

TORINO, 03/12/2022

Pregnancy and systemic right
ventricle: a combination that takes
your breath away!

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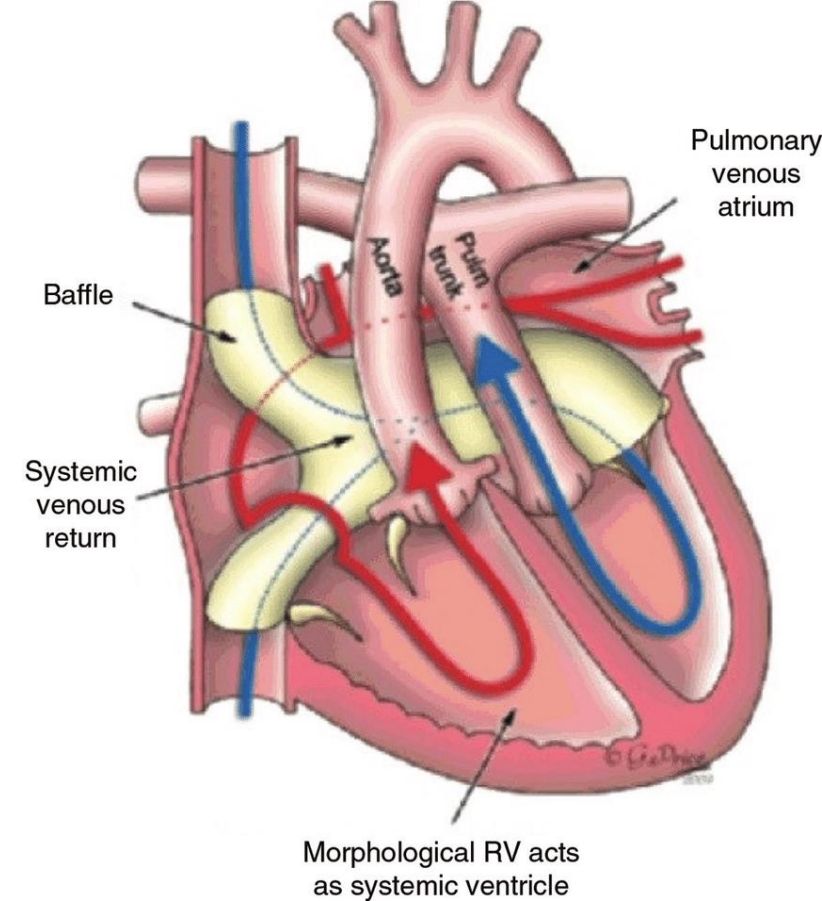
S.C. Cardiologia Pediatrica e centro GUCH

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Ospedale Infantile Regina Margherita

SARA, 34 y.o.

- **d-TGA and mitral cleft**
- 9 m.o. atrial switch with Mustard procedure
- 2014, 27 y.o. pulmonary venous pathway stenosis and post-capillary PH → Balloon angioplasty (Padova H.)
- 2019, 32 y.o. CMR:
 - systemic **RV hypertrophied with normal EF (68%)**.
 - Sub-pulmonary LV with normal EF.
 - moderate MR (cleft). **Moderate TR.**
 - LVOT obstruction with systolic anterior motion of the MV
 - **Moderate stenosis of pulmonary venous pathway.**



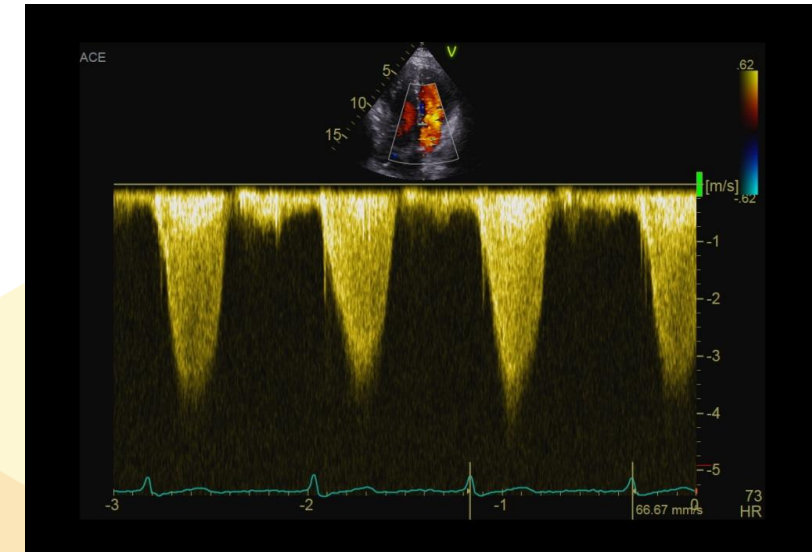
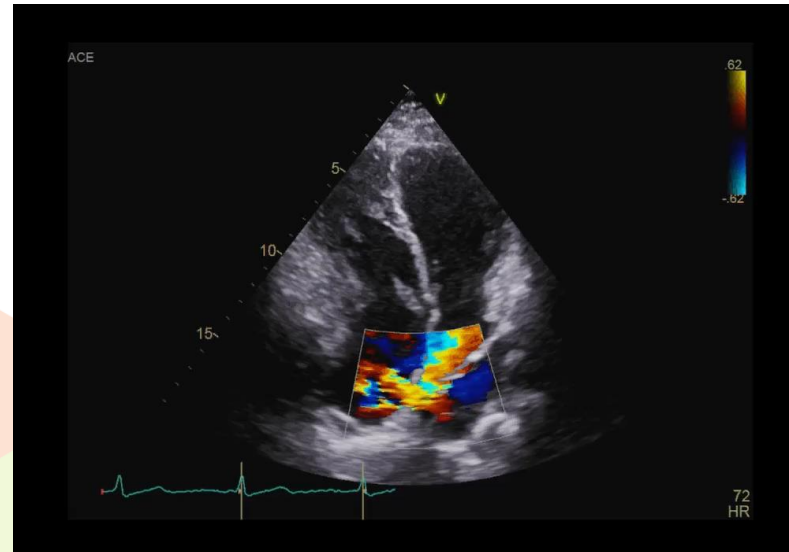
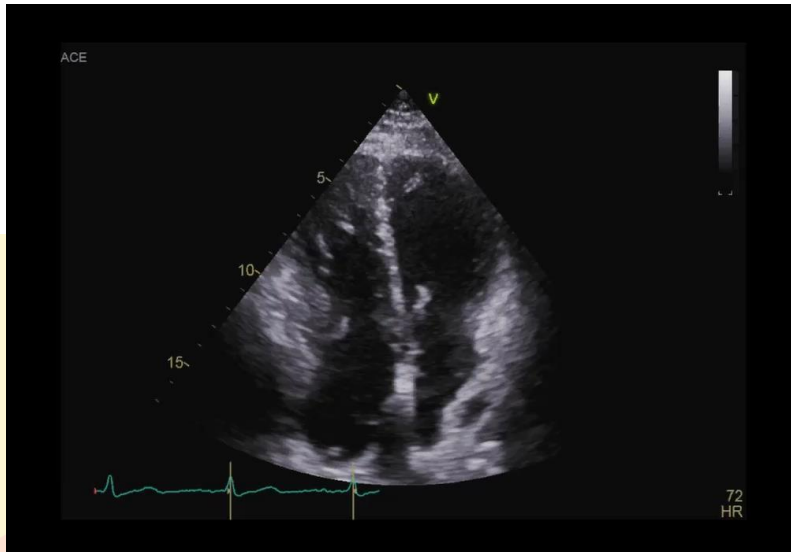
- **Lost at f-up**

