

**CORSO DI FORMAZIONE SIEC
in
ECOCARDIOGRAFIA
PEDIATRICA**

Firenze Hotel Michelangelo

18-20 febbraio 2016

Patologia Ostruttiva dell'Efflusso Destro
Stenosi polmonare
Atresia Polmonare a setto intatto

Dr Nadia Assanta

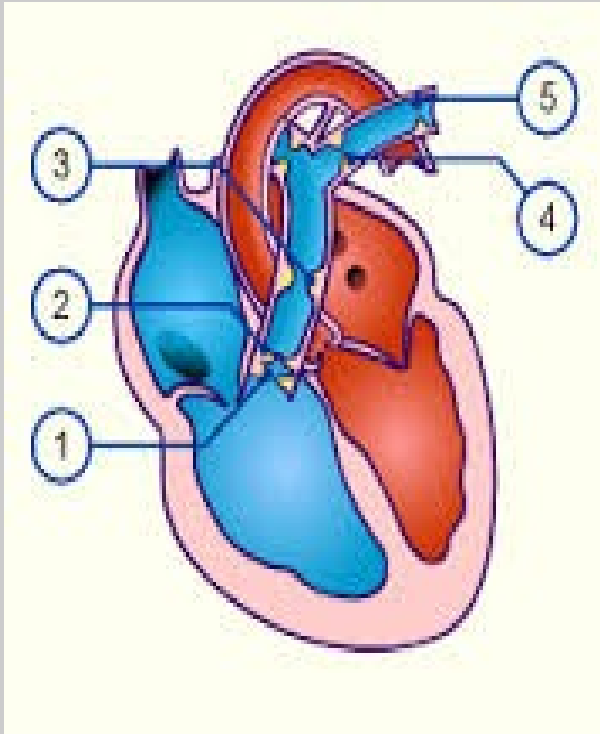
Ospedale del Cuore -FTGM-Massa

STENOSI POLMONARE

Varianti anatomiche

Ostruzione al flusso dal Vdx al circolo arterioso polmonare:

- Subvalvolare (infundibulare)
 - Valvolare
 - Sopravalvolare
 - Rami polmonari
-
- 10% di tutte le CHD
 - M=F



STENOSI POLMONARE

ISOLATA O ASSOCIATA A SINDROMI



S. di Noonan



S. di Williams



S. di Alagille

STENOSI POLMONARE

Diagnosi prenatale

Forme critiche dotto di Botallo dipendenti



STENOSI POLMONARE

Valvolare

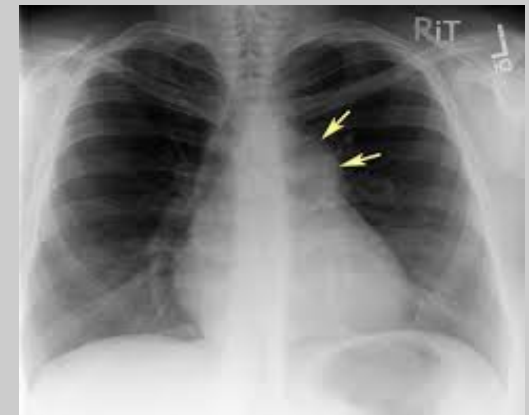
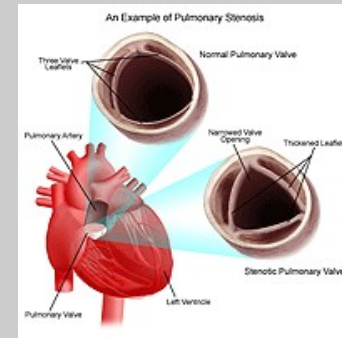
- Generalmente isolata
- Talora associata a DIA
- Anatomia: Doming o displasia della valvola polmonare
- Dilatazione post stenotica dell'arteria polmonare
- Ventricolo destro ipertrofico
- Insufficienza tricuspидale da sovraccarico pressorio Vdx



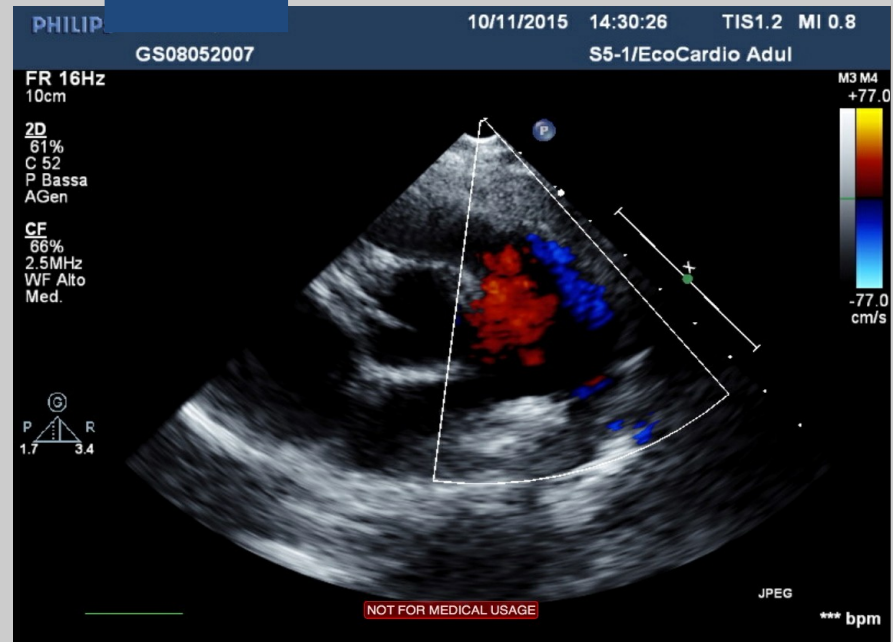
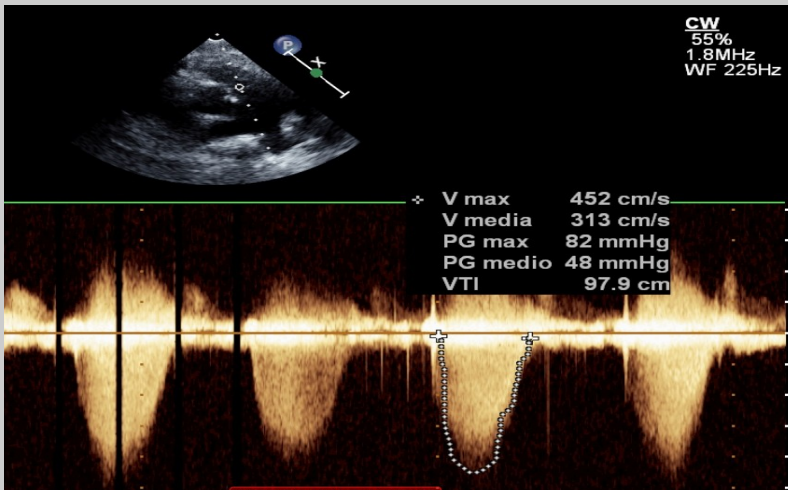
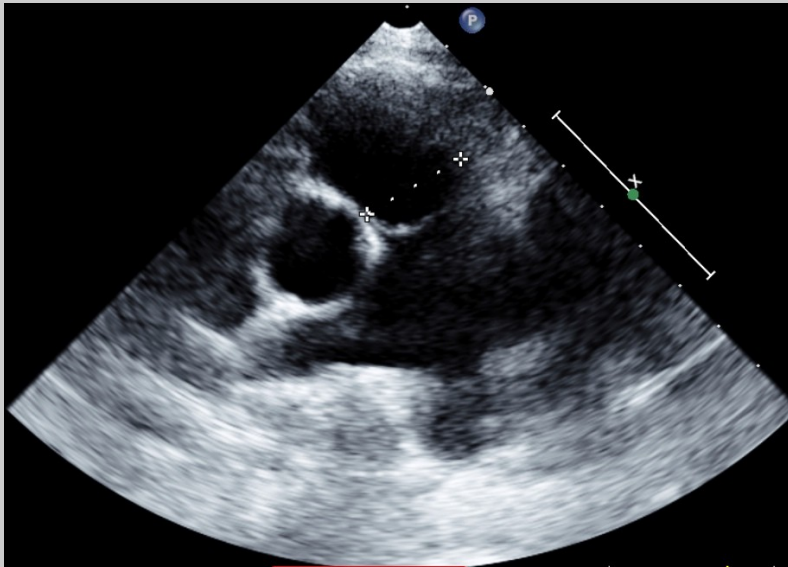
STENOSI VALVOLA POLMONARE

Valvola Polmonare

- Ispessita
- Displastica
- Doming sistolico
- Bicuspide o con fusione delle commissure
- Turbolenza al Doppler e gradiente anterogrado
- IP variabile
- Dilatazione post stenotica della AP



STENOSI POLMONARE

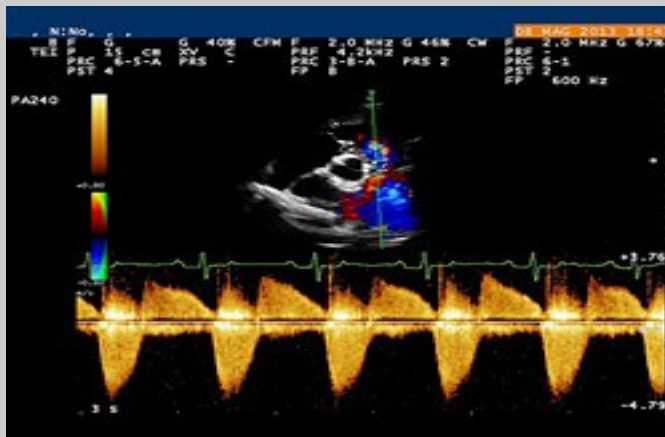


STENOSI VALVOLA POLMONARE

Valvola Polmonare

- Stenosi polmonare lieve : PG <50 mmhg
- Stenosi polmonare moderata : PG 50-80 mmhg
- Stenosi polmonare severa: PG >80 mmhg

- Importante la dimensione dell'anello polmonare

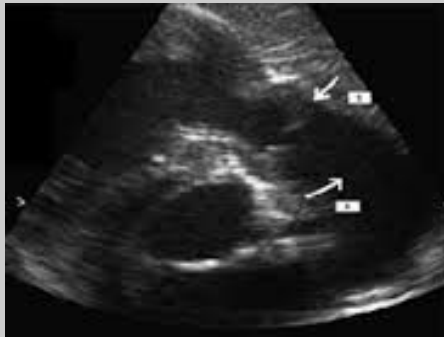


STENOSI VALVOLA POLMONARE

Valvola Polmonare

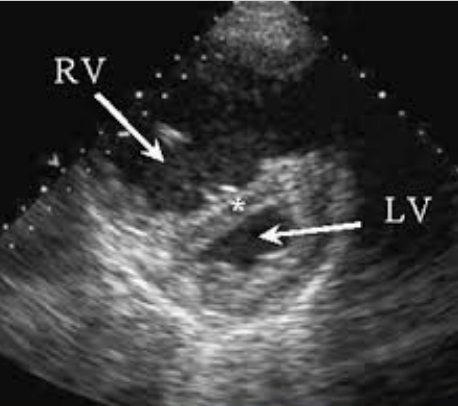
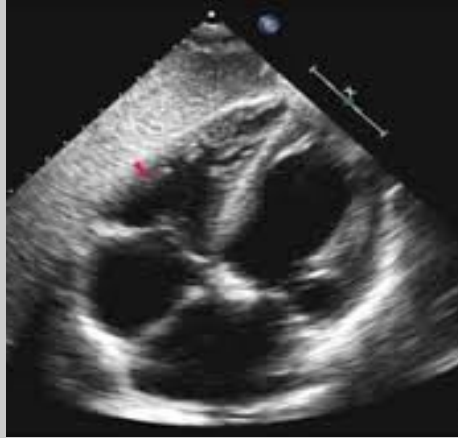
CLASSIFICAZIONE ANATOMICA: La stenosi polmonare presenta due sottotipi anatomici:

- TIPO A (più frequente) : caratterizzato dalla fusione dei lembi polmonari in presenza di un annulus polmonare di normali dimensioni e frequente dilatazione postvalvolare.
- TIPO B: caratterizzato da ipoplasia dell'annulus della valvola polmonare (con lembi valvolari rudimentali spesso fusi) e talvolta anche da ipoplasia dell'arteria polmonare.



