IX Congresso
Novas Fronteiras em Medicina Cardiovascular
22 a 24 de Fevereiro 2019
The Village Praia D'El Rey - Óbidos

RICA HF team
Departamento Coração e Vasos
Hospital de Santa Maria
Hospital Pulido Valente
Faculdade de Medicina Lisboa

Clinical Cases: dealing with controversies

An uncommon case of HFrEF

Joana Rigueira,
Inês Aguiar-Ricardo

2019

Clinical Case

- 19 year-old female patient
- **Evacuated from S. Tomé and Principe for heart failure**

- No previous medical history known
  - No history of viral infection in the previous months

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Current History

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Asymptomatic

September 2018

Childbirth 1st son

10 days

Heart failure symptoms
Dyspnoea
Orthopnea
Peripheral edema

Hospital admission

Noradrenaline and diuretics

TTE: Large VSD; LV dilation; Moderate depression of LVEF

Discharge after 10 days
NYHA III

Medication: furosemide 40mg (1id), spironolactone 25mg (1id)

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Current History and physical examination

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- Arrived to Portugal in December→ Hospital Emergency Department

Admission:

- GCS 15
- BP **97/66** mmHg HR **102bpm, NSR**;
- TT 37ºC, **Sat O2 (O2 2l/min)=96%, acyanotic**
- Regular rate and rhythm, normal S1, **↑S2**, no S3, **pansystolic heart murmur (III/VI) in precordial region** (+ left sternal border)
- **Bibasilar inspiratory crackles**
- No jugular venous distension, nor hepatomegaly, nor peripheral oedema
- No digital clubbing

UTIC

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EKG

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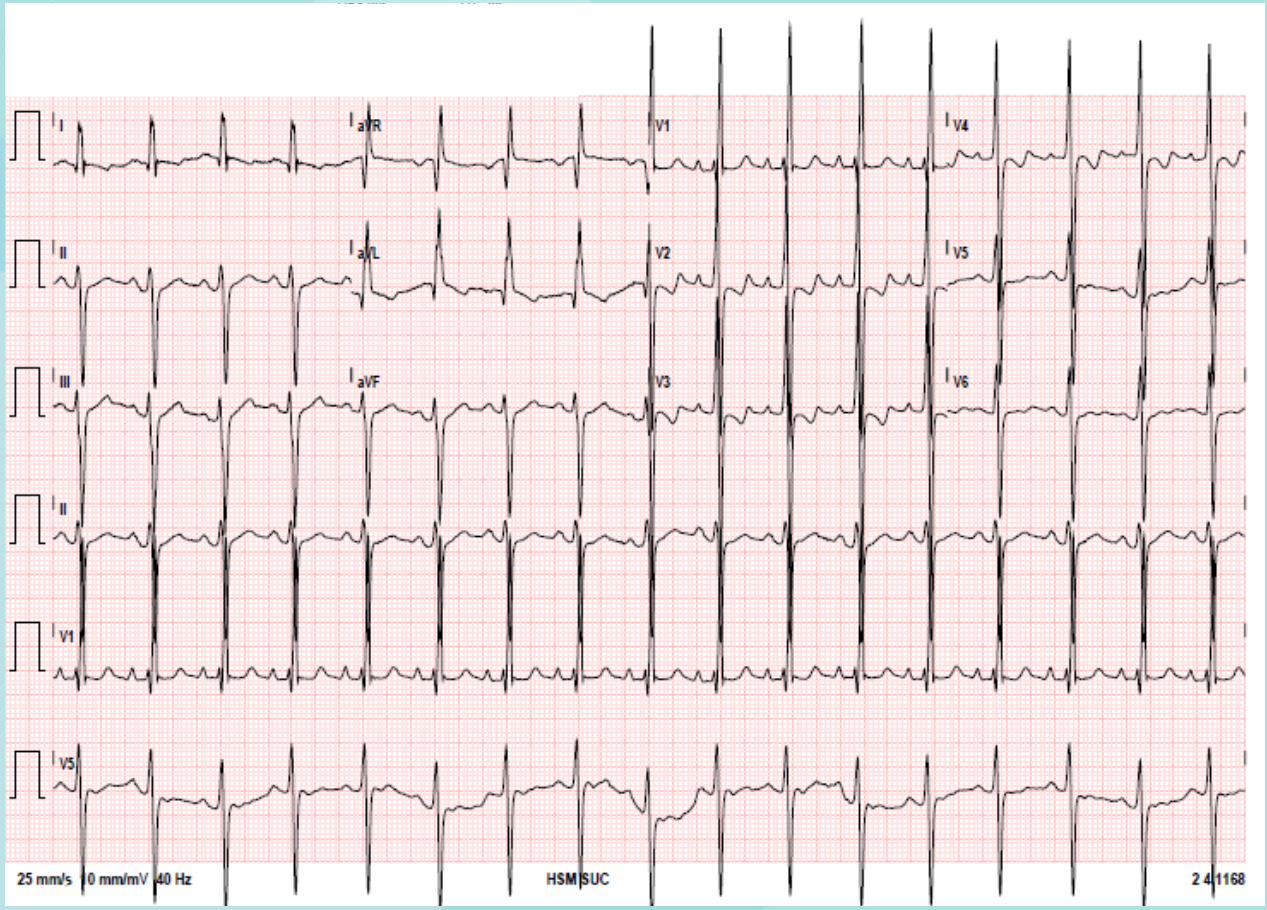

