



A.Pijuan-Domènech
Unitat de Cardiopaties Congènites de 'Adolescent i l'Adult
UCCAA, ACOR Barcelona
Hospital Vall d'Hebron

30 years old

D.o.B. 22/01/1980

Diagnosis of congenital heart disease at neonatal period

Cyanosis

Suspected critical pulmonary stenosis



Neonatal Cardiac Catheterization:

- Pulmonary stenosis
- Suprasystemic right ventricular pressures
- Moderate tricuspid regurgitation
- •Right to left shunt at atrial level, through patent foramen ovale (PFO)
- Post-stenotic dilatation of pulmonary trunk
- •PDA not patent



Surgery 25/01/1980 (3 days old)

- Sternotomy
- Bicaval clamping
- Pulmonary arteriotomy
- Pulmonary valvotomy

Surgical findings: puntiform orifice

Postoperative course: uneventful



- •Follow up at the Cardiology Pedriatic unit and transferred to ACHD
- ·Normal development: active life, asymptomatic
- •On follow up moderate to severe pulmonary regurgitation (PR) and mild right ventricular (RV) dilatation was present
- 2004: (24 y old):pregnancy completed

No cardiac or obstetric complications except

NYHA Class II at the end of pregnancy

Spontaneous vaginal delivery

Baby girl: 2900 g, APGAR 9/10



Question number 1

Which is the commonest residual lesion after surgical repair of pulmonary stenosis or Tetralogy of Fallot?

- 1. Pulmonary stenosis
- 2. Pulmonary regurgitation
- 3. Residual lesions are extremely infrequent and patients do not need follow up
- 4. In pulmonary stenosis, surgically relieved residual lesions are not common



Question number 1

Which is the commonest residual lesion after surgical repair of pulmonary stenosis or Tetralogy of Fallot?

- 1. Pulmonary stenosis
- 2. Pulmonary regurgitation
- 3. Residual lesions are extremely infrecuent and patients do not need follow up
- 4. In pulmonary stenosis, surgically relieved residual lesions are not common



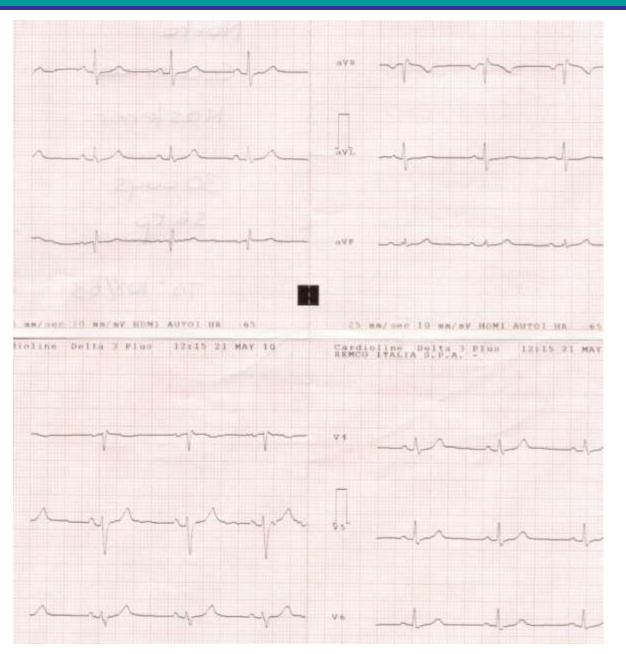
2009 : 29 years old

Asymptomatic, NYHA I

- BP 110/60 mmHG, O2saturation air room 95%
 2/6 systolic murmur, single 2ond heart sound,
 2/4 diastolic murmur
- ECG: SR 65 bpm.Right axis deviation +90.rSR

And patient wants to be pregnant.....