- 02/2021 f-up: 34 y.o.,
important dyspnea, **NYHA III**

8 weeks pregnant

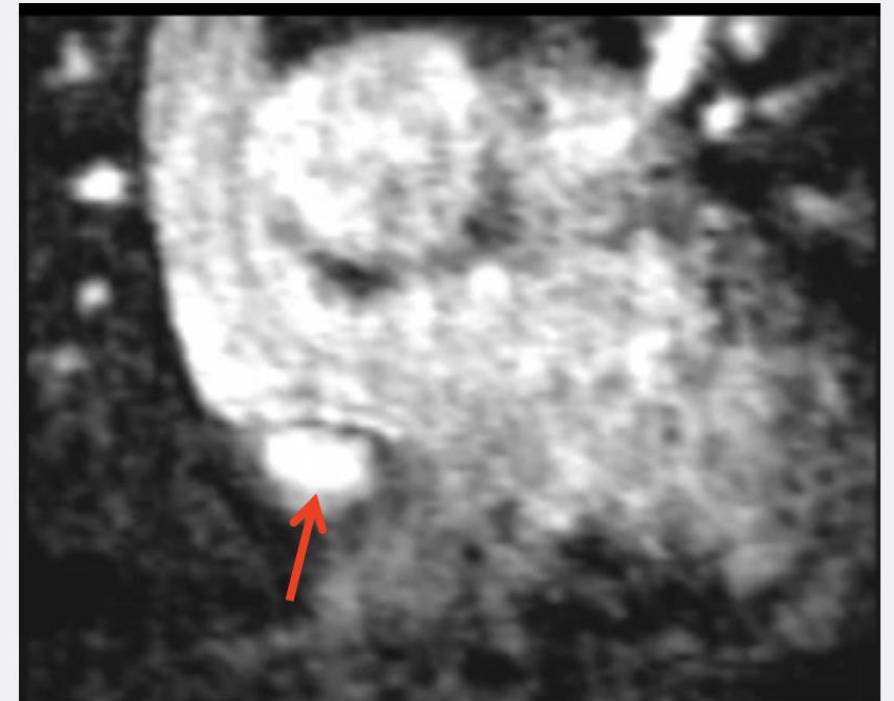
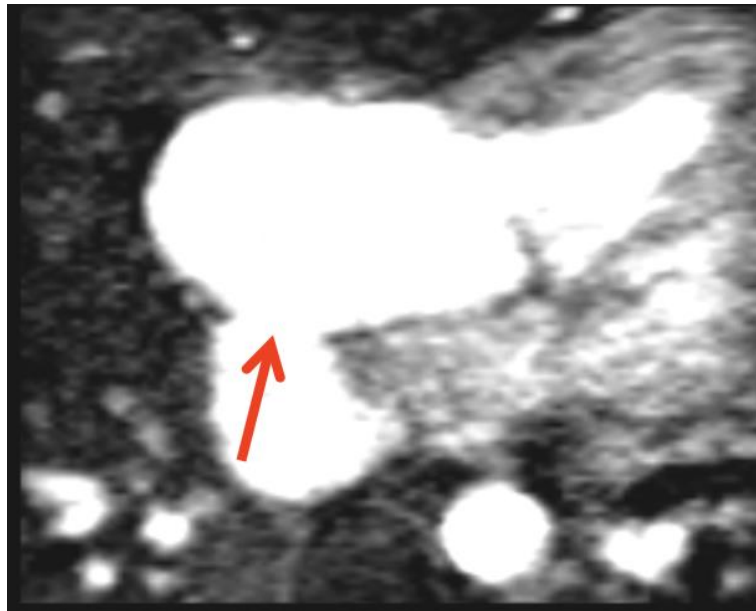


- Echo: Systemic RV normal EF. Subpolm LV 68%.
moderate-severe TR, mild-moderate MR
worsening of pulmonary venous return tunnel stenosis
subpulmonary LV with mild obstruction due to SAM



2021 CMR:

- Subpulmonary LV: EDV 78 ml/m², EF 70%
- Systemic RV: EDV 67 ml/m², EF 67%, hypertrophic
- Moderate LVOT obstruction with systolic anterior motion of the MV
- ***pulmonary venous returns directed to the right atrium, with obstruction at the outlet in the atrium with a pre-stenotic ectasia.***
- No evidence of atrial baffle detachment

