Percorsi Pediatrici di Val di Noto

IL NEONATO CARDIOPATICO: INTERAZIONE GESTIONALE TRA NEONATOLOGO CARDIOLOGO E PEDIATRA

Maria Pia Calabro'

14 marzo 2015

Cardiopatie Congenite
Incidenza

9 su 1000 nati vivi

- **Circa 1/4 delle Cardiopatie congenite (2.6‰ nati vivi) sono "malformazioni gravi", dotto-dipendenti o comunque tali da richiedere un cateterismo cardiaco e/o un intervento chirurgico entro il primo mese di vita (CCHD = *Critical congenital Heart Disease*)**

- **Tra tutte le malformazioni congenite, le cardiopatie congenite sono la causa più frequente di mortalità neonatale.**

CCHD → Diagnosi tardiva o mancata: 7 su 100.000 n.v.
(Aamir T et Al: Acta paediatr. 2007; 96:1146-1149)

CCHD ⟶ diagnosi tardiva (post-dimissione): 25% dei casi ***(Brown KL: Heart 2006;92: 1298-1302)***

CCHD ⟶ diagnosi tardiva o mancata è la causa di 1-2 decessi su 100.000 nati
(Chang RK: Circulation 2007; 2: 376)

Missed Diagnosis of Critical Congenital Heart Disease

Rucy-Kang R et Al.: Arch pediatr Adolesc Med 2008; 162: 969-974

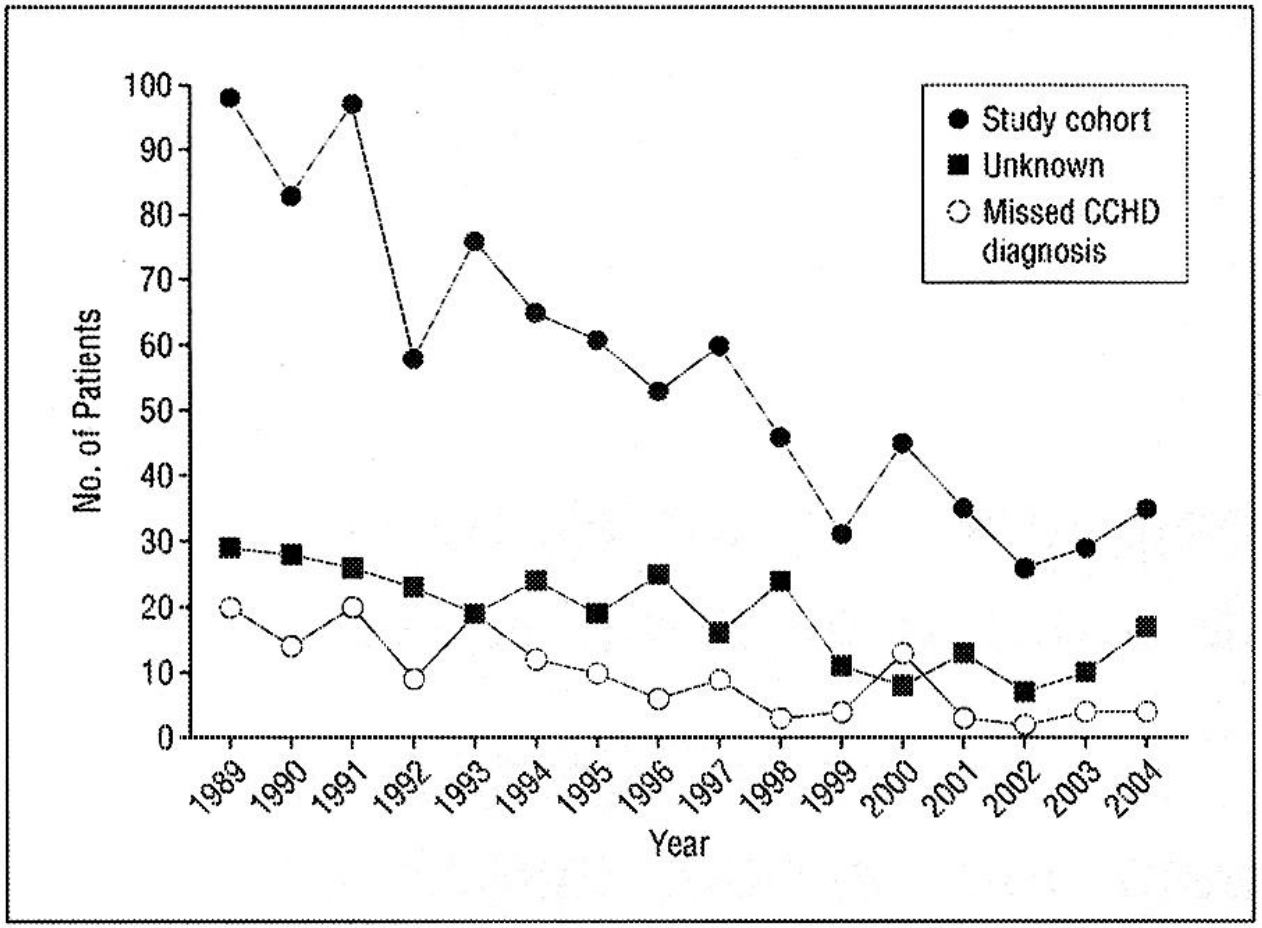


Figure 2. Time trend of the number of patients who died of critical congenital heart disease (CCHD) at 1 to 364 days of age without surgery. Study cohort consists of 898 patients who fulfilled the initial selection criteria.

Diagnosi di CCHD: “Customary Practice”

PRENATAL ULTRASOUND:

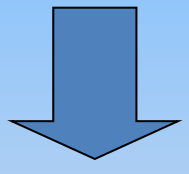
“….numerous studies have reported that even when fetal ultrasound is routinely performed during pregnancy, fewer than 50% of cases of CCHD are identified” !!!

(Mahle et Al.: Circulation 2009; 120: 447-458)

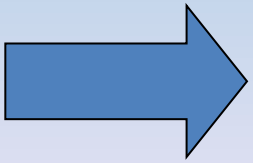
Diagnosi di CCHD: “Customary Practice”

IL NEONATOLOGO E LA VALUTAZIONE DELL’APPARATO CARDIOVASCOLARE NEL NEONATO

NON SI PUO’ PRESCINDERE DA UN ATTENTO ESAME CLINICO CHE INDIRIZZA VERSO L’ESECUZIONE DEGLI ADATTI ESAMI STRUMENTALI



- **ISPEZIONE**
- **PALPAZIONE**
- **ASCOLTAZIONE**



- **SATURAZIONE O2**
- **ECG**
- **RX del torace**
- **ECOCARDIOGRAMMA**

Diagnosi di CCHD: “Customary Practice”

“...Skilled physical examination, a sensitive and specific screening tool in older children, does not always distinguish between neonates with and without congenital heart disease…”

(Mahle et Al.: Circulation 2009; 120: 447-458)

Il “soffio cardiaco” nei primi giorni di vita *(0.6-4.2% dei neonati*) non è sempre indicativo di cardiopatia congenita

Soffio sistolico eiettivo basale del neonato (fisiologica stenosi relativa dei rami polmonari rispetto al tronco, con inserzione ad angolo acuto)

Soffio sistolico da insufficienza tricuspidalica (ipertensione polmonare fisiologica nelle prime ore di vita).

Soffio sistolico apicale da insufficienza mitralica (ischemia transitoria del muscolo papillare).

Soffio sistolico da presenza di falso tendine ventricolare sinistro.

Gravi cardiopatie congenite quali la “trasposizione delle grandi arterie a setto integro” possono non manifestarsi con un soffio cardiaco.

La valutazione di un soffio cardiaco non può MAI prescindere da una attenta valutazione clinica completa del neonato.

"Presentation of congenital heart disease in infancy: implications for routine examination"

Wren C. et Al.: Arch Dis Child Fetal Neonatal Ed 1999;80:F49-F53

- **More than half of babies with undiagnosed congenital heart disease which comes to light in infancy are missed by routine neonatal examination and more than one third by the 6 week examination**

- **Parents, community midwives, health visitors, general practitioners and paediatricians should recognise that a normal neonatal examination does not guarantee that the baby is normal and certainly does not exclude life threatening cardiovascular malformation**

- **Follow up of babies with murmurs without arranging for an early definitive (echocardiographic) diagnosis is of little value and can be risky**

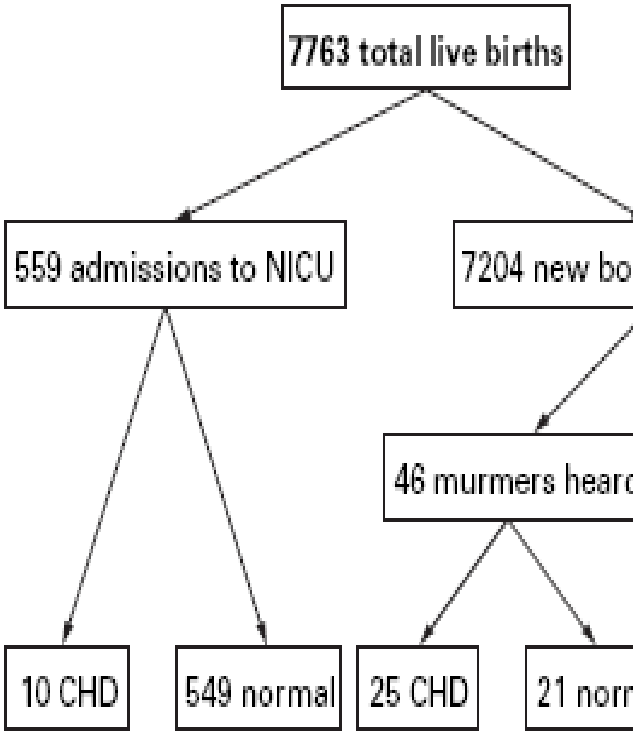
- **Babies with murmurs at neonatal or 6 week examinations should be referred for early paediatric cardiological evaluation. This will result either in a definitive diagnosis of congenital heart disease or in authoritative reassurance of normal cardiac anatomy and function**

- **Babies with Down's syndrome have a high prevalence of congenital heart disease and all should be referred for early echocardiographic examination**

Arch Dis Child Fetal Neonatal Ed 1999;80:F43–F45

Prevalence and clinical significance of cardiac murmurs in neonates

Sean B Ainsworth



Key messages

- The prevalence of murmurs detected at routine examination of neonates is less than 1%.
- About half of murmurs are due to an underlying cardiovascular malformation
- Early referral of all newborn babies with murmurs for definitive diagnosis is recommended
- The absence of a murmur does not exclude serious heart disease

Cite this as: *BMJ* 2008;337:a3037
doi:10.1136/bmj.a3037

RESEARCH

Impact of pulse oximetry screening on the detection of duct dependent congenital heart disease: a Swedish prospective screening study in 39 821 newborns

Pulse oximetry screening detects **100%** of infants with pulmonary duct dependent circulation and, when combined with routine clinical examination, **92%** of all infants with duct-dependent circulation.

As inpatient maternity stays have reduced, an increasing proportion of babies with duct dependent pulmonary circulation leave hospital undetected

Pulse oximetry screening performed both preductally and postductally detects 100% of infants with pulmonary duct dependent circulation and, when combined with routine clinical examination, detects 92% of all infants with duct dependent circulation before hospital discharge, and has a higher detection rate than physical examination alone

Introduction of pulse oximetry screening is cost neutral in the immediate perspective, as each additional case that receives a timely diagnosis costs the same as the treatment of a child that is readmitted in circulatory collapse, but there are probably additional long term cost benefits from reduced neurological morbidity

AHA/AAP Scientific Statement

Role of Pulse Oximetry in Examining Newborns for Congenital Heart Disease

A Scientific Statement From the American Heart Association and American Academy of Pediatrics

William T. Mahle, MD, FAHA, FAAP, Chair; Jane W. Newburger, MD, MPH, FAHA, FAAP; G. Paul Matherne, MD, FAHA, FAAP; Frank C. Smith, MD; Tracey R. Hoke, MD, FAAP; Robert Koppel, MD, FAAP; Samuel S. Gidding, MD, FAHA, FAAP; Robert H. Beekman III, MD, FAHA, FAAP; Scott D. Grosse, PhD; on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Interdisciplinary Council on Quality of Care and Outcomes Research; and the American Academy of Pediatrics Section on Cardiology and Cardiac Surgery, and Committee on Fetus and Newborn

Sensitivity for detecting CCHD: 69.6%
False positive screens occurred in only 0.035%

"...Future studies in larger populations...are needed to determine wheter this practice should become standard of Care in the routine assessment of the neonate..."

were used. In an analysis of pooled studies of oximetry assessment performed after 24 hours of life, the estimated sensitivity for detecting CCHD was 69.6%, and the positive predictive value was 47.0%; however, sensitivity varied dramatically among studies from 0% to 100%. False-positive screens that required further evaluation occurred in only 0.035% of infants screened after 24 hours.

Conclusions—Currently, CCHD is not detected in some newborns until after their hospital discharge, which results in newborns after 24 hours of life, but before hospital discharge, may detect CCHD. Routine pulse oximetry performed after 24 hours in hospitals that have on-site pediatric cardiovascular services incurs very low cost and risk of harm. Future studies in larger populations and across a broad range of newborn delivery systems are needed to determine whether this practice should become standard of care in the routine assessment of the neonate. (***Circulation***. **2009;120:447-458.**)

INDICE DI PERFUSIONE PERIFERICA: UN NUOVO PARAMETRO DI VALUTAZIONE EMODINAMICA NEI NEONATI

- **Indice di Perfusione Periferica (IPP), derivato dal segnale fotoelettrico pletismografico del pulso-ossimetro, è il rapporto tra la componente pulsatile (arteriosa) e non pulsatile (derivata da altri tessuti, quale la componente venosa, ossea, connettiva) della luce che raggiunge il rilevatore del dispositivo.**

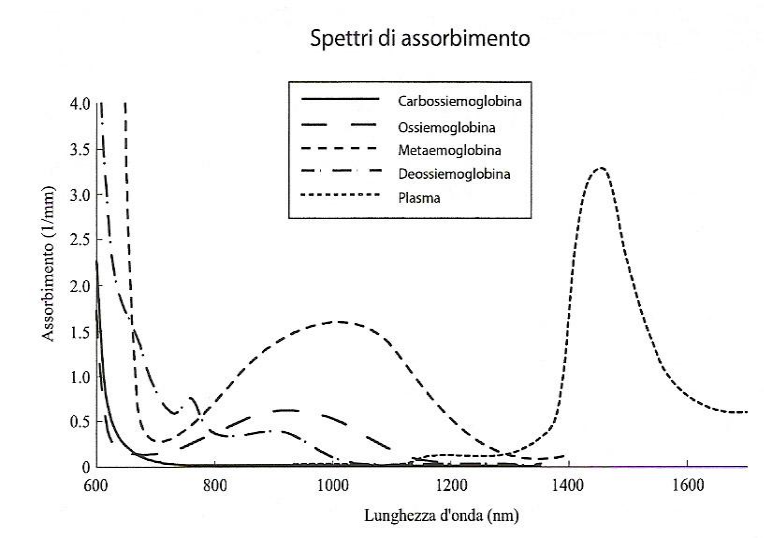
• l'IPP riflette istantaneamente le variazioni del flusso sanguigno periferico, anche nel neonato in condizioni critiche.

