

2016 ASE State of the Art Echocardiography Course | Tucson, AZ

Carcinoid

Sunday, February 14, 2016 | 7:00 – 7:15 PM | 15 min

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NYU
SCHOOL OF
MEDICINE



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Disclosures

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Speakers Bureau
Philips, Medtronic

CARCINOID TUMOR



Siegfried Oberndorfer
(1876 – 1944)
German pathologist

In 1907, Siegfried Oberndorfer coined the term *Karzinoide* (based on *καρκινώδης*, later anglicized as carcinoid)

Karzinoide Tumoren des Dünndarmes.
[Carcinoid tumors of the small intestine]
Frankfurter Zeitschrift für Pathologie
1907;1:426-429.

Carcinoid = ‘carcinoma like’

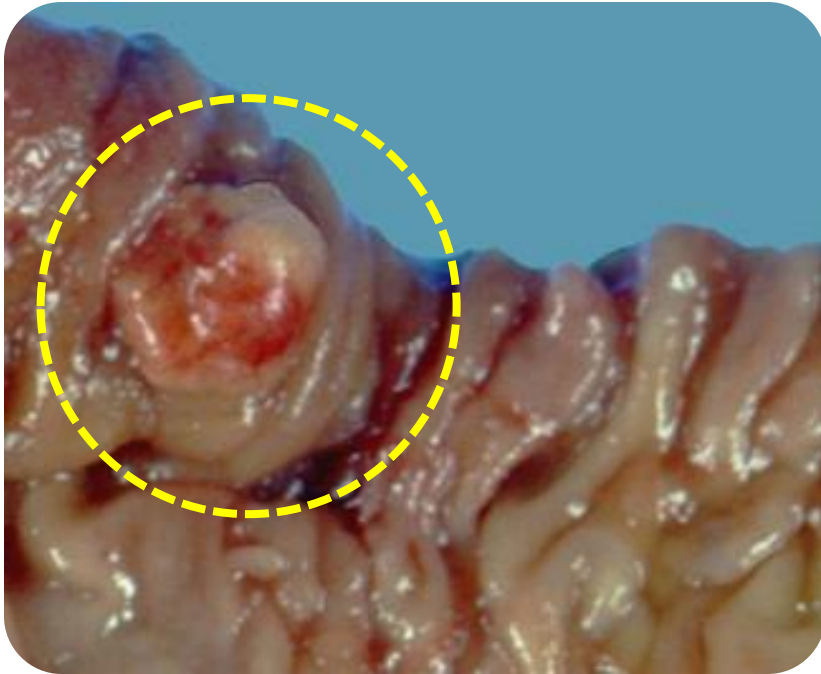
Histologically, a malignant tumor...

... but clinically often behaves like a benign nonmetastatic tumor

CARCINOID TUMOR VS. CARCINOID HEART DISEASE

What's the difference?

CARCINOID TUMOR



Carcinoid of the small intestine

- **Malignant tumor** of neuroendocrine cells in the gut or bronchial tree
- **Slow growing** and occasionally metastatic
- Secretes **vasoactive substances & hormones** including serotonin (5-HT) resulting in **carcinoid syndrome**:
 - Facial flushing
 - Diarrhea
 - Carcinoid heart disease

What portion of the heart gets involved depends on the primary tumor's location:

- **Intestinal** carcinoid >> Preferential **RIGHT** heart involvement
- **Bronchial** carcinoid >> Preferential **LEFT** heart involvement

CARCINOID HEART DISEASE

Carcinoid heart disease is **NOT** a cardiac malignancy.



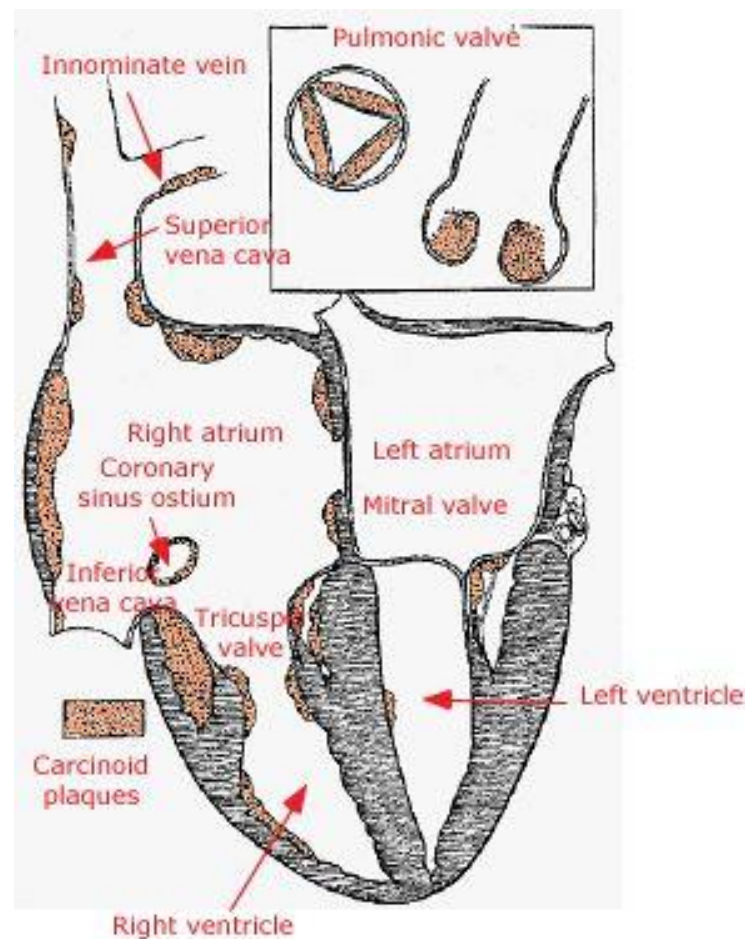
Carcinoid heart disease is a **paraneoplastic syndrome** caused by tumor-derived vasoactive substances.

Paraneoplastic manifestation of a carcinoid tumor

- Mural plaques
- **Valvulitis >> Stenosis + Regurgitation**

Because intestinal carcinoid are more common than bronchial ones, carcinoid disease involves primarily the **right heart**.

Carcinoid heart disease is often associated with **liver metastases** of an intestinal carcinoid.



CARCINOID HEART DISEASE

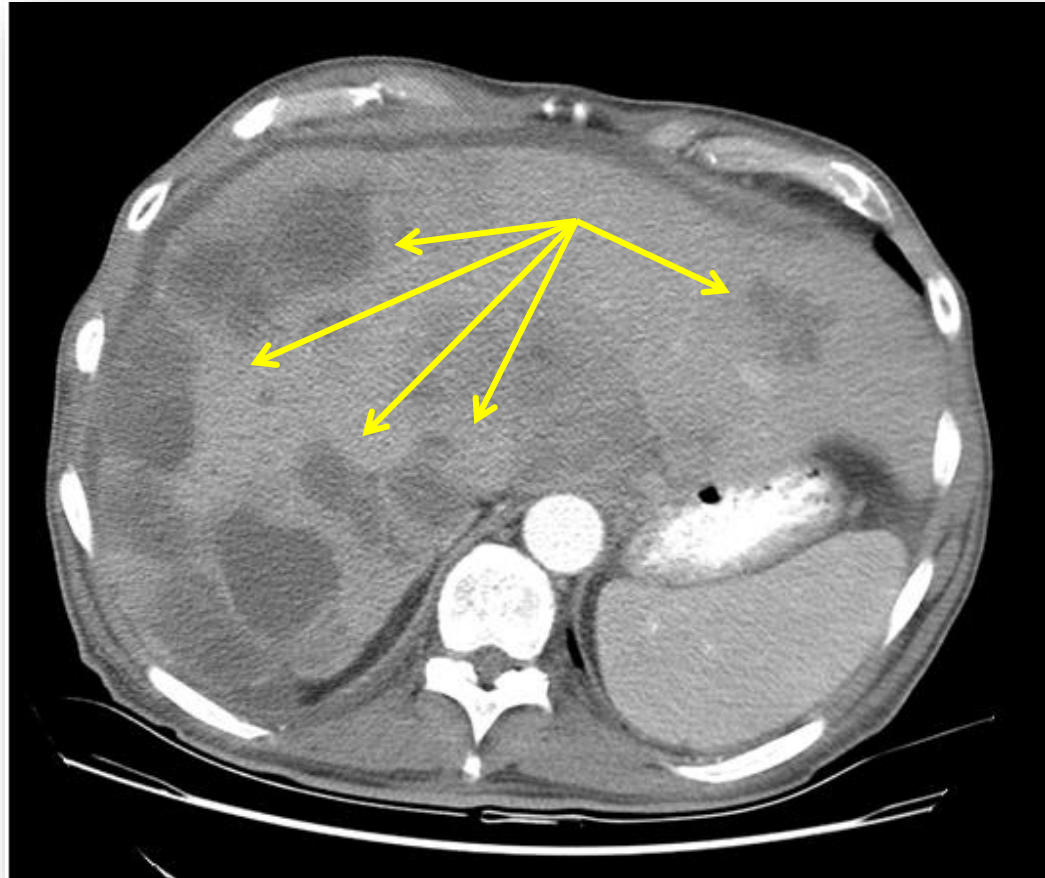
Case #1

CLINICAL PRESENTATION

56-year-old previously healthy man

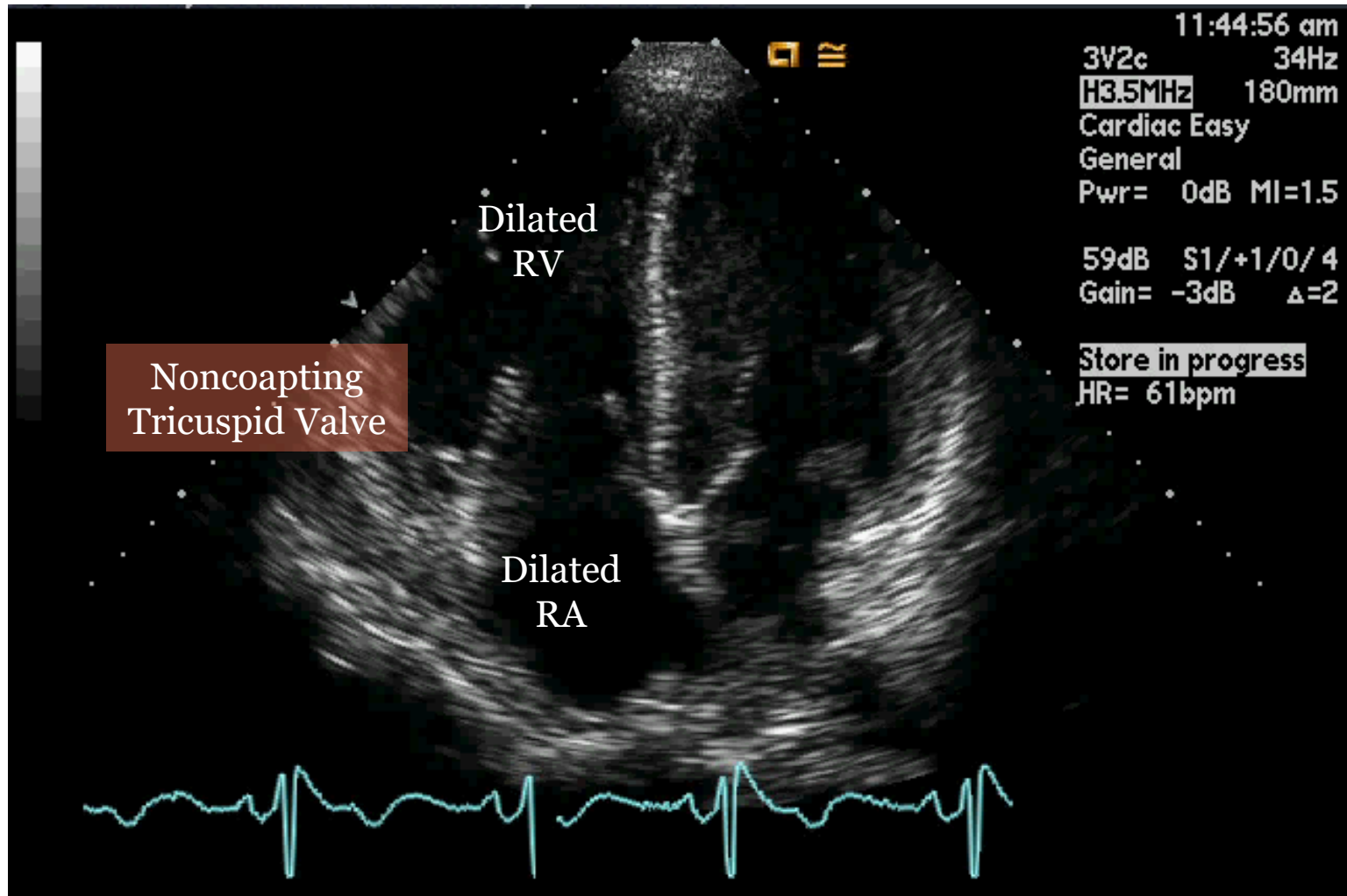
- Presented with right upper quadrant pain
- Referred for abdominal CT