STENOSI POLMONARE

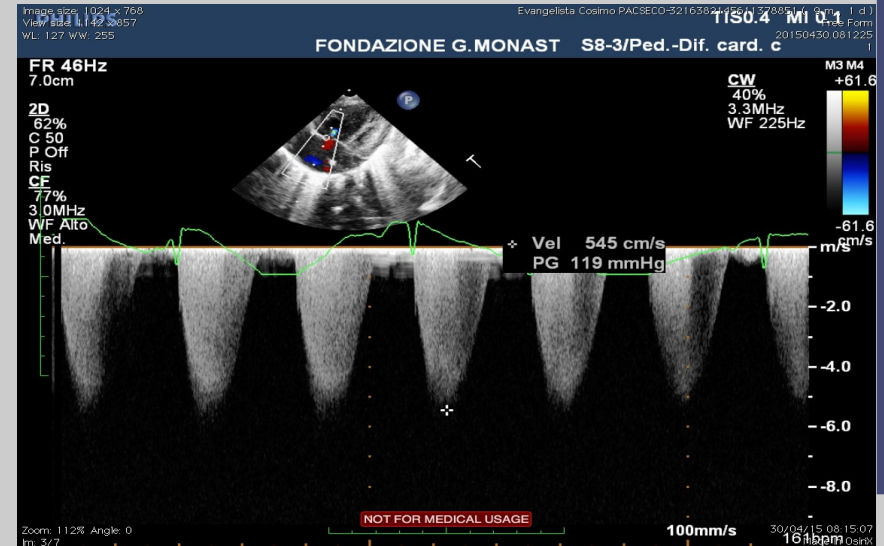
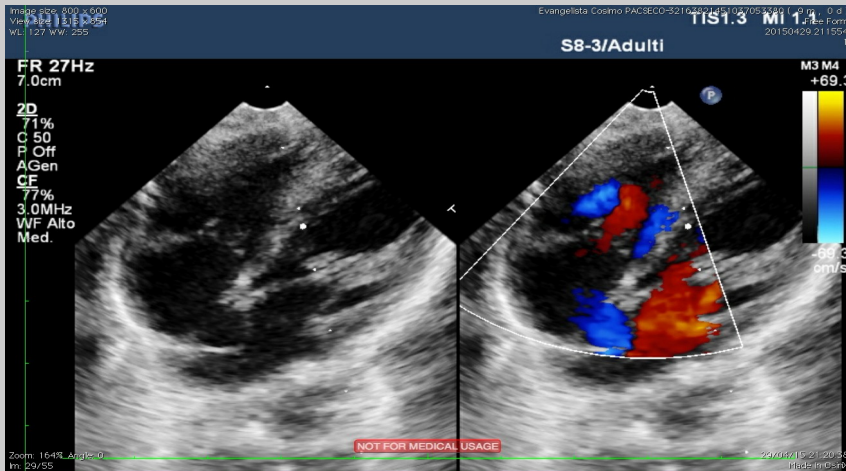
Ventricolo destro



- Dilatato
- Ipertrofico
- Sovraccarico pressorio con tipico movimento del setto IV

STENOSI POLMONARE

Valvola tricuspide

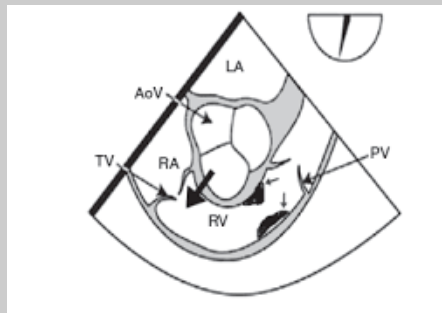
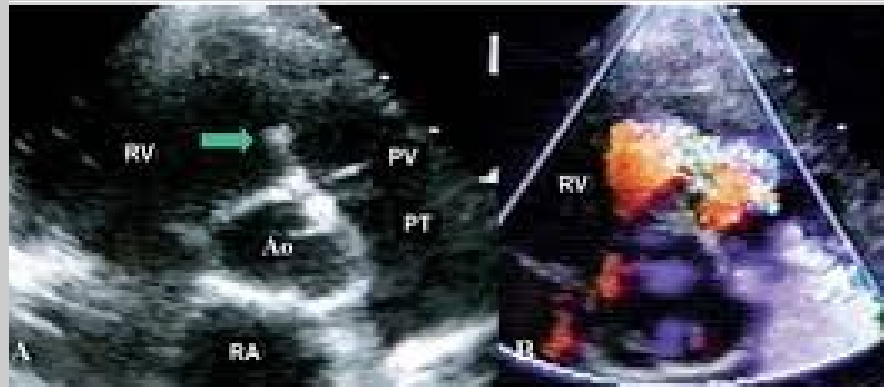
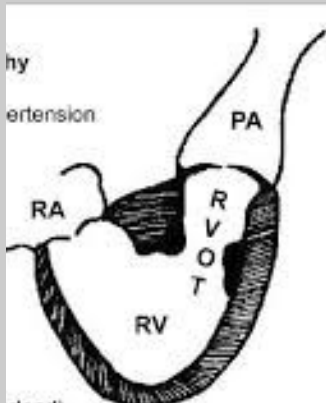
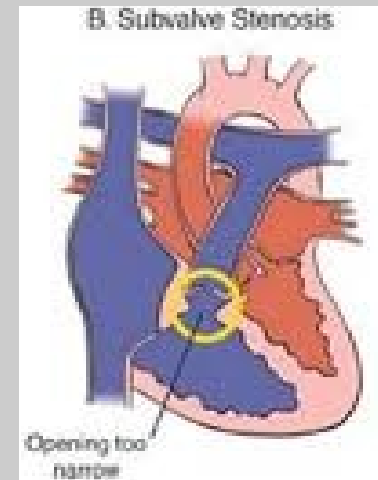


- Insufficienza tricuspide moderata-severa
- Elevate pressioni in ventricolo destro

STENOSI POLMONARE

Infundibulare

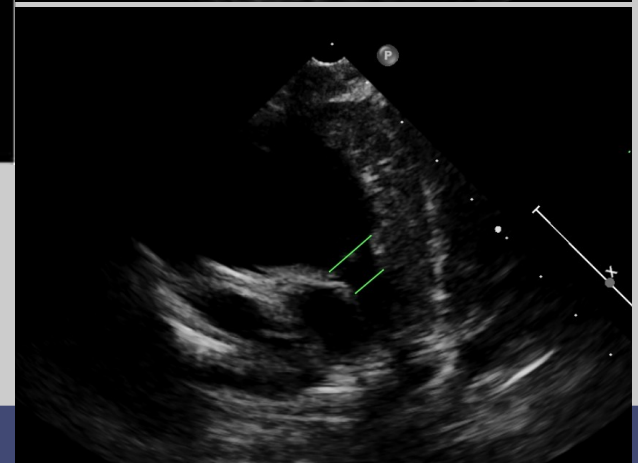
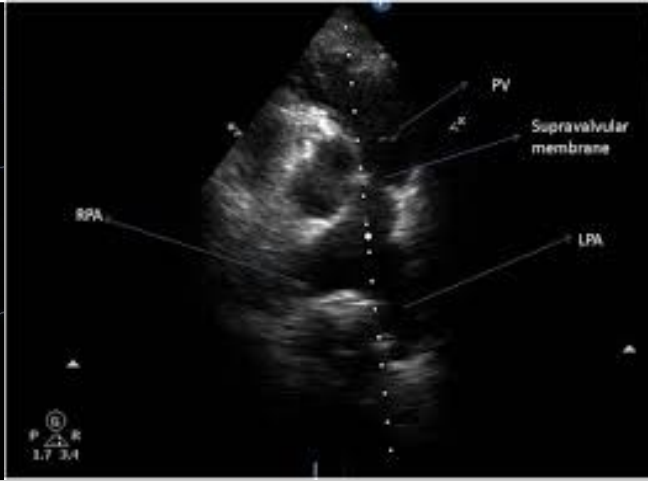
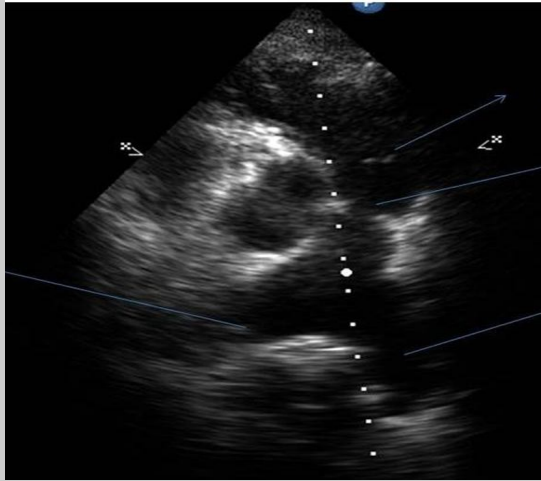
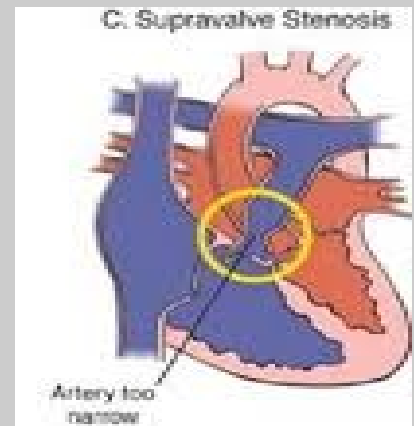
- Al di sotto dell'anello valvolare
- Spesso associata a difetto interventricolare
- Tipica la stenosi infundibulare della TOF



STENOSI POLMONARE

Sopravalvolare

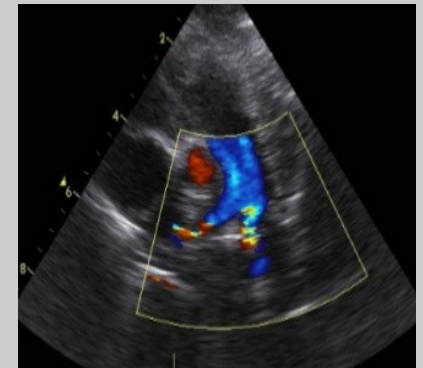
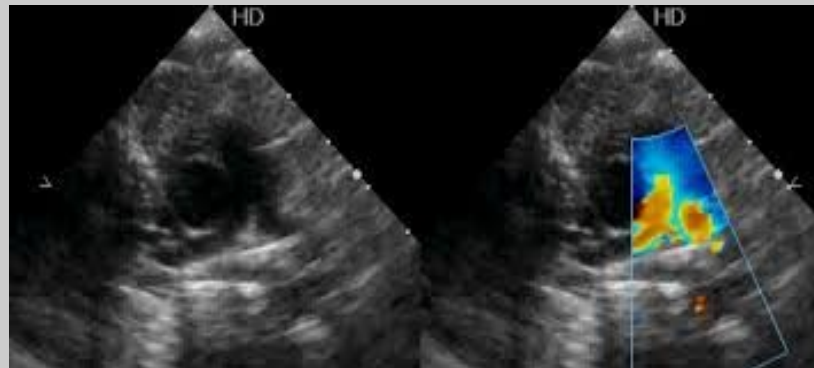
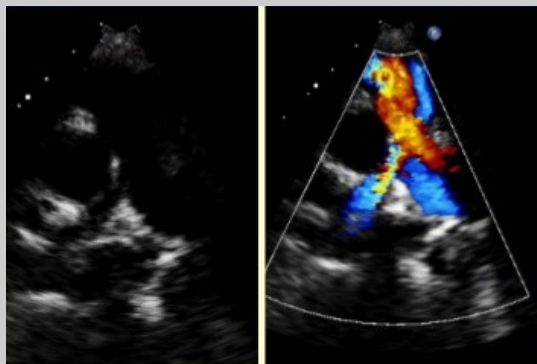
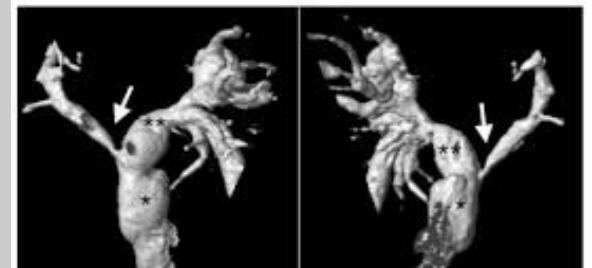
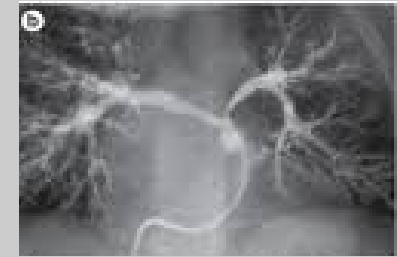
- Al di sopra dell'anello valvolare
- Spesso associata a S di Noonan o Williams
- Non dilatazione post stenotica



STENOSI POLMONARE

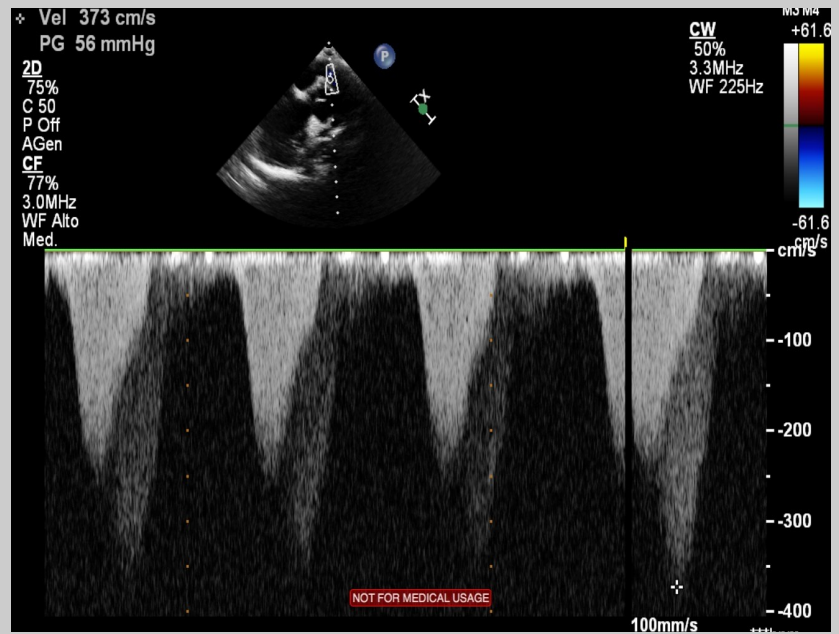
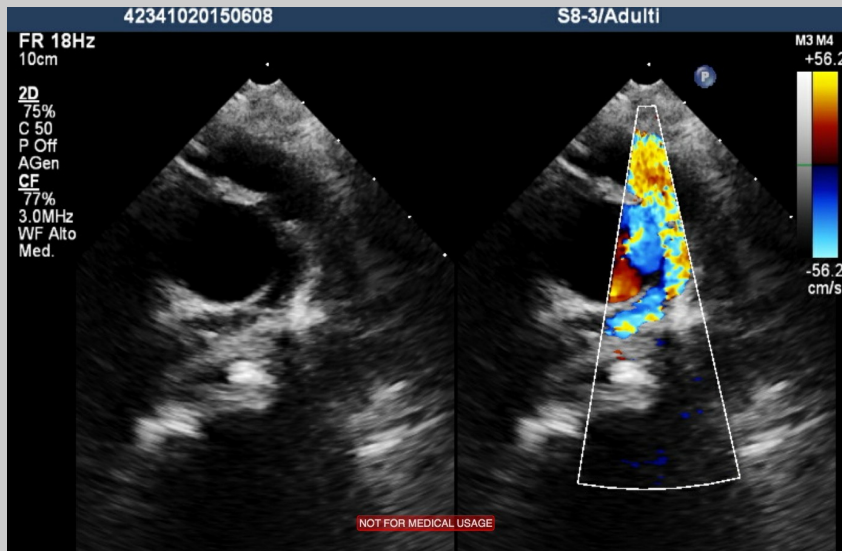
Rami polmonari

- Ipoplasia rami polmonari (S di Williams)
- Stenosi isolata rara (ramo polm sinistro)
- Associata ad altra CHD (es TOF)
- RMN o AngioTAC



STENOSI POLMONARE

Rami polmonari



STENOSI POLMONARE: trattamento

Table 13 Indications for intervention in right ventricular outflow tract obstruction

Indications	Class ^a	Level ^b
RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is >64 mmHg (peak velocity >4m/s), provided that RV function is normal and no valve substitute is required	I	C
In valvular PS, balloon valvotomy should be the intervention of choice	I	C
In asymptomatic patients in whom balloon valvotomy is ineffective and surgical valve replacement is the only option, surgery should be performed in the presence of a systolic RVP >80 mmHg (TR velocity >4.3 m/s)	I	C
Intervention in patients with gradient <64 mmHg should be considered in the presence of: <ul style="list-style-type: none"> • symptoms related to PS or, • decreased RV function or, • double-chambered RV (which is usually progressive) or, • important arrhythmias or, • right-to-left shunting via an ASD or VSD. 	IIa	C
Peripheral PS, regardless of symptoms, should be considered for repair if >50% diameter narrowing and RV systolic pressure >50 mmHg and/or lung perfusion abnormalities are present	IIa	C

^aClass of recommendation.

