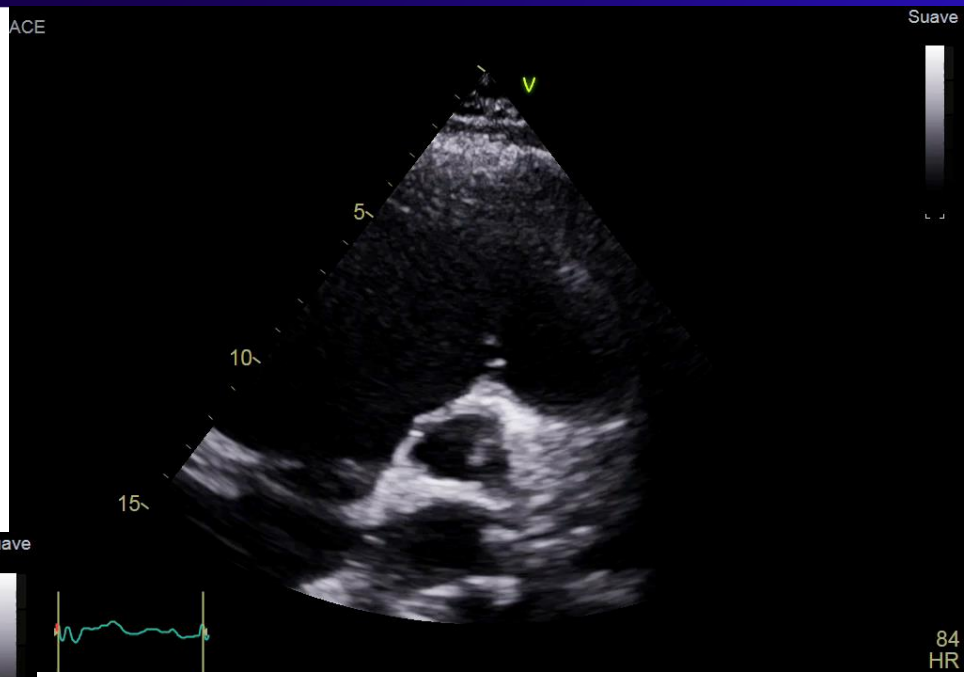
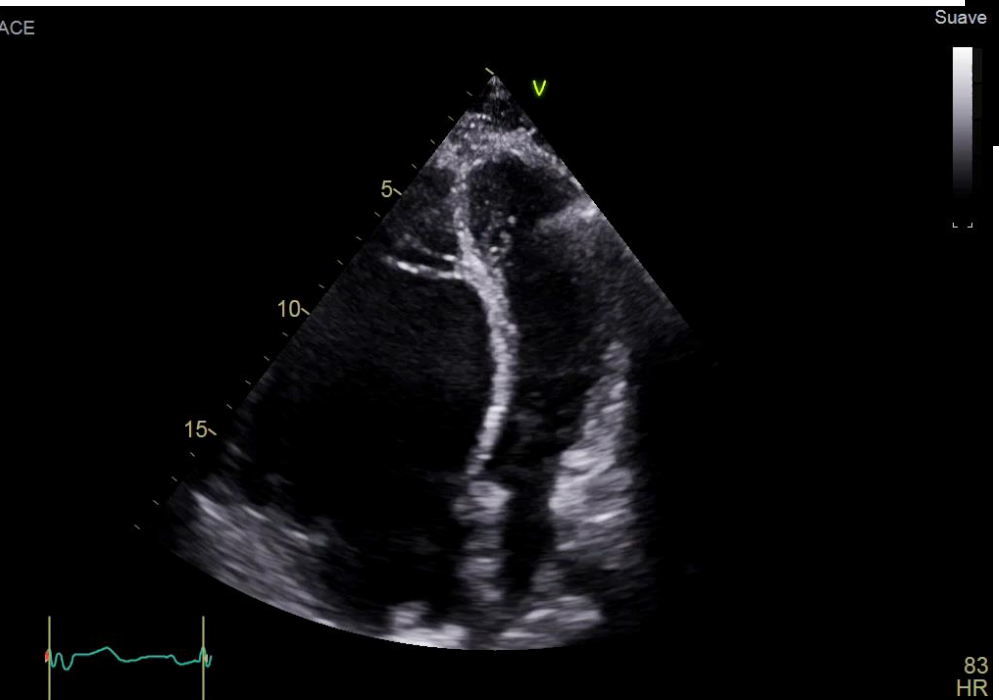
Right-sided ventricular failure due to Ebstein's anomaly