ABDOMINAL CT

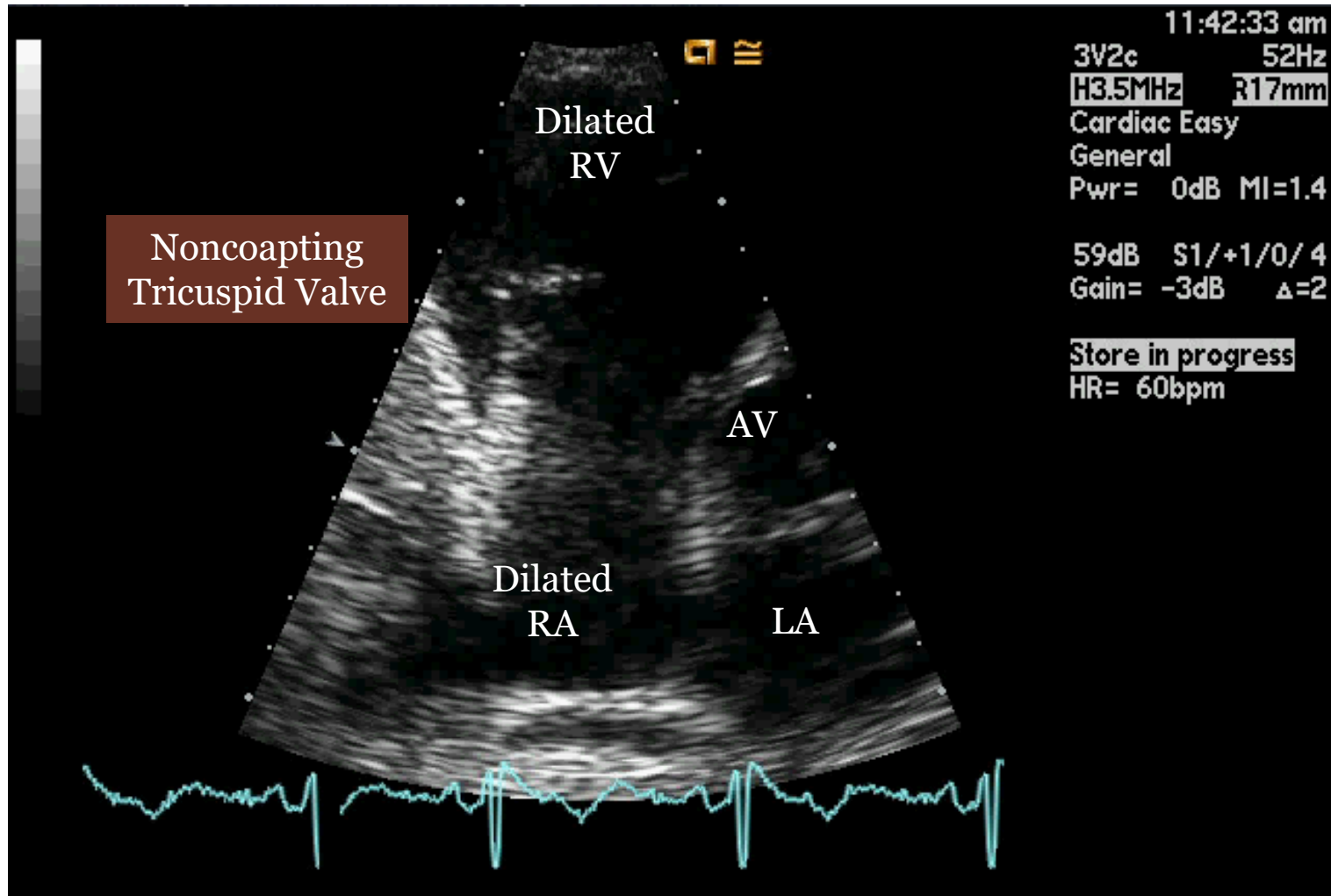


Multiple liver metastases of an unknown primary tumor

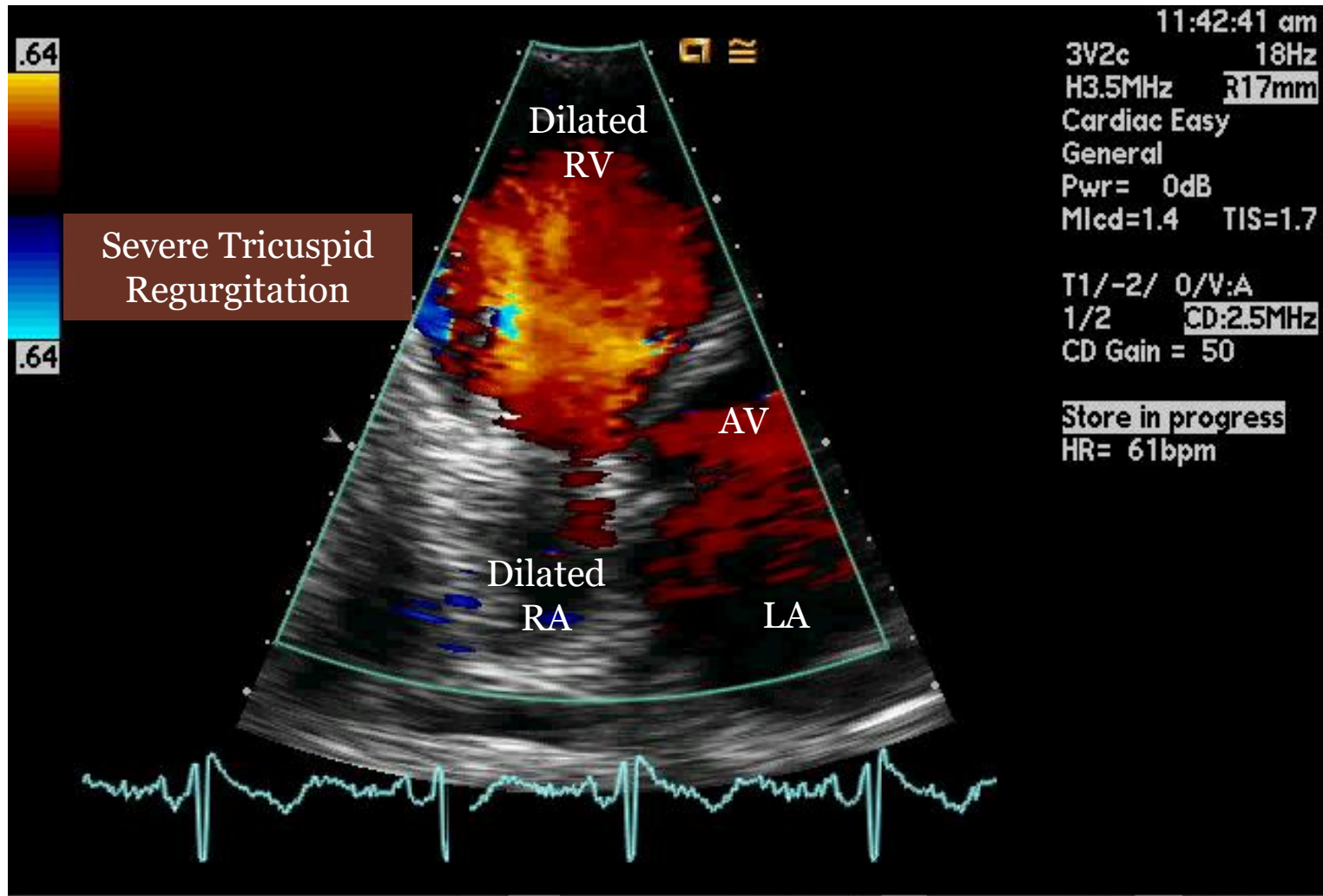
TRANSTHORACIC ECHOCARDIOGRAM | APICAL 4-CHAMBER VIEW



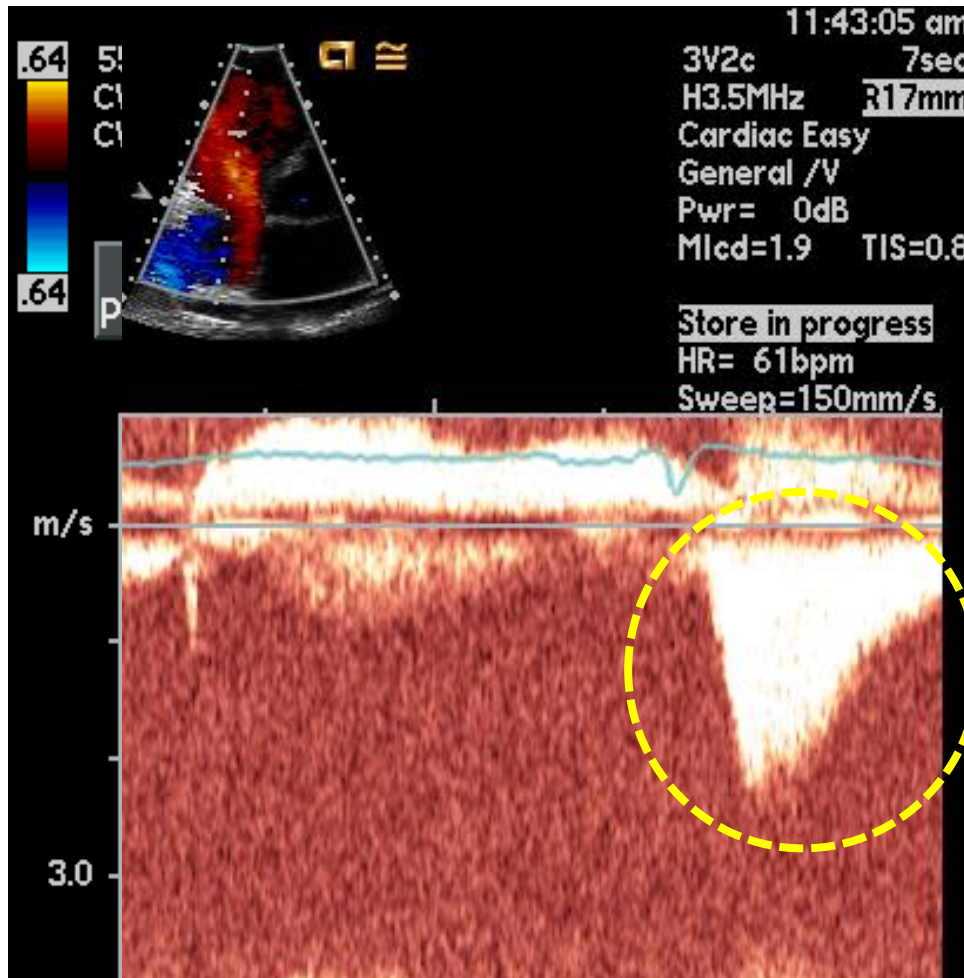
TRANSTHORACIC ECHOCARDIOGRAM | PARASTERNAL SHORT-AXIS VIEW



TRANSTHORACIC ECHOCARDIOGRAM | PARASTERNAL SHORT-AXIS VIEW

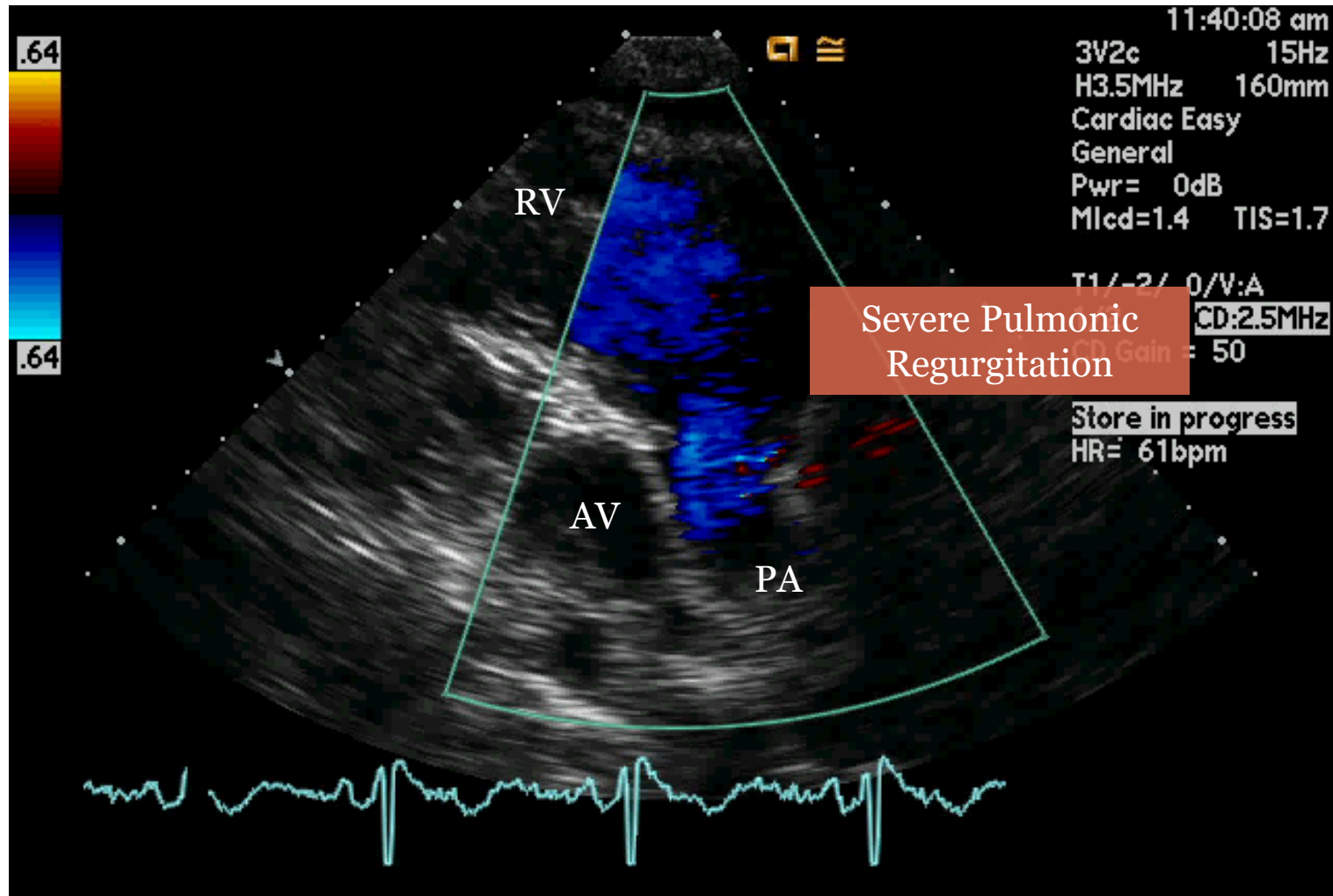


TRANSTHORACIC ECHOCARDIOGRAM | CW DOPPLER OF TRICUSPID VALVE

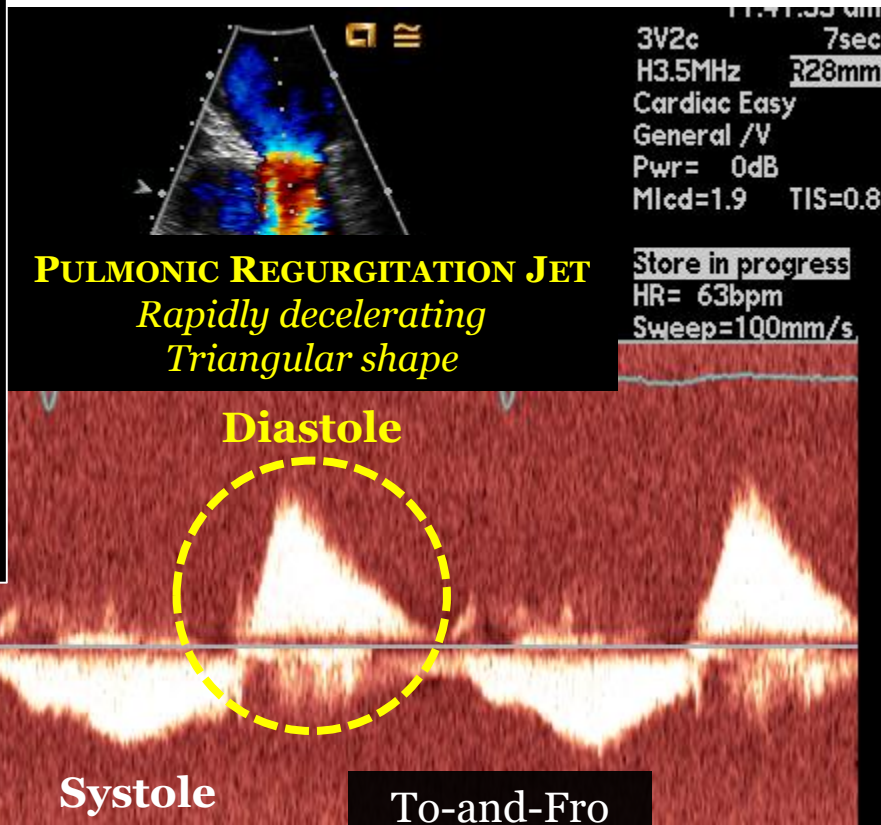
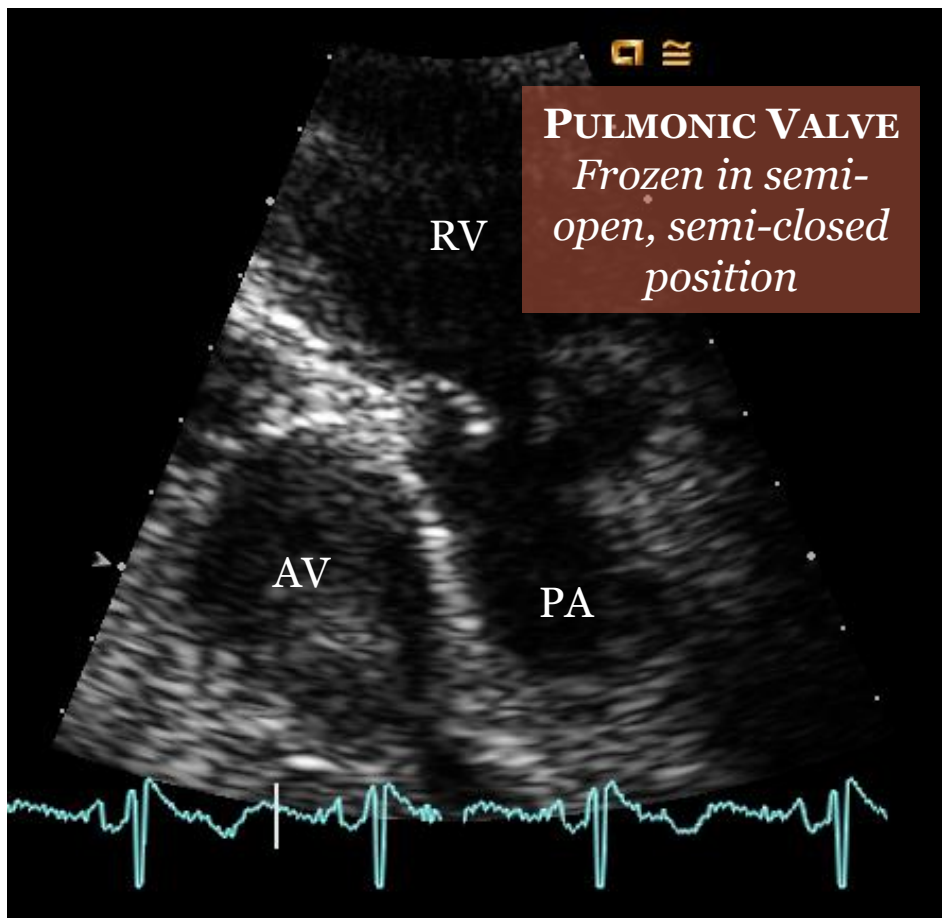


TR JET
Low velocity
Triangular shape

TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC VALVE



TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC VALVE



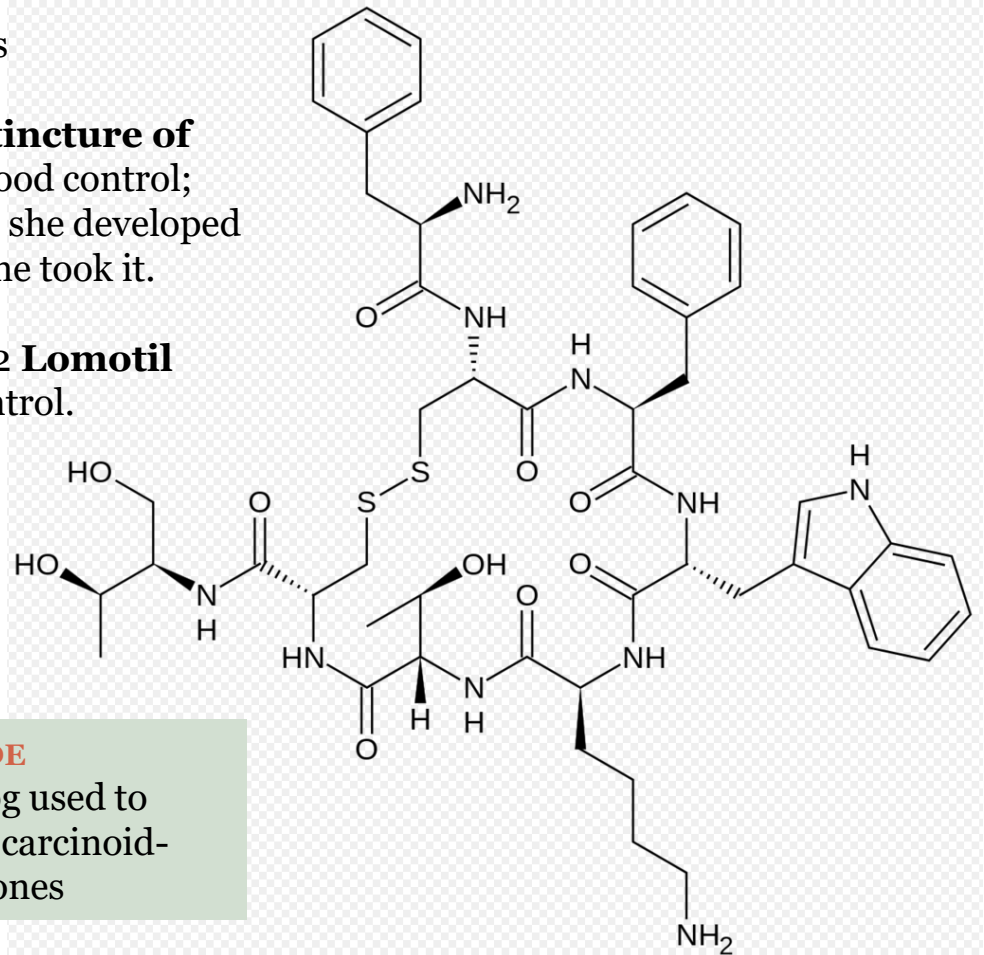
CARCINOID HEART DISEASE

Case #2

CLINICAL PRESENTATION

71-year-old female with 8-year history of metastatic carcinoid

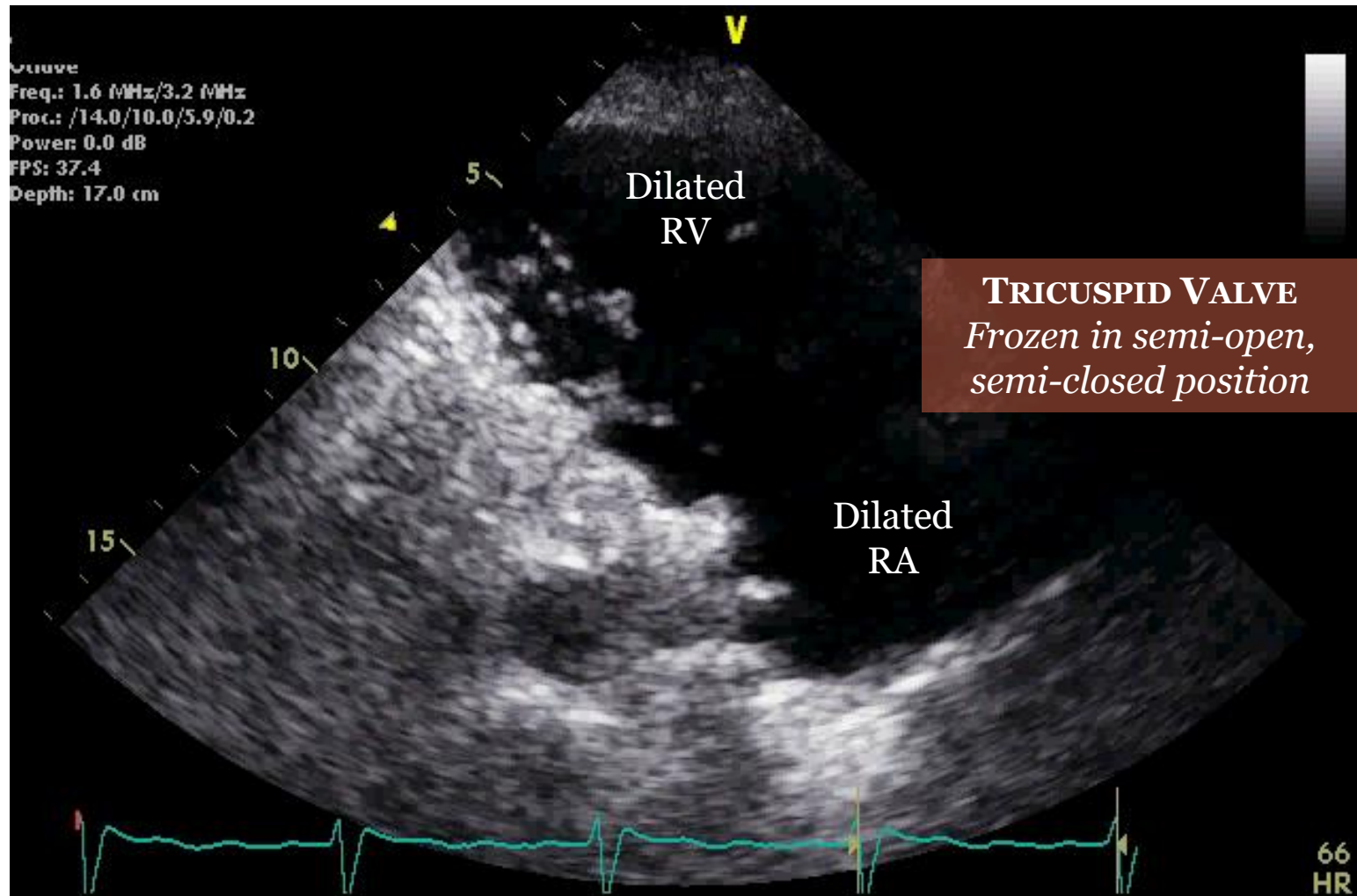
- Severe abdominal pain & increased diarrhea x 2 weeks
- On **somatostatin** analog for many years
- For the last 3 years, she has been taking **tincture of opium** for her diarrhea with relatively good control; however, she stopped 4 days ago because she developed severe right upper quadrant pain when she took it.
- Since then, she has been taking at least 12 **Lomotil** antidiarrheal pills with poor diarrhea control.