^bLevel of evidence.

ASD = atrial septal defect; PS = pulmonary stenosis; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; RVP = right ventricular pressure; TR = tricuspid regurgitation; VSD = ventricular septal defect.

ESC guideline 2010

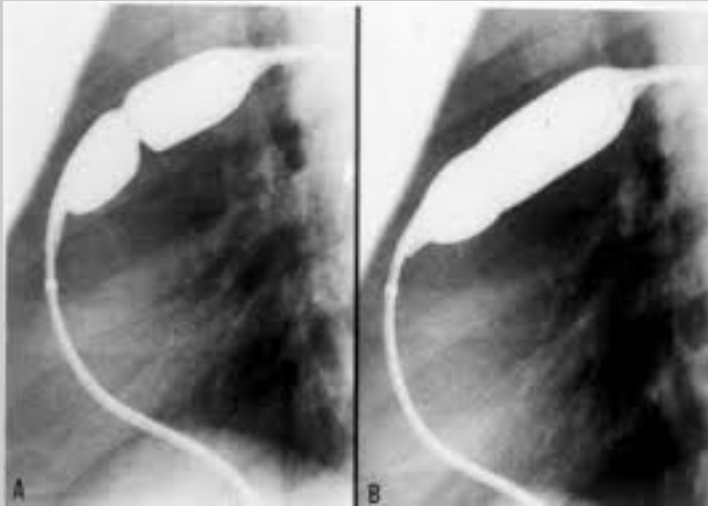
STENOSI POLMONARE

Trattamento

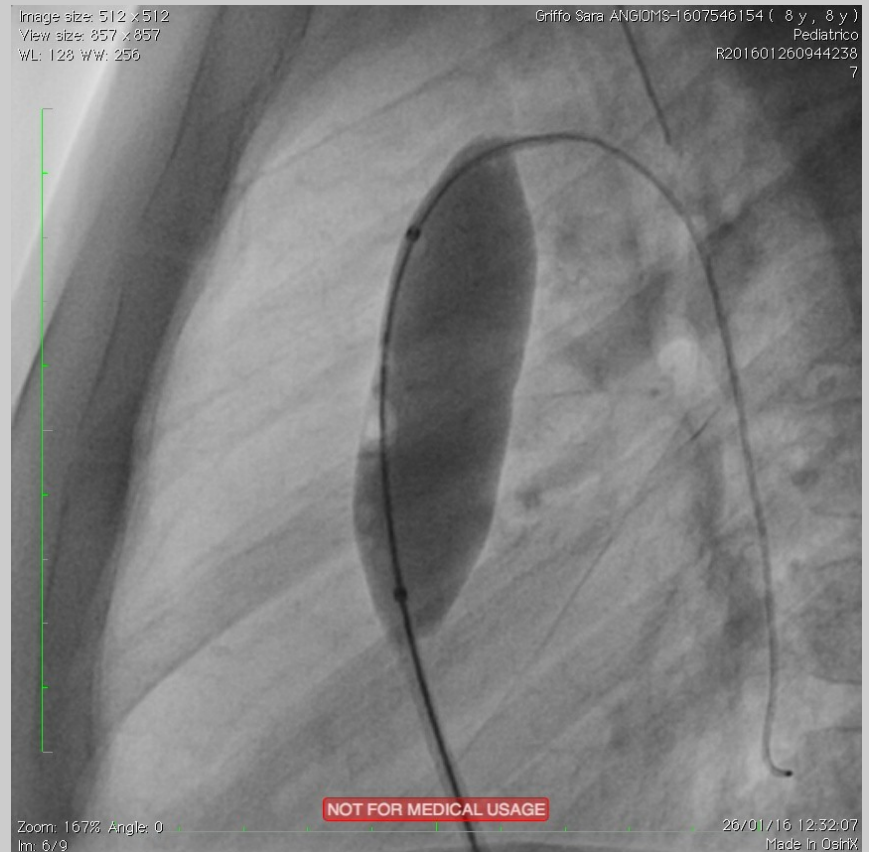
- Valvuloplastica polmonare percutanea (dilatazione con palloncino) nelle SP valvolari con buon anello
- Valvulotomia chirurgica
- Valvuloplastica chirurgica
- Transannular patch (Ipoplasia dell'anello)
- Allargamento con patch (Arteria polmonare e rami)
- Angioplastica e/o stent per stenosi rami polmonari

STENOSI POLMONARE

Trattamento SP

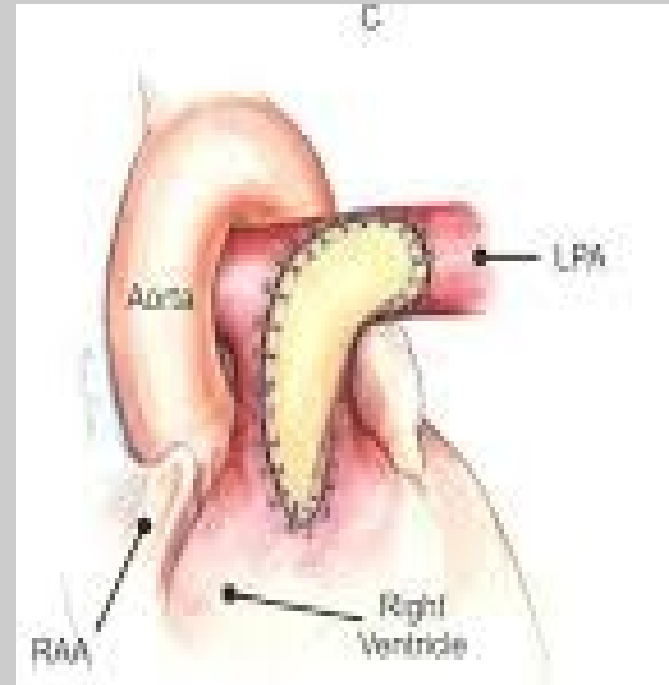
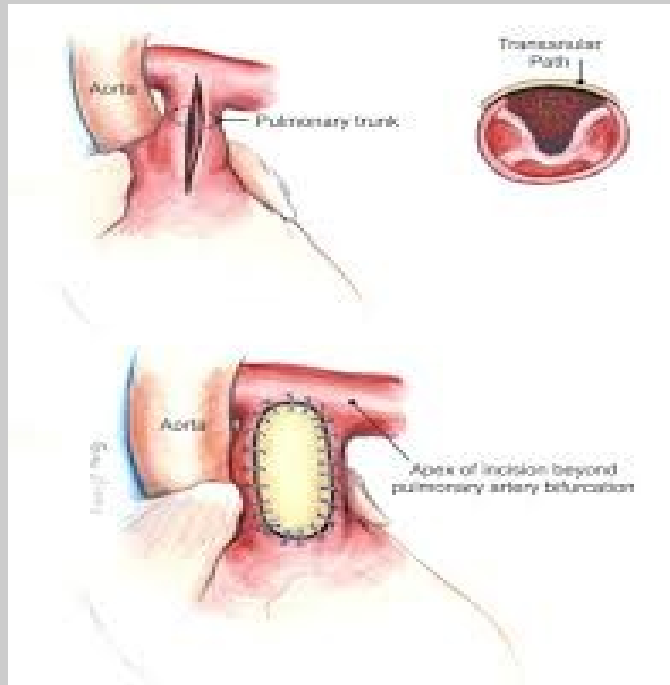


- Angioplastica con palloncino percutanea



STENOSI POLMONARE

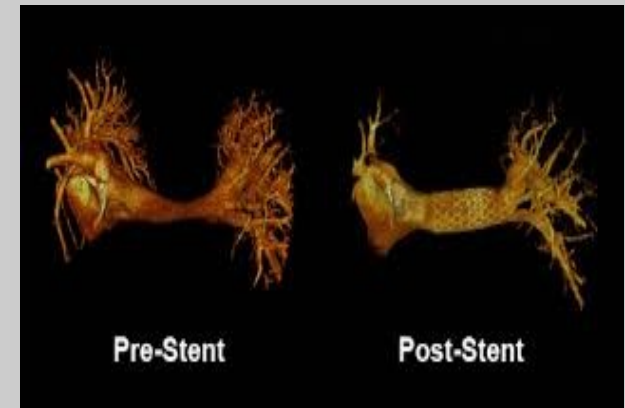
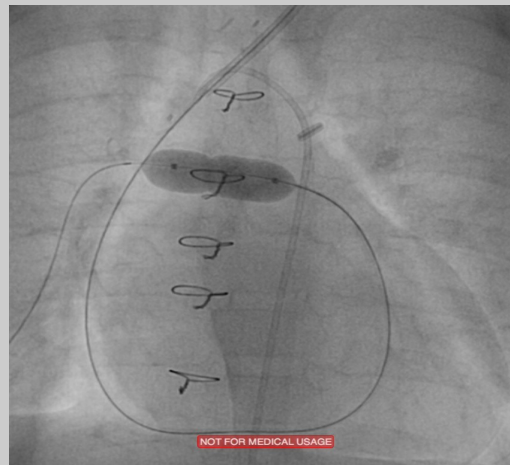
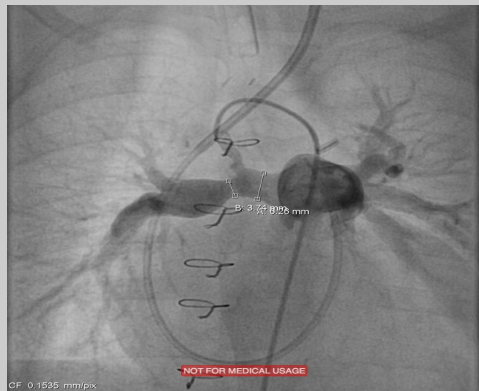
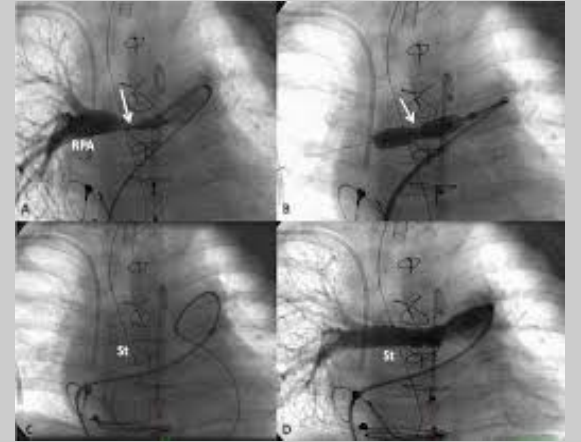
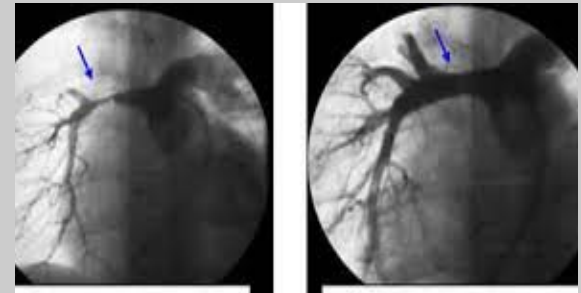
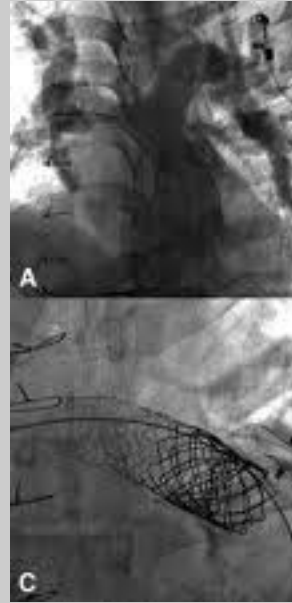
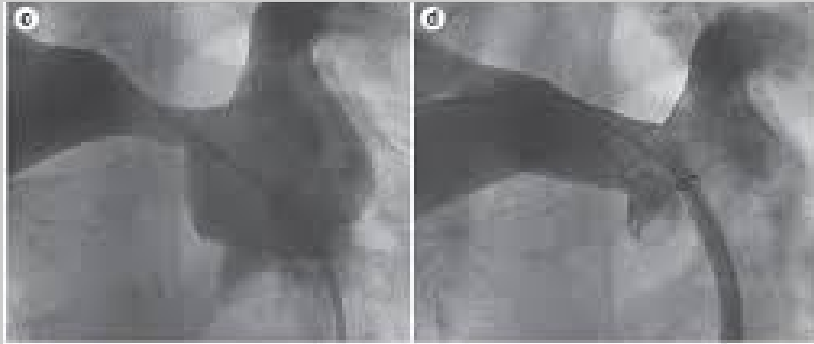
Trattamento chirurgico