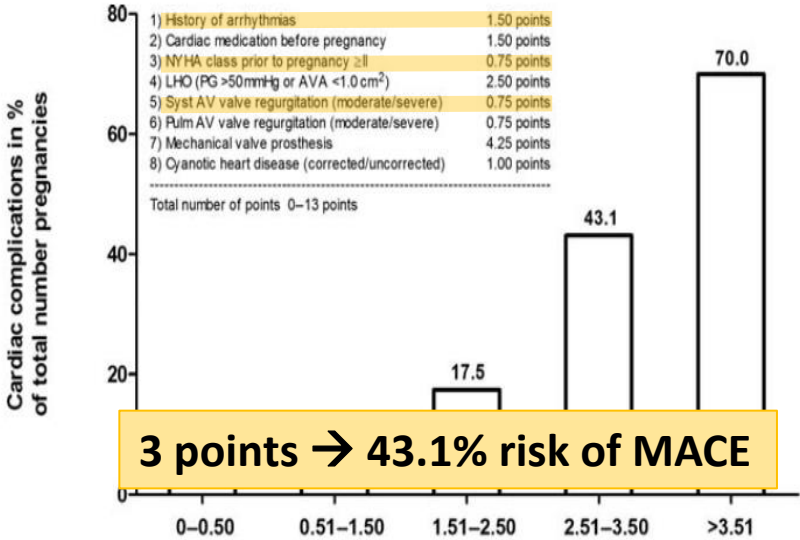


Assessing the maternal risk of MACE:

ZAHARA risk score

Europ. Heart J. (2010) 31, 2124–2132

1302 pregnancies, only CHD



ROPAC risk chart

European J of HF (2016) 18, 522–523

2742 pts

58% CHD

Table 3 Risk chart for a cardiac event

Pre-pregnancy:	Emerging countries				Advanced countries			
	No	AF	HF	HF+AF	No	AF	HF	HF+AF
	35	73	65	90	35	73	65	90
	25	62	58	84	25	62	58	84
	11	37	29	67	11	37	29	67
	5	22	16	49	5	22	16	49
	5	20	15	46	5	20	15	46

MACE

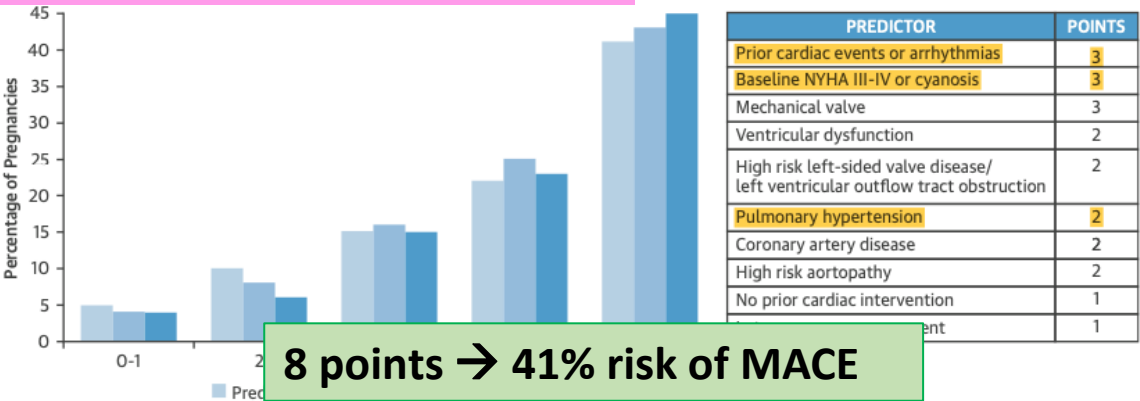
Risk of cardiovascular events
between 41% – 65%

CARPREP risk score

JACC 2018;71:2419–30

1938 pregnancies

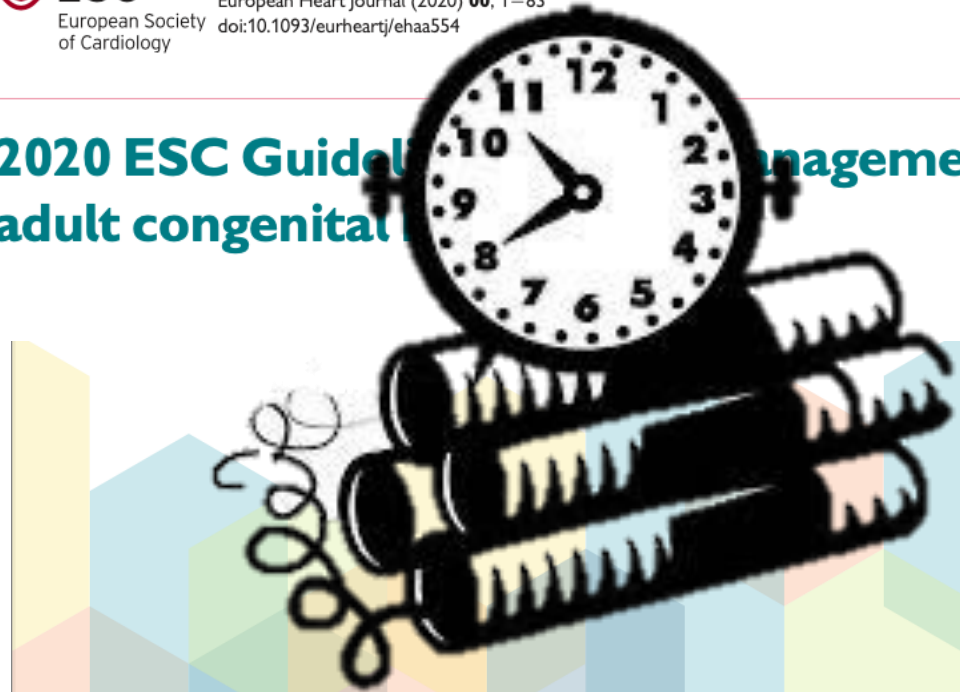
63% CHD and acquired HD/arrhythmias



2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%
Counselling	Yes	Yes	Yes	Yes: expert counselling required	Yes: pregnancy contraindicated: if pregnancy occurs, termination should be discussed

