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**Clinical case: the features of the disease in patient with 20 years
history of idiopathic pulmonary arterial hypertension**

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Survival in patients with IPAH



- IPAH (REVEAL)
- ◆ IPAH > 65 years (COMPERA)
- ◆ IPAH with comorbidity (Swedish registry)

	1 year	3 years	5 years
IPAH (REVEAL)	91%	74%	65%
IPAH > 65 years (COMPERA)	90%	68%	
IPAH with comorbidity (Swedish registry)	85%	71%	59%

Disease history of the patient

Dyspnea on moderate exertion

Echo:

RA area = $7,1 * 5,5 \text{ cm}^2$;
RV ant-post d.= 5,2 cm,
PA truncus =3,5 cm,
RPA and LPA=2,0 cm, systPAP =60 mm Hg.;
IVC =2,5 cm, collapse > 50%.
Small pericardial effusion

Liver ultrasound–venous congestion of the liver;

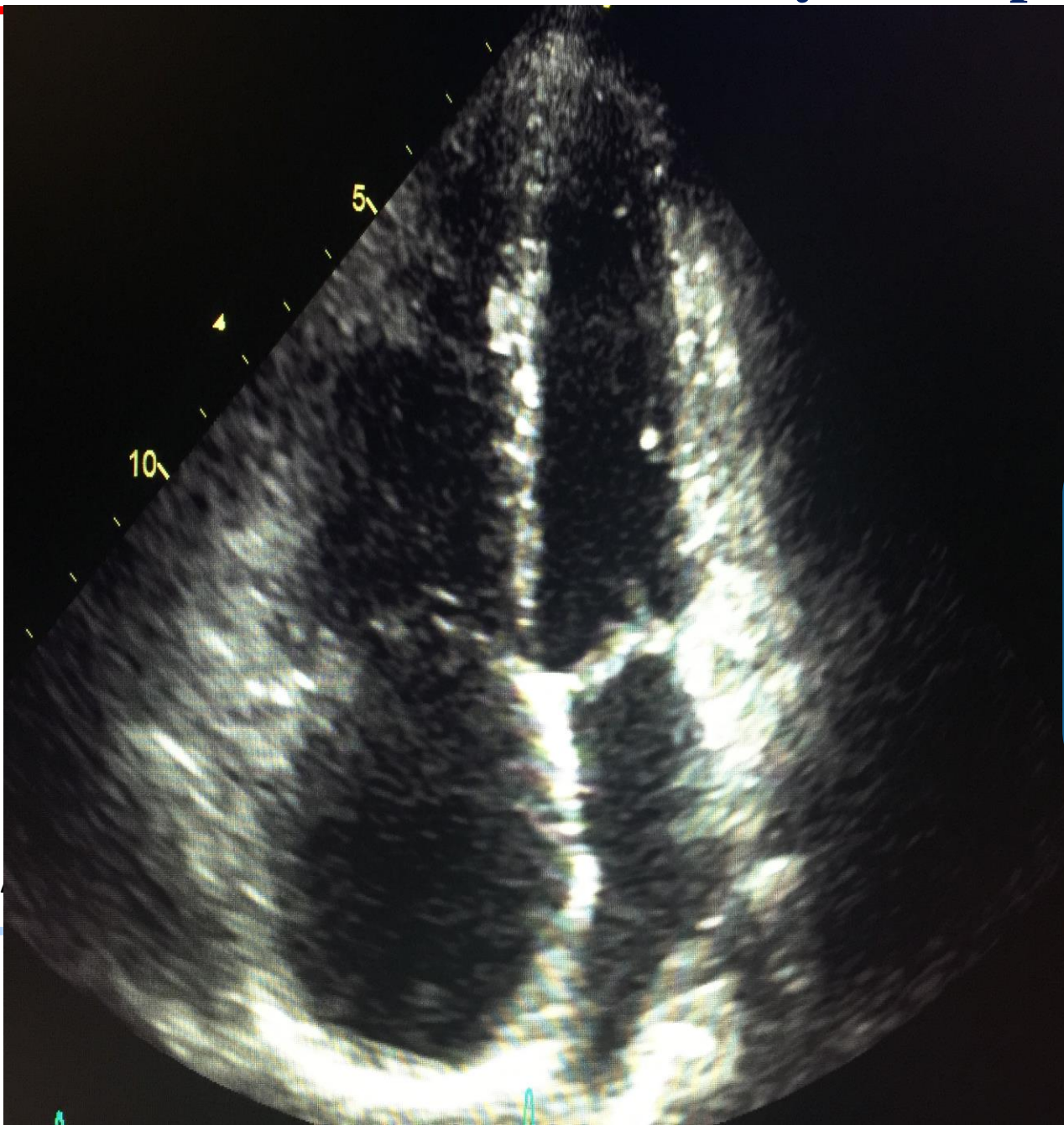
CT with pulmonary angiography:

↑ PA truncus from 4,2 to 4,5 cm,
RPA =3,2 cm, in RPA organized thrombotic masses with calcification sites spreading to segmental and subsegmental branches;

Warfarine (INR 2,5-3,5);
Spironolactone 25 mg/day
Torasemide 2,5 mg/day.

2012

55 years



Disease history of the patient

Dry cough in
dusty rooms or
during wet
weather
➔ COPD

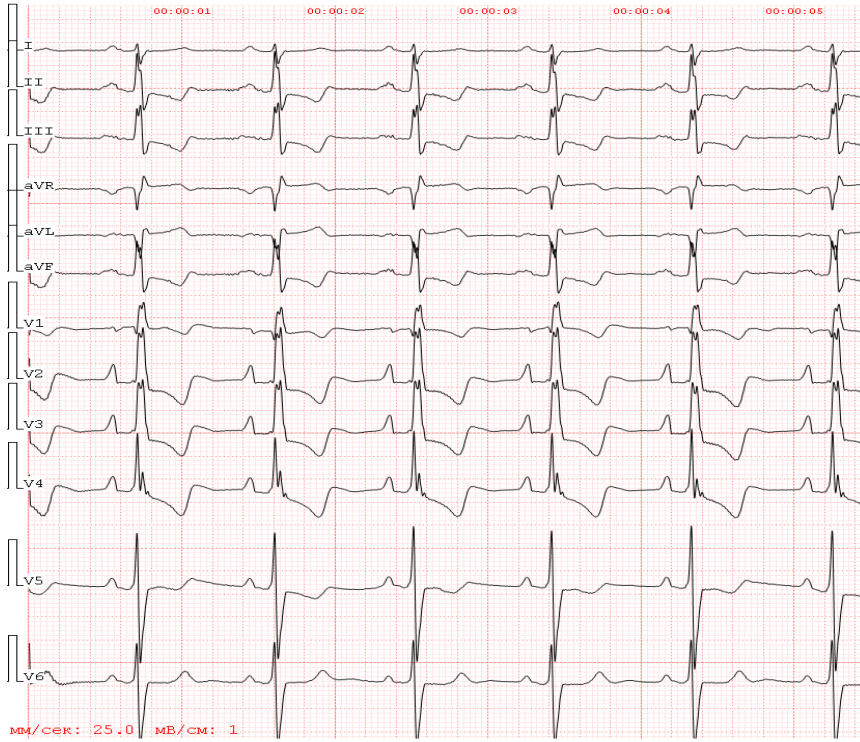


- Formoterol 12
mkg +Budesonide
400 mkg 2 doses
inhaled

Age: 57 years

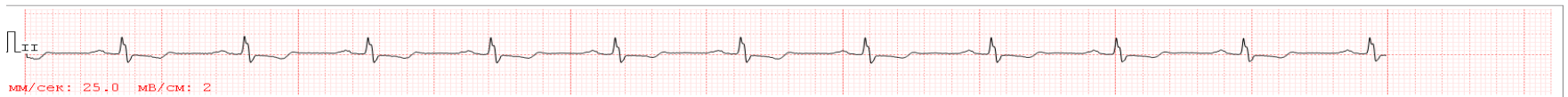
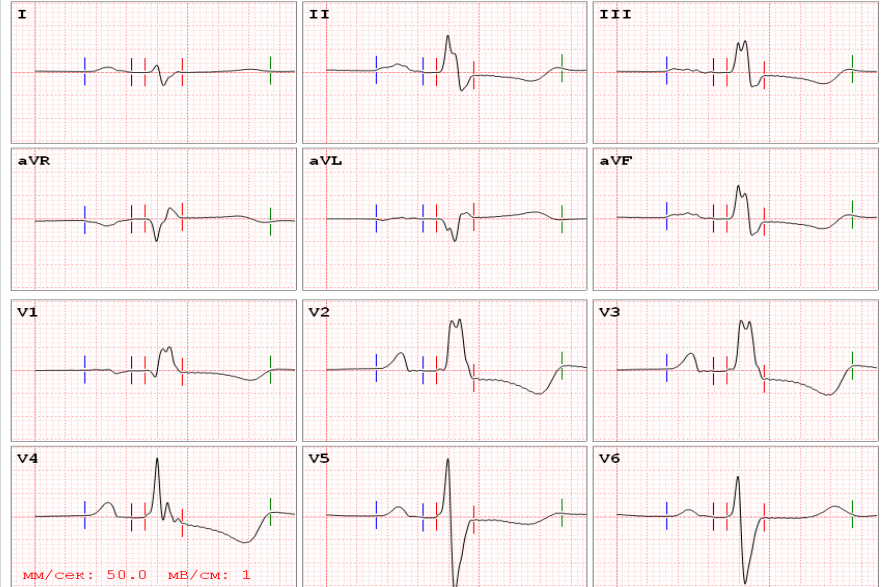
2014

ECG



QRS	122 мс	QT	412 мс	P ось	56 °
P	152 мс	QTc	422 мс	QRS ось	104 °
PQ	194 мс			T ось	-66 °

Ритм синусовый, ЧСС 65 уд. в минуту. Изменение предсердного компонента по типу P-pulmonale. Отклонение электрической оси сердца вправо. Признаки изменения миокарда вследствие выраженной гипертрофии правого желудочка.