Case presentation

- 36 years-old male
- Surgically corrected atrial septum defect
- Ebstein's anomaly (Carpentier type D)





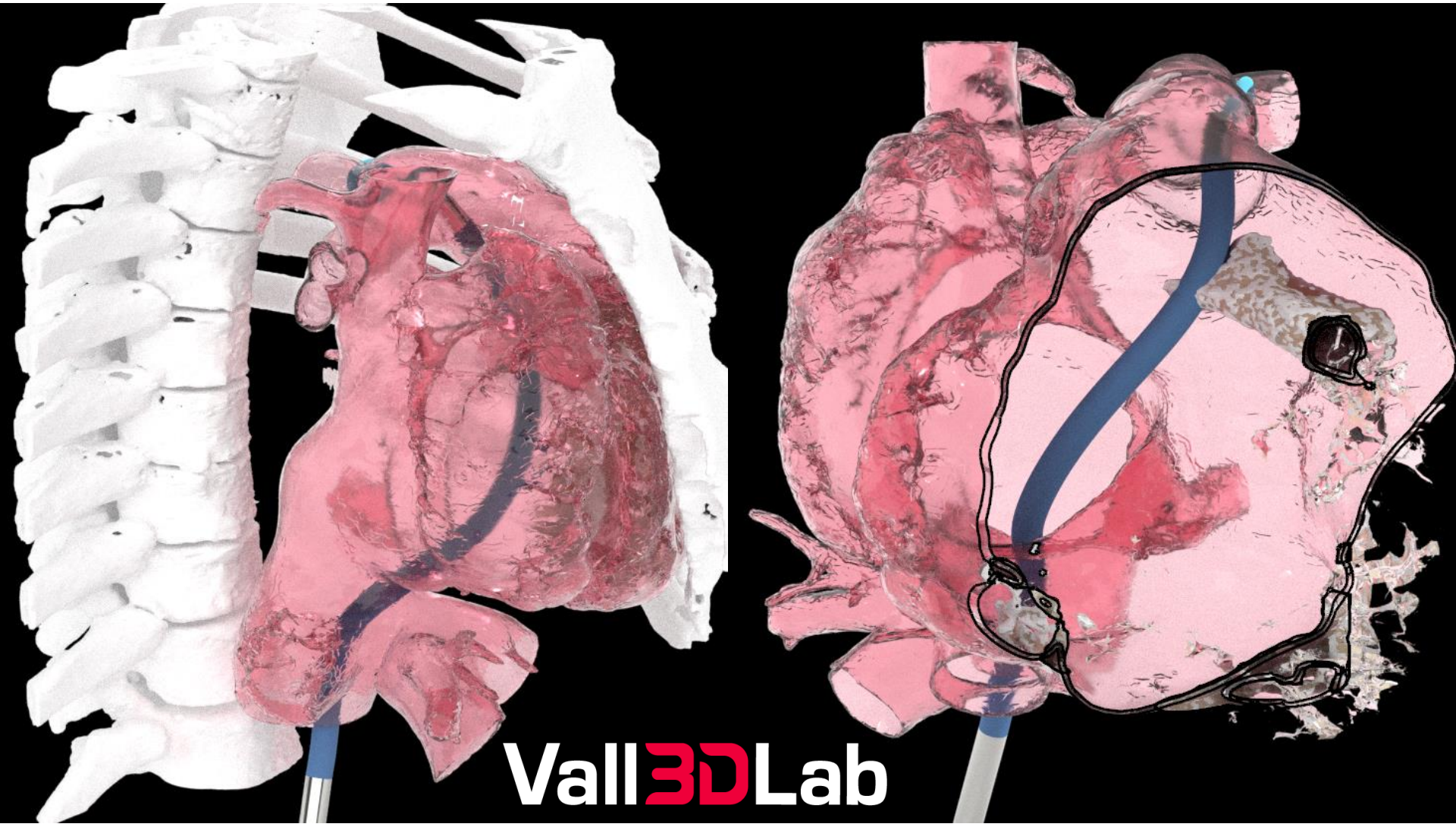
- Repetitive ambulatory levosimendan infusions for congestive right heart failure

