: Thyroid gland enlarged, situated in typical place. Homogenous parenchyma, granulomatous, with hyper and hypoechogenus areas. No volumetric growth found.

ABDOMINAL ULTRASOUND **from 08/11/17**

Liver: Right lobe – 132,5 (N-till 150mm), left lobe thickness -73,8 (N- till 65mm)
Increased echogenicity, structure is homogenous. Ductular system is not changed.
Gall bladder: 76,2*19,5 mm. Wall thickness is increased, normal shape, homogenous contain with small calculi 1-1,5 mm long, pair of them – 6mm long.
Pancreas: head – 22,2 mm (N-24-30mm), tail – 29 mm (N – 17-28), body – 18,2 mm (N-12-17). Increased parenchyma echogenicity, homogenous. Ductular system is not changed.
Spleen: 39.7*102.5cm. Normal echogenicity, structure is homogenous.
Right kidney: 100.5*40.2mm, V=82cm/c, normal echogenicity, structure is homogenous, not changed.
Left kidney: 103.7*40.9mm, V=84cm/c, normal echogenicity, structure is homogenous, not changed.
Conclusion: Diffuse changes of pancreas parenchyma. Slight hepatomegaly. Calculous chronic cholecystitis.

COMPLETE DIAGNOSIS of our patient is:

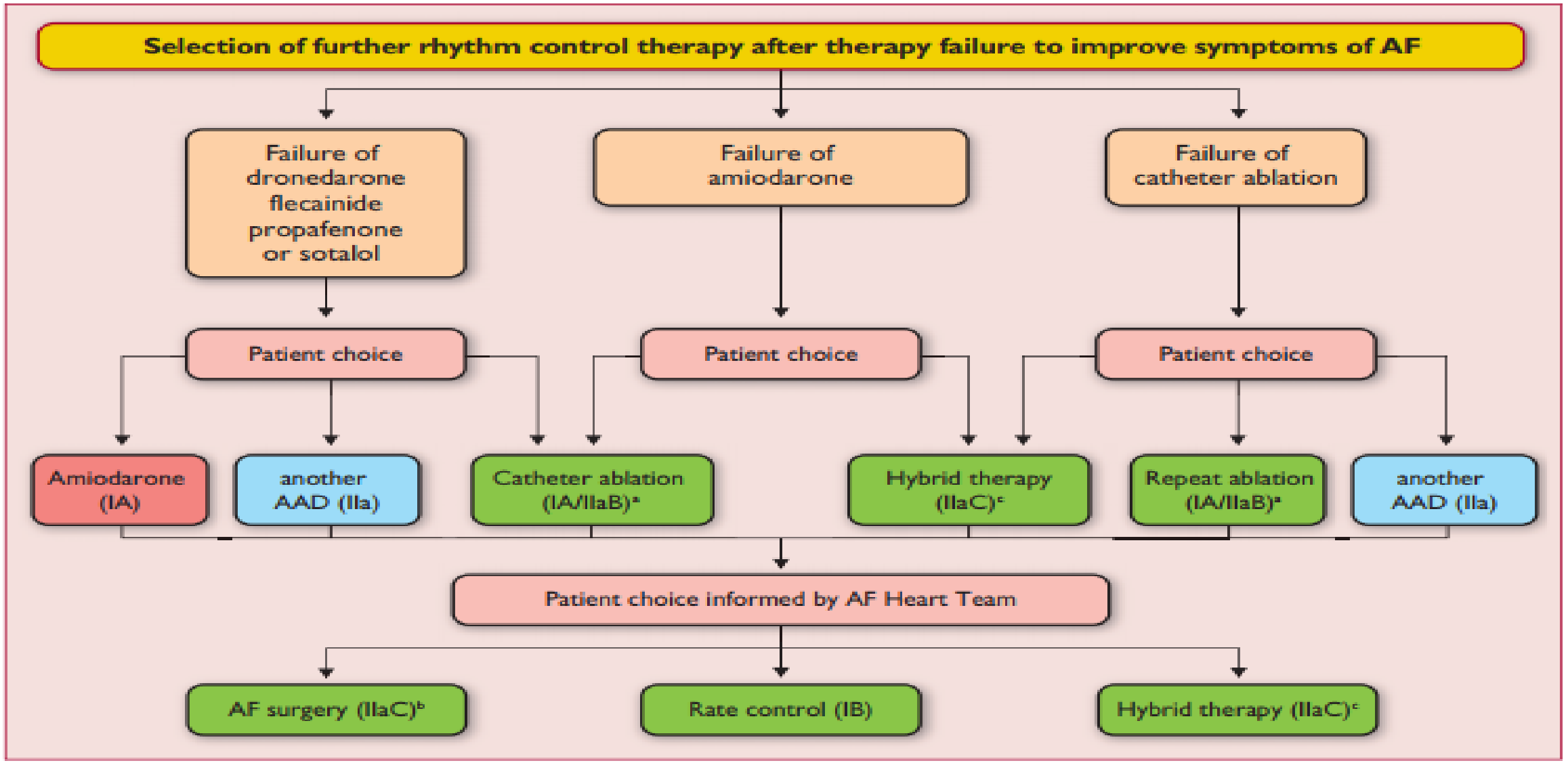
Main: Acquired heart defect: tetralogy of Fallot, state after radical correction (1984, 1990). Complete RBBB, left anterior LBB hemi - block. Atrial fibrillation, persistent form, paroxysm 06-nov-2017. Dilation of ascending aorta, right heart chambers. Tricuspid valve regurgitation II degree, pulmonary trunk valve regurgitation I degree. Chronic Heart Failure 0 stage

