

Le cardiopatie congenite in pazienti adulti

Non sempre tutto è facilmente riconoscibile.

*Presentazioni cliniche inusuali,
complicanze nascoste e killer silenziosi*

Berardo Sarubbi

Unità Dipartimentale Cardiopatie Congenite dell'Adulto

Ospedale Monaldi - Napoli

www.berardosarubbi.it



Caso clinico 1: una cardiopatia congenita complessa

Caso clinico 1

- N.R. anni 17
- IV classe NYHA
- Emoftee. Pnx spontaneo.
- Episodi ripetuti di fibrillazione ventricolare: cardioversione con DC shock esterno.
- Sat O2 50 mmHg.
- PAS 90/45 mmHg. Cardiopatia congenita complessa.

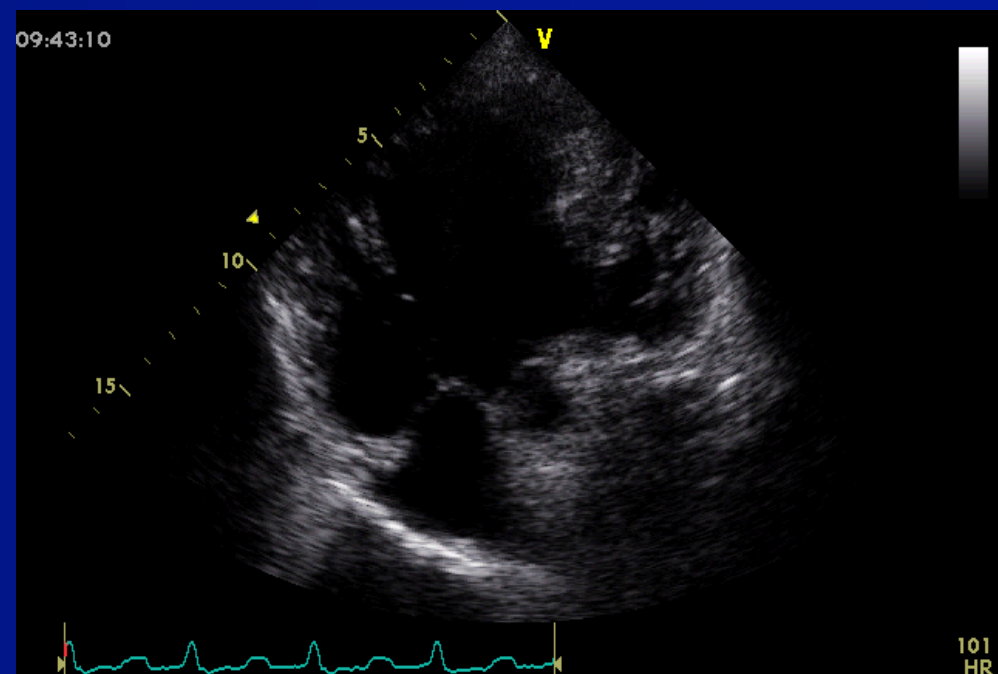
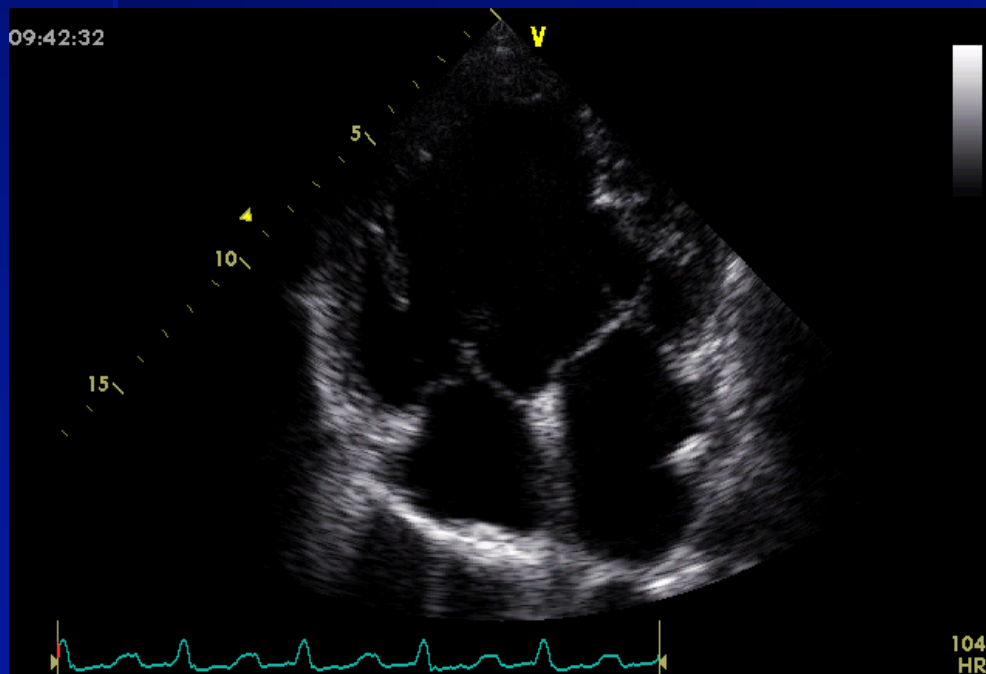
Caso clinico 1

- N.R. anni 17

Doppia entrata in ventricolo unico.

Camera accessoria anteriore sn.

Ampio forame bulboventricolare.



Caso clinico 1

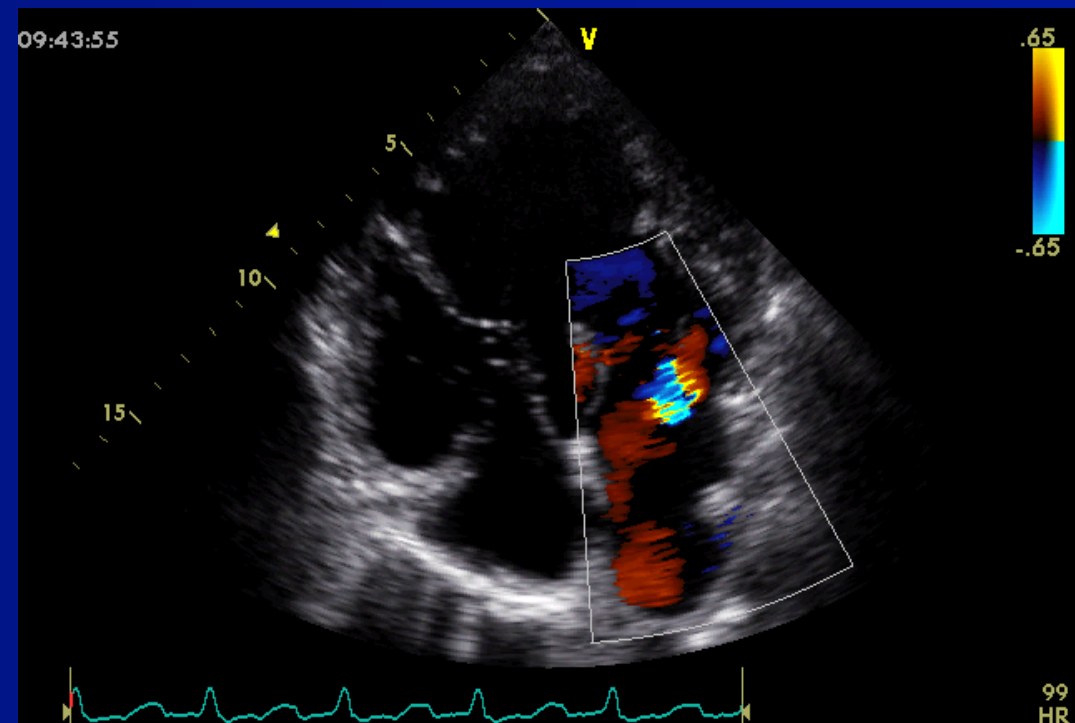
- N.R. anni 17

Doppia entrata in ventricolo unico.

Camera accessoria anteriore sn.

Ampio forame bulboventricolare.

Insufficienza severa della valvola AV sn.



Caso clinico 1

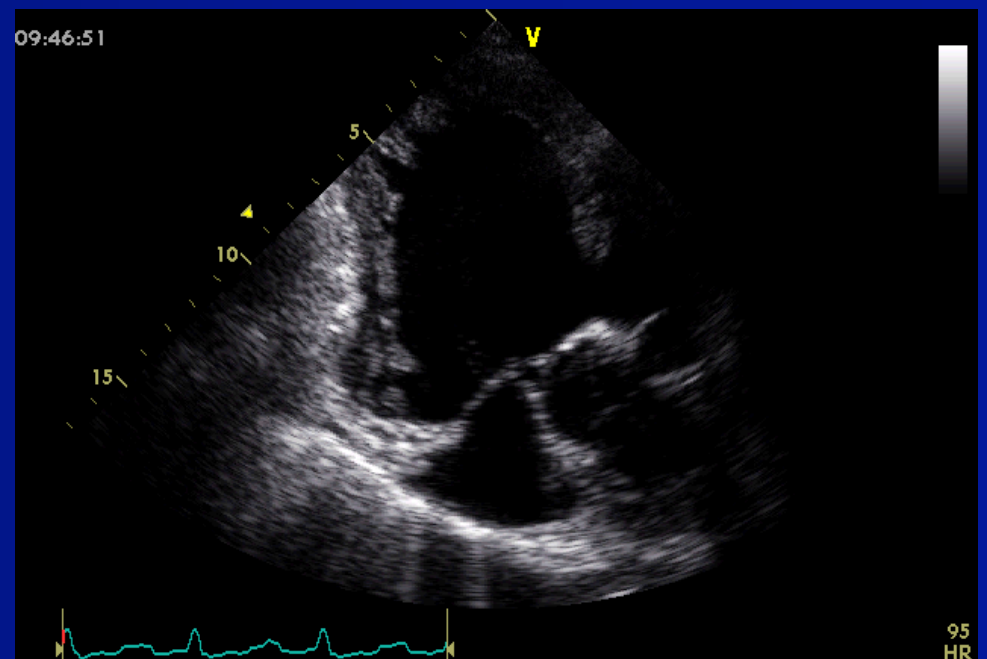
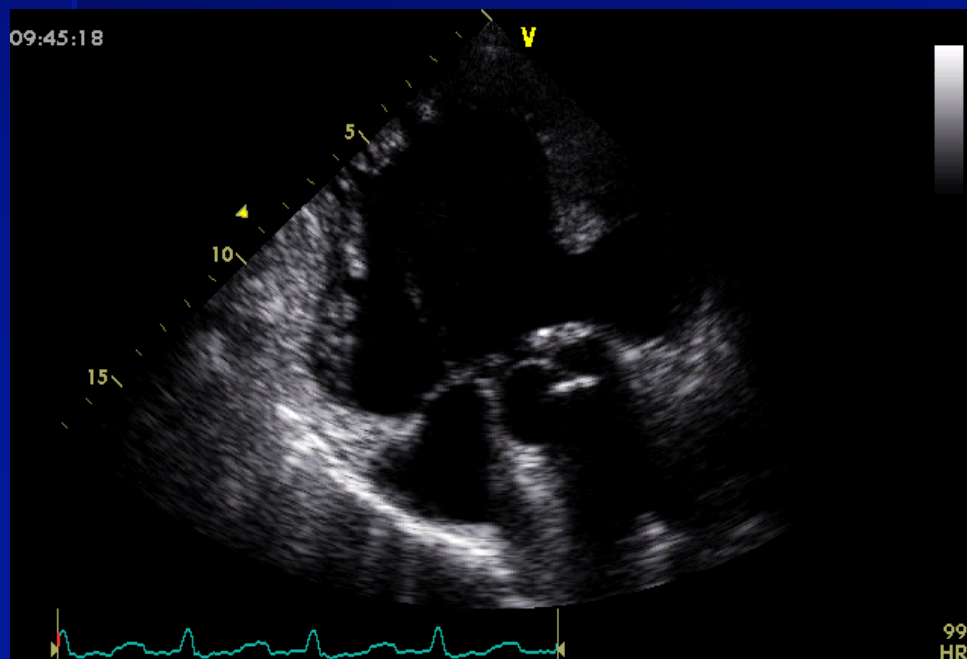
- N.R. anni 17

Doppia entrata in ventricolo unico.

Camera accessoria anteriore sn.

Ampio forame bulboventricolare.

Vasi malposti (polmonare post-dx, aorta ant-sn).