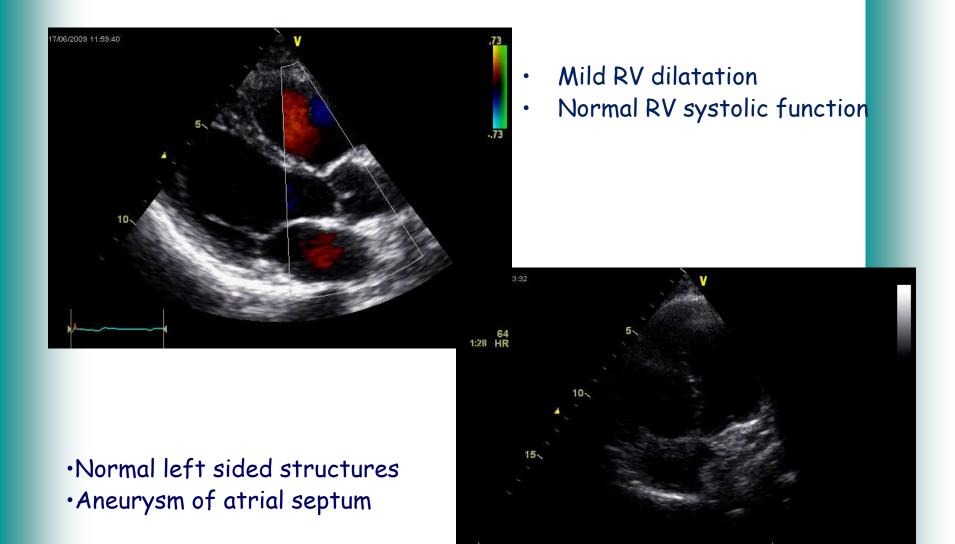






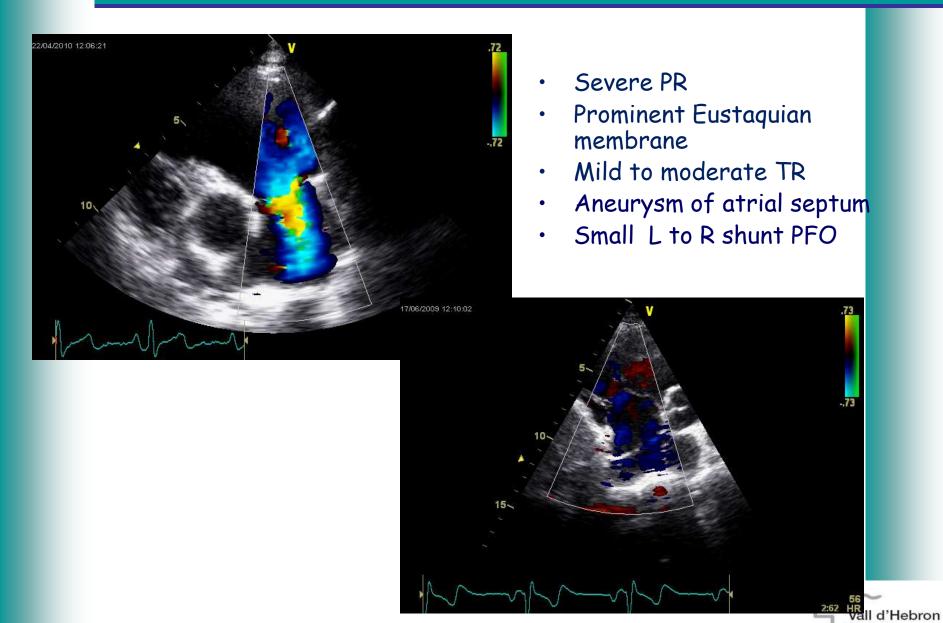






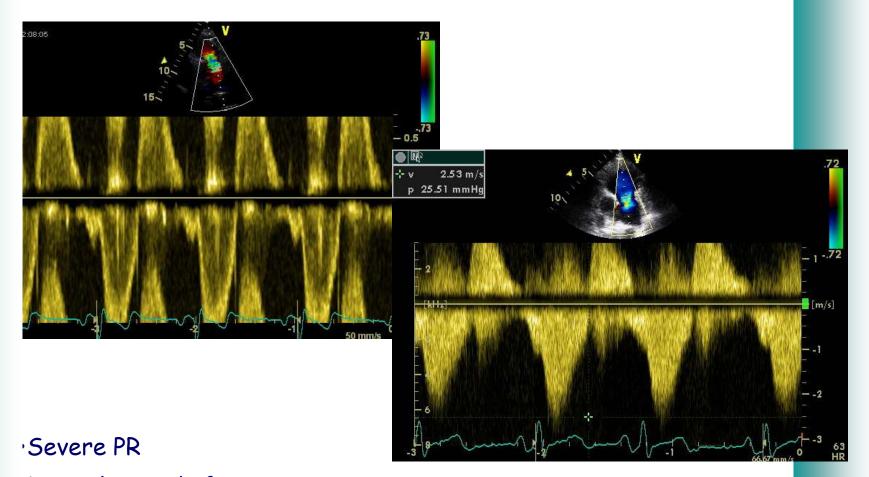


Vall d'Hebron





Hospital



- Diastolic RV disfunction
- Restrictive RV physiology
- RV to RA pressure 25 mmHG



Question number 2

What kind of risk do you think has got the patient, the fetus and neonate if pregnancy?

- 1. PR is a risk factor for maternal complications, as well as prior cardiac events
- 2. Prior cardiac events, are also predictors of fetal and neonatal complications, mainly intrauterine growth retardation and prematurity
- 3. Global risk of transmission or recurrence of CHD is around 4%
- 4. There is a 9-10% risk of maternal complications, mainly heart failure or arrythmia, usually respond to treatment
- 5. All previous sentences are correct



Question number 2

What kind of risk do you think has got the patient, the fetus and neonate if pregnancy?

- 1. PR is a risk factor for maternal complications, as well as prior cardiac events
- 2. Prior cardiac events, are also predictors of fetal and neonatal complications, mainly intrauterine growth retardation and prematurity
- 3. Global risk of transmission or recurrence of CHD is around 4%
- 4. There is a 9-10% risk of maternal complications, mainly heart failure or arrythmia, usually respond to treatment
- 5. All previous sentences are correct



Question number 3

So what do we answer to the patient, can I become pregnant?

- 1. No, you can not be pregnant
- 2. Yes, go ahead, no problem at all
