SIEC-2015 Napoli
TEACHING :
GROWN-UP CONGENITAL HEART DISEASE (GUCH)
Parte1

- Diagnosi occasionale in età adulta

  P. Piovesana (Camposampiero)

Cardiopatie congenite in età adulta

Cinquant'anni fa, prima dell'avvento della cardiochirurgia, solo il 20 % dei cardiopatici congeniti sopravvivevano all'età pediatrica, diventando adulti

Possiamo distinguere **cardiopatie semplici** come il Difetto Interventricolare (DIV), il Difetto Interatriale (DIA) e il Dotto Pervio (DA) e **cardiopatie complesse** come la Tetralogia di Fallot (TF), la Trasposizione dei grandi vasi (TGV), la Coartazione Aortica (CoAo) e l'Atresia della Tricuspide.

Le prime raramente si manifestano clinicamente alla nascita, mentre le seconde possono dare cianosi ("bambino blu") o precoce scompenso in modo anche drammatico.

- Gli **adulti con cardiopatie congenite** mostrano un profilo di presentazione tardiva complesso e del tutto differente, ad esempio, da quelli con cardiopatia ischemica acquisita. Esiste quasi sempre, infatti, un **lungo periodo di compensazione cardiovascolare** prima che si manifestino sintomi evidenti, pur in presenza di importanti sequele e/o residui anatomici e funzionali

- La percezione dei sintomi, in pazienti con cardiopatie congenite croniche, può anche essere misconosciuta.

- **>>>>>>> (scompenso congestizio/ IPTPol/ Aritmie)**

- Per seguire questi malati e deciderne il programma terapeutico si deve avere una **conoscenza approfondita fisiopatologica, clinica e diagnostica e delle cardiopatie congenite native**

- Rispondere ai quesiti circa **l'idoneità fisico-sportiva e lavorativa**,.

- Affrontare le specifiche condizioni tipiche delle cardiopatie congenite, quali **ipertensione polmonare**, **aritmie, endocardite infettiva, cianosi, scompenso cardiaco**, **gravidanza, disordini ematologici, chirurgia extracardiaca, anomalie** della funzione respiratoria, aspetti psicologici, cognitivi, etici e sociali.
- >> Centri di Riferimento GUCH

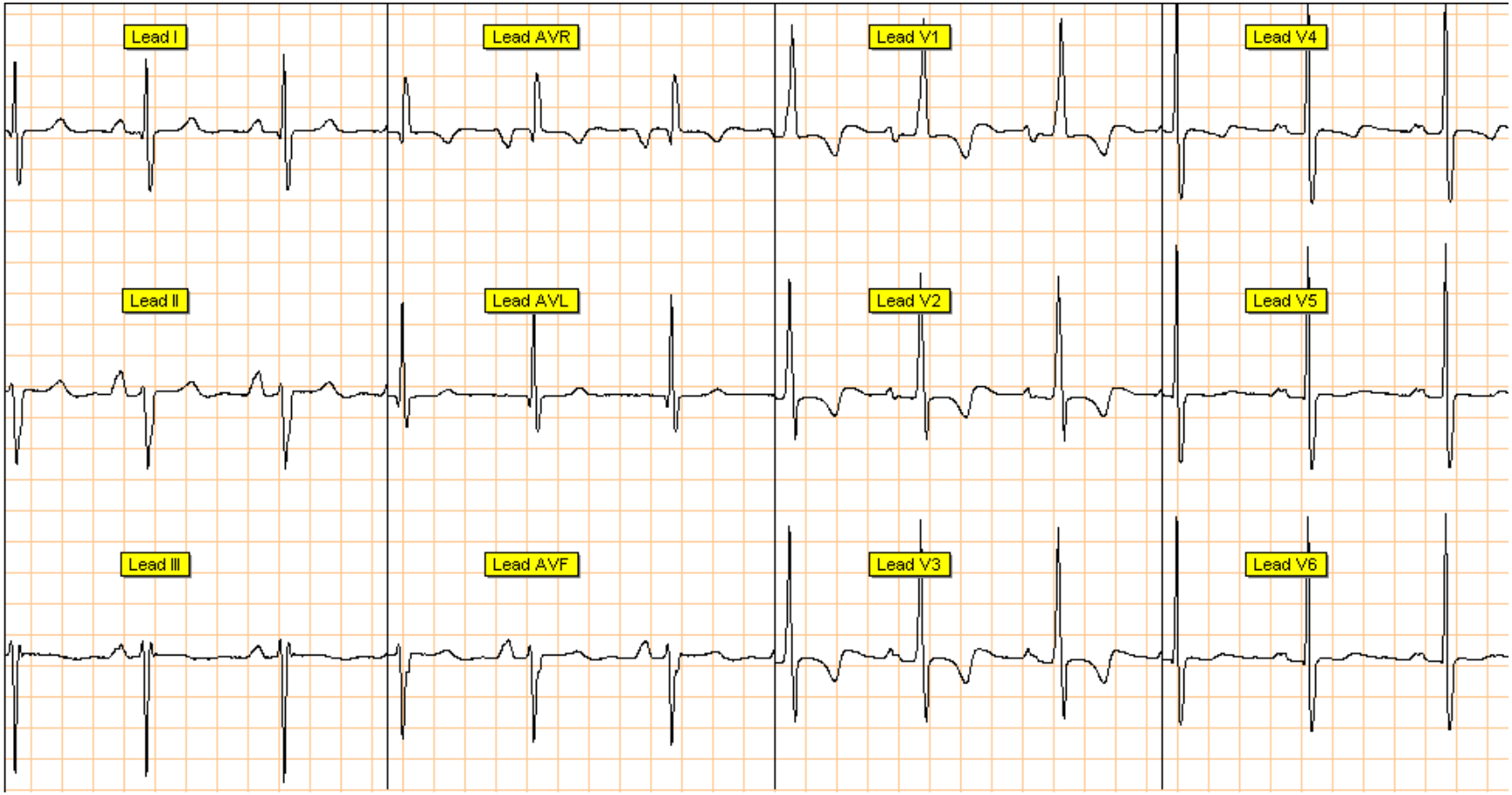
Approccio al paziente con S. di Einsenmenger

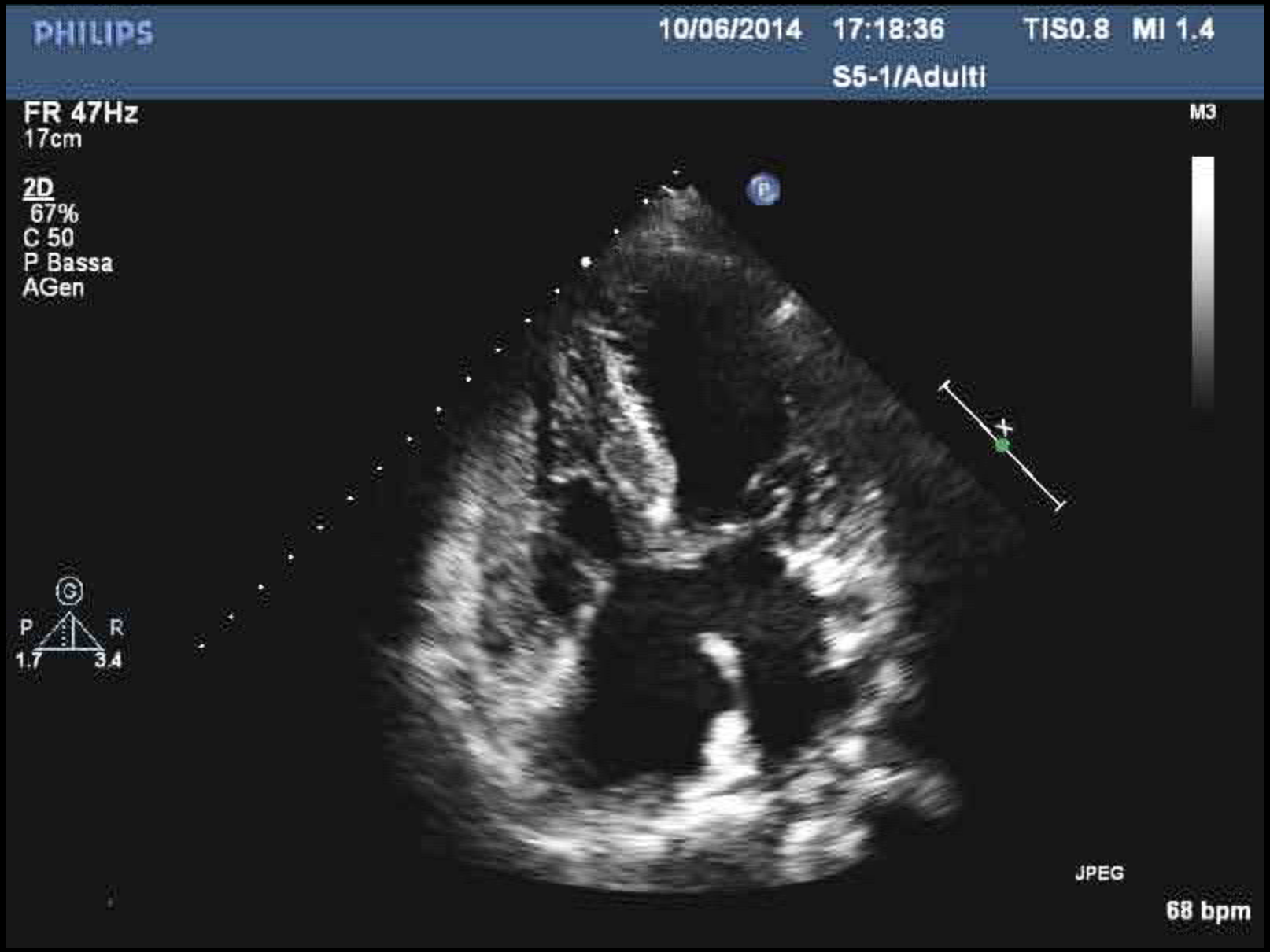
*Systemic-to-pulmonary communication, pulmonary arterial disease, and cyanosis is called Eisenmenger syndrome.

- Negli adulti la più comune causa di cianosi da malattia congenita di cuore è la Sindrome di Eisenmenger (Ipertensione polmonare) oltre alla Tetralogia di Fallot (Stenosi polmonare).

* CAV-DIV-PDA-DIA

S.L. n. 20.5.68 S.Down
Dispnea da sforzo

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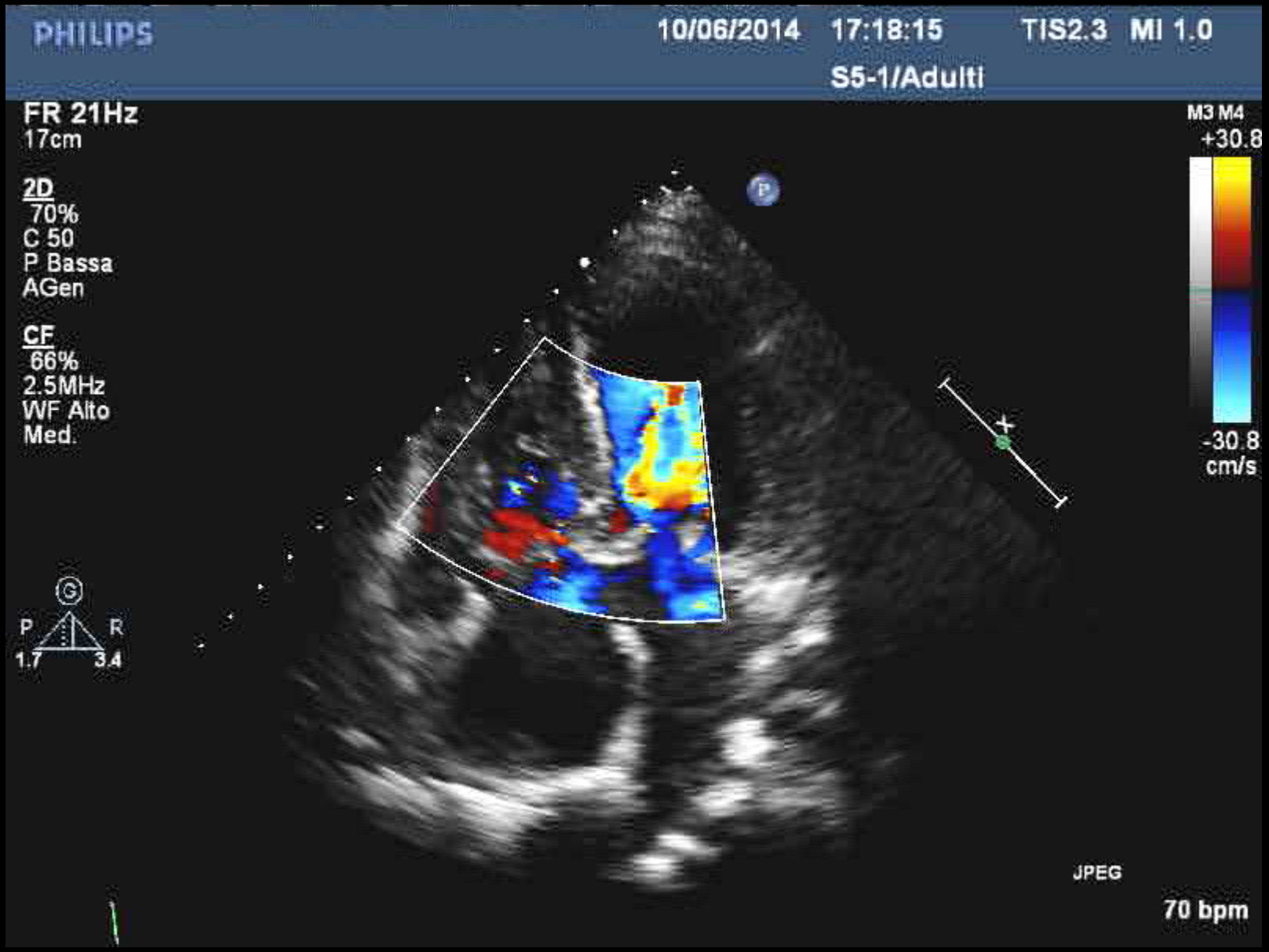
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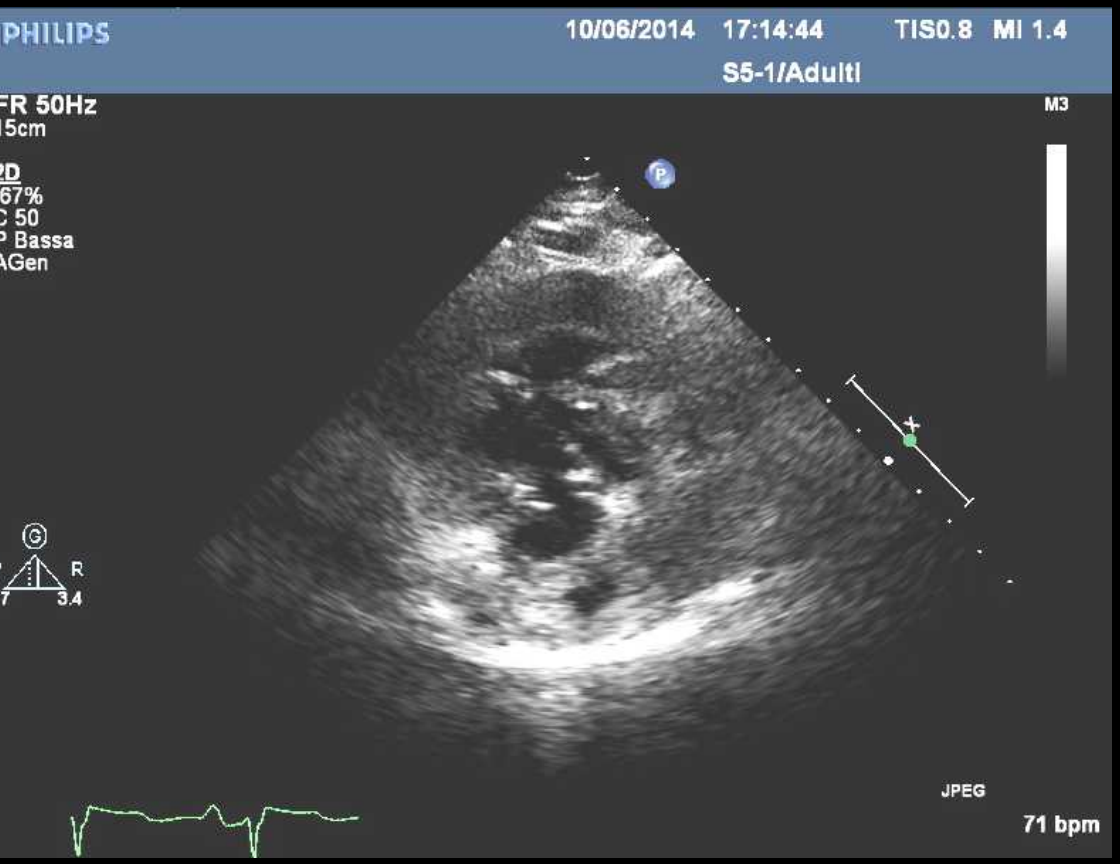
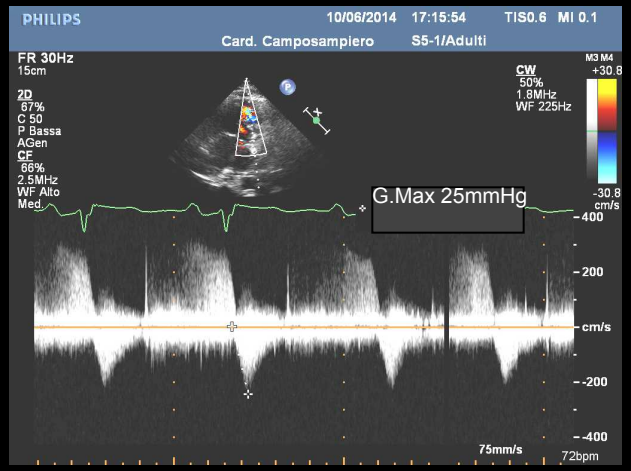
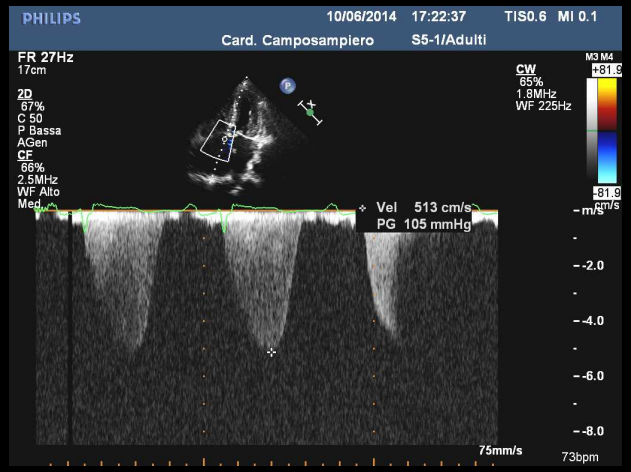






PAPS = 105+10
PPTD = 25+10
PPm = 55mmHg

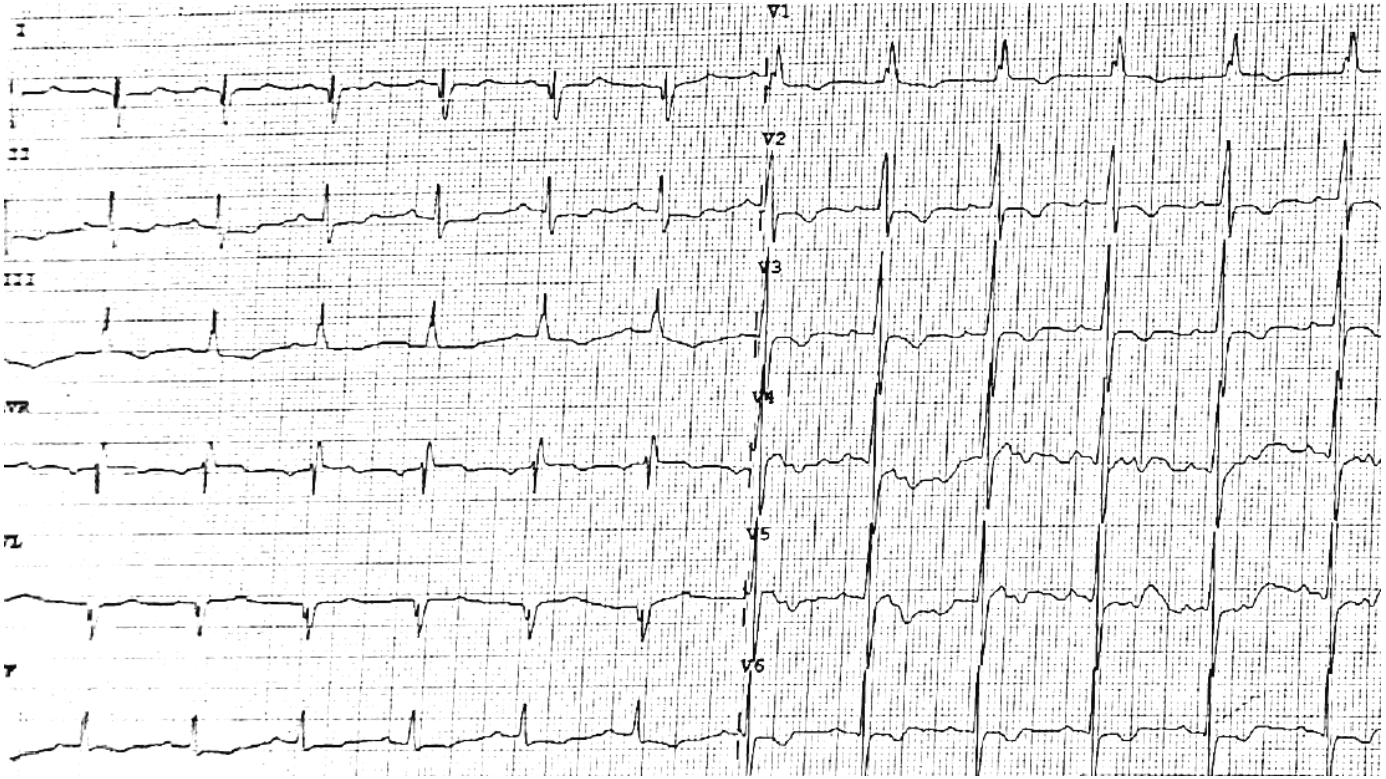
Iter diagnostico-terapeutico out-come

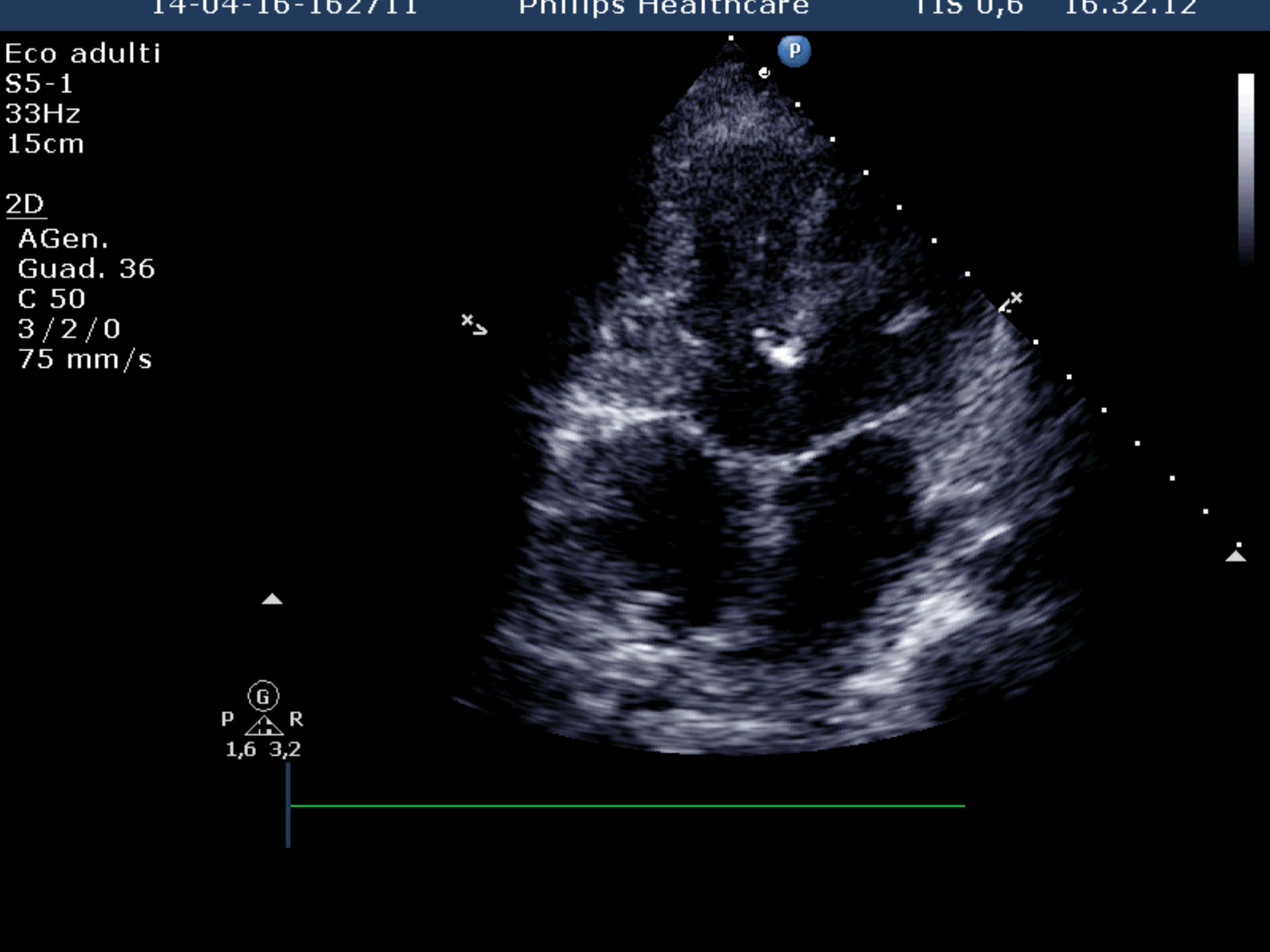
- Ecocardiogramma / valutazione clinica
- Cateterismo cardiaco
- Presa in carico pz in Centro di riferimento per la diagnosi e cura IPP
- Trattamento ipertensione polmonare con farmaci vasodilatatori polmonari .
- Test di valutazione funzionale

>>>> Miglioramento clinico-strumentale

C.A. S.Down.n.1.1.47 Non prec.card.noti Comparsa edemi declivi ed acrocianosi Diagnosi di DIV