REVIEW

Noninvasive monitoring of peripheral perfusion

Alexandre Lima
Jan Bakker

Intensive Care Med (2005) 31:1316–1326
DOI 10.1007/s00134-005-2790-2



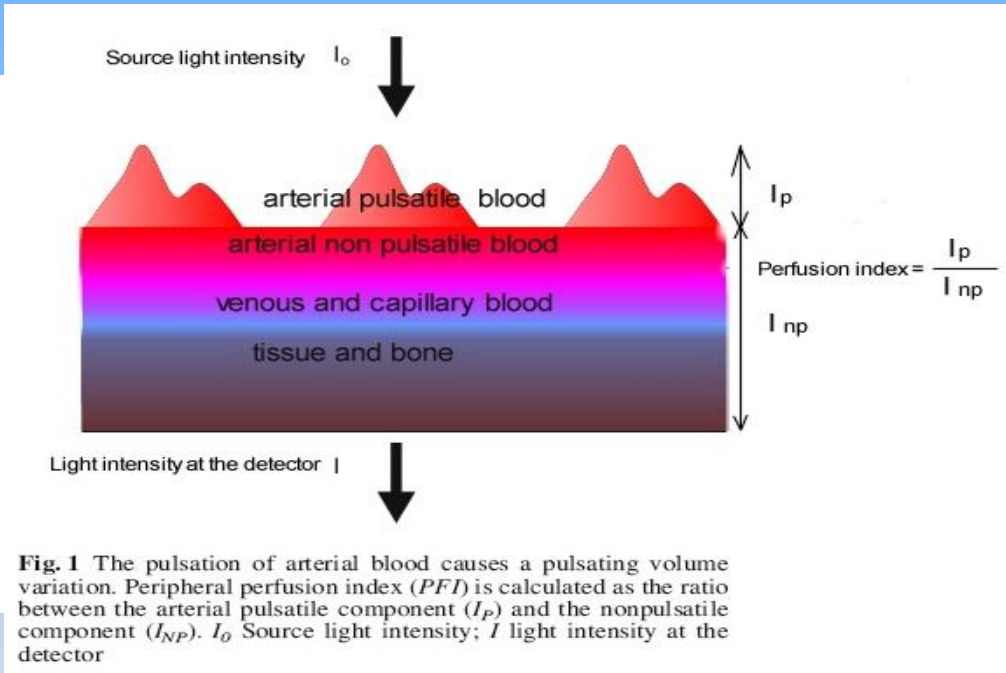
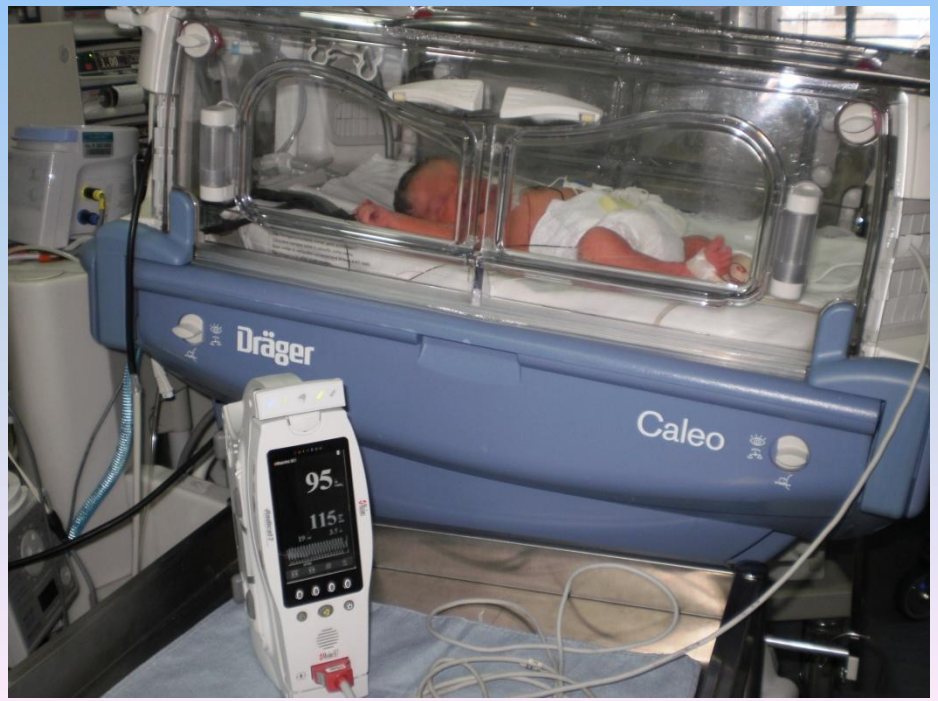


Fig. 1 The pulsation of arterial blood causes a pulsating volume variation. Peripheral perfusion index (*PFI*) is calculated as the ratio between the arterial pulsatile component (I_P) and the nonpulsatile component (I_{NP}). I_0 Source light intensity; I light intensity at the detector

Durante il ciclo cardiaco la variazione dell'assorbimento della luce è una funzione della quantità di sangue presente nel tessuto durante la pulsazione

P.P.I.: Peripheral Perfusion Index



Noninvasive peripheral perfusion index as a possible tool for screening for critical left heart obstruction

A de-Wahl Granelli, I Östman-Smith (ingegerd.ostman-smith@pediat.gu.se)
Department of Paediatrics, The Institute of Clinical Sciences, The Sahlgrenska Academy, Gothenburg University, Sweden

Acta Pædiatrica 2007 **96**, pp. 1455–1459

Figure 1 Application sites: right hand (preductally) and either foot (postductally).

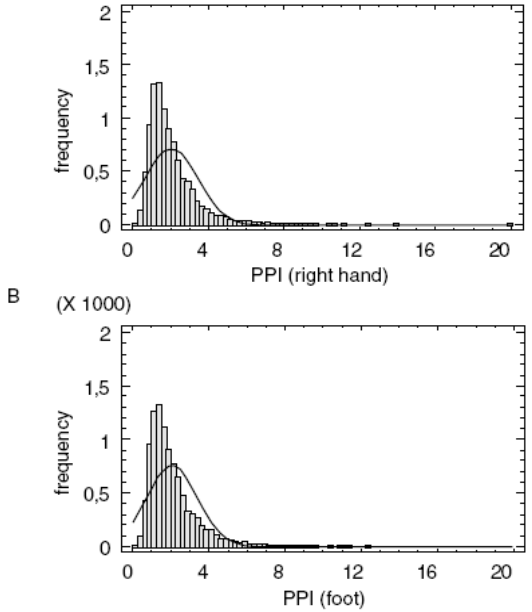


Figure 2 The distribution of PPI right hand (A) and foot (B) values.

CONCLUSIONS

We have established reference values for PPI (right hand and foot) in normal newborns between 1 and 120 h of age. Lower PPI values than 0.70 may indicate illness. Including cut-off values for PPI in pulse-oximetry screening for duct-dependent congenital heart disease is a promising tool for improving the detection of critical congenital heart disease with duct-dependent systemic circulation.

ECOCARDIOGRAMMA

DA ESEGUIRE OGNI VOLTA CHE SI SOSPETTA UNA PATOLOGIA ORGANICA CONGENITA O ACQUISITA DEL CUORE.

PUO' FORNIRE INFORMAZIONI SU:

- **ANATOMIA DEL CUORE E GROSSI VASI**
- **CINETICA E FUNZIONE VENTRICOLARE**
- **PRESENZA DI VERSAMENTO PERICARDICO**
- **PRESENZA DI MASSE (TROMBI O TUMORI CARDIACI)**
- **LOCALIZZAZIONE DI CATETERI INTRAVASALI**
- **VALUTAZIONE DEL FLUSSO NEL DOTTO ARTERIOSO E NEL FORAME OVALE**

Quando eseguire un ecocardiogramma in un neonato:

- **Quando l'anamnesi familiare è positiva per cardiopatie congenite complesse**
- **Nel caso di cianosi persistente nonostante l'esclusione di patologie polmonari**
- **Quando è presente un soffio cardiaco**
- **Quando la saturazione di O2 è costantemente inferiore al 95% dopo le prime 24 ore di vita**
- **In un neonato sindromico o con malformazioni congenite**

RIASSUMENDO…

CARDIOLOGO PEDIATRA

Attenta valutazione clinica neonatologica:

- **ispezione**
- **palpazione dei polsi**
- **ascoltazione del cuore**
- **saturazione O2**
- **PPI**

ECG
Telecuore
Ecocardiogramma (*)

(*) L'ecocardiogramma **NON PUO' ESSERE CONSIDERATO ESAME DI SCREENING**. Il neonatologo può acquisire competenze per eseguire alcune semplici valutazioni cardiologiche all'ecocardiogramma (valutazione delle camere cardiache, della contrattilità e della cinetica globale, del dotto arterioso, dello shunt al livello del forame ovale, della pressione polmonare)

Perché è importante una diagnosi precoce in assenza di diagnosi prenatale

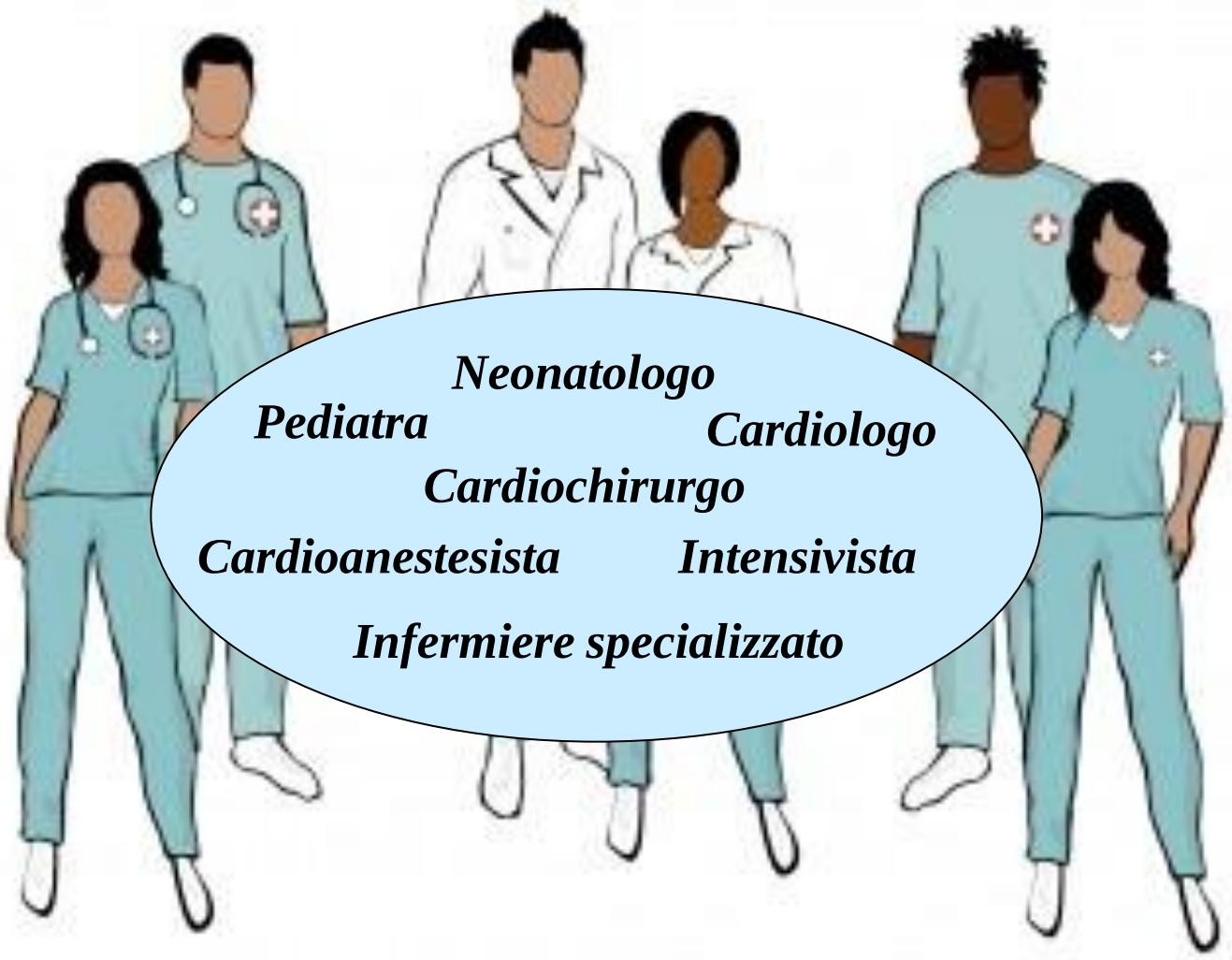
Difficile sospettare CC al momento del parto perché la circolazione transizionale ritarda la comparsa dei sintomi.