Caso clinico 1

- N.R. anni 17

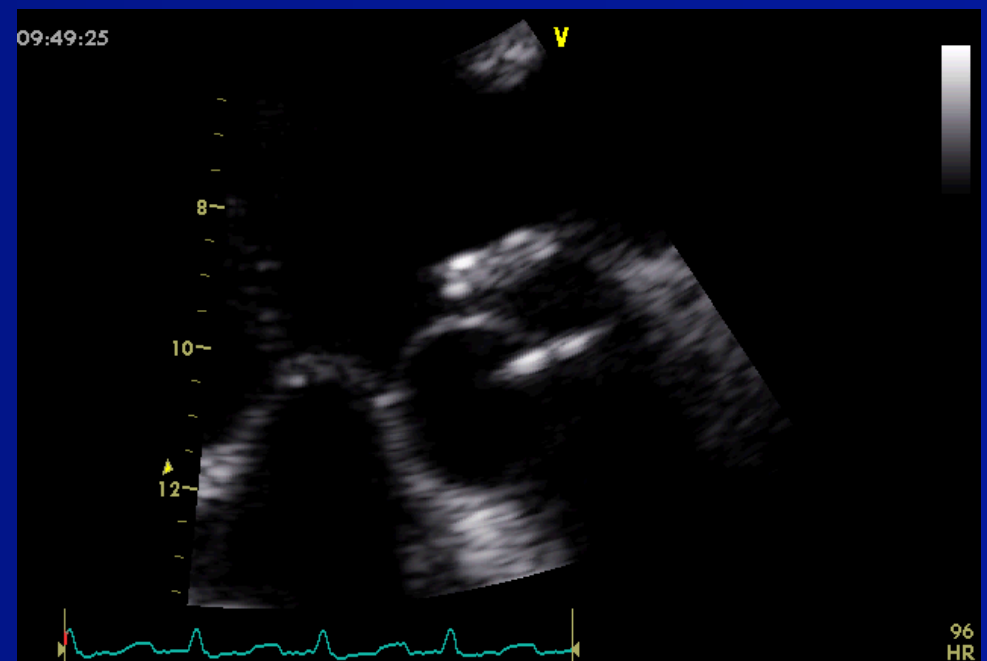
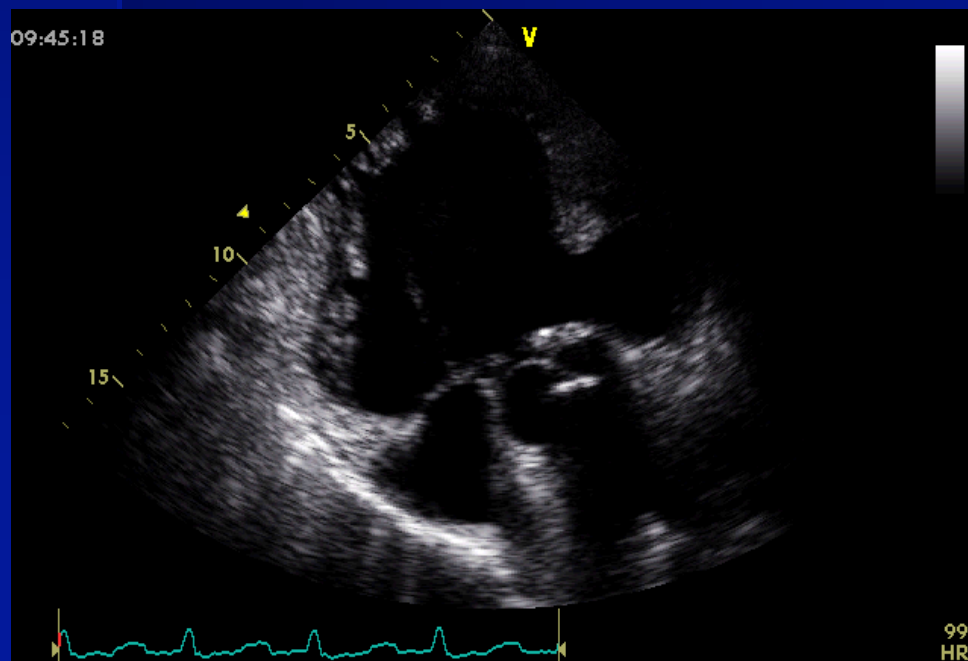
Doppia entrata in ventricolo unico.

Camera accessoria anteriore sn.

Ampio forame bulboventricolare.

Vasi malposti.

Stenosi sottovalvolare polmonare.



Caso clinico 1

- N.R. anni 17

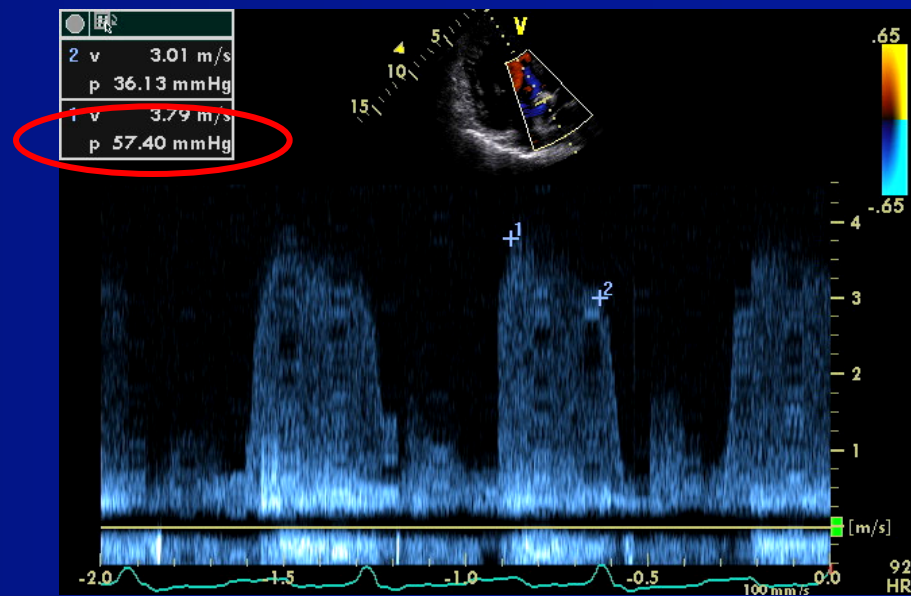
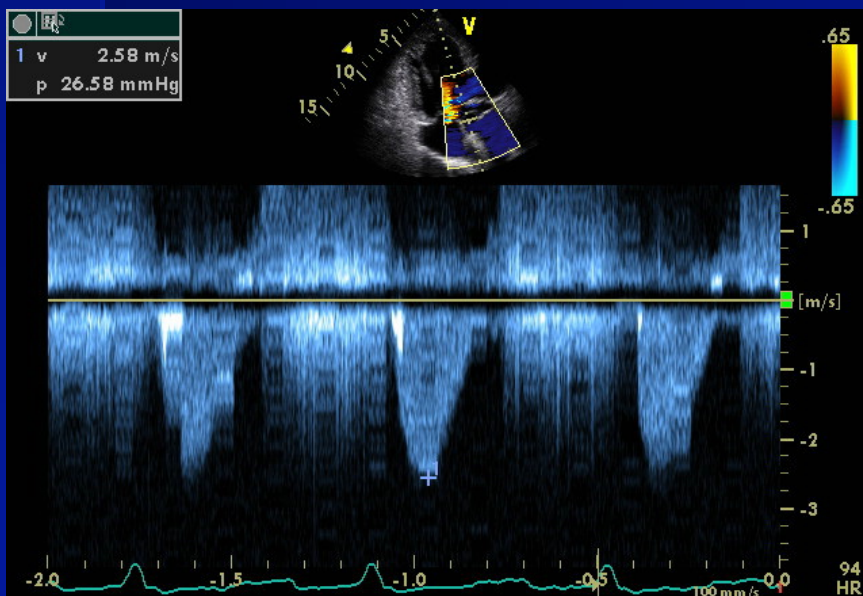
Doppia entrata in ventricolo unico.

Camera accessoria anteriore sn.

Ampio forame bulboventricolare.

Vasi malposti.

Stenosi sottovalvolare polmonare “non-protettiva”. Ipertensione polmonare.



Caso clinico 1

- N.R. anni 17

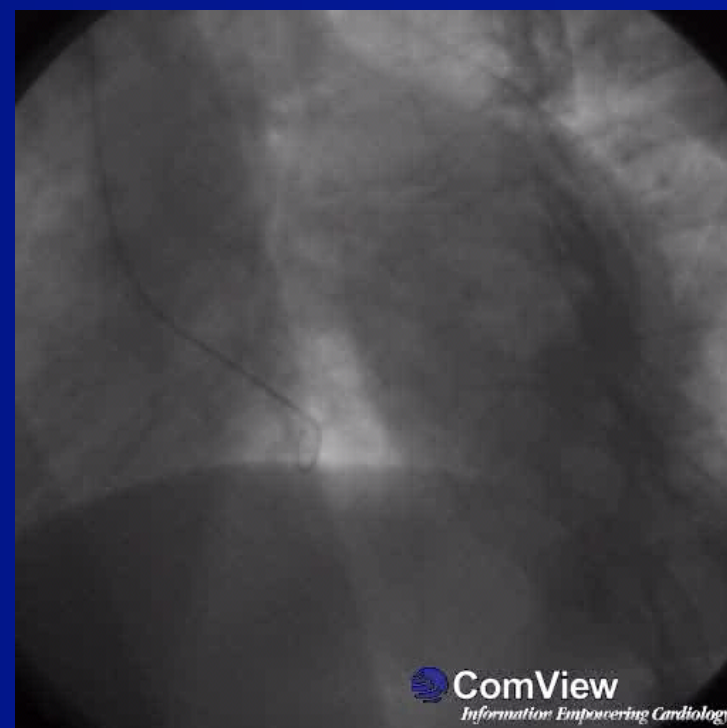
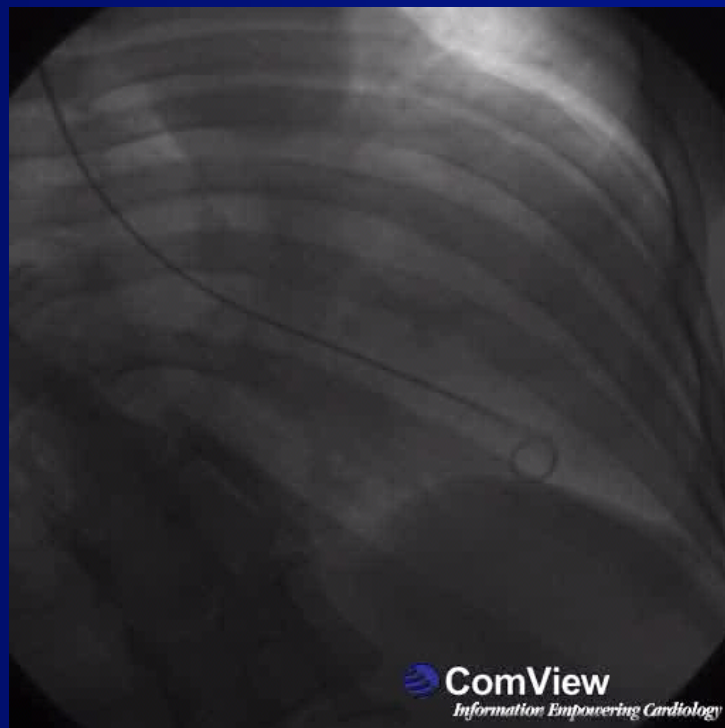
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Vasi malposti.

Ipertensione polmonare.



Caso clinico 1

- N.R. anni 17

Doppia entrata in ventricolo unico.

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Ampio forame bulboventricolare.

Vasi malposti.

Ipertensione polmonare.

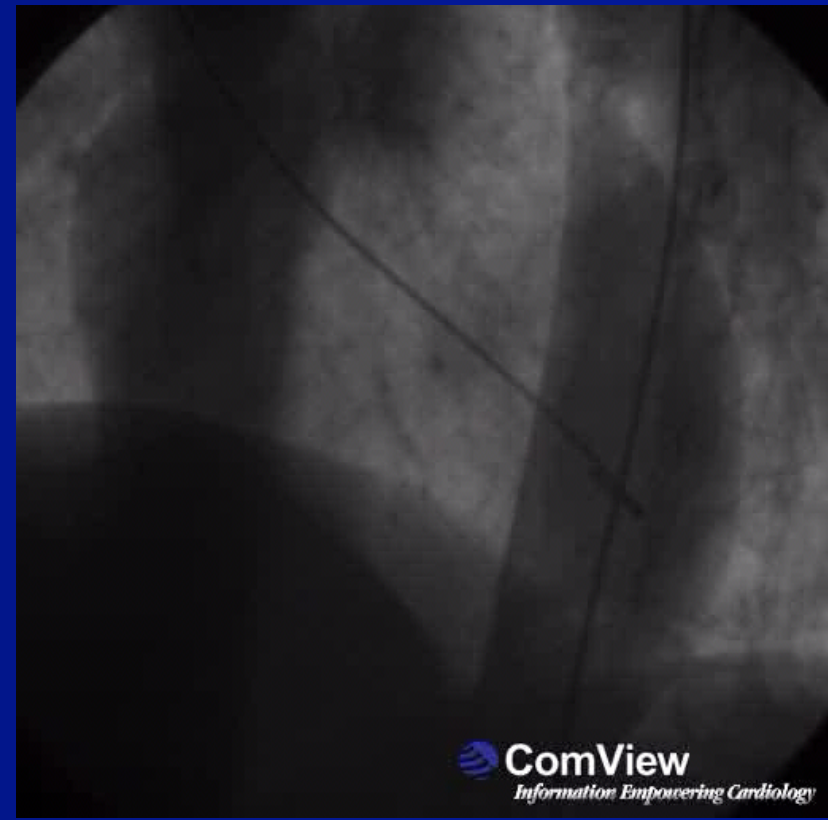
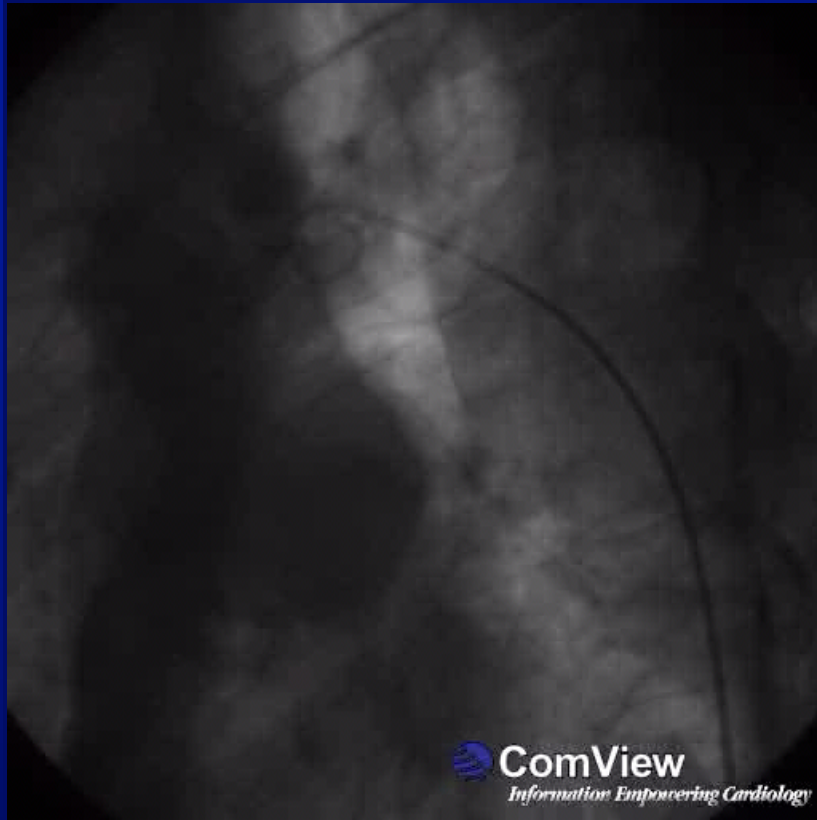
A 3 anni follow up.

Ottimizzazione terapia di background + bosentan (125 mg x 2) +
epoprostenolo (12 ng/kg/min).

III classe funzionale.