- 3. There is some risk, we will discuss it next year, today the outpatient clinic is very busy
- 4. There is some risk, we will perform some more tests and discuss with you issues regarding risks for you and your baby at the preconceptional clinic



Question number 3

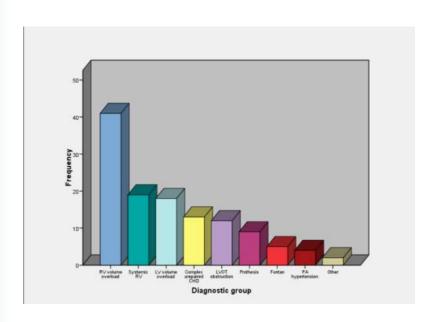
So what do answer to the patient, can I become pregnant?

- 1. No, you can not be pregnant
- 2. Yes, go ahead, no problem at all
- 3. There is some risk, we will discuss it next year, today the outpatient clinic is very busy
- 4. There is some risk, we will perform some test to evaluate you and discuss with you issues regarding risks for you and your baby at the preconceptional clinic



Our plan...

Book cardiac MRI, exercise test MVO2 Discuss risk in Preconceptional assessment clinic







Hospital

Vall d'Hebron

Our plan...

Book cardiac MRI, exercise test Preconceptional assessment clinic

Patient's plan...

She felt ok to be pregnant Pregnancy went on and controlled locally



36 week pregnant

- Followed up locally, normal fetal growth, no obstetric complications
- Progressive shortness of breath from 30th week, and transferred to emergency room

Physical findings:

BP 100/60 mmHG,
Mild cyanosis, Air room O2 saturation 87%
Ankle edema
No respiratory difficulty at rest, chest clear
Normal first single 2 heart sound a 95 bpm,
2/6 systolic murmur, short diastolic murmur 2/4
ECG: SR at 80.Right axis deviation, rSR pattern



Question number 4

Which do you think is the most plausible diagnosis?