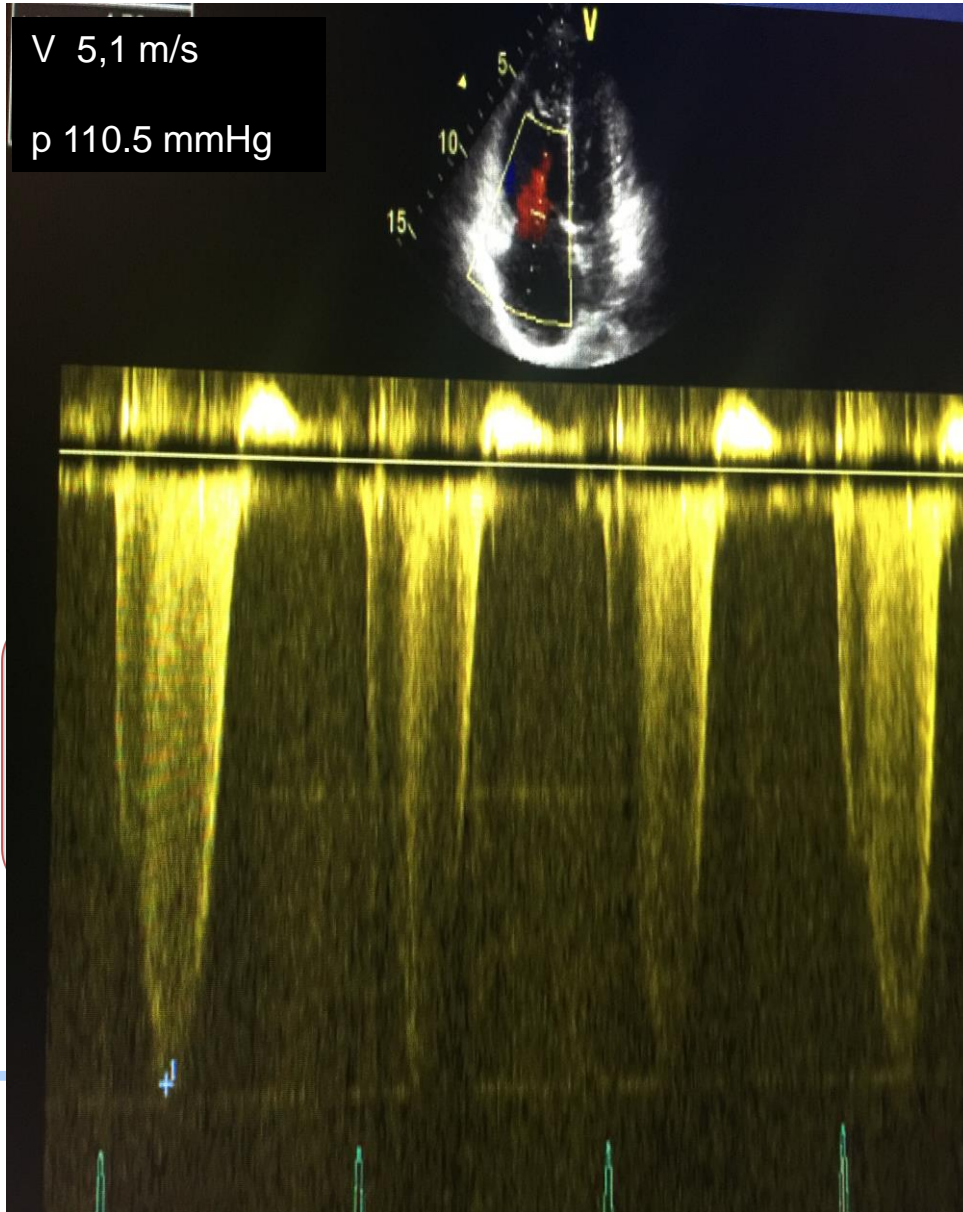
СС: 65 ВЧ: 70.0Гц НЧ: 3.2сек Режктор: Да Тремор: Нет Фильтр ИКС: Нет Баз. линия: Да

Стр: 2/3

Age: 58 years

2015

Disease history of the patient



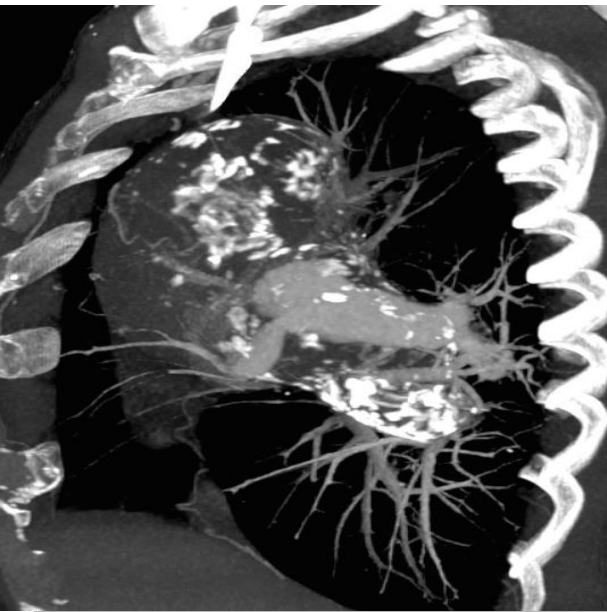
Dyspnea progression

6MWD = 415 m;
dyspnea by Borg scale =5;
Echo: systPAP = from 60 to 130 mmHg; PA truncus from 3,5 to 4,6 cm, RPA = 7,5 cm;
Moderate pericardial effusion.
Lung X-Ray: Coef. Mur to 69% (in 2014 $r=49\%$), Coef. Lupi to 48% (in 2014 $r=42\%$)

