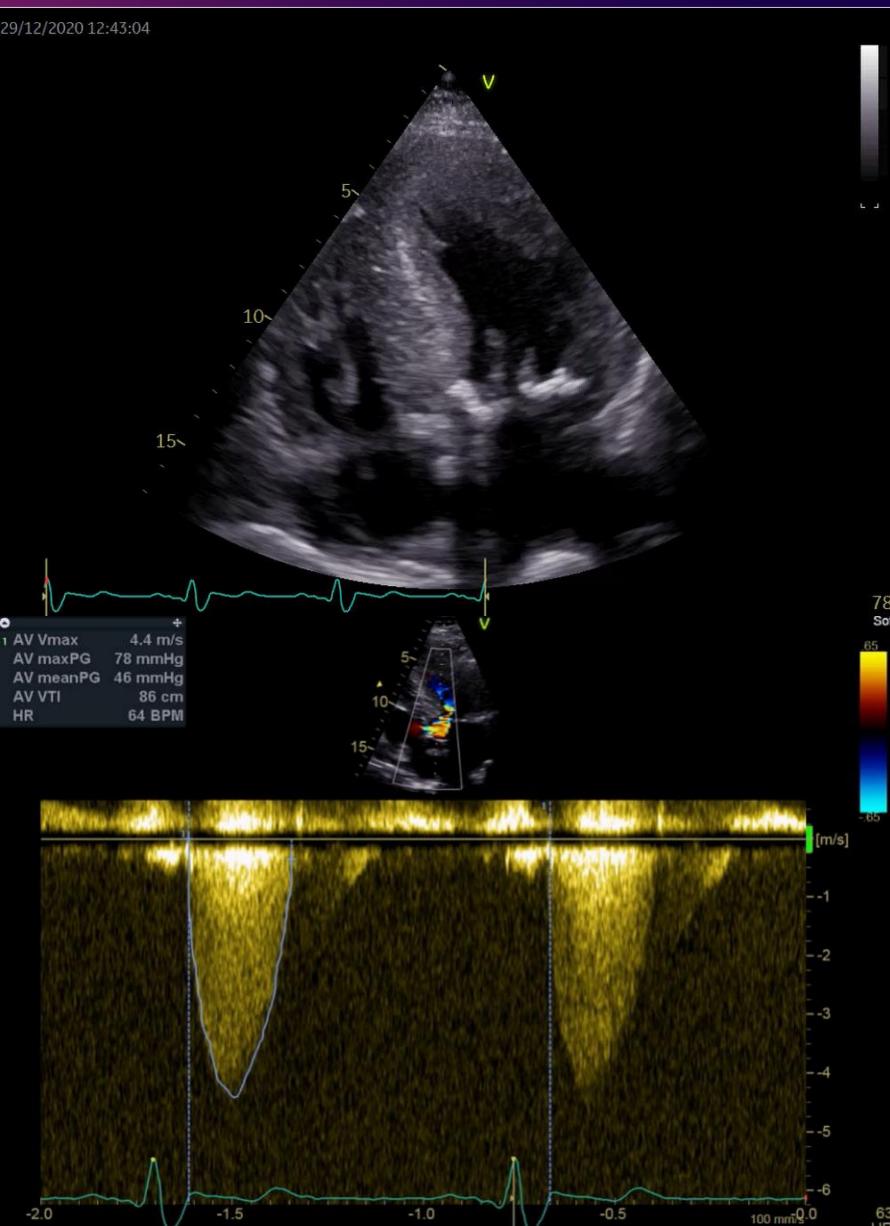




TAVI in extreme left ventricular hypertrophy (Fabry disease)