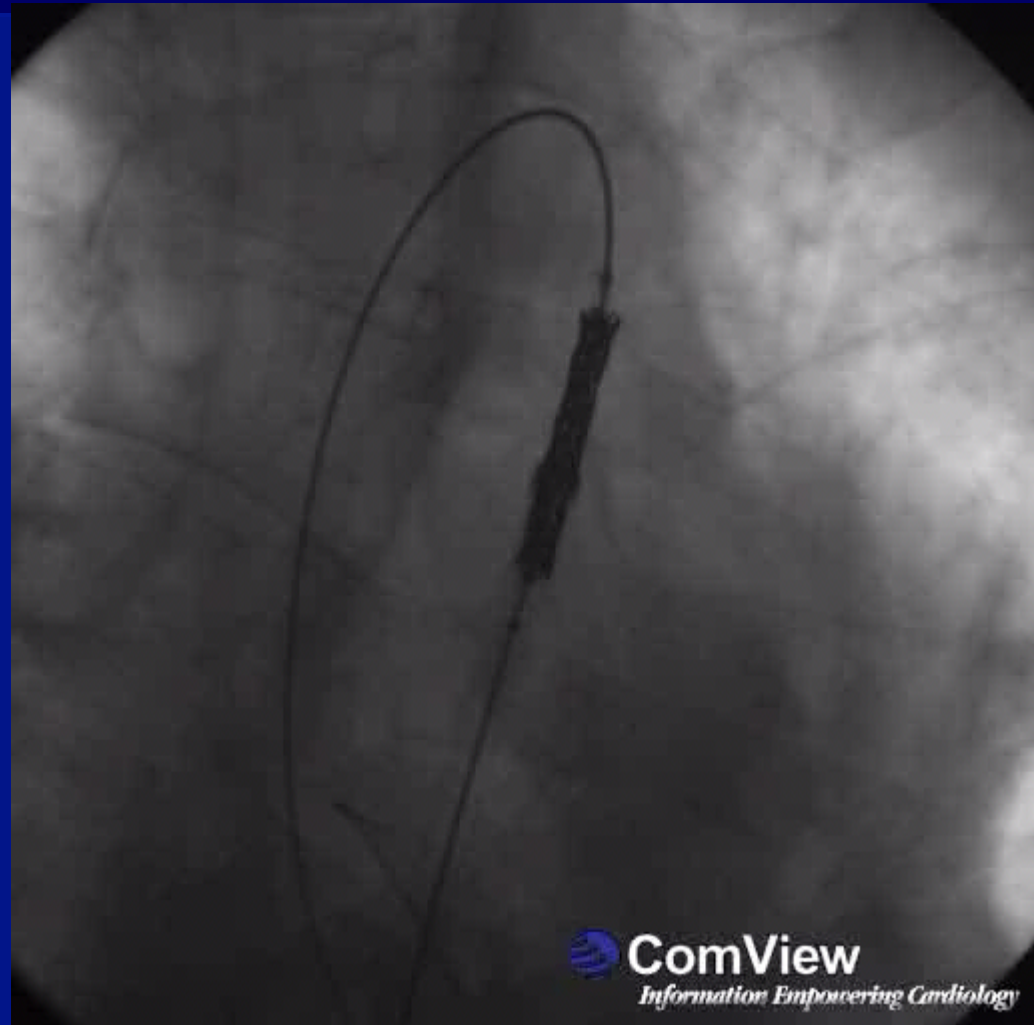
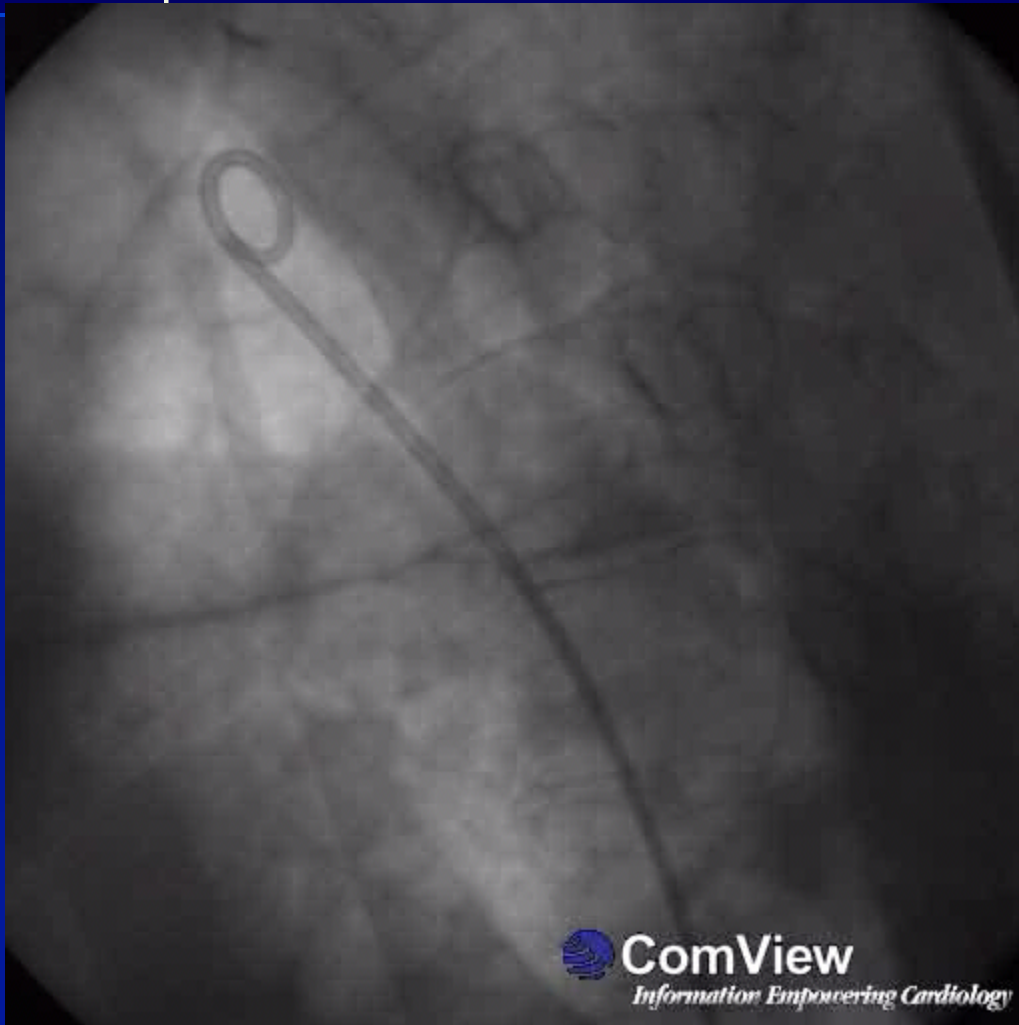
Caso clinico 2: management complesso di CHD combinate

- A.G. anni 39
- Coartazione aortica + DIV.



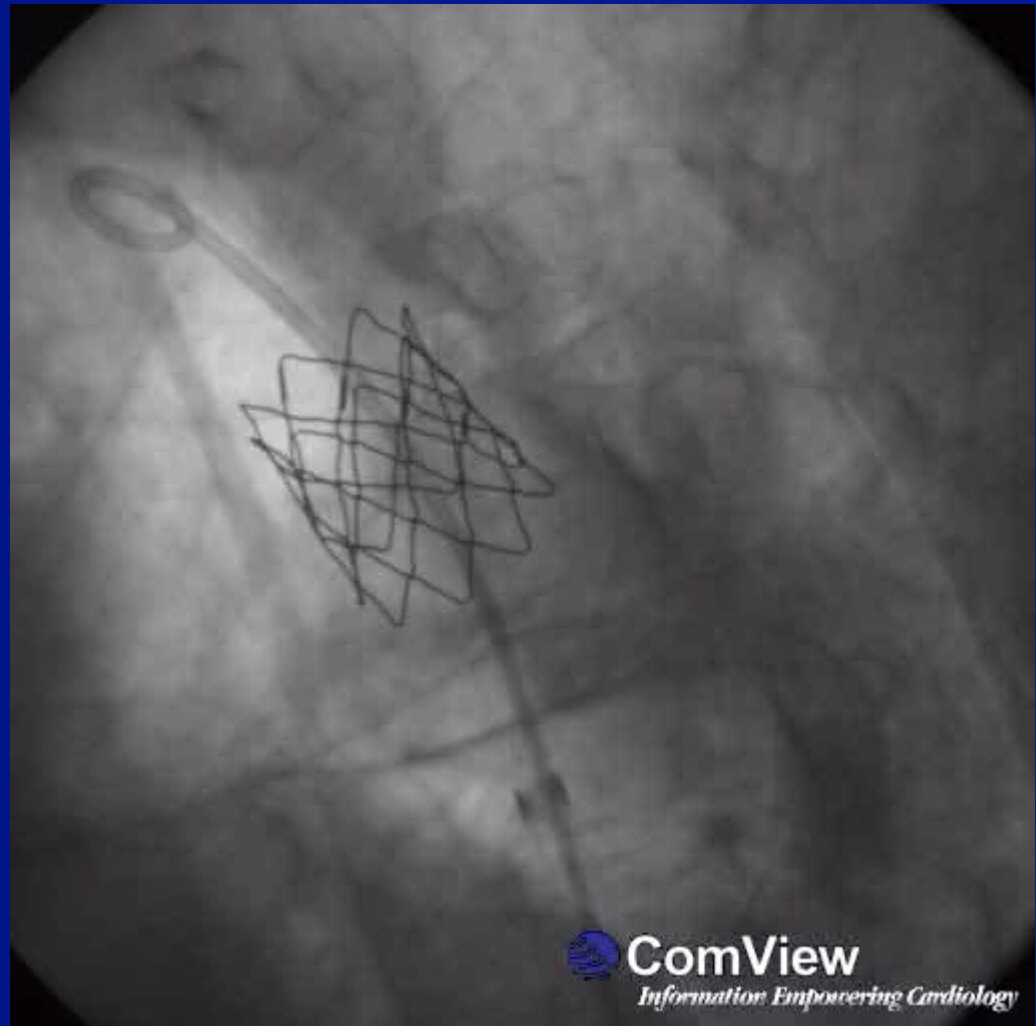
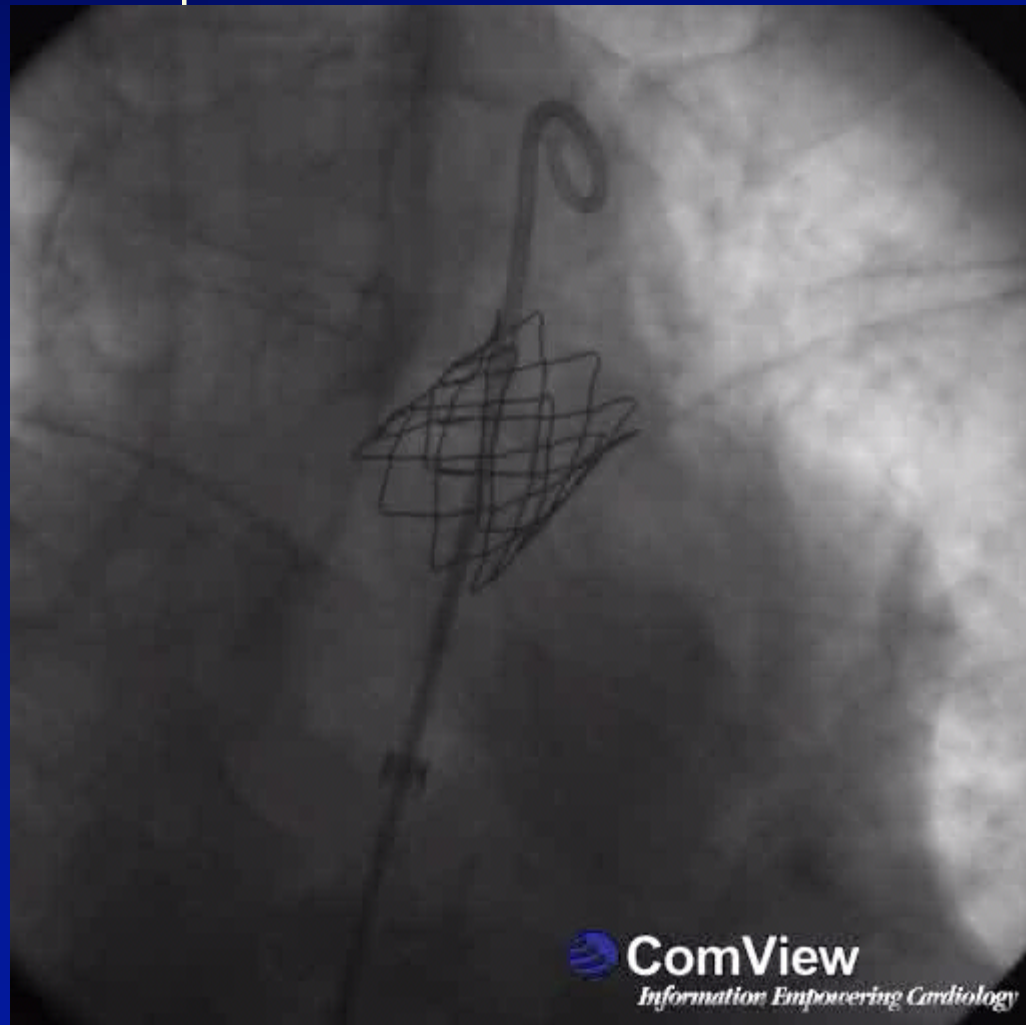
Caso clinico 2