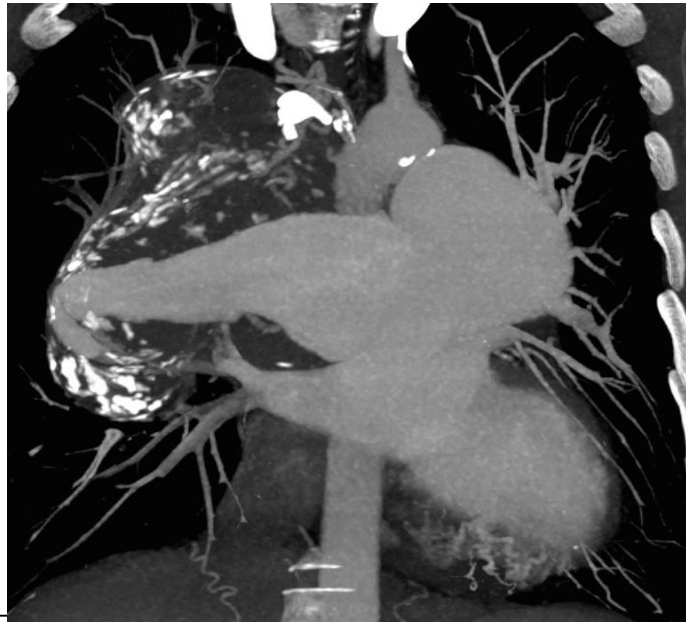
2016

Age: 60 years

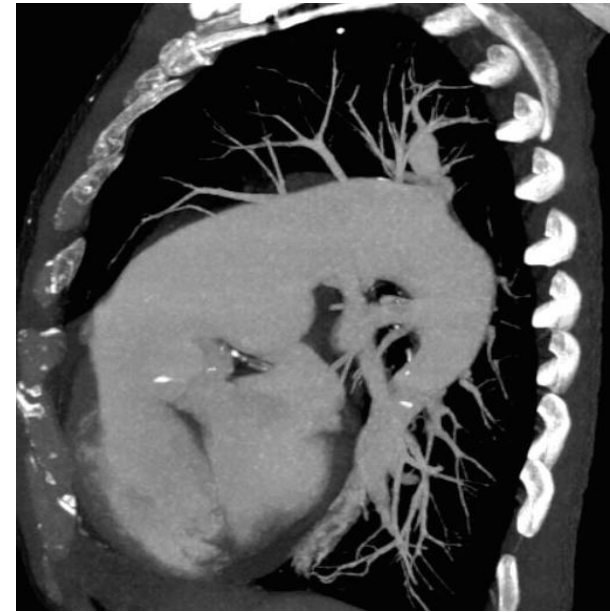
CT with pulmonary angiography 2016 year: PA truncus to 5,5 cm, RPA from 7,2 to 8,8 cm, LPA 4,2 cm



Right PA sagittal
plane



Frontal plane
of RPA and LPA



Left PA sagittal
plane

Disease history of the patient

Dry cough in
dusty rooms or
during wet
weather
→ COPD



- Formoterol 12 mkg
+Budesonide 400 mkg 2 doses inhaled

Age: 57 years

2014

6MWD = 466 m; dyspnea by Borg scale =3;
Echo:
PA truncus from 3,5 to 4,0 cm, RPA =from 2 to 5,8 cm.
Lung CT angiopulmonography:
RPA =7,2 cm, LPA =4,0 cm + rope-formed thrombosis in LPA