- ECG

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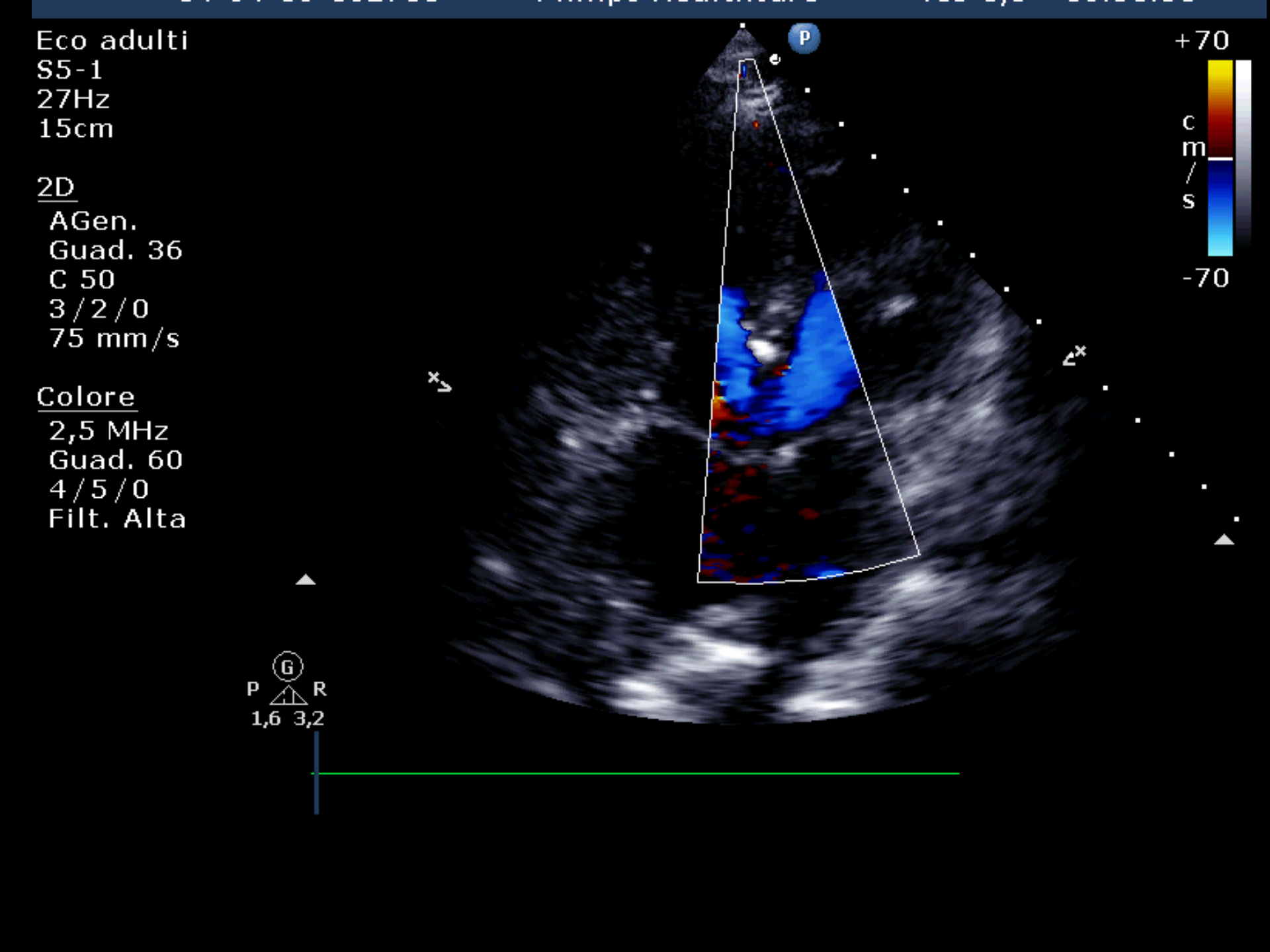
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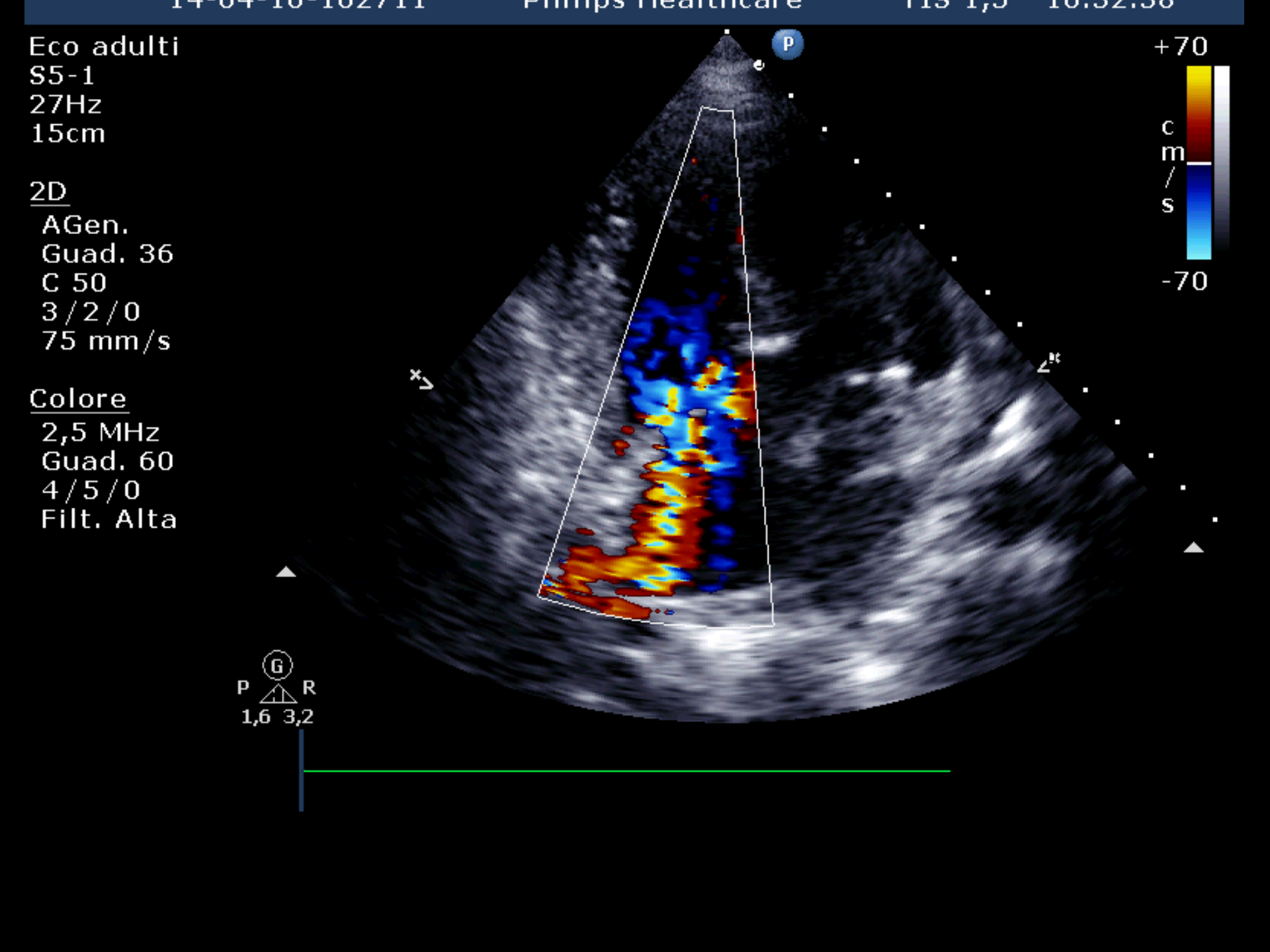
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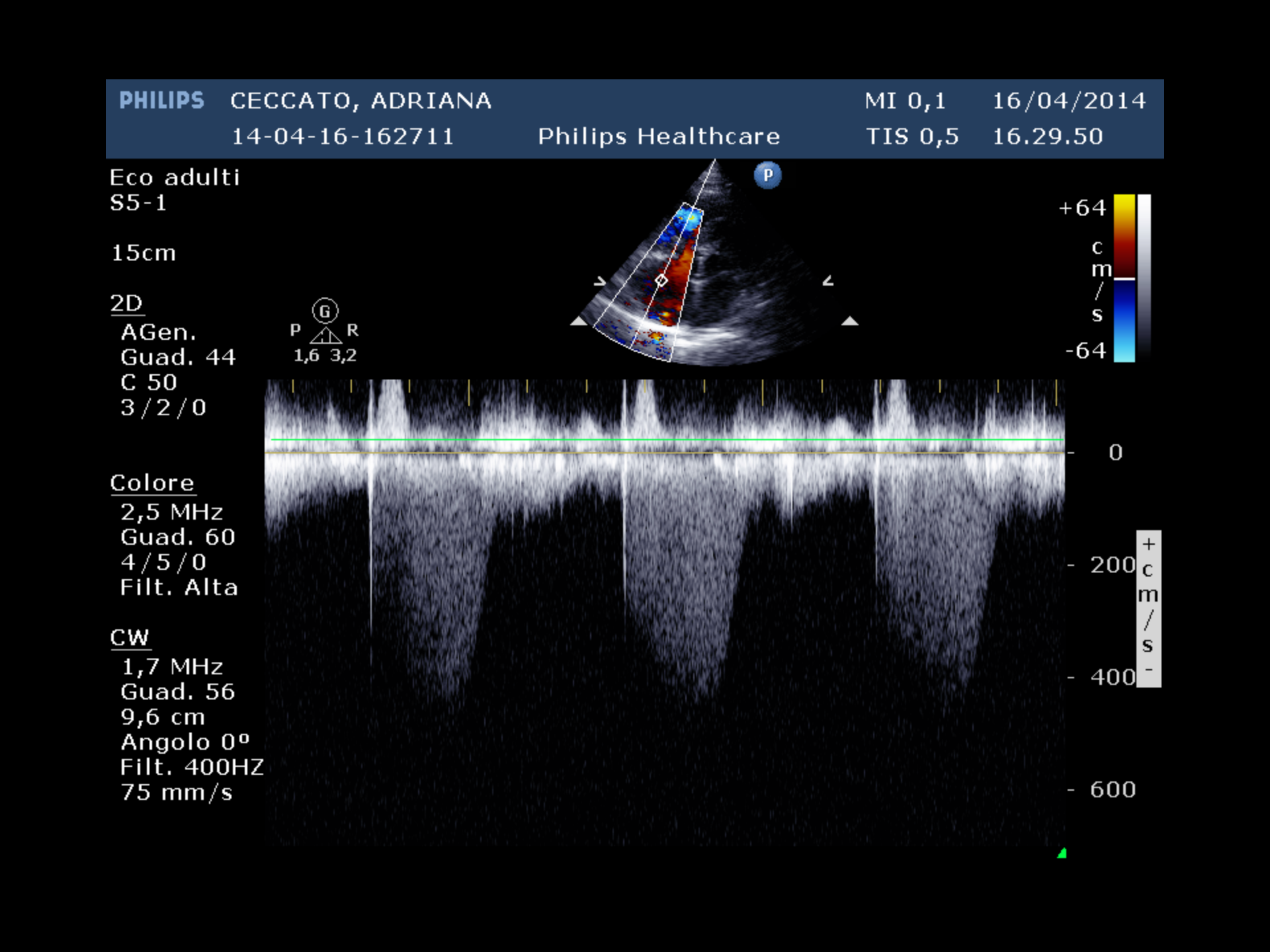
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+ cm/s -

Iter ed outcome

- Ecocardiogramma/ valutazione clinica
- Ricovero per trattamento scompenso cardiaco
- Rifiuto eseguire cateterismo cardiaco da parte tutor legale
- Impossibilità del trattamento con vasodilatatori polmonari
- Exitus dopo 8 mesi per Shock cardiogeno

Approccio al Pz con S. di E.

- Invio /Contatto con Centro di riferimento per il trattamento dellle Card.Congenite dell'Adulto.

Problematiche specifiche

- ●**Rischio infettivo** : Profilassi antibiotica dell'endocardite batterica
- ● **Policitemia** : As indicated in the 2008 American College of Cardiology/American Heart Association (ACC/AHA) guidelines, therapeutic **phlebotomy is for hemoglobin greater than 20 g/dL and hematocrit >65 percent**, associated with headache, increasing fatigue, or other symptoms of hyperviscosity in the absence of dehydration or anemia. Repeated routine phlebotomies are **not** recommended because of the risk of iron depletion, decreased oxygen-carrying capacity, and stroke.
- ● **Rischio emorragico aumentato** dovuto a frequenti e numerose anormalita' dei fattori emostatici. Terapia antiaggregante ed anticoagulante da utilizzare con estrema cautela.
- ●Insufficienza renale associata con iperuricemia e proteinuria
- ●**Rischio di embolia paradossa** (stroke ed ascessi cerebrali).

Medical management of Eisenmenger syndrome

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary
A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease)
Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

- **Pulmonary vasodilator therapy** may improve hemodynamics and quality of life in some patients.
- Caution when undertaking noncardiac surgery
- Specific attention to hematologic issues
- Heart and lung transplantation, or lung transplantation with repair of the cardiac defect
- Hemoglobin, platelet count, iron studies, creatinine, and uric acid
- Digital oximetry, both with and without supplemental oxygen therapy.
- Patients with Eisenmenger syndrome should seek prompt attention for arrhythmias and infections.

Things to avoid in Eisenmenger

- **Pregnancy** (Fixed arterial resistance , Pre-load >>>)
- **Volume depletion** (the associated fall in systemic vascular resistance can result in an increase in the right-to-left shunt with subsequent reduction in cerebral blood flow and possible cardiovascular collapse due to reduced cardiac output)
- Isometric exercise (increase right to left shunt)
- High altitude (low oxygen tension leads to further desaturation)
- **Iron deficiency may be present in patients with Eisenmenger syndrome, resulting in relative anemia even in the face of elevated hemoglobin levels**, and must be managed with care.
- Pulmonary artery thrombosis is common among adults with Eisenmenger syndrome. The benefit of anticoagulation has not been proven
- **Any surgical procedure, even if relatively minor, is potentially life-threatening**

CASO CLINICO

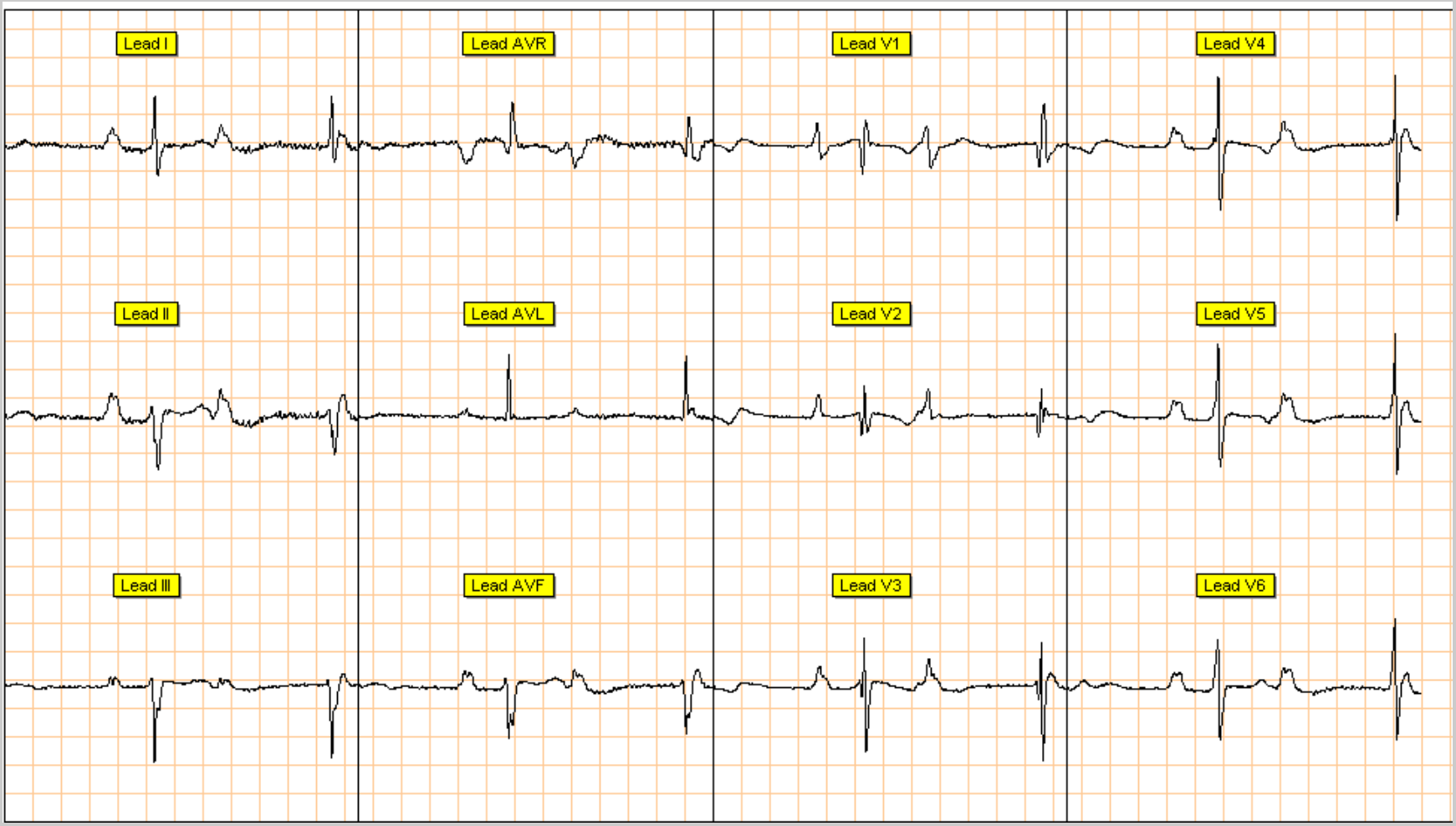
Paziente di sesso femminile,45 anni, razza africana

Non precedenti di rilievo

3 gravidanze normodecorse

Accesso in PS in Aprile 2012 per crisi ipertensiva

Riscontro ECG di blocco atrio-ventricolare completo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Lead II
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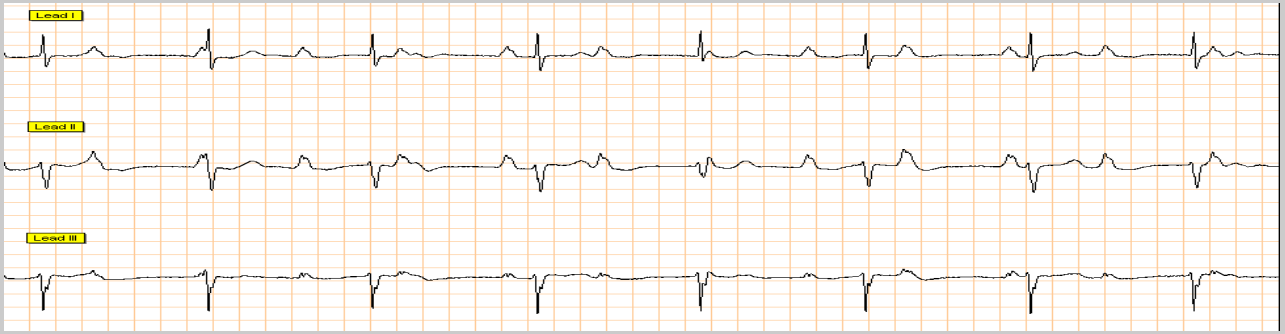
Lead III
Lead AVF
Lead V3
Lead V6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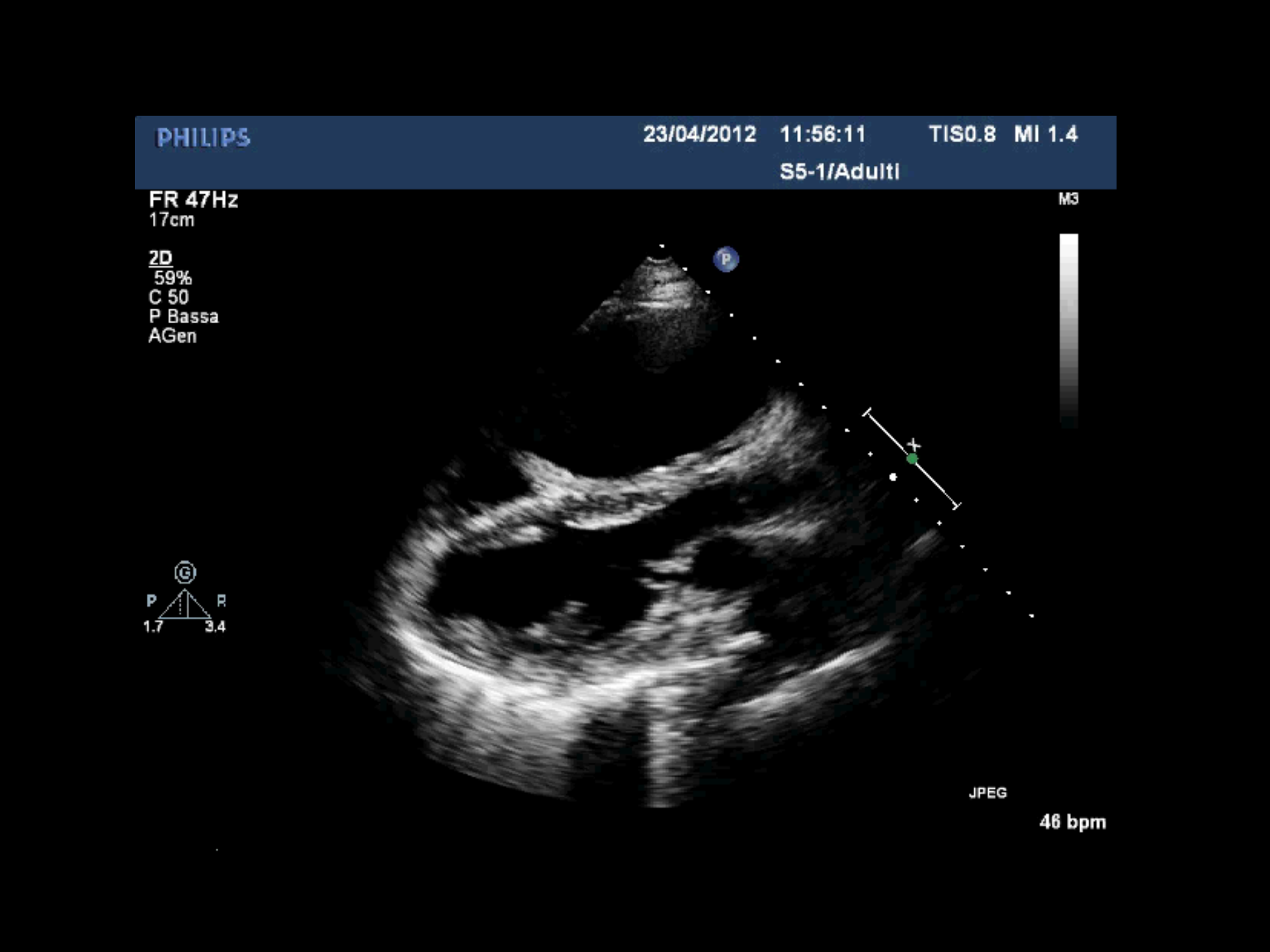
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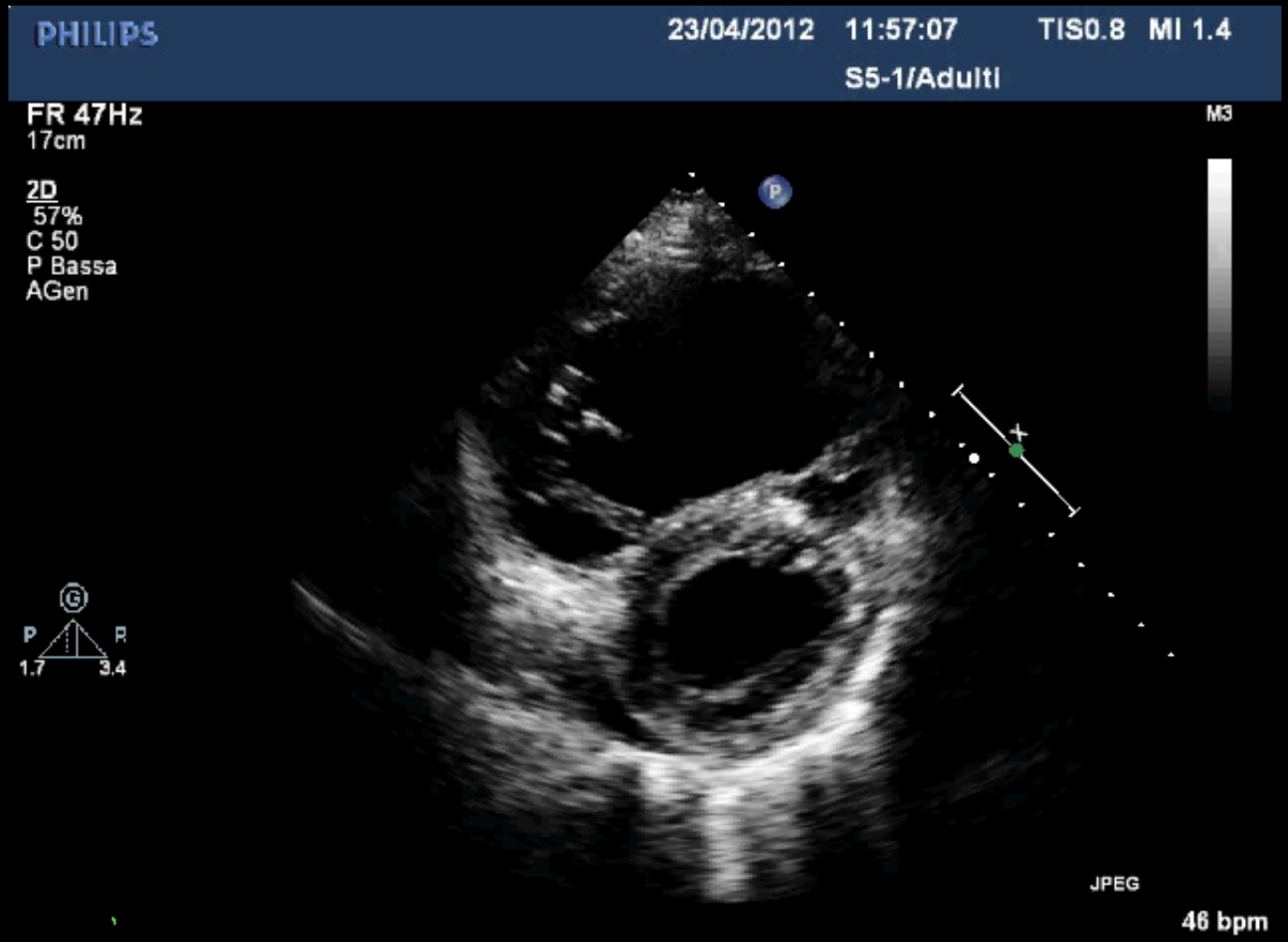
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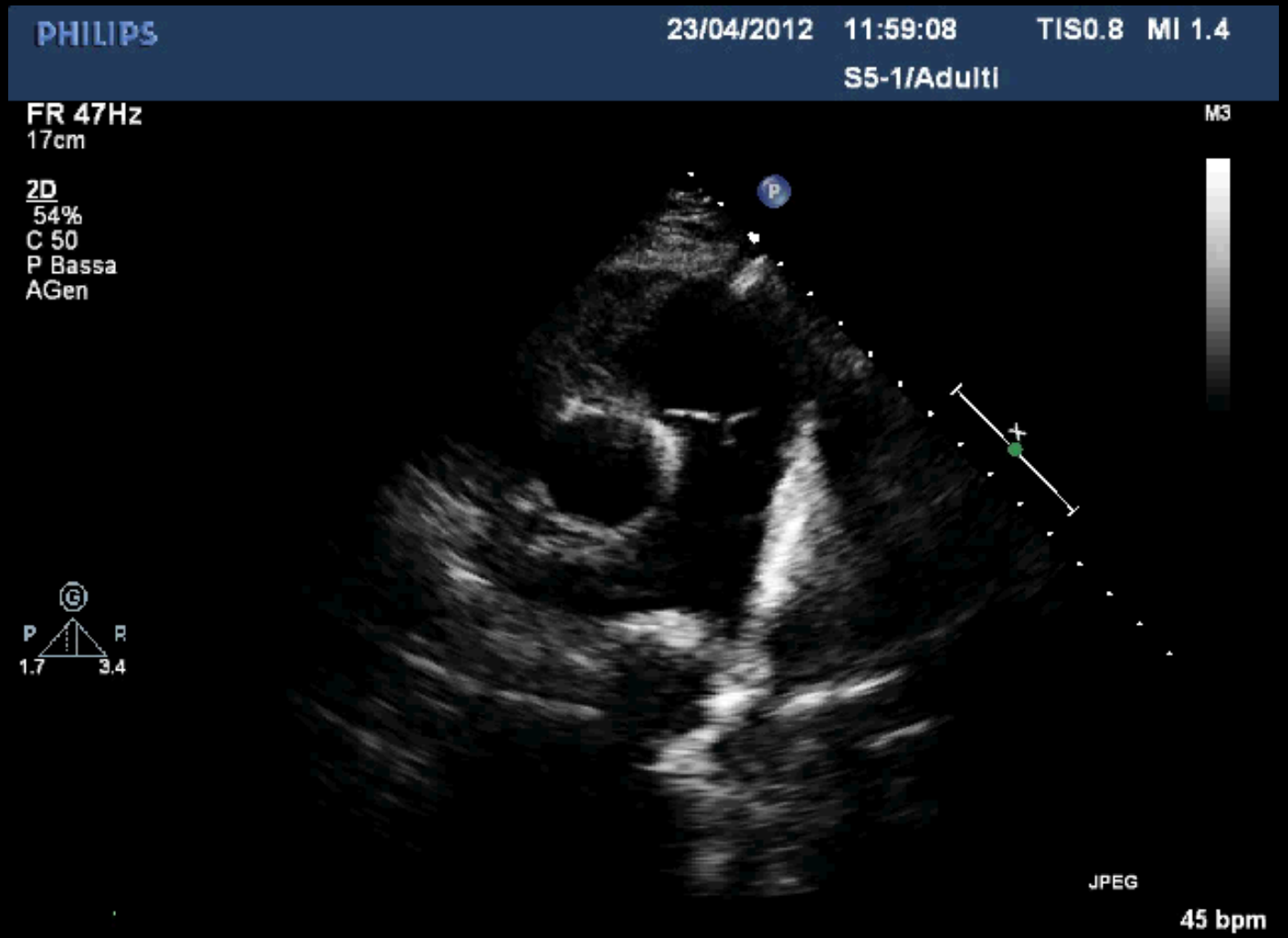
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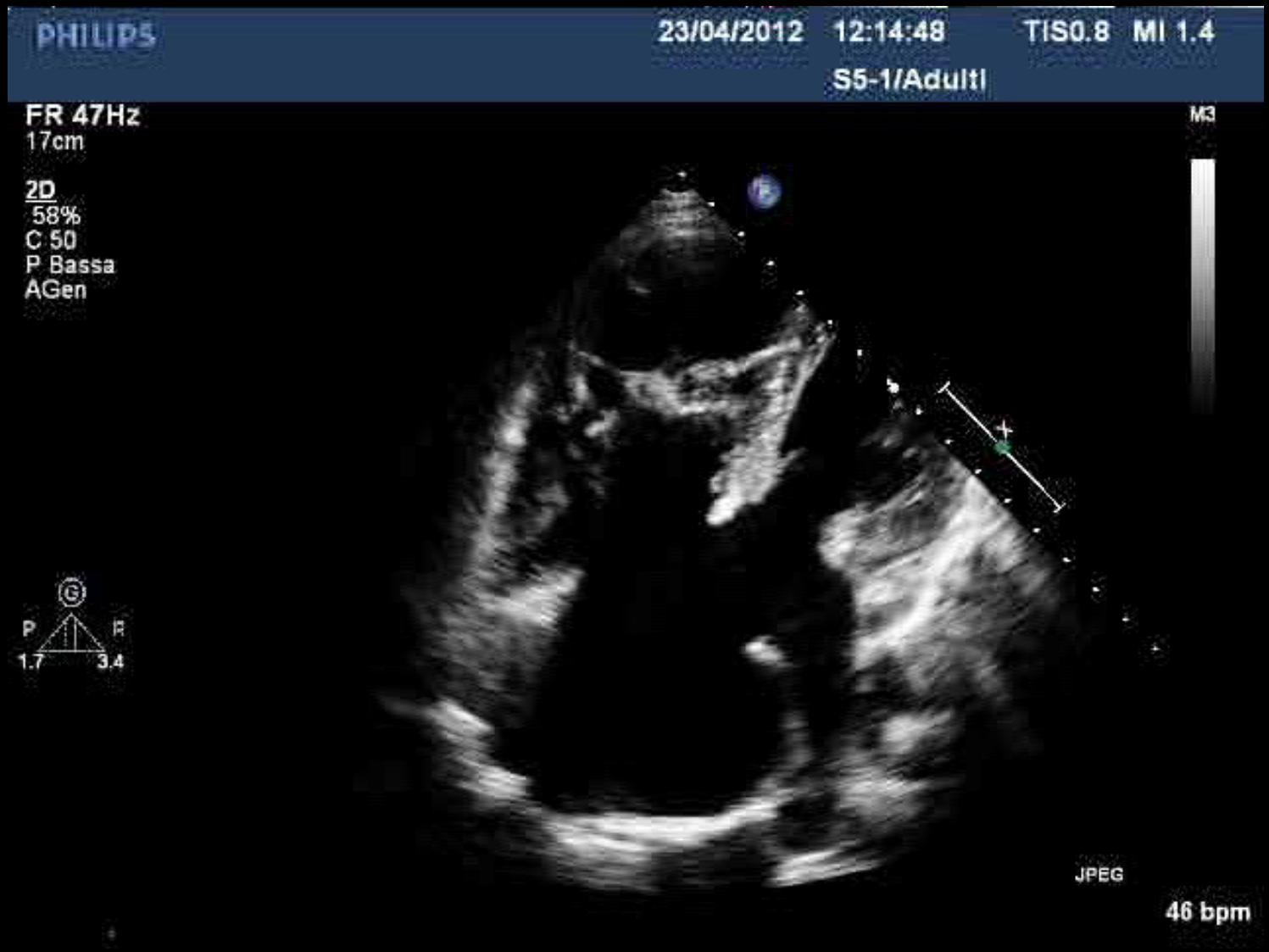
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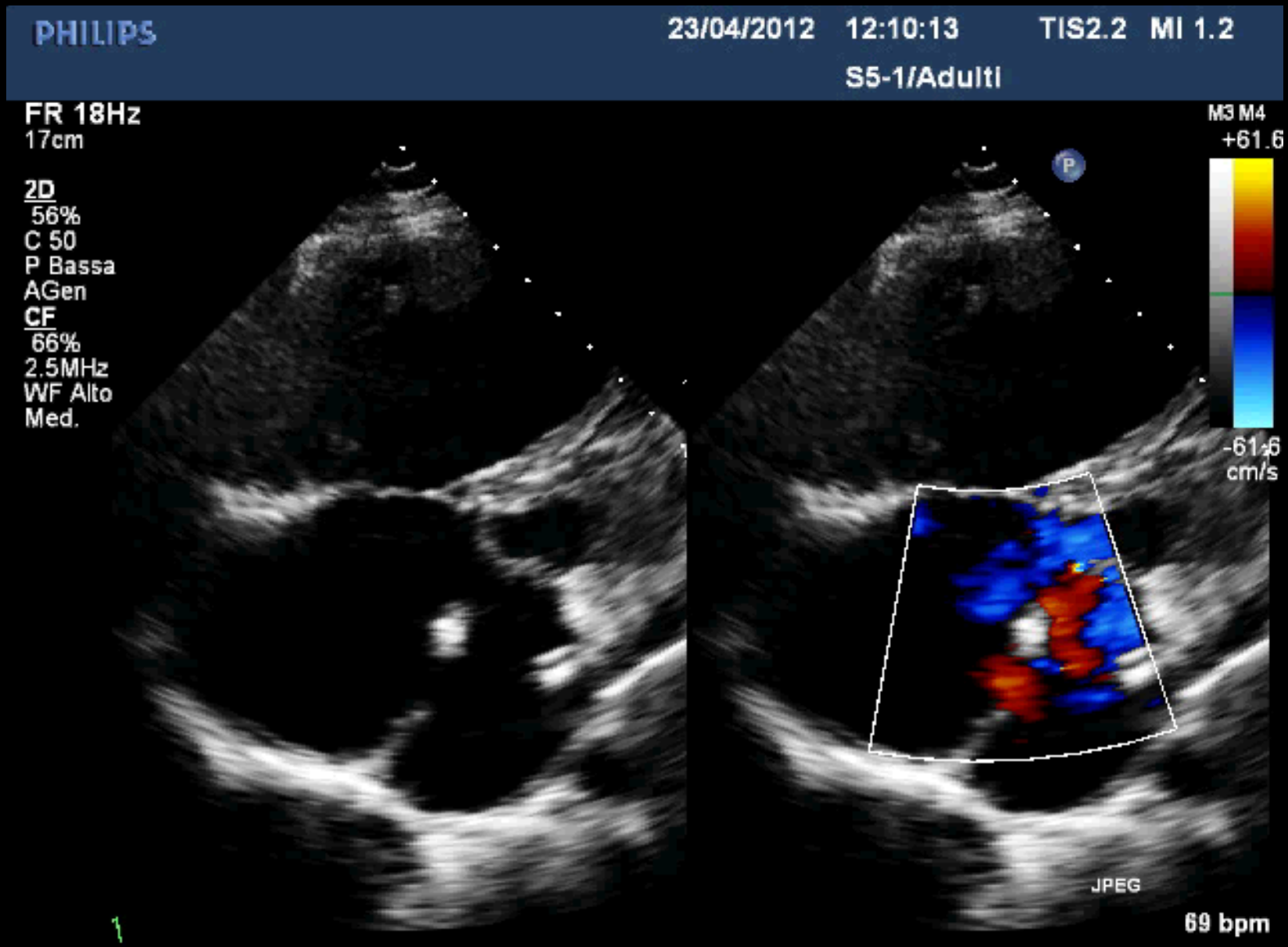
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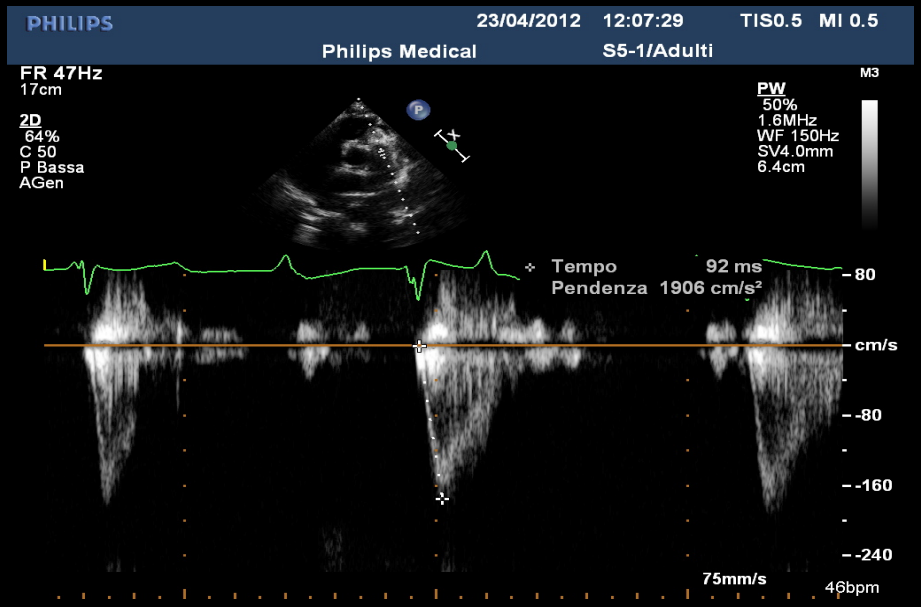
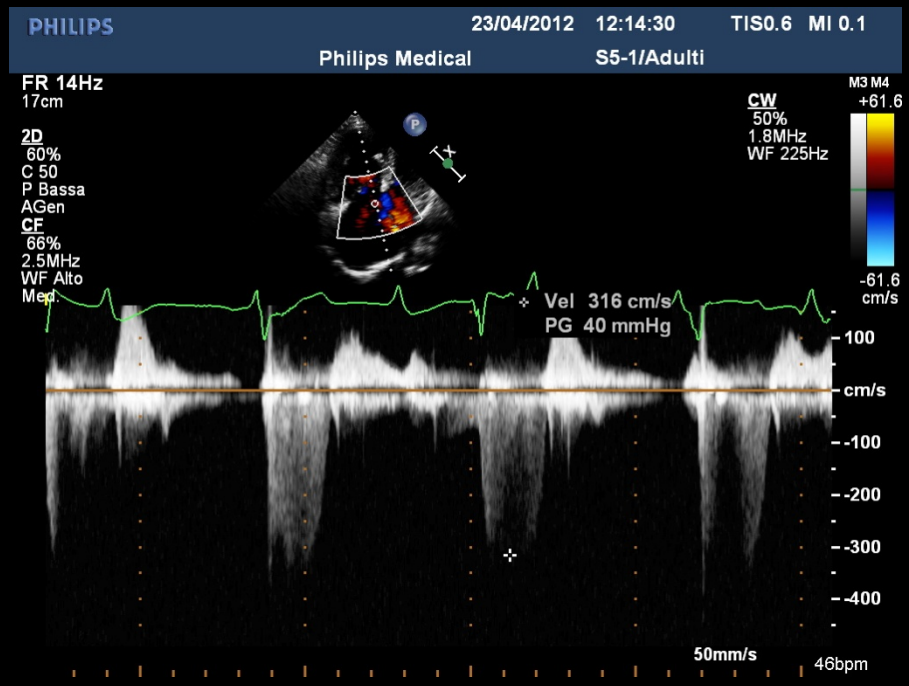
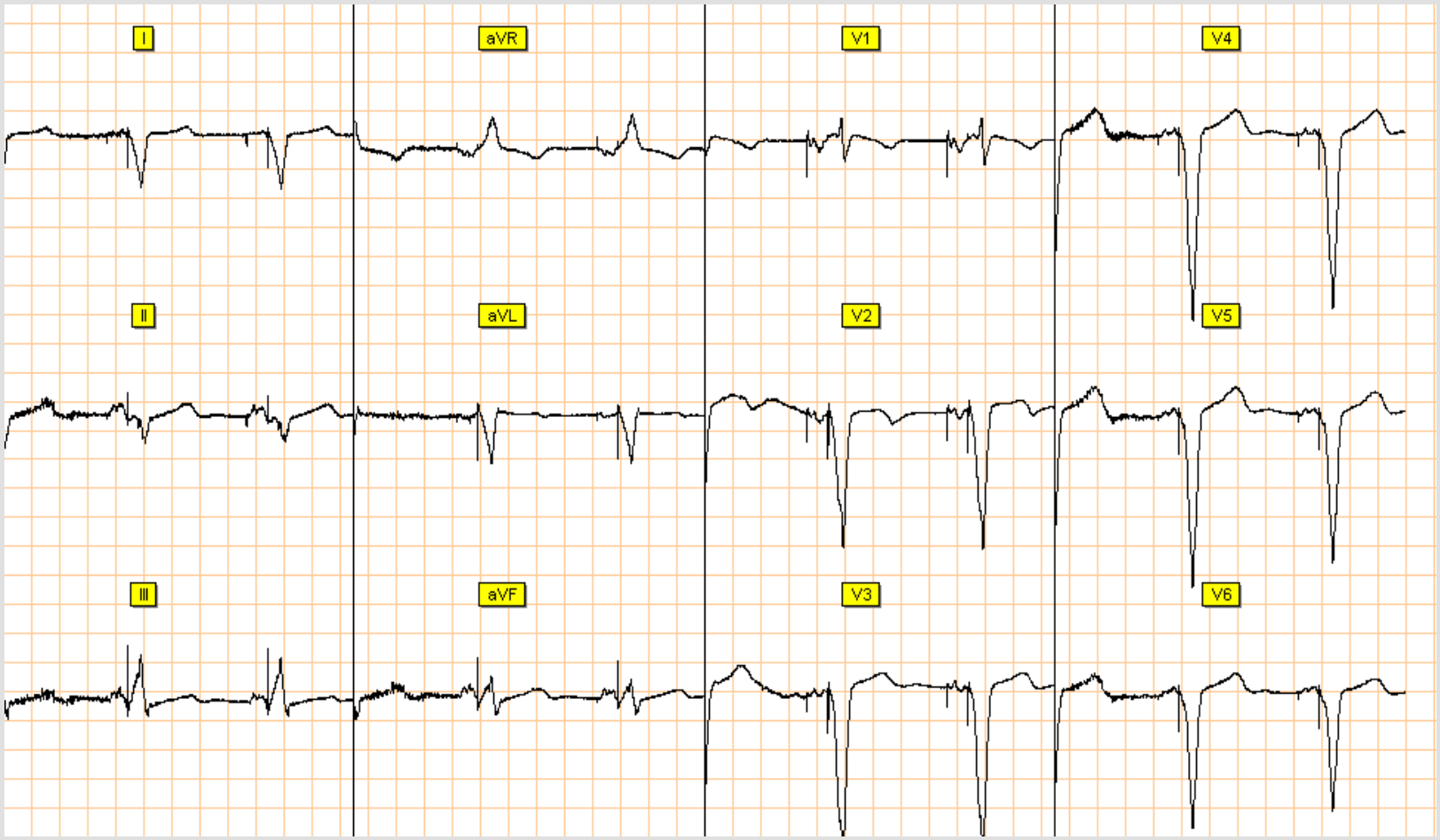