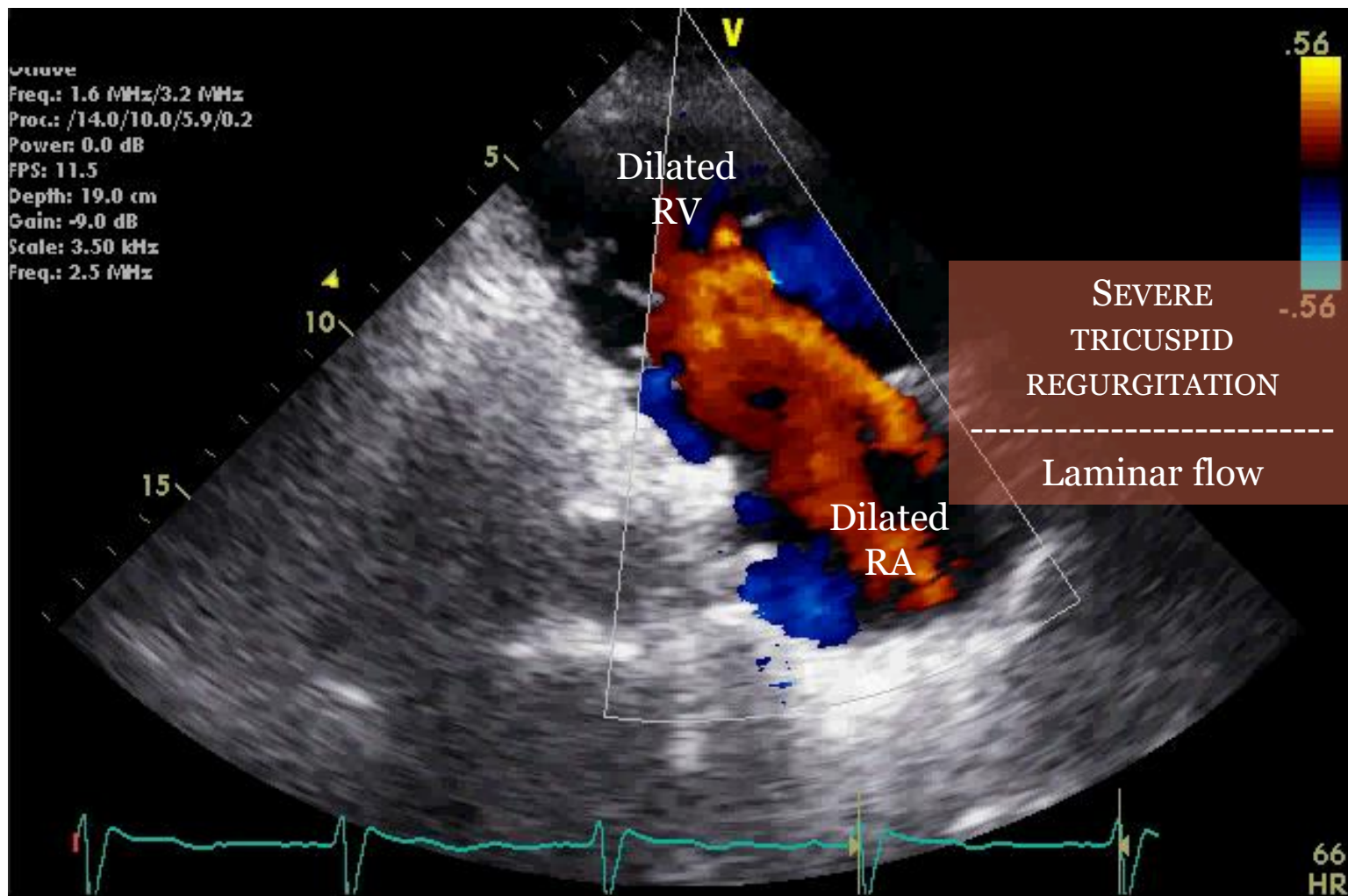
OCTREOTIDE

Somatostatin analog used to counteract effects of carcinoid-derived hormones

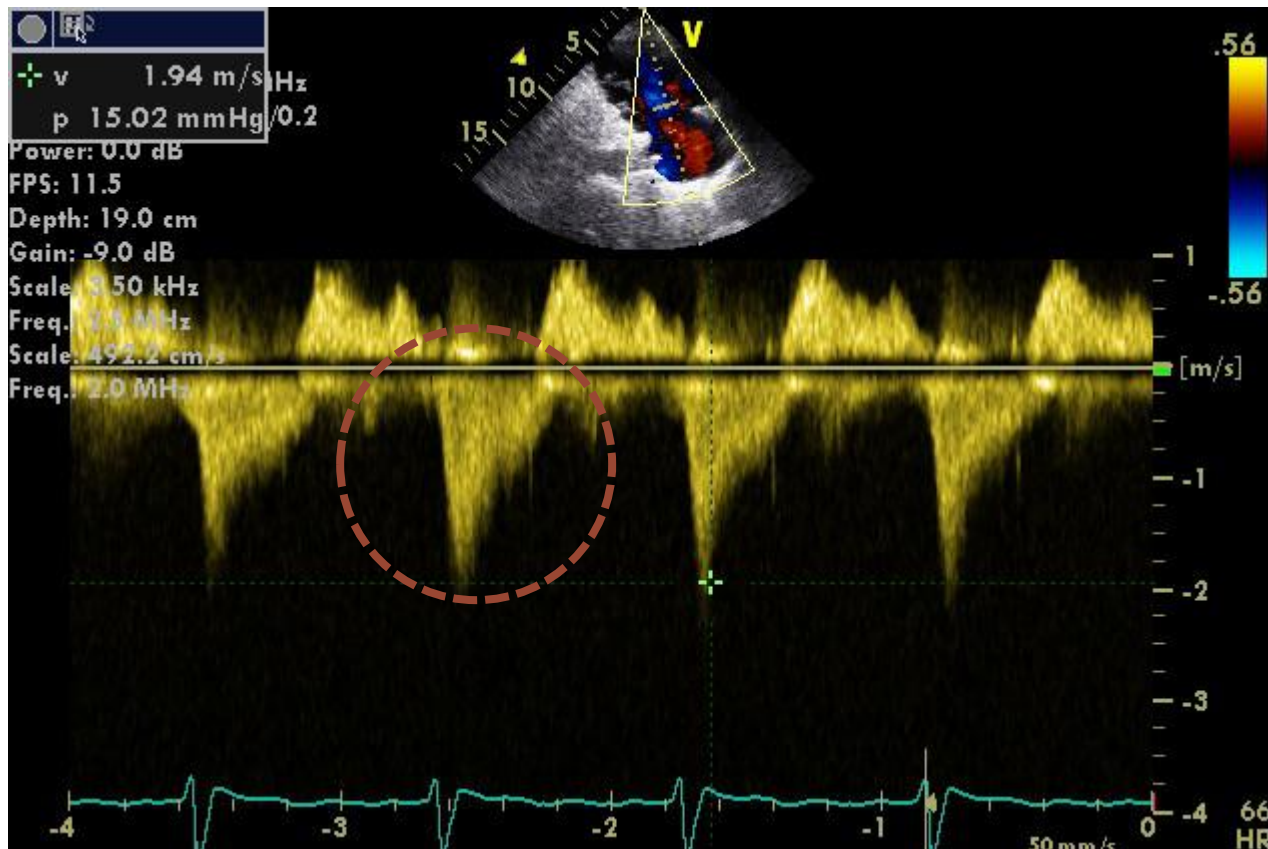
TRANSTHORACIC ECHOCARDIOGRAM | RV INFLOW VIEW



TRANSTHORACIC ECHOCARDIOGRAM | RV INFLOW VIEW



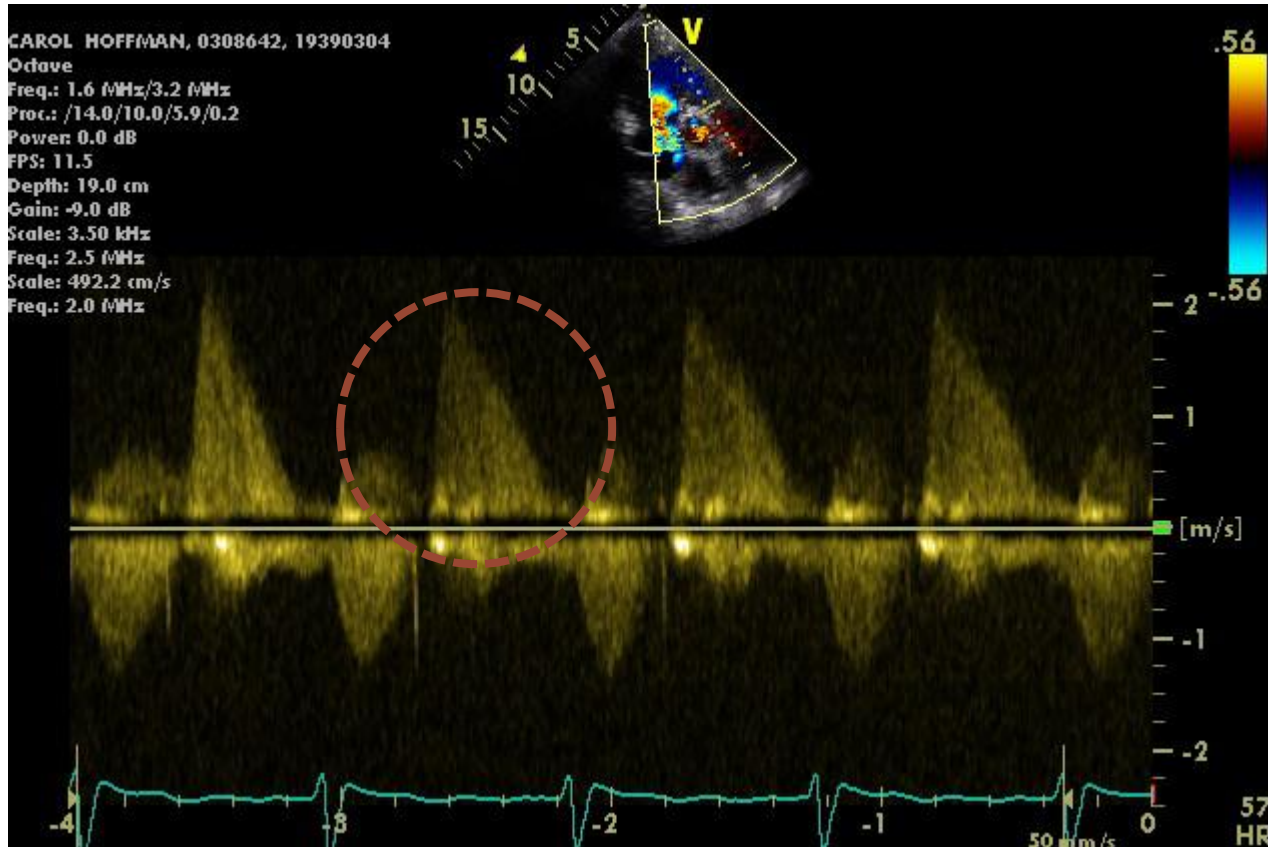
TRANSTHORACIC ECHOCARDIOGRAM | CW DOPPLER OF TRICUSPID VALVE



TRICUSPID REGURGITATION JET

Low velocity
Triangular shape

PULMONIC REGURGITATION JET *Rapidly decelerating*



CARCINOID HEART DISEASE

Case #3

CLINICAL PRESENTATION

57-year-old female with shortness of breath over the past 6 weeks.