- Tadalafil → sildenafil 60 mg/day;
- Torasemide up to 10-15 mg/day,
- Spironolactone → eplerenone 25 mg

Age: 58 years

2015

Dyspnea progression

6MWD = 415 m;
dyspnea by Borg scale =5;
Echo: systPAP = from 60 to 130 mmHg; PA truncus from 3,5 to 4,6 cm, RPA = 7,5 cm;
Moderate pericardial effusion.
Lung X-Ray: Coef. Mur to 69% (in 2014 Γ =49%), Coef. Lupi to 48% (in 2014 Γ =42%)
CT with pulmonary angiography: PA truncus to 5,5 cm, RPA from 7,2 to 8,8 cm, LPA 4,2 cm

Sildenafil →
riociguat 7,5 mg/day
+
Iloprost 40 mkg inhaled

Age: 60 years

2016

Disease history of the patient

05.03.2017

Age:61years

Excessive static load →

- shortness of breath at rest,
- pressing pains behind the sternum in a horizontal position

07.03.2017

Hospitalization in PH
Expert center

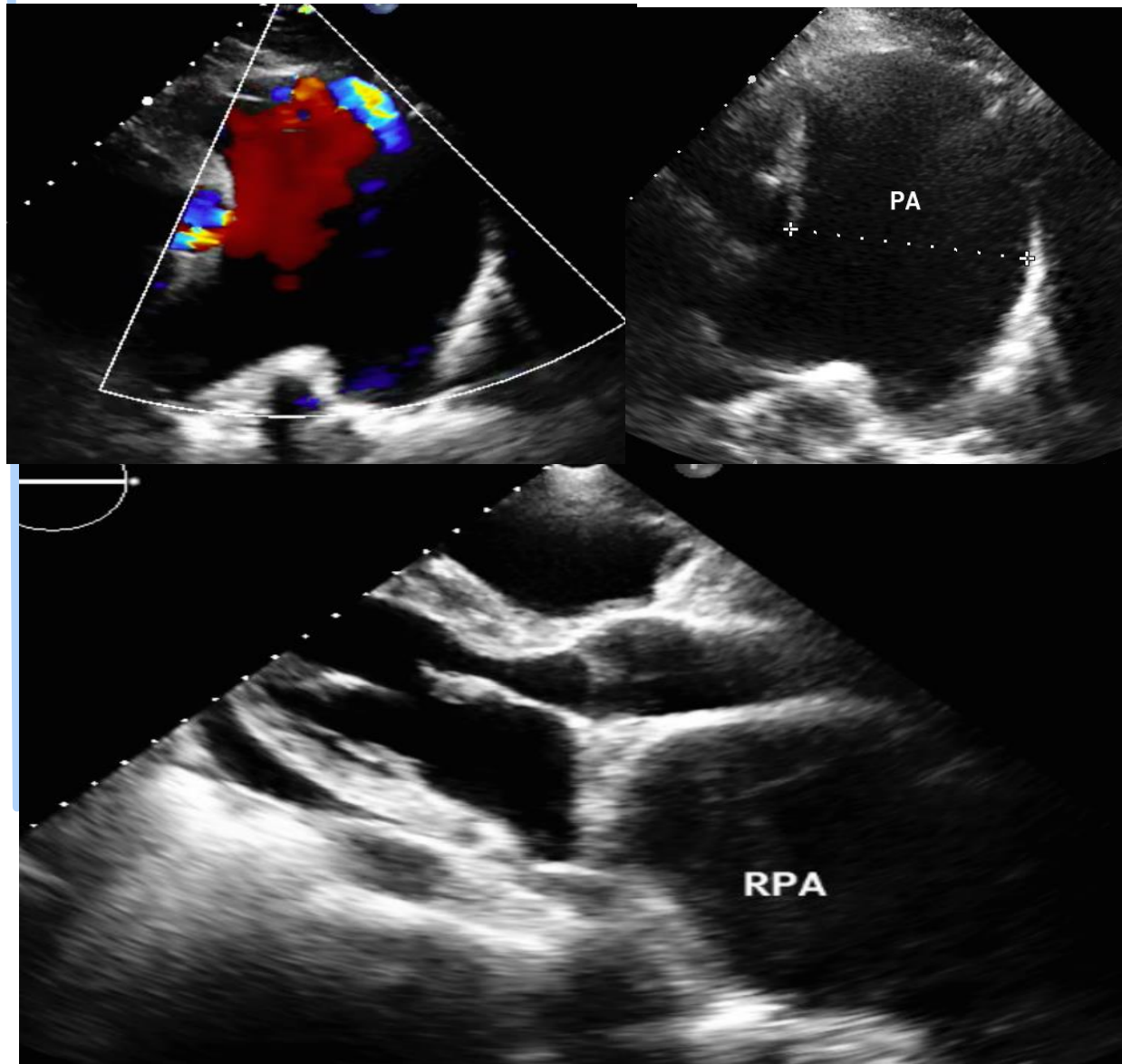
ECHO 07.03. 2017 г.