Dealing with controversies: An uncommon case of HFrEF

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Transthoracic echo

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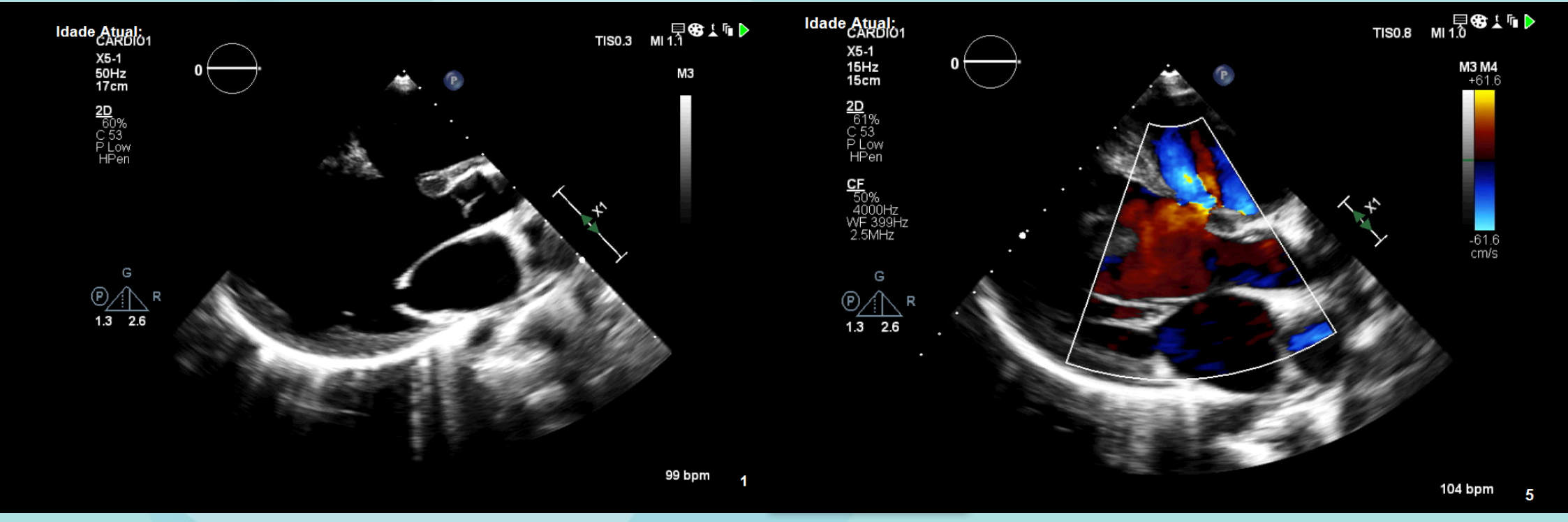