Esistono cardiopatie difficili da sospettare/diagnosticare che possono costituire una emergenza → difetti extracardiaci (RVATP , Anomalie arco aortico)

Quando sintomi compaiono tardivamente il neonato può essere già stato dimesso

Nelle CC dotto dipendenti alla comparsa dei sintomi vi è un rapido decadimento delle condizioni generali → alto rischio danno d'organo

TEAM MULTIDISCIPLINARE



CARDIOPATIE CONGENITE "DOTTO-DIPENDENTI"

Flusso polmonare dotto-dipendente

- ✓ Atresia polmonare a setto integro
- ✓ Atresia polmonare con DIV
- ✓ Atresia della tricuspide
- ✓ Tetralogia di Fallot grave
- ✓ Stenosi polmonare critica

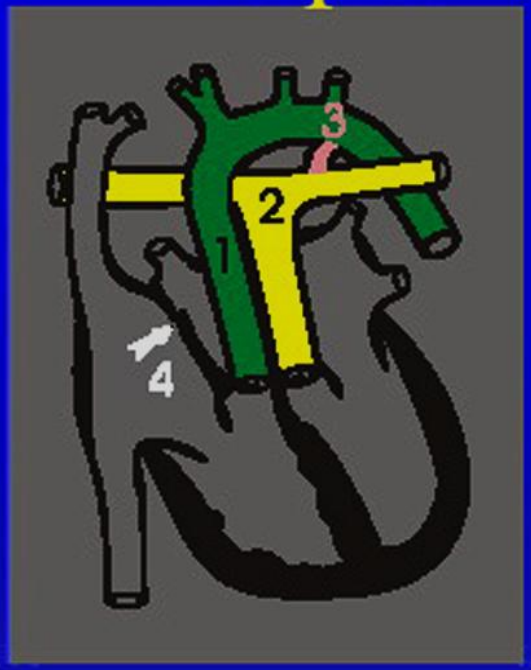
Flusso sistemico dotto-dipendente

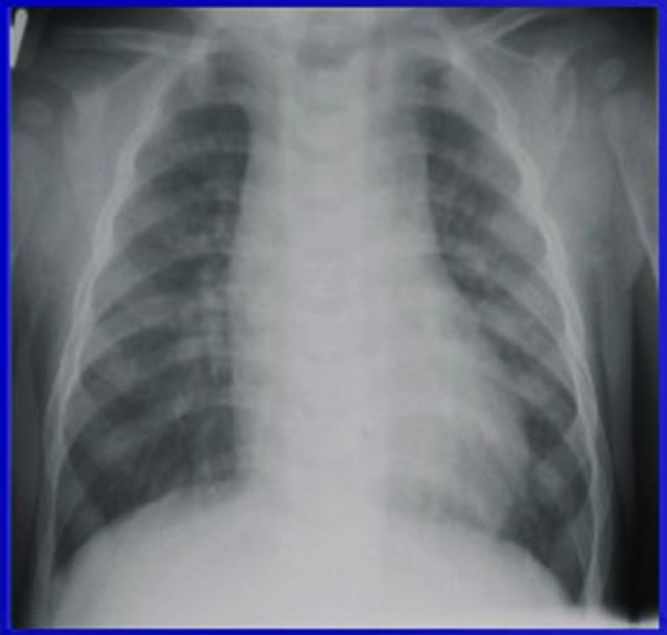
- ✓ Sindrome del cuore sinistro ipoplasico
- ✓ Interruzione dell'arco aortico
- ✓ Coartazione aortica critica
- ✓ Stenosi aortica critica

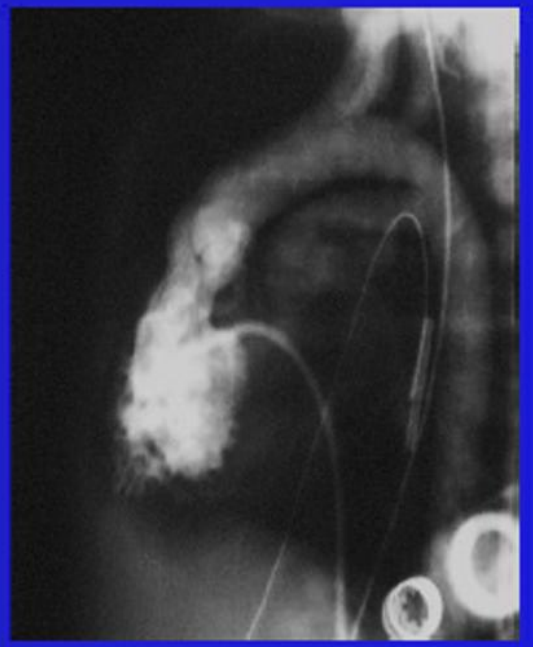
Mixing dotto-dipendente

- ✓ Trasposizione completa delle grandi arterie

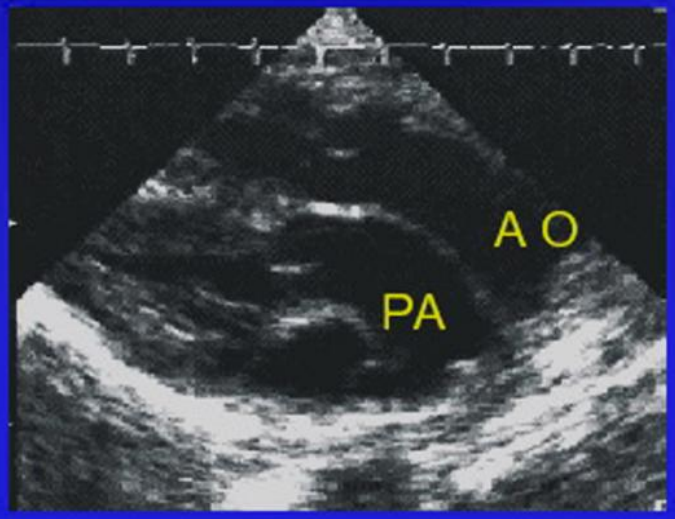
Trasposizione delle grandi arterie

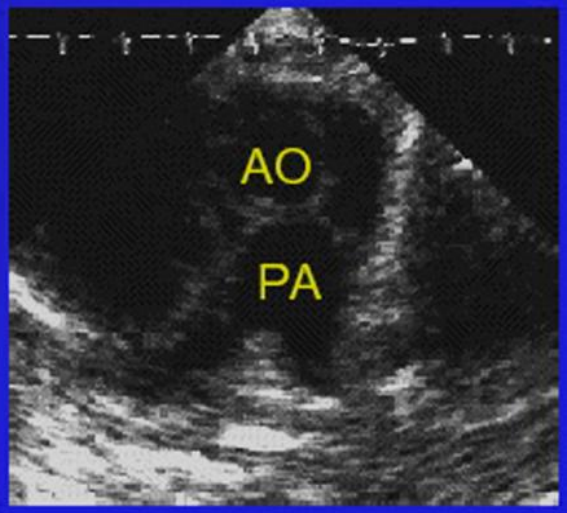






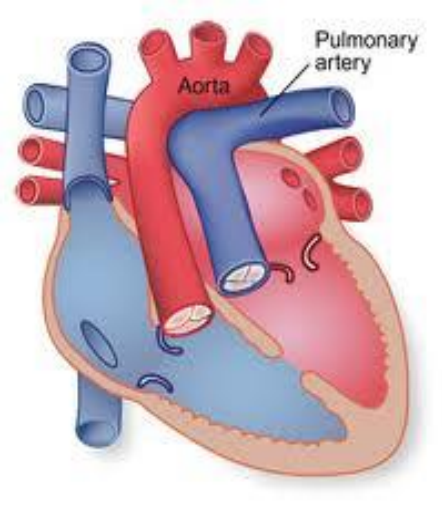
1. Aorta anteriore, dal ventricolo destro
2. Polmonare posteriore, dal ventricolo sinistro.
3. PDA.
4. Forame ovale pervio





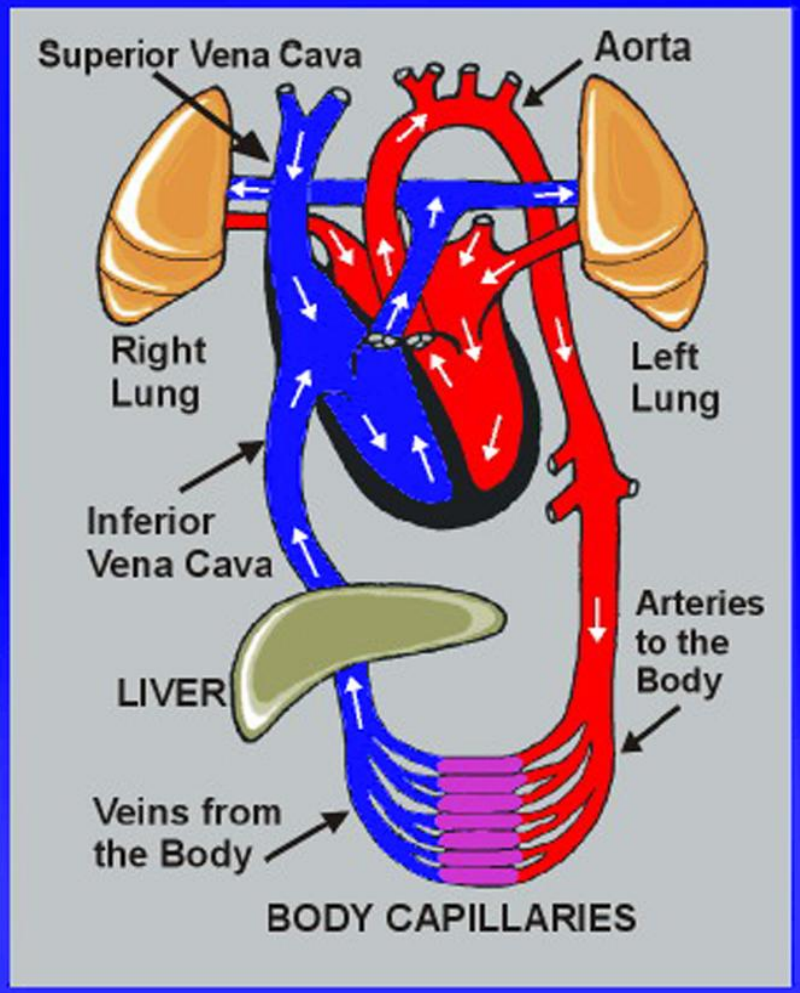
TRASPOSIZIONE DELLE GRANDI ARTERIE
Forme anatomiche

- TGA a SETTO INTEGRO *(EMERGENZA!)*
- TGA + DIV
- TGA + DIV + Stenosi Polmonare
- TGA + DIV + CoAo *(EMERGENZA!)*

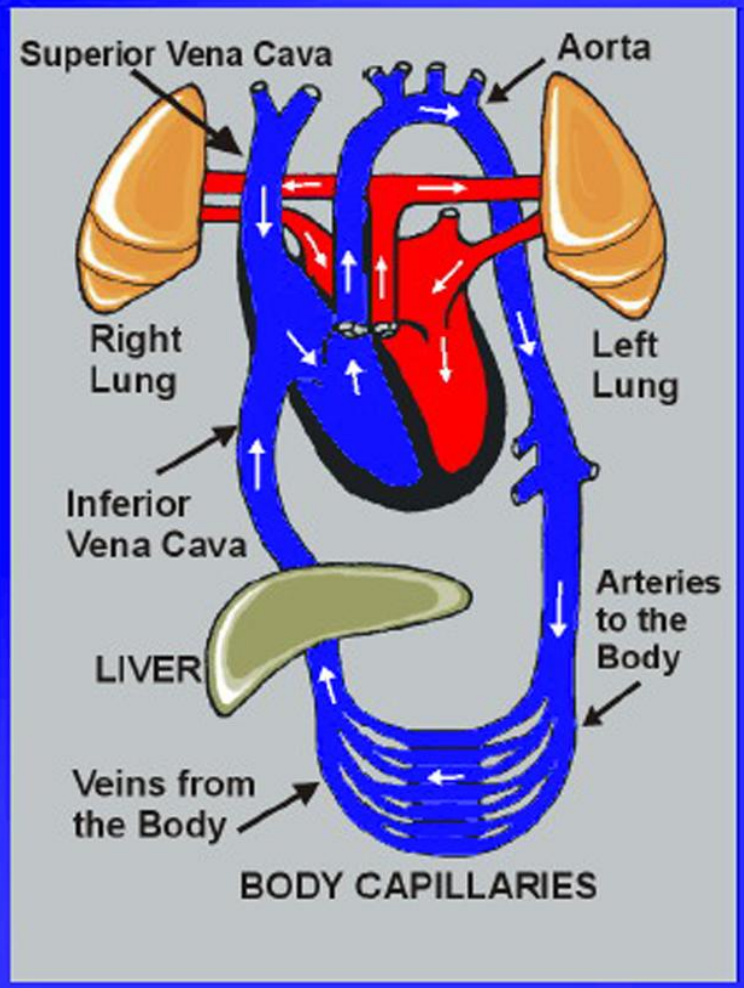


FISIOPATOLOGIA

Trasposizione delle grandi arterie



Circolazione normale
(in serie)

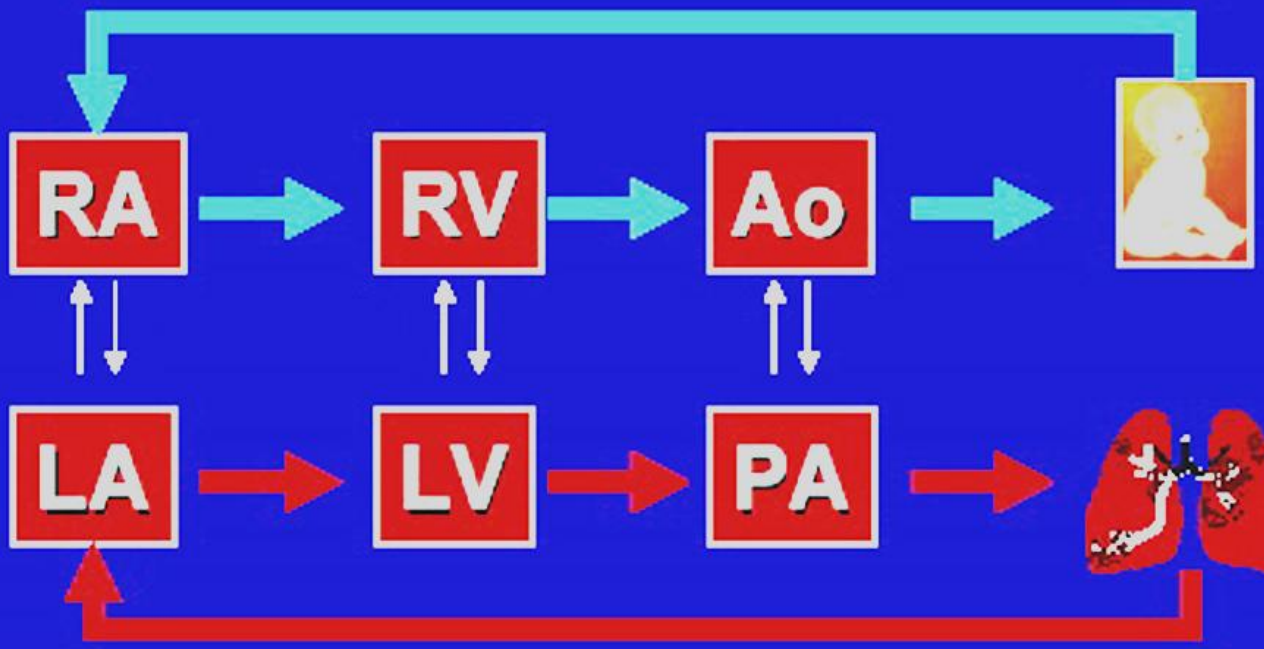


TGA
(Circolazione in parallelo)