PAPS 55mmHg
PAm 30 mmHg
(Dynamic Pulmonary Hypertension)

Trasferimento in Cardiochirurgia dove viene sottoposta ad Intervento di correzione chirurgica (chiusura del DIA ostium primum e ostium secundum + plastica mitralica) + impianto di pace-maker epicardico DDD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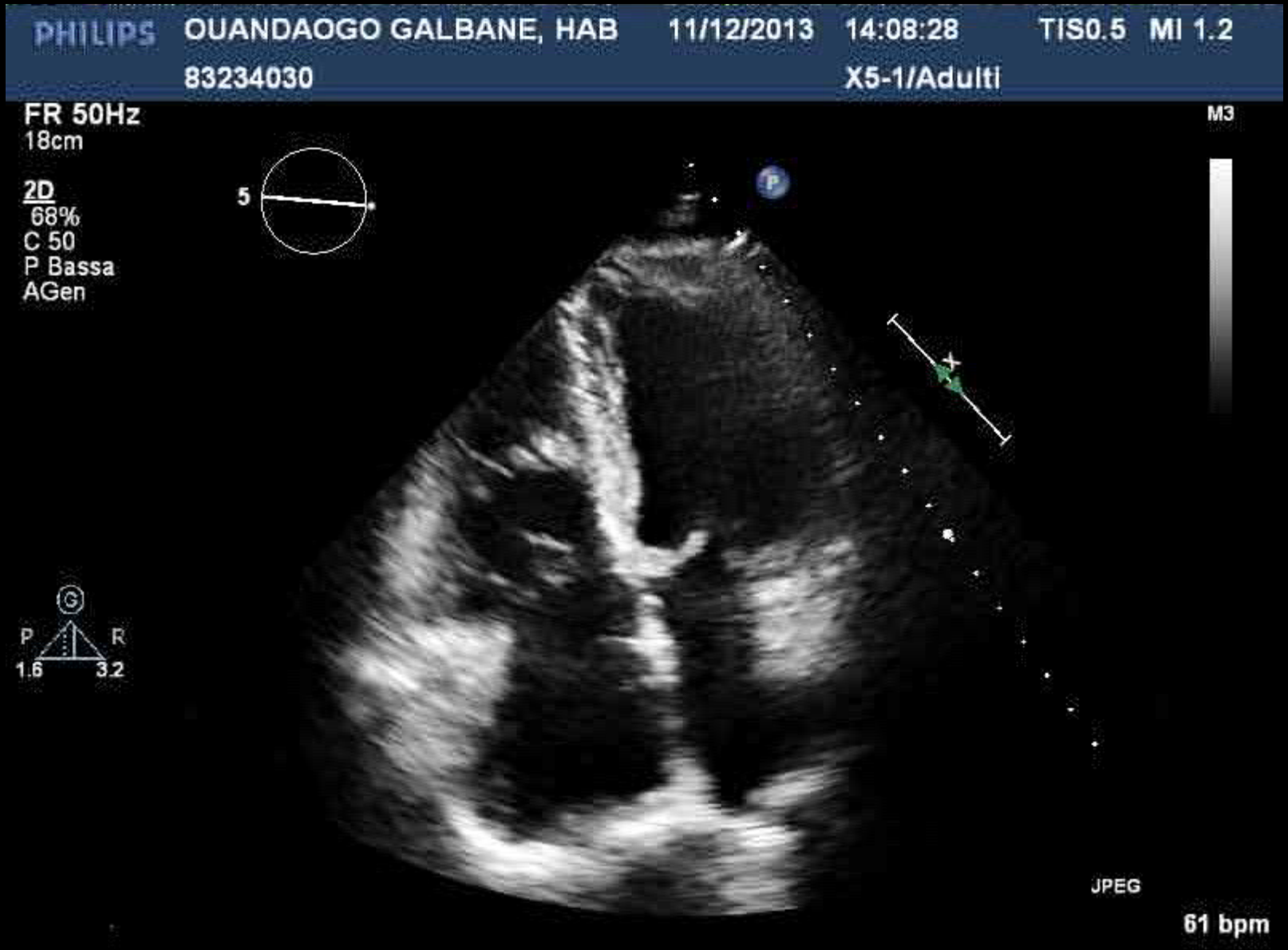
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61 bpm

TOF

- Sebbene molto rara, TOF rimane la malformazione cianotizzante più frequente nell'adulto senza precedente intervento chirurgico.
- L'intervento correttivo e' ancora possibile in età adulta

Tetralogy of fallot repair in patients 40 years or older.
Attenhofer Jost CH, Connolly HM, Burkhart HM, Scott CG, Dearani JA, Carroll AJ, Tajik
Mayo Clin Proc. 2010;85(12):1090.

- OBJECTIVE: To report the outcomes of patients with tetralogy of Fallot (TOF) undergoing surgical repair at age 40 years or older.
- PATIENTS AND METHODS: We reviewed records of patients (age,≥40 years) who underwent TOF repair from January 1, 1970, through December 31, 2007. Symptoms, palliative procedures, surgical reports, and long-term outcomes were analyzed.
- RESULTS: Fifty-two patients (30 men [58%]) had surgery at a mean±SD age of 50±8 years; 27 (52%) had prior palliative surgery at a mean±SD age of 17±11 years. Procedures for TOF repair included pulmonary valve replacement (n=10), transannular patch (n=10), and native pulmonary valve preservation (n=32). The 30-day mortality rate was 6% (stroke, n=2; ventricular fibrillation, n=1). A mean±SD follow-up of 14.9±9.3 years was feasible in 48 of 49 survivors; improvement in functional class was observed in 42 patients. Reoperation was performed in 7 patients (4 for pulmonary regurgitation). Twenty-nine patients died (mean±SD age, 65±12 years); causes of death were cardiac (n=7), noncardiac (n=4), and unknown (n=18). Mean±SD age at death was younger in patients with previous palliation (59±11 years vs 70±12 years; P=.03). The 10-year survival rate was lower than expected compared with an age- and sex-matched population (73% vs 91%; P<.001).
- CONCLUSION: Complete repair of TOF in patients 40 years or older is feasible but carries increased operative risk. Surgical survivors have improvement in functional class; however, survival remains lower than expected. Reduced survival and **need for reoperation emphasize the importance of pulmonary valve replacement at the time of initial repair** and long-term follow-up.

Tetralogy of fallot repair in patients 40 years or older.
Attenhofer Jost CH, Connolly HM, Burkhart HM, Scott CG, Dearani JA, Carroll AJ, Tajik
Mayo Clin Proc. 2010;85(12):1090.

TABLE 4. **Follow-up Data**[a,b]

Variable	All patients (N=48)	Previous palliation		*P* value
		Yes (n=25)	No (n=23)	
Duration of follow-up (y)	14.9±9.3	12.4±6.7	17.5±10.9	.05
Postoperative improvement	42 (88)	22 (88)	20 (87)	>.99
NYHA functional class				.06
I	22 (46)	8 (32)	14 (61)	
II	21 (44)	15 (60)	6 (26)	
III	4 (8)	1 (4)	3 (13)	
IV	1 (2)	1 (4)	0 (0)	
Reoperation	7 (15)	5 (20)	2 (9)	.42
Interval between surgery and death (y)	16±11	12±8	20±12	.06
Age at death (y)	65±12	59±11	70±12	.03

[a] NYHA = New York Heart Association.
[b] Categorical data are provided as number (percentage) of patients and continuous data as mean ± SD.

Miglior prognosi nei pazienti non trattati in precedenza con palliazione
(anastomosi sistemico-polmonari per correggere cianosi)
Fallot rosei
(minore compromissione anatomo-funzionale)

DIFETTI INTERATRIALI DELL’ADULTO

ASD Natural History

- Difetto del Setto Interatriale (ASD) la più frequente cardiopatia congenita dell'adulto (oltre alla Valvola Ao bicuspide).
- Decorso asintomatico
- Complicazioni come primo sintomo

  (aritmie sopraventricolari, scompenso dx, ipertensione polmonare,embolia paradossa)

Difetto interatriale : Storia naturale

- Nei pz con moderato-grande ASD non operati lo shunt puo' aumentare con l'eta'.

  Nelle forme gravi
- Intorno ai 40 anni >>scompenso congestizio
- Aritmie sopraventricolari da stretching atriale
- <10% dei casi >>>Ipertensione polmonare

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary
A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease

Indications for closure and medical management of atrial septal defects in adults

- The main indication for atrial septal defect (ASD) closure is right ventricular enlargement with or without symptoms
- ASD closure is also reasonable in patients with paradoxical embolism or documented orthodeoxia-platypnea

Orthodeoxia-platypnea

- Rare disorder characterized by both arterial desaturation (orthodeoxia) and dyspnea (platypnea) in the upright position with improvement in the supine position. This syndrome has been described with ASD and PFO.

Atrial Septal Defect and pulmonary hypertension

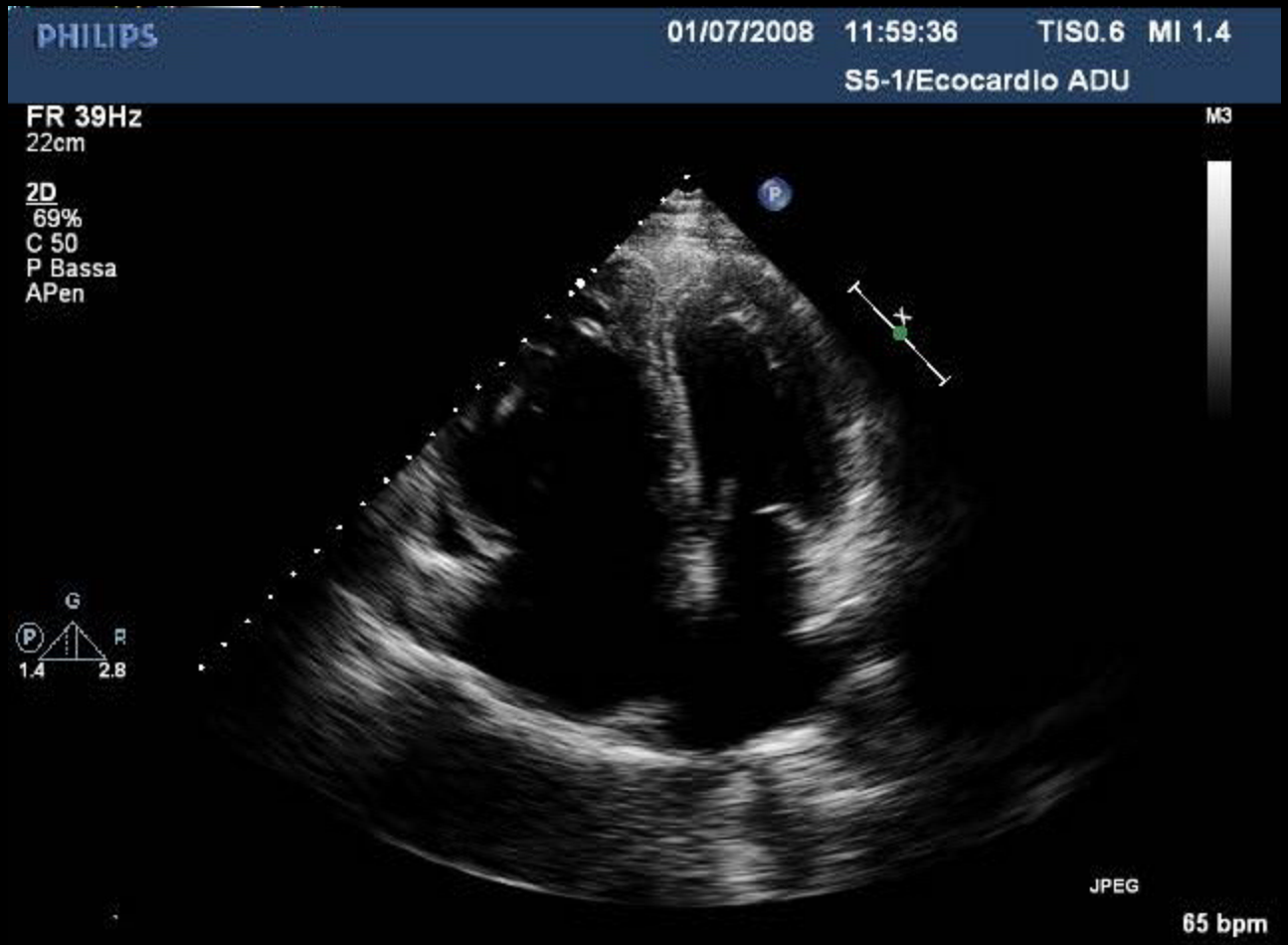
- ASD closure is indicated in patients with associated right ventricular volume overload regardless of symptoms if pulmonary vascular resistance **(PVR) is <5 Wood units**.
- The efficacy of ASD closure is uncertain but may be considered in select patients with net left to right shunt (Qp:Qs> 1.5) who have PVR ≥5 Wood units but <2/3 systemic vascular resistance **or** pulmonary artery pressure <2/3 systemic pressure
- In contrast, patients with severe irreversible pulmonary hypertension (PVR >2/3 systemic vascular resistance or pulmonary artery pressure > 2/3 systemic arterial pressure) should not undergo ASD closure

Severe irreversible pulmonary hypertension and atrial septal defect

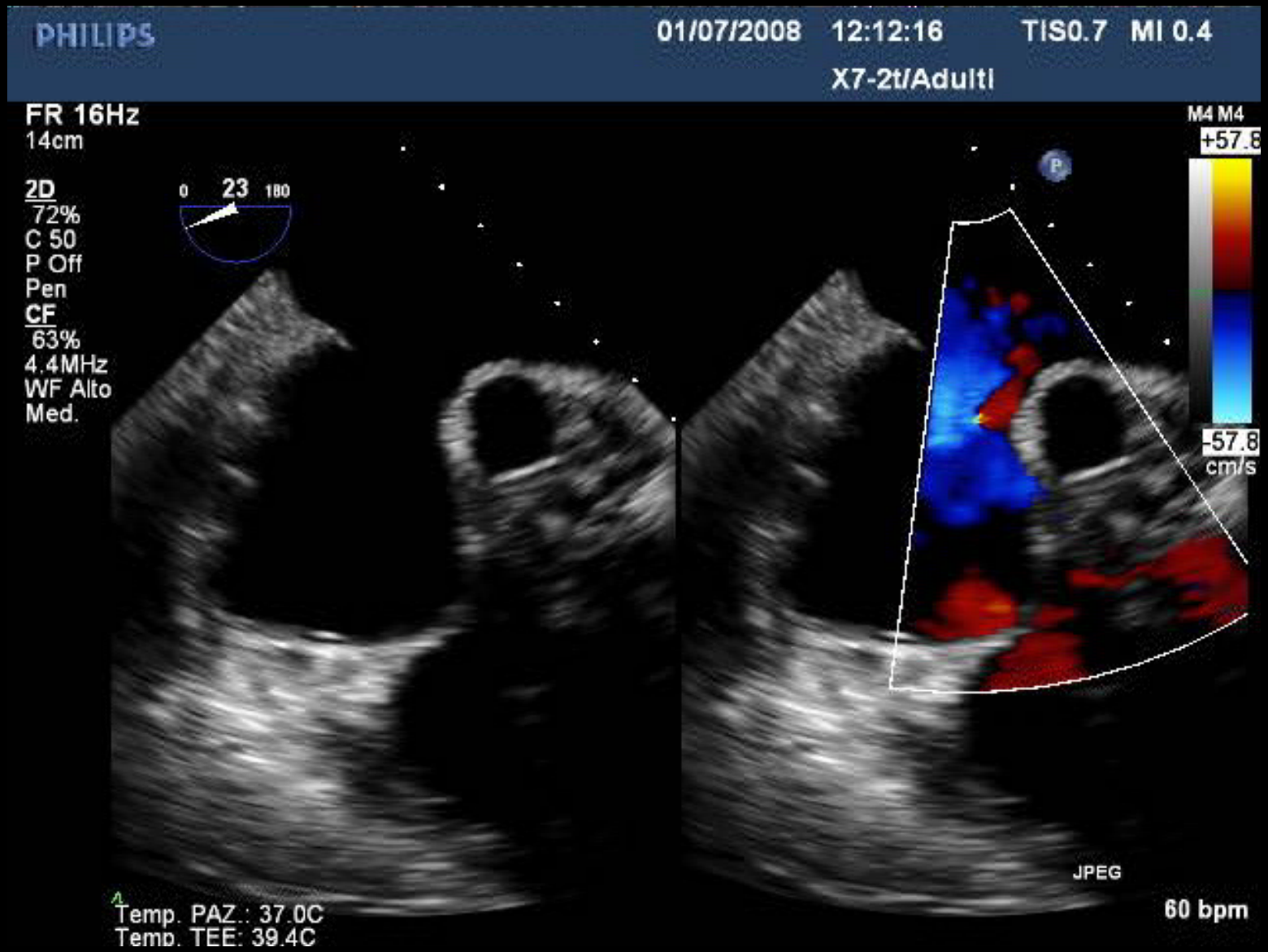
(Pulmonary vascular resistance greater than 1200 dyne sec cm-5m2 (15 Wood units)

- Not ASD closure
- ASD and maintenance of interatrial communication provides a mechanism to maintain cardiac output at the expense of desaturation and this may be advantageous. (Eisenmenger syndrome)

C.R a 60 Invio per FA e ritenzioen idrica



Cateteterismo cardiaco: PAPS 55mHg , RVP 5 U.W. Qp/QS 2,5



Br Heart J. Mar 1994; 71(3): 224–228.
PMCID: PMC483657

Natural history of secundum atrial septal defect in adults after medical or surgical treatment: a historical prospective study

D Shah, M Azhar, C M Oakley, J G F Cleland, and P Nihoyannopoulos

- *Objective*—To compare outcome in patients with medically treated secundum atrial septal defect (ASD) first diagnosed after the age of 25 with the long-term outcome in a similar group of patients after surgical closure.
- *Design*—A historical, prospective, unrandomised study.
- *Setting*—A tertiary referral centre.
- *Patients*—All patients with ASD followed up since 1955 who fulfilled the entry criteria and had reached a current age of over 45 years—that is, 34 medical and 48 surgical patients with a mean follow up of 25 years.
- *Main outcome measures*—Survival, symptoms, and complications.
- *Results*—There was no difference in survival or symptoms between the two groups and no difference in the incidence of new arrhythmias, stroke or other embolic phenomena, or cardiac failure. No patient in either group developed progressive pulmonary vascular disease.
- **Conclusion—Outcome in adults with ASD was not improved by surgical closure. Because progressive pulmonary vascular disease did not develop in any of these patients its prevention is not a reason for advising closure of ASD in adults.**

Surgical Treatment for Secundum
Atrial Septal Defects in Patients 40 Years Old
A Randomized Clinical Trial

Fause Attie, MD, FACC, Martı´n Rosas, MD, PHD, Nuria Granados, MD, Carlos Zabal, MD,
Alfonso Buendı´a, MD, Juan Caldero´n, MD

Jacc 2001,38,7.