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Caso clinico 2

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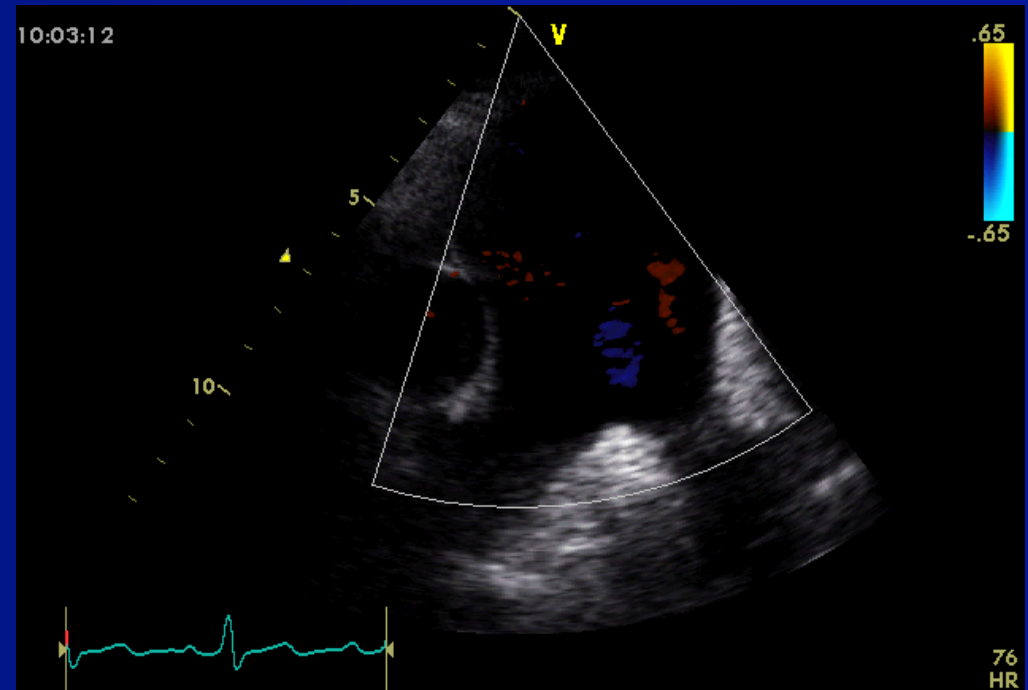
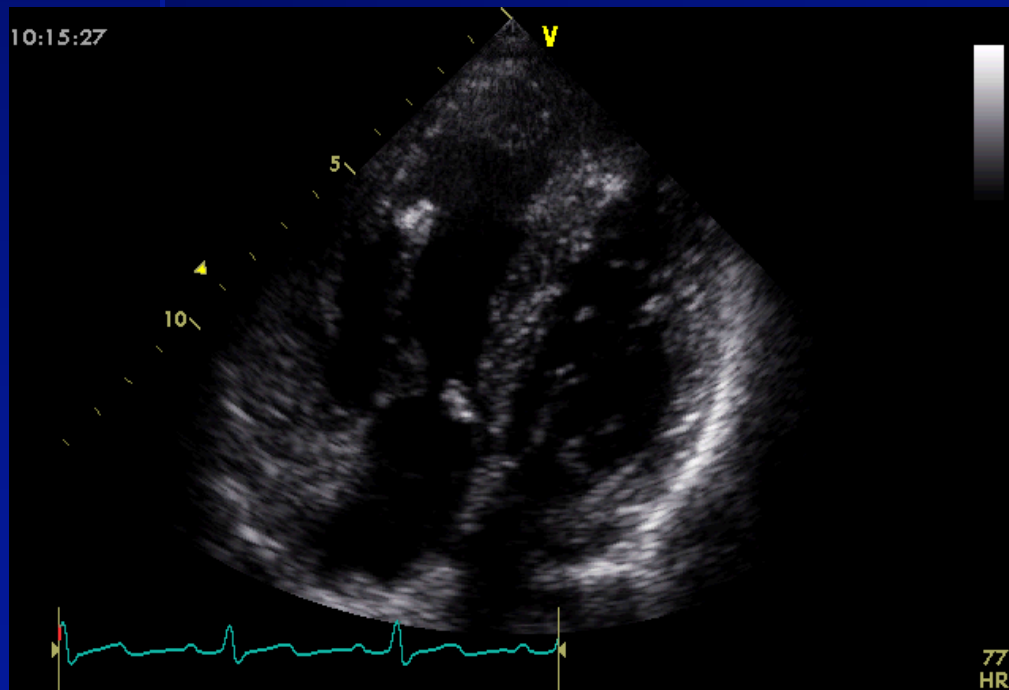


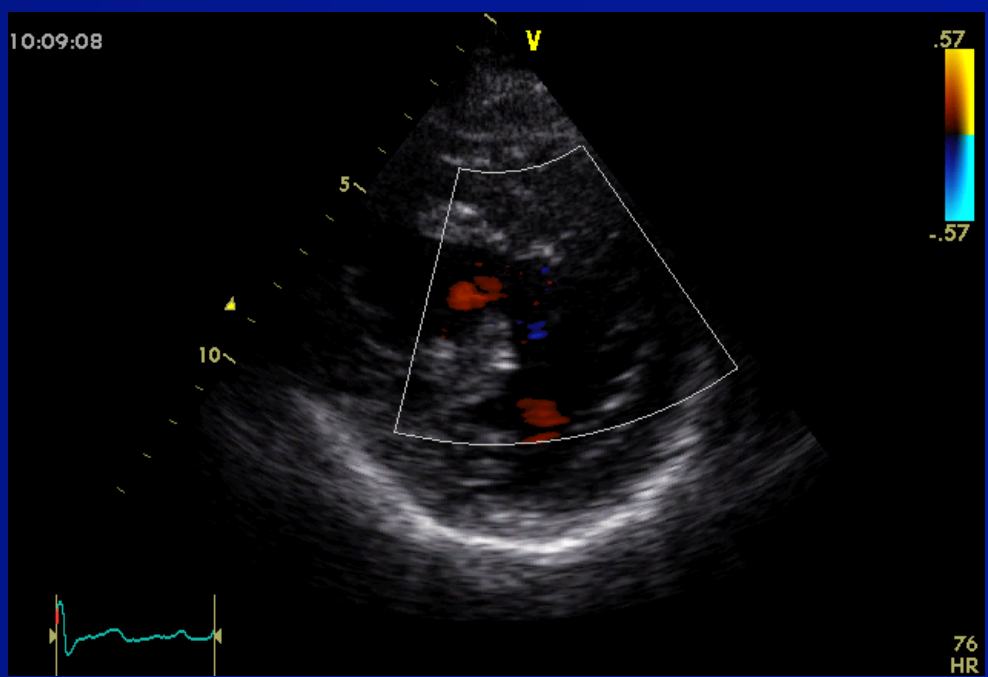
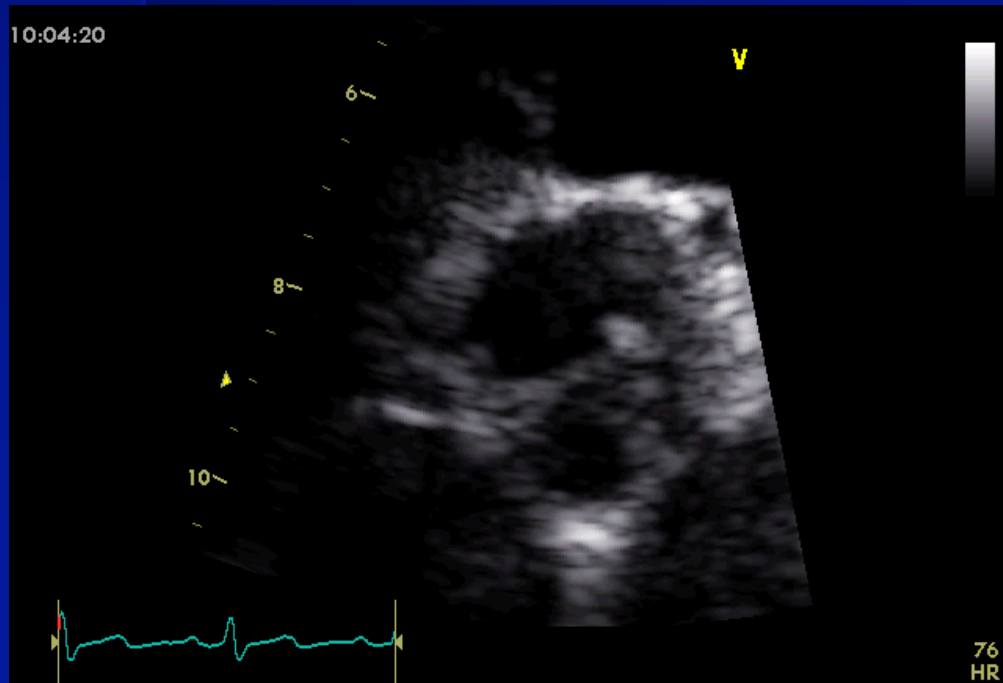
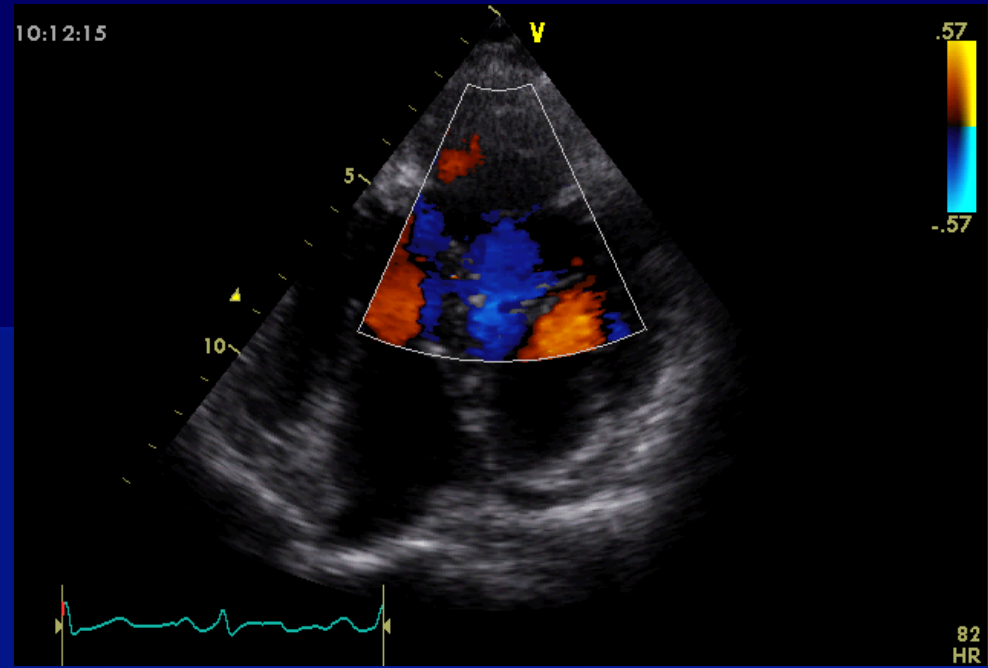
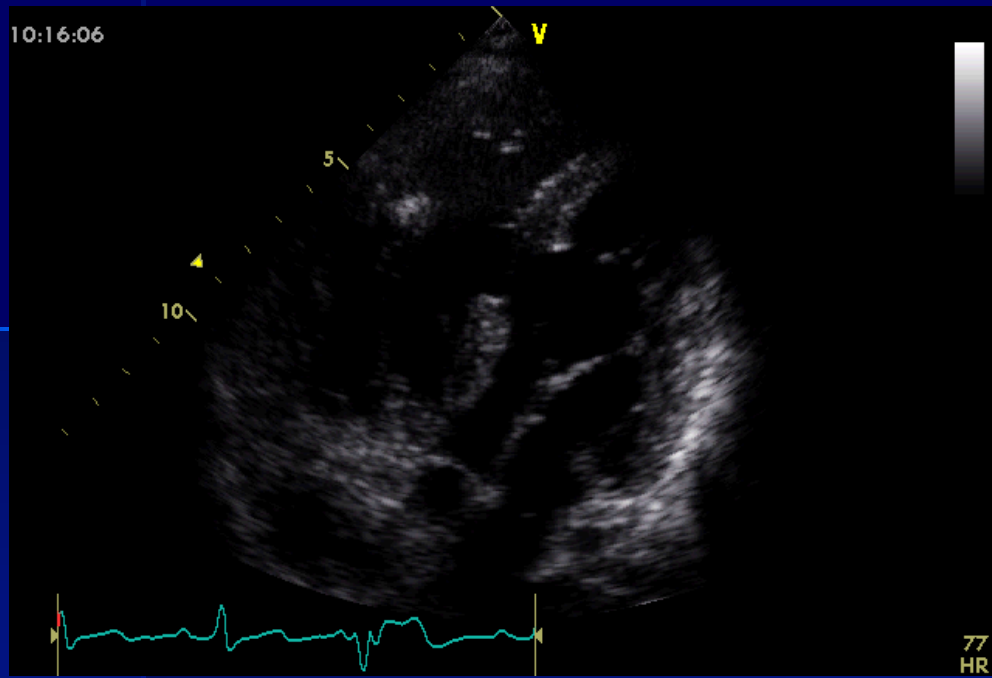
Caso clinico 2

- A.G. anni 39
- Coartazione aortica.

Dilatazione e ipertrofia VD.

Marcata dilatazione polmonare.





Caso clinico 2

- A.G. anni 39

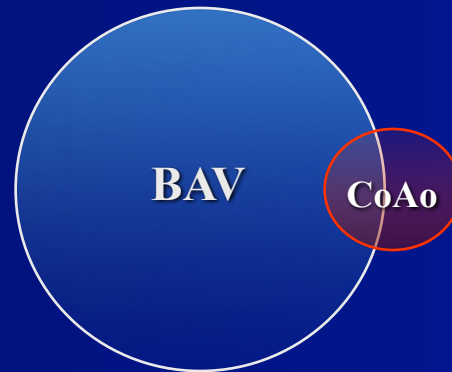
SINDROME DELLA COARTAZIONE AORTICA:

- Coartazione aortica
- Difetto interventricolare (del terzo medio del setto anteriore).
- Valvola aortica bicuspid.

Aorta bicuspid e coartazione



- **Pazienti con aorta bicuspid presentano una coartazione nel 6% dei casi.**
- **Il 50% (20-85%) dei pazienti con coartazione presentano una bicuspidia aortica.**
- **Complicanze nel 33% dei pazienti con BAV + CoAo**



Nistri S, Heart 1999;82:19-22

Keane MG, Circulation 2000;102:35-39

Fedak P, Circulation 2002;106:900-4

Aorta bicuspide: quando si associa a valvulopatia, quando a coartazione?