Clinical course

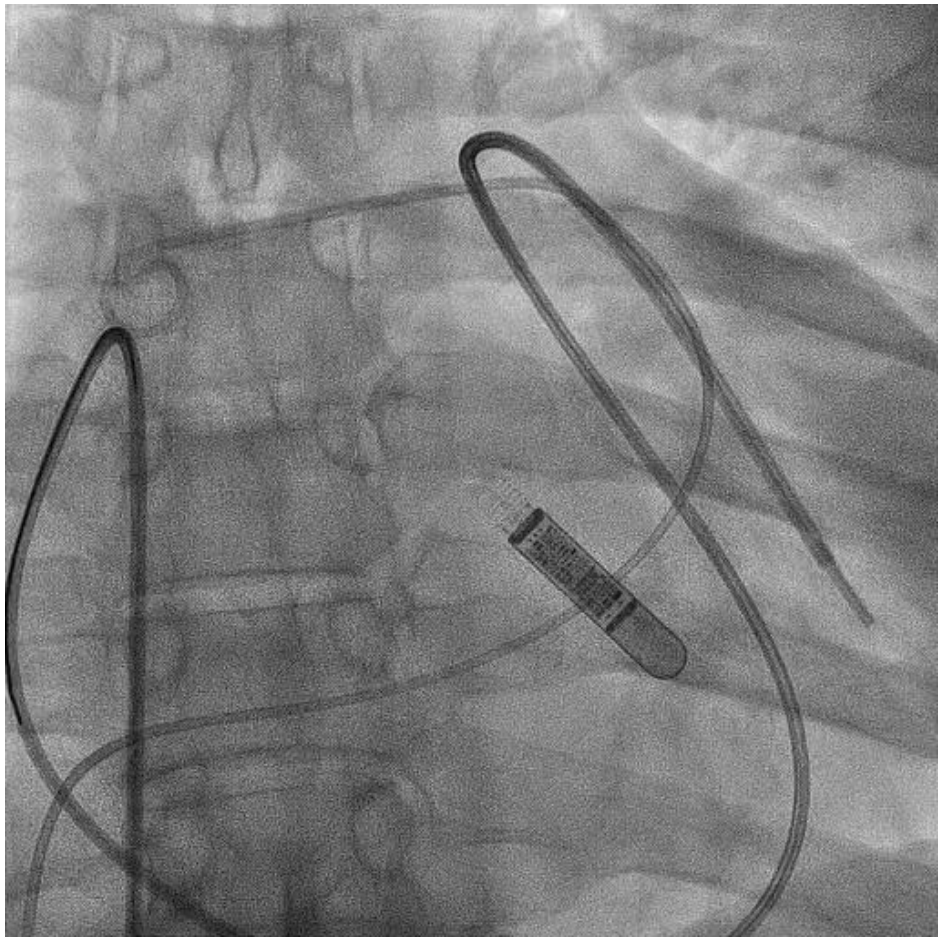
- January – April:
  - Circulatory driven syncope
  - Three re-admissions due to congestive right heart failure