OBJECTIVES

To examine whether surgical treatment of secundum atrial septal defects (ASDs) in patients 40 years old improves their long-term clinical outcome.

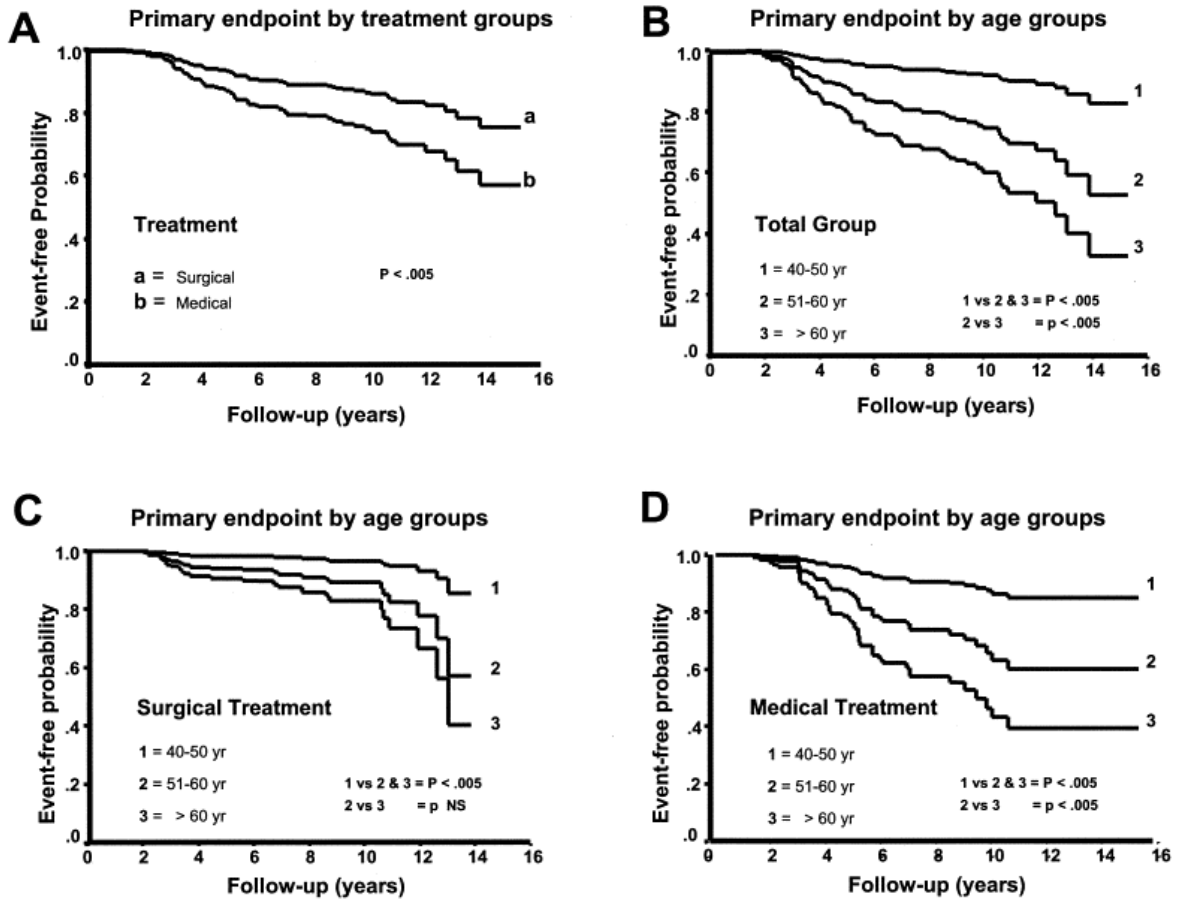
- BACKGROUND

Surgical treatment of secundum ASDs in adults 40 years old is a subject of controversy because of the perception of good long-term clinical outcomes in patients with unrepaired ASDs and the lack of data from randomized trials.

- CONCLUSIONS **Surgical closure was superior to medical treatment in improving both the composite of major cardiovascular events and overall mortality in patients 40 years old with secundum ASDs.**
- This superiority was related to the mean pulmonary artery pressure, age at diagnosis and cardiac index. **Because of the higher risk of morbidity and mortality, we believe that anatomic closure should always be attempted as the initial treatment for ASDs in adults 40 years old with pulmonary artery systolic pressure <70 mm Hg and a QP/QS output ratio 1.**7. The operation must be performed as soon as possible

Surgical treatment for secundum atrial septal defects in patients>40 years old. A randomized clinical trial.

AAttie F, Rosas M, Granados N, Zabal C, Buendía A, Calderón JJ Am Coll Cardiol. 2001;38(7):2035.



The multivariate analysis, adjusted by age at entry, mean pulmonary artery pressure and cardiac index, demonstrated significant differences between the study groups

EFFECT OF DEFECT CLOSURE ON RISK OF ATRIAL ARRHYTHMIAS

- **Incidence of atrial flutter/fibrillation in adults with atrial septal defect before and after surgery.Berger F, Vogel M, Kramer A, Alexi-Meskishvili V, Weng Y, Lange PE, Hetzer R Ann Thorac Surg. 1999;68(1):75.**

BACKGROUND: There is controversy about the benefit of surgical repair for atrial septal defect in adults, especially its effect on the incidence of supraventricular dysrhythmias, atrial flutter and fibrillation. We studied their incidence before and after operation.

METHODS: We examined surface and 24-hour Holter electrocardiograms before, early (between 3 and 7 days), and late (more than 6 months) after operation, performed at age 42.2 years (range, 18.5 to 74.9 years), in 211 adults with atrial septal defect. Patients were arbitrarily divided into three groups: age 18 to 40 years (n = 101), age 40 to 60 years (n = 83), and age more than 60 years (n = 27). All consecutive patients operated on between January 1988 and December 1996 and having a pulmonary to systemic flow ratio of 1.5:1 or greater were included in this study.

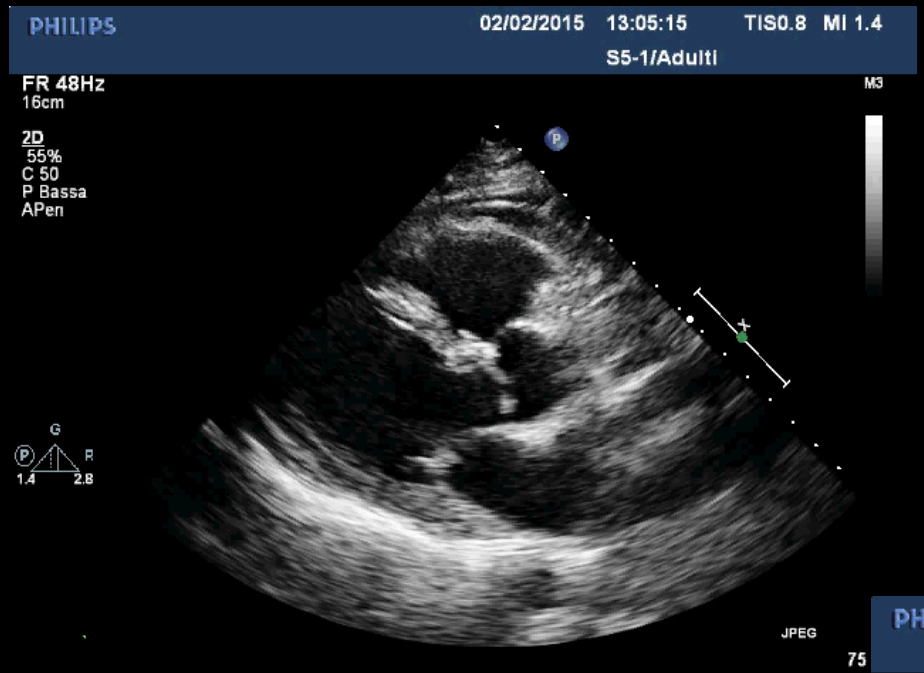
RESULTS: The age of patients without arrhythmias before or after atrial septal defect closure (39+/-13 years) was significantly lower than that of patients with flutter (54+/-12 years) or fibrillation (59+/-8 years). The incidence of atrial flutter was influenced by surgical repair as atrial flutter converted to sinus rhythm late after operation in 10 of 18 patients. However, there was no change in the incidence of atrial fibrillation before (n = 28) and after (n = 21) operation.

CONCLUSIONS: **Our data show that surgical correction of atrial septal defect leads to regression of the incidence of atrial flutter but not fibrillation**. Thus, surgical repair of atrial septal defect to abolish supraventricular tachyarrhythmias in adults is warranted, but in patients with fibrillation, it may have to be combined with a Maze operation in the future.

ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease: Executive Summary
A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines for the Management of Adults With Congenital Heart Disease

Effect of Age

- Younger patients have better long-term outcomes following surgical ASD closure than older patients , but both younger patients as well as older patients (over 40 or 60 years old) with appropriate indications benefit from ASD closure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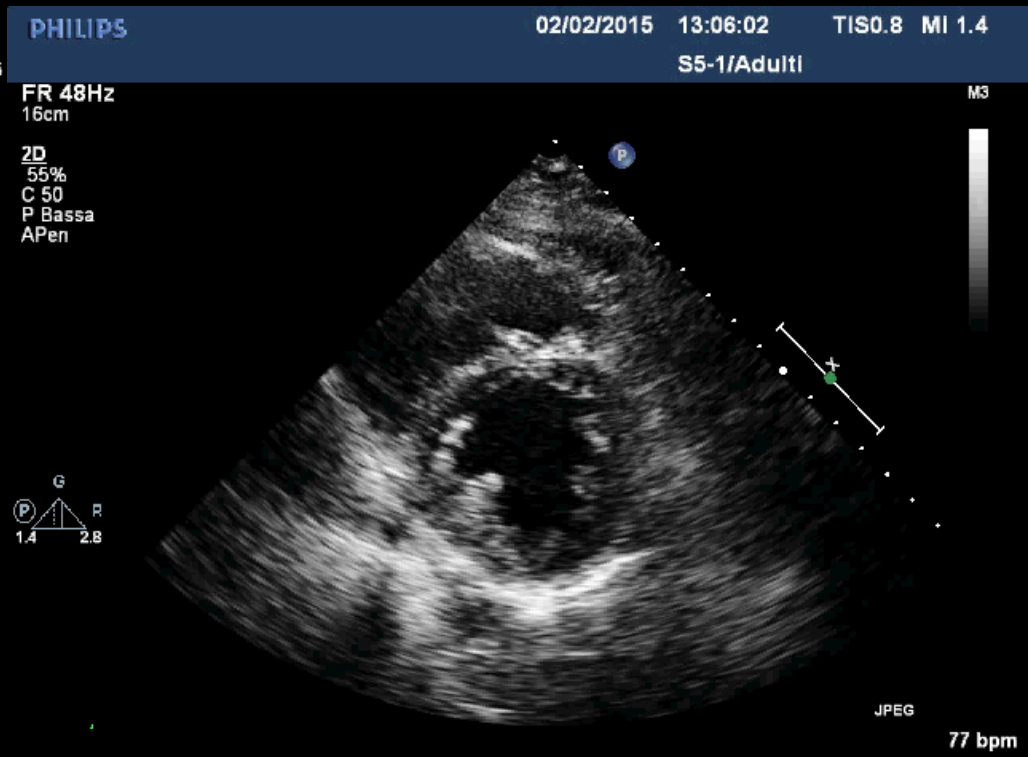
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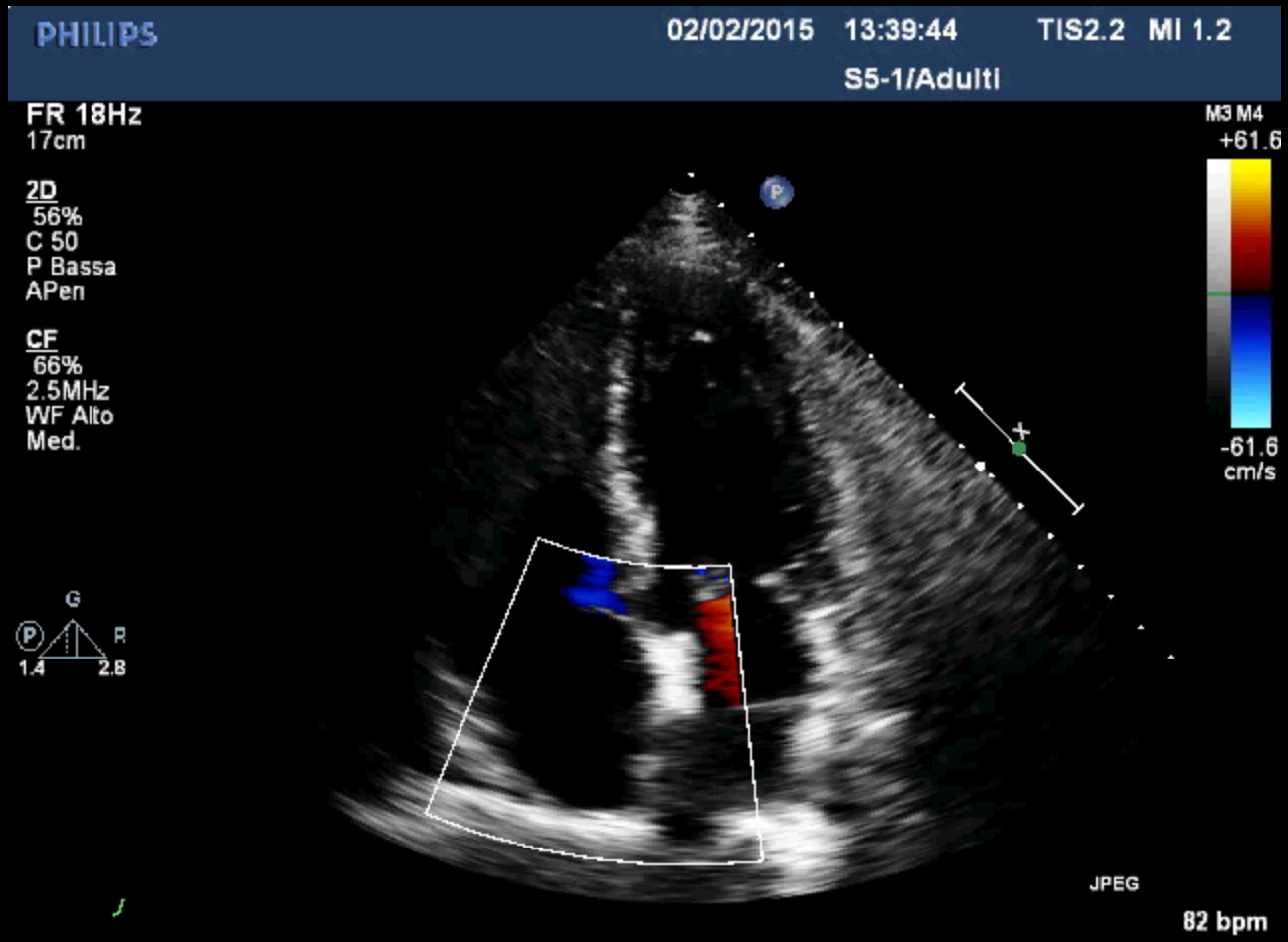
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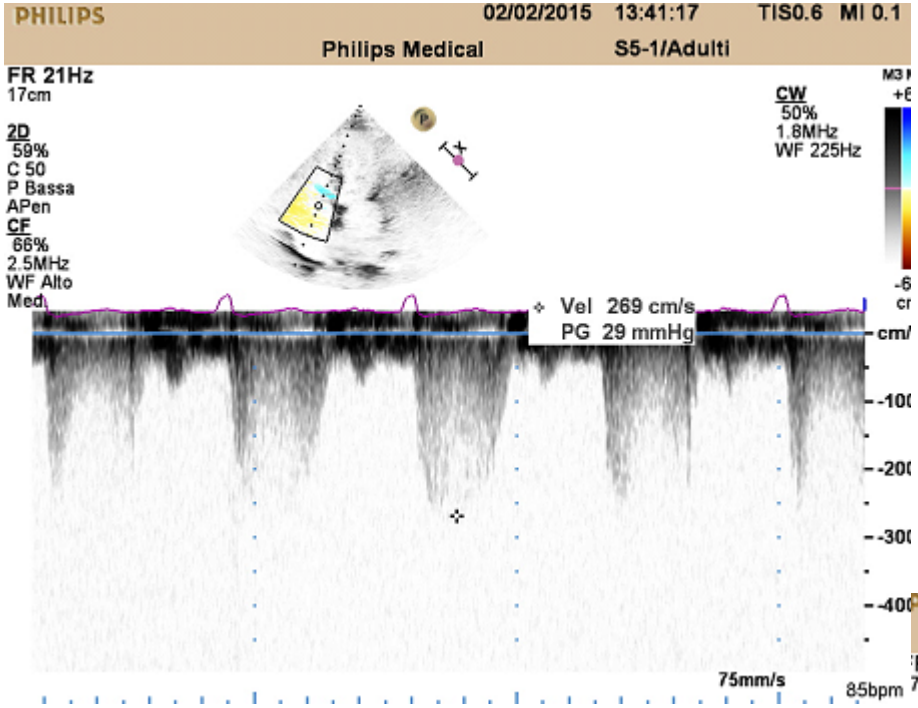
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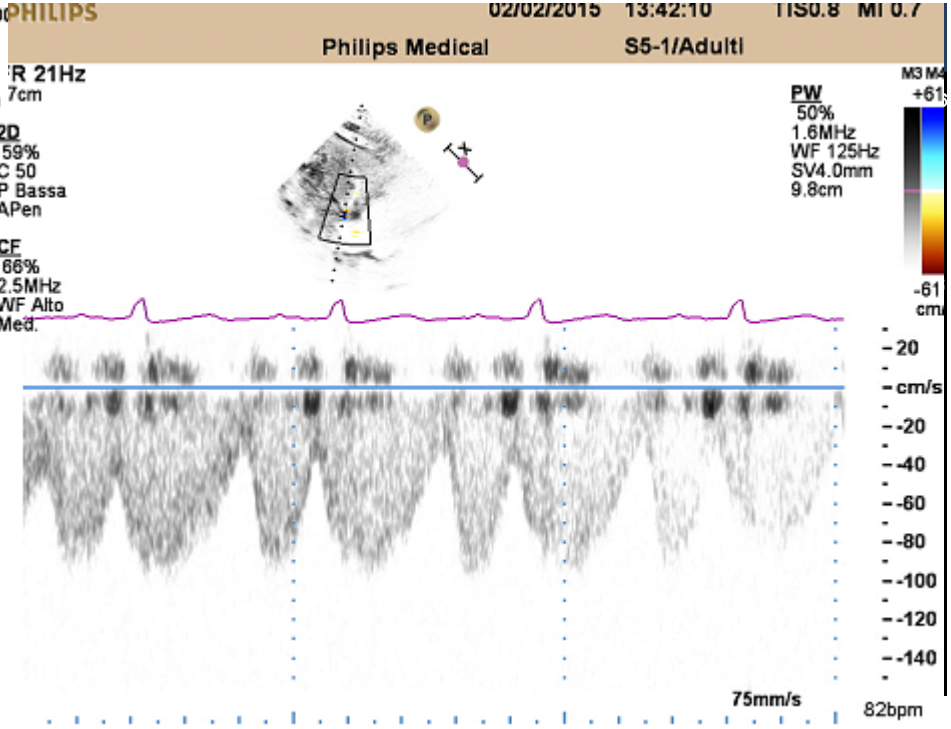
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Surgical vs percoutaneous closure of ASD

- Observational studies comparing surgical and percutaneous transcatheter closure of secundum ASD suggest that mortality rates are similar, the rate of procedural success is comparable or slightly better with surgery, and the rate of early complications and length of hospital stay is reduced with the percutaneous approach

Percutaneous versus surgical closure of secundum atrial septal defect: Comparison of early results and complication Butera, MD, PhDa,Mario Carminati, MDa, Alessandro Frigiola, MDb

Am.H.J 2006

- **Background**

  Surgical closure of atrial septal defect (ASD) provides excellent results. Given the increasing popularity of percutaneous techniques, a comparison between the 2 methods is needed.
- **Methods**

  Between December 1988 and June 2003, we performed 1284 procedures in 1268 consecutive patients with isolated secundum ASD. Five hundred and thirty-three patients underwent surgical repair of ostium secundum ASD (group A). Seven hundred and fifty-one consecutive patients underwent percutaneous ASD closure (group B). The following outcomes were studied: mortality, morbidity, hospital stay, and efficacy.
- **Conclusions**

  Percutaneous ASD closure provides, in experienced hands and in highly specialized centers, excellent results with a lower complication rate and requires a shorter stay in hospital.

Clinical implications

In our study, percutaneous closure of ASD appears to be **safer** than surgical closure; furthermore, in our analysis this technique needs a **shorter time spent in hospital**. It is also possible to speculate that the shorter postoperative convalescence after transcatheter ASD closure could lessen the **social impact** (ie, work absence).

Another important advantage is related to the lesser psychological impact of percutaneous techniques. In fact, the **absence of skin scars**, shorter hospitalization, and avoidance of admission to the intensive care unit are widely appreciated by patients and parents.

There may also be some advantages during the follow-up. First, the **absence of a scar on atrial myocardium may reduce the incidence of arrhythmias**. Secondly, **bypass surgery is complicated by a late decline in cognitive function** as shown by Selnes et al in patients undergoing CABG. Even in pediatric patients, there is some evidence that bypass surgery could be related to a slightly poorer neuropsychological outcome in the follow-up. Thirdly, from our study, it appears that, in the current era, most ASD are suitable for transcatheter closure.

Resolution of Right Heart Enlargement After Closure of Secundum Atrial Septal Defect With Transcatheter Technique

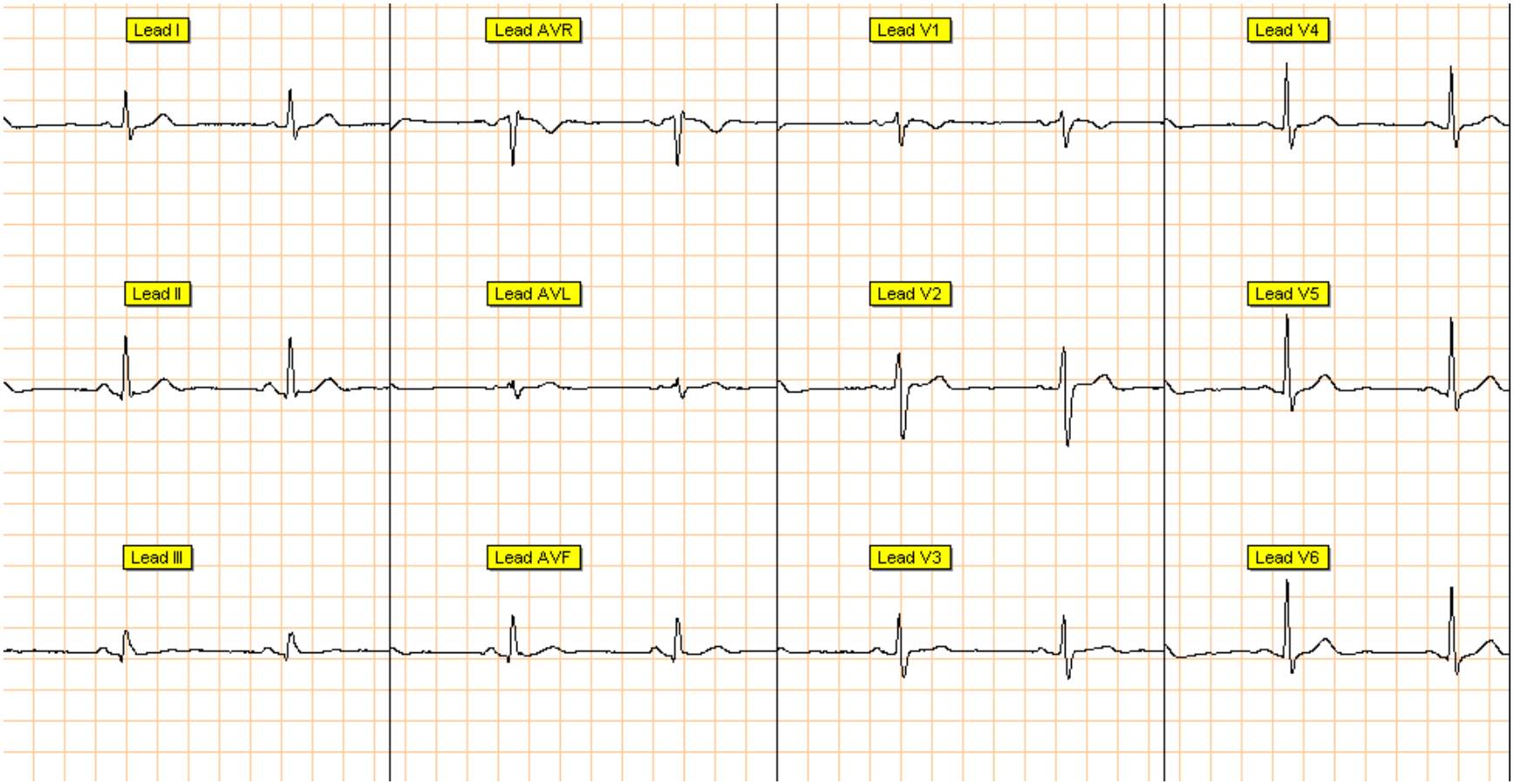
Henry W. Kort, MD, David T. Balzer, MD, Mark C. Johnson, MD

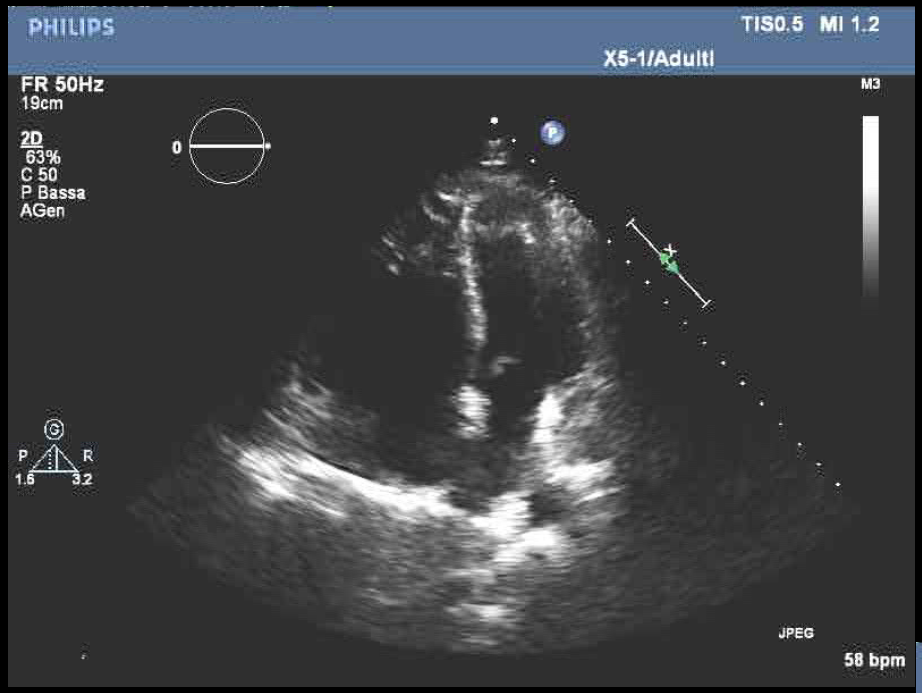
Journal of the American College of Cardiology Vol. 38, No. 5, 2001

- **OBJECTIVES** The purpose of this study was to prospectively characterize the reduction in right atrial (RA)
- area and right ventricular (RV) volume after transcatheter closure of atrial septal defect (ASD)
- and to investigate factors that may predict magnitude of resolution in right heart enlargement.
- **METHODS** Transthoracic echocardiography was performed in 38 patients undergoing transcatheter. closure of ASD. The RA area and RV volume were measured prior (n 38), within 24 hours (n 37), at 3 to 6 months (n 24), at 12 months (n 20) and at 24 months (n 10) after closure of ASD. the study group was compared to a control group of 19 patients with structurally normal hearts.