2020 ESC Guidelines for the management of adult congenital heart disease



Recommendations for intervention in transposition of the great arteries after atrial switch operation

Recommendations	Class ^a	Level ^b
Indications for surgical intervention		
In symptomatic patients with pulmonary venous atrium obstruction, surgical repair (catheter intervention rarely possible) is recommended.	I	C
In symptomatic patients with baffle stenosis not amenable to catheter intervention, surgical repair is recommended.	I	C
Indications for catheter intervention		
In symptomatic patients with baffle stenosis, stenting is recommended when technically feasible.	I	C

CLINICAL AND TECHNICAL PROBLEMS

1st trimester pregnancy – fetal protection

United States Nuclear Regulation Commission (USNRC)

1- degrees of radiation exposure

fetus radiation dose < 50 mGy is considered safe

2- gestational age

embryo/fetus most susceptible during organogenesis
(2 to 7 weeks gestational age)

Access difficulties

1- Reach the Landing zone

Difficult through the retrograd way AO-RV-RA-LA-PV
(long way)

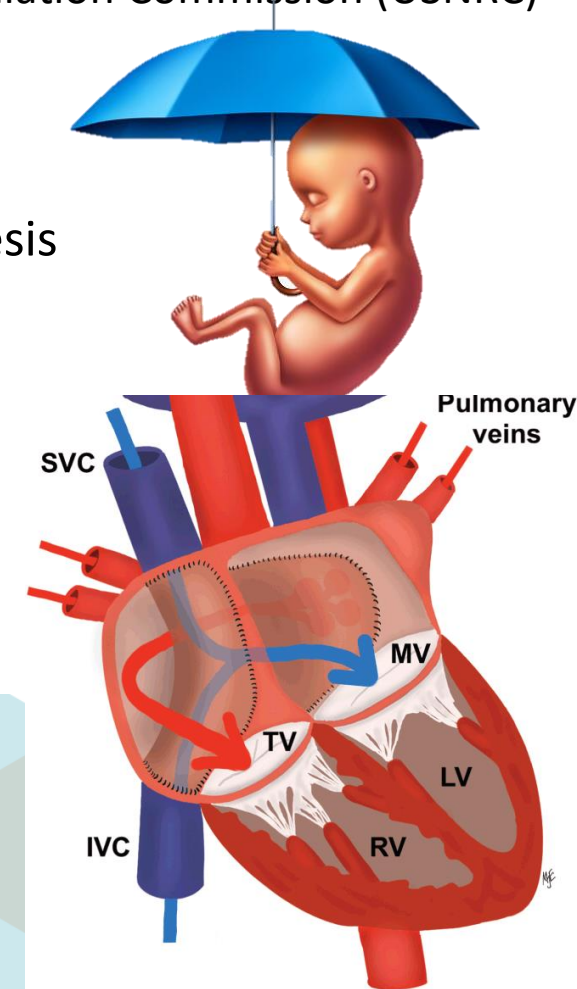
Maybe better with a Trans-septal puncture?

2- Long Distance

12-14F **catheter length > 90 cm** (for CP stent) **not available.**
Delivery with Nudel system?

3- Diameter

Balloon Expandable Endoprosthesis Gore Viabahn VBX not available > 11mm diameter-
Post dilatation max 16 mm → **risk of displacement or embolization**