- Allargamento con Patch

STENOSI POLMONARE

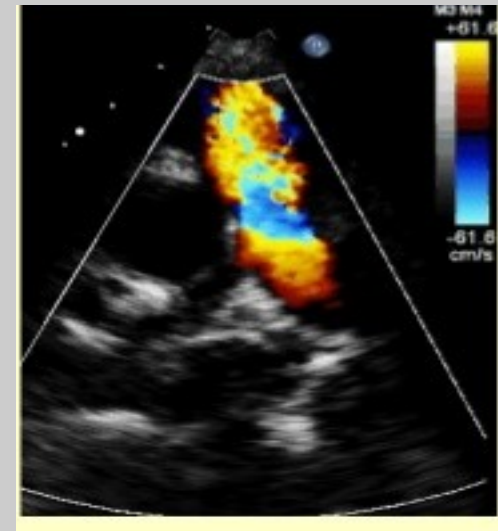
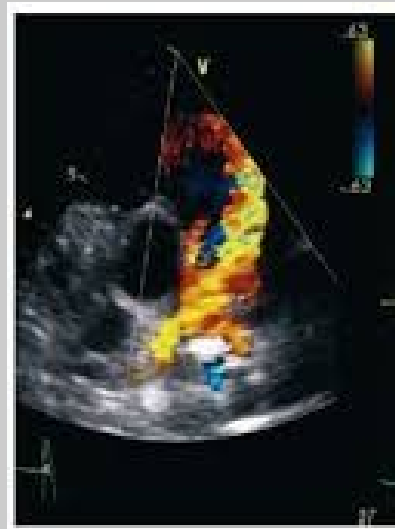
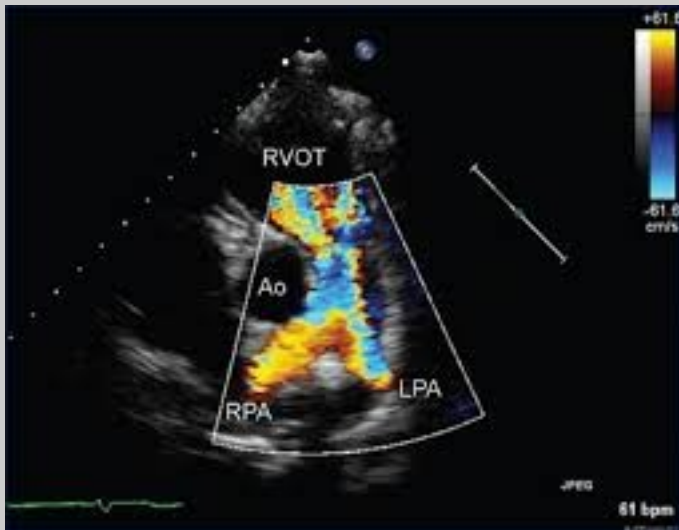
Trattamento rami



STENOSI POLMONARE

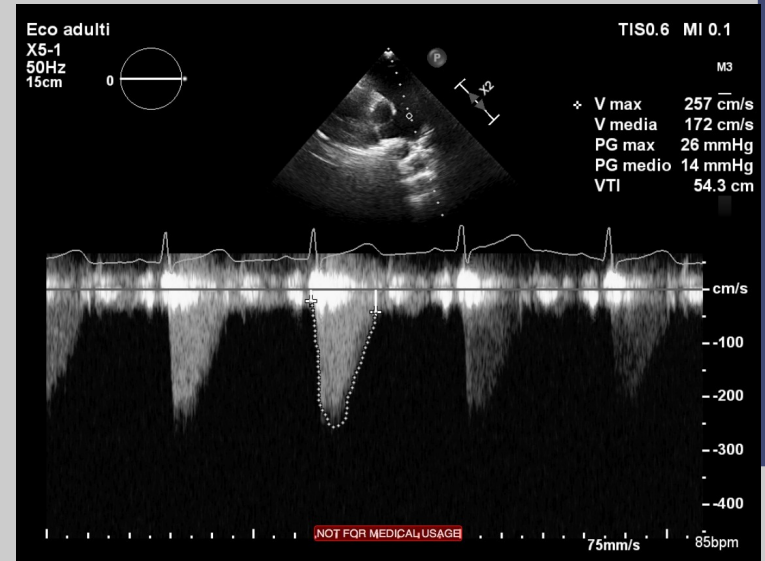
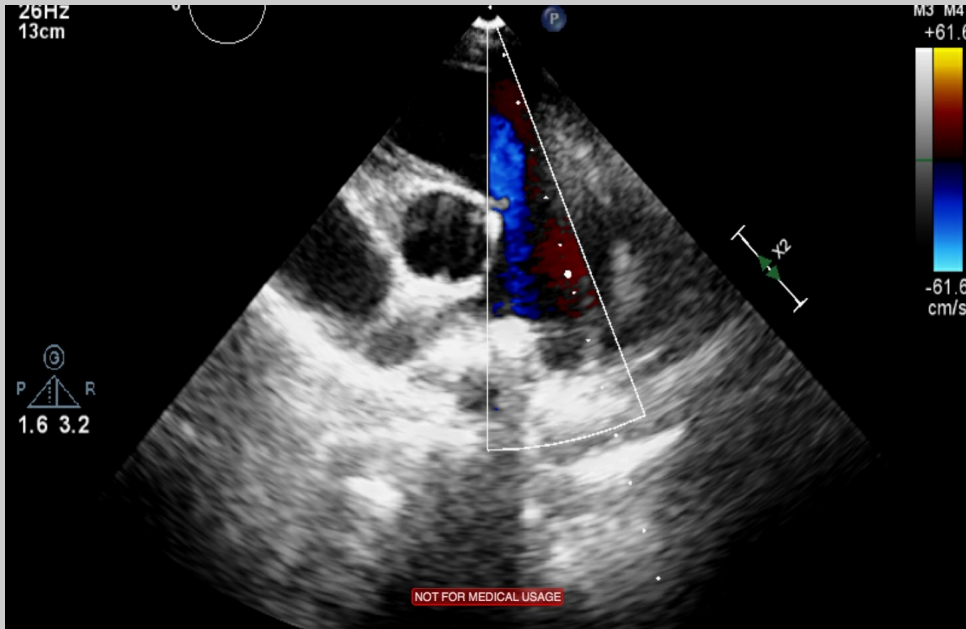
Valutazione post trattamento

- Dimensioni e contrattilità del Vdx
- Gradiente anterogrado residuo
- Insufficienza polmonare
- Rami polmonari



STENOSI POLMONARE

Valutazione post trattamento



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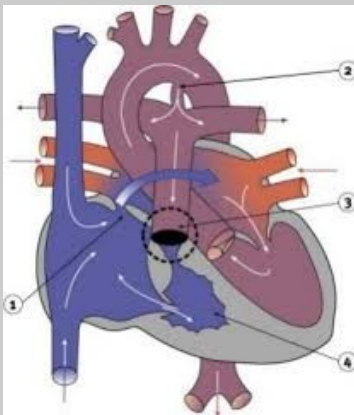
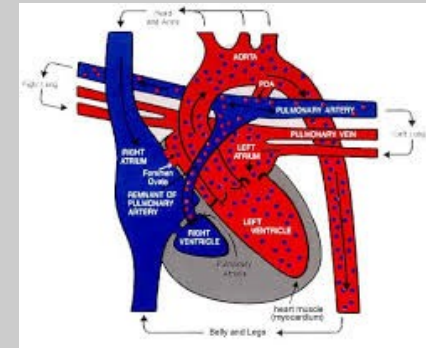
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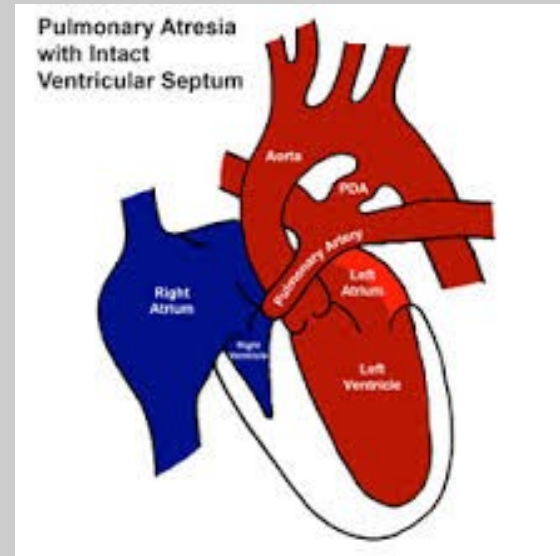
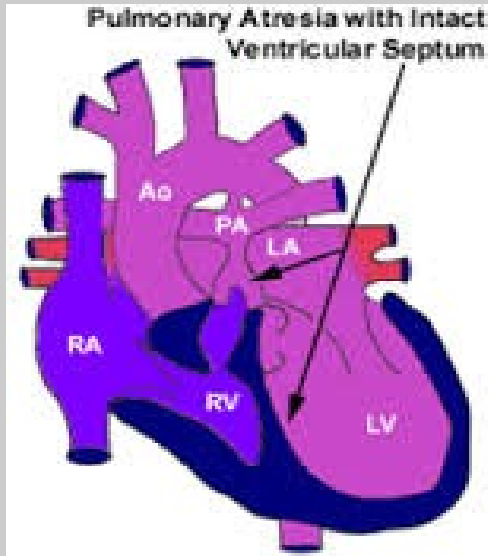
Ospedale del Cuore -FTGM-Massa

ATRESIA POLMONARE A SETTO INTATTO

- Malformazione cardiaca con atresia della valvola polmonare, setto interventricolare intatto e vario grado di ipoplasia del Vdx e della tricuspide
- Circa 2% delle cardiopatie congenite
- M=F
- Cardiopatia critica dotto dipendente
- Importanza di una corretta diagnosi prenatale e neonatale



ATRESIA POLMONARE A SETTO INTATTO



- Notevole variabilità dell'anatomia in base a:
 - Dimensioni e anatomia del Vdx
 - Dimensioni e morfologia valvola Tricuspide
 - Sinusoidi coronarici con circolazione Vx dipendente
 - Generalmente arteria e rami polmonari di buon calibro perchè perfusi dal dotto

ATRESIA POLMONARE A SETTO INTATTO

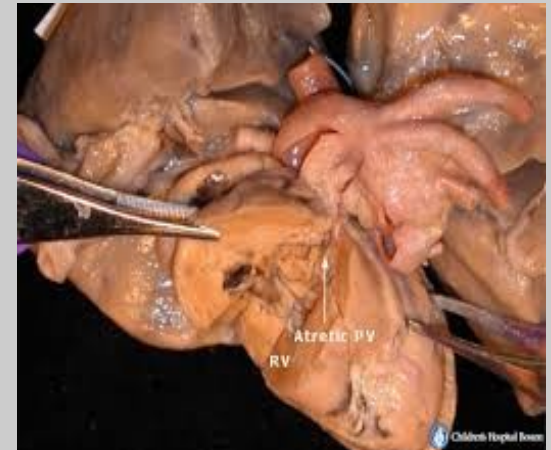
Ventricolo Destro

Valutare dimensioni e la struttura anatomica del Vdx:

- Tripartito: ha tutte e tre le sue componenti (inlet, pars trabecolata, outlet)
- Bipartito: inlet ed outlet
- Unipartito: solo inlet

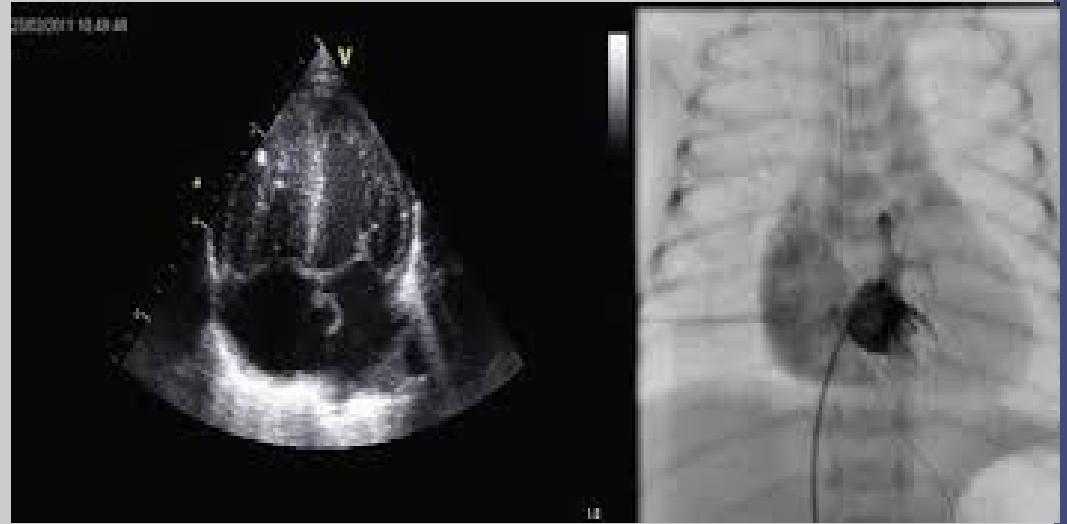
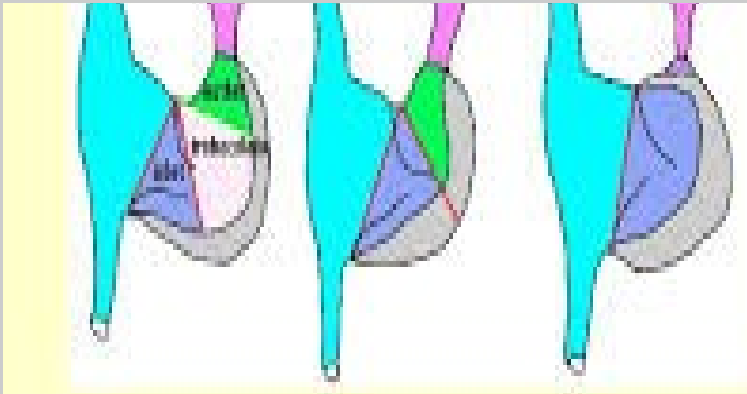
Valutare dimensioni e morfologia valvola tricuspide:

- Utile *Z value* della tricuspide (stretta correlazione tra le dimensioni della TV e le dimensioni del Vdx)
- Spesso displasica
- Insufficienza valvolare in relazione anche alla pressione in Vdx

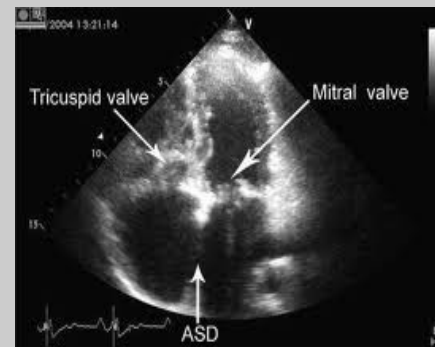


ATRESIA POLMONARE A SETTO INTATTO

Ventricolo Destro

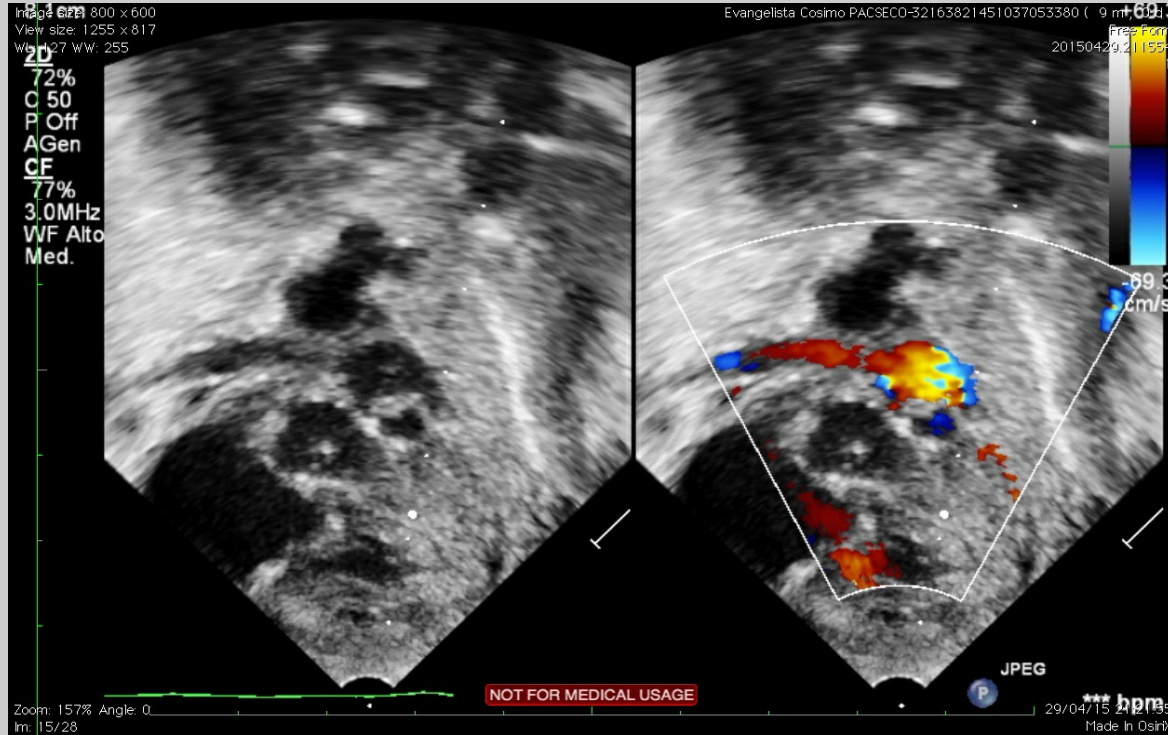
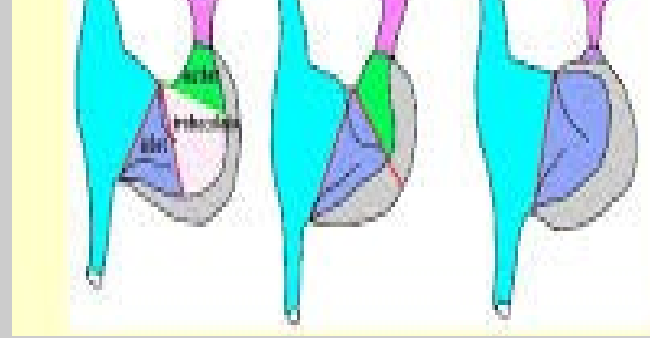


- TV Z score < -4 : correzione monoventricolare
- TV Z score $> -4 < -2$ correzione ad 1,5 ventricolo
- TV Z Score > -2 correzione biventricolare



ATRESIA POLMONARE A SETTO INTATTO

Ventricolo Destro



ATRESIA POLMONARE A SETTO INTATTO

Circolazione Coronarica

Connessione circolo coronarico-ventricolo destro

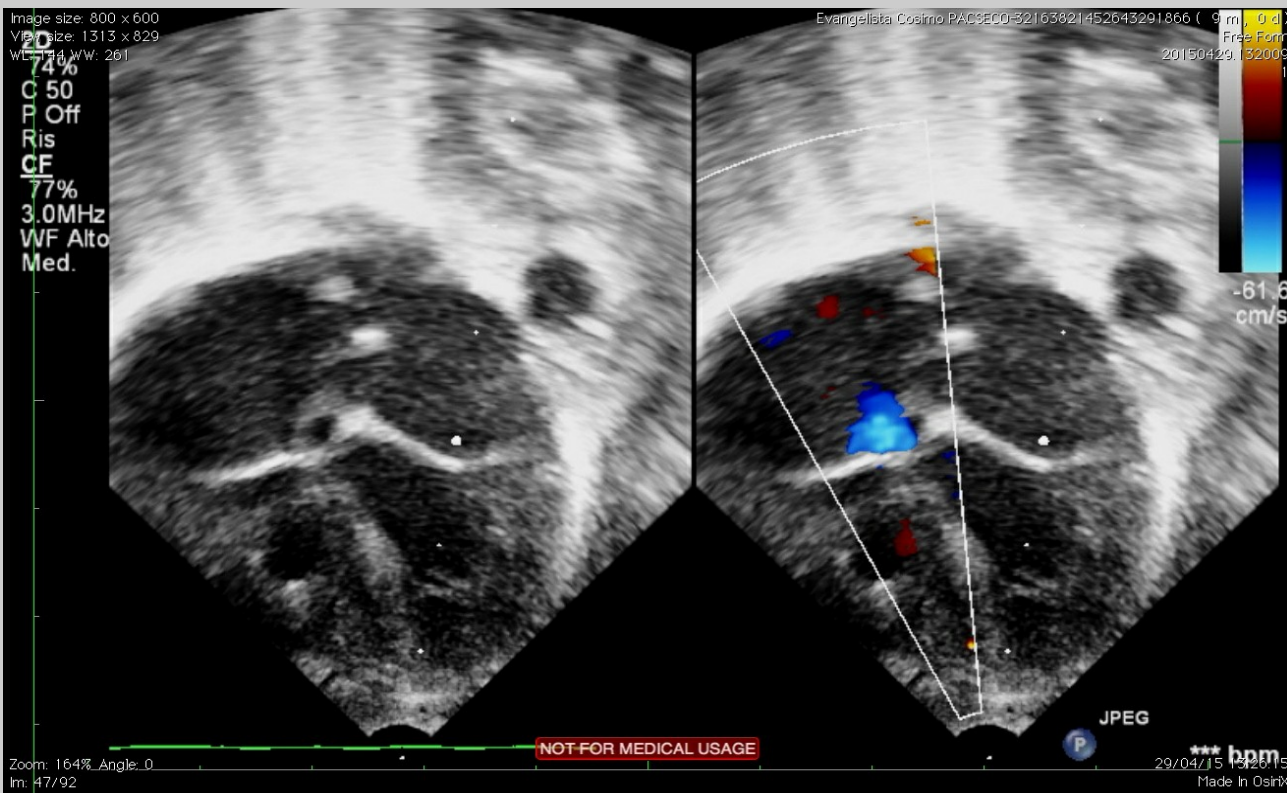
- Assente connessione circolo coronarico-ventricolo dx
- Solo fistole coronariche
- Fistole coronariche più stenosi su singola arteria coronarica
- Fistole più stenosi su multiple arterie coronariche
- Circolazione coronarica Vdx dipendente: in alcune zone del miocardio la perfusione coronarica dipende dal flusso retrogrado dal vdx durante la sistole



RVDCC: correzione monoventricolare

ATRESIA POLMONARE A SETTO INTATTO

Circolazione Coronarica

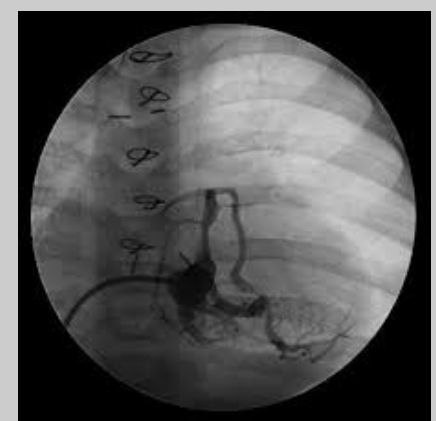
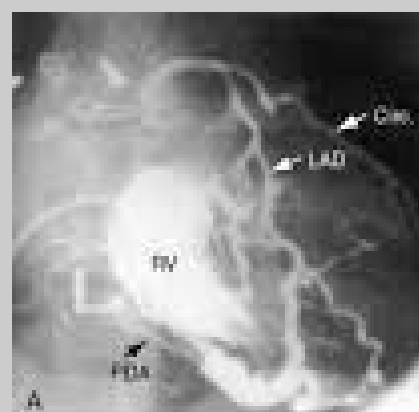
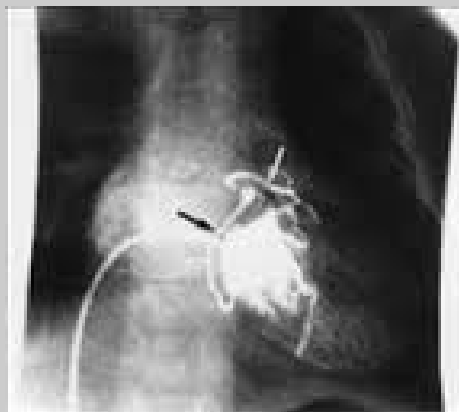


ATRESIA POLMONARE A SETTO INTATTO

Circolazione Coronarica

Circolazione Coronarica Vdx Dipendente

Fistole tra Vdx e coronarie associate ad ostruzioni al flusso coronarico. Alcune zone del miocardio hanno una circolazione coronarica dipendente non dalla pressione diastolica aortica, ma dal flusso coronarico retrogrado dal vdx durante la sistole. Rischio di infarto o morte alla decompressione del vdx



ATRESIA POLMONARE A SETTO INTATTO

Trattamento

- Valvulotomia e valvuloplastica percutanea
- Valvulotomia chirurgica
(con o senza shunt o stent nel dotto di Botallo)
- Solo Shunt se CCVDD o severa ipoplasia Vdx



- Correzione monoventricolare (Glenn e Fontan)
- Correzione ad 1,5 ventricolo (Glenn e flusso anterogrado)
- Correzione biventricolare (correzione anatomica)

ATRESIA POLMONARE A SETTO INTATTO

Trattamento

Pulmonary atresia with intact ventricular septum percutaneous radiofrequency-assisted valvotomy and balloon dilation versus surgical valvotomy and blalock taussig shunt

JACC 2000;35

Pulmonary atresia with intact ventricular septum: Strategy based on right ventricular morphology

J Thor and Cardio surgery 2003

Pulmonary atresia with intact ventricular septum: initial management

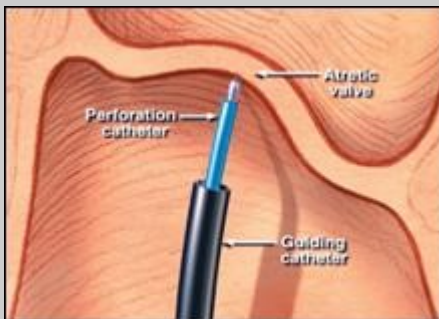
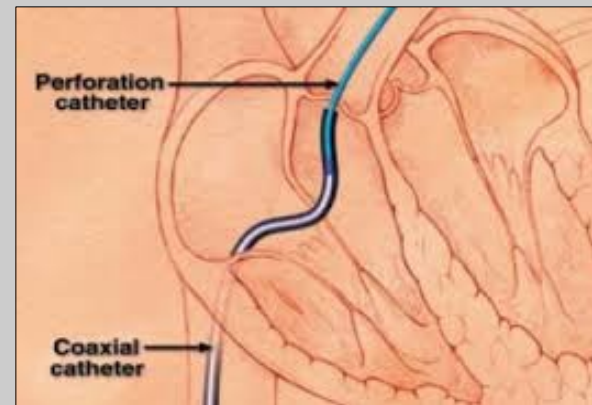
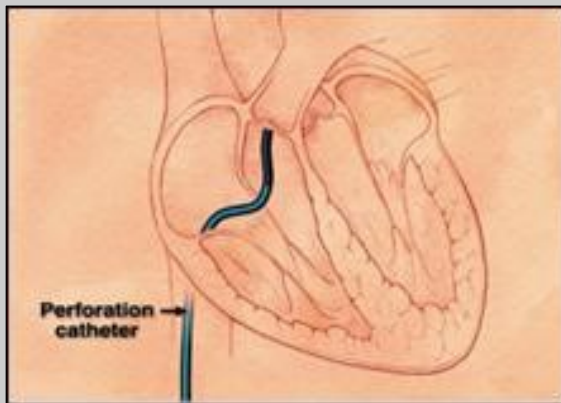
Ann Thor Surg 2006