- Previously unlimited exercise tolerance
- Progressive bilateral **leg edema**, **increasing abdominal girth** and **loose bowel movement**.
- Recently noticed intermittent **flushing** and **shivering** without fever



OCTREOTIDE ABDOMINAL CT SCAN

*Multiple foci of uptake within the liver and in a calcified mesenteric mass compatible with **octreotide avid tumors** such as **carcinoid**.*

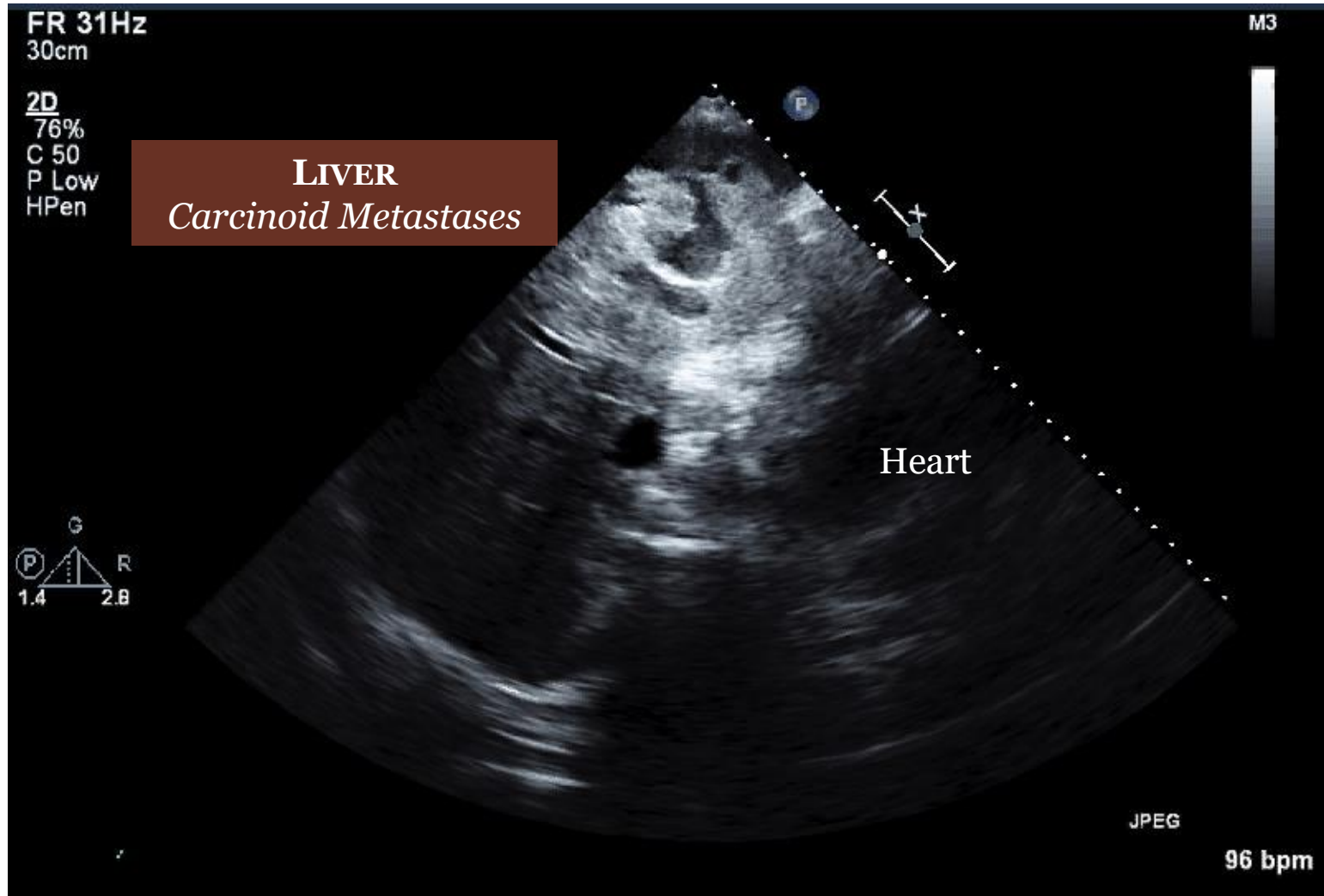
24-HOUR URINE TEST

Test Name	Value	Abnormal Flag	Range	Unit
Time (Hours)	24			
Total Volume	2450			
5-HIAA	79.2			mg/L
5-HIAA, mg/day	194	H	0-15	mg/d
5-HIAA Urine, mg/g CRT	189	H	0-14	mg/g
5HIAA Interpretation	See Note			
Urine Creatinine	42			mg/dL
Creatinine mg/day	1029		500-1400	mg/d

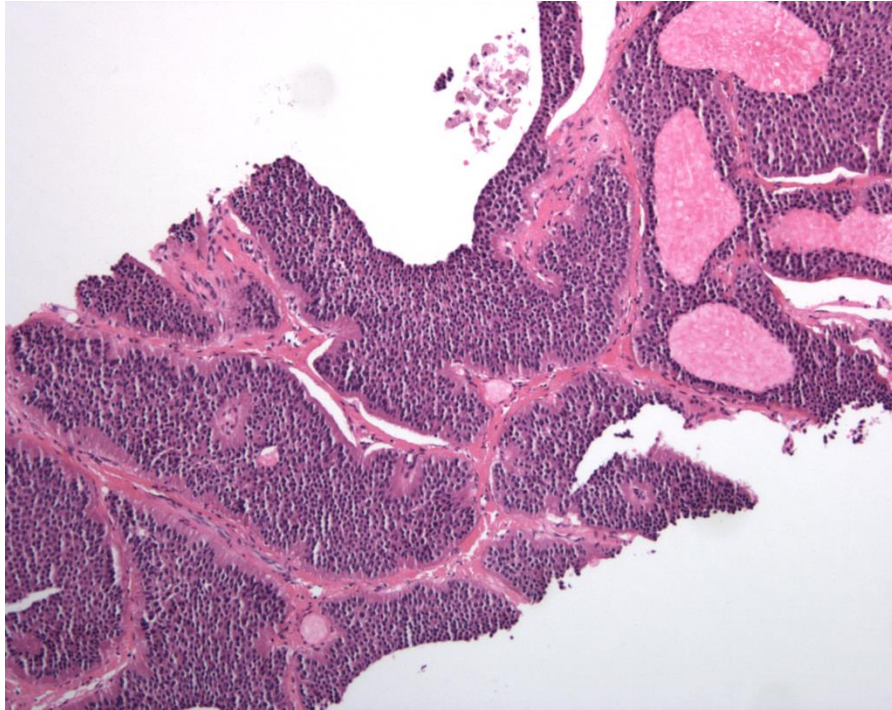
*Increased urinary excretion of serotonin metabolites, consistent with **carcinoid syndrome**.*

TRANSTHORACIC ECHOCARDIOGRAM

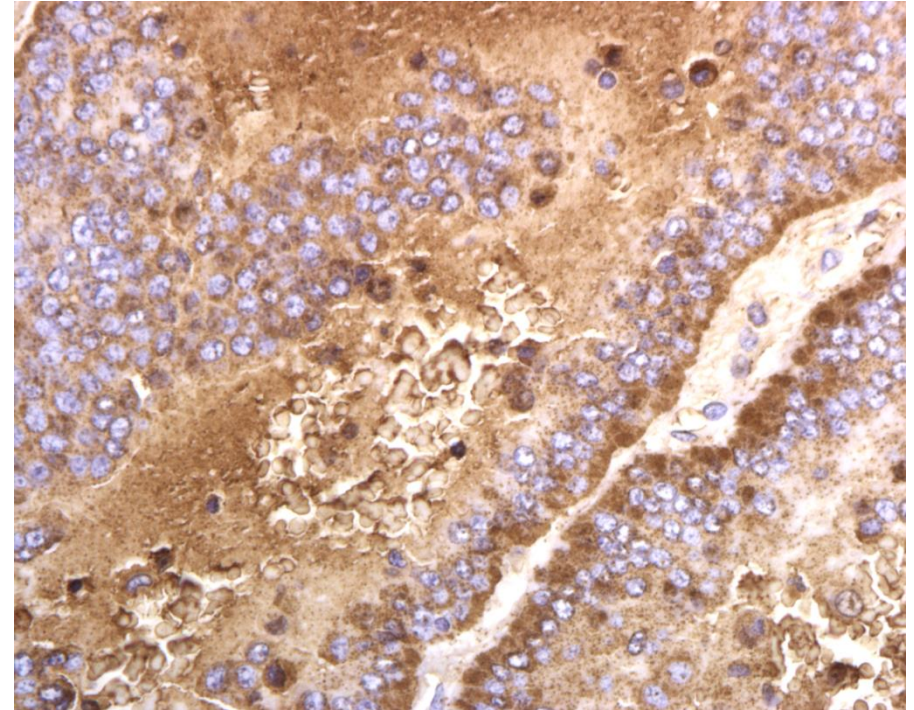
TTE showed **severe tricuspid** and **pulmonic regurgitation**... and something else in subcostal view



LIVER BIOPSY



CARCINOID TUMOR
Metastatic well differentiated
neuroendocrine tumor



CARCINOID TUMOR
Positive staining for **chromogranin**
consistent with carcinoid

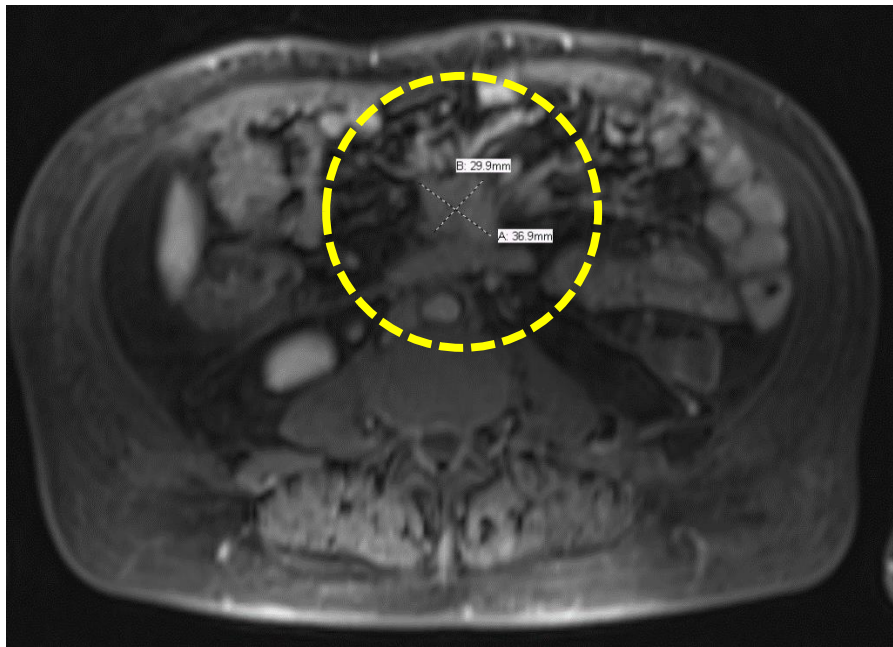
CARCINOID HEART DISEASE

Case #4

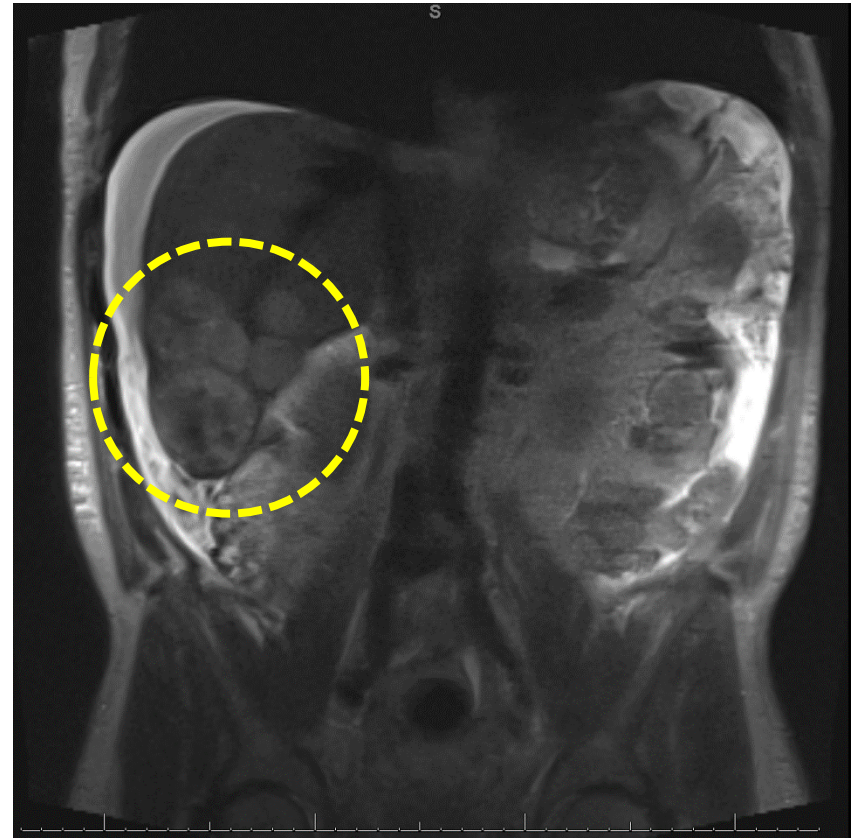
CLINICAL PRESENTATION

64-year-old men with recent weight loss and bilateral lower extremity edema

- A year ago, he noticed some flushing and rosy appearance to his cheeks
- Six months ago he started having diarrhea
- **Abdominal MRI**
 - Mesenteric soft tissue mass
 - Multiple hepatic metastases
- **Liver biopsy**
 - Carcinoid tumor