TGA: Fisiopatologia

Entità del mixing intercircolatorio in funzione di:

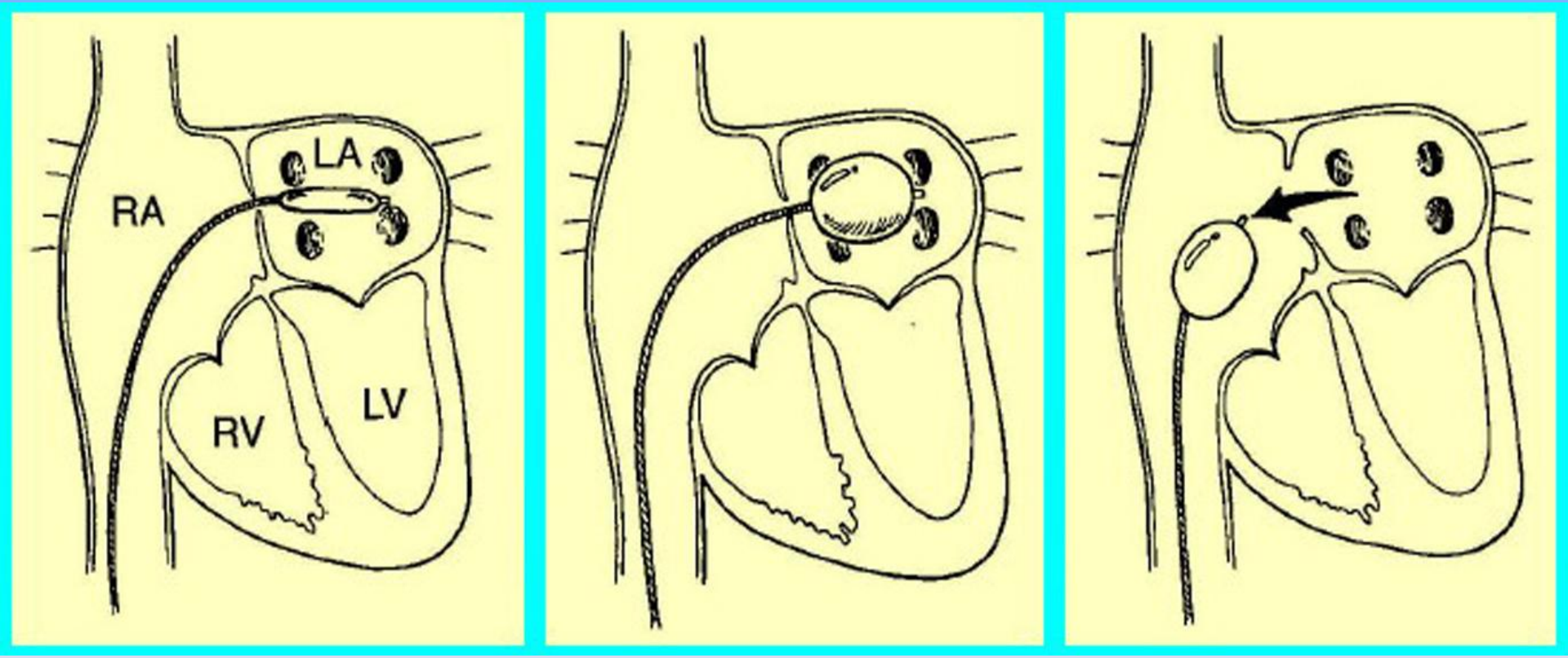
comunicazioni tra i due circuiti



1. posizione
2. calibro
3. numero

TRASPOSIZIONE DELLE GRANDI ARTERIE

ATRIOSETTOSTOMIA (Rashkind)



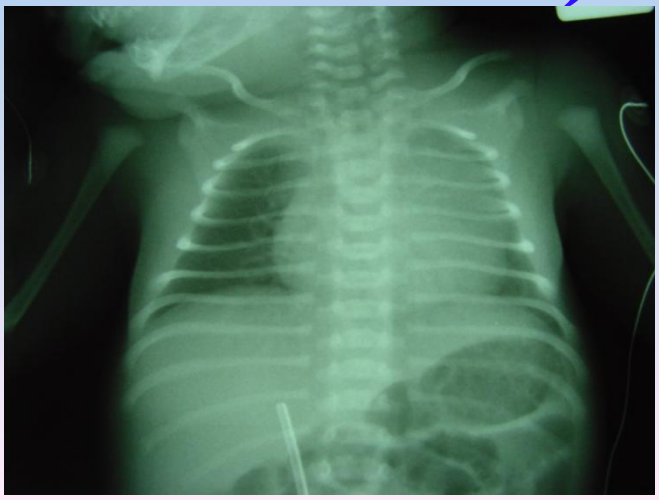
TRASPOSIZIONE DELLE GRANDI ARTERIE

Circoli in parallelo

QUADRO CLINICO

- ***Cianosi*** ⟶
- ***Soffio ?*** ⟶

- ***Ipossia normocapnica (resistente all'O2)***
- ***Rx Torace:*** ⟶



iperafflusso polmonare

TRASPOSIZIONE DELLE GRANDI ARTERIE

MANAGEMENT

- ***CORREZIONE ACIDOSI***
- ***CORREZIONE IPOTERMIA***
- ***PGE1 (0.01-0.05 γ/kg/min)***
- ***SEDAZIONE***
- ***INTUBAZIONE***
- ***TRASFERIMENTO IMMEDIATO***

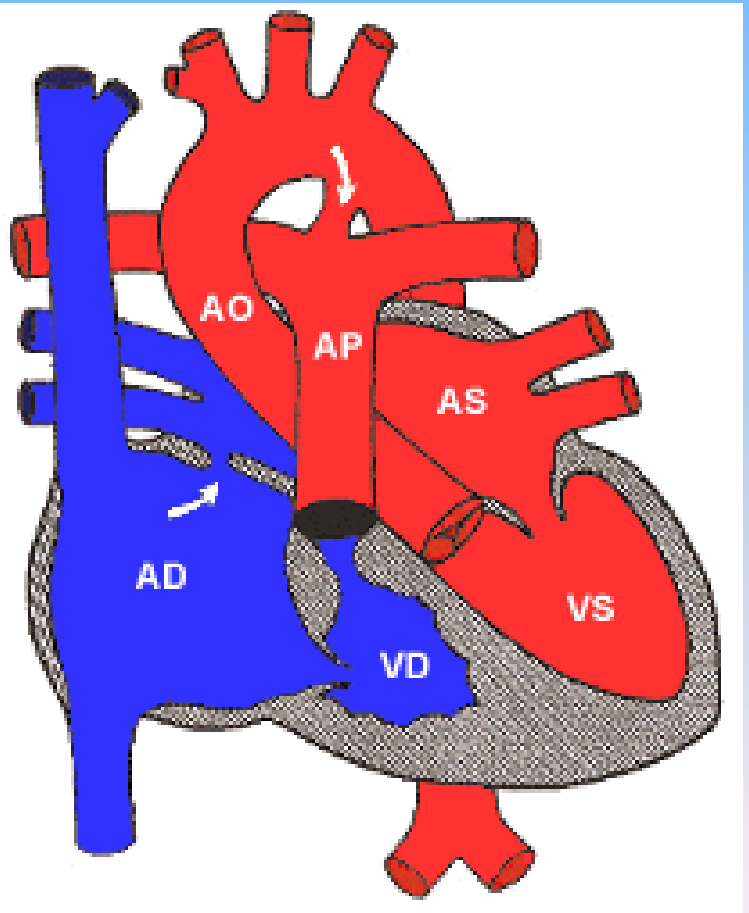
Cardiopatie Congenite con Dotto – Dipendenza del Circolo Polmonare

- ***Atresia polmonare a SIV integro***
- ***Atresia polmonare con DIV***
- ***Atresia della tricuspide***
- ***Ebstein (AP funzionale)***
- ***Tetralogia di Fallot grave***
- ***Stenosi polmonare critica***
- ***Cardiopatie complesse con atresia polmonare***

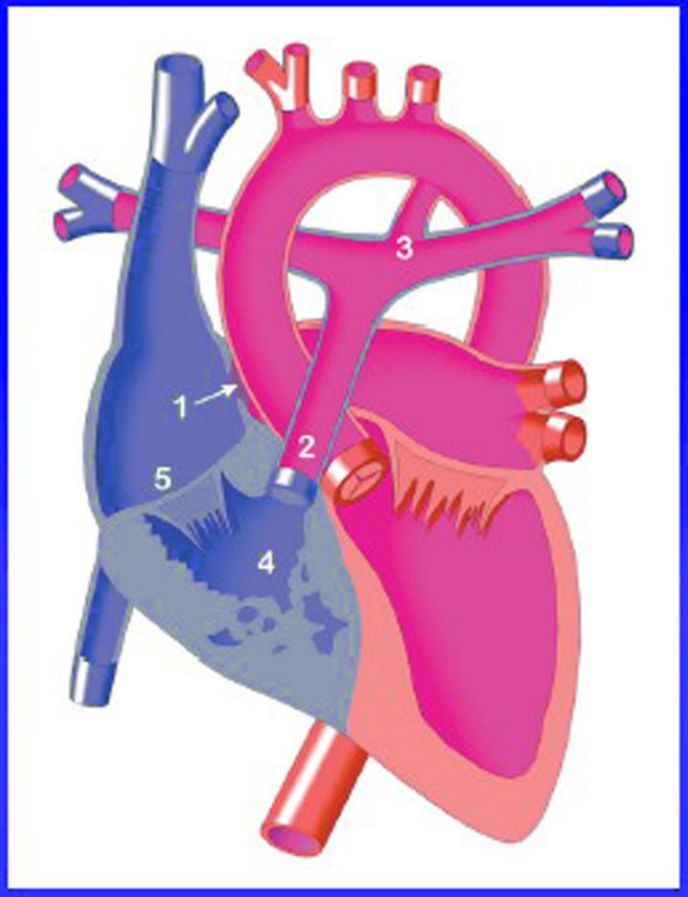
CIANOSI! →

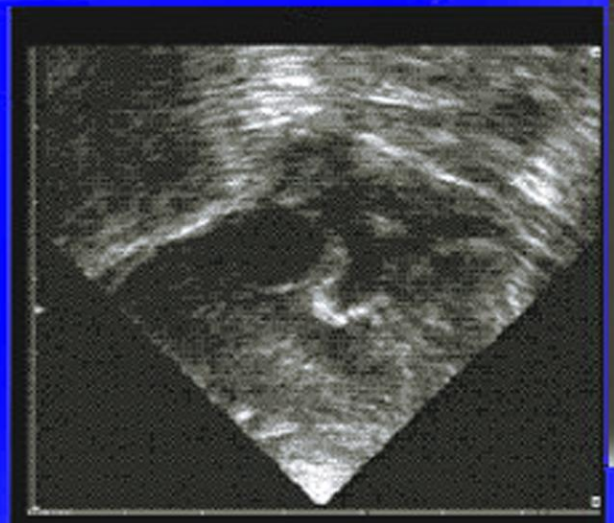
DOTTO-DIPENDENZA POLMONARE
Fisiopatologia

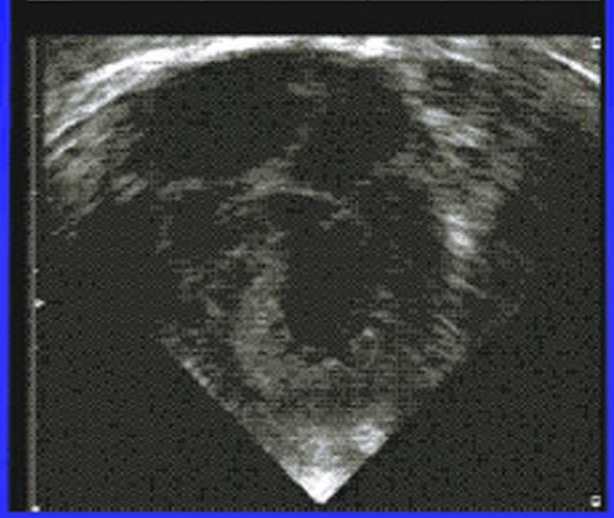
- **Ostruzione critica all' efflusso del ventricolo "polmonare"**
- **Dimensioni del dotto arterioso**