ATRESIA POLMONARE A SETTO INTATTO

Trattamento

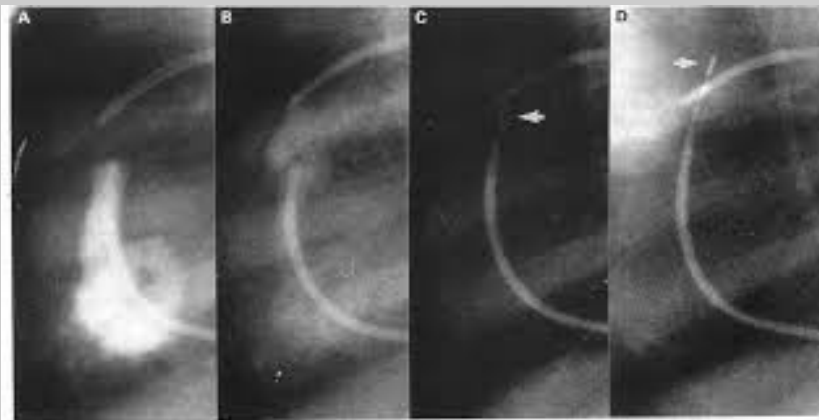
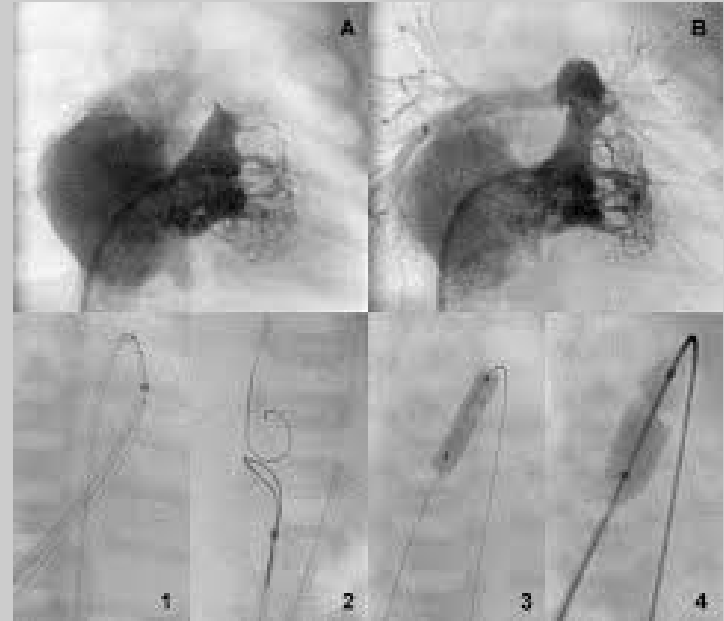
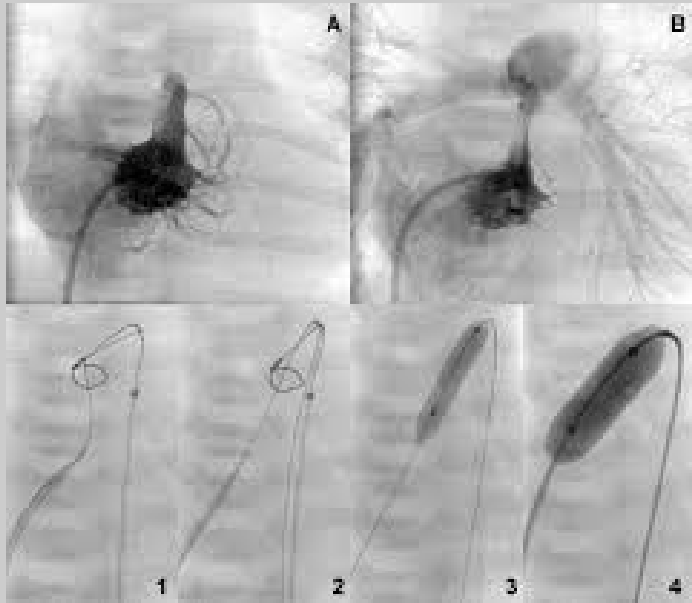
Pulmonary atresia with intact ventricular septum percutaneous radiofrequency-assisted valvotomy and balloon dilation versus surgical valvotomy and blalock taussig shunt

JACC 2000;35



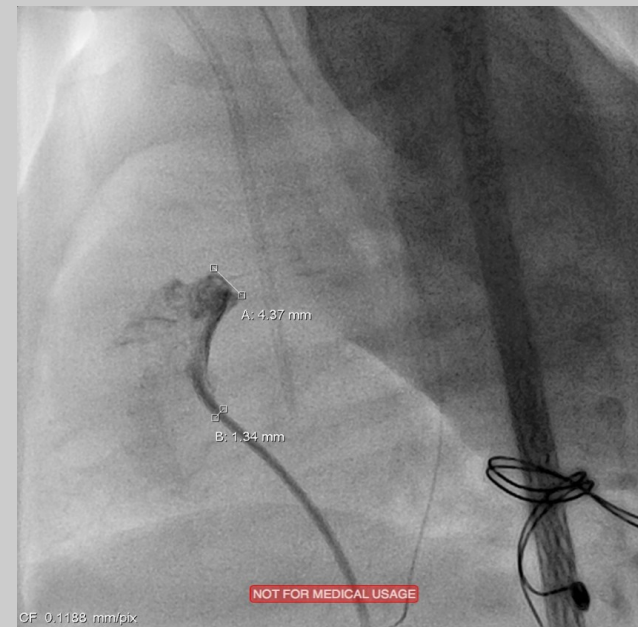
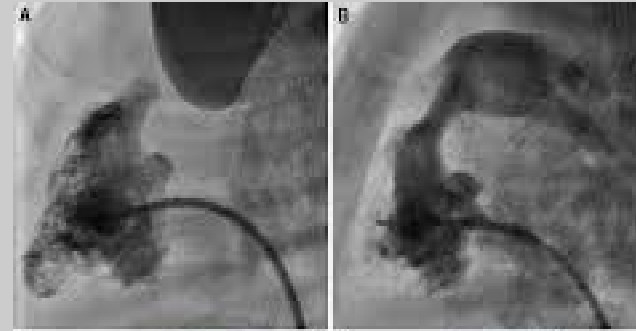
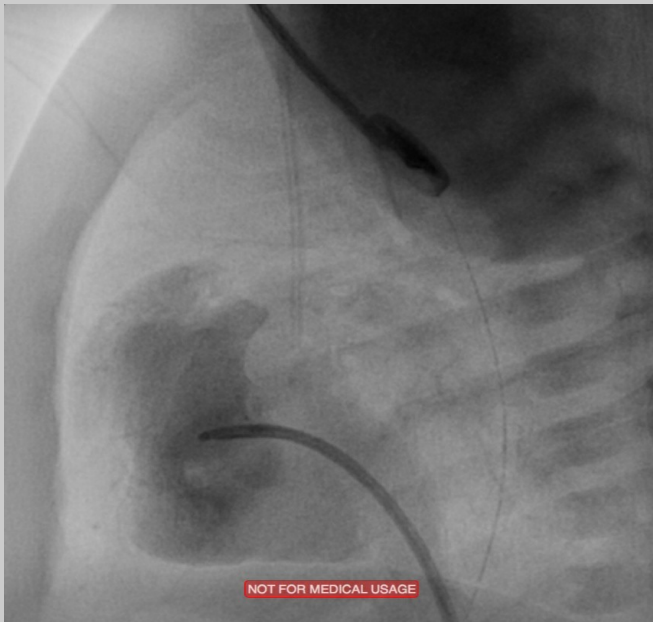
ATRESIA POLMONARE A SETTO INTATTO

Trattamento Percutaneo



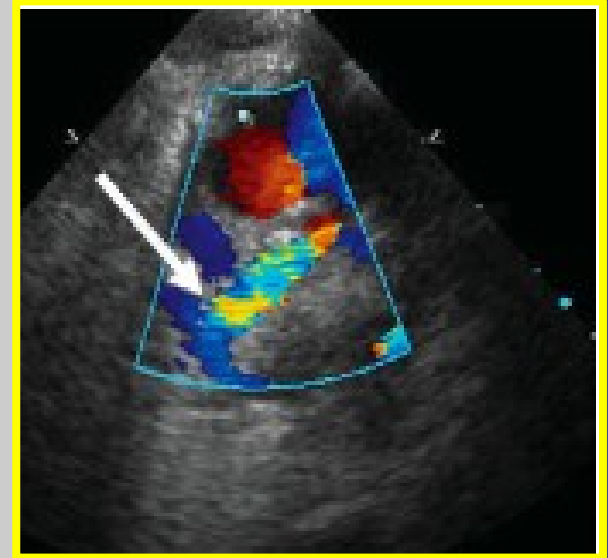
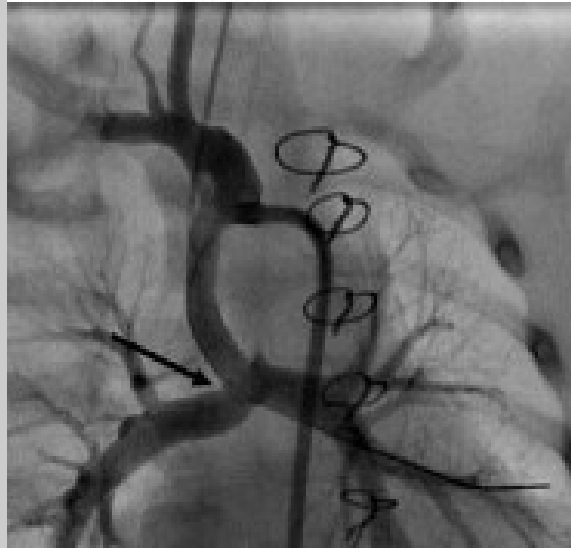
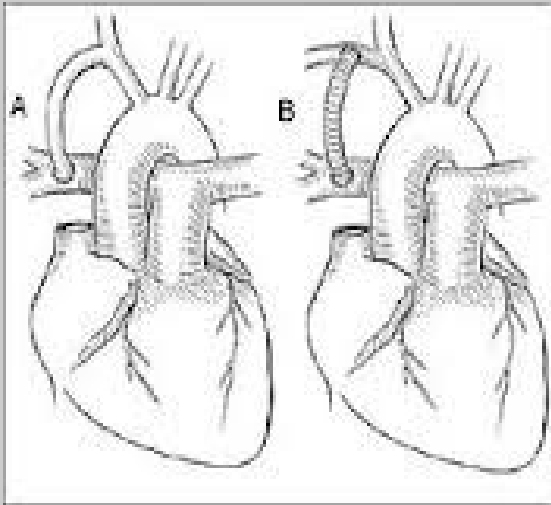
ATRESIA POLMONARE A SETTO INTATTO

Trattamento Percutaneo



ATRESIA POLMONARE A SETTO INTATTO

Trattamento Chirurgico

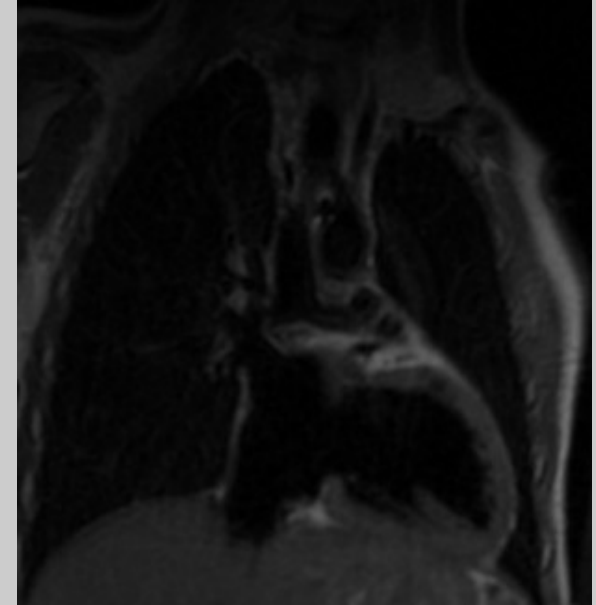
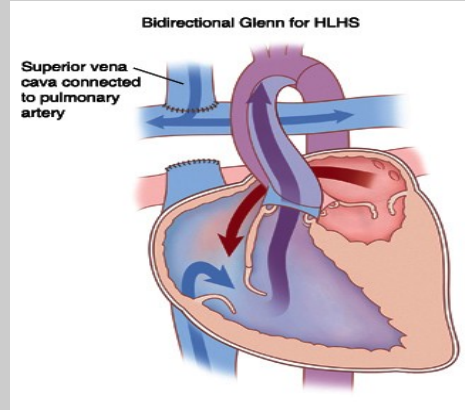
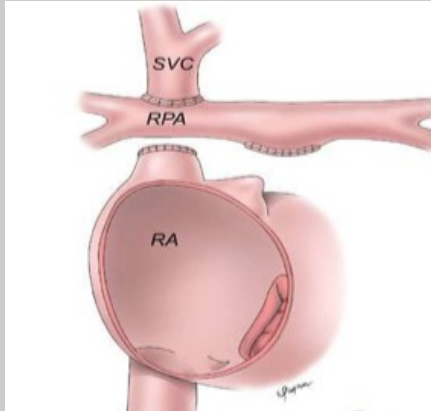


Shunt sistemico polmonare:

- Connessione tra arteria anonima o succlavia e arteria polmonare omolaterale
- Epoca neonatale