- **CONCLUSION Closure of secundum ASD results in decreased indexed RV volume comparable to that in control subjects at 24 months following closure. Indexed RA area remains increased compared to that in control subjects but does decrease over time. Decrease in RA area is inversely proportional to age at time of ASD closure. Long-term follow-up is required to evaluate the clinical impact of persistently increased RA size**.

MT donna 62 anni
invio per iter operatorio
(adenoma sella turcica)

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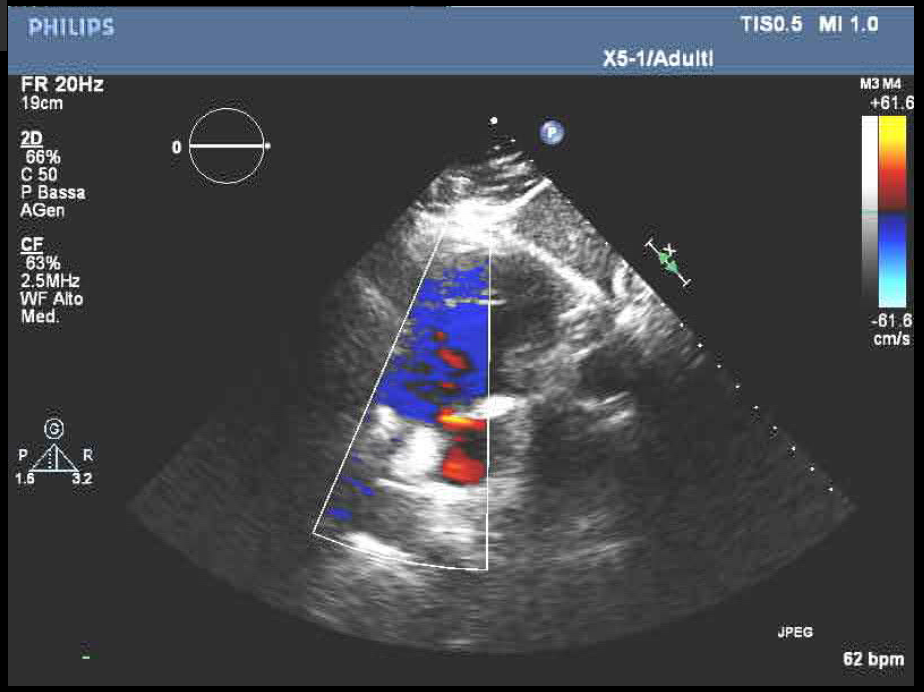
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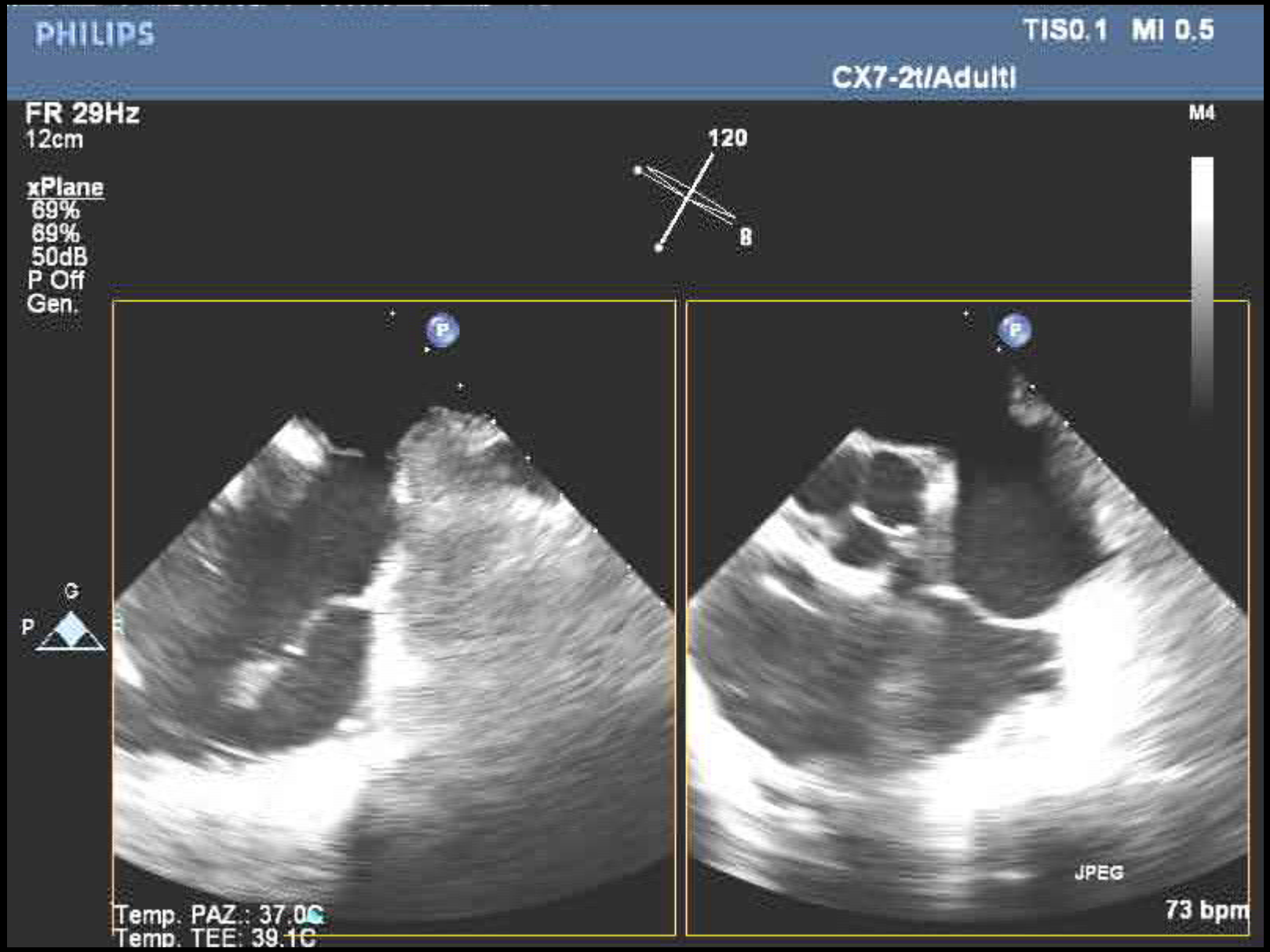
62 bpm

Problema : paziente non accetta intervento chirurgico valutata opzione interventistica

- Distinguere tra Ostium 2 alto / difetto tipo seno-venoso
- Importante localizzare bene la sede del difetto e la fattibilità della chiusura percutanea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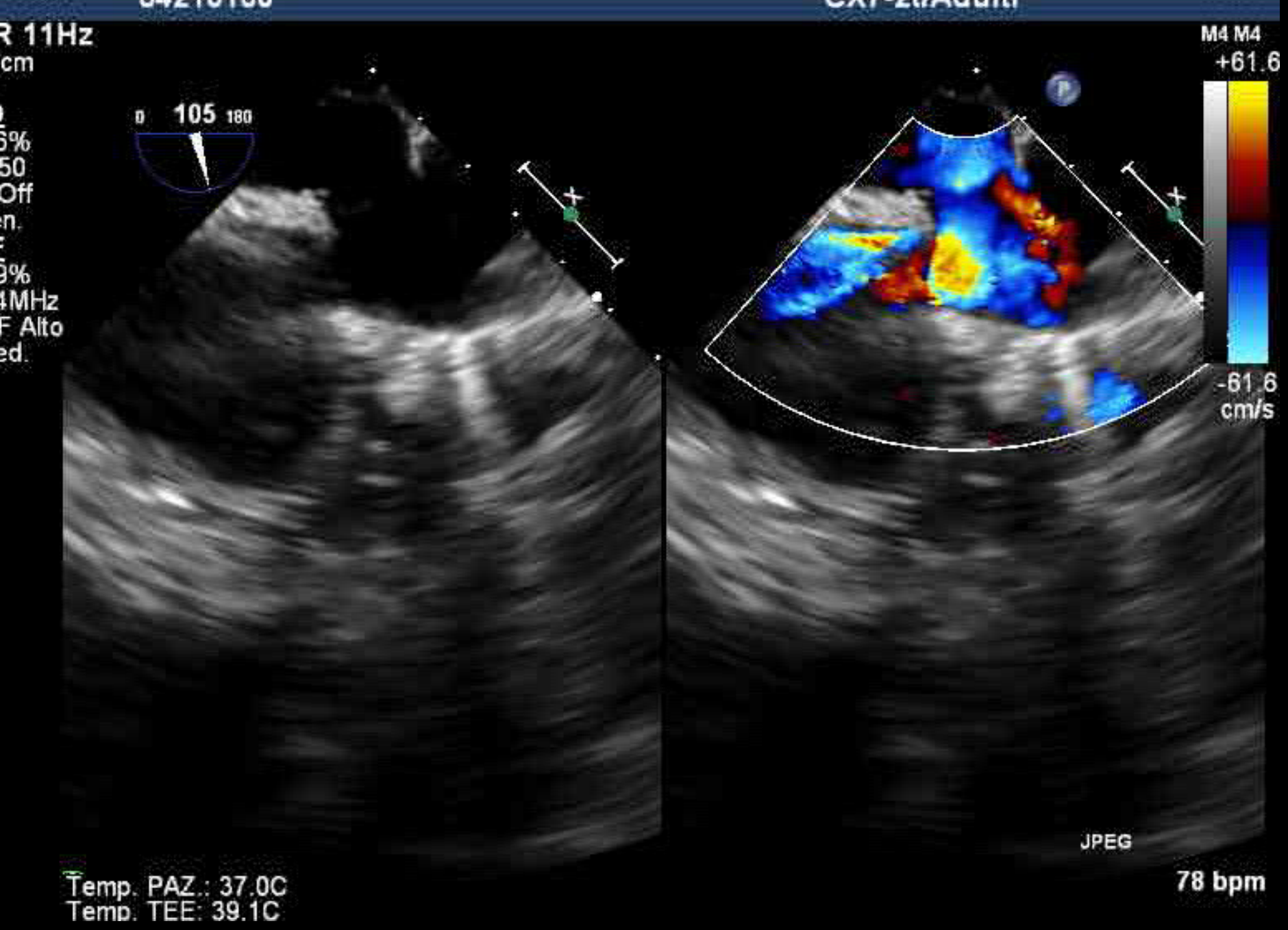
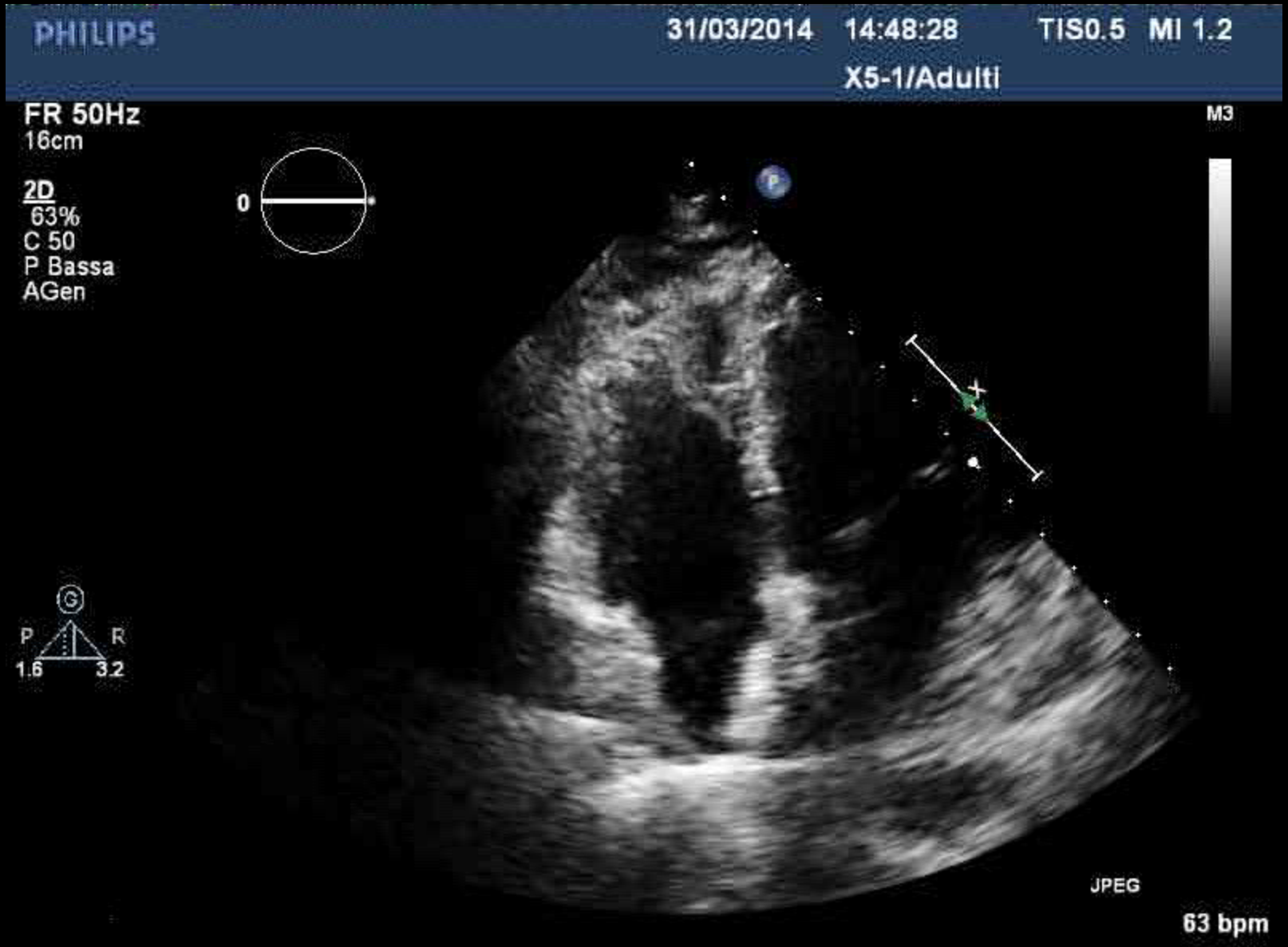


Difetto tipo Seno-Venoso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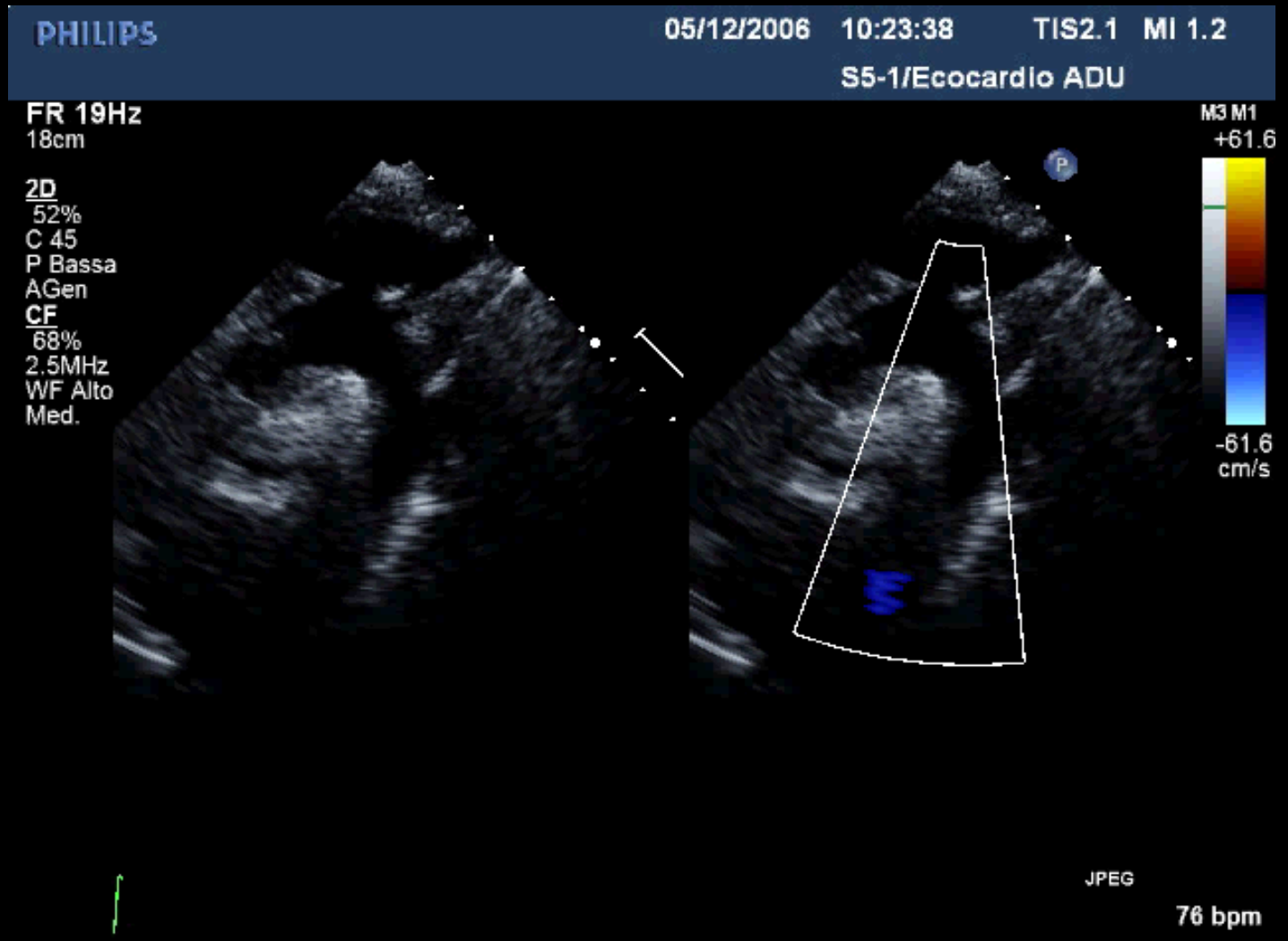
DIA dell'adulto

Pressioni e resistenze polmonari
Localizzazione DIA
Dimensioni DIA
Fattibilità chiusura percutanea
Escludere la presenza di DPAP

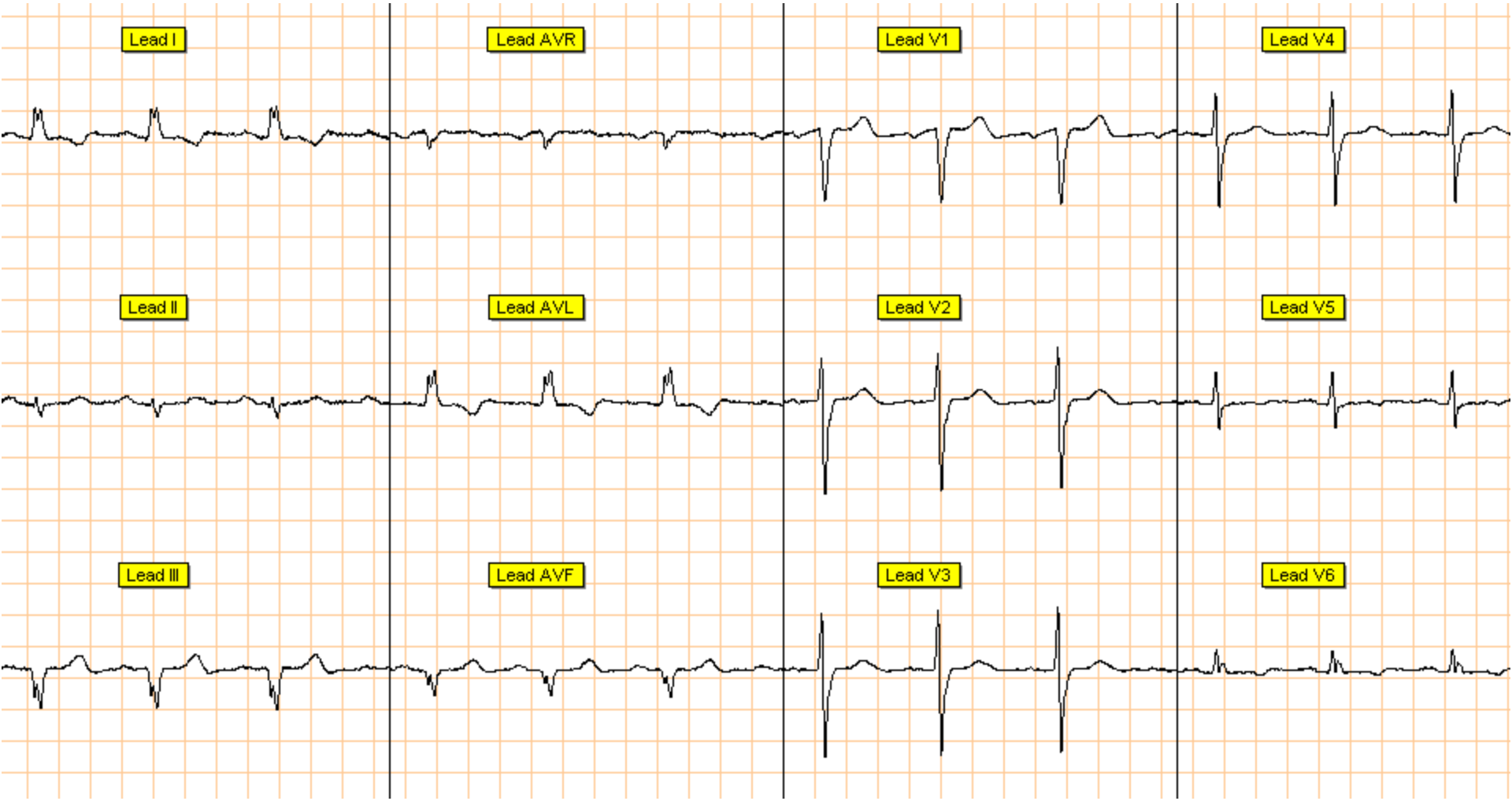
Escludere la presenza di DPAP



Patologie rare e complesse

Paziente di 68 anni ,maschio

- Iperteso in trattamento
- Alcolinizzazione nodulo tiroideo 10aa prima
- Esegue visita per iter pre-operatorio
  (TURP)
- Asintomatico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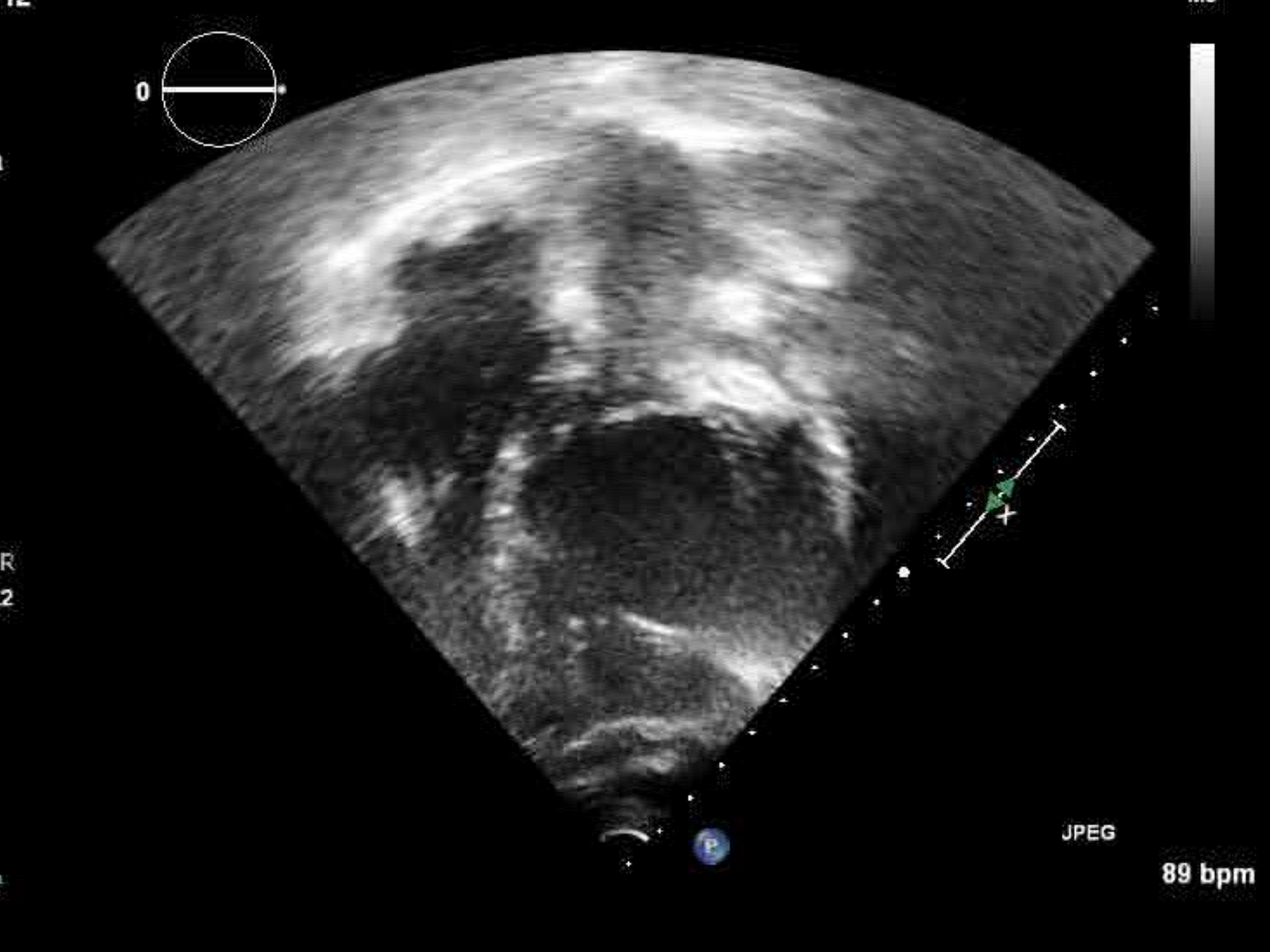
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Trasposizione dei grandi Vasi

Storia naturale D-TGA

(Normale concordanza AV/ Discordanza V/A)

- In assenza di adeguato tratttamento i pazienti affetti muoiono entro l'anno di vita.

  Il 30 percento nella prima settimana di vita,
  il 50 percento nel primo mese

Storia naturale della L-TGA (trasposizione corretta:Discordanza A/V e V/A)

- L'incidenza annuale è stata stimata in circa 1/33.000 nati vivi e la malattia rappresenta circa lo 0,05% di tutte le cardiopatie congenite
- **Il quadro clinico e l'età di esordio dipendono dalle malformazioni presenti.** (VSD sono presenti nel 70 to 80 % pazienti con L-TGA**.** DIV,SPO,Ostruzione all'efflusso polmonare nel 30 - 60 % ,Insuff. Tricuspidale (systemic atrioventricular) > 90% della L-TGA)
- La Trasp-corretta può esitare nel progressivo rigurgito delle valvole atrioventricolari e nell'insufficienza del ventricolo sistemico
- Il rischio di ricorrenza tra i fratelli è 2,6-5,2%.
- In presenza di anomalie del sistema di conduzione, può essere **indicato l'impianto di un pacemaker**, ad esempio per la presa in carico del blocco atrioventricolare di secondo o terzo grado di tipo avanzato
- Trattamento medico dello scompenso.

Storia naturale L-TGA

- < 20 percent delle L-TGA si presentano come forma isolata
- Il Blocco AV completo e' l'aritmia piu' frequente nella L-TGA e si manifesta con bradicardia, astenia , ridotta tolleranza allo sforzo.

Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study.