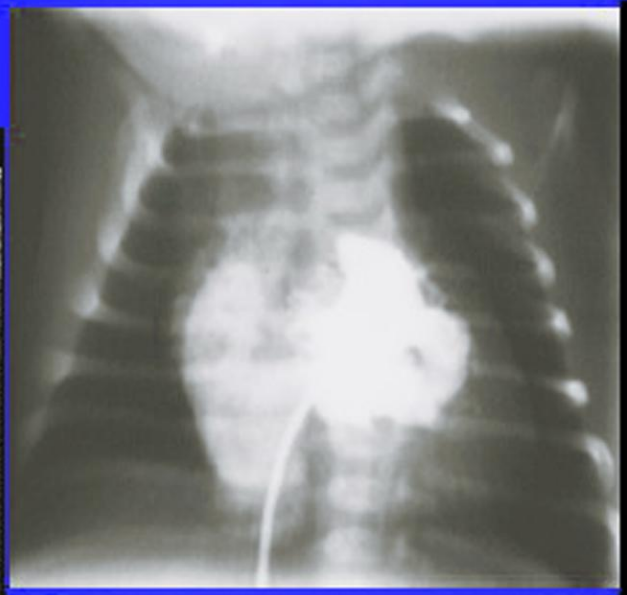


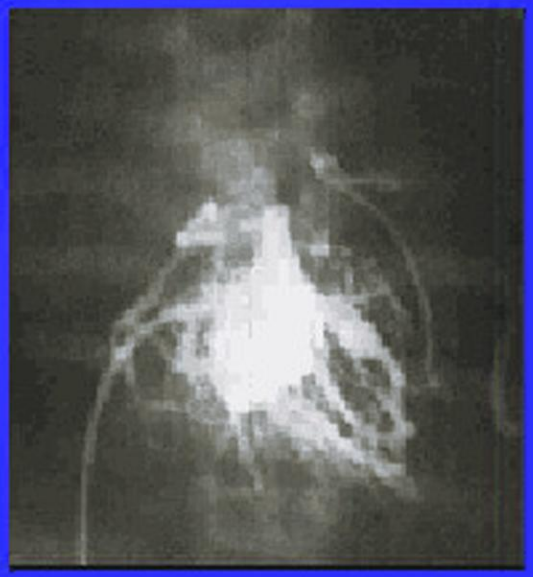
Atresia polmonare con setto integro



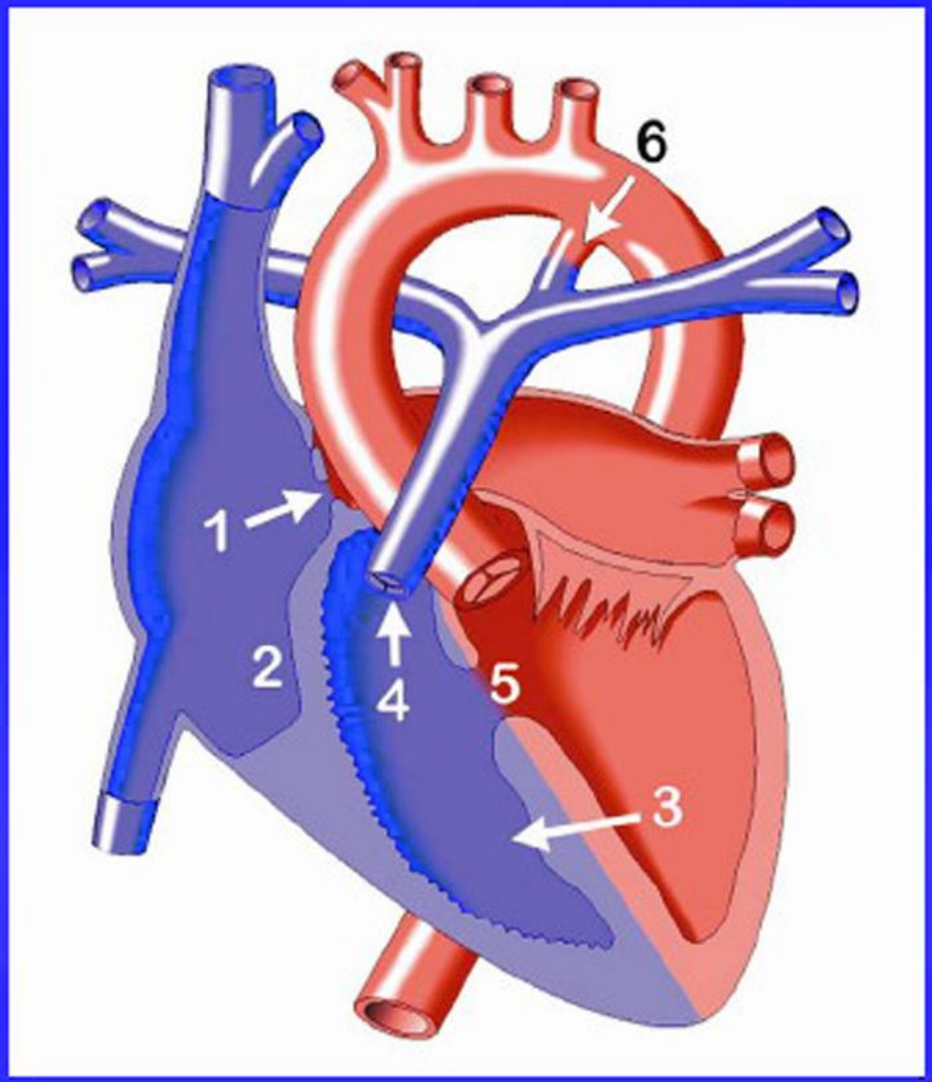


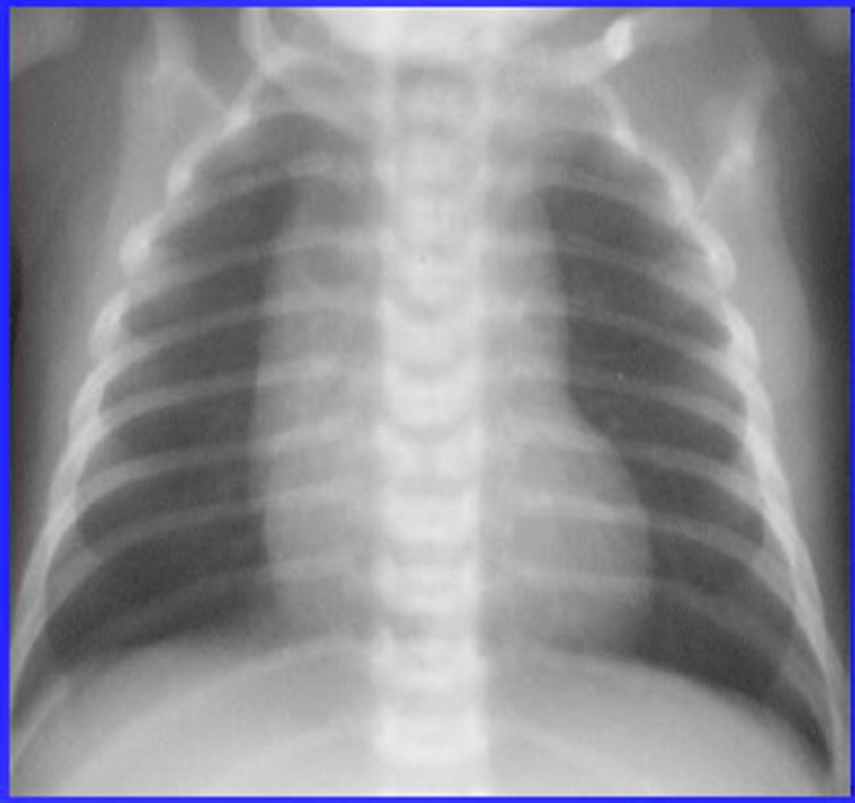






Atresia della Tricuspide





1. Atrial septal defect
2. Atretic (missing) tricuspid valve
3. Hypoplastic right ventricle
4. Pulmonary stenosis
5. Ventricular septal defect
6. Patent ductus arteriosus

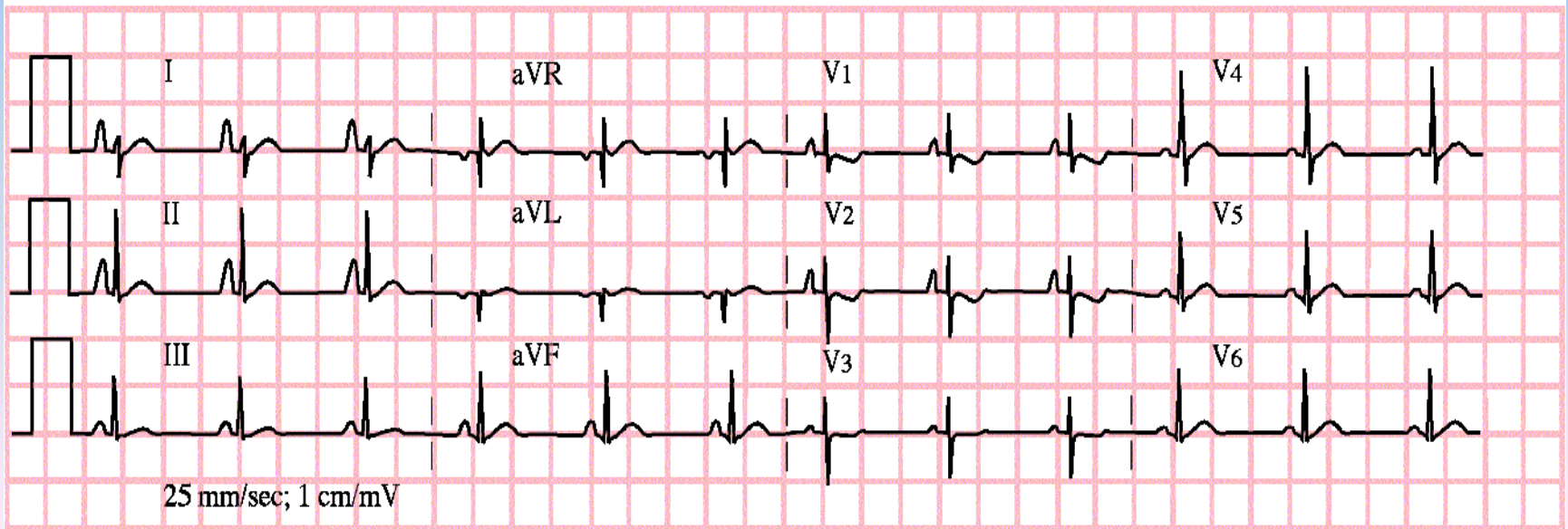
Dotto-dipendenza del circolo polmonare

Clinica: - *cianosi*

Ascoltazione: - soffio sistolico
- 2 tono polmonare diminuito o “unico”

ECG: - ingrandimento atriale dx, deviazione assiale dx

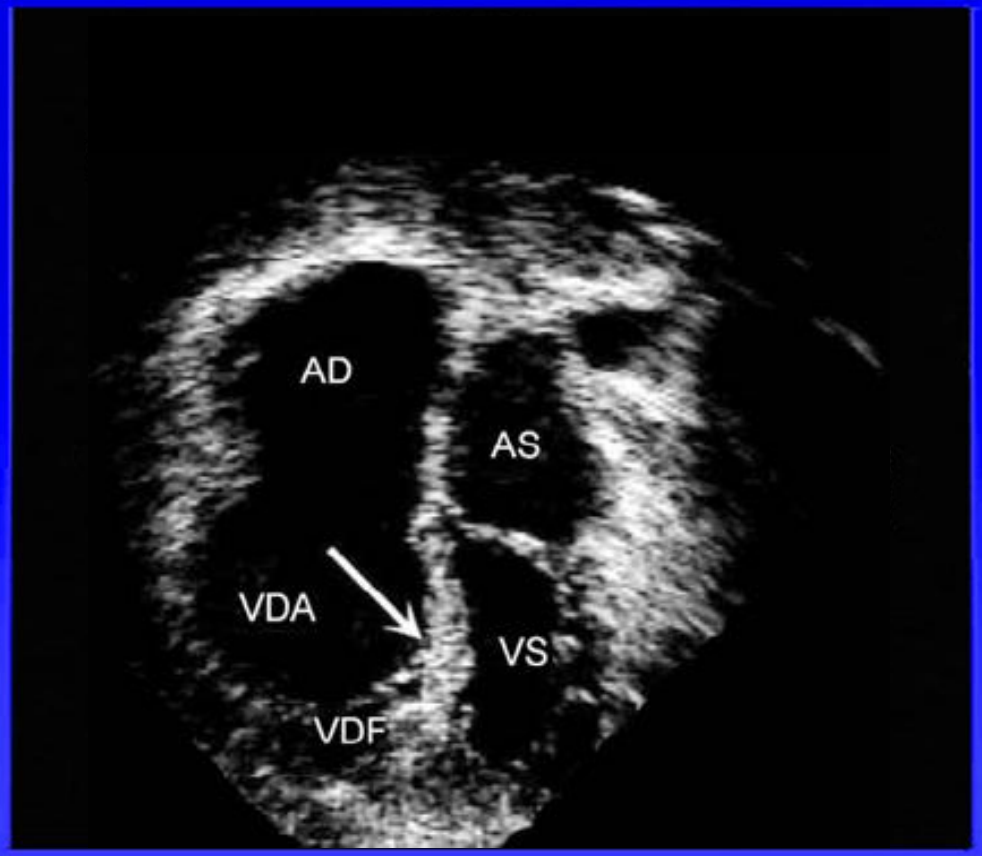
Radiologia: - campi polmonari ipovascolarizzati

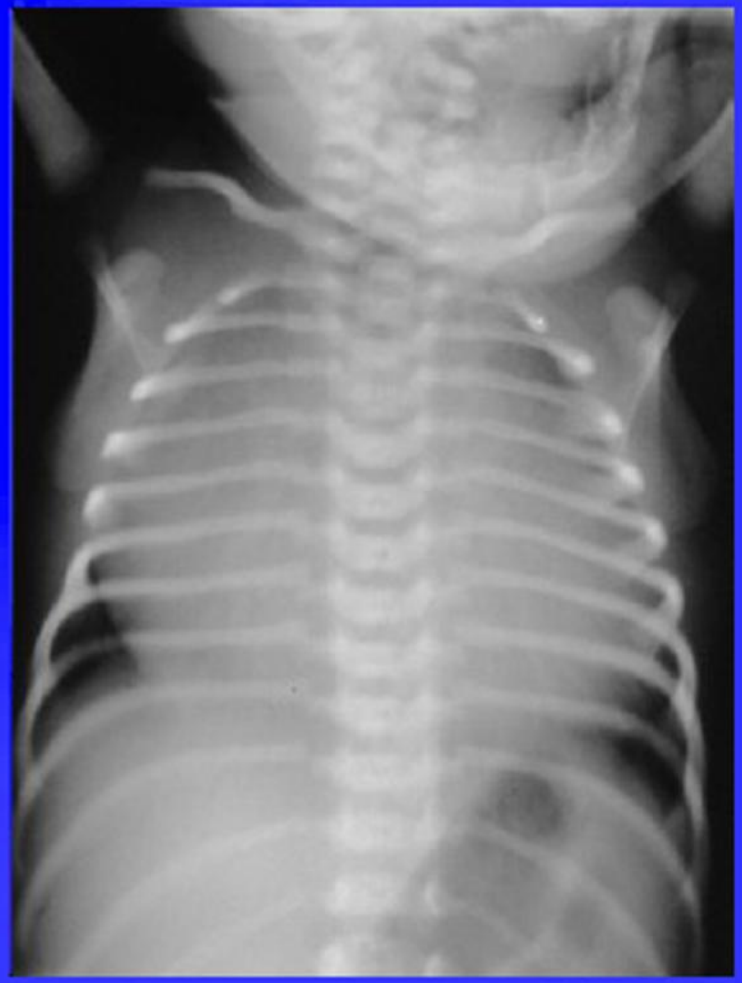


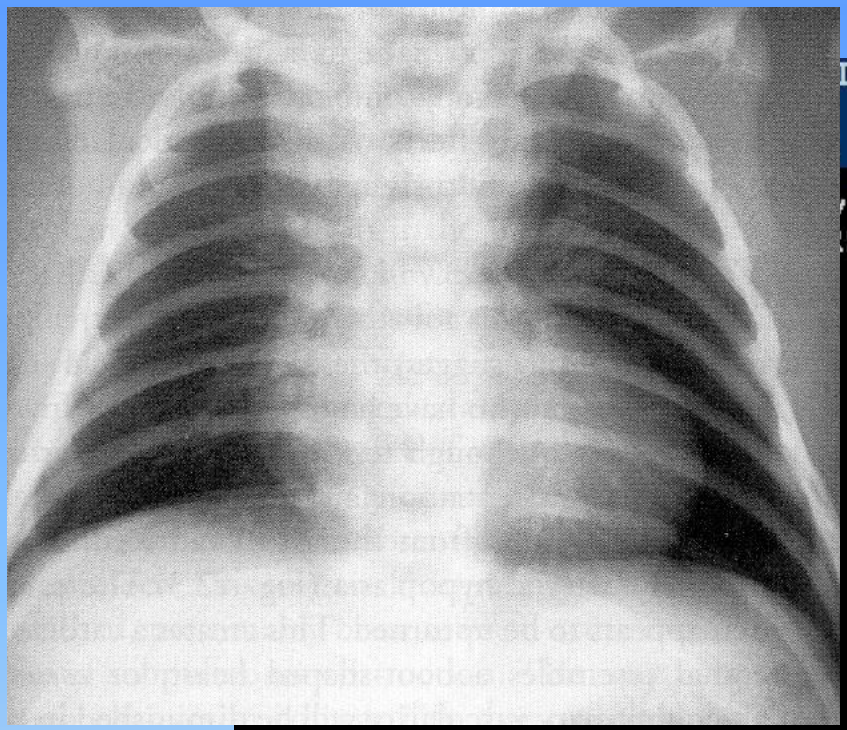
onare

DIAGNOSI ECOCARDIOGRAFICA!