- 1. Acute pulmonary embolism
- 2. Desaturation during last trimester of pregnancy is normal, patient has normal findings for a pregnant woman
- 3. Acute pneumonia
- 4. Volume overload, diastolic RV failure with inversion of shunt through a PFO and subsequent desaturation



Question number 4

Which do you think is the most plausible diagnosis

- 1. Acute pulmonary embolism (PE)
- 2. Desaturation during last trimester of pregnancy is normal, patient has normal findings for a pregnant woman
- 3. Acute pneumonia
- 4. Volume overload, diastolic RV failure with inversion of shunt through a PFO and subsequent desaturation



Question number 5

If the patient had not cardiac disease
Which do you think is the most plausible diagnosis?

- 1. Acute pulmonary embolism (PE)
- 2. Desaturation during last trimester of pregnancy is normal, patient has normal findings for a pregnant woman
- 3. Acute pneumonia
- 4. Volume overload, diastolic RV failure with inversion of shunt through a PFO and subsequent desaturation



Question number 5

If the patient had not cardiac disease
Which do you think is the most plausible diagnosis?

- 1. Acute pulmonary embolism
- 2. Desaturation during last trimester of pregnancy is normal, patient has normal findings for a pregnant woman
- 3. Acute pneumonia
- 4. Volume overload, diastolic RV failure with inversion of shunt through a PFO and subsequent desaturation



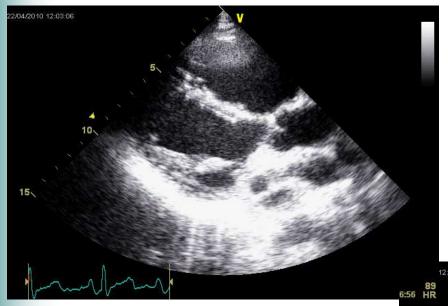
<u>Blood tests</u>: Hb 10,3g/L, Ht 30,8%, White cells 10.100, Platelets 237.000; QT 112% D-dimer: 2610, Glu 95; Urea 23; Cr 0,68; Na 140; K 3,47

CUS: compression ultrasound leg vein imaging normal, no thrombus

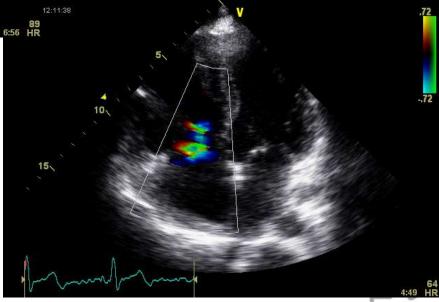
Echocardiogram:

- Right ventricular dilatation
- Normal RV systolic function
- Normal left sided structures
- Free PR, no progression of TR
- Peak TR= 2,2 m/s, no pulmonary arterial hypertension
- Marked RV diastolic dysfunction





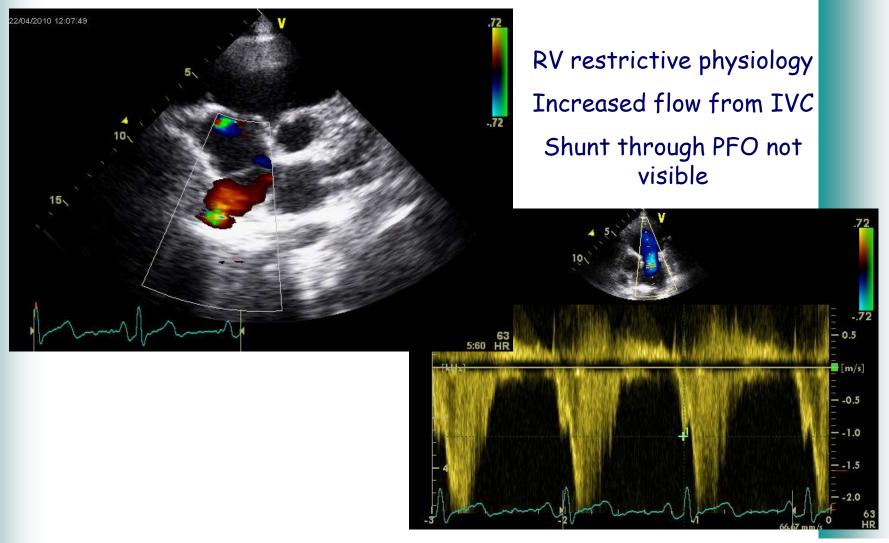
- ·Normal left sided structures
- ·Mild to moderate TR, free PR
- Normal RV systolic function
- ·RV dilatation