Cardiac model with 3D printing

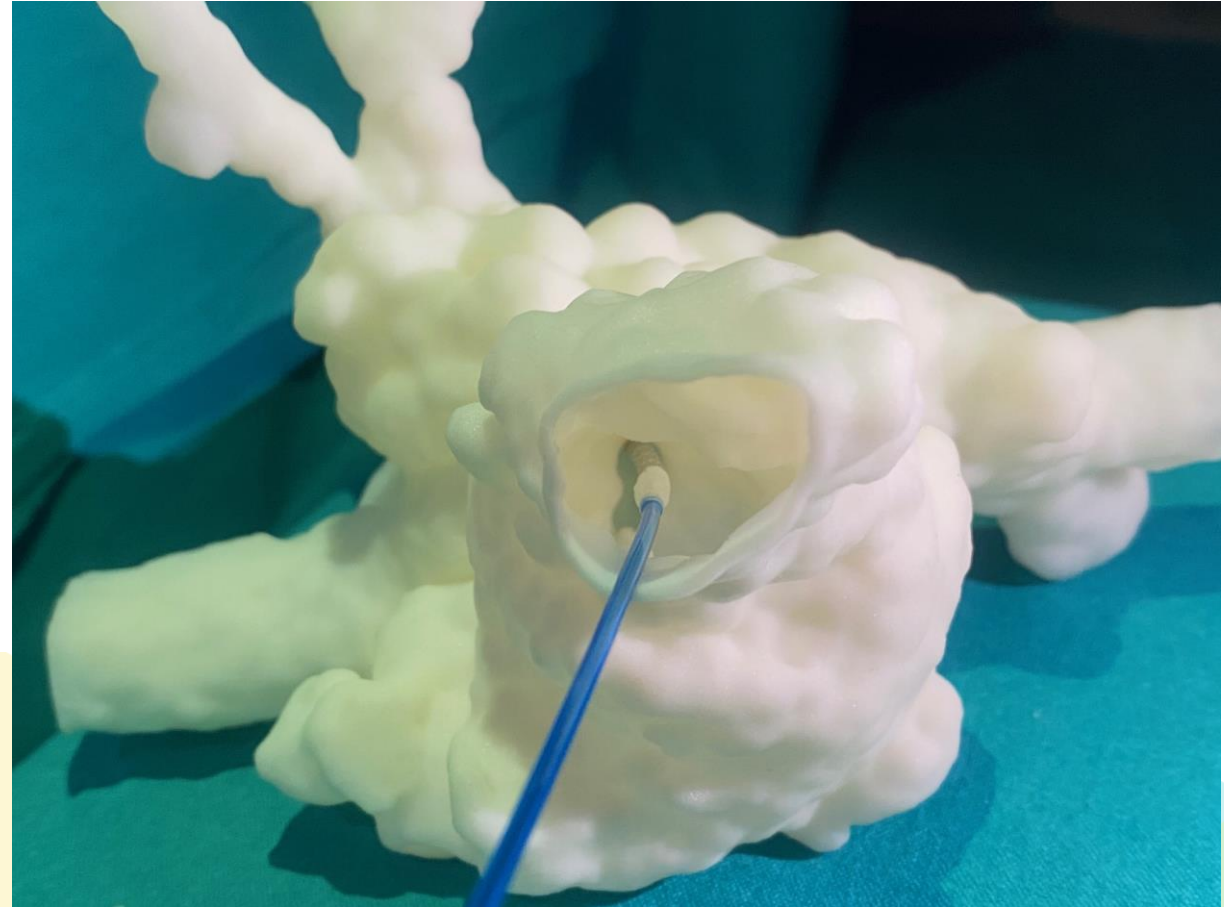
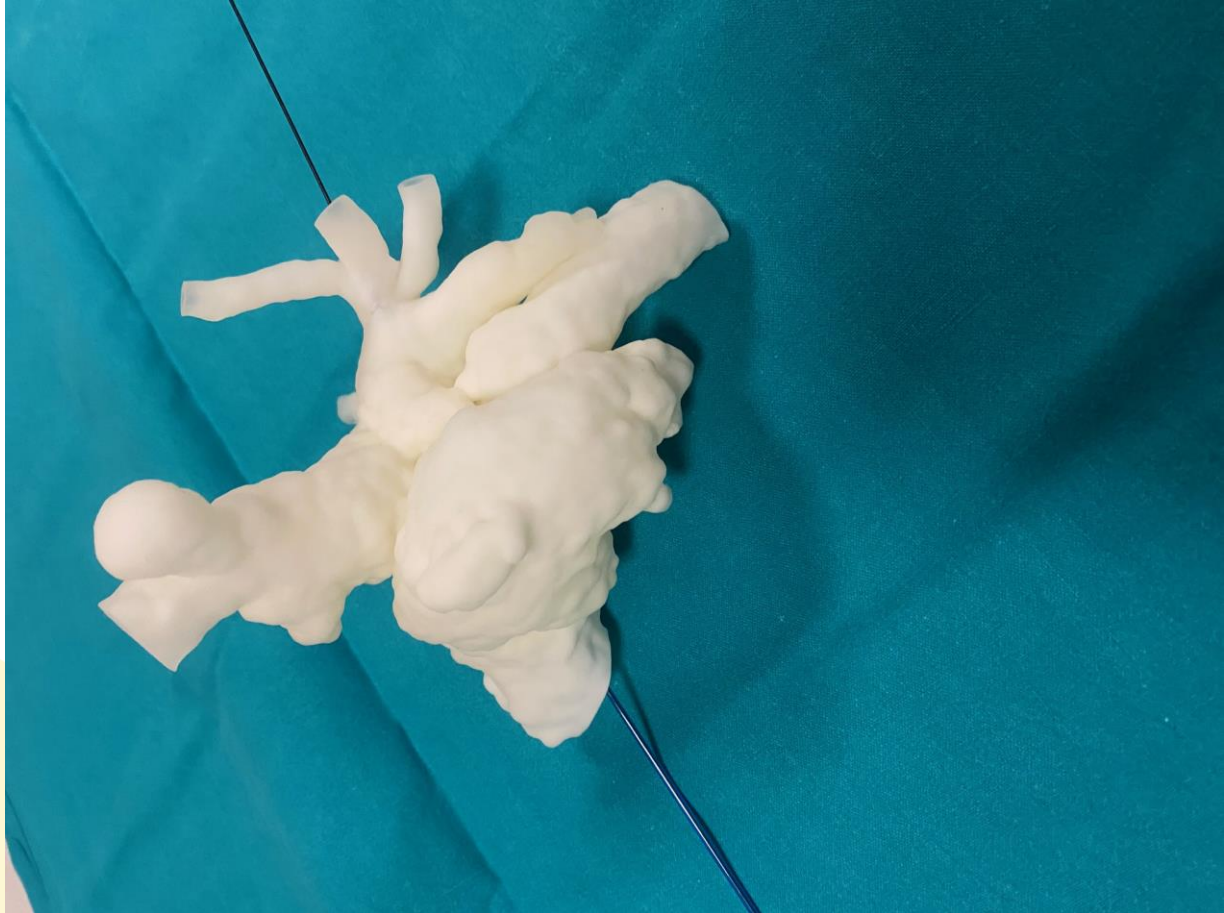


Fondazione IRCCS
Policlinico San Matteo

Sistema Socio Sanitario



Regione
Lombardia



courtesy of Dr. Annoni

Cath lab accurate planning with 3D Cardiac model

Trans-septal puncture

- Not suitable from IVC: puncture in landing zone
- Not suitable from SVC: high risk to go outside the heart