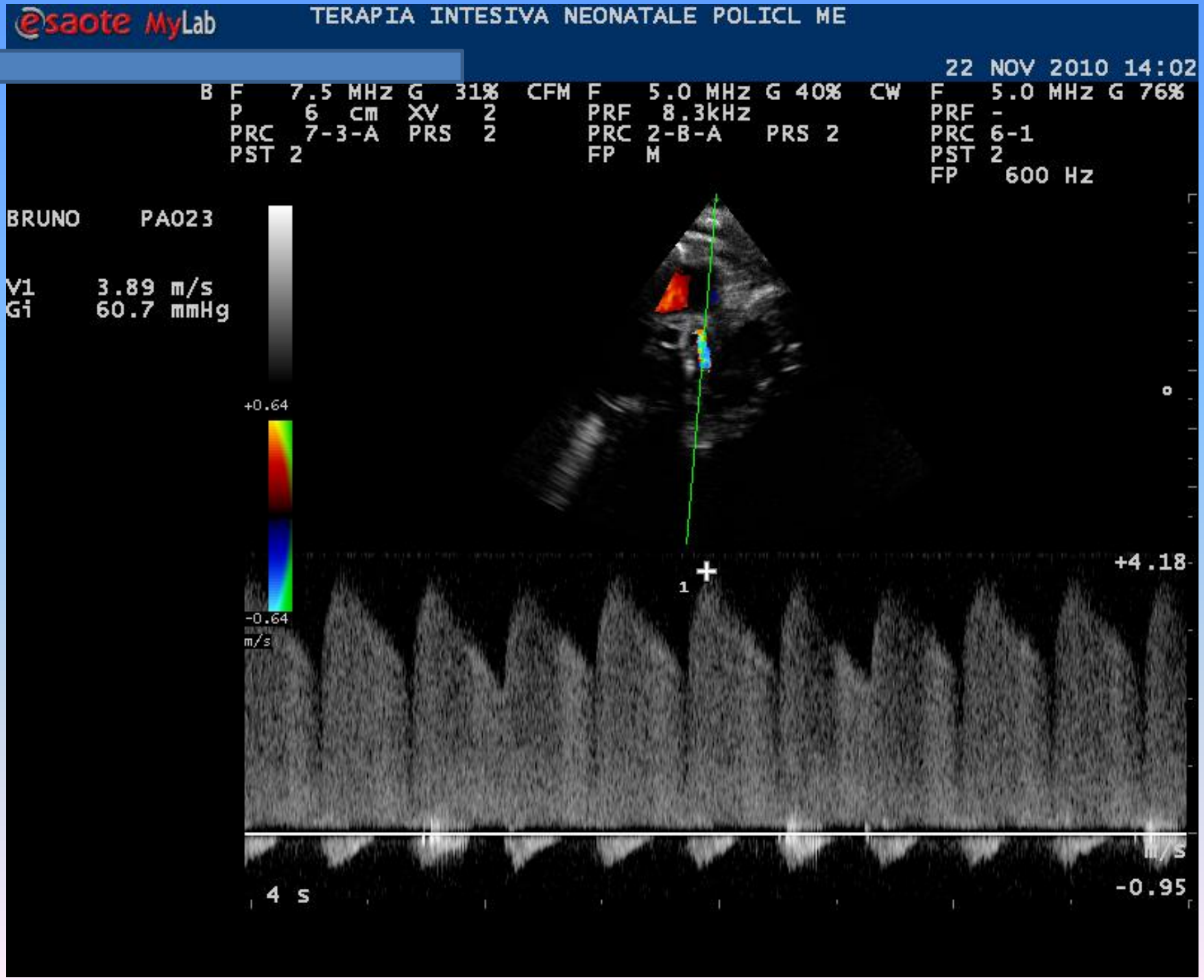
MALATTIA DI EBSTEIN



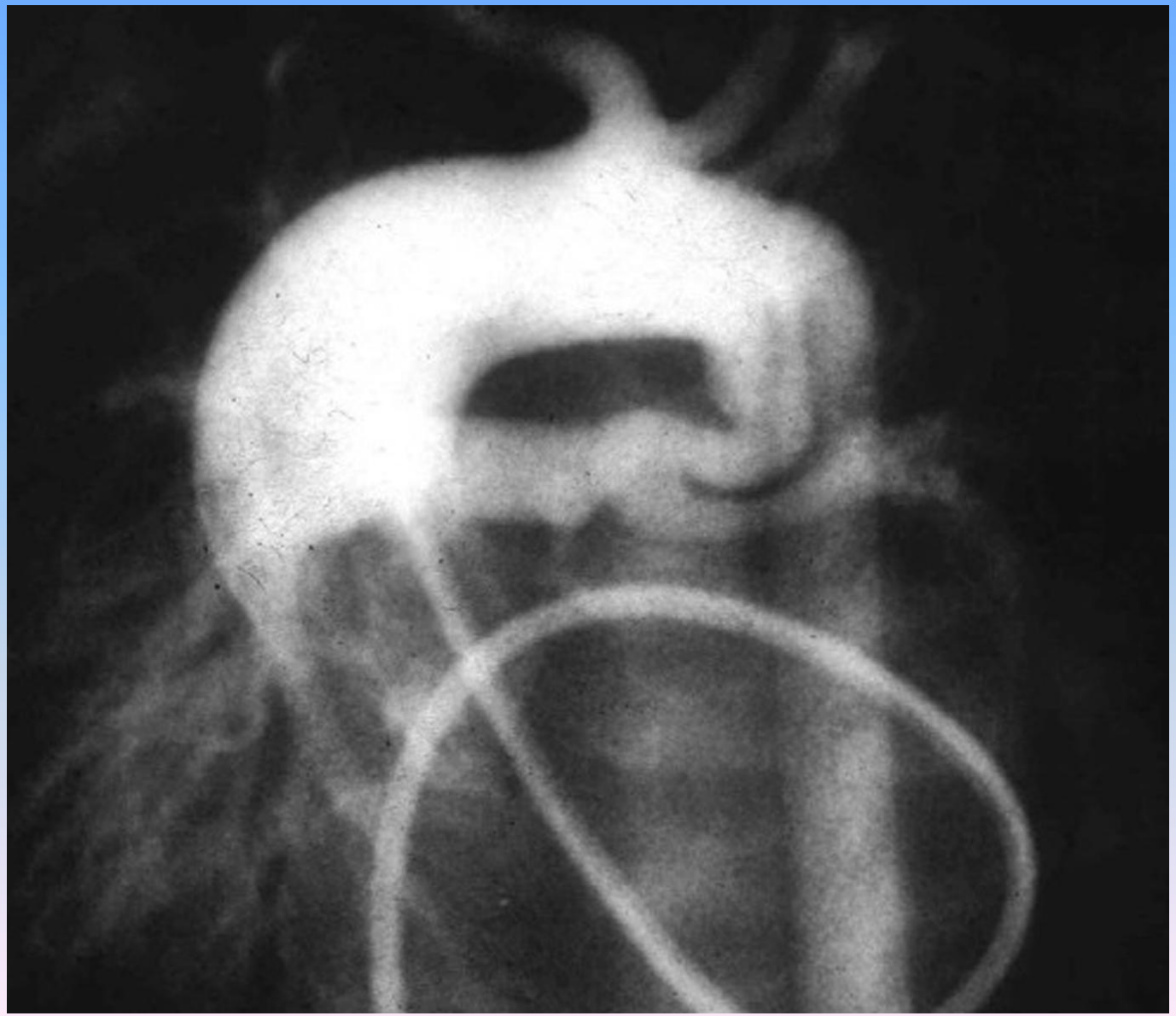




INTESIVA NEONATALE POLICL ME
22 NOV 2010 14:12
10%
2
S M
VD
AD
AO
AP


esaote MyLab
TERAPIA INTESIVA NEONATALE POLICL ME
22 NOV 2010 14:02
B F 7.5 MHz G 31% CFM F 5.0 MHz G 40% CW F 5.0 MHz G 76%
P 6 cm XV 2 PRF 8.3kHz PRF -
PRC 7-3-A PRS 2 PRC 2-B-A PRS 2 PRC 6-1
PST 2 FP M PST 2
FP 600 Hz
BRUNO PA023
V1 3.89 m/s
Gi 60.7 mmHg
+0.64
-0.64
m/s
+4.18
m/s
-0.95
4 s

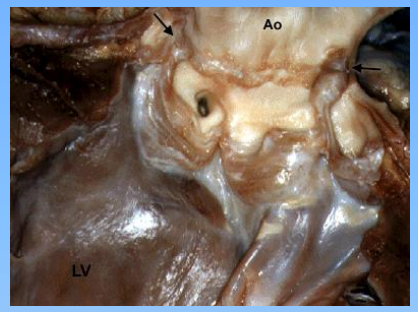
DOTTO DIPENDENZA POLMONARE



dotto-dipendenza del circolo polmonare
MANAGEMENT

- ***CORREZIONE ACIDOSI***
- ***CORREZIONE IPOTERMIA***
- ***PGE1 (0.01-0.05 γ/kg/min)***
- ***SEDAZIONE***
- ***INTUBAZIONE***
- ***TRASFERIMENTO DIFFERIBILE***
- ***TRATTAMENTO (emodinamica interventistica e/o chirurgia)***

Cardiopatie Congenite con Dotto – Dipendenza del Circolo Sistemico

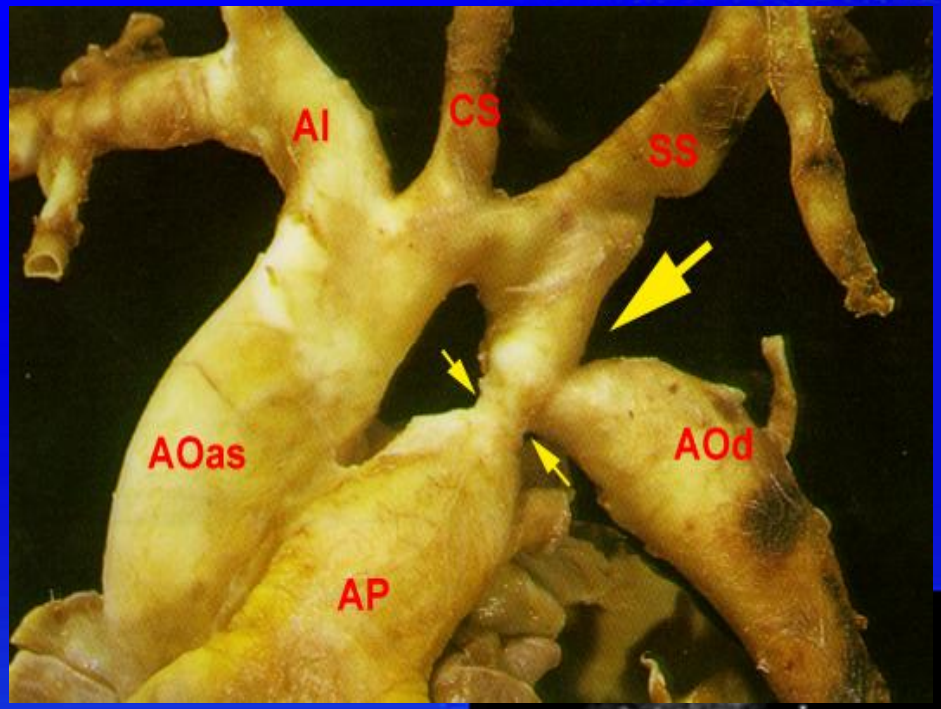


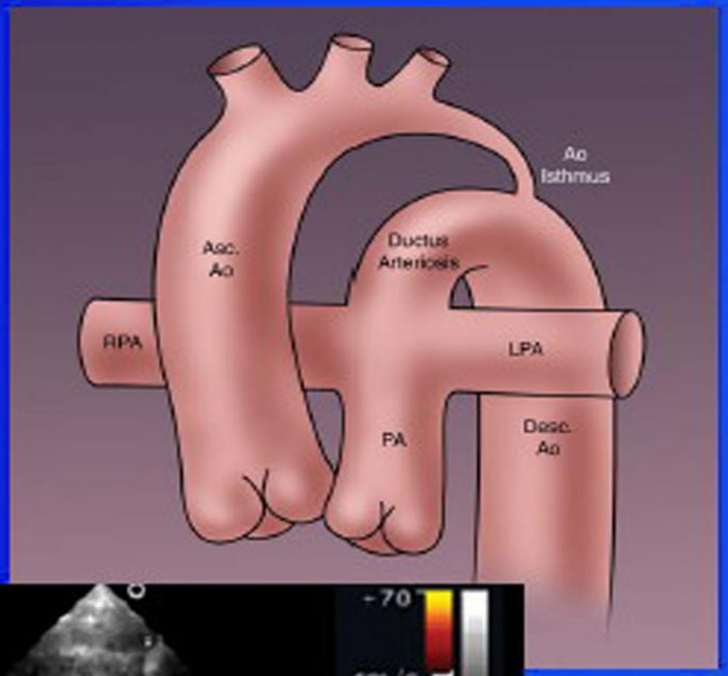
- **Stenosi aortica critica** ⟶
- **Coartazione aortica critica**
- **Interruzione dell'arco aortico**
- **Sindrome del cuore sinistro ipoplasico**