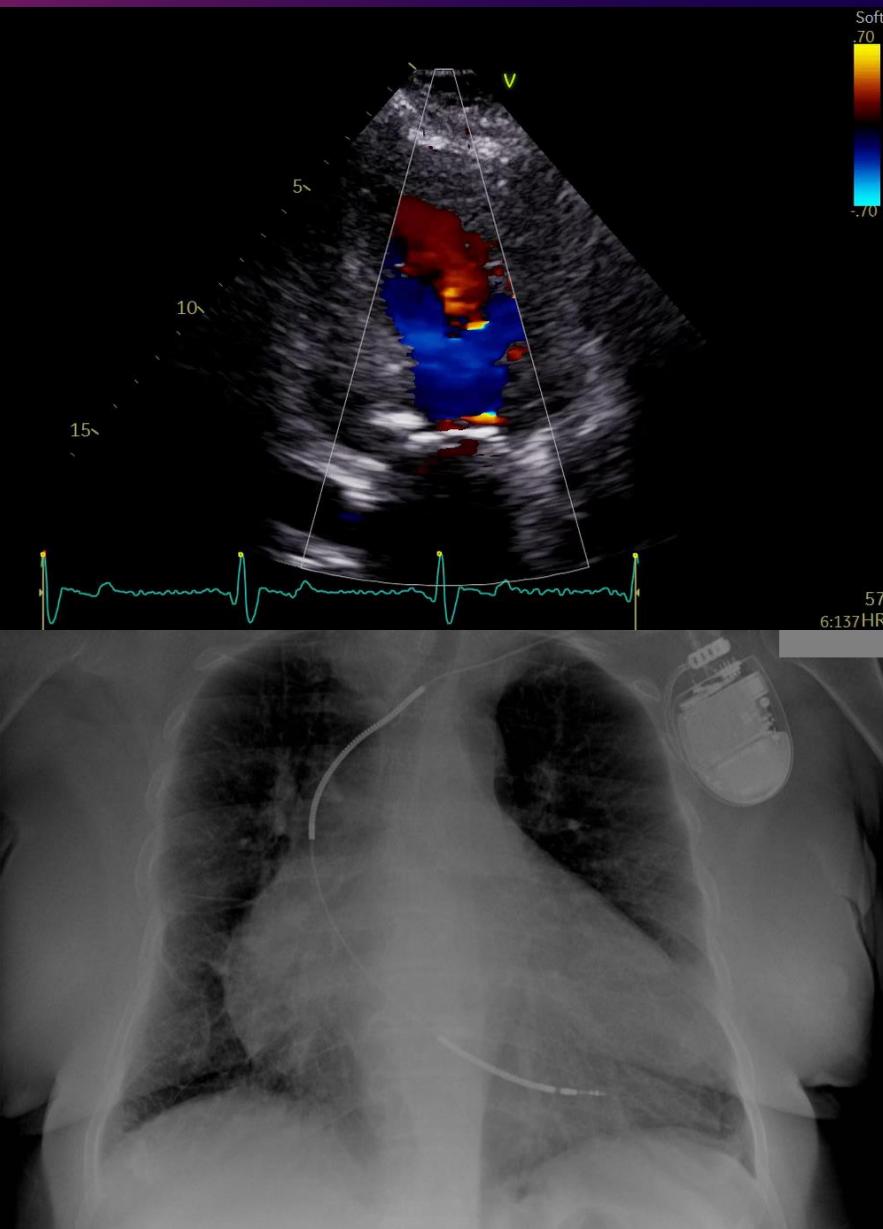
- 66-year old female with Fabry disease (treated since 2009).
- Admitted due to chest pain and exertional dyspnea.
- Ischemic heart disease (NSTEMI and PCI of the right coronary artery and first marginal branch in 2020), non-sustained ventricular tachycardia, and ICD implantation (2020), type 2 diabetes, arterial hypertension, depression, poor mobility after cerebrovascular insult, right bundle branch block.
- Declined for surgery.