Hospital

Vall d'Hebron





Question number 6

Which of the following is not indicated for our patient?

- 1. Fetal monitoring
- 2. Admission, bed rest, and fluid restriction
- 3. ACE inhibitors
- 4. Prophylactic use of LMH and care of venous lines, including use of air filters
- 5. 1 to 4 are all correct



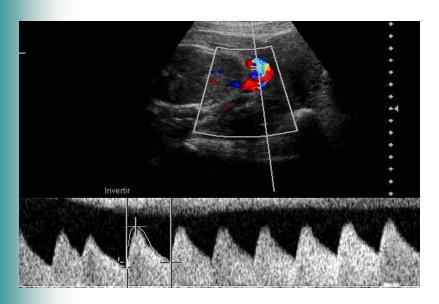
Question number 6
Which of the following is not indicated for our patient?

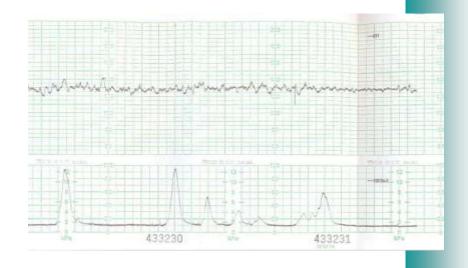
- 1. Fetal monitoring
- 2. Admission, bed rest, and fluid restriction
- 3. ACE inhibitors
- 4. Prophylactic use of LMH and fastidious care of lines, including use of air filters
- 5. 1 to 4 are all correct



Treatment:

- Admission
- Bed rest, 02 supplementation, restriction of fluids,
- LMH
- Fetal monitoring
- Air filter





During admission patient had uneventful course, fetal status correct

Question number 7

Which mode of delivery would you choose?

- 1. Emergent C-section
- 2. Elective next day C-section after pulmonary maturation
- 3. Vaginal delivery without epidural anastesia to avoid hipotension
- 4. Spontaneus onset of labour with epidural anaesthesia, limit second stage of delivery to minimise pain and anxiety



Question number 7

Which mode of delivery would you choose?

- 1. Emergent C-section
- 2. Programmed next day C-section after pulmonary maturation
- 3. Induced vaginal delivery without epidural anastesia to avoid hipotension
- 4. Spontaneus onset of labour, limit second stage of delivery with epidural anaesthesia to minimise pain and anxiety



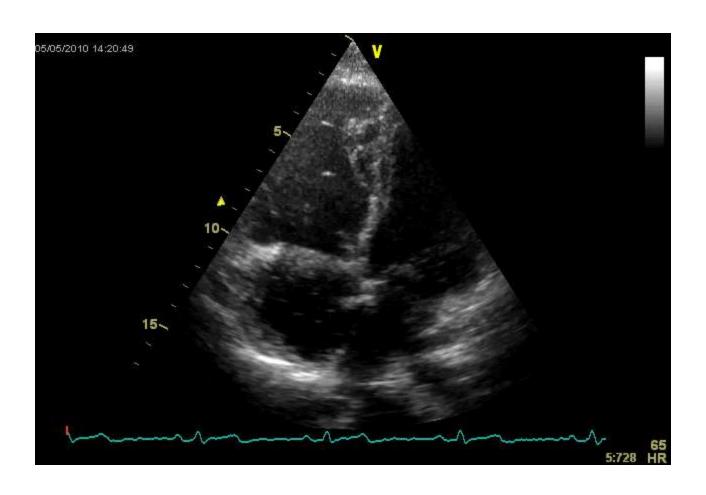
Delivery

- 38,5w epidural anesthesia,
- Spontaneus vaginal delivery,
- Baby boy 2930g, APGAR 9/10