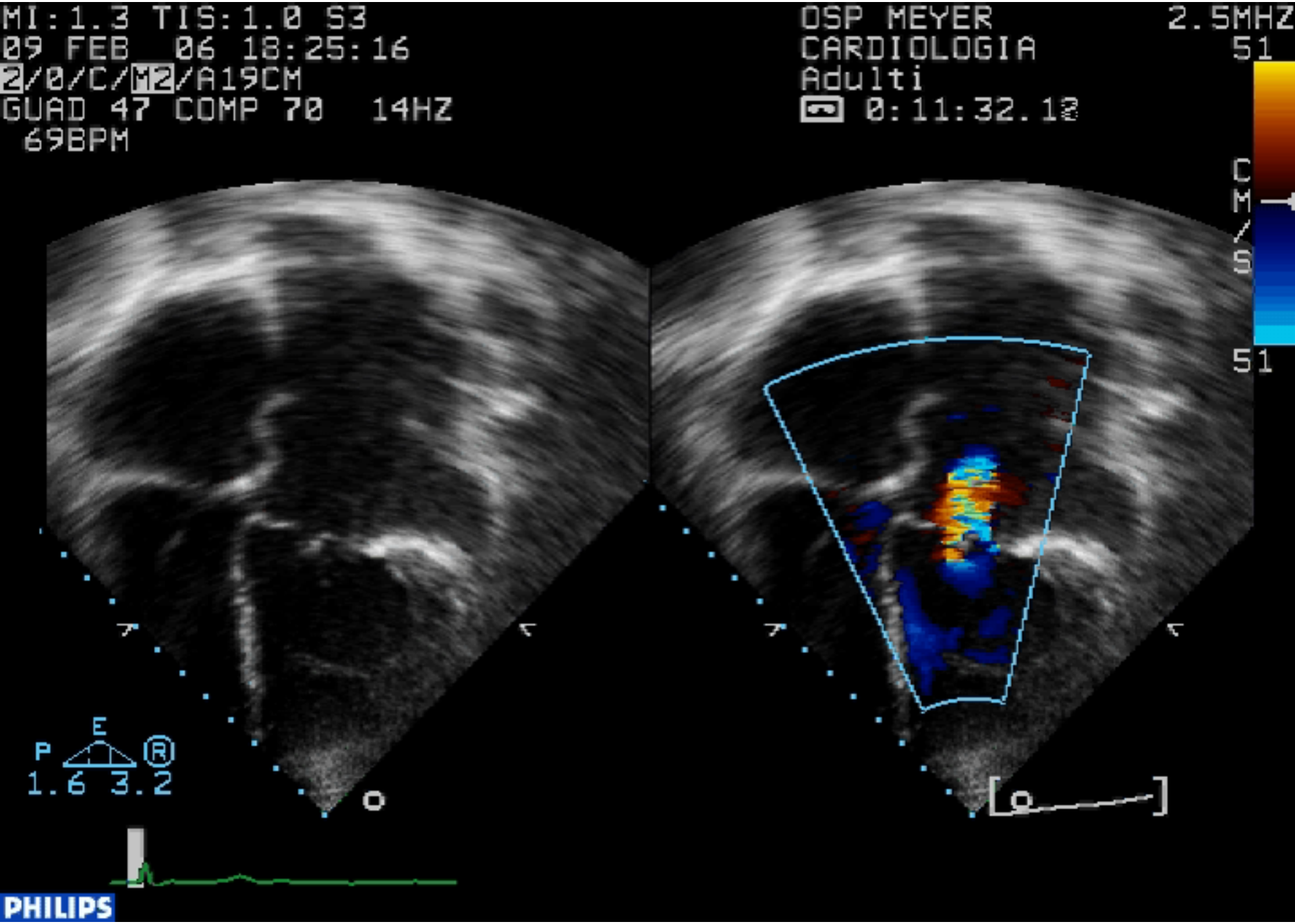
Graham TP Jr,, Sutherland JL

J Am Coll Cardiol. 2000;36(1):255.

OBJECTIVES: The purpose of this study was to determine long-term outcome in adults with congenitally corrected transposition of the great arteries (CCTGA),

BACKGROUND: Patients with CCTGA have the anatomical right ventricle as their systemic pumping chamber, with ventricular dysfunction and CHF being relatively common in older adults.

CONCLUSIONS: Patients with CCTGA **are increasingly subject to CHF with advancing age**; this complication is extremely common by the fourth and fifth decades. **Tricuspid (systemic atrioventricular) valvular regurgitation** is strongly associated with RV (anatomical right ventricle connected to aorta in CCTGA patients; systemic ventricle in CCTGA) dysfunction and CHF; whether it is causative or a secondary complication remains speculative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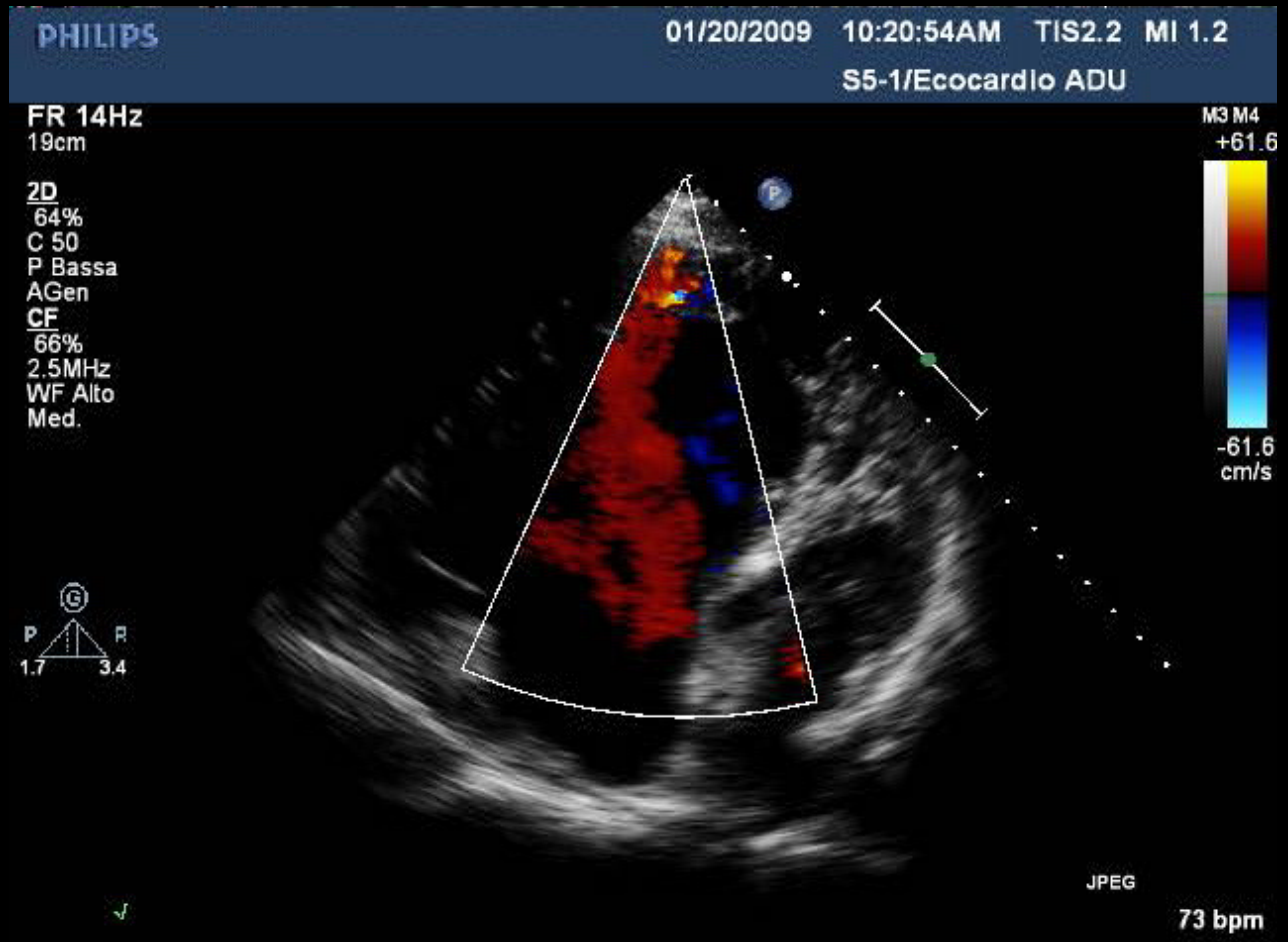
Anomalia di Ebstein

Clinical presentation of Ebstein’s anomaly

- The clinical presentation of Ebstein's anomaly varies widely, ranging from symptomatic neonate to the asymptomatic adult.
- In general, **symptoms vary according to the anatomic variation** (ie, the degree of displacement and the functional status of the tricuspid-like leaflets).
- Patients **with mild apical displacement** and mild dysfunction of the tricuspid valve may remain asymptomatic until an advanced age.
- In contrast, patients **with marked tricuspid leaflet displacement** or abnormal leaflet attachment may have **severe regurgitation** and **right-sided heart failure**, elevated right atrial pressures, and **significant right-to-left interatrial shunting**. The anomaly may be fatal if severe cardiomegaly and heart failure are present .
- Patients with Ebstein's anomaly who have an interatrial communication are **at risk for paradoxical embolization, brain abscesses, and sudden death.**

aa45, ictus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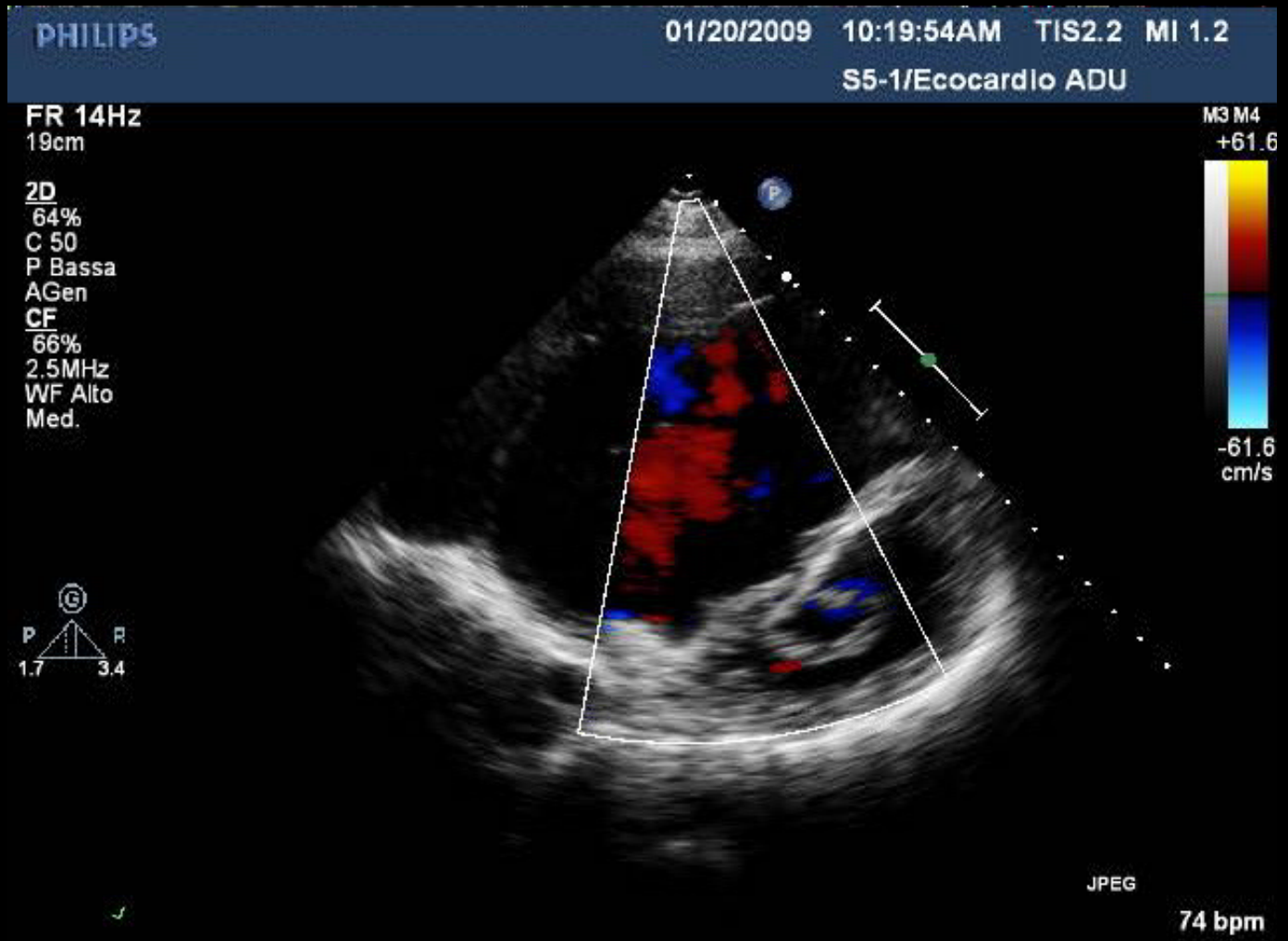
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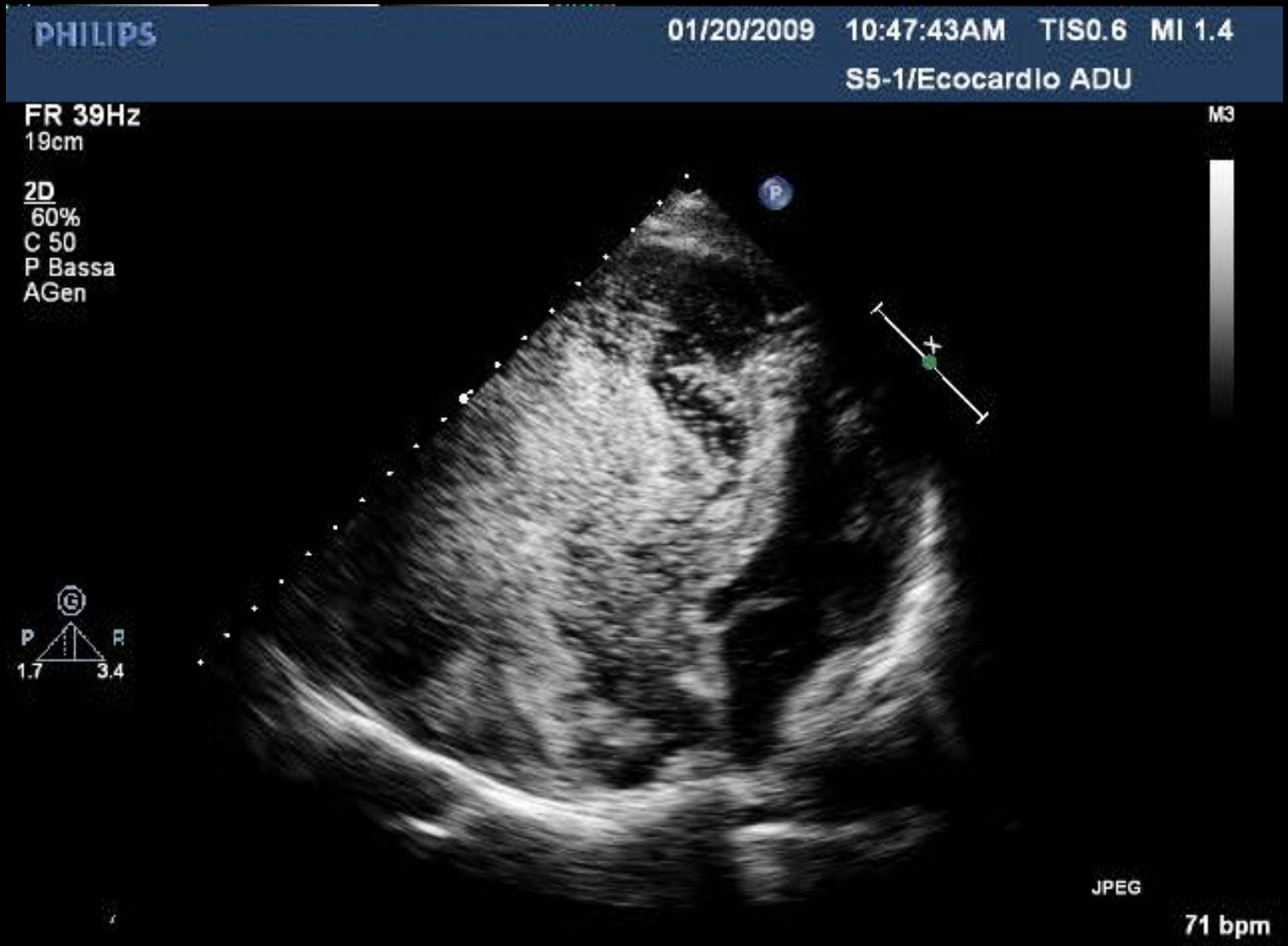
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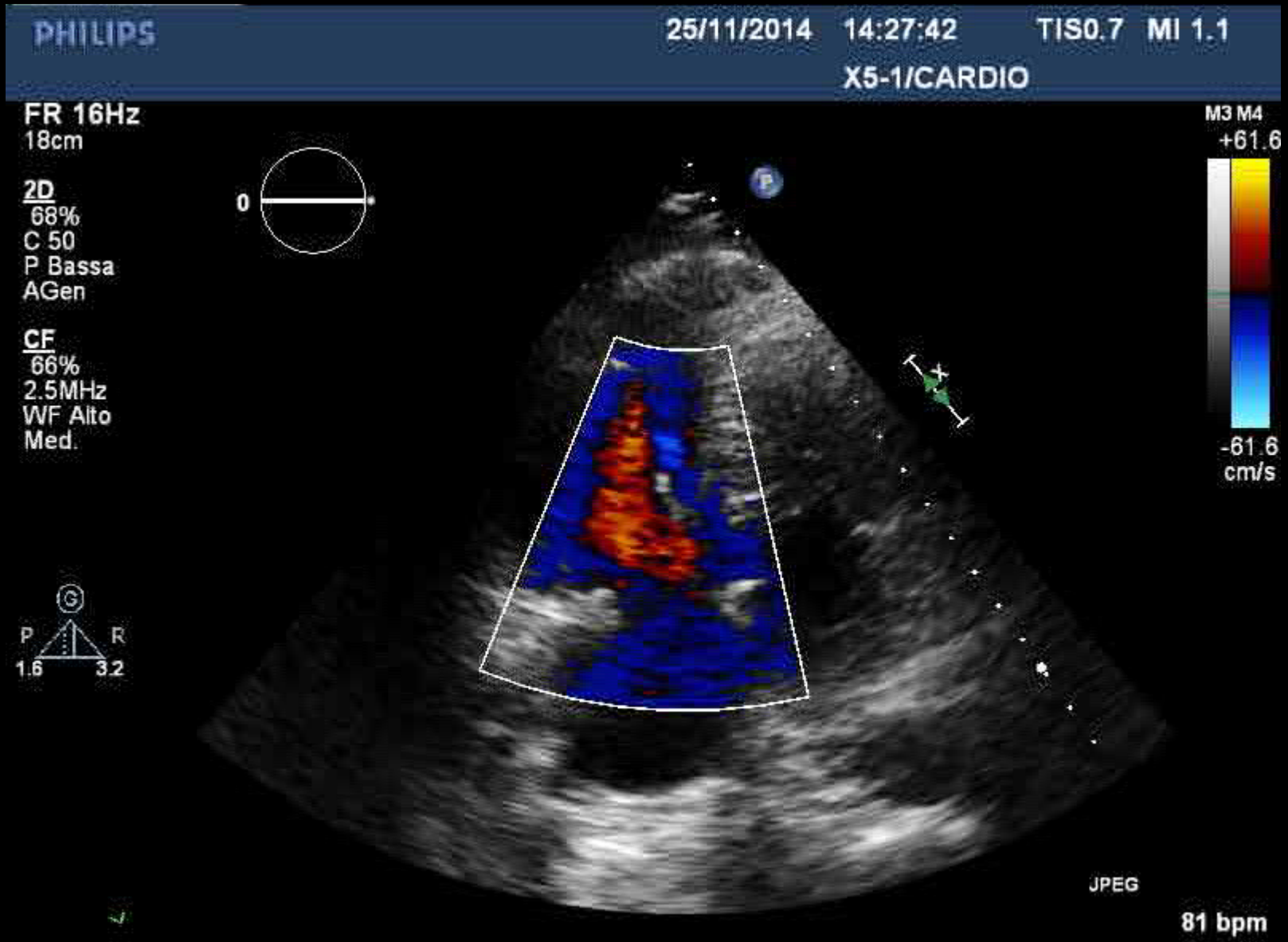
Reccomendations for Ebstein

- Surgical tricuspid valve re-repair or replacement is recommended for symptoms, severe tricuspid regurgitation with progressive RV dilation or systolic dysfunction or arrhythmias
- Acyanotic women with Ebstein's anomaly usually tolerate pregnancy, provided that sinus rhythm is maintained.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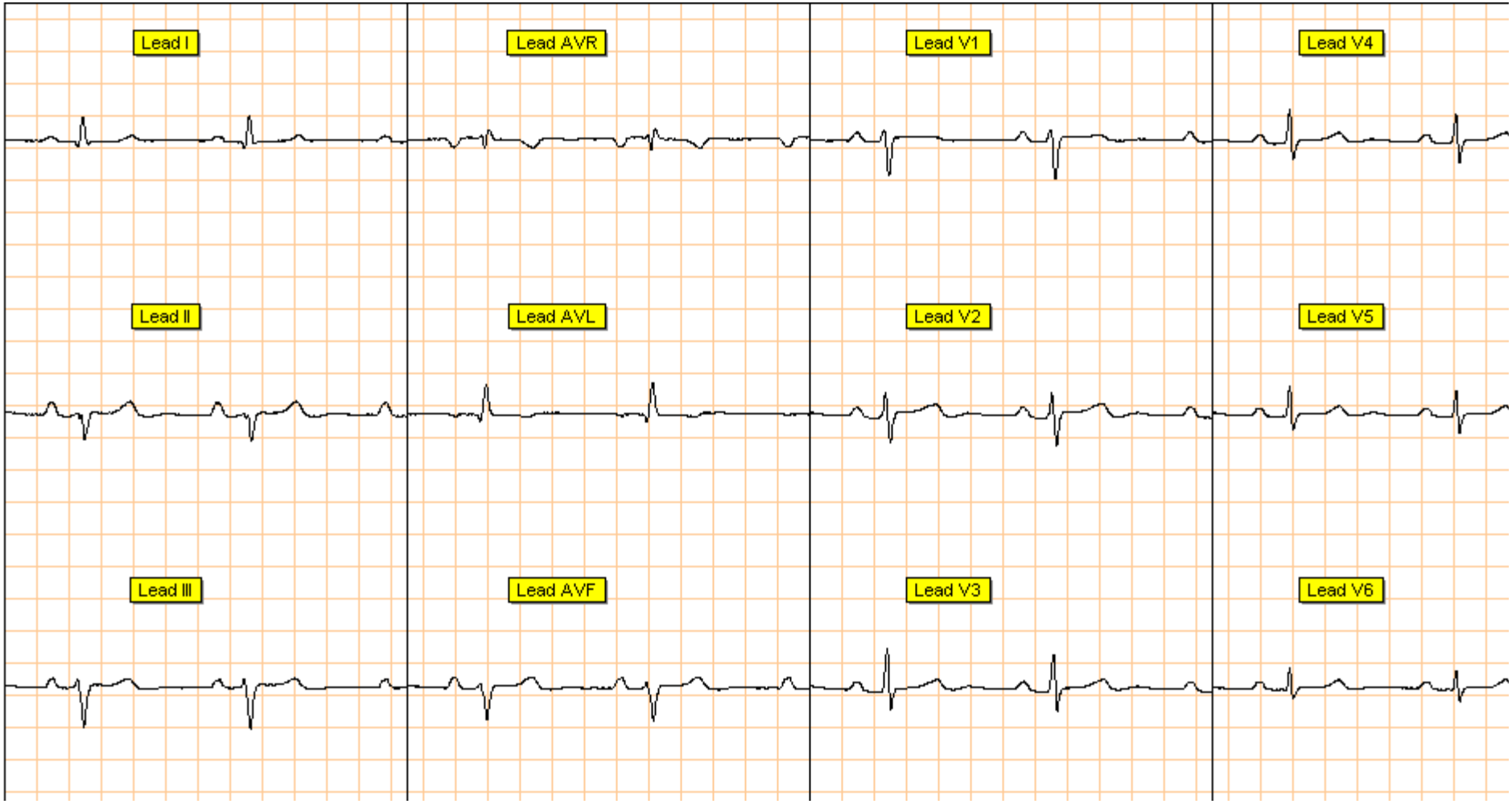
18cm
2D
68%
C 50
P Bassa
AGen
CF
66%
2.5MHz
WF Alto
Med.
0
M3 M4
+61.6
-61.6
cm/s
G
P
R
1.6
3.2
JPEG
81 bpm

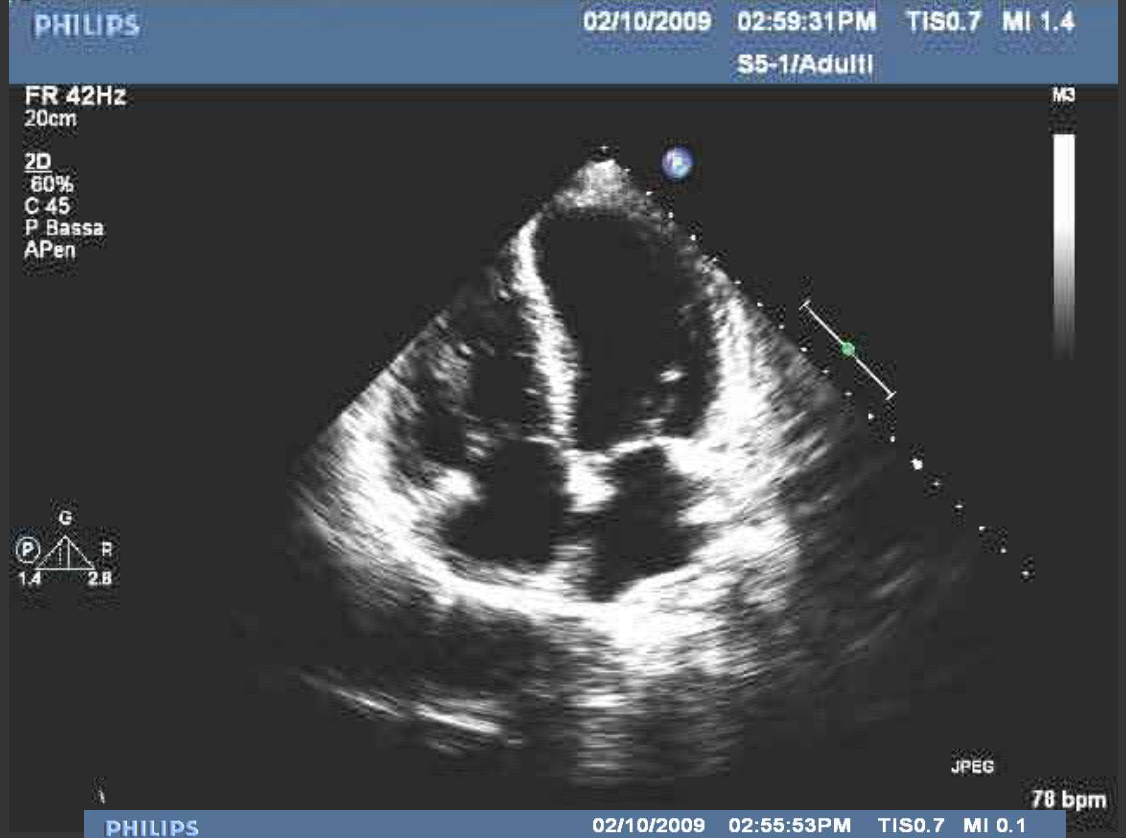
Stenosi polmonare

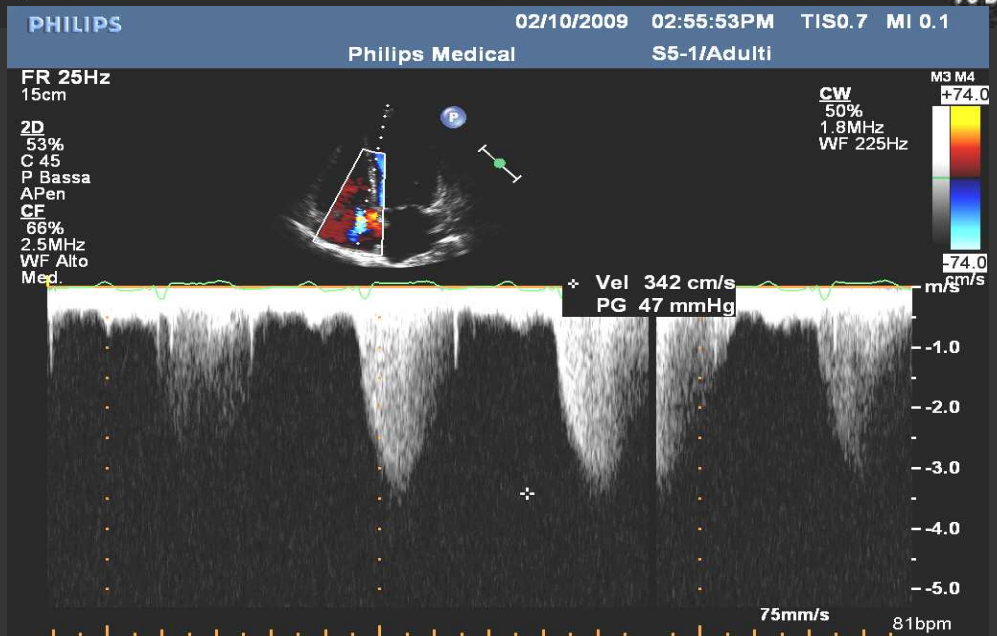
Clinical manifestations and diagnosis of pulmonic stenosis in adults