Transthoracic echocardiography

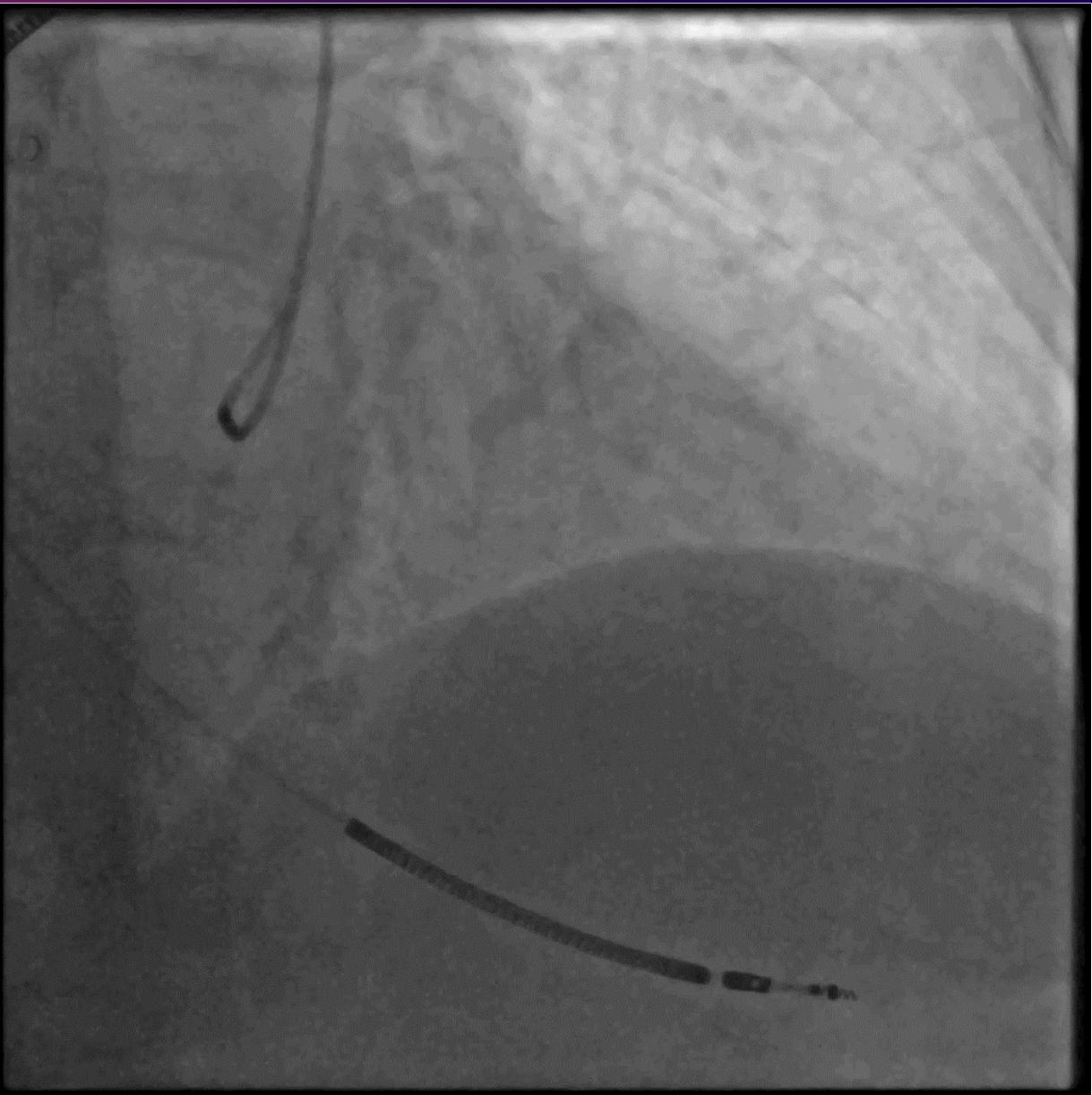


- Septum: 2,6 cm, inferolateral wall: 2,0 cm.
- Ejection fraction 57 %.
- Mean aortic gradient 46 mm Hg.
- Systolic pulmonary pressure: 35 mm Hg.

Transthoracic echocardiography and chest x-ray



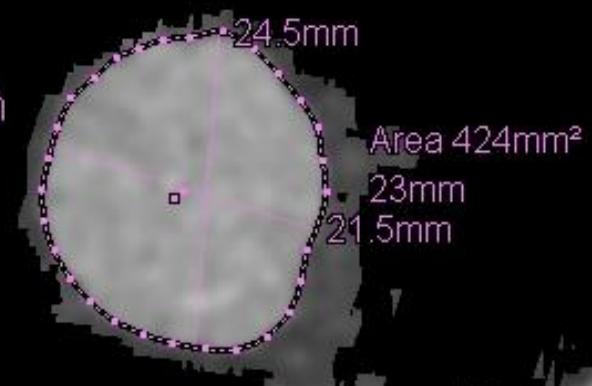
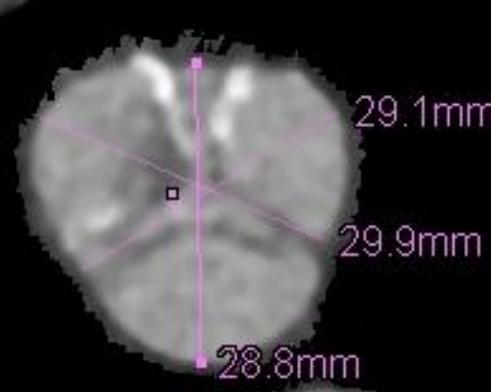
- Intracavitary gradient in the apical part of the left ventricle (40 mm Hg).
- Big heart – small cavities.