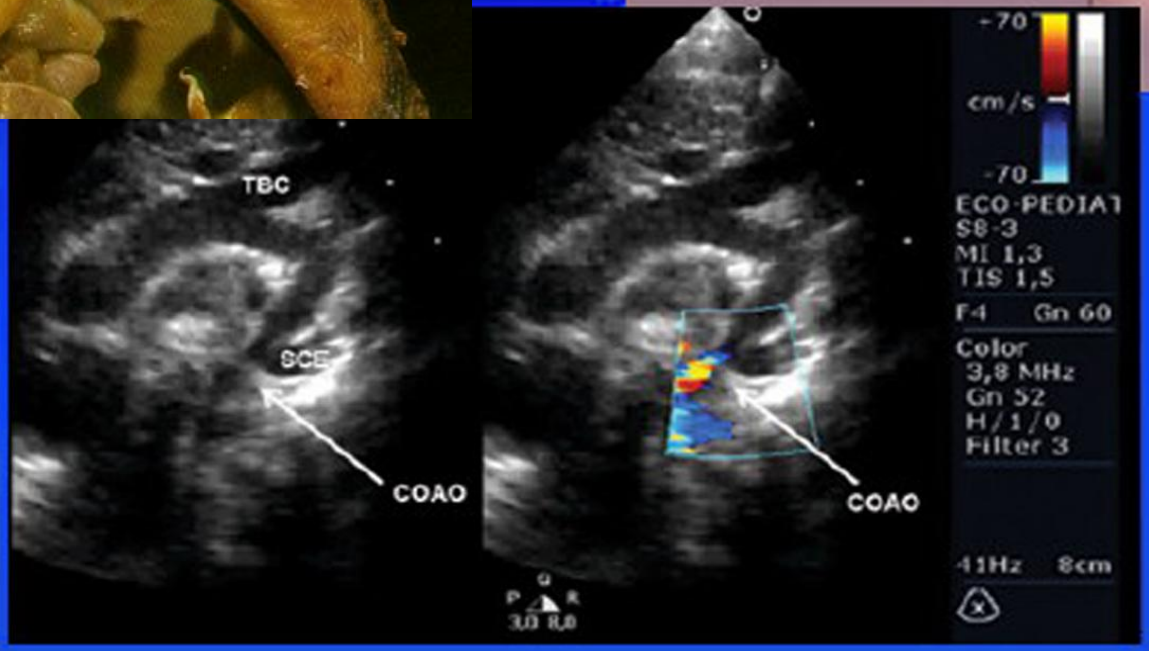
↓

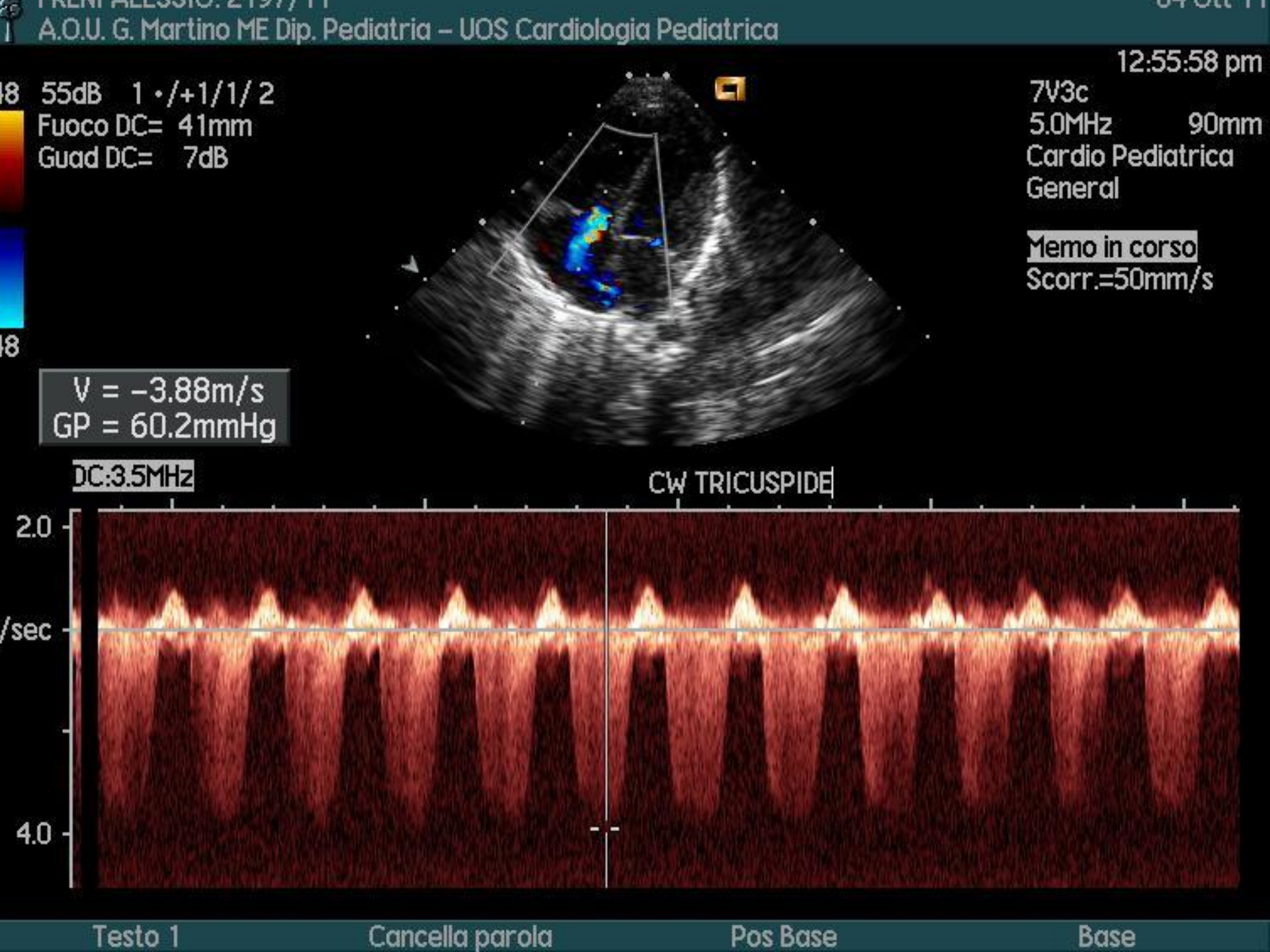
Scompenso cardiaco ⟶ **Shock cardiogeno** ⟶