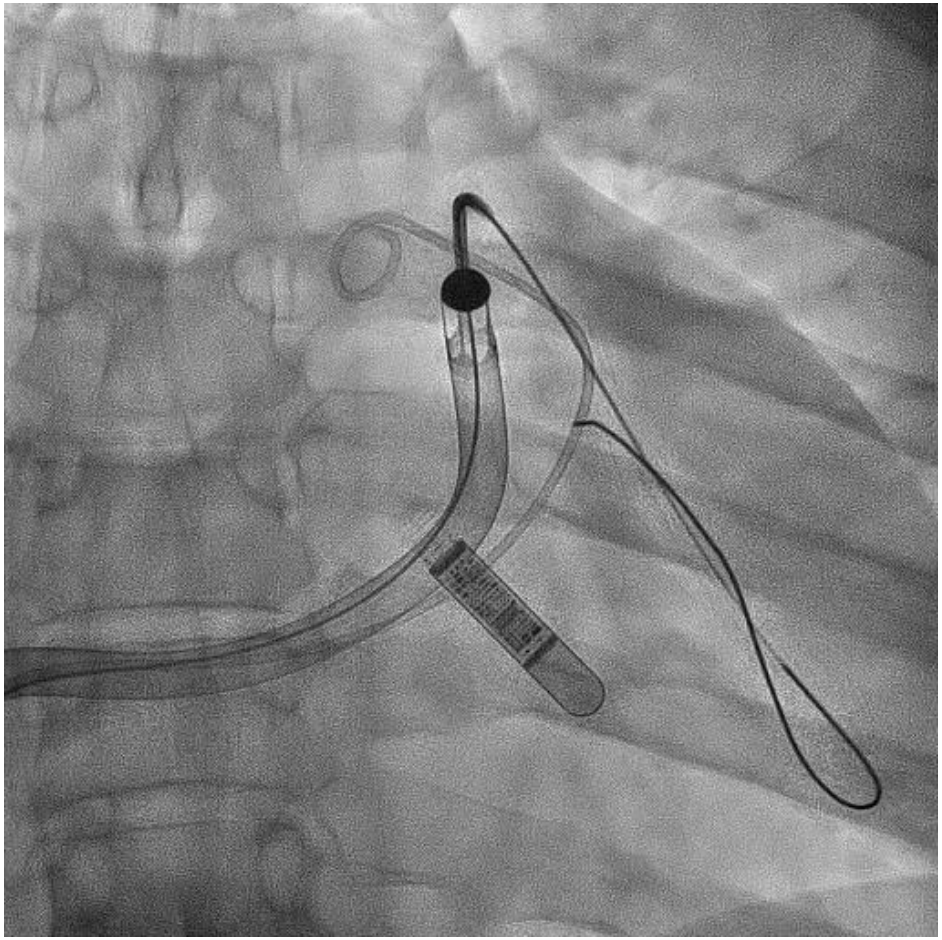
- June:
  - Admission to the ICU due to congestive right heart failure refractory to medical treatment in INTERMACS profile 2
  - Heart-team discussion:
    - Pre-trasplant study
    - **Percutaneous mechanical circulatory support**
    - Right catheterization: **postcapilary pulmonary hypertension** (median pulmonary artery pressure 24 mmHg, pulmonary wedge presure 17 mmHg, transpulmonary gradient 7 mmHg)