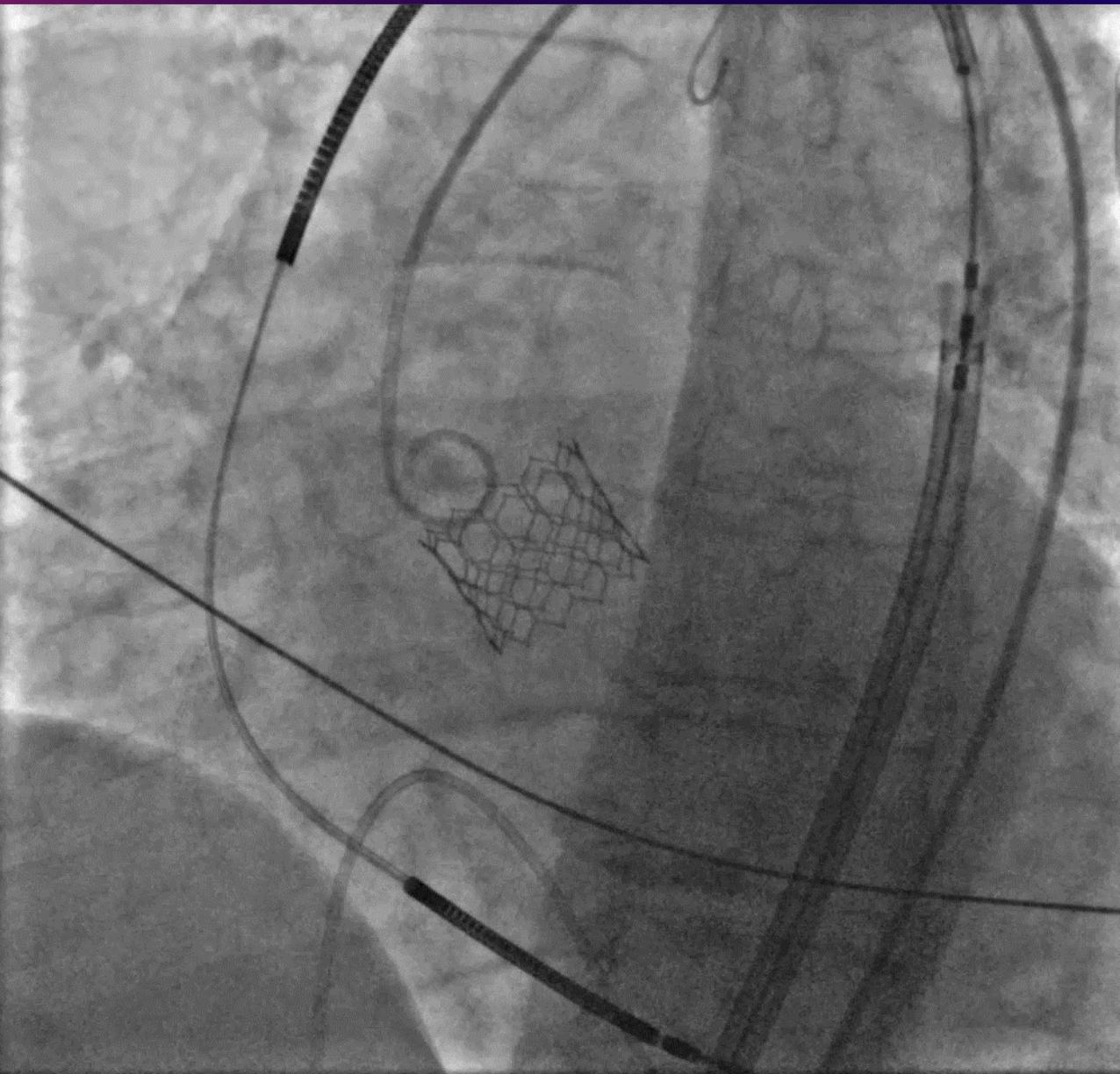


- Wide coronary arteries with diffuse stenosis of distal left anterior descending coronary artery.