Dealing with controversies: An uncommon case of HFrEF

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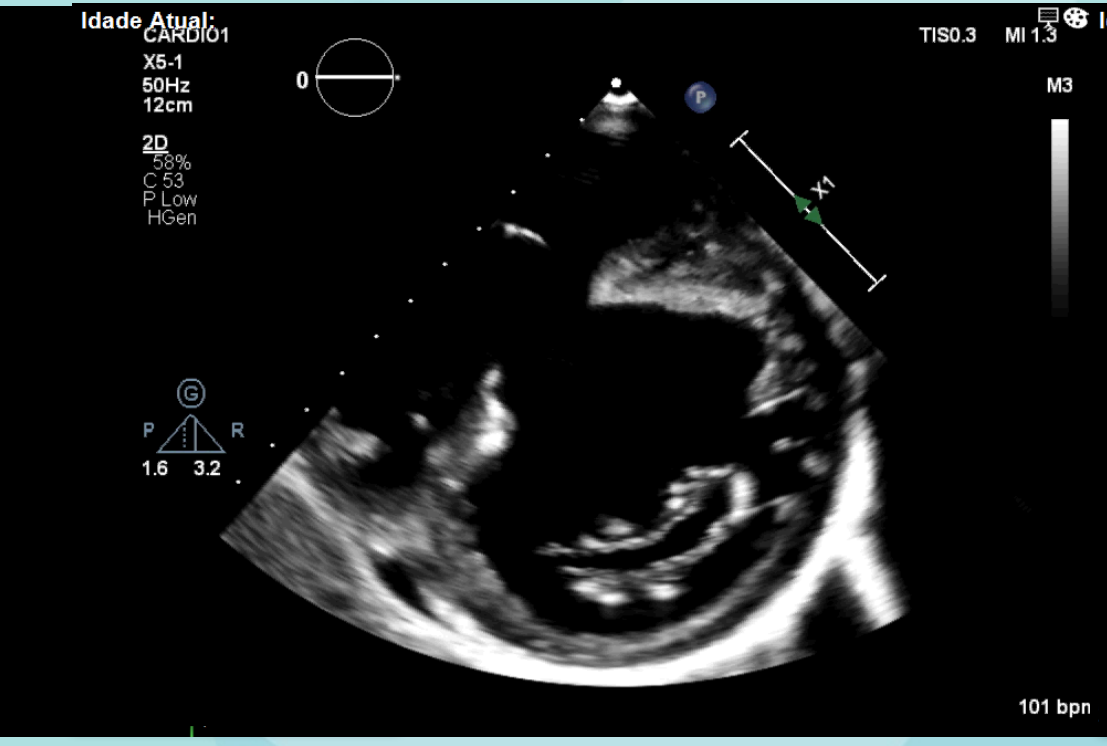