- Pulmonic stenosis in children is usually associated with a **benign clinical course.**
- Survival into adulthood is common in patients with pulmonic stenosis
- **Calcification of the valve with age** may result in decreased valve mobility and increased obstruction
- The patient who is initially asymptomatic may begin to experience symptoms that vary from **mild exertional dyspnea** to signs and **symptoms of right heart failure**
- The severity of pulmonic stenosis is classified by major society guidelines using the following criteria (mild <3m/sec,moderate ,severe > 4m/s)
- **Pregnancy safe** for mild.moderate stenosis
- **Baloon valvotomy for severe stenosis or moderate sintomatic**

Paz 48a, femmina , malattia connettivo, Ridotta capacità funzionale

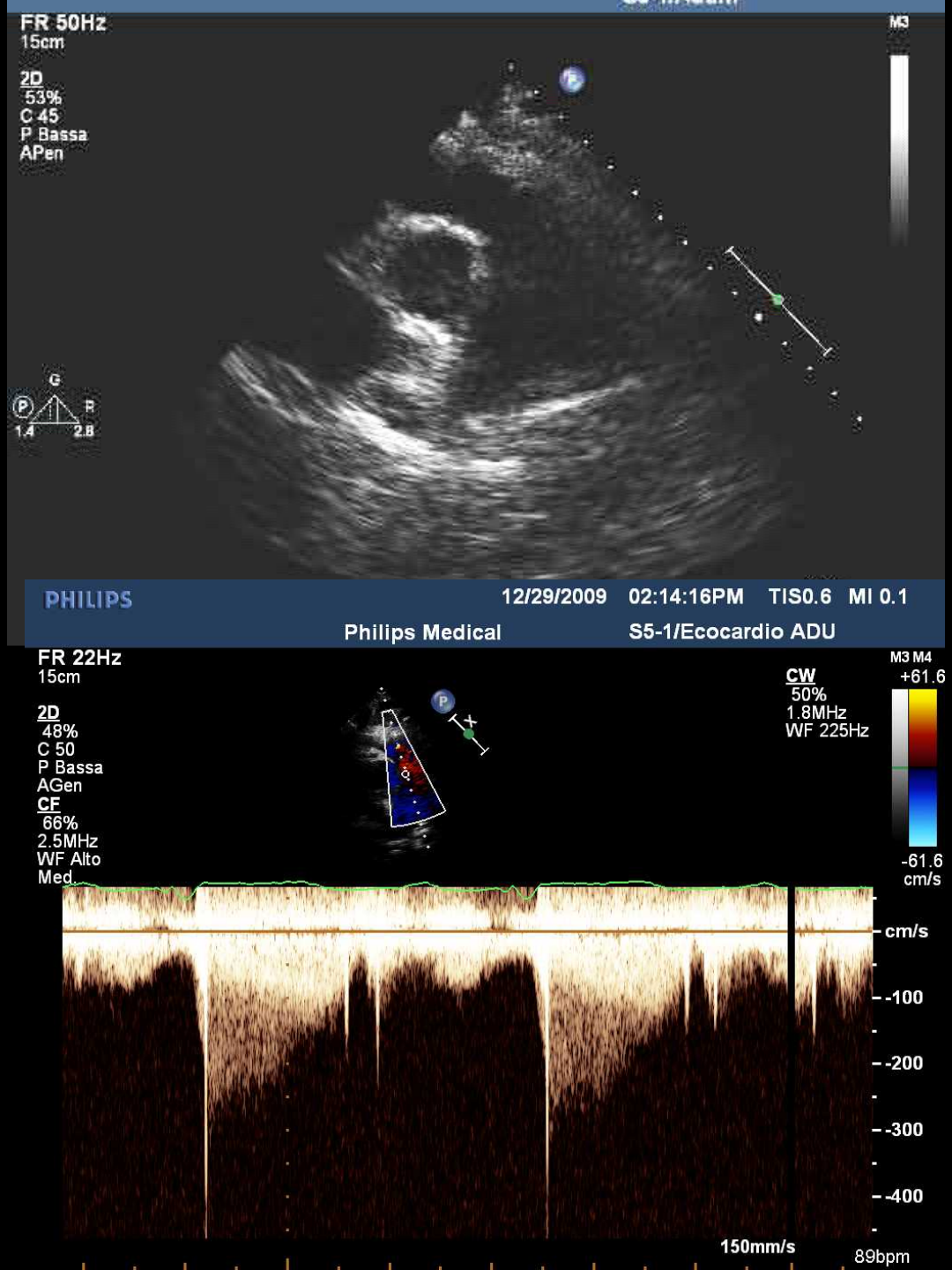



PHILIPS
02/10/2009 02:59:31PM TIS0.7 MI 1.4
S5-1/Adulti
FR 42Hz
20cm
2D
60%
C 45
P Bassa
APen
M3
G
P
R
1.4
2.8
JPEG
78 bpm

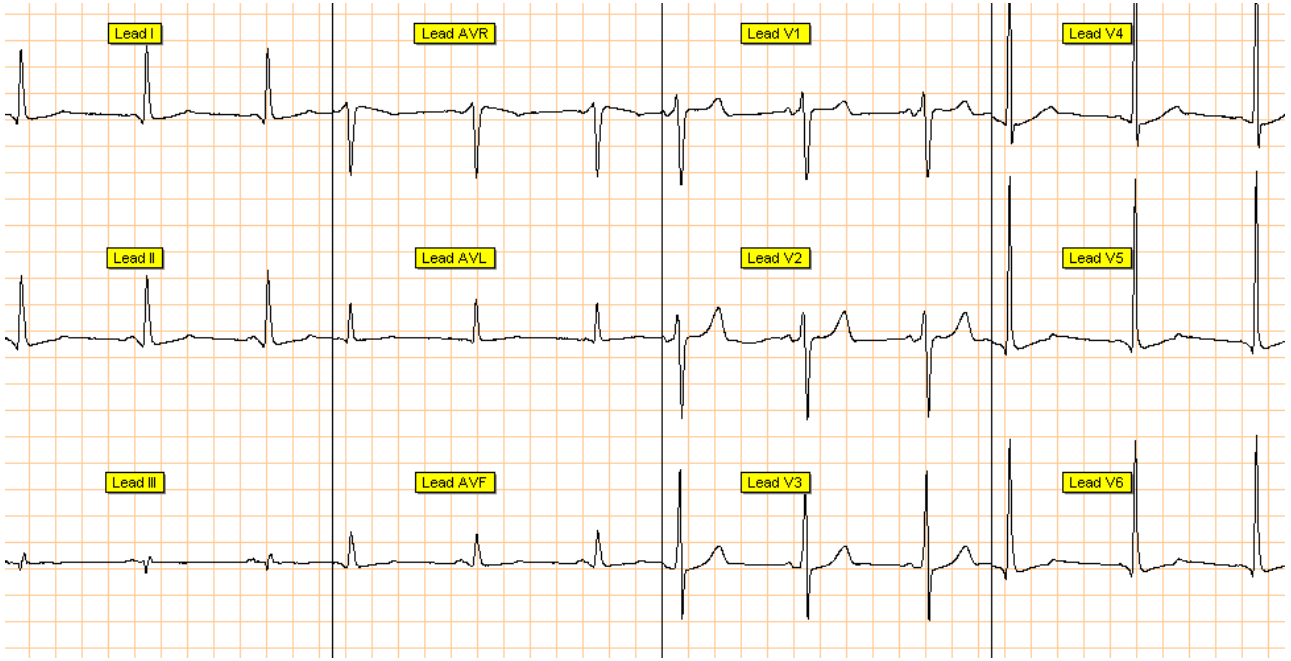

PHILIPS
02/10/2009 02:55:53PM TIS0.7 MI 0.1
Philips Medical
S5-1/Adulti
FR 25Hz
15cm
2D
53%
C 45
P Bassa
APen
CF
66%
2.5MHz
WF Alto
Med.
CW
50%
1.8MHz
WF 225Hz
M3 M4
+74.0
-74.0
cm/s
Vel 342 cm/s
PG 47 mmHg
m/s
-1.0
-2.0
-3.0
-4.0
-5.0
75mm/s
81bpm

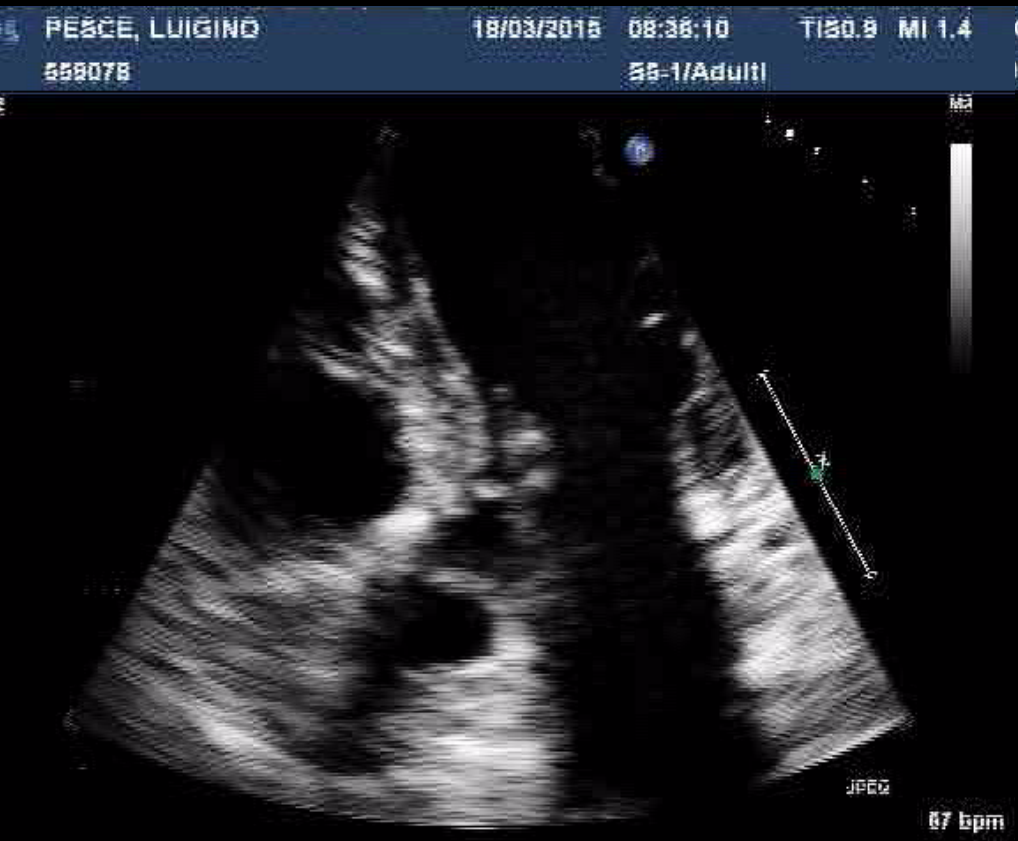
Considerata per studio emodinamico in vista di trattamento con Antagonisti dell’endotelina

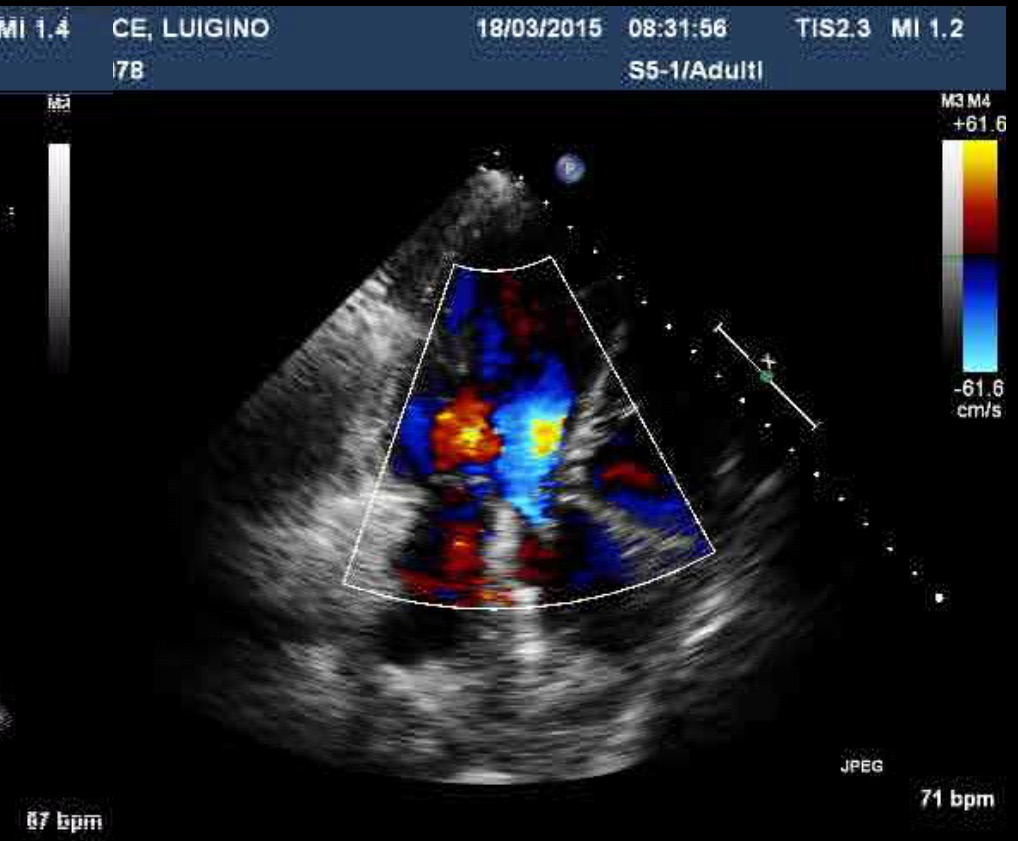
Al cateterismo riscontro di valori pressori polmonari normali e normali resistenze polmonari


FR 50Hz
15cm
2D
53%
C 45
P Bassa
APen
M3
G
P
R
1.4
2.8
PHILIPS
12/29/2009 02:14:16PM TIS0.6 MI 0.1
Philips Medical
S5-1/Ecocardio ADU
FR 22Hz
15cm
2D
48%
C 50
P Bassa
AGen
CF
66%
2.5MHz
WF Alto
Med
CW
50%
1.8MHz
WF 225Hz
M3 M4
+61.6
-61.6
cm/s
cm/s
-100
-200
-300
-400
150mm/s
89bpm

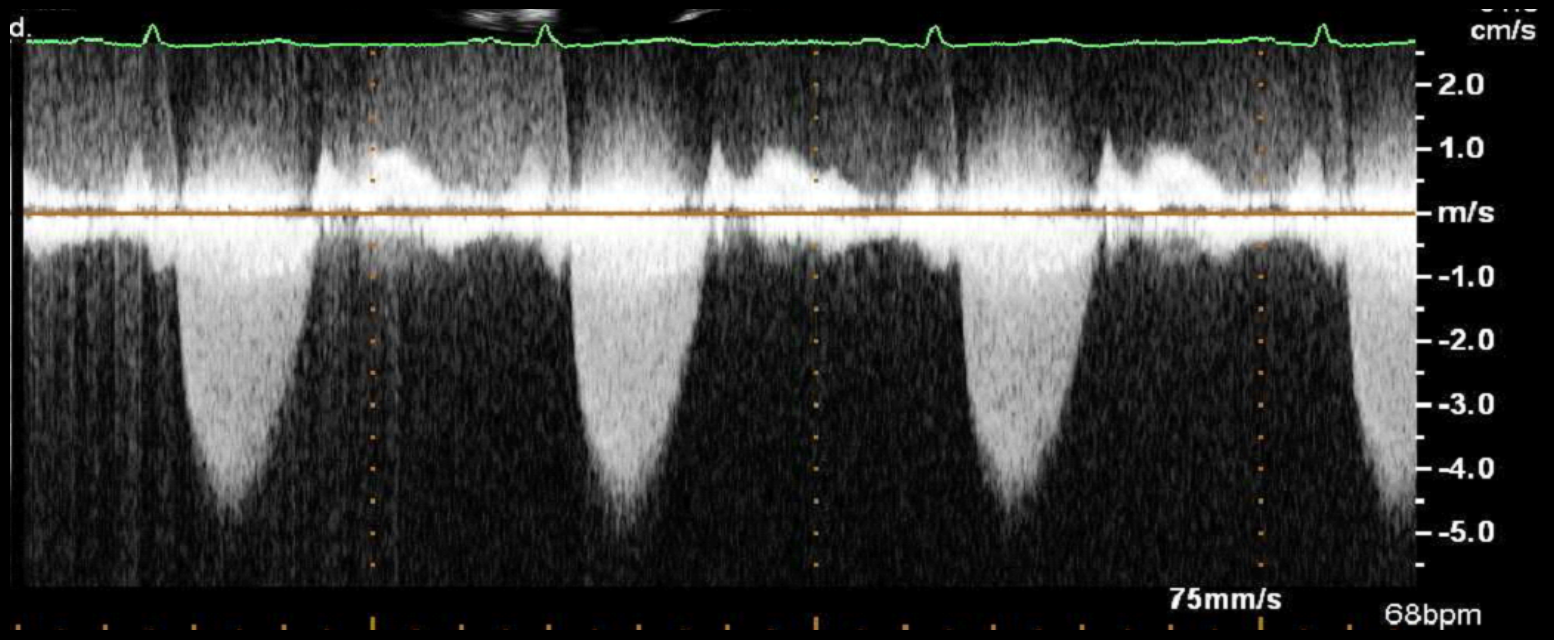
aa. 65 inviato da oncologo per chemioterapia (linfoma)
Non precedenti cardiologici
PAO 130/70 Soffio sistolico 2-3/6 basale.




PESCE, LUIGINO
559078
18/03/2015
08:36:10
TIS0.9 MI 1.4
S5-1/Adult
JPEG
67 bpm


CE, LUIGINO
78
18/03/2015
08:31:56
TIS2.3 MI 1.2
S5-1/Adult
M3 M4
+61.6
-61.6
cm/s
JPEG
71 bpm

G.Max 80mmhg
Gm 50mmHg

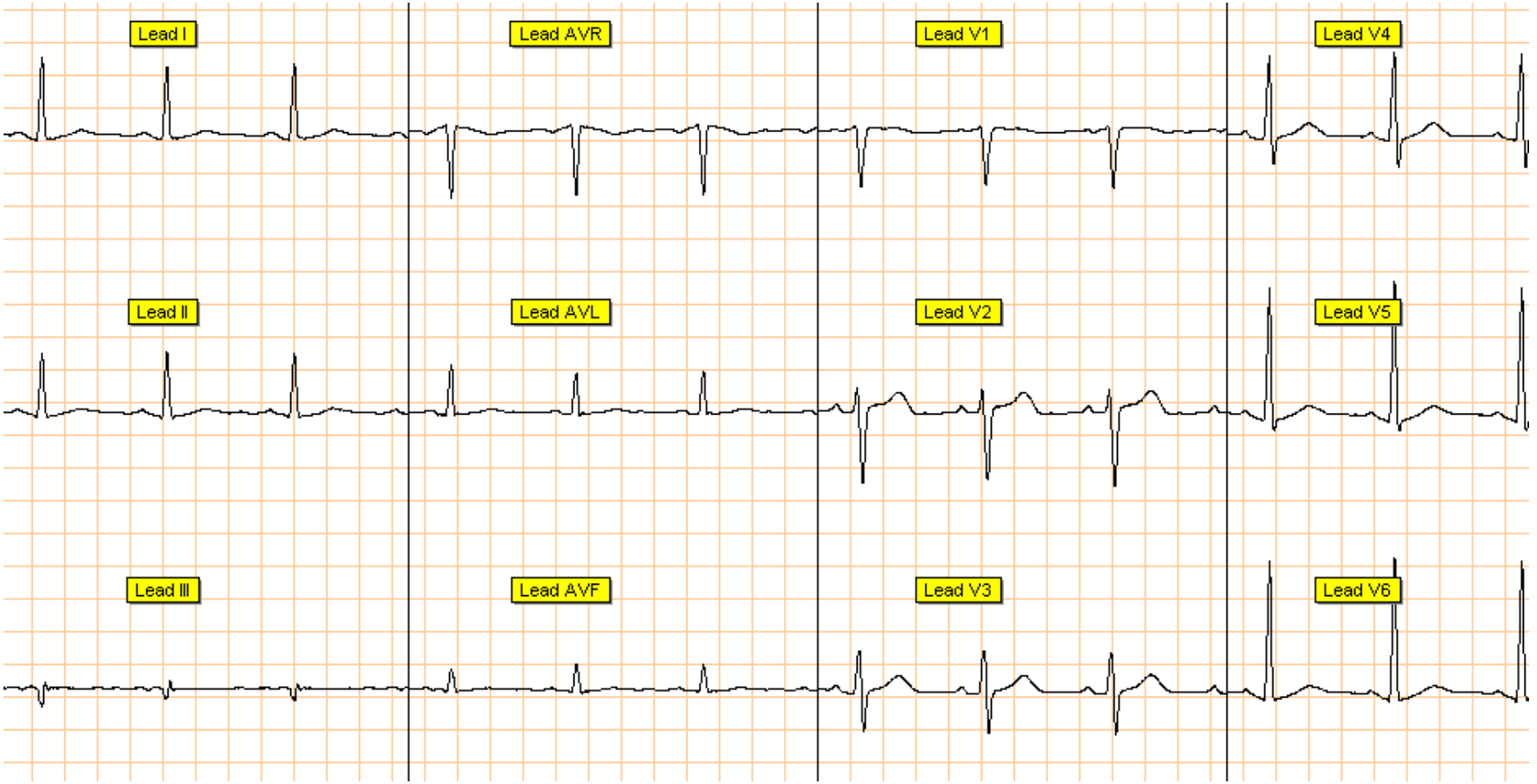


Subvalvular aortic stenosis

- A variety of lesions can cause subvalvar AS including a **thin membrane** (most common), thick fibromuscular ridge, diffuse tunnel-like obstruction, abnormal mitral valve attachments, and occasionally, accessory endocardial cushion tissue.
- It develops and progresses over time so that most experts in the field consider it an **acquired rather than a congenital lesion**. It has been proposed that subvalvar AS is due to progressive left ventricular outflow tract (LVOT**) thickening and scarring due to turbulence caused by an underlying abnormality in LVOT architecture**
- The diagnosis of subvalvar AS is made by echocardiography. Electrocardiogram and chest radiography are not diagnostic
- The definitive treatment for subvalvar AS is **surgical correction** of the obstruction
- **Subvalvar AS does not respond to balloon dilation.**
- **Many centers would not recommend surgery for peak gradients <40 mmHg alone, as many remain stable for many years.**
- Complications that occurred primarily in earlier surgical case series included mitral valve damage, AR, ventricular septal defect (VSD) creation, or bundle branch or complete heart block

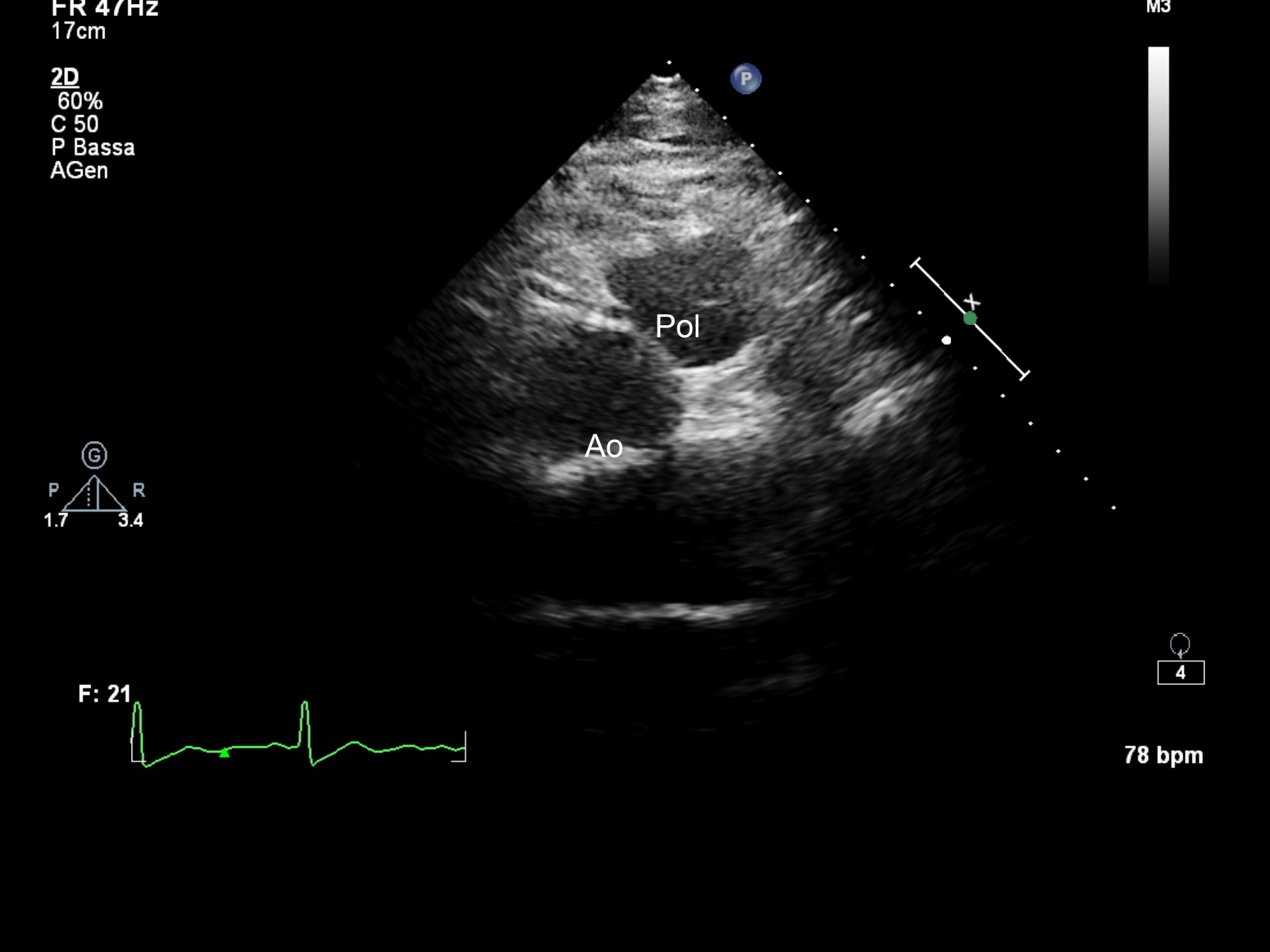
Diagnosi inaspettate

Camionista croato,55aa,arresto cardiaco scaricando un carico pesante dal camion
Rianimato dal 118 (13 DC-shock +RCP) accorso rapidamente .