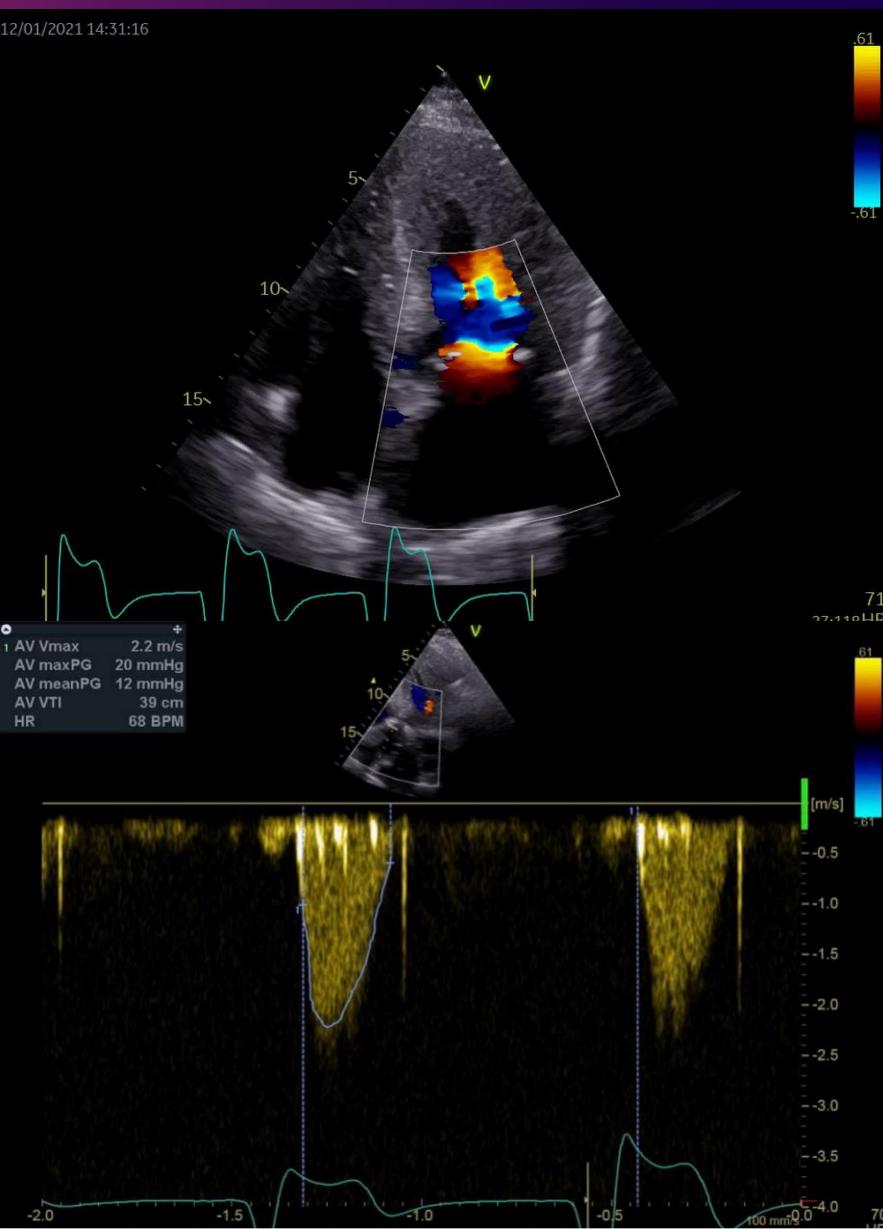
- Extreme left-ventricular hypertrophy with a small volume.
- Narrow, cone-shaped outflow tract.





- Good position of the Sapien 3 23 mm prosthesis, not protruding much into the left ventricular outflow tract.
- Minimal paravalvular regurgitation.

Transthoracic echocardiography post TAVI



- Minimal paravalvular regurgitation.

- Mean aortic gradient
12 mm Hg.

- Complete atrioventricular block immediately after balloon inflation.
- Transitory ischemic attack and a cerebrovascular insult (posterior cerebral artery) with transitory loss of sight on days 1 and 2 after TAVI.
- Good recovery in a few days, left hospital on day 7 after TAVI.

- TAVI in a patient with Fabry disease and severe left ventricular hypertrophy is feasible.
- Special attention should be given to the left ventricular outflow tract anatomy to avoid unwanted prosthesis migrations during implantation.
- The chance of complete heart block after TAVI is probably increased since patients with Fabry disease are prone to bradycardic heart rhythm disturbances.