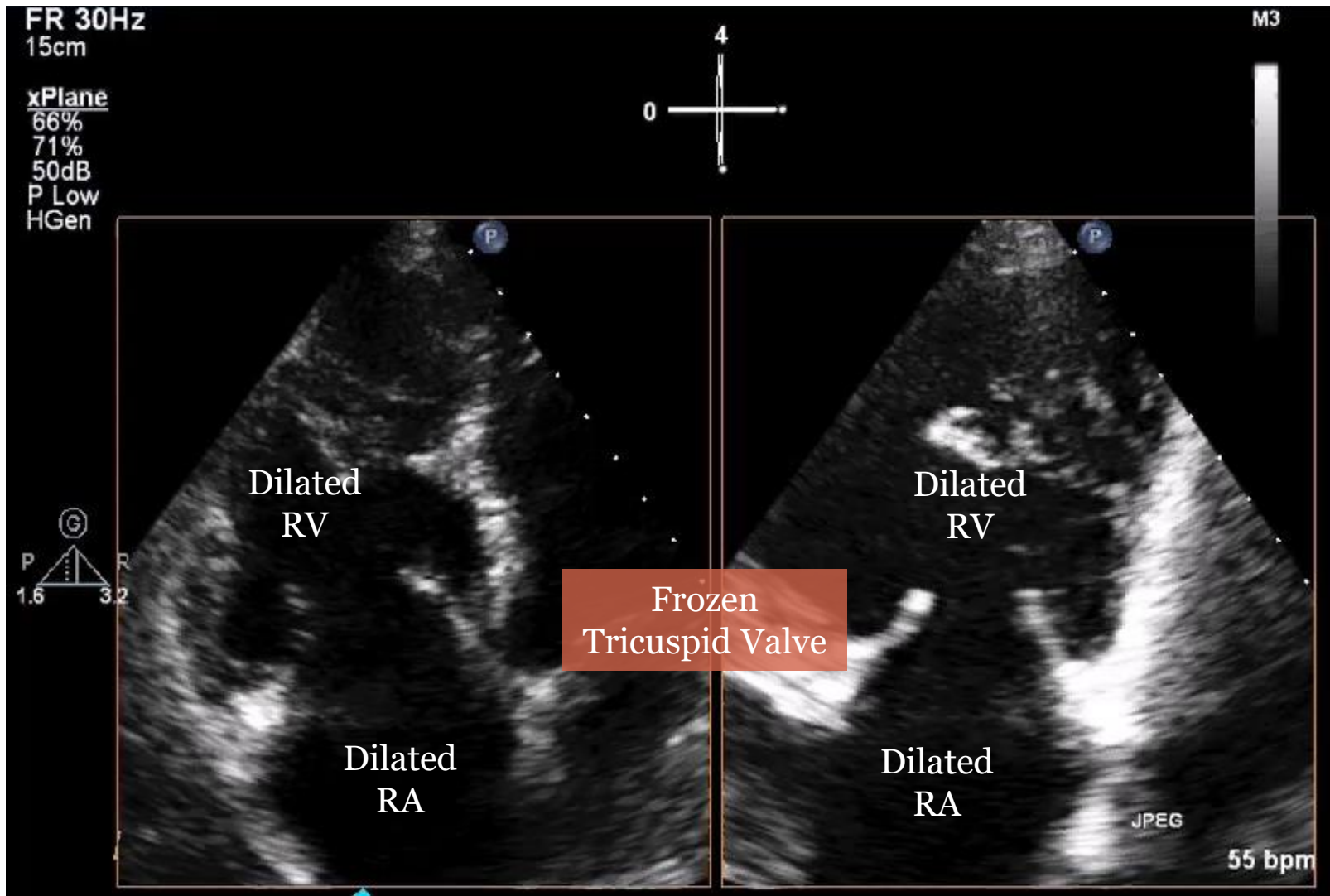


MRI | MESENTERIC CARCINOID

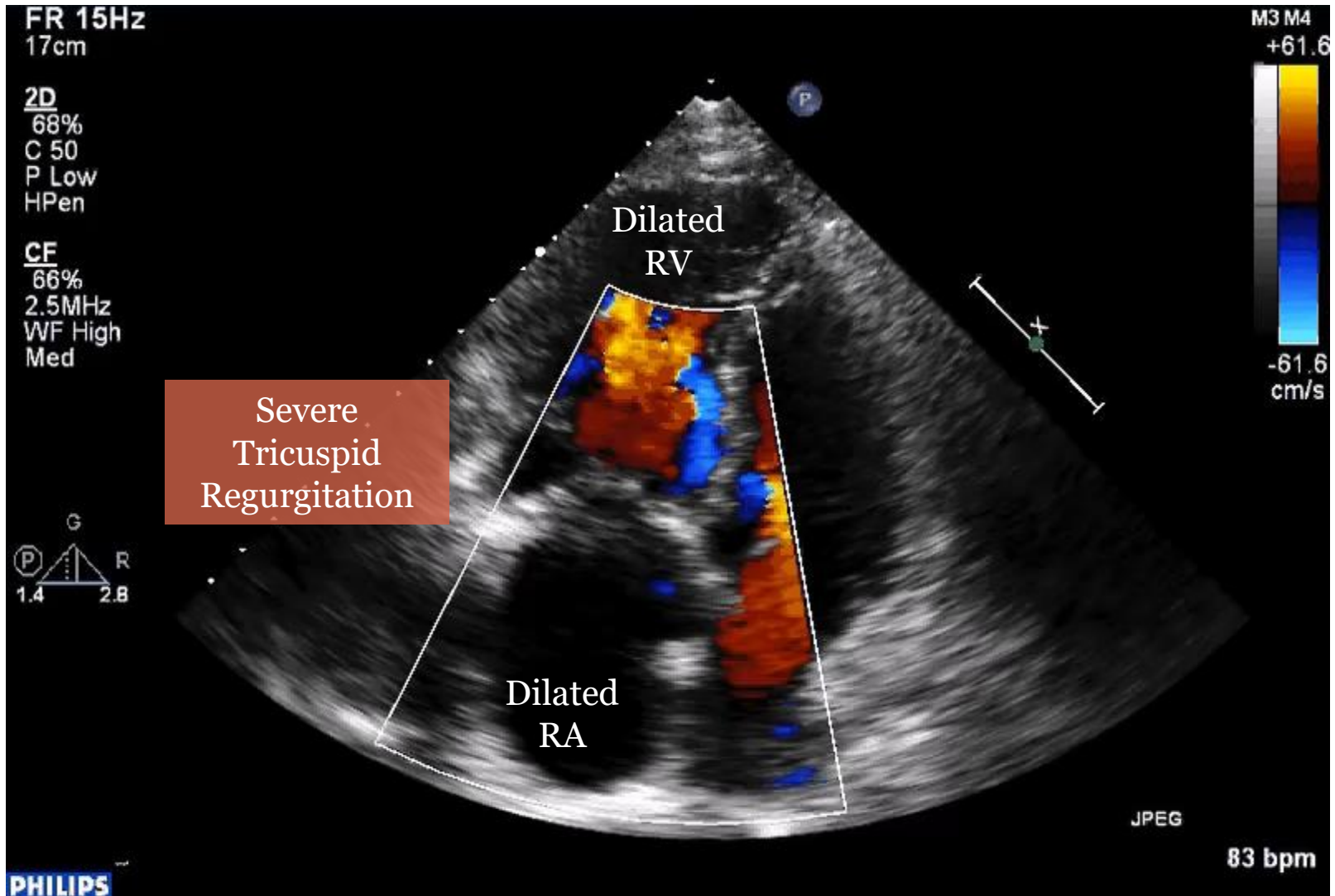


MRI | INNUMERABLE LIVER METASTASES

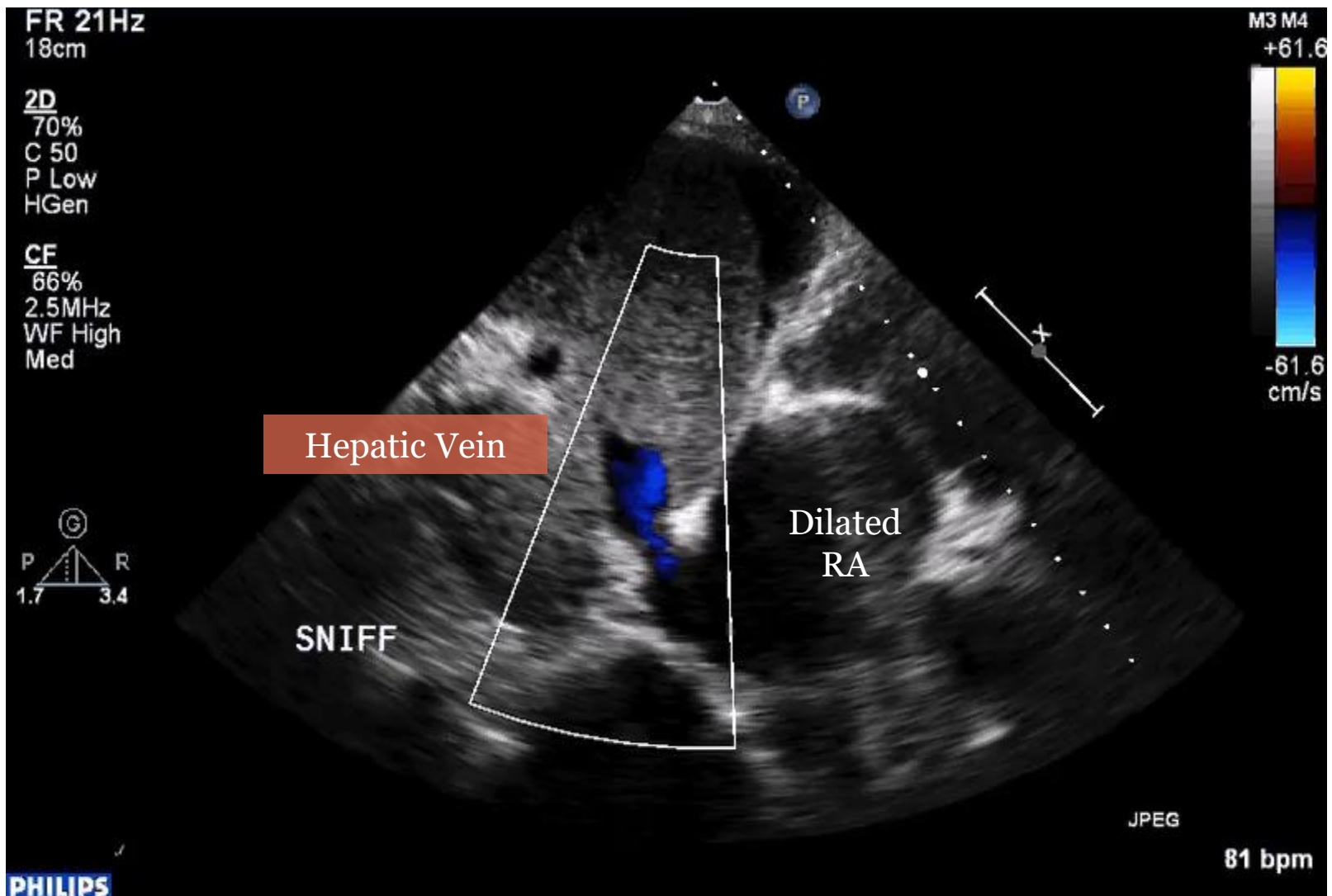
3D TRANSTHORACIC ECHOCARDIOGRAM | BIPLANE VIEW