Simulation Stent Release

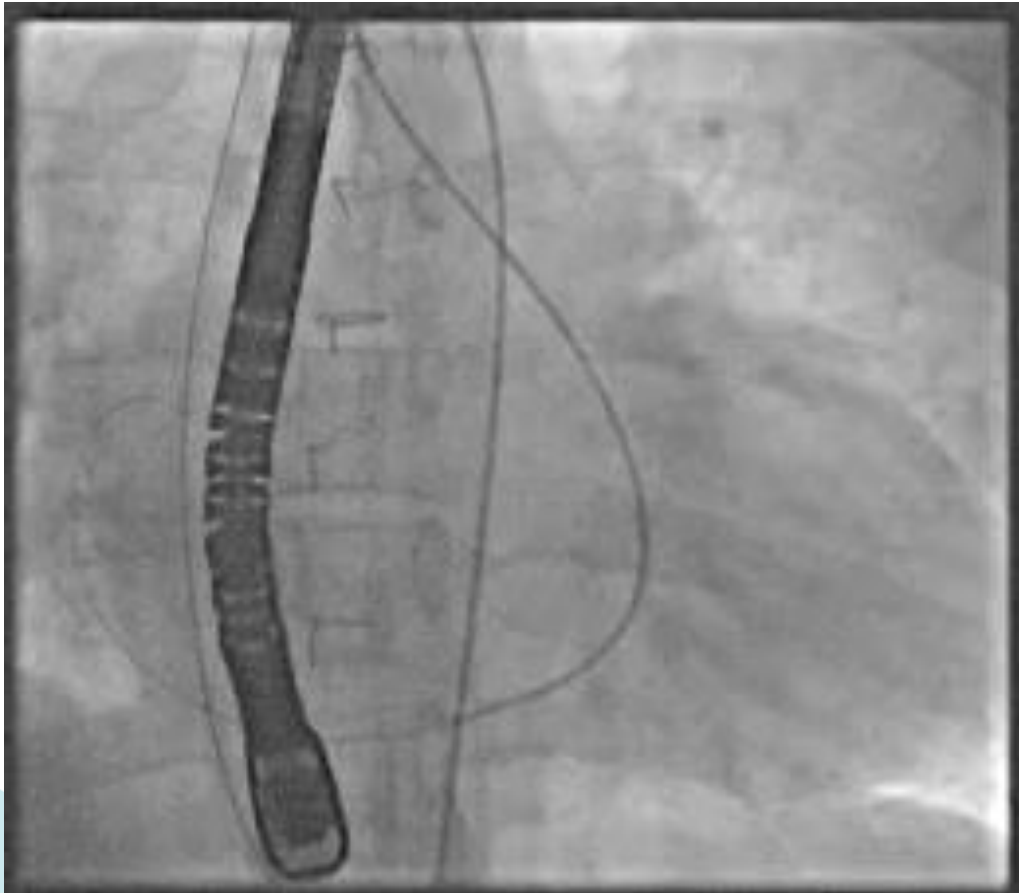
- **NuDEL CP Stent Delivery System** (diam 18 lugh 34mm)
- **GORE VIABAHN VBX Balloon Expandable Endoprosthesis** (diam 11 lugh 29mm e 39mm) → R of embolization
- **BAILOUT SCENARIOS: simple angioplasty** (Bard Atlas Gold)



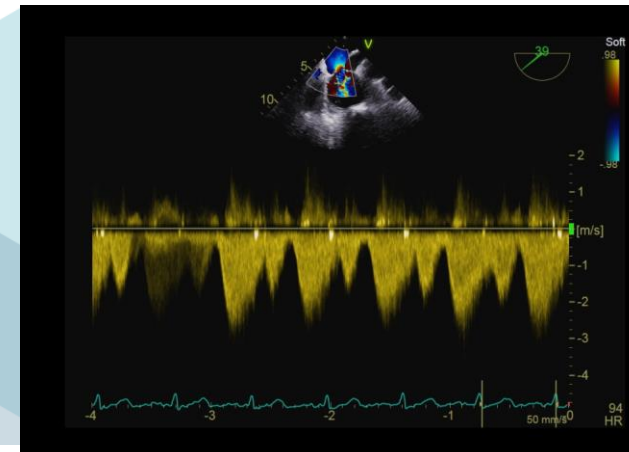
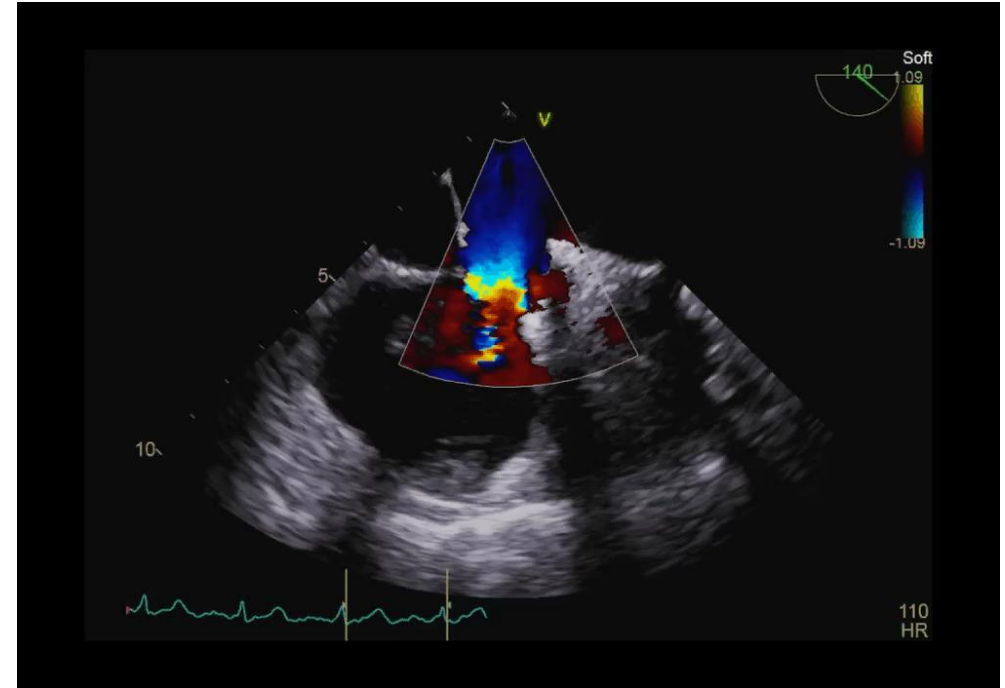
courtesy of Dr. Annoni