3-D reconstruction of RV for Impella RP implantation

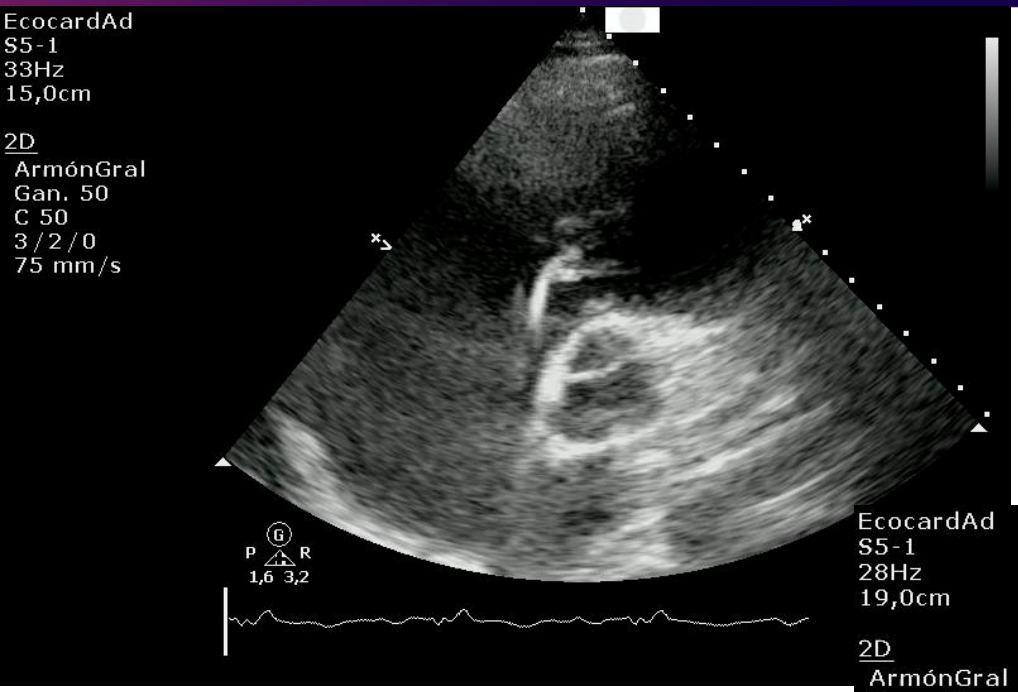


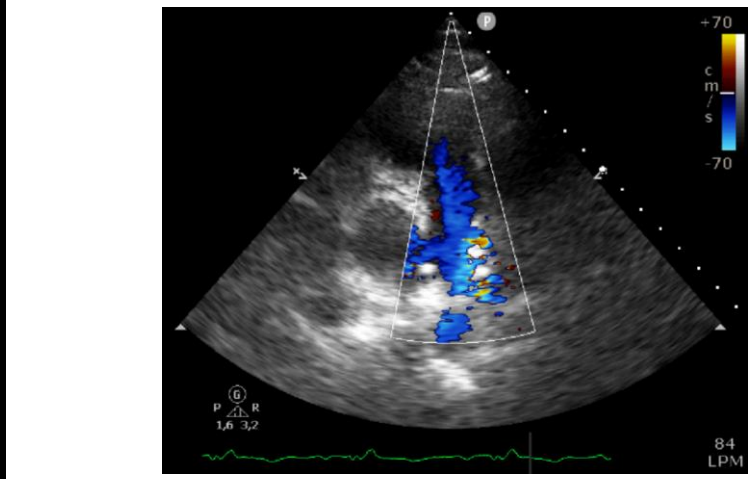
Fluoroscopic-guided Impella RP deployment