S RA = 36 cm²,

systPAP = 145 mm Hg,

aneurysmal expansion of the PA
truncus (5.6 cm) / RPA (10.2 cm);

In pericardium up to 300 ml of fluid



CT with pulmonary angiography

07.03.2019:

RPA 10 cm, dissection of RPA intima



Sagittal plane of RPA



Axial plane (moderate amount of fluid in pericardium)

RPA dissection
with heart hemotamponade
+



Pericardiocentesis
(up to 1 liter of blood);

AF paroxysm with
ventricular rate up to 160 beat/min
(! Thrombocytopenia $70 \cdot 10^9/l$)
+



-Diltiazem 240 mg / day,
-Apixaban 10 mg/day

Chronic bronchitis exacerbation,
the bronchospasm,
the subfebrile fever



Antibiotic therapy
bronchodilators,
parenteral dexametazone

-Riociguat 4.5 mg → 7.5 mg / day,
-Iloprost 40-60 mg / day,
-Macitentan 10 mg,
-Ibuprofen 200-400 mg / day (for a month),
-Torasemide 15 -20 mg,
-Eplerenone 50 mg,
-Formoterole + budesonide up to 2 inhalation / day,
-Oxygen therapy 2-4 l /min



Waiting list on lung transplantation

Risk assessment in March 2017

Determinants of prognosis (estimated 1- year mortality)	Low risk < 5%	Intermediate risk 5-10%	High risk > 10 %
Clinical signs of right heart failure	Absent	Absent	Present

Diagnosis: IPAH, WHO FC IV

WHO FC	I, II	III	IV
6MWD	> 440 m	165-440 m	< 165 m

PAH specific treatment:

- Riociguat 7.5 mg / day,
- Iloprost 40-60 mg / day,
- Macitentan 10 mg/day

Imaging (echocardiography, CMR)	RA area < 18 cm ² No pericardial effusion	RA area 18-26 cm ² No or minimal pericardial effusion	RA area > 26 cm ² Pericardial effusion
Hemodynamics	RAP < 8 mmHg CI > 2,5 l/min/m ² Svo ₂ > 65%	RAP 8-14 mmHg CI > 2,0 -2,4 l/min/m ² Svo ₂ 60 - 65%	RAP > 14 mm Hg CI > 2,0 l/min/m ² Svo ₂ < 60%

Last hospitalization - November 2018

- **Dyspnea on minimal exertion**
- **Shortness of breath**
- **Dry cough**
- **PH FC IV (WHO)**

ECG – AF with VR 75 beats/min.