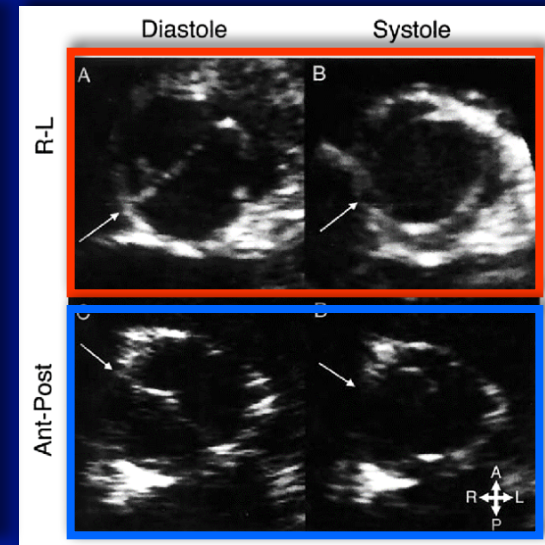
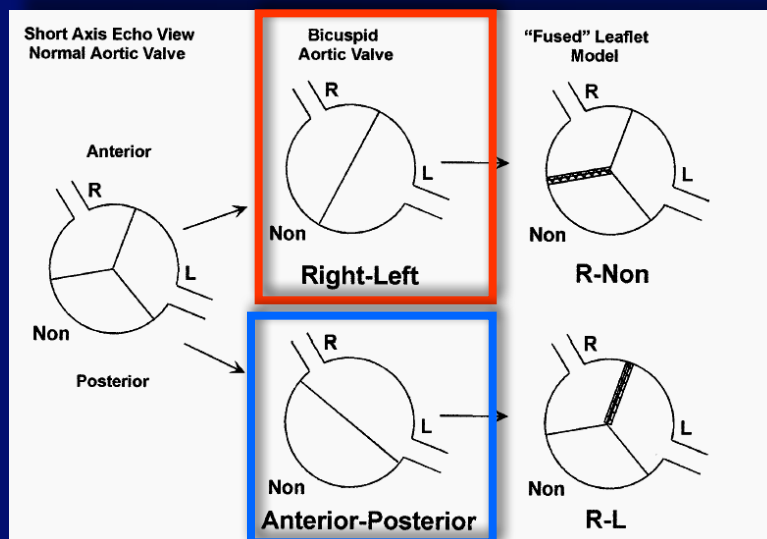
Morphology of Bicuspid Aortic Valve in Children and Adolescents

Susan M. Fernandes, MHP, PA-C,* Stephen P. Sanders, MD,† Paul Khairy, MPH, MD,*
 Kathy J. Jenkins, MD, MPH,* Kimberlee Gauvreau, ScD,* Peter Lang, MD,* Hilary Simonds, MS,*
 Steven D. Colan, MD*

J Am Coll Cardiol 2004;44:1648 –51

Conclusions: “Analysis of bicuspid aortic valve morphology is of clinical and prognostic relevance. Fusion of the right-coronary and non-coronary leaflets was associated with more significant valve pathology, whereas fusion of the right-coronary and left-coronary leaflets was associated overwhelmingly with aortic coarctation and less aortic valve pathology”.

n = 1,135 patients



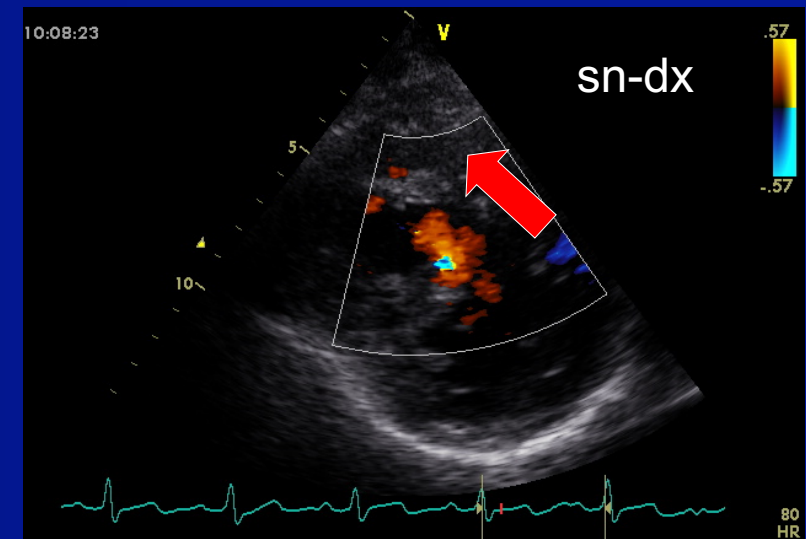
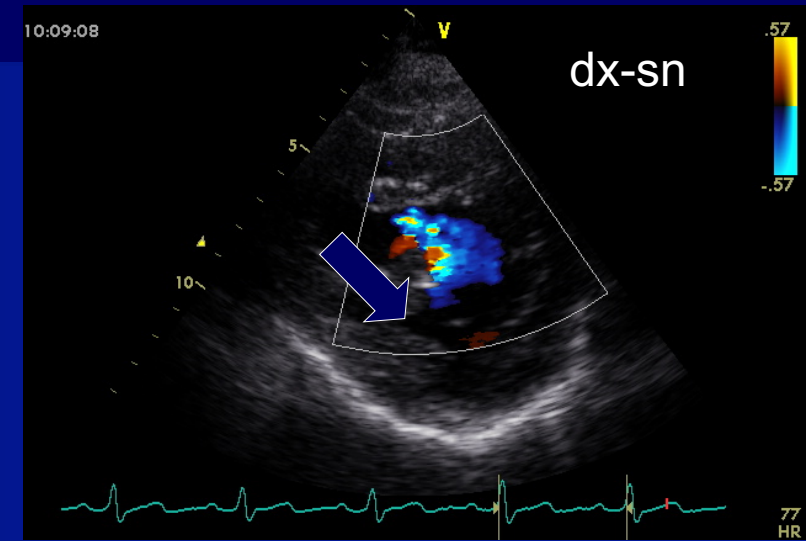
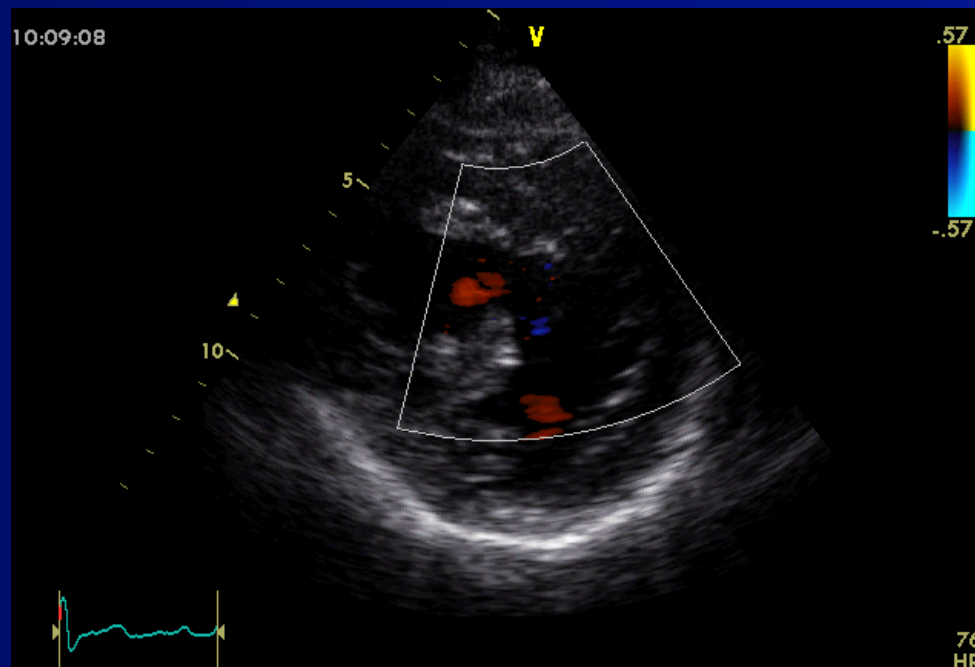
valve pathology

aortic coarctation

Caso clinico 2

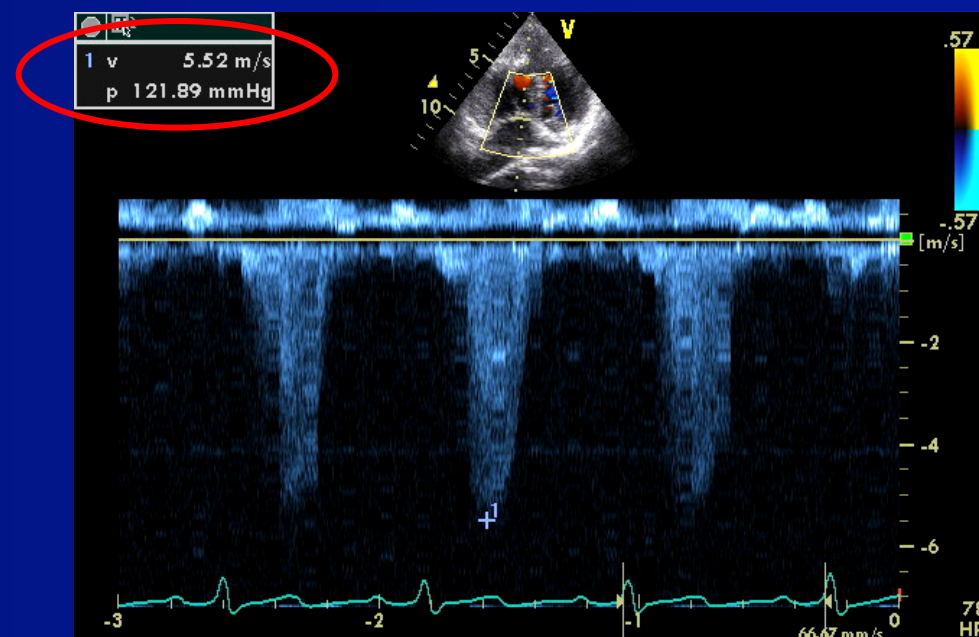
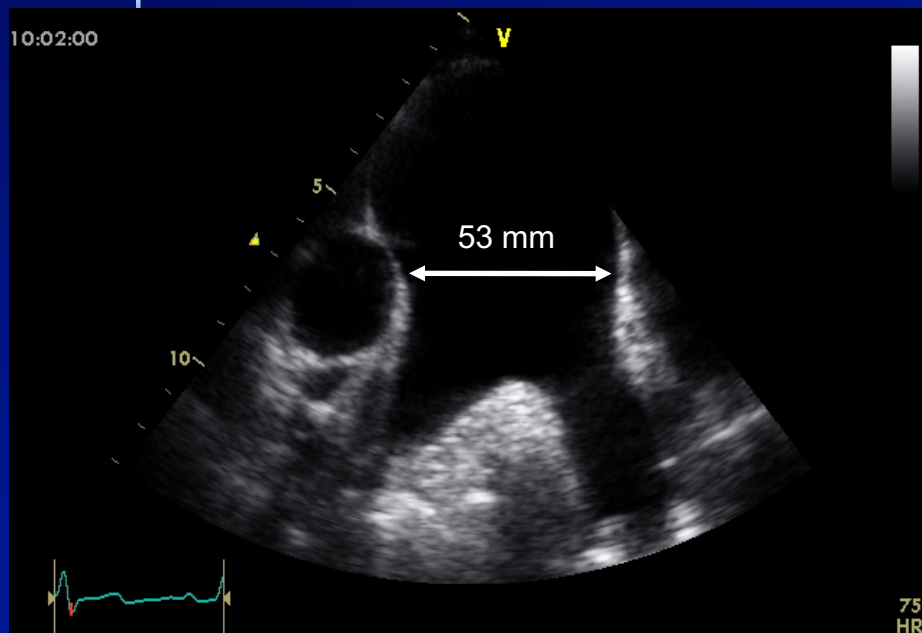
- A.G. anni 39
- Sindrome della coartazione aortica.

Ipertensione polmonare.



Caso clinico 2

- A.G. anni 39
- Sindrome della coartazione aortica.
- Ipertensione polmonare.



A 3.5 anni follow up.

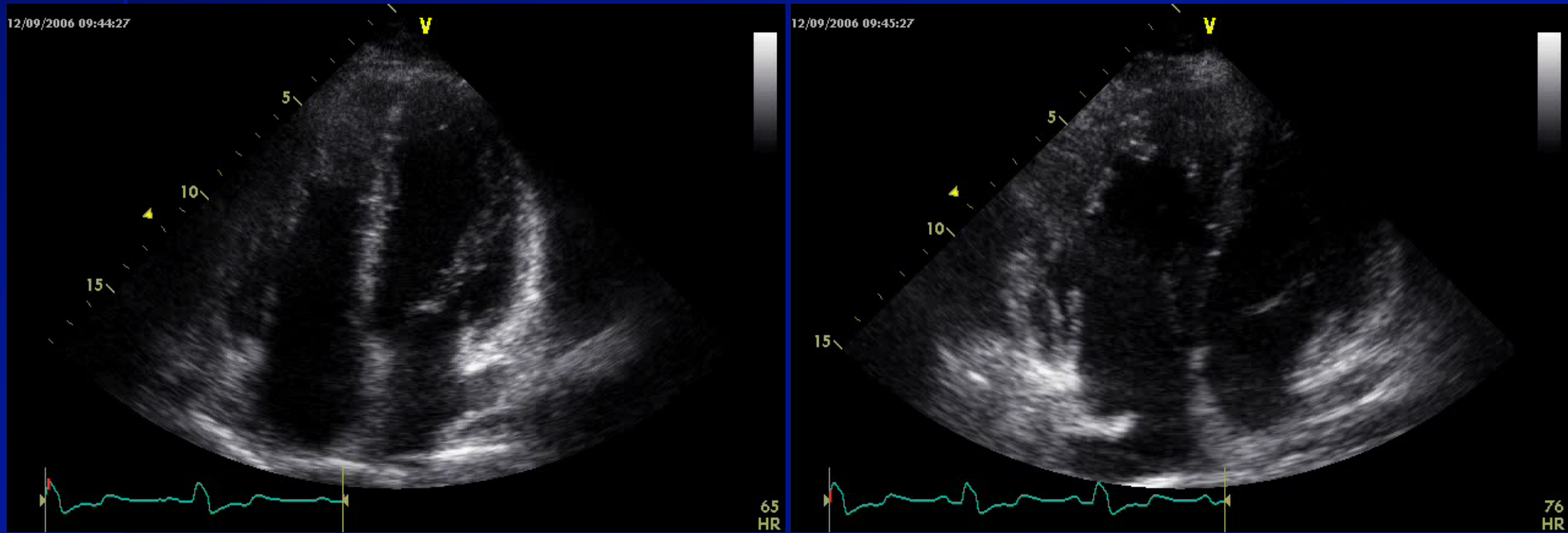
Ottimizzazione terapia di background + bosentan (125 mg x 2) + sildenafil (20 mg x 3 al di).