ATRESIA POLMONARE A SETTO INTATTO

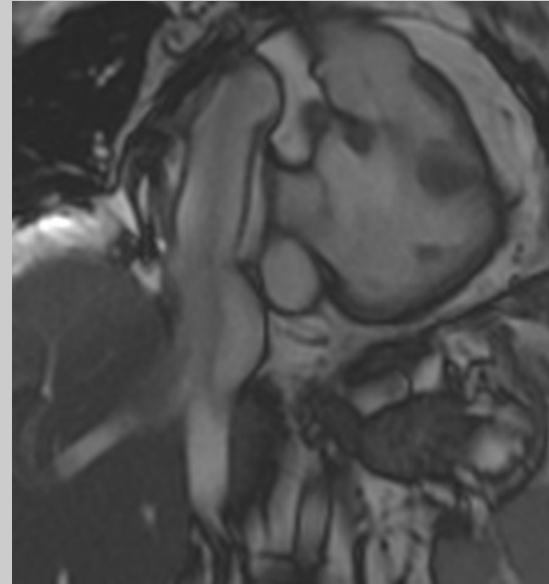
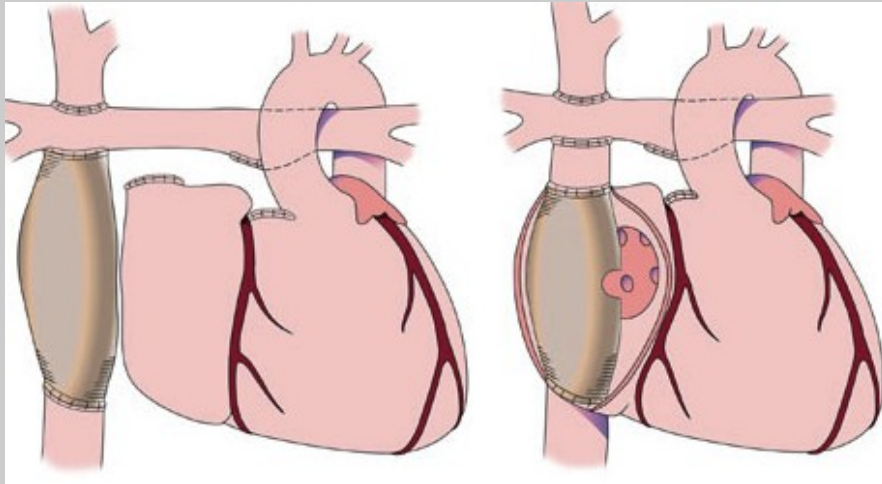
Trattamento Chirurgico: Glenn



- **Intervento di Glenn:** connessione della VCS con l'arteria polmonare omolaterale
- Primo step dell'intervento di Fontan
- Come correzione ad 1,5 ventricolo con flusso anterogrado polmonare

ATRESIA POLMONARE A SETTO INTATTO

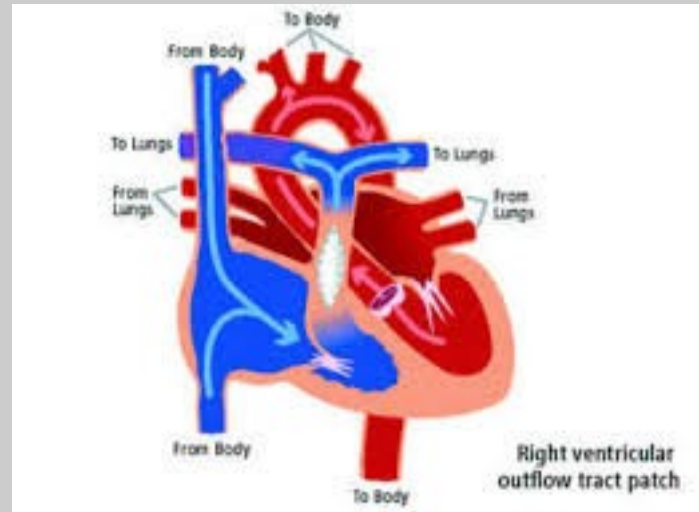
Trattamento Chirurgico: Fontan



- Intervento di Fontan modificata (Total cavopulmonary connection) : VCS e VCI connesse direttamente alla polmonare omolaterale

ATRESIA POLMONARE A SETTO INTATTO

Trattamento Chirurgico: Correzione anatomica



- Ricostruzione dell'efflusso destro con patch o condotto Vdx-AP

ATRESIA POLMONARE A SETTO INTATTO

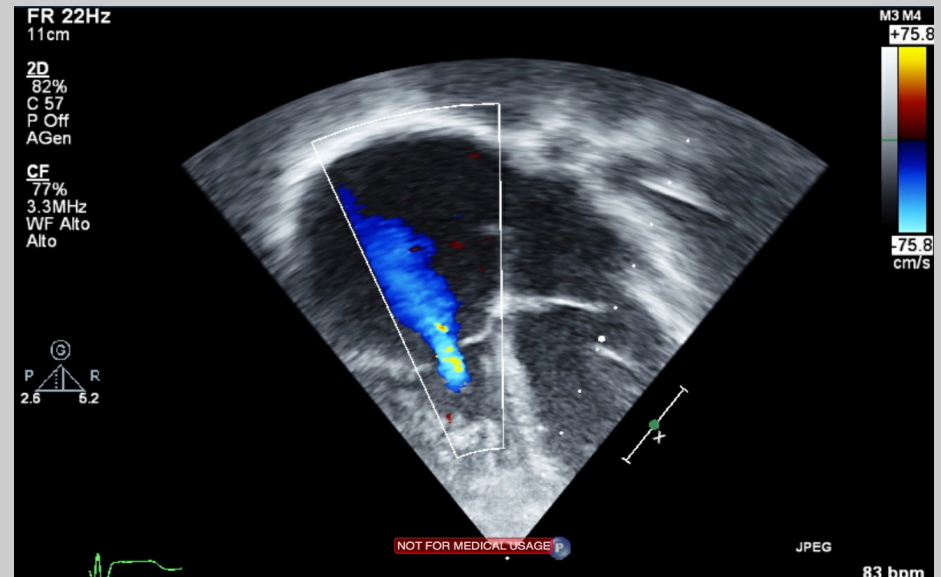
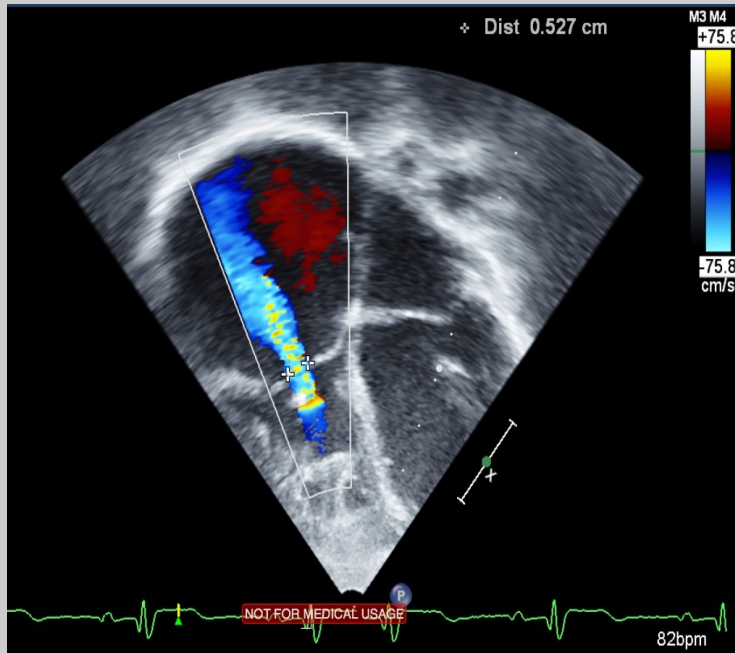
VALUTAZIONE ECO POST PROCEDURA

- **Da cosa si parte:** atresia polmonare, tricuspide displasica, Vdx ipoplasico
- **Cosa è stato fatto:** shunt, Glenn, Fontan, correzione anatomica
- **Cosa cercare:**
 - Valvola tricuspide
 - Ventricolo destro
 - Efflusso destro



ATRESIA POLMONARE A SETTO INTATTO

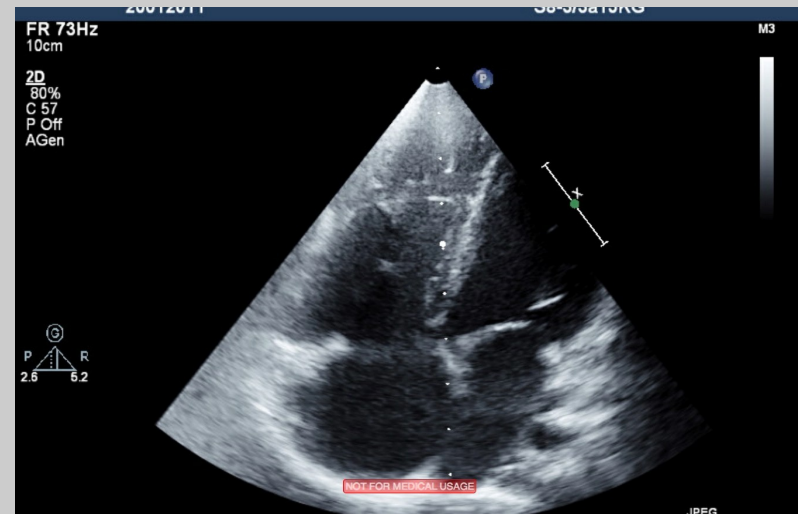
VALUTAZIONE ECO POST PROCEDURA



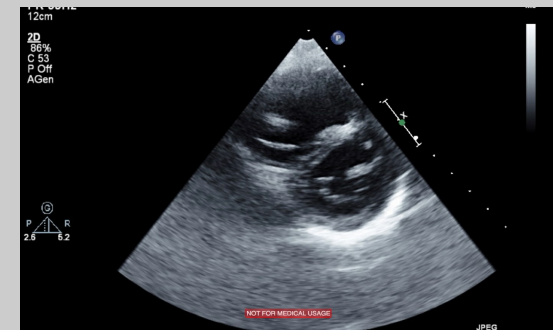
- Valvola tricuspide

ATRESIA POLMONARE A SETTO INTATTO

VALUTAZIONE ECO POST PROCEDURA



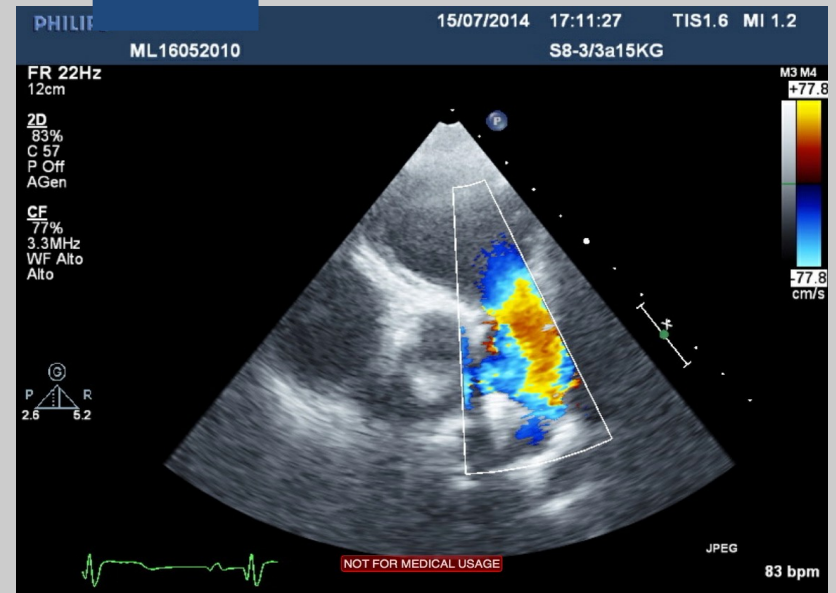
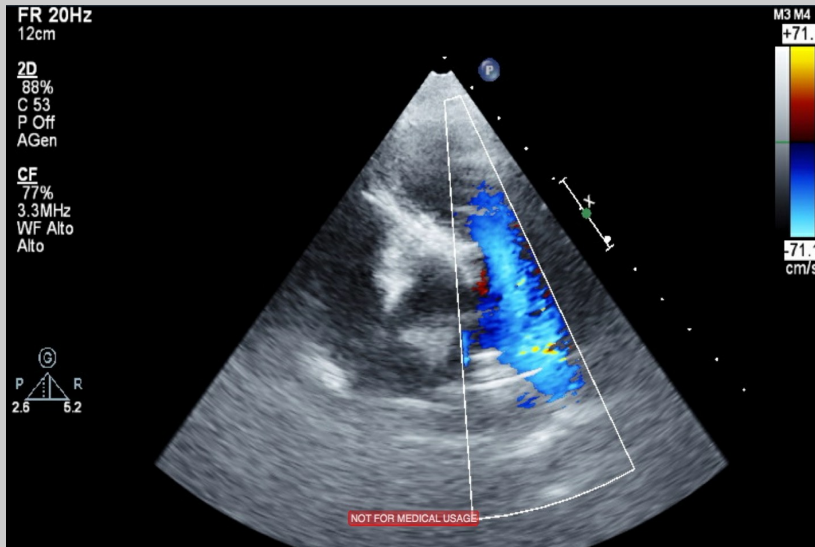
■ **Ventricolo destro**