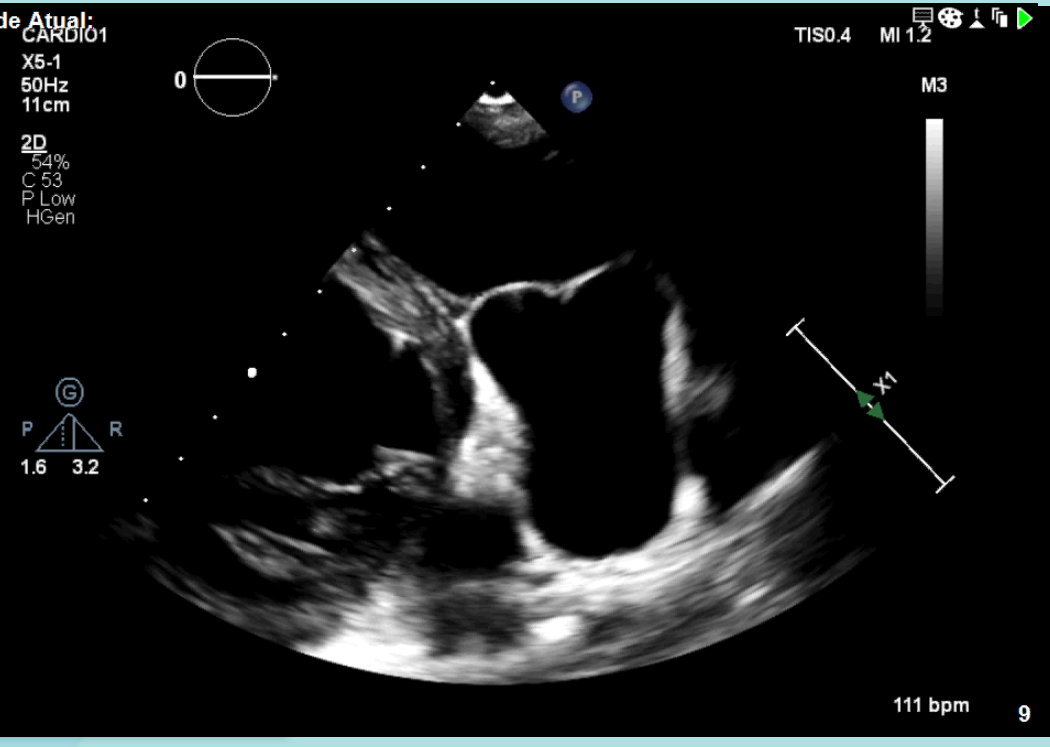

Pulmonary artery trunk= 35mm

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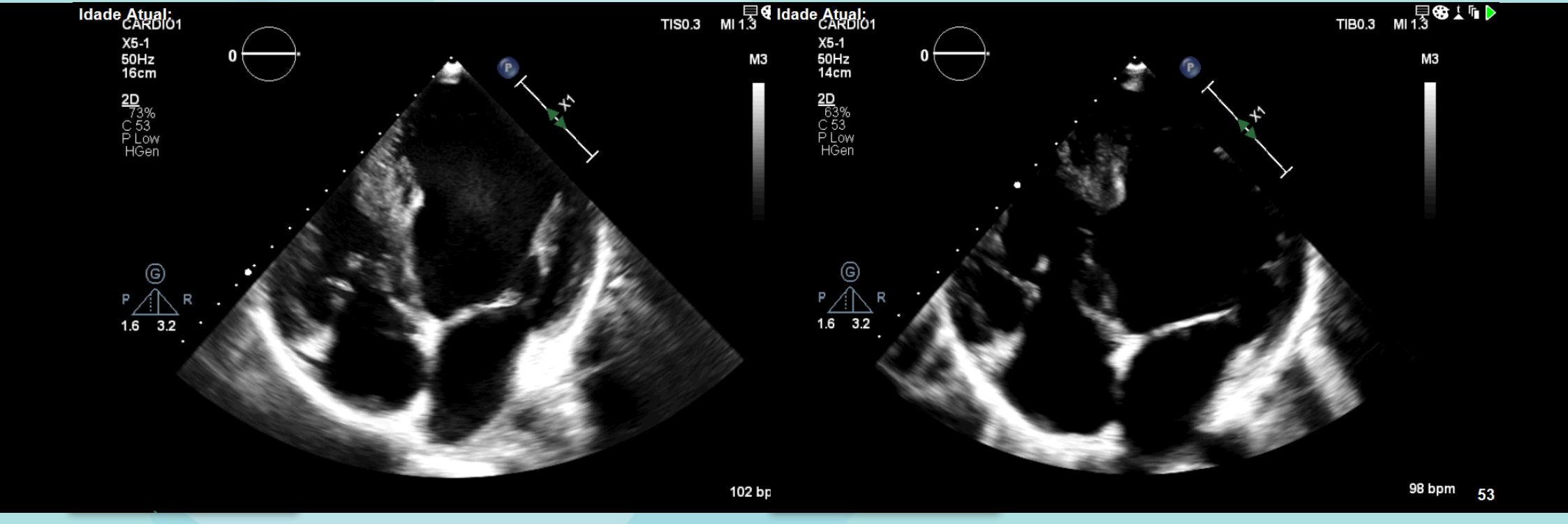