II classe funzionale.

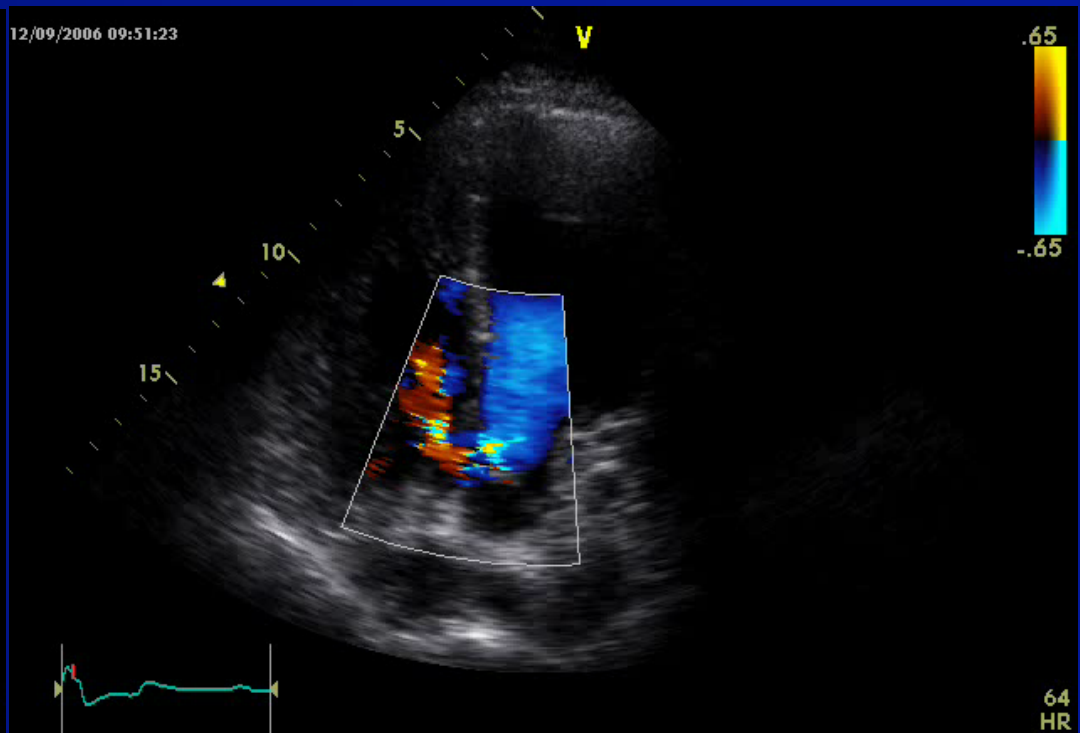
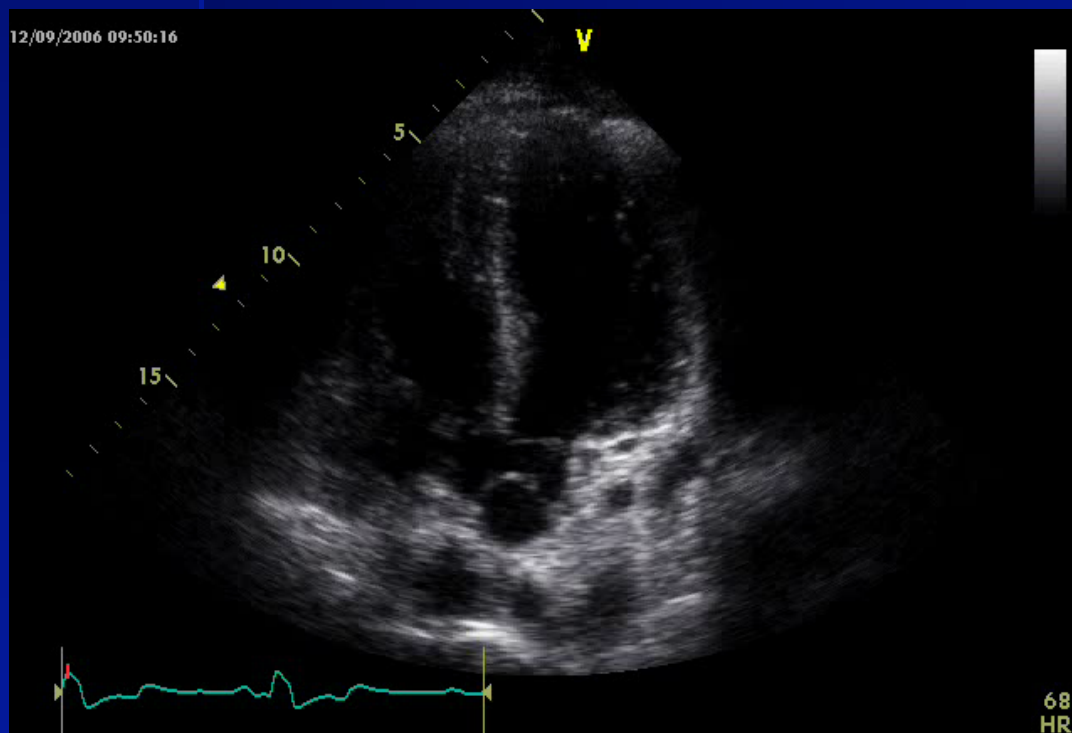
Caso clinico 3: una strana forma di "ipertensione polmonare"

- AS anni 43
- Sesso M
- II classe NYHA
- Diagnosi di DIV in età pediatrica.
- Cateterismo cardiaco nel 1982 (19 a): DIV + ipertensione polmonare.
- Sat O2 95%.
- PAS 110/60 mmHg.

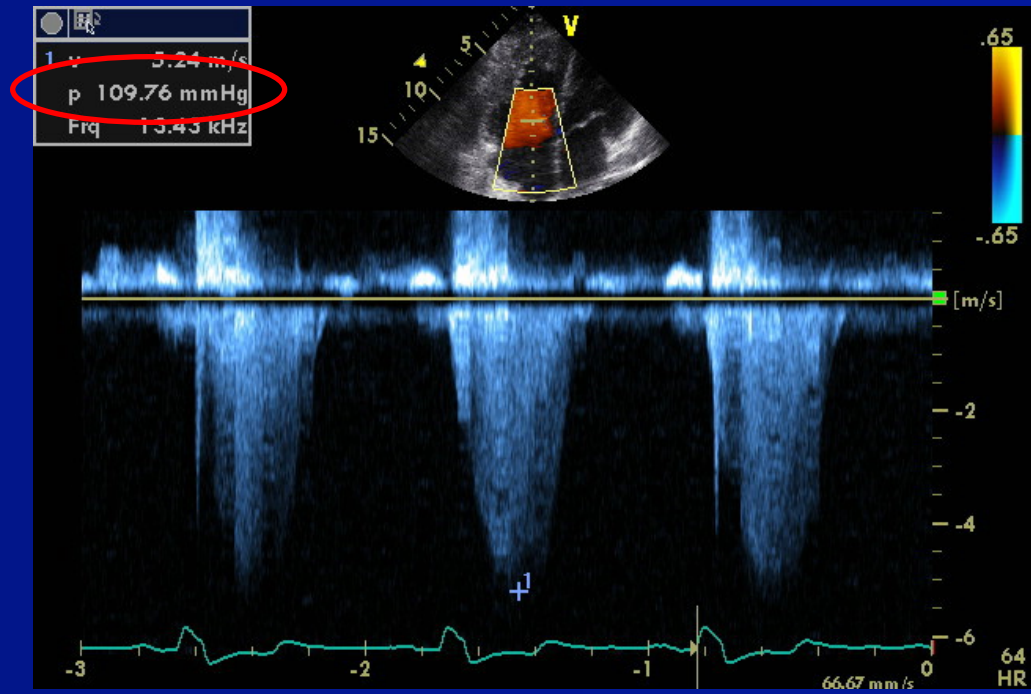
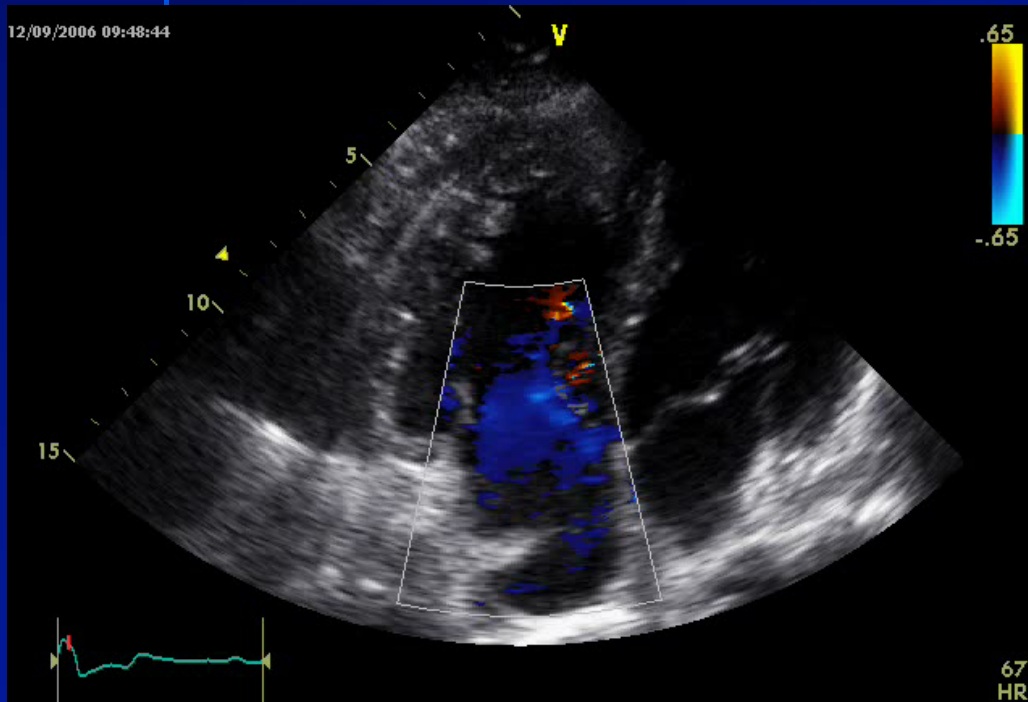
4 camere apicale



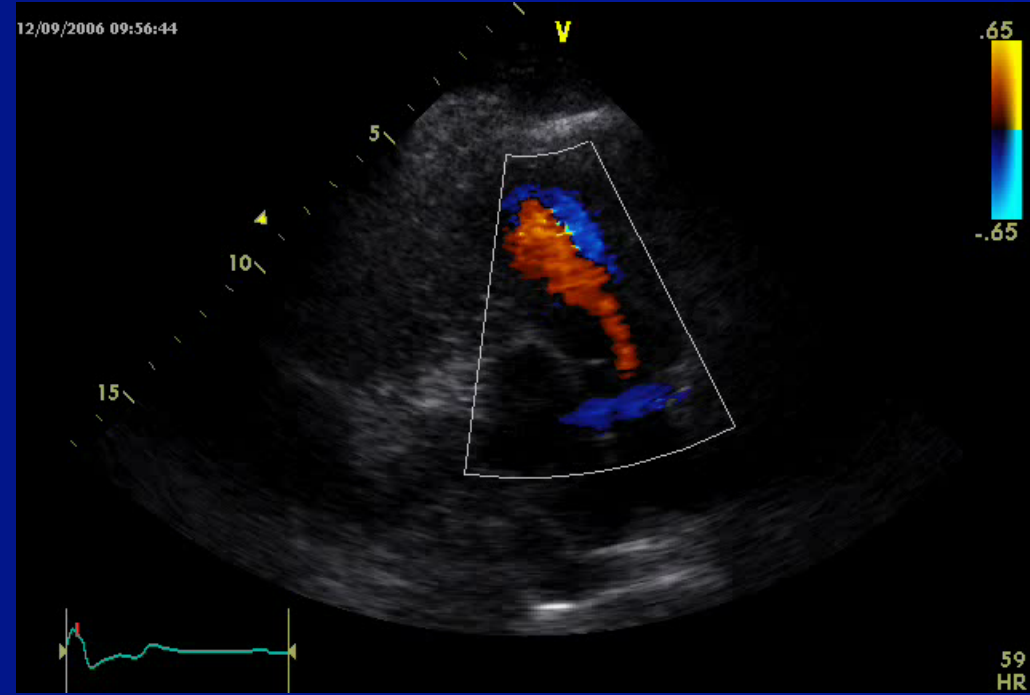
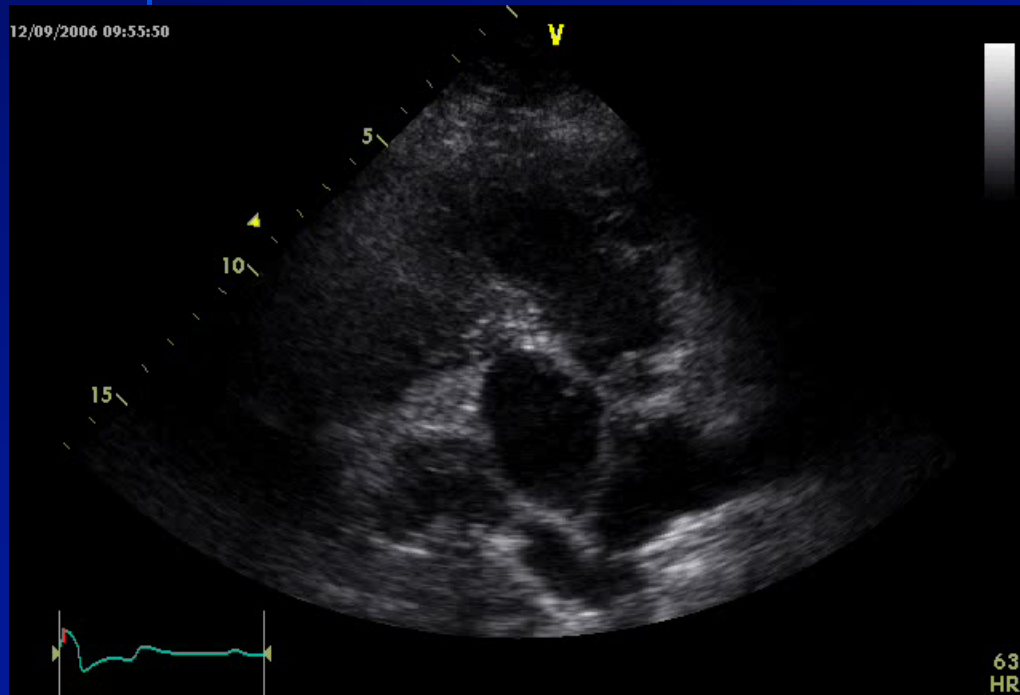
5 camere apicale: DIV perimembranoso