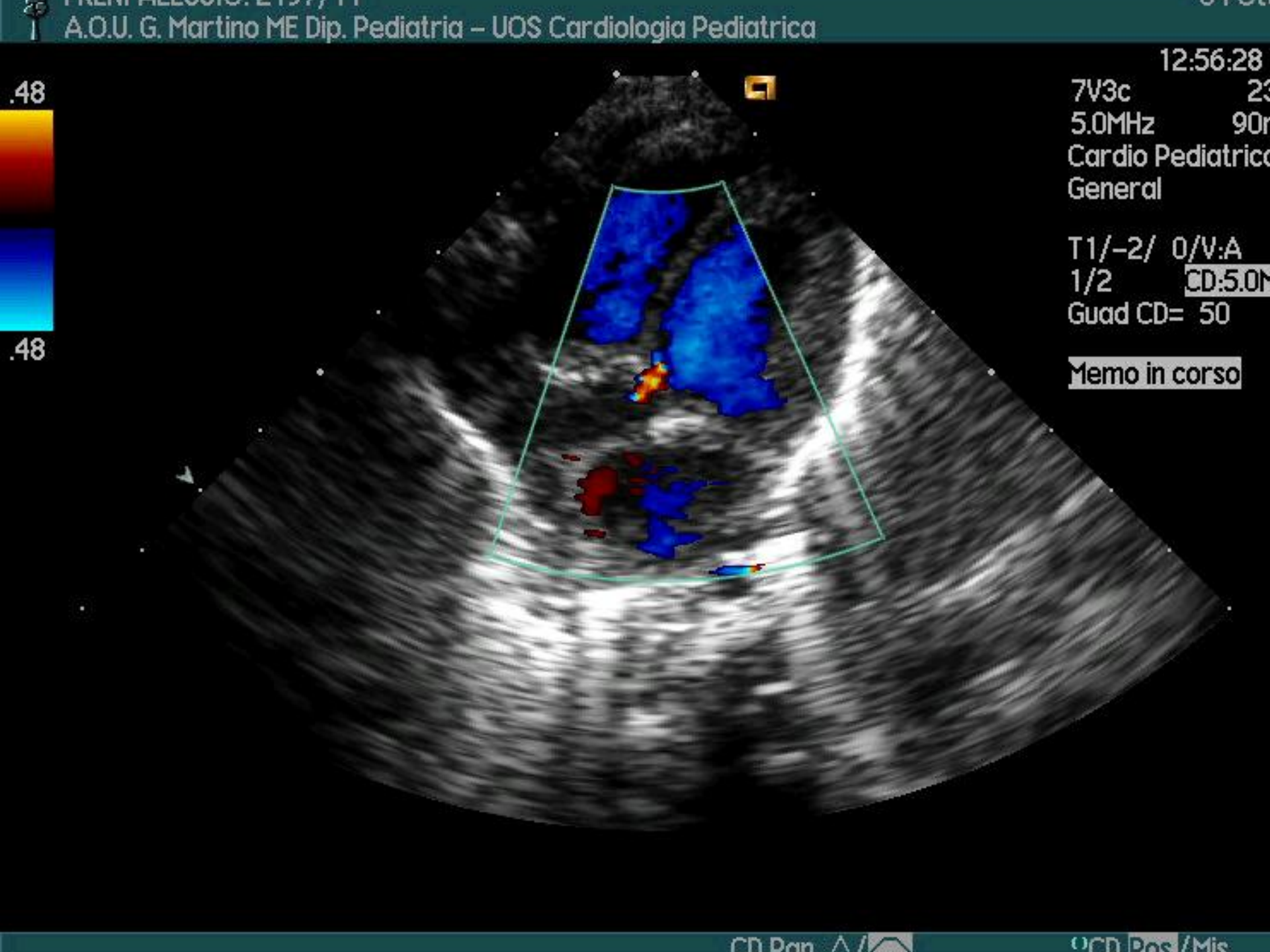
Coartazione aortica

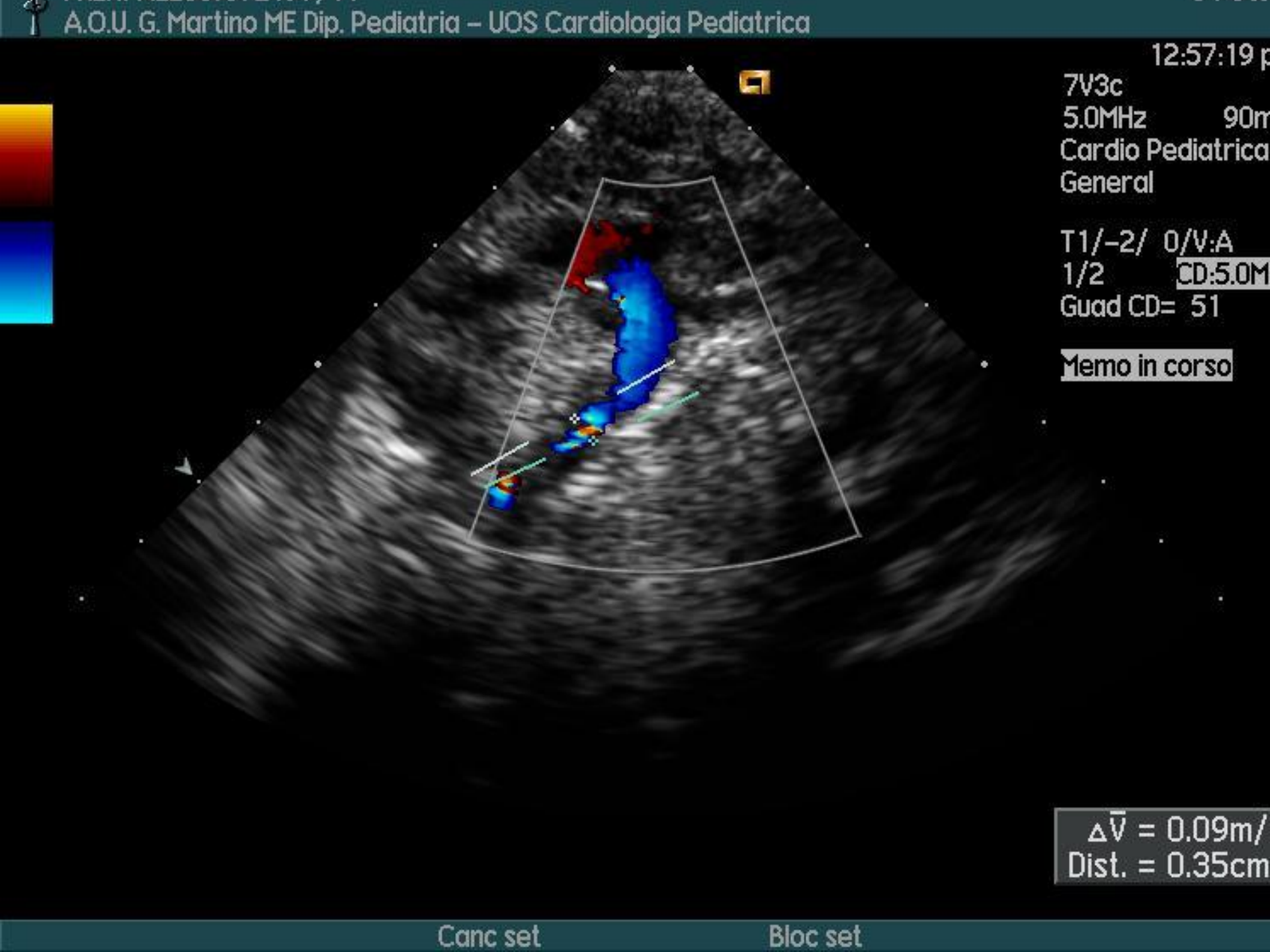


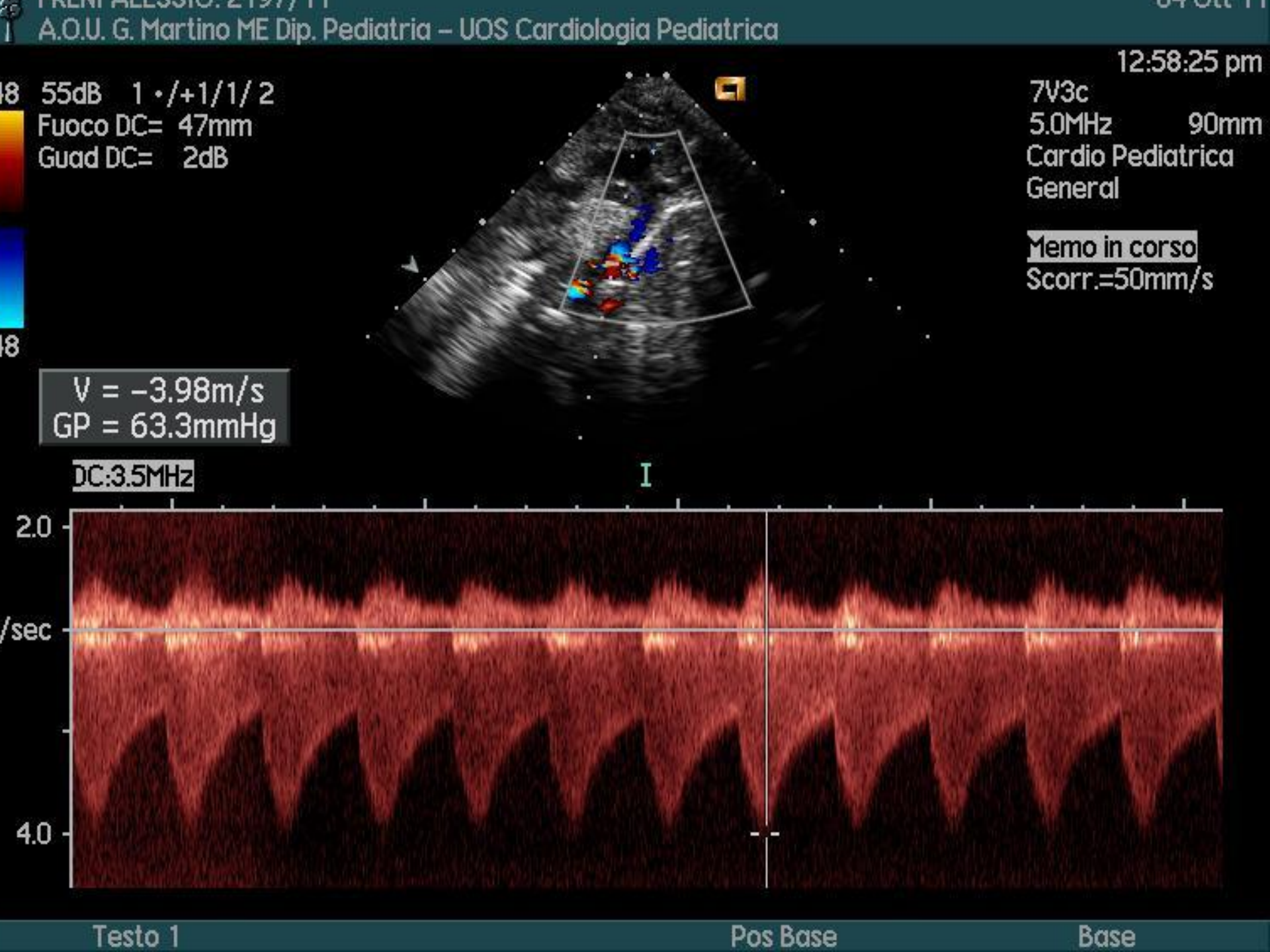




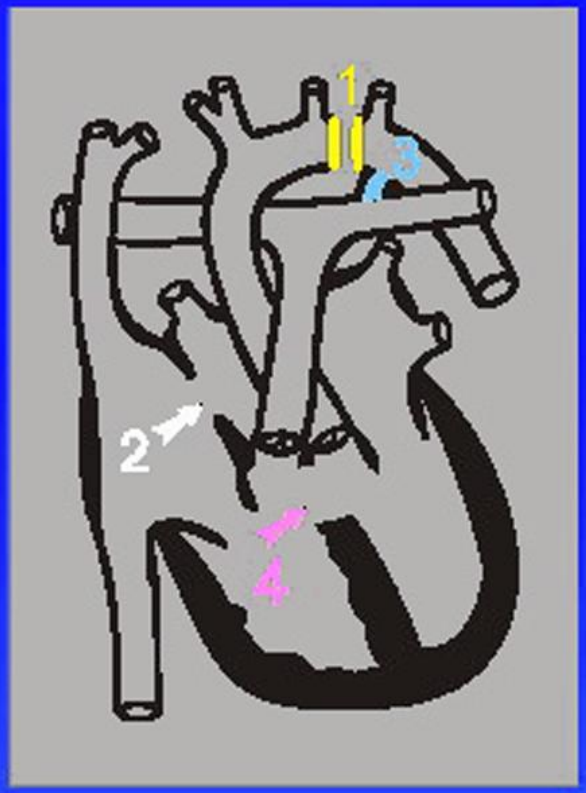

A.O.U. G. Martino ME Dip. Pediatria – UOS Cardiologia Pediatrica
12:55:58 pm
55dB 1 •/+1/1/ 2
Fuoco DC= 41mm
Guad DC= 7dB
7V3c
5.0MHz 90mm
Cardio Pediatrica
General
Memo in corso
Scorr.=50mm/s
V = -3.88m/s
GP = 60.2mmHg
DC:3.5MHz
CW TRICUSPIDE
2.0
/sec
4.0
Testo 1
Cancella parola
Pos Base
Base

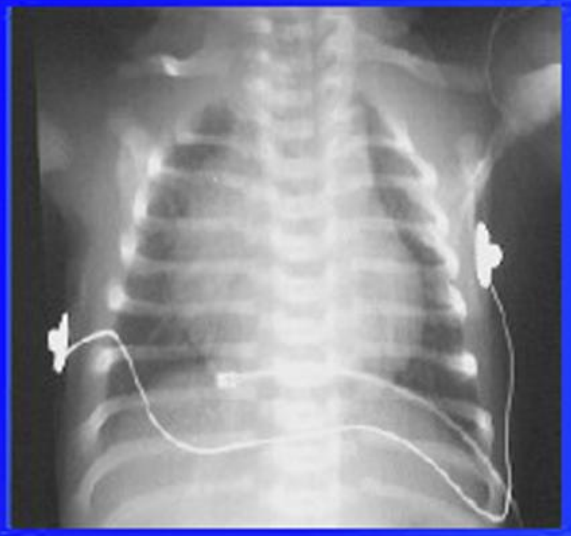

A.O.U. G. Martino ME Dip. Pediatria – UOS Cardiologia Pediatrica
12:56:28
7V3c
5.0MHz
Cardio Pediatrica
General
T1/-2/ 0/V:A
1/2
Guad CD= 50
Memo in corso
.48
.48

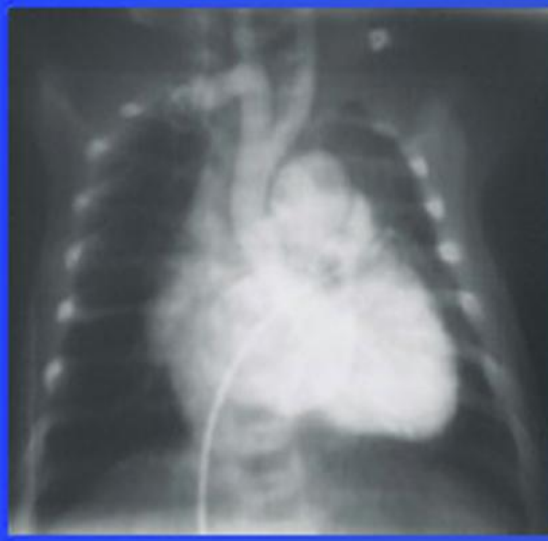

A.O.U. G. Martino ME Dip. Pediatria – UOS Cardiologia Pediatrica
12:57:19 p
7V3c
5.0MHz 90m
Cardio Pediatrica
General
T1/-2/ 0/V:A
1/2 CD:5.0M
Guad CD= 51
Memo in corso
ΔV̄ = 0.09m/
Dist. = 0.35cm
Canc set
Bloc set


A.O.U. G. Martino ME Dip. Pediatria – UOS Cardiologia Pediatrica
12:58:25 pm
55dB 1 •/+1/1/ 2
Fuoco DC= 47mm
Guad DC= 2dB
7V3c
5.0MHz 90mm
Cardio Pediatrica
General
Memo in corso
Scorr.=50mm/s
V = -3.98m/s
GP = 63.3mmHg
DC:3.5MHz
2.0
/sec
4.0
Testo 1
Pos Base
Base

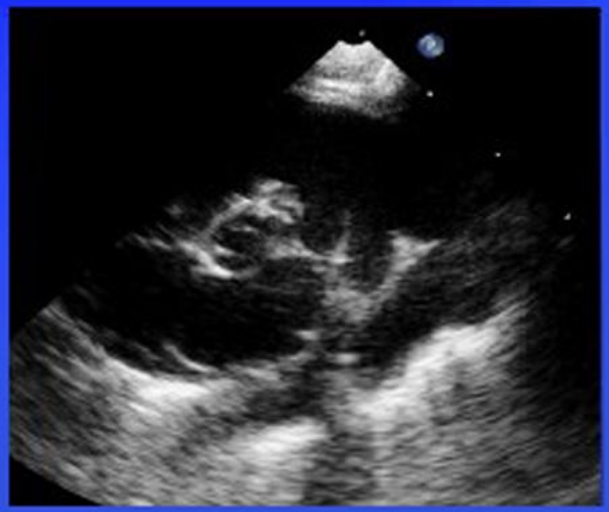
Interruzione dell' arco aortico

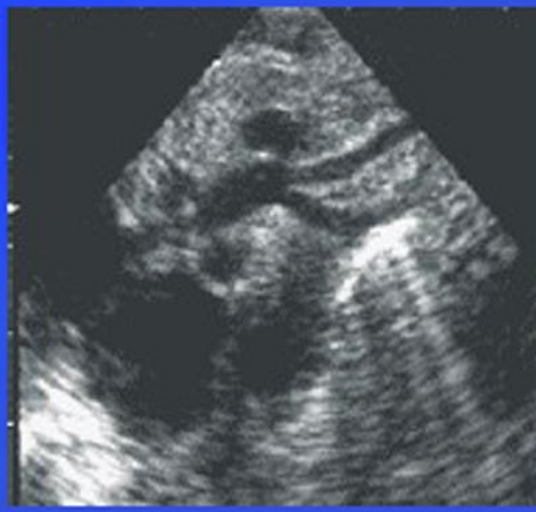




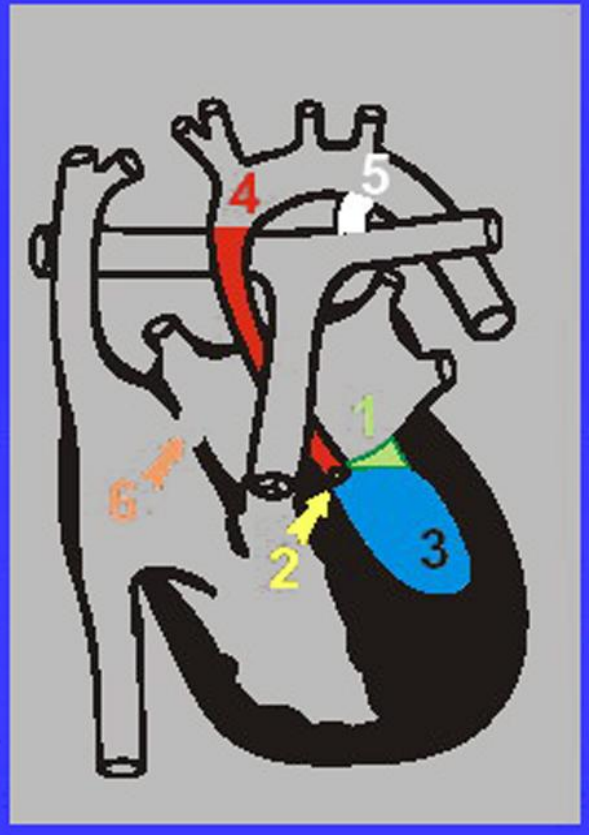


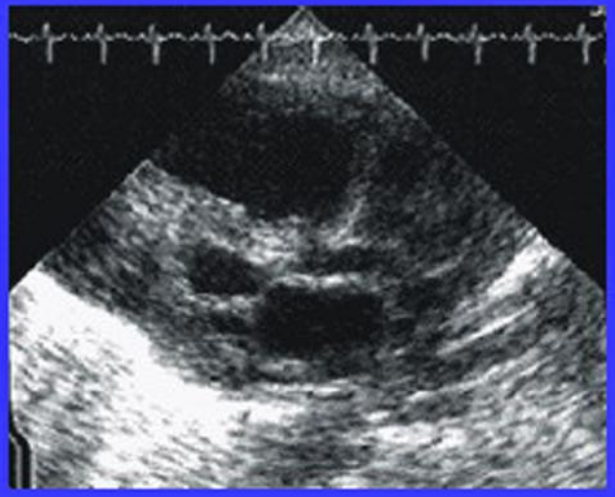
1. Discontinuity of the aortic arch between the left carotid and left subclavian arteries
2. Atrial septal defect
3. Patent ductus arteriosus
4. Ventricular septal defect



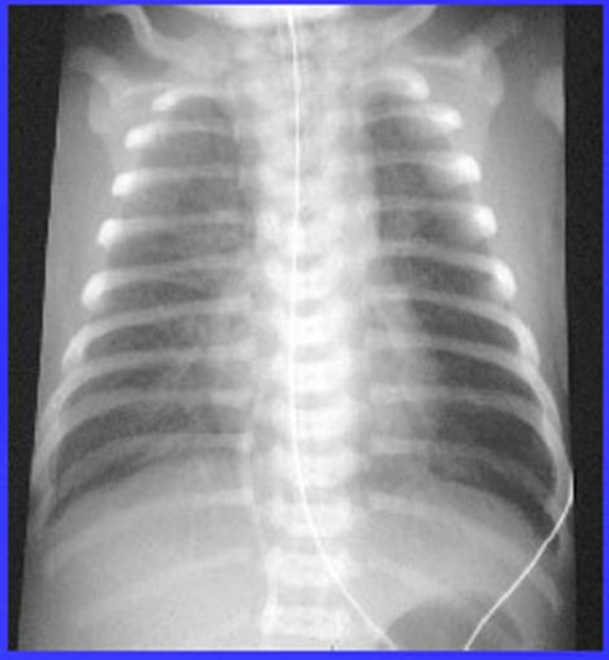


Sindrome del cuore sinistro ipoplasico