LV EDV = 144mL/m2; EDD (PLAX) =53mm/m2

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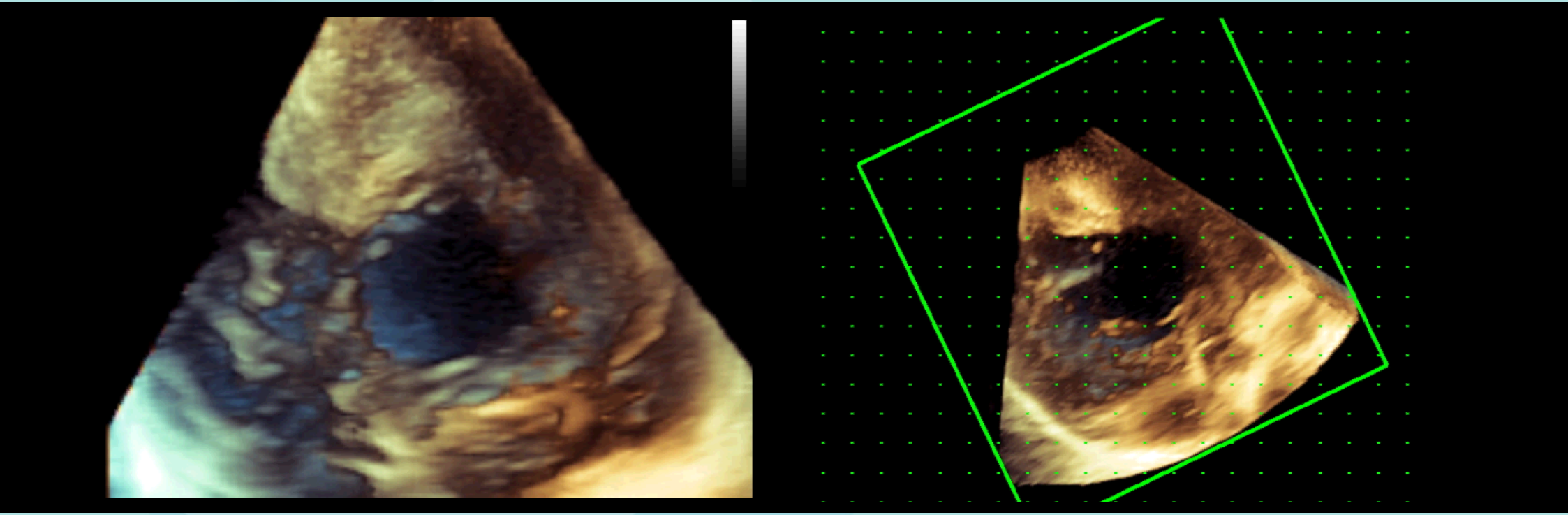

26 x 26 mm Area= 5.6cm^2

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Blood analysis

Hb	14.8g/dL
RBC	4,9x10^12/L
Htc	44.7%
Leukocytes	5.86 x10^9/L
Neutrophils	45%
Platelets	249 x10^9/L
INR	1.14
Urea	24 mg/dL
Creatinine	0,67 mg/dL
eGFR	130 mL/min/1.73

ALT	31 U/L
AST	27 U/L
T Bil	0-48 mg/dL
LDH	252 U/L
GGT	44U/L
CK	59 U/L
TnT-hs	9 ng/L
NTproBNP	**4321 pg/mL**
CRP	**3,27mg/dL**
Na^+/K^+ /Mg^{2+}	141/3.9/2.1

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Evolution

- IV Diuretics→ PO diuretics
- No need of vasopressor drugs
- NYHA II

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Questions at this point

- **LVEF dysfunction cause**
  - LV volume overload (VSD)?
  - Peripartum cardiomyopathy?
  - Coronary heart disease (congenital…)?
  - Other

- **Most appropriate treatment**
  - Medical therapy
  - VSD closure?

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LVEF dysfunction investigation

- **Blood tests results**
  - HIV neg
  - Calcium normal
  - Thyroid function tests normal

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LVEF dysfunction investigation