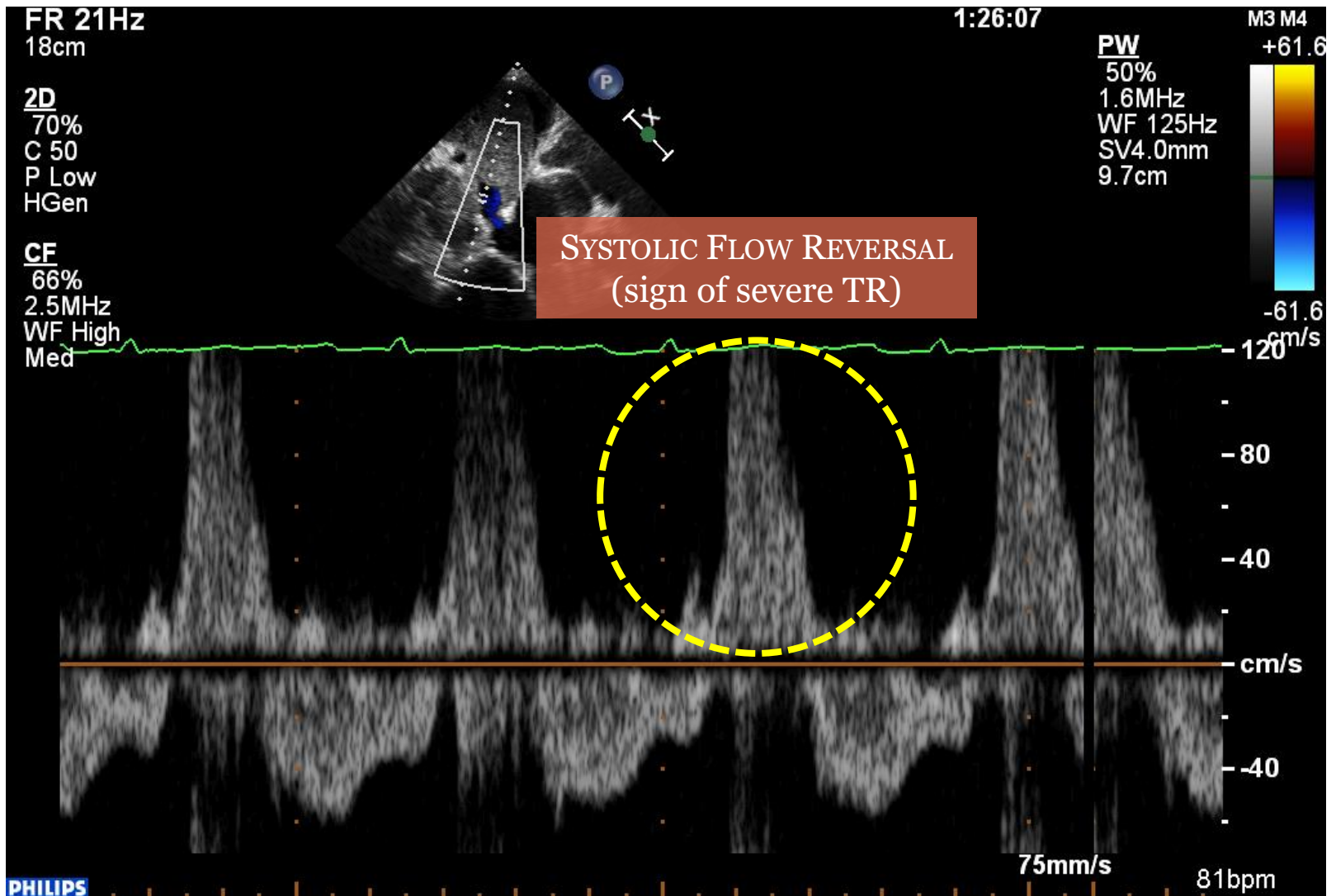
TRANSTHORACIC ECHOCARDIOGRAM | APICAL 4-CHAMBER VIEW



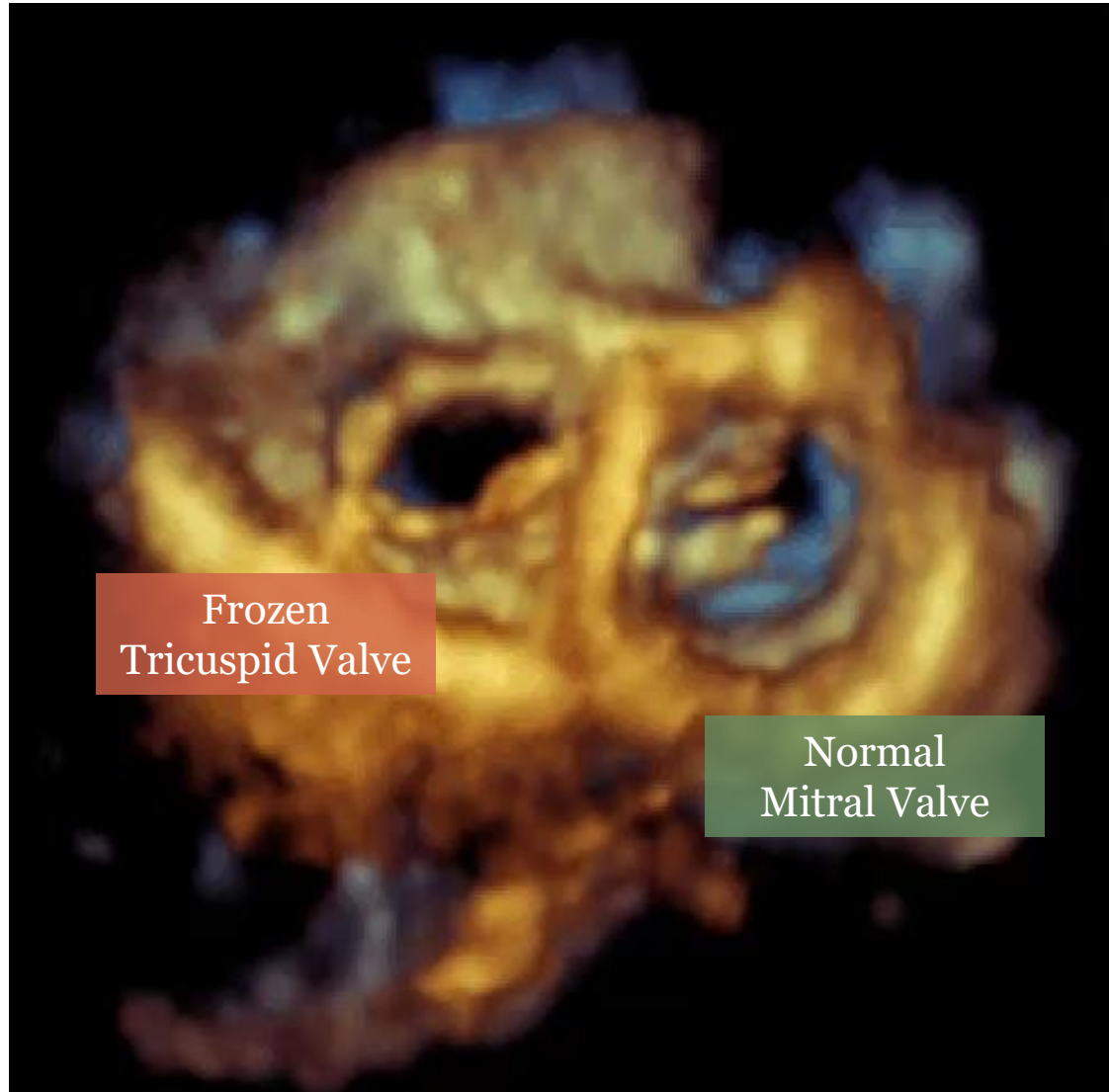
TRANSTHORACIC ECHOCARDIOGRAM | SUBCOSTAL VIEW