Postpartum period:

- ·Postpartum period
- ·Peripheral edemas disapeared
- •O2 Sat went back to 93-94%
- Breast feeding

Thransthoracic contrast Echo was performed



·Basal right to left shunt through PFO



Question number 8

Which do you think is the correct answer regarding postpartum period?

- 1. 1 day stay at the hospital and discharge, cardiac problems disease occur almost only at delivery
- 2. Surgical pulmonary valve replacement and PFO closure at inmediate peripartum period
- 3. Careful postpartum follow up, balance fluid shift, mantaining LMH
- 4. Careful postpartum follow up, balance fluid shift, mantaining LMH and outpatient appointment during first month



Question number 8

Which do you think is the currect answer regarding postpartum period?

- 1. 1 day stay at the hospital and discharge, cardiac problems disease occur almost only at delivery
- 2. Surgical pulmonary valve replacement and PFO closure at inmediate peripartum period
- 3. Careful postpartum follow up, balance fluid shift, mantaining LMH
- 4. Careful postpartum follow up, balance fluid shift, mantaining LMH and outpatient appointment during first month



Question number 9

Which do you think are kee issues in pregnancy and CHD?

- 1. Prepregnancy counselling
- 2. Multidisciplinary team management
- 3. Fetal echocardiography to all patients as risk of transmission is 5-10%
- 4. Careful plan for labour, high level of maternal survelliance during postpartum
- 5. 1 to 4 are correct



Question number 9

Which do you think are kee issues in pregnancy and CHD?

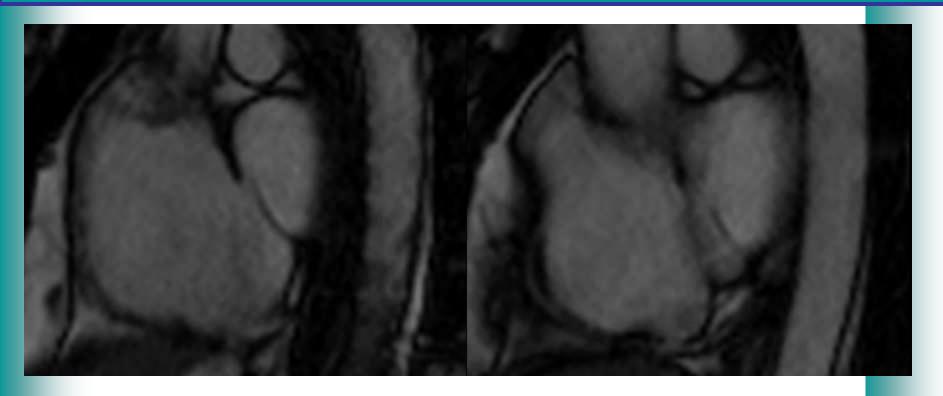
- 1. Prepregnancy counselling
- 2. Multidisciplinary team
- 3. Fetal echocardiography to all patients as risk of transmission is 5-10%
- 4. Careful plan for labour, spontaneous onset and high level of maternal survelliance during postpartum
- 5. 1 to 4 are correct



Slide for guidelines if published on Sunday regarding

Pre-pregnancy risk assessment Vaginal delivery is first choice..





6 months later: NYHA II, O2 Sat 93-94%

MRI: RVEF 61 % Regurgitant fraction PR 38%

RA dilatation 24 cm2,LVEF 58%

Exercise test MVO2: duration 9 min, 75% of predicted

Accepted for FFO closure and PVR



THANK YOU!!