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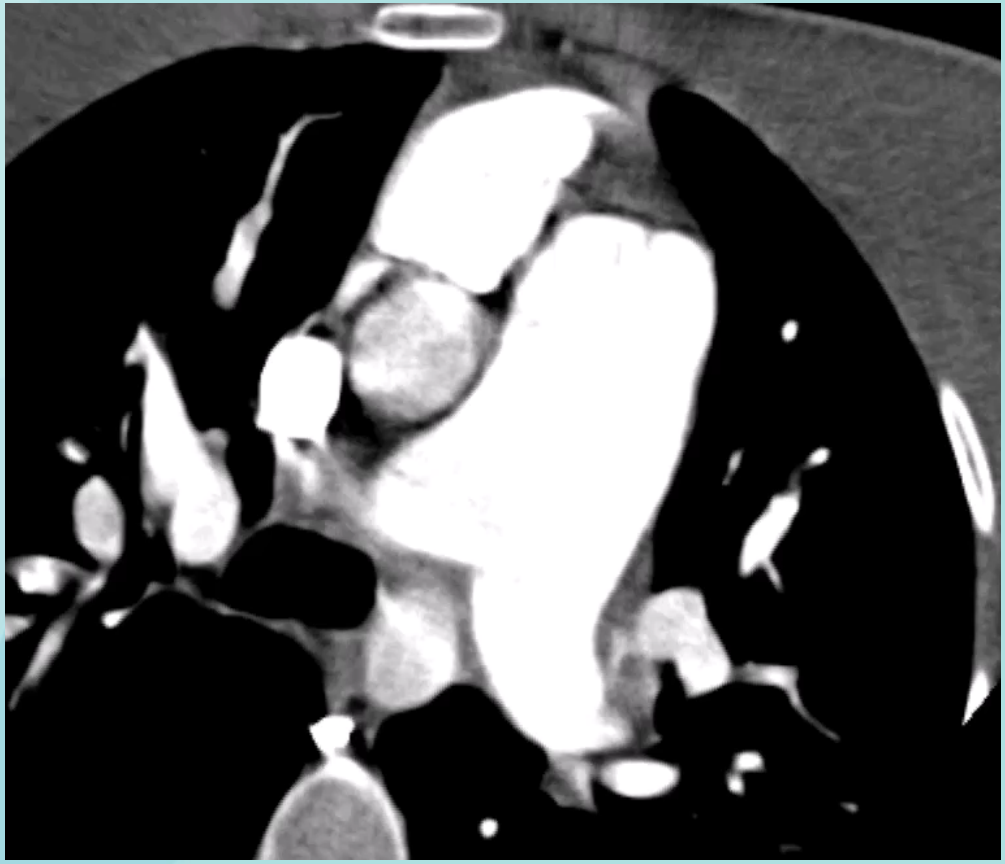
- CT coronary angiography



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LVEF dysfunction investigation

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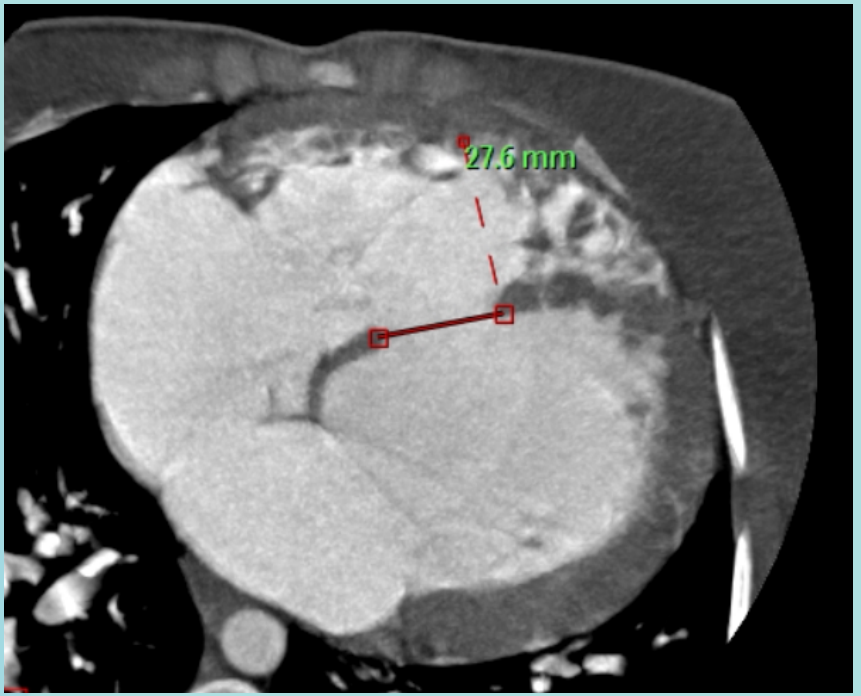
- CT coronary angiography



- Coronary arteries with normal origin without obstructive disease
- VSD with 2.8cm
- Left-right shunt

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Right heart catheterization

Basal study	
Aorta (s/d/m)	119 / 66 / 88
PA (s/d/m)	117 / 56 / **75**
RA (a/v/m)	11 / 6 / 6
PAWP (mmHg)	**10**
TPG (mmHg)	65
Cardiac output (Fick)	2.13 (l/min)
Cardiac index Fick	1.63 (l/min/m²)
Qp / Qs Fick:	**1.57**
SVR (Fick)	38,5 Wood U
PVR (Fick)	**19.3 Wood U / 25.2 uW.m2**