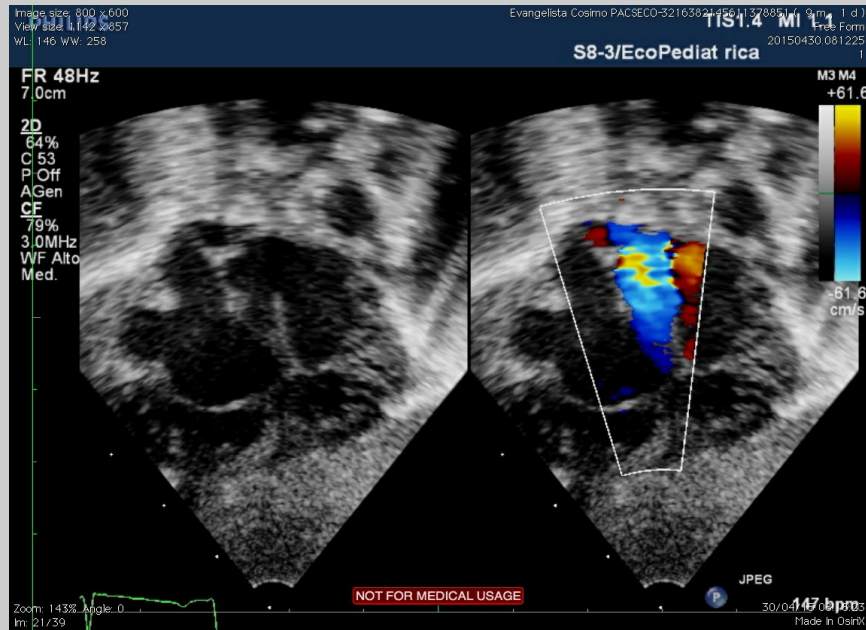
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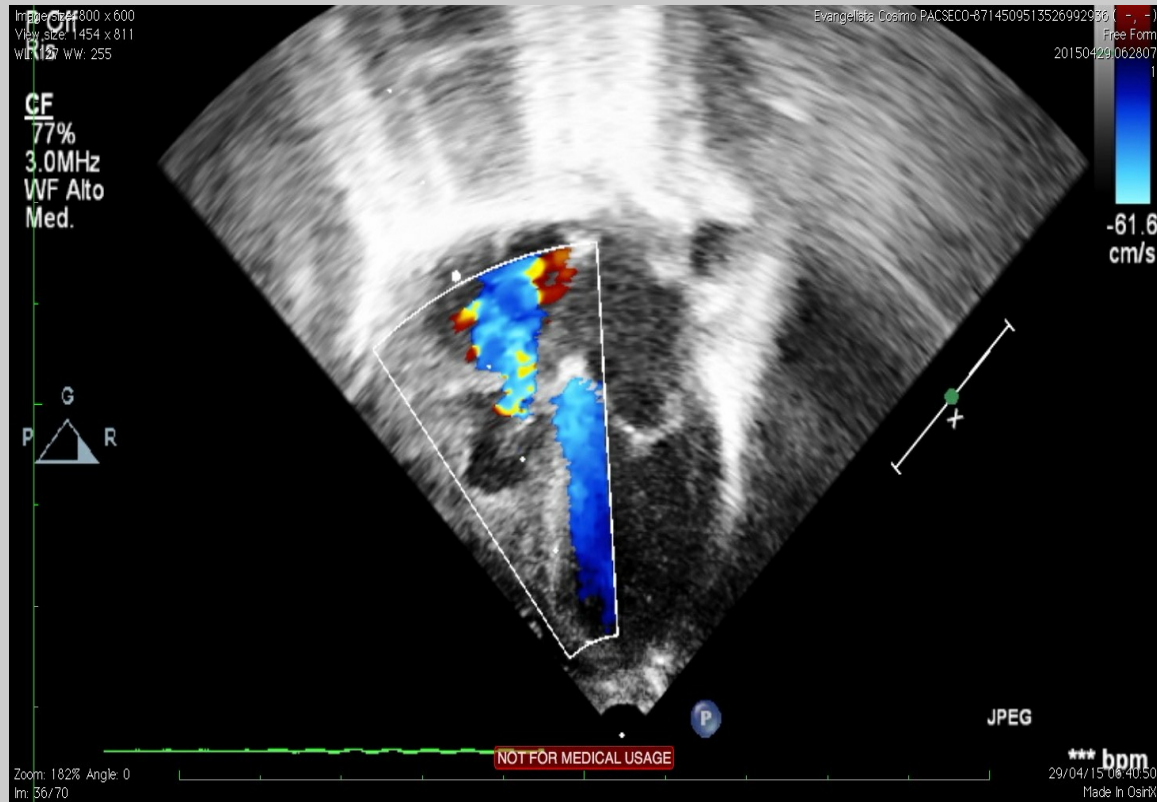
- Efflusso destro



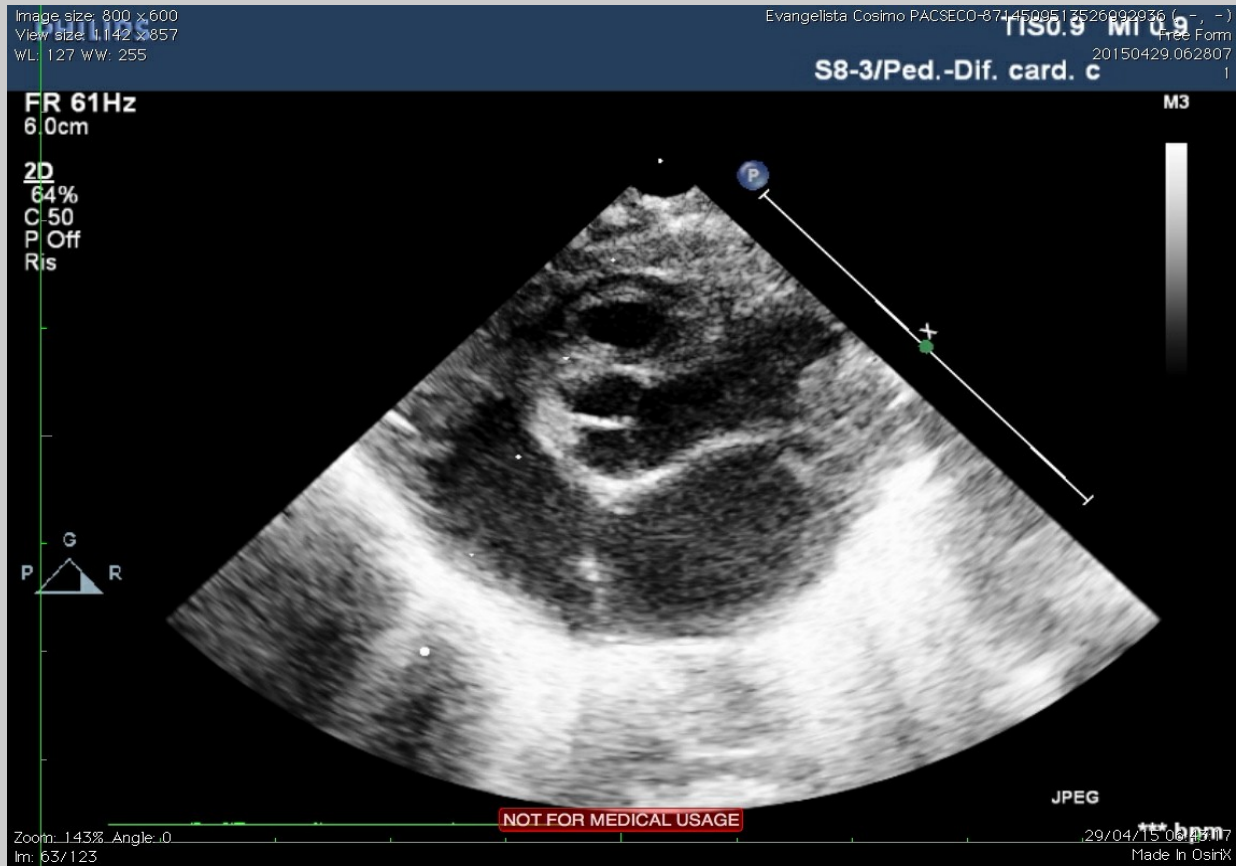
ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA



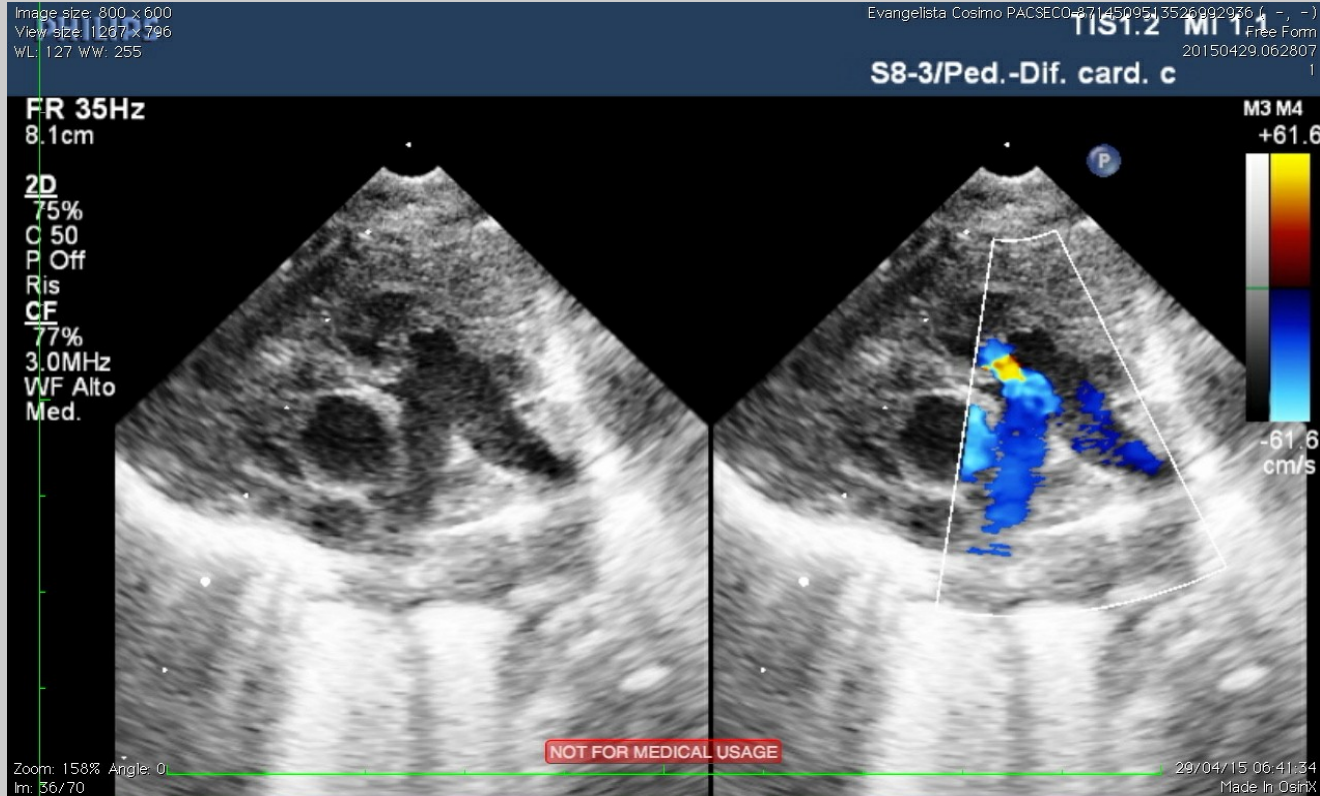
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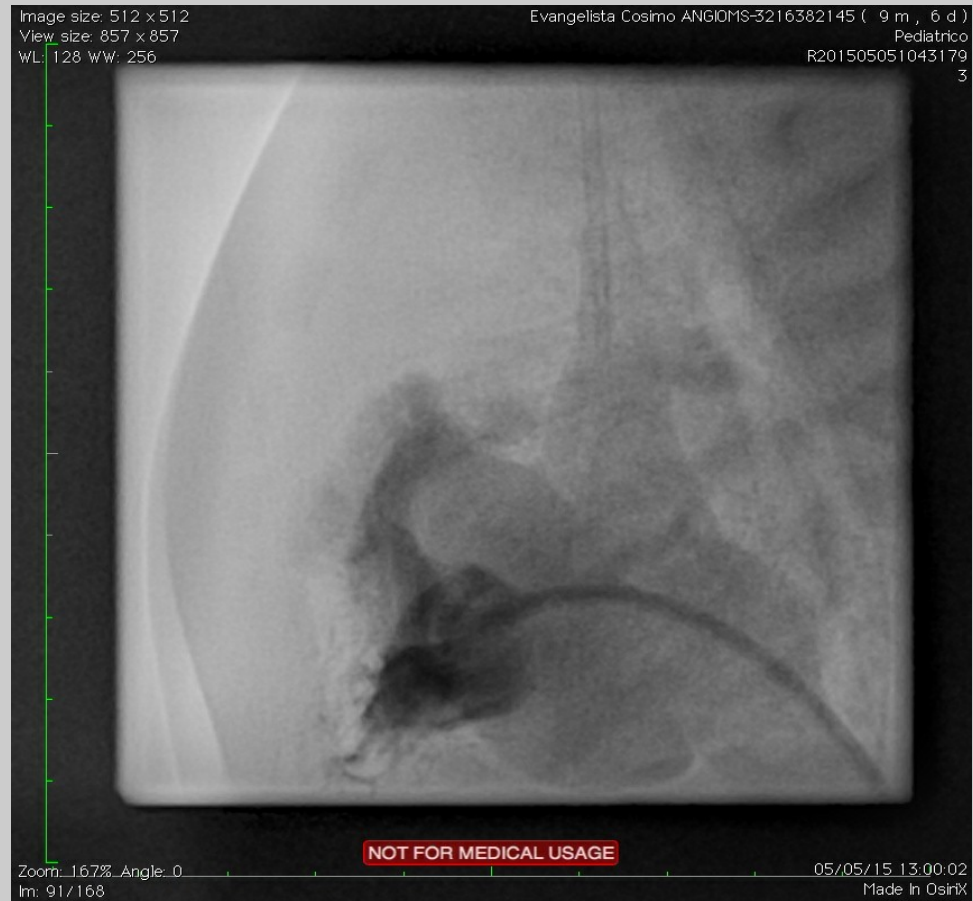
ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA



ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA



ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA



ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA



ATRESIA POLMONARE A SETTO INTATTO/ SP CRITICA