ECHO –

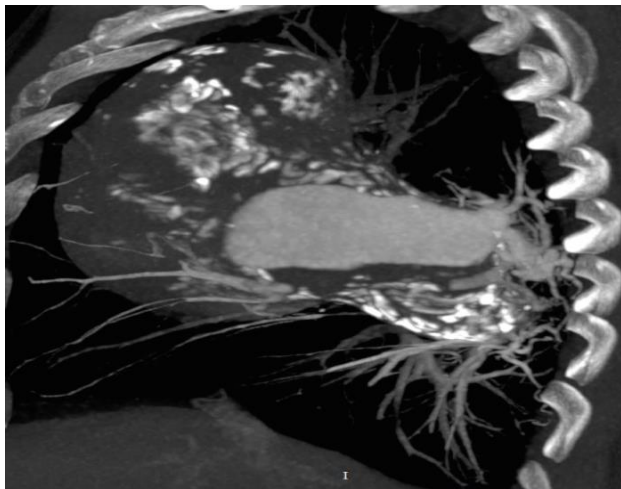
RA area=44 cm²; RPA=11,2 cm;

RV dimension=5,2cm.

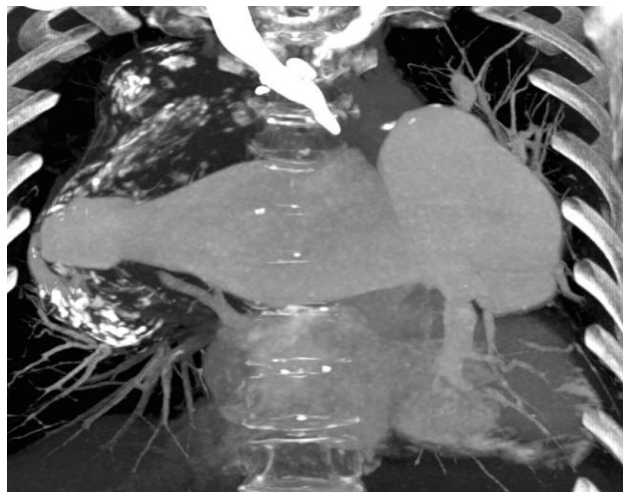
IVC : 2,1 / collapse 0,7 cm;

No pericardial effusion.

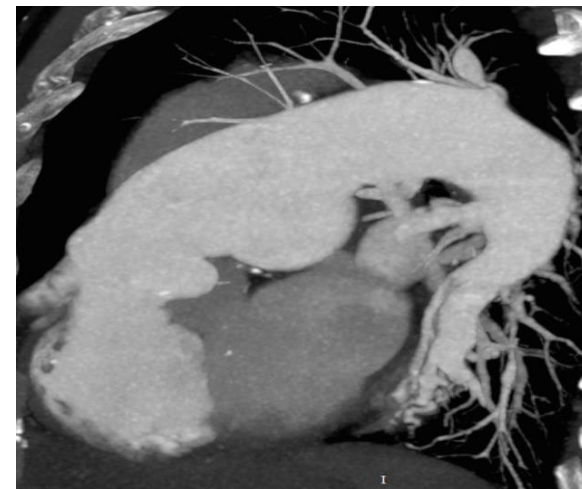
Lung CT with pulmonary angiography – no dynamics.



Right PA sagittal plane



Frontal plane of RPA and LPA



Left PA sagittal plane

Pulmonologist's recommendations:

Spiriva respimat 2,5 mkg/dose (2 doses)/day

Symbicort Turbuhaler 160/4,5 mkg/dose (2 doses TD)

Summary and conclusion

- Patient with verified IPAH diagnosis with + AVT
- 20 years follow-up (Department of Pulmonary Hypertension)
- Thus, this unique clinical case reflects the importance of dynamic control and timely escalation / optimization of PAH-specific therapy, as well as adequate control of the hypercoagulation state, heart failure signs and compensation of concomitant diseases in IPAH patient with complex phenotype.



Thank you for attention!