PROCEDURE

14 weeks of pregnancy General anesthesia TOE-guided procedure

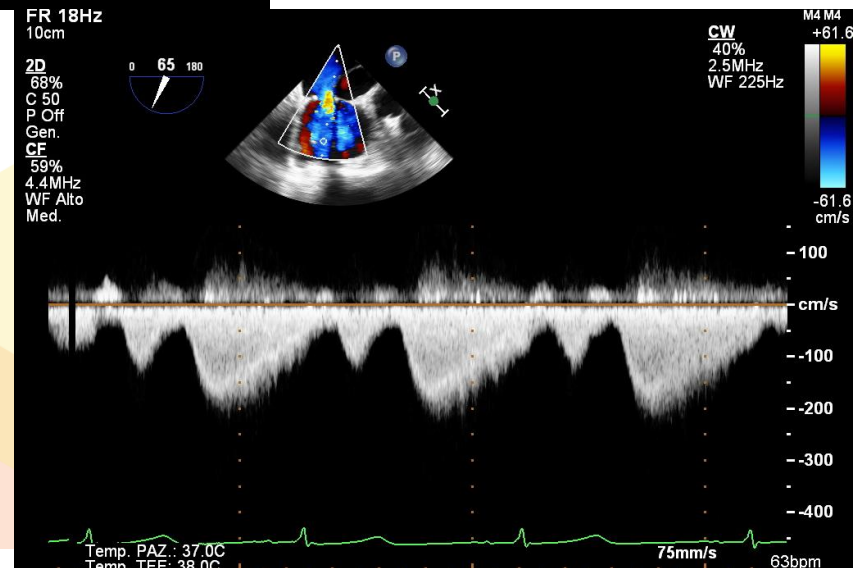
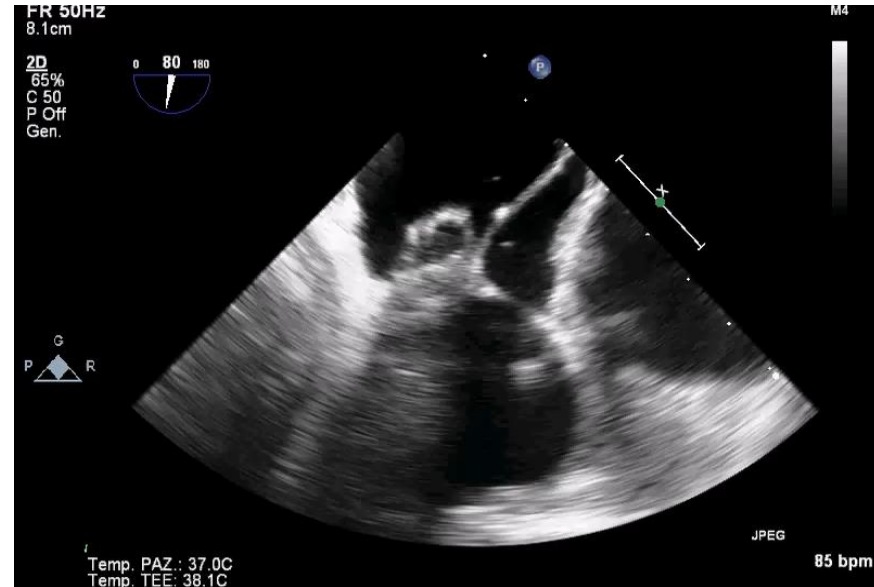