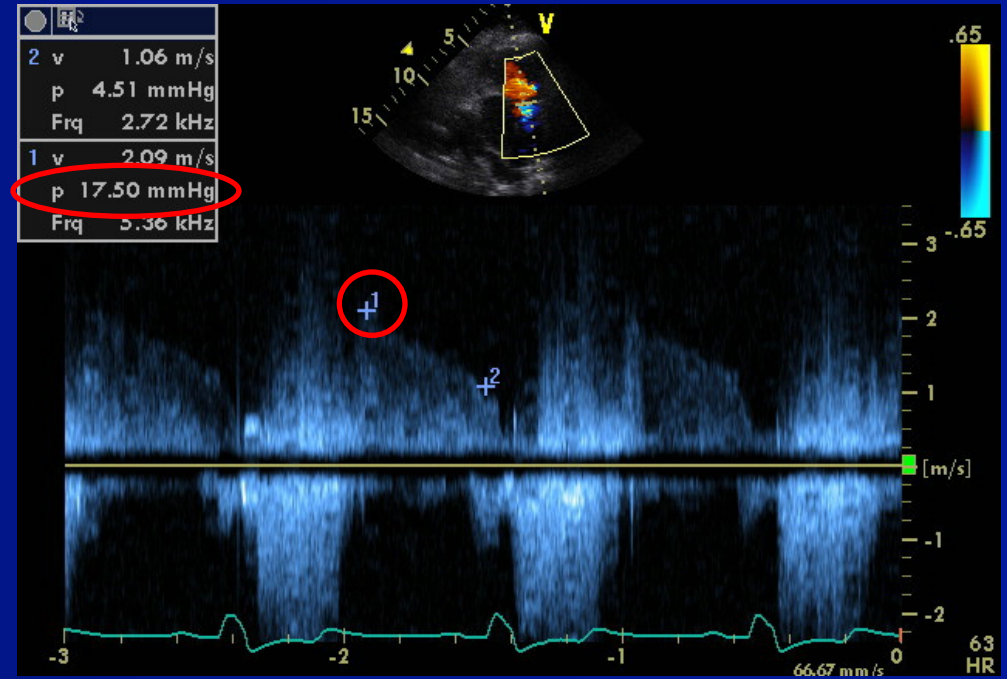
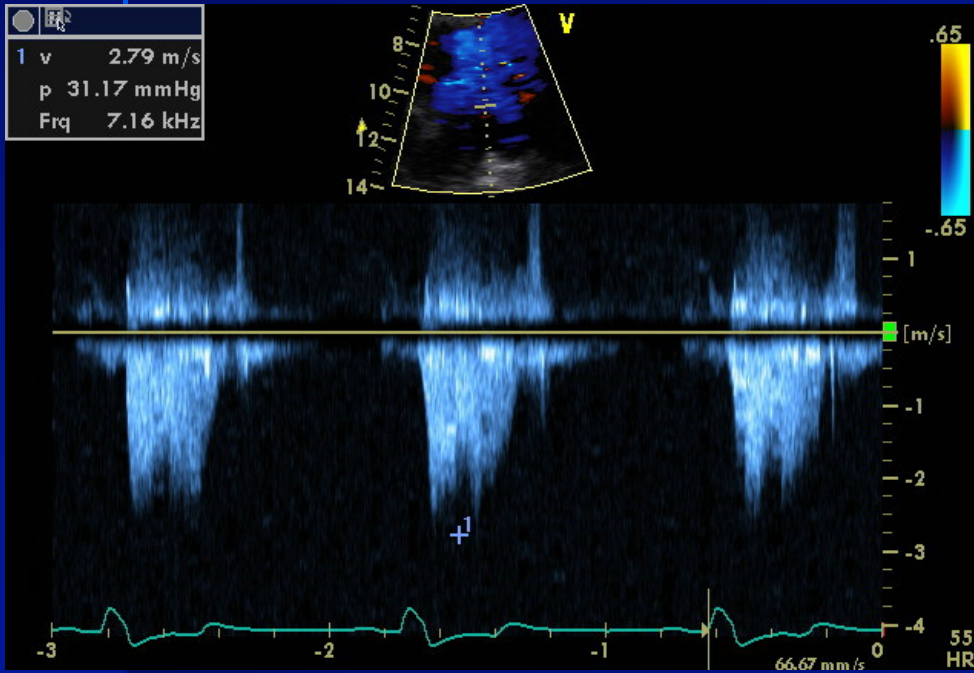
IT ed elevate pressioni



Parasternale short axis: piano valvolare Ao



Parasternale short axis

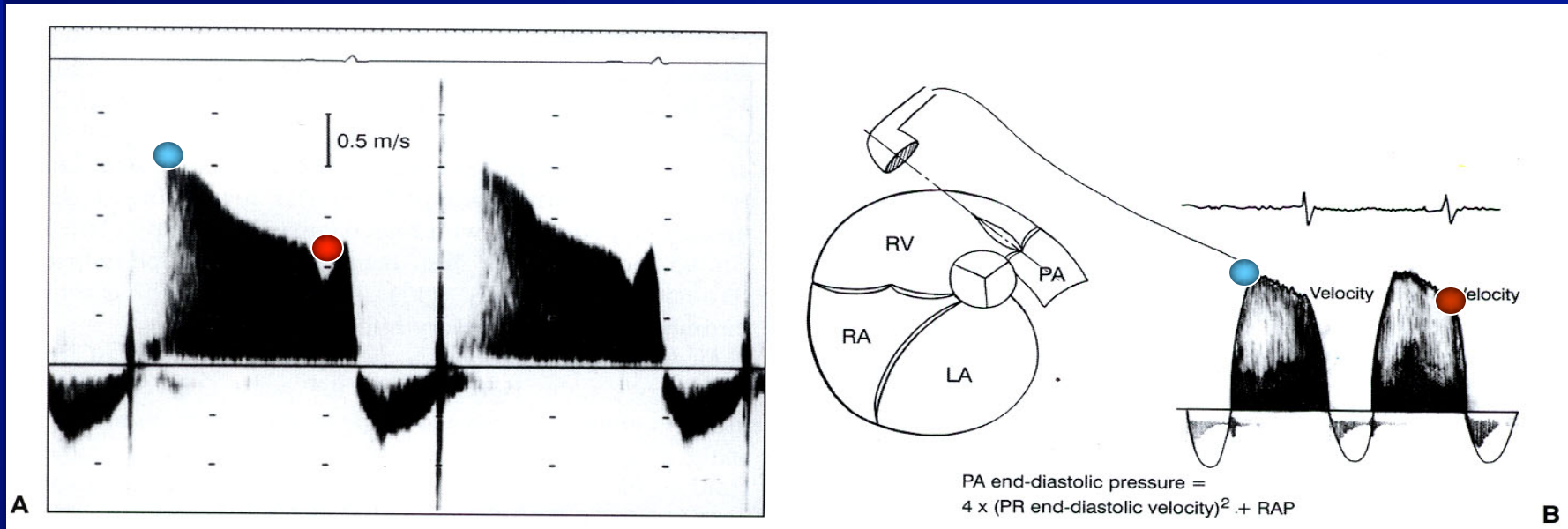


Pressione polmonare media e diastolica

Pressione polmonare media = Press. Protodiastolica ●

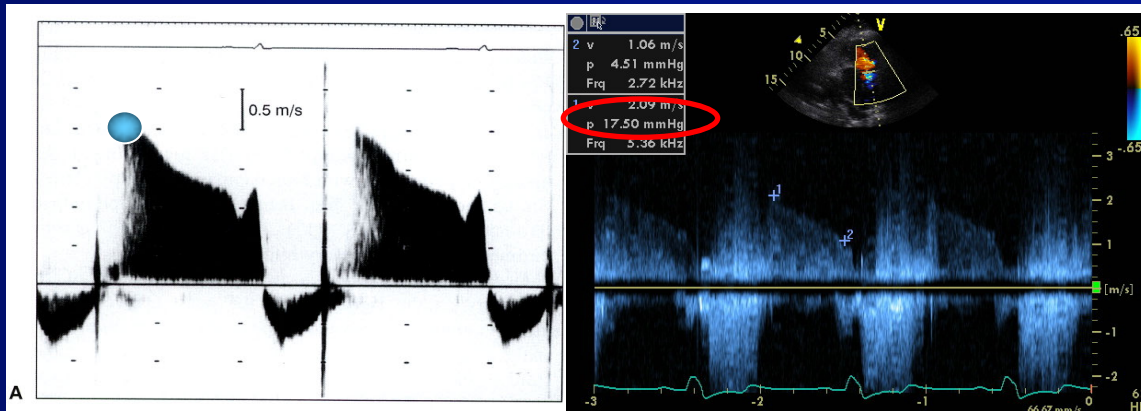
(Masuyama)

Pressione polmonare diastolica = Press. Telediastolica ● + RAP



Pressione polmonare media e diastolica

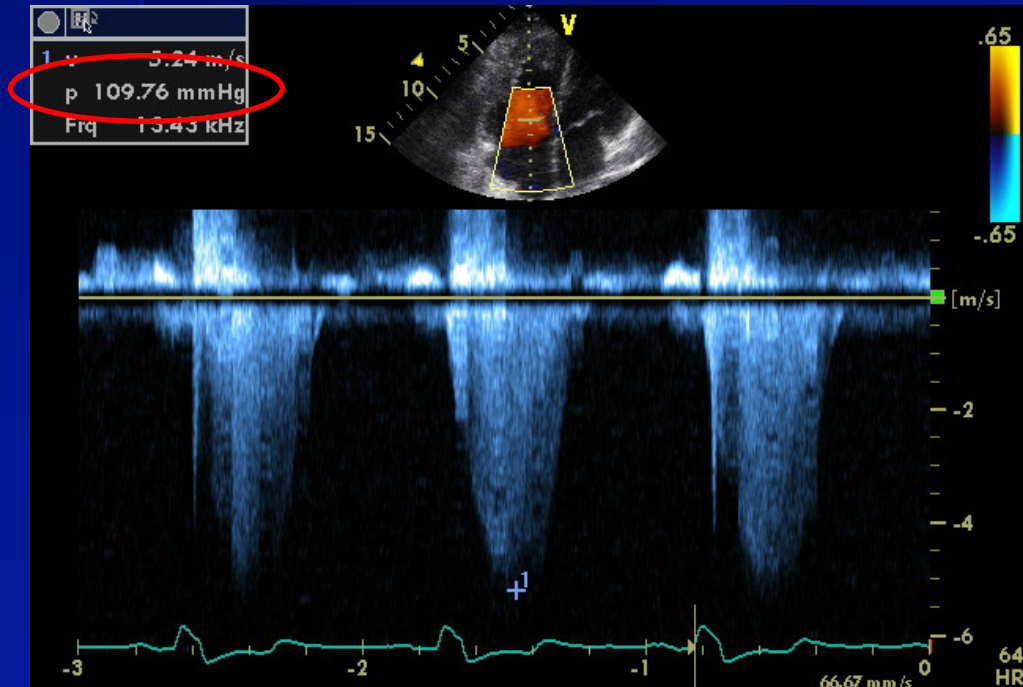
Pressione polmonare media = Press. Protodiastolica ○



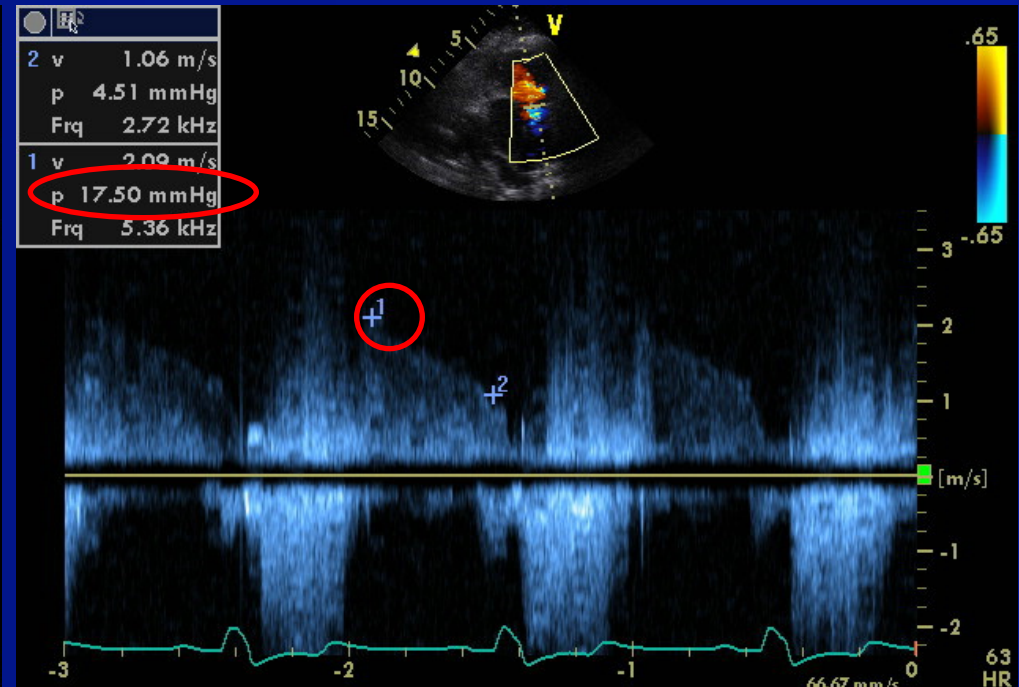
mPAP: 17 mmHg



Quale pressione polmonare?