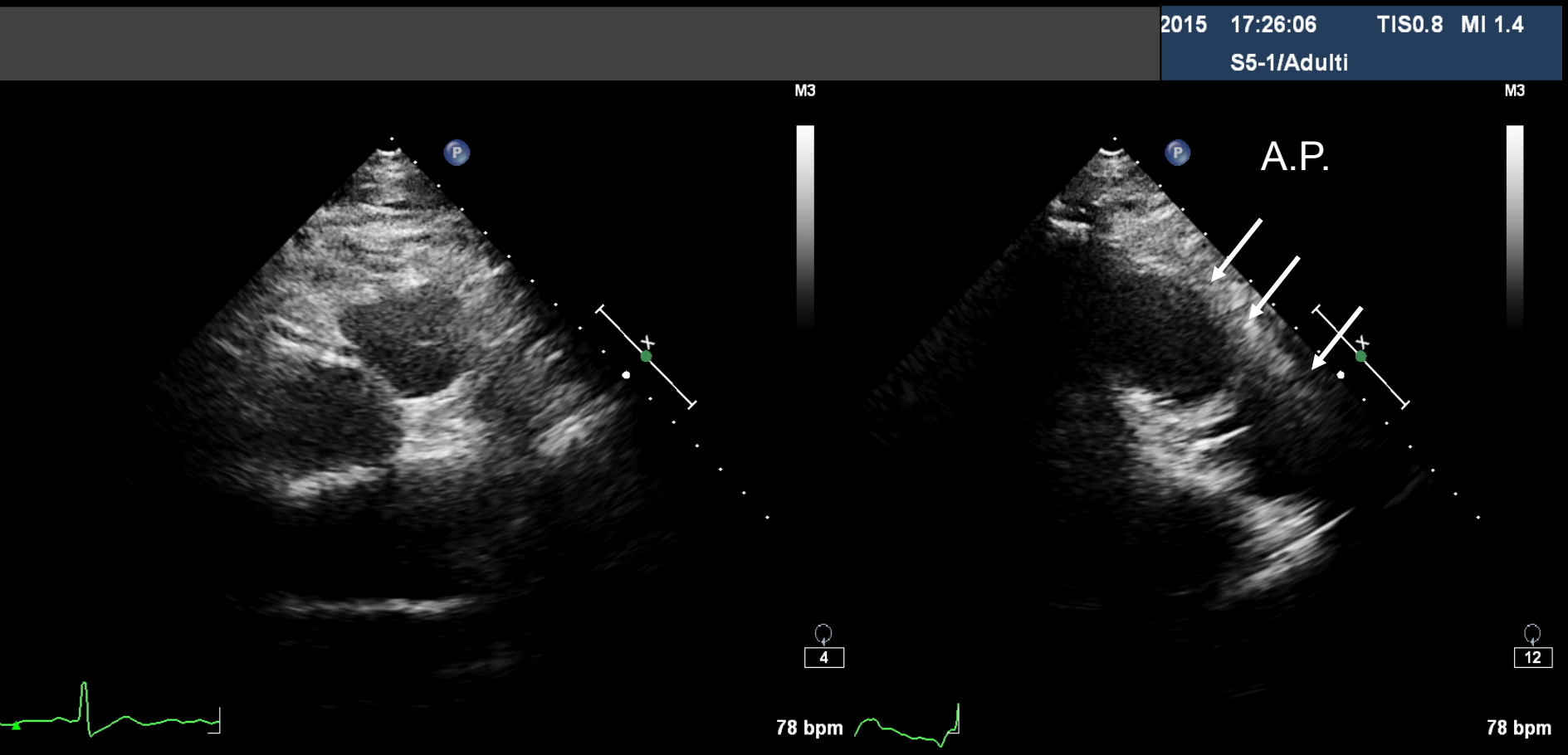
Valutazione clinico-anamnestica

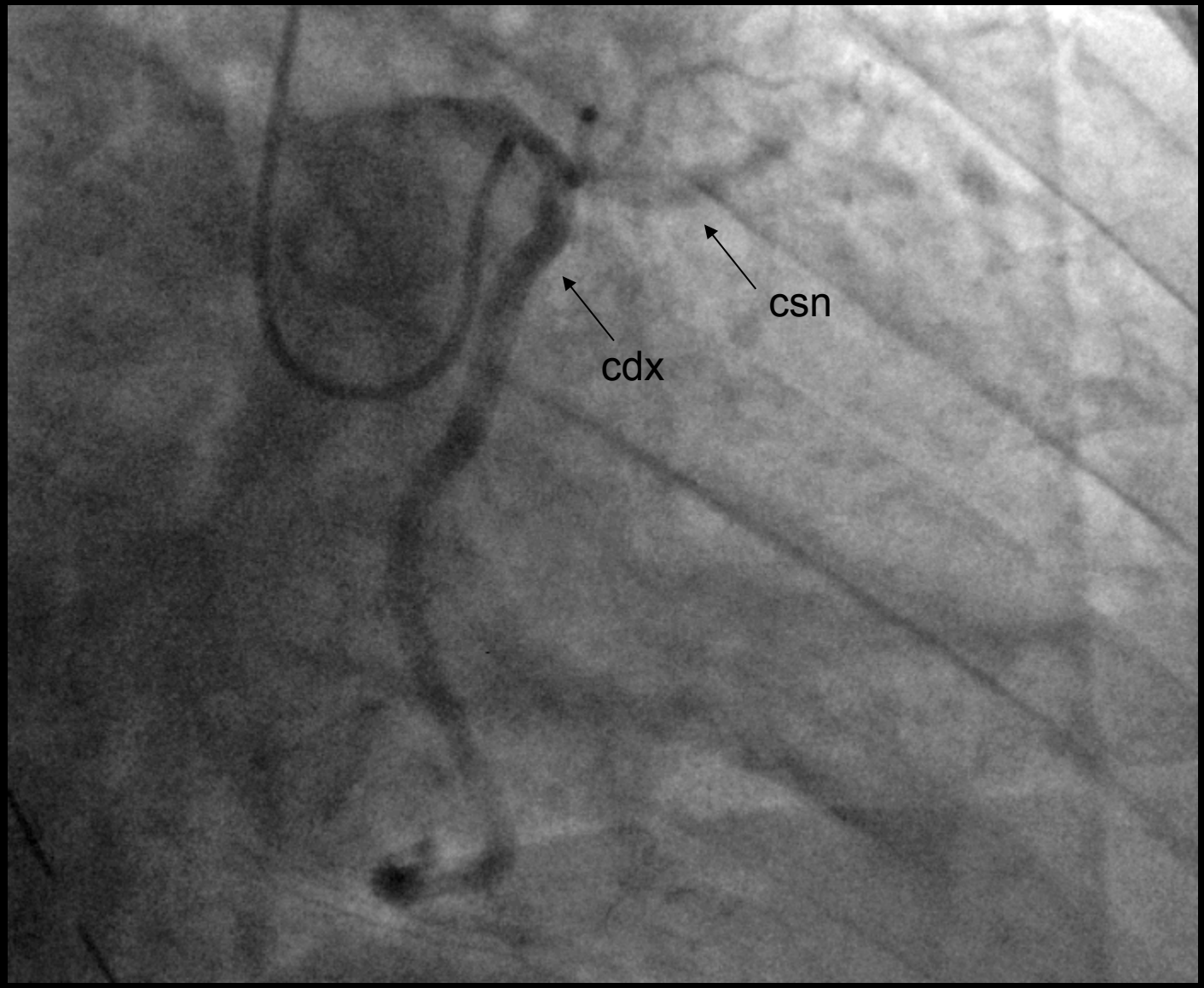
- Nessun precedente cardiologico
- Familiarità negtiva
- ECG del 2010 normale eseguito per iter operatorio (int.ortopedico)
- Esami tossicologici eseguiti in RIA negativi

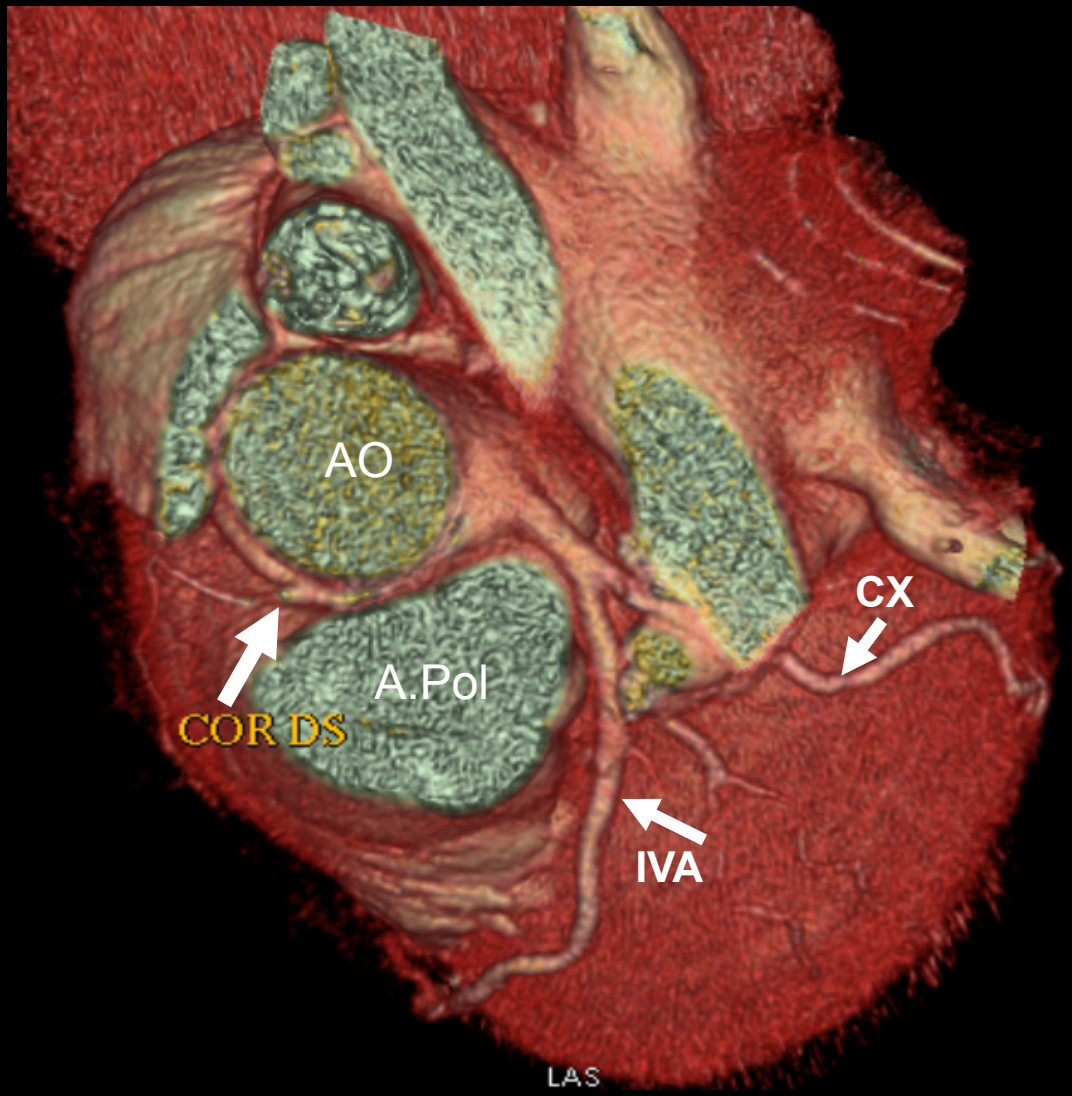

FR 47Hz
17cm
2D
60%
C 50
P Bassa
AGen
M3
Pol
Ao
G
P
R
1.7
3.4
F: 21
4
78 bpm

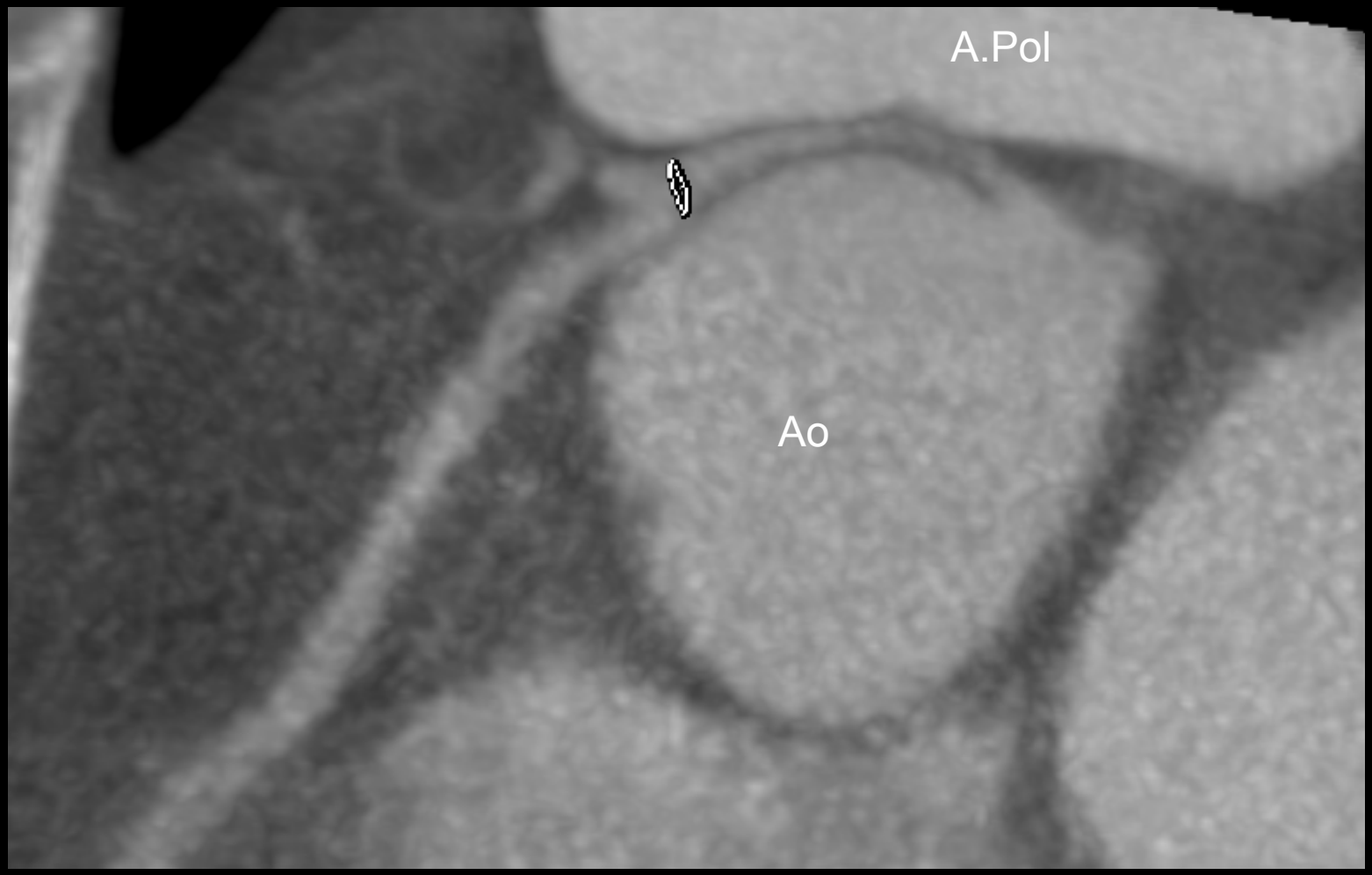
Asse corto

Asse lungo



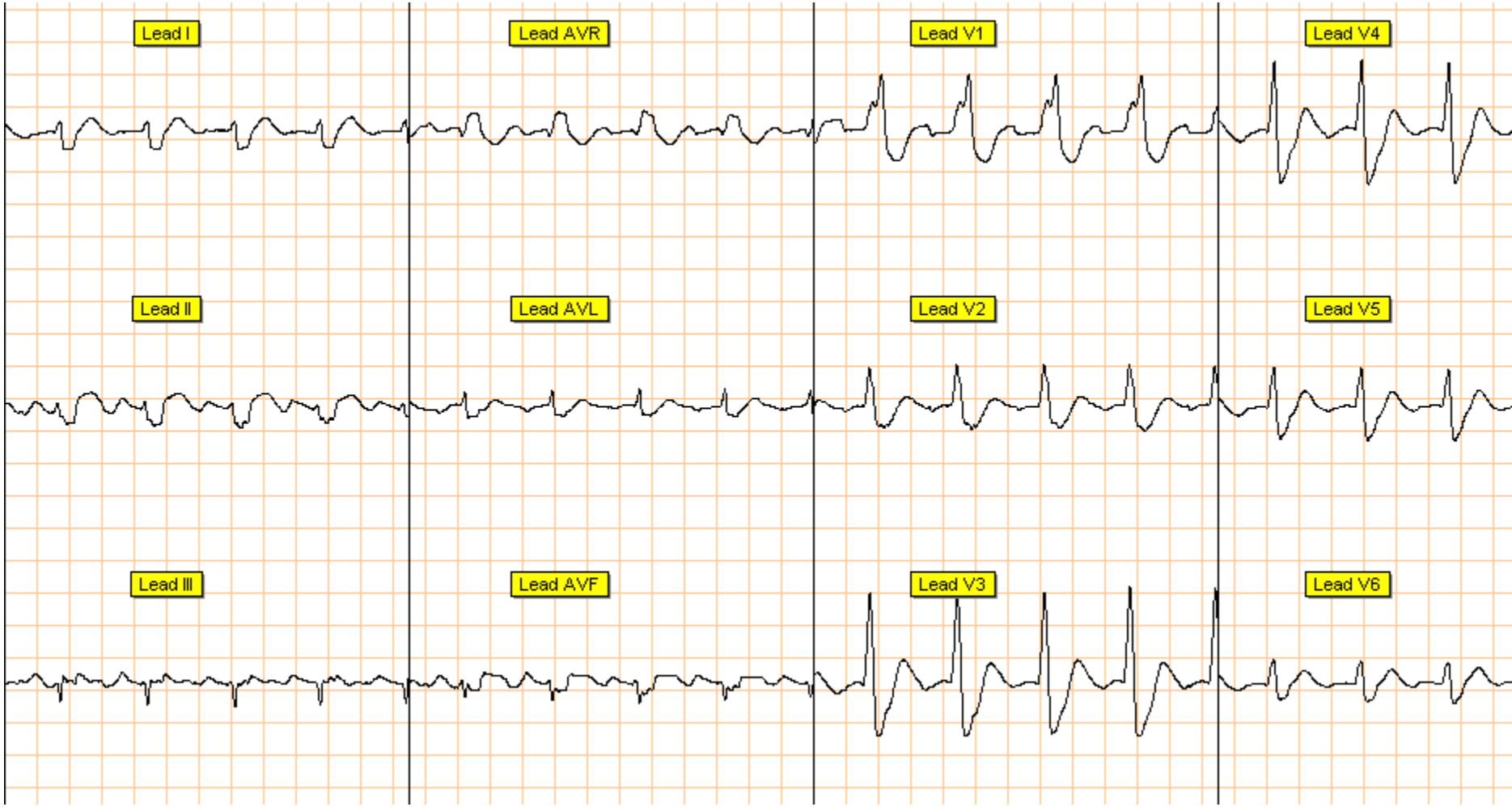

csn
cdx

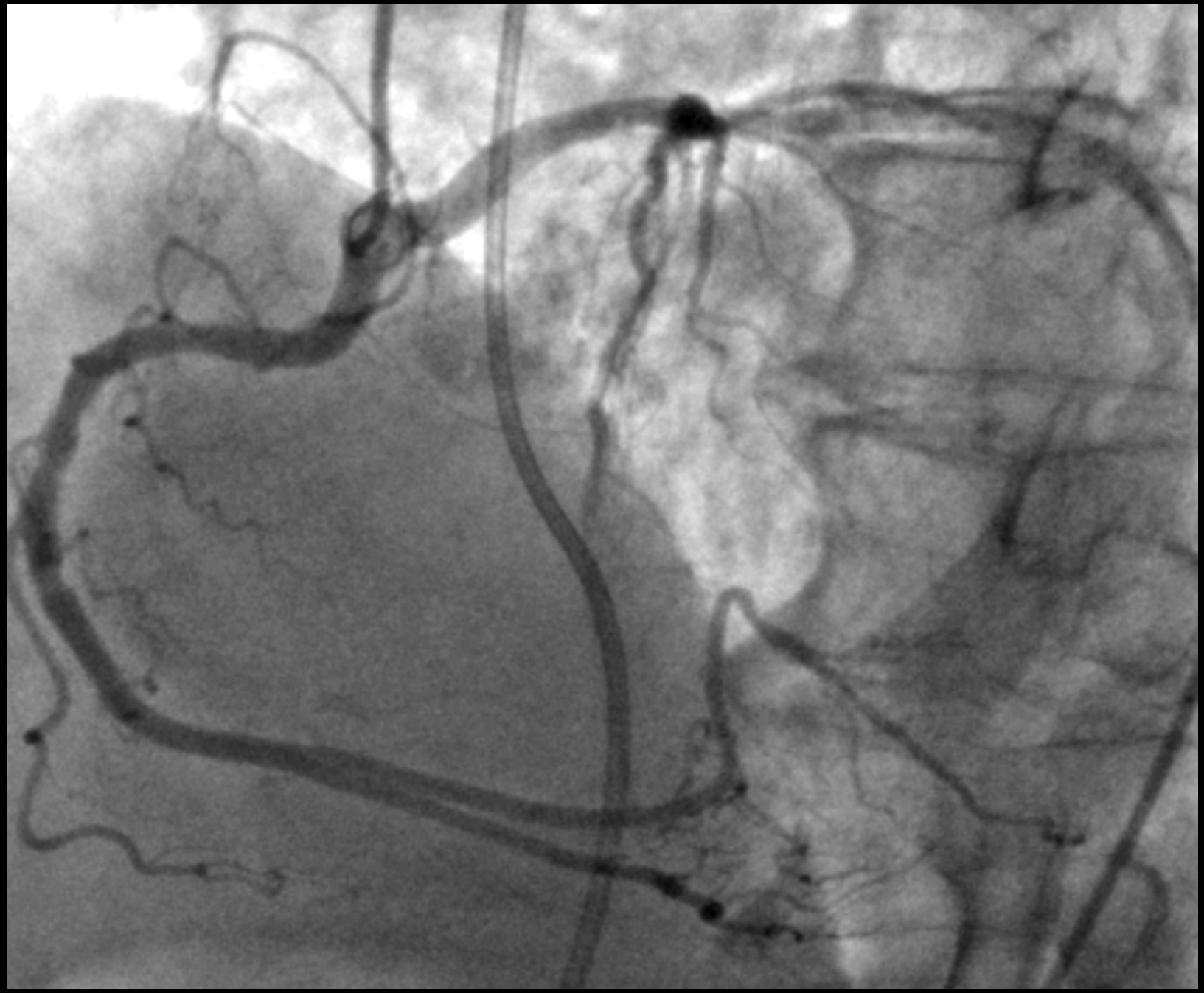

AO
A.Pol
COR DS
CX
IVA
LAS

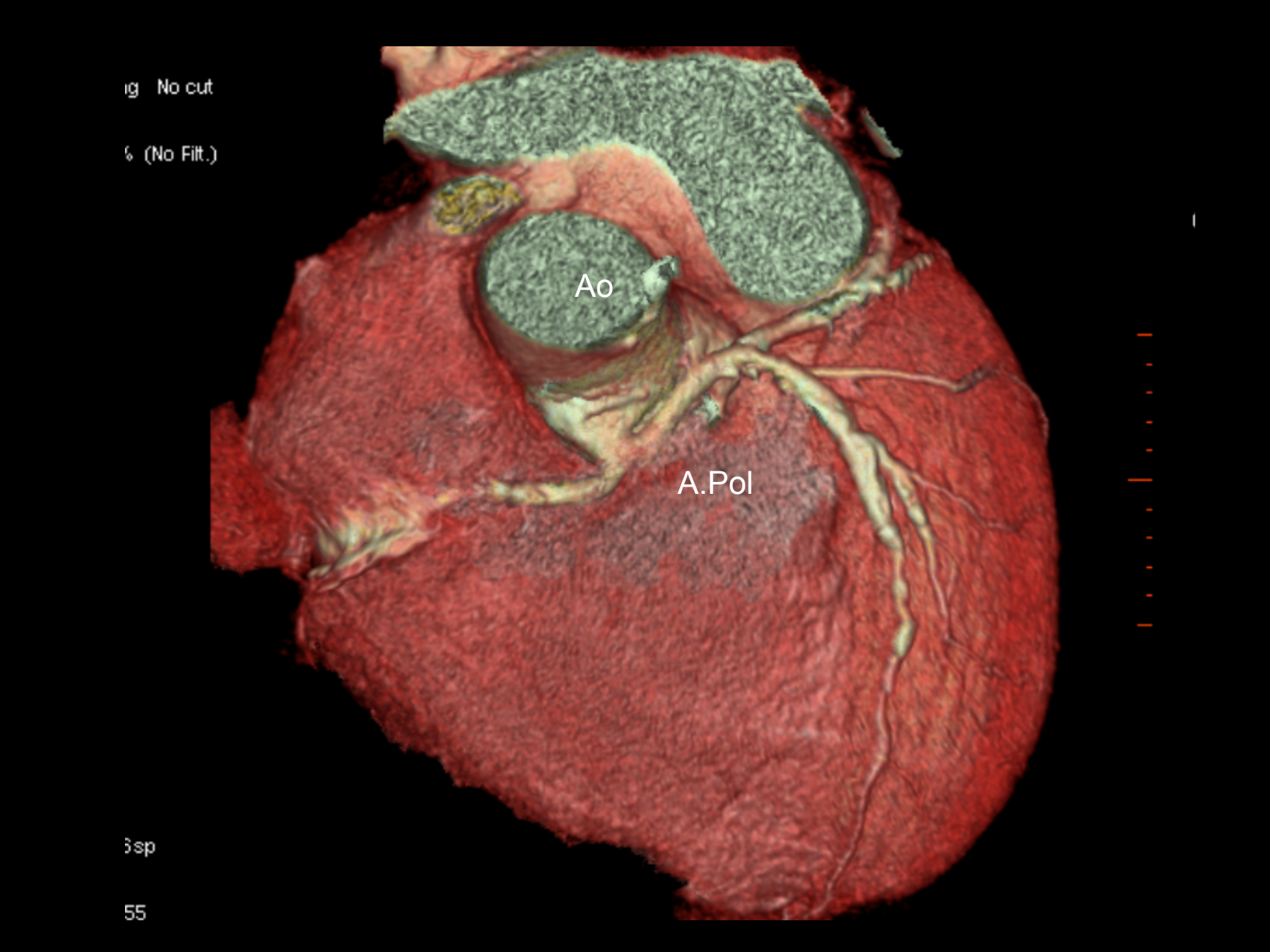

A.Pol
Ao

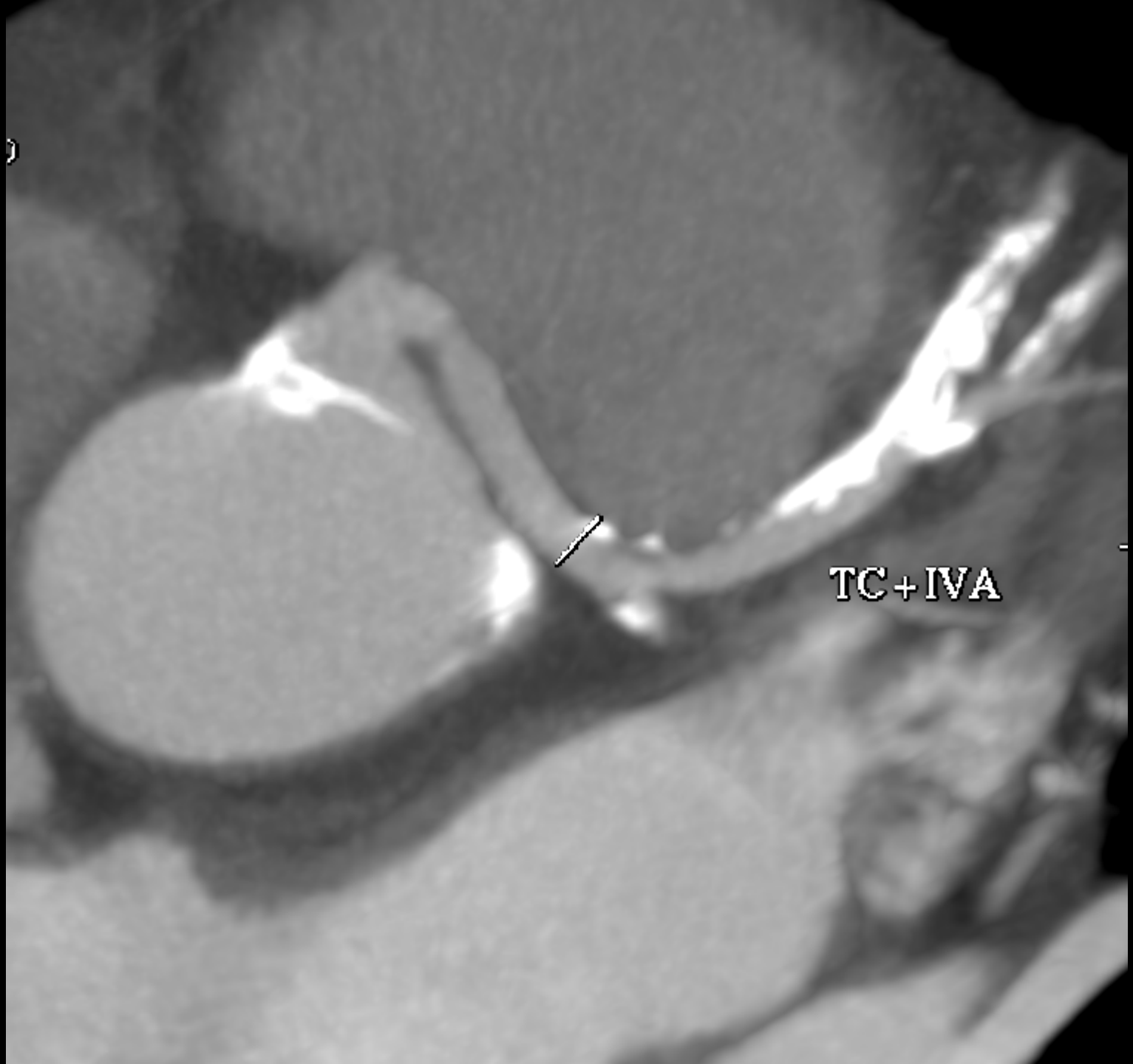
F.L. 83y ex atleta olimpico
Ricoverato in RIAN. per insufficienza respiratoria

All'Ecocardio : Stenosi aortica serrata >TAVI






ig No cut
% (No Filt.)
Ao
A.Pol
Sp
55


TC+IVA

8.5. Recommendations for Congenital Coronary Anomalies of Ectopic Arterial Origin

Class I

1. The evaluation of individuals who have survived unexplained aborted sudden cardiac death or with unexplained life-threatening arrhythmia, coronary ischemic symptoms, or LV dysfunction should include *assessment of coronary artery origins and course.* (*Level of Evidence: B*)

2. CT or magnetic resonance angiography is useful as the initial screening method in centers with expertise in such imaging. (*Level of Evidence: B*)

3. Surgical coronary revascularization should be performed in patients with any of the following:

a. Anomalous left main coronary artery coursing between the aorta and pulmonary artery. (*Level of Evidence: B*)

b. Documented coronary ischemia due to coronary compression (when coursing between the great arteries or in intramural fashion). (*Level of Evidence: B*)

c. Anomalous origin of the right coronary artery between aorta and pulmonary artery with evidence of ischemia

Conclusione

- Nonostante il grande sviluppo della diagnostica non invasiva come screening per la diagnosi precoce, alcune cardiopatie congenite anche complesse possono manifestarsi in eta' adulta , soprattutto se con emodinamica favorevole o in popolazioni non adeguatamente controllate dal punto di vista sanitario.
- Indipendentemente dall'età l'opzione chirurgica/ riparativa o con terapia medica avanzata è quasi sempre percorribile e non può essere scartata a priori.
- Percorso selettivo o con la collaborazione di Centri di riferimento in possesso dei requisiti culturali e tecnici adeguati

  (Eco-Ped/Emod/RMN/CardioTC/UTIC/CCH)

Grazie per l’ascolto