TRANSTHORACIC ECHOCARDIOGRAM | HEPATIC VEIN



3D TRANSTHORACIC ECHOCARDIOGRAM | LV PERSPECTIVE

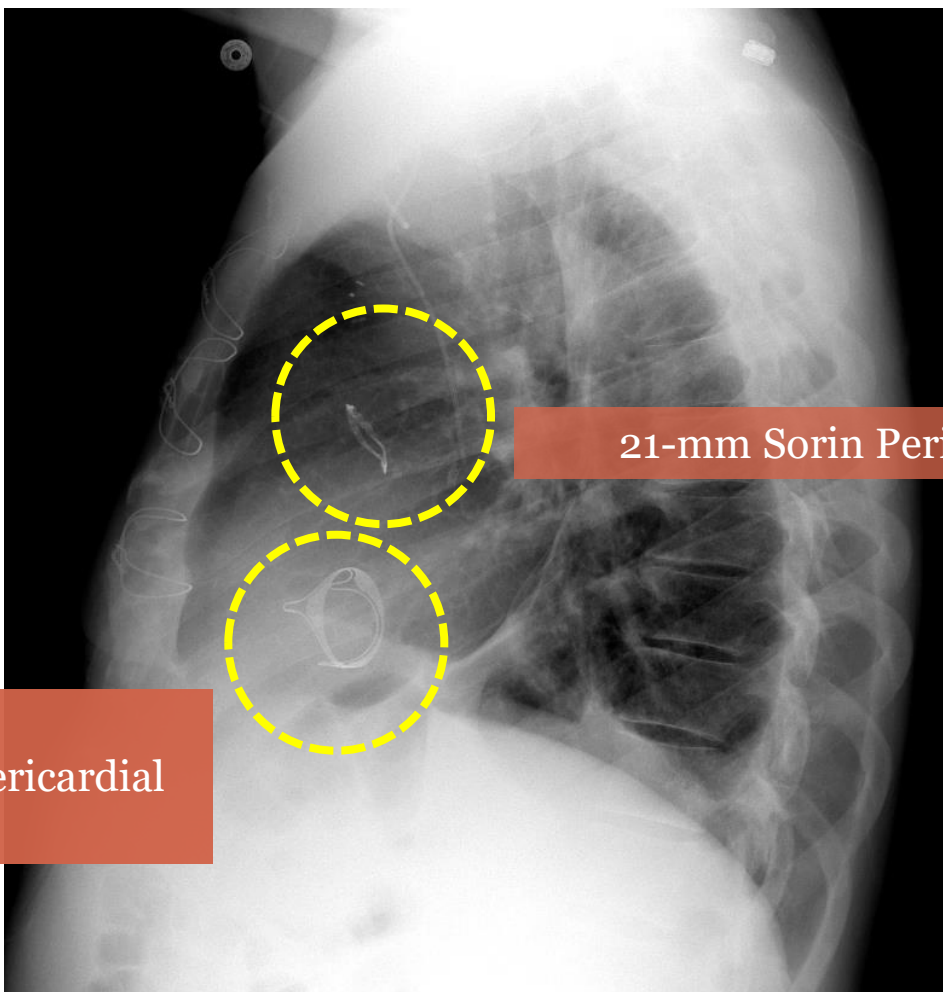


SURGERY FOR CARCINOID VALVE DISEASE

Underwent surgical replacement of both tricuspid & pulmonic valve



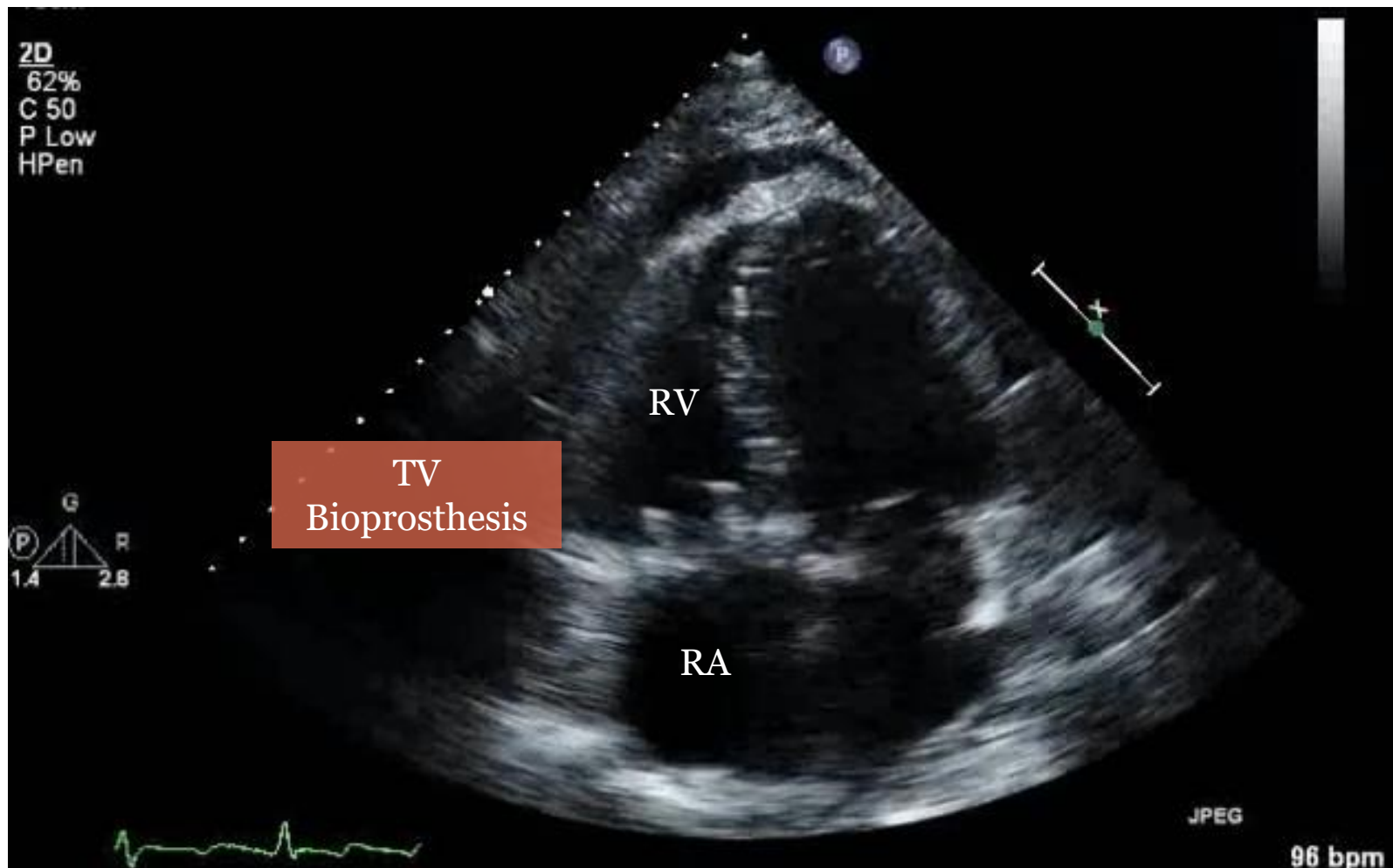
33-mm
Edwards Magna Pericardial
TVR



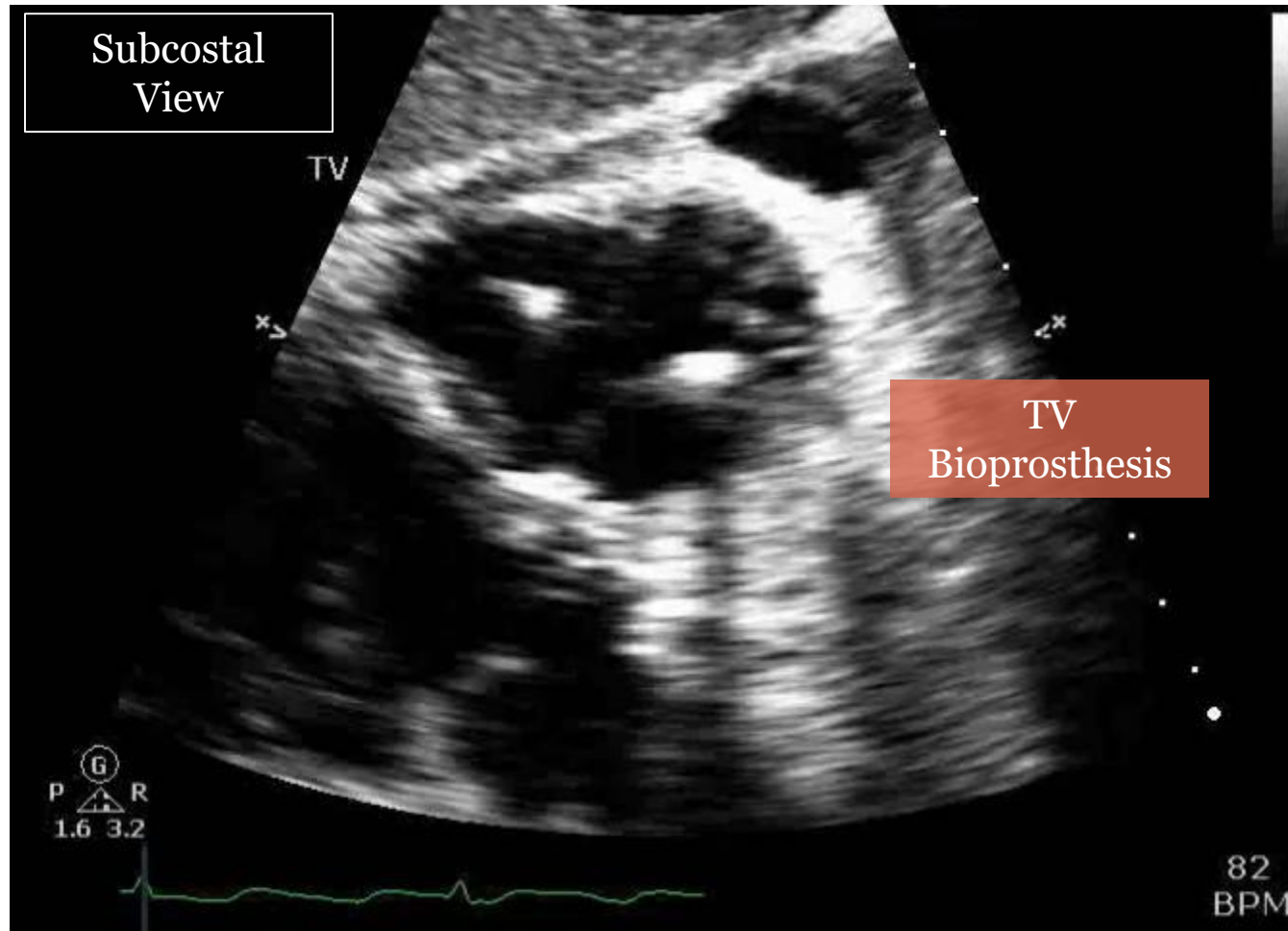
21-mm Sorin Pericardial PVR



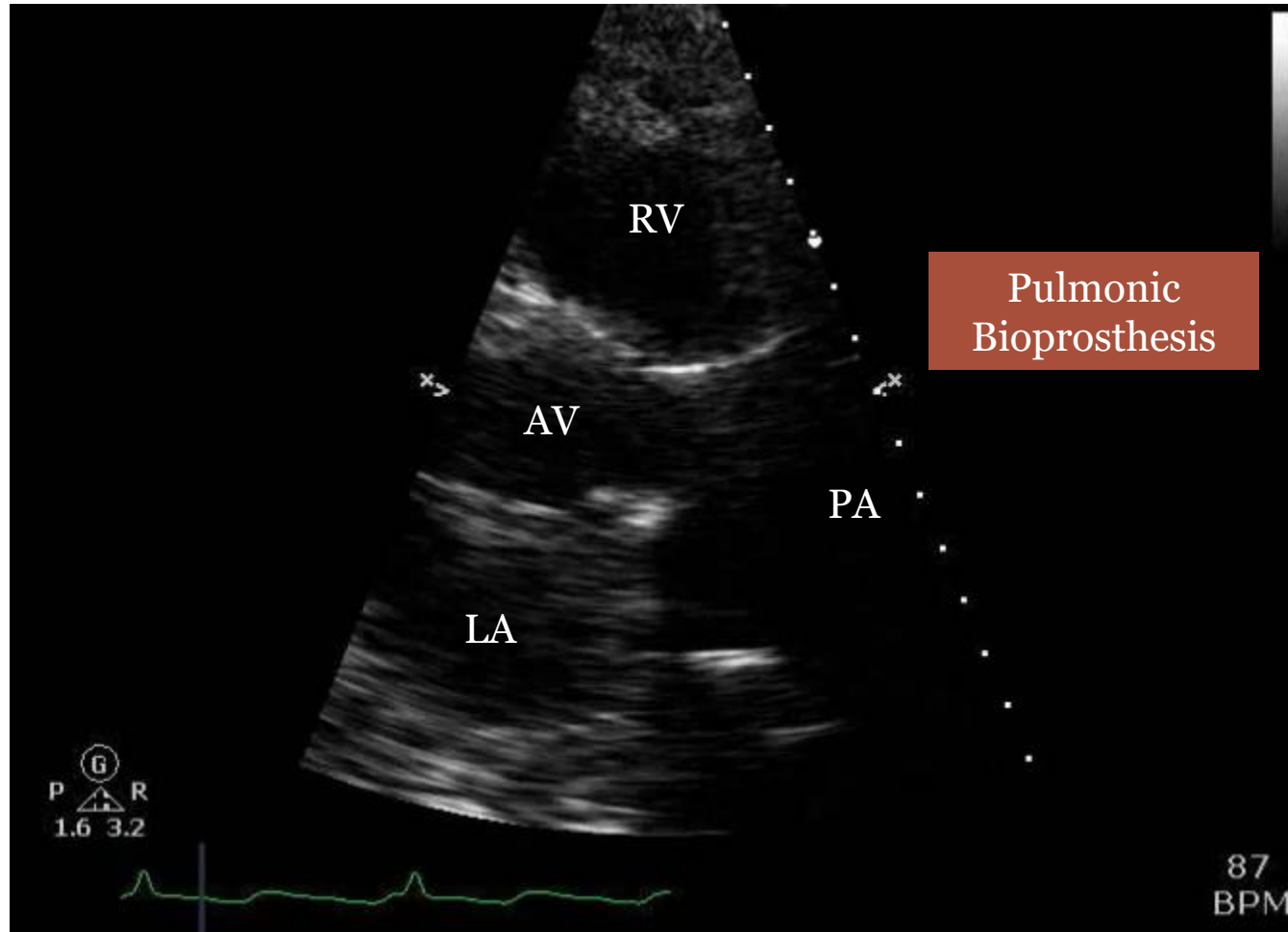
TRANSTHORACIC ECHOCARDIOGRAM | TRICUSPID BIOPROSTHESIS



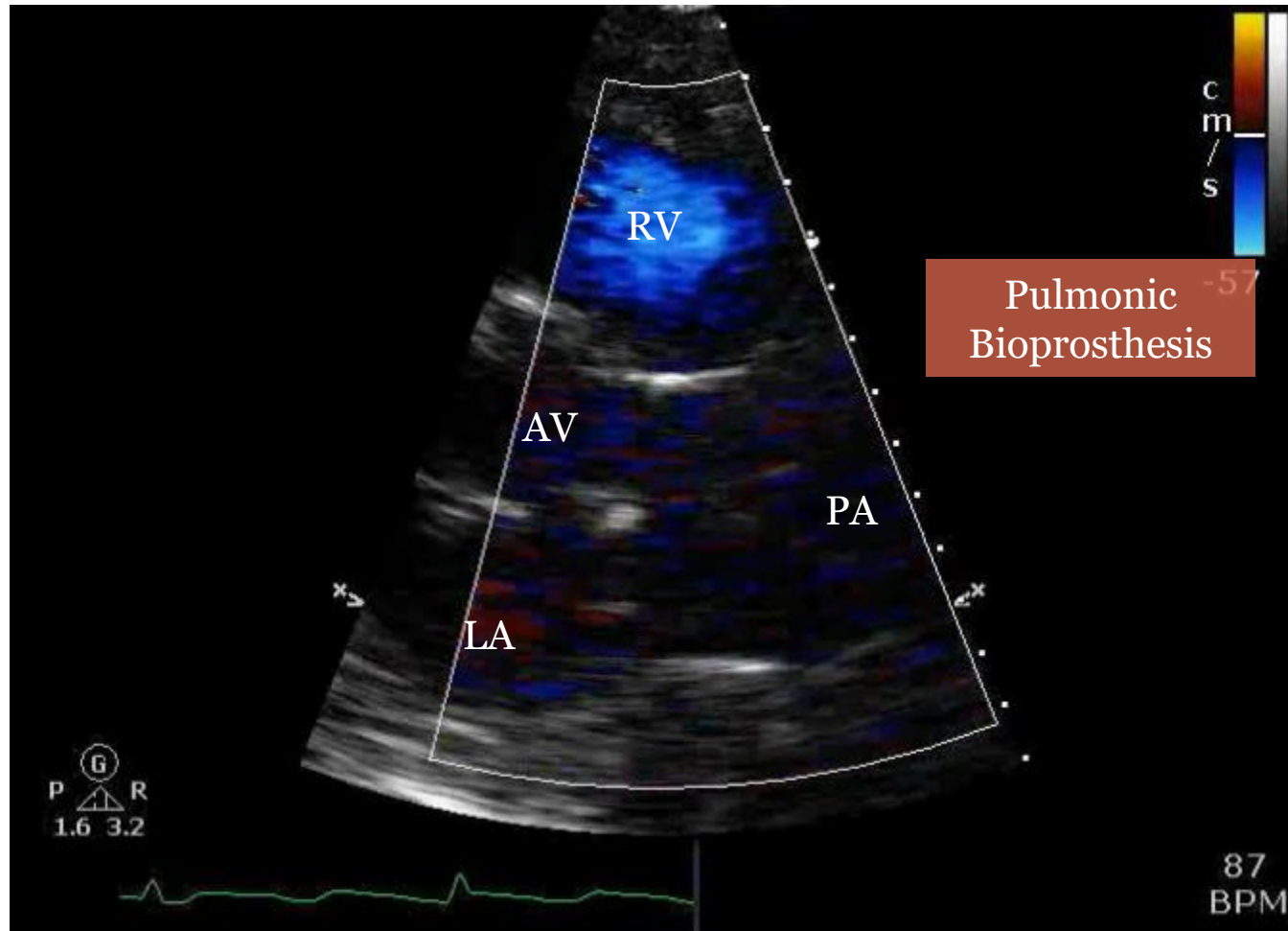
TRANSTHORACIC ECHOCARDIOGRAM | TRICUSPID BIOPROSTHESIS



TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC BIOPROSTHESIS



TRANSTHORACIC ECHOCARDIOGRAM | PULMONIC BIOPROSTHESIS



CASE #4 EPILOGUE

At present, 4 years after TVR & PVR,
patient is doing well on octreotide.

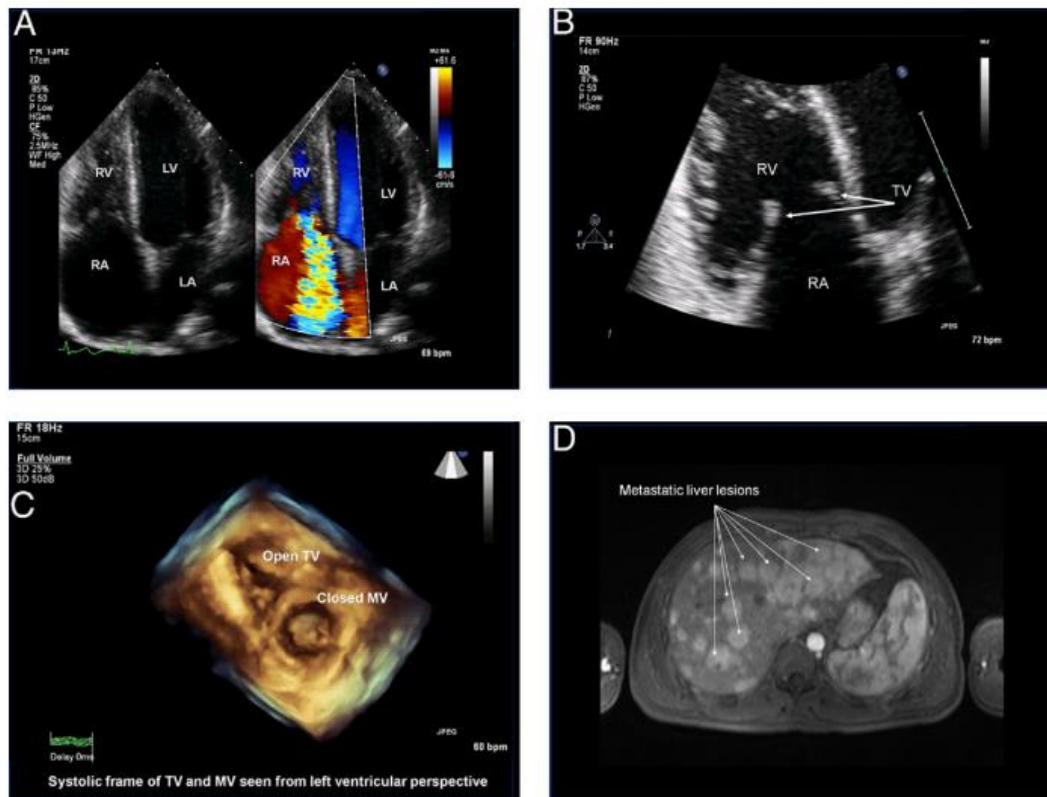
CARCINOID HEART DISEASE

Case #5

Carcinoid Heart Disease

Susie N. Hong, MD, Muhamed Saric, MD, PhD, Itzhak Kronzon, MD

New York, New York



J. Am. Coll. Cardiol. **2010**;55;1996.

Thank you

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New York University Medical Center