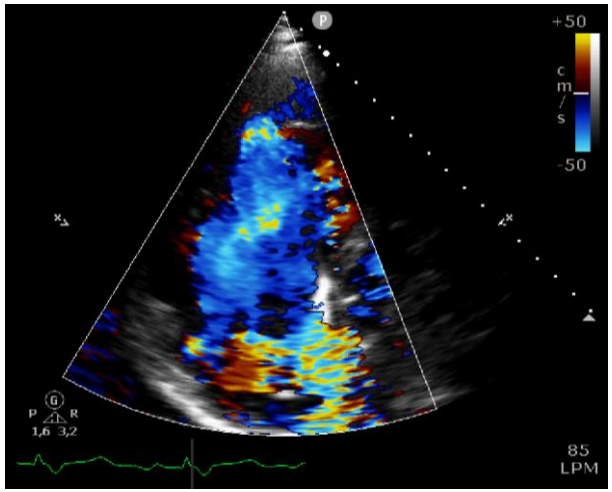


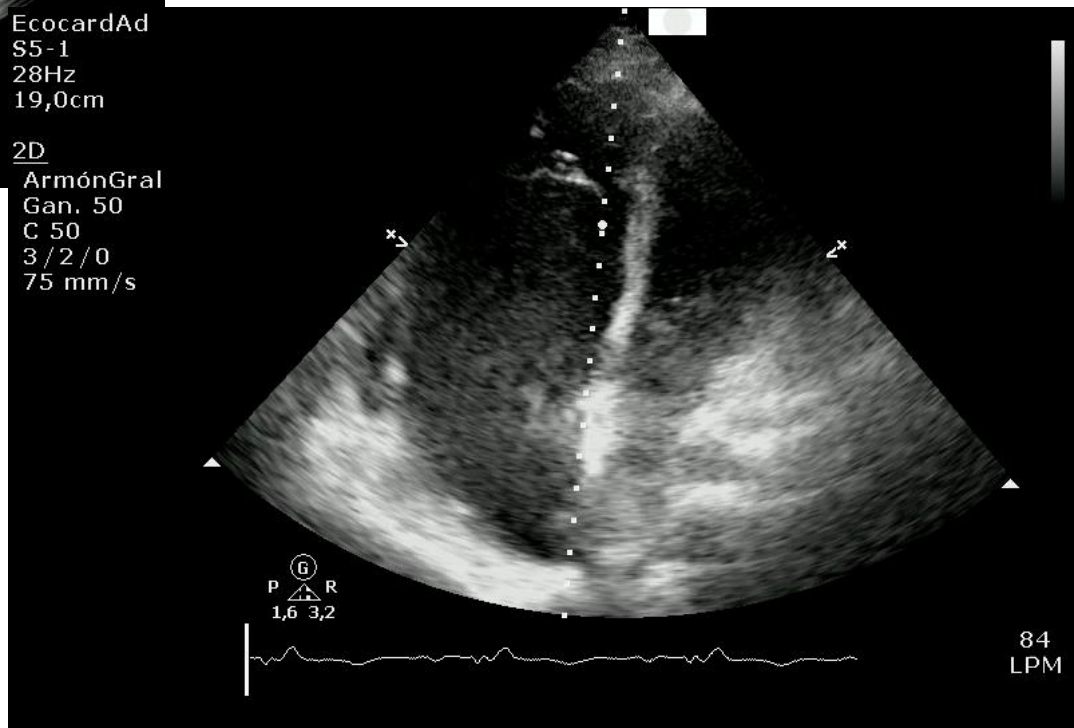


TTE after Impella RP implantation









Biochemical and hemodynamic measurements after 15 days of initiation of Impella RP support

	Day 0	Day 1	Day 5	Day 15
Creatinine (mg/dL)	1.4	1.2	0.8	0.6
Bilirrubine (mg/dL)	2.5	2.6	2	1.1
Central venous pressure (mmHg)	19	17	10	7
Cardiac index (L/min/m2)		1.7	2.9	3.4
Pump flow (L/min)		3	3	3

Improvement in hemodynamics, responsiveness to diuretics and laboratory values of end-organ function after Impella RP insertion.

Impella RP was set to motor level of P6 with approximately 3-3.2 L/min calculated flow

Outcome

- 15th day of support:
  - Drop off in the calculated flow and increase of energy consumption
  - Laboratory signs of hemolysis, with normalization of all parameters with volume loading and increase of anticoagulation
- 30th day of support:
  - Device displacement to right ventricle
  - Bed-side reposition failure → bed-side removal of device
  - Inclusion on the elective transplant waiting list

To the best of our knowledge, this is the first-in-the-world case report of Impella RP implantation in a patient with congenital heart disease and massive tricuspid regurgitation