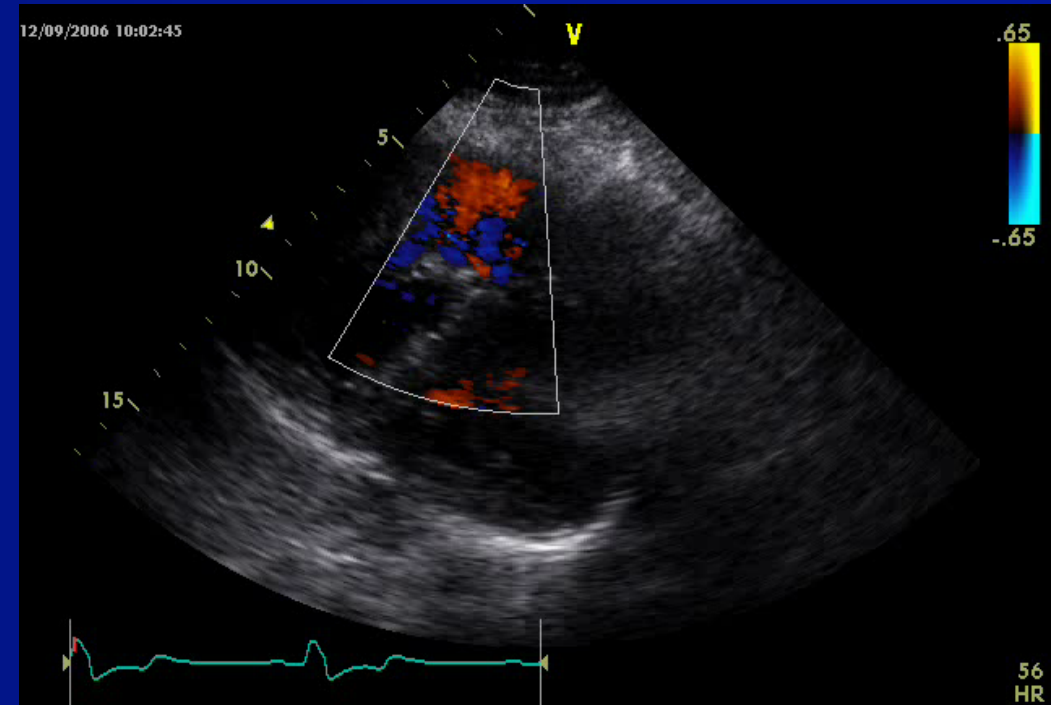
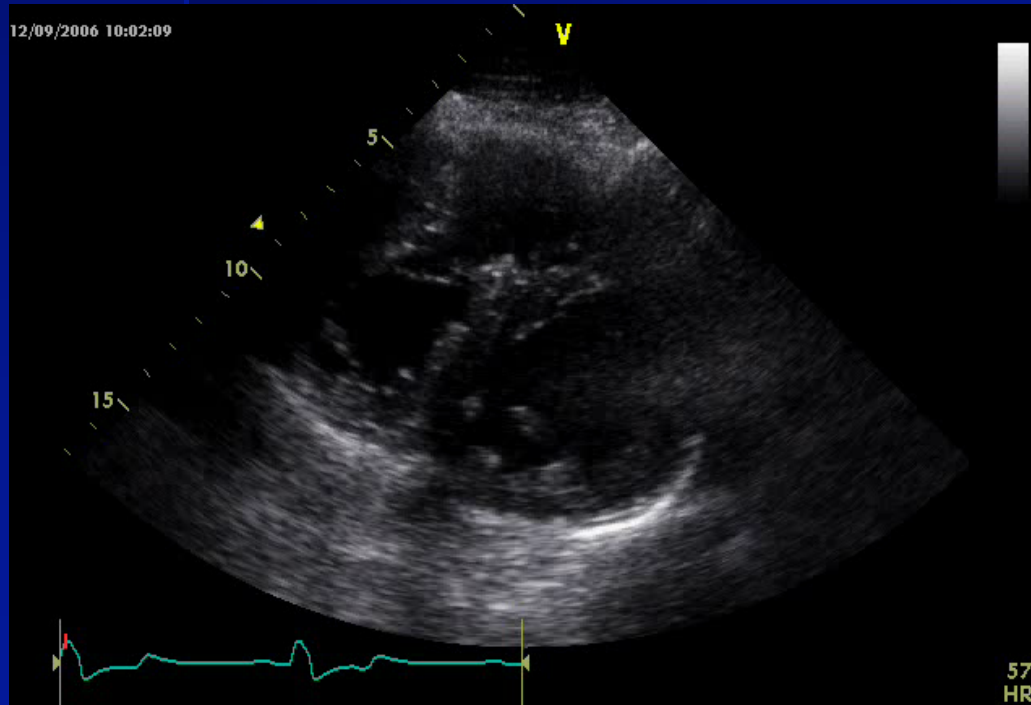


PAP sistolica 119 mmHg

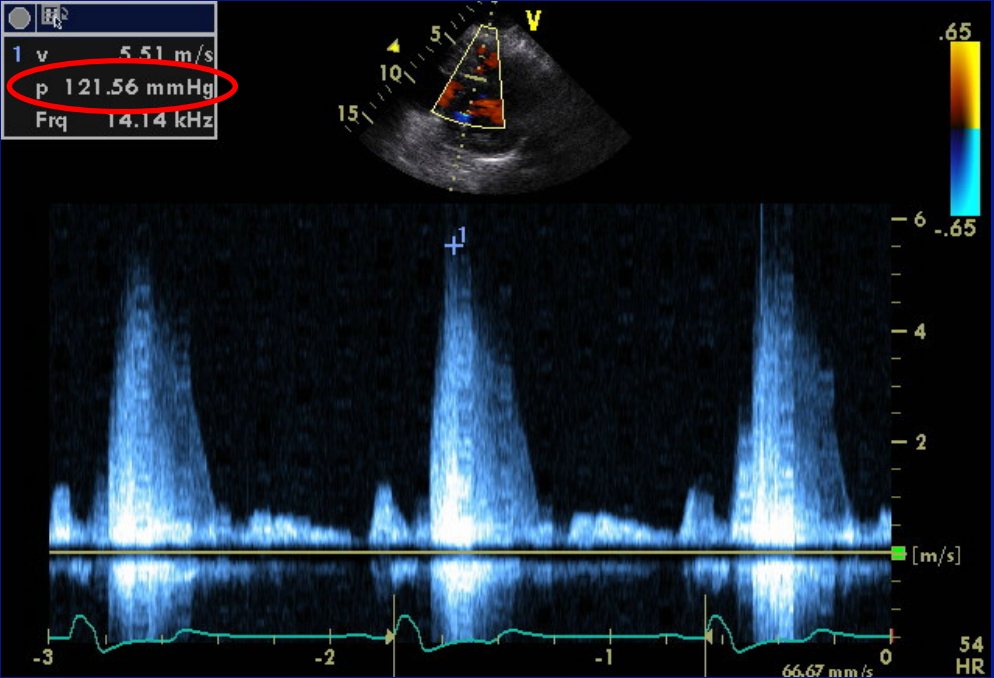
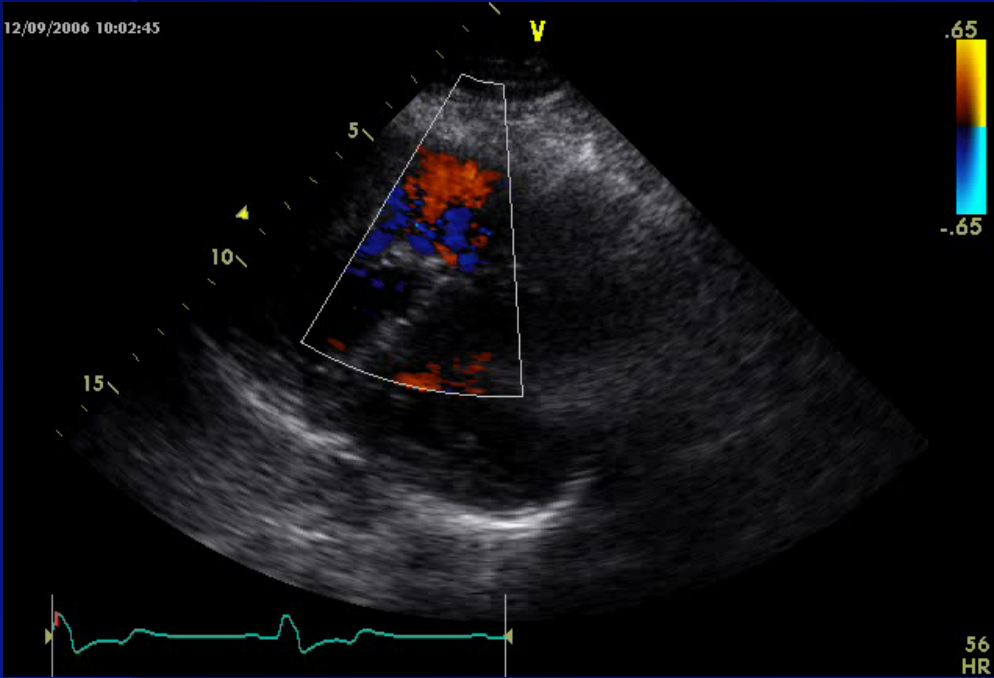
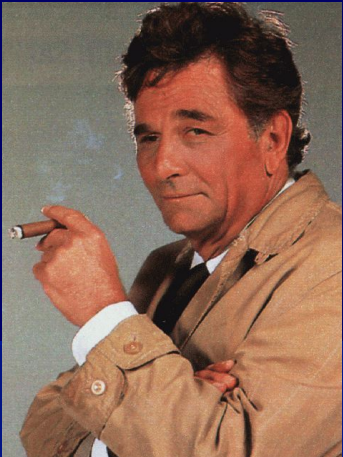


PAP media 17 mmHg

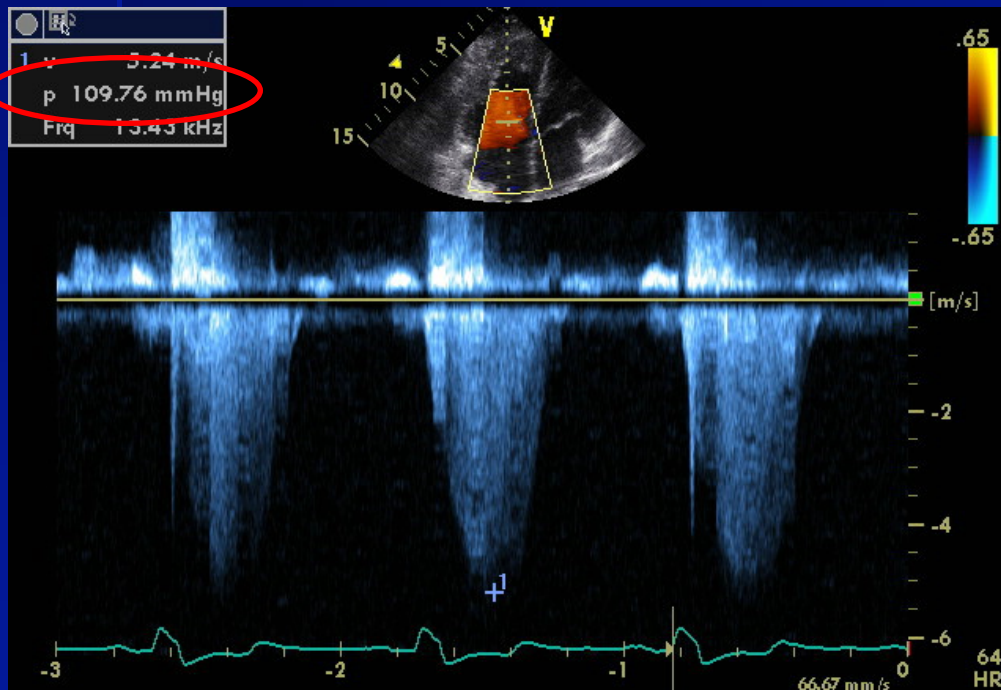
Parasternale short axis: ventricoli



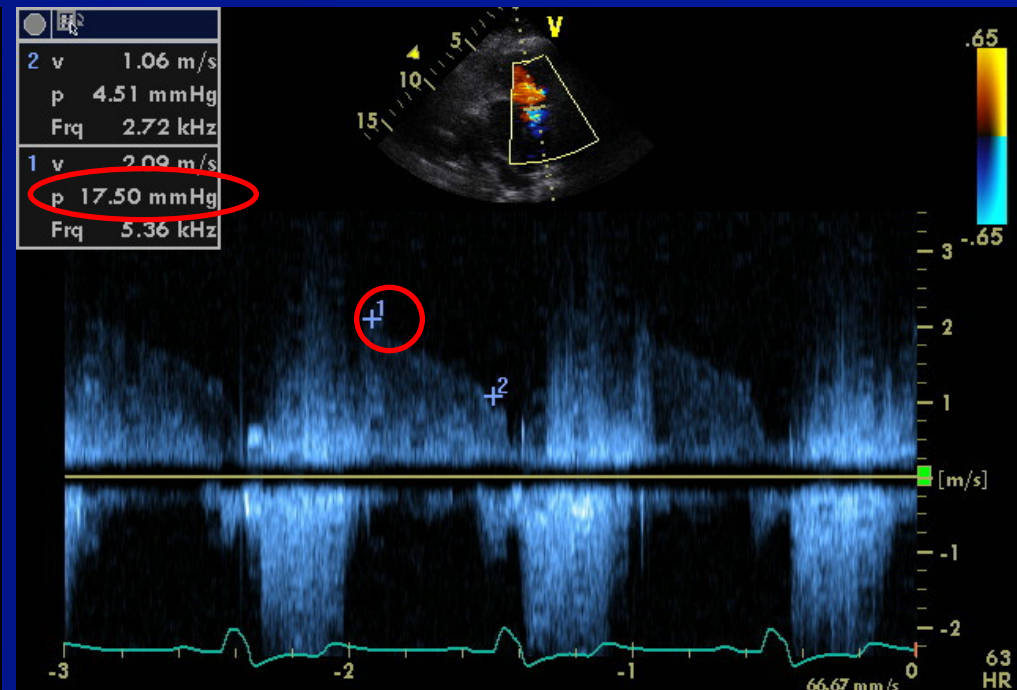
Caso 3



Quale pressione polmonare?



PAP sistolica 119 mmHg



PAP media 17 mmHg

Caso clinico 3: una strana forma di "ipertensione polmonare": conclusioni

NON è ipertensione polmonare!

Diagnosi di DIV + stenosi medioventricolare destra (reattiva).

Ricaduta clinica:

NON vanno dati farmaci specifici per la PAH (bosentan, sildenafil ...)

Terapia: chirurgica