- ✓ Fluoro: 37 min
- ✓ Angio: NO
- ✓ Contrast: 30ml



PROCEDURE

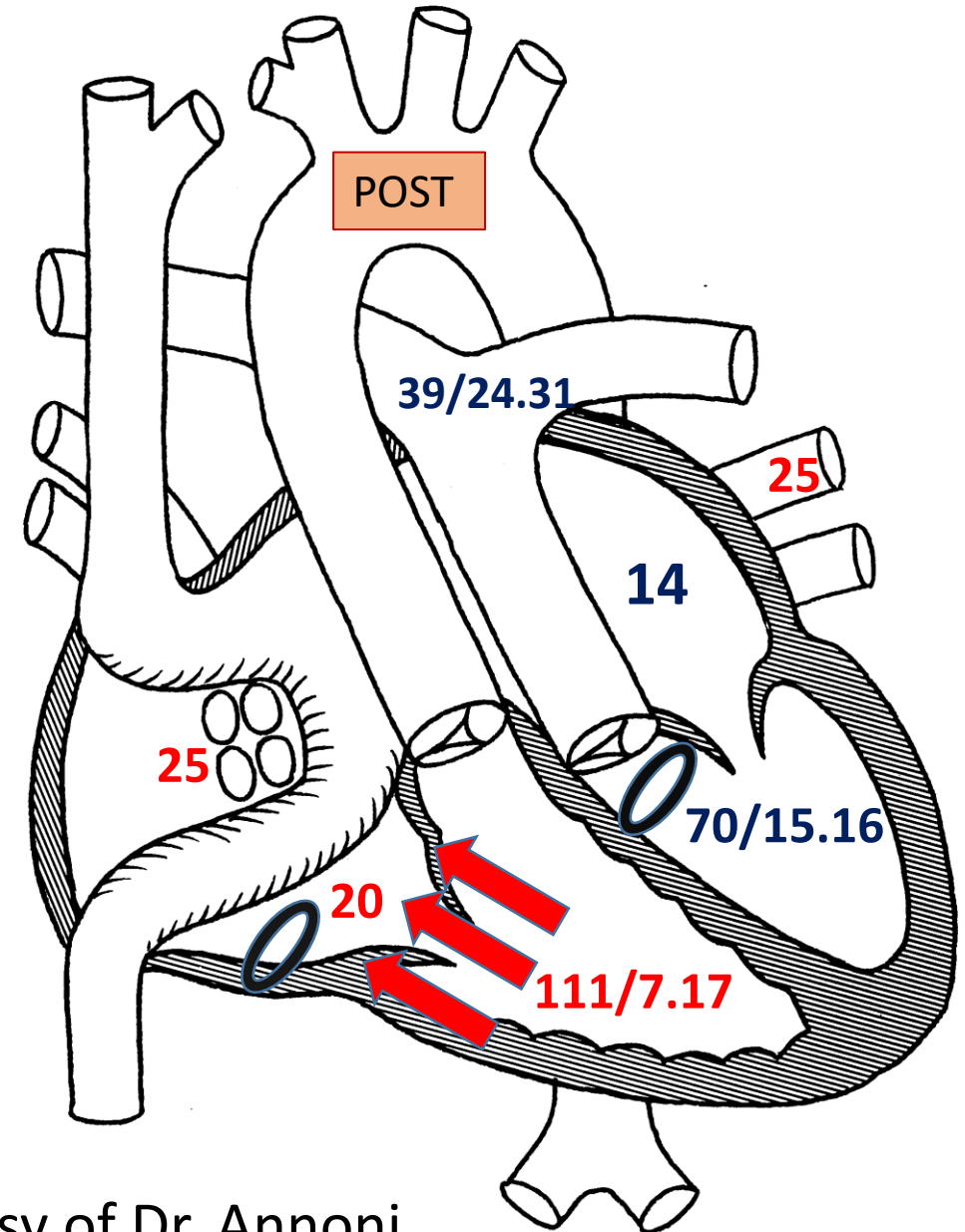
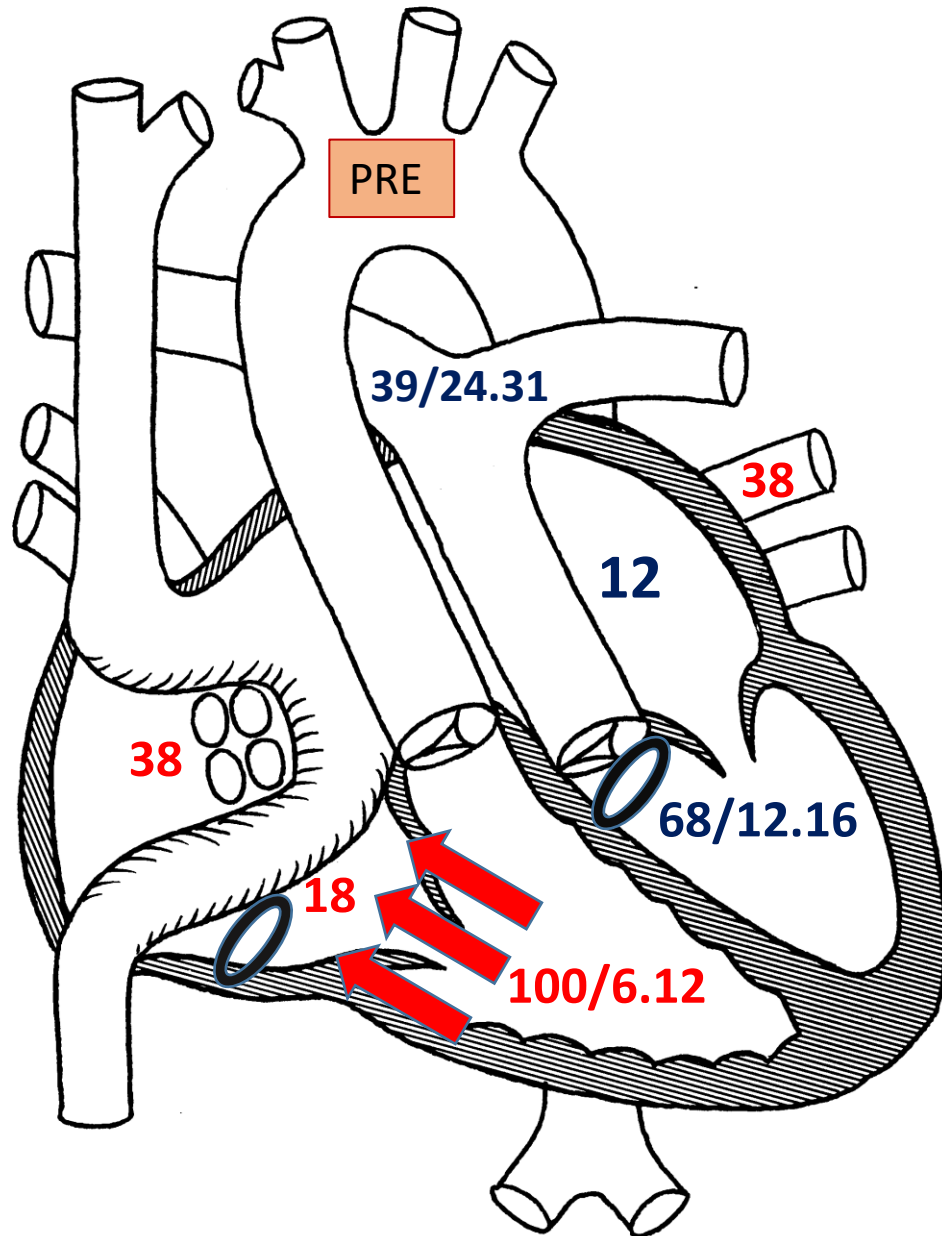
14 weeks of pregnancy General anesthesia TOE-guided procedure ✓
✓
✓ Fluoro: 37 min
Angio: NO
Contrast: 30ml



courtesy of Dr. Annoni

RESULTS

Started ASA



courtesy of Dr. Annoni

F-up

- **Strict f-up during the pregnancy**
- Several SVT → treated with metoprolol 25 mg x 2 → 50 mg + 25 mg
 - Last 20 days amiodarone has been added, 5 days/week
- At **31 w + 1** → For worsening HF, Killip class III → C-section

Newborn of 2000 g

Apgar score 7,8



Last F-up 10/2022

- NYHA II, rare palpitations Meds: furosemide 25 mg bid, metoprolol 50 mg + 25 mg
- Holter ECG: RS, rare VPC e SVPC, no pauses.
- **Echo: RV with mild dysfunction** (FAC 35%, S' lat 7 cm/s) **TR moderate-severe pulmonary venous return tunnel stenosis with 20 mmHg** (difficult to measure)
LV with EF 60% LVOT obstruction with SAM (60/30 mmhg).



1. Catheterization to evaluate pulmonary venous return stenosis?
2. Add a ACEi/ARB/ARNI?
3. Tricuspid valve surgery ?



AC/DC

ACHD cardiologist

Gynaecologist

Internal medicine
specialist

Anesthesiologist

Neonatologist



Thank you!