After NO 30ppm	
Aorta (m)	72
PA (s/d/m)	117 / 53 / 72
RA (m)	6
TPG (mean)	61
Pulmonary output	4.75
AP saturation	85%
Ao Saturation	90%
Qp / Qs Fick	**2.1**
PVR (Fick)	**12,8 Wood U**

4- Reduction of pulmonary vascular resistance > 20%

VENTRICULAR SEPTAL DEFECT CLOSURE– *YES OR NO?*

RVPi > 8 uW.m^2

RVP > 4,6 uW

4 uW.m^2 ≤ **RVPi** ≤ 8 uW.m^2

2,3 uW ≤ **RVP** ≤ 4,6 uW

Do not close

'Treat and repair'

Parcial closure

RVPi < 4 uW.m^2

RVP < 2,3 uW

VENTRICULAR SEPTAL DEFECT CLOSURE– *YES OR NO?*

ESC European Society of Cardiology

European Heart Journal - Case Reports (2018) **2**, 1–7
doi:10.1093/ehjcr/yty033

CASE SERIES

Treat-and-repair strategy is a feasible therapeutic choice in adult patients with severe pulmonary arterial hypertension associated with a ventricular septal defect: case series

Satoshi Akagi[1]*, Shingo Kasahara[2], Toshihiro Sarashina[1], Kazufumi Nakamura[1], and Hiroshi Ito[1]

Case 1

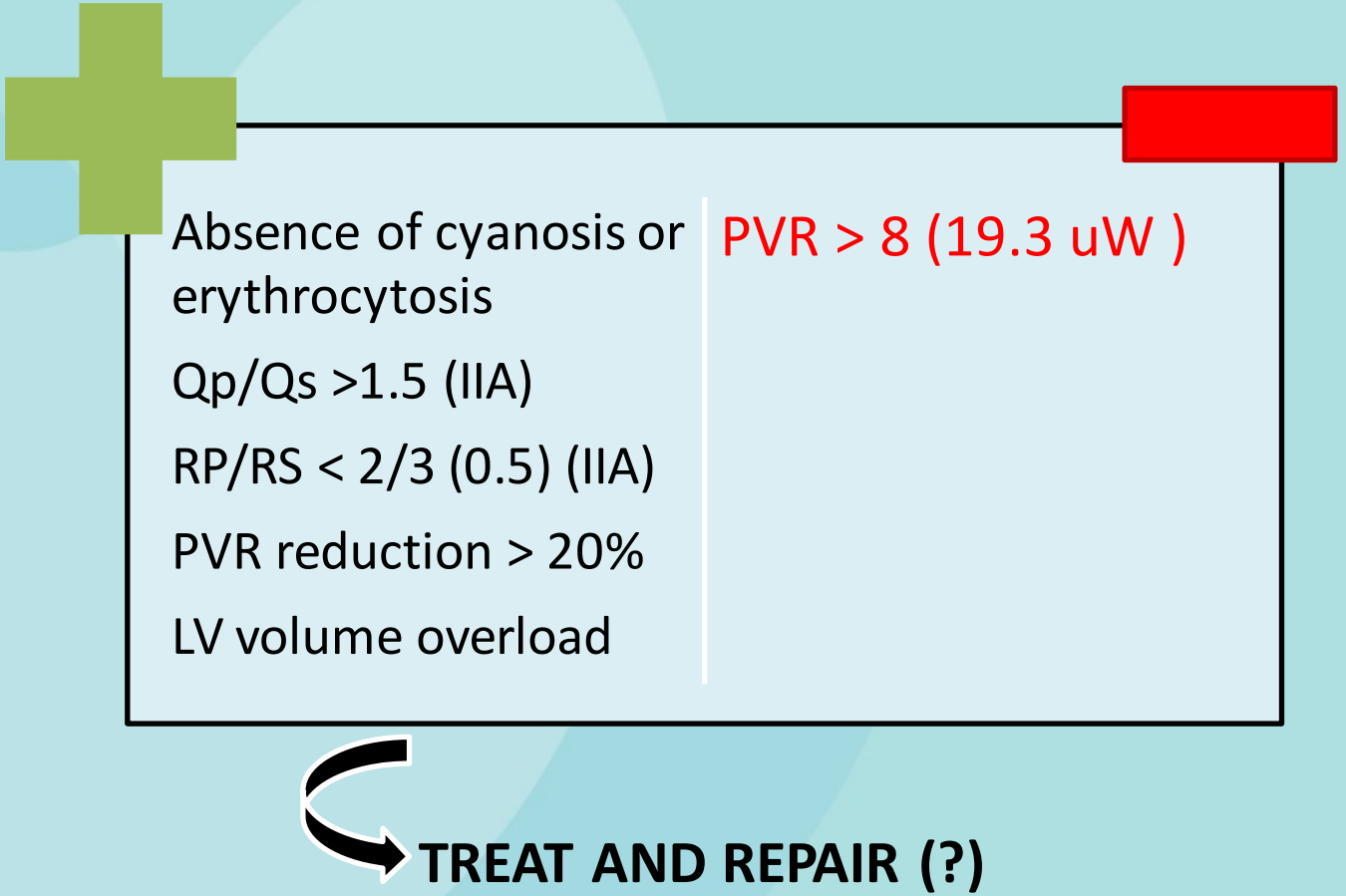
At birth	Diagnosis of ventricular septal defect (VSD)
49 years of age	Diagnosis of pulmonary arterial hypertension (PAH). Mean pulmonary artery pressure (PAP) of 76 mmHg and indexed pulmonary vascular resistance (PVRi) of 15.8 Wood units/m^2. Started treatment with sildenafil (60 mg/day) and ambrisentan (5 mg/day)
3 months later	Addition of epoprostenol and titration of the dose
12 months later	Mean PAP of 58 mmHg, PVRi of 8.8 Wood units/m^2. Repair of VSD
22 months later	Mean PAP of 42 mmHg and PVRi of 11.5 Wood units/m^2 (sildenafil at 60 mg/day, ambrisentan at 5 mg/day, and epoprostenol at 24 ng/kg/min)