Concomitant diseases: Chronic calculous cholecystitis. Thyroid hyperplasia I degree, euthyroid state.

MEDICATIONS PRESCRIBED IN HOSPITAL

- clopidogrel 75 mg 1 time\day
- enoxaparinum natrium 0,8ml (80mg) 2 times a day subcutaneous
- rivaroxaban 20 mg 1 time\day
- carvedilol 6,25 mg 2 times/day
- amiodaron 200 mg 3 times a day with HR control
- asparginat K-Mg 20,0 ml on 100ml of Ringer’s solution 0,9% IV 1 time\day
- pantoprazol 40 mg 2 times\day
- tiotriazolin (metabolic) 4,0 ml IV 1 time\day

Choice of rhythm control therapy following treatment failure



AAD = antiarrhythmic drug; AF = atrial fibrillation; PVI = pulmonary vein isolation.
[a]catheter ablation should target PVI. IA for paroxysmal AF, IIaB for persistent and long-standing persistent AF.
[b]AF surgery may be PVI (e.g. in paroxysmal AF) or maze surgery (e.g. in therapy-refractory or persistent and long-standing persistent AF).
[c]Hybrid therapy involves combination of antiarrhythmic drugs, catheter ablation, and/or AF surgery.

http://www.heartrhythmalliance.org/files/files/afa/for-clinicians/2016%20ESC%20Guidelines.pdf

RECOMMENDATIONS:

Despite treatment prescribed (improvement of symptoms, decreased HR <110), patient still has atrial fibrillation and rhythm wasn't restored.

- Patient has enlargement and changes in thyroid gland structure. Would be reasonable to recommend thyroid hormones evaluation before surgical treatment (TSH, T3, T4 levels) to disapprove thyroid function alteration as a cause of AF appearance.
- Clinical risk scores for bleeding: CHA2DS2-VASc=1, HASBLED=1, EHRA=II. Patent was recommended to continue therapy with rivaroxaban 20 mg /daily for stroke prevention.
- Long-term pharmacological rate control with beta-adrenoreceptor blocker carvedilol 6,25 *2 times/daily in combination with amiodarone 200 mg 2 times/daily (a target heart rate ,80 b.p.m. at rest and ,110 b.p.m. during moderate exercise)
- Consultation of arhythmologist and synchronized direct current electrical cardioversion (and is the method of choice in severely haemodynamically compromised patients with new-onset AF). Pre-treatment with amiodarone (requiring a few weeks of therapy) can improve the efficacy of electrical cardioversion.
- For cardioversion of AF/atrial flutter, effective anticoagulation is recommended for a minimum of 3 weeks before cardioversion.
- Catheter ablation of symptomatic paroxysmal AF is recommended to improve AF symptoms in patients who have symptomatic recurrences of AF on antiarrhythmic drug therapy (amiodarone, dronedarone, flecainide, propafenone, sotalol) and who prefer further rhythm control therapy, when performed by an electrophysiologist who has received appropriate training and is performing the procedure in an experienced centre.

CONCLUSION

Cardiac surgery has transformed the outcome for patients with tetralogy of Fallo. However, there are problems with late morbidity and mortality primarily due to right ventricular dysfunction, exercise intolerance, arrythmia, and sudden cardiac death. The most common form seen in patients with tetralogy of Fallot is atrial fibrillation although atrial flutter and other forms of atrial arrhythmias also occur.

Despite good progress in the management of patients with atrial fibrillation, this arrhythmia remains one of the major causes of stroke, heart failure, sudden death, and cardiovascular morbidity, which requires not only heart rate control but also prevention of tromboembolic complications. In other words, the main violin played leading role in tetralogy Fallo patient health’s piece will be atrial fibrillation which should be managed accourding to the latest guidelines for its management.

Thank you