Table I Haemodynamic change in Patient 1

	First evaluation		Post-treatment		Post-operation
	Baseline	Oxygen	Baseline	Oxygen	Baseline
mPAP (mmHg)	76	73	58	72	42
mSAP (mmHg)	94	87	69	70	80
PAWP (mmHg)	11	9	9	11	9
Qp (L/min/m^2)	4.11	5.11	5.54	8.53	2.86
Qs (L/min/m^2)	2.44	2.18	2.51	2.62	2.86
Qp/Qs	1.68	2.34	2.20	3.26	1
PVRi (WU m^2)	15.8	12.5	8.8	7.2	11.5
PVRi reduction rate (%)	—	-21	—	-18	—
SVRi (WU m^2)	39.3	37.2	25.1	23.3	26.6
Rp/Rs	0.40	0.33	0.35	0.31	0.43
Rp/Rs reduction rate (%)	—	-18	—	-11	—
SaO_2	97	99	92	99	98

mPAP, mean pulmonary artery pressure; mSAP, mean systemic arterial pressure; PAWP, pulmonary artery wedge pressure; Qp, body surface area (BSA) indexed pulmonary blood flow; Qs, BSA indexed systemic blood flow (cardiac index); PVRi, BSA indexed pulmonary vascular resistance; SVRi, BSA indexed systemic vascular resistance; WU, wood unit; SaO_2, arterial oxygen saturation.

VENTRICULAR SEPTAL DEFECT CLOSURE– *YES OR NO?*



Evolution

- Discussed with Pulmonary Hypertension Team
  - → Bosentan started in hospital and titrated to 125mg (1/2+1/2) with tolerance (but with low blood pressure profile)
  - → Furosemide 40mg (1 + ½)
  - → Spironolactone 25mg

- Re-evaluation in outpatients clinic

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Follow up: Outpatients clinic

- OMS class I-II
- BP 105/73mmHg HR 100bpm
- Acyanotic, SatO2 (No additional O2)=98%
- Pulmonary auscultation unremarkable
- No right heart failure signs

- Sildenafil 12.5mg (8/8h) was added to bosentan
  → Repeat the right heart catheterization after 3 months
    → Decision of possible VSD closure with fenestrated device

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Clinical case summary

- 19 year-old female patient from S. Tomé and Principe;
- Heart failure (NYHA III) after childbirth;
- Large ventricular septal defect, L→R shunt; LV dysfunction
- Precapillary PH. ↓ pulmonary vascular resistance > 20%

- **LV dysfunction cause** → volume overload; peripartum cardiomyopathy?
- **Treatment** → Trial with bosentan + sildenafil → repeat right catheterization→ possible closure with fenestrated device

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Thank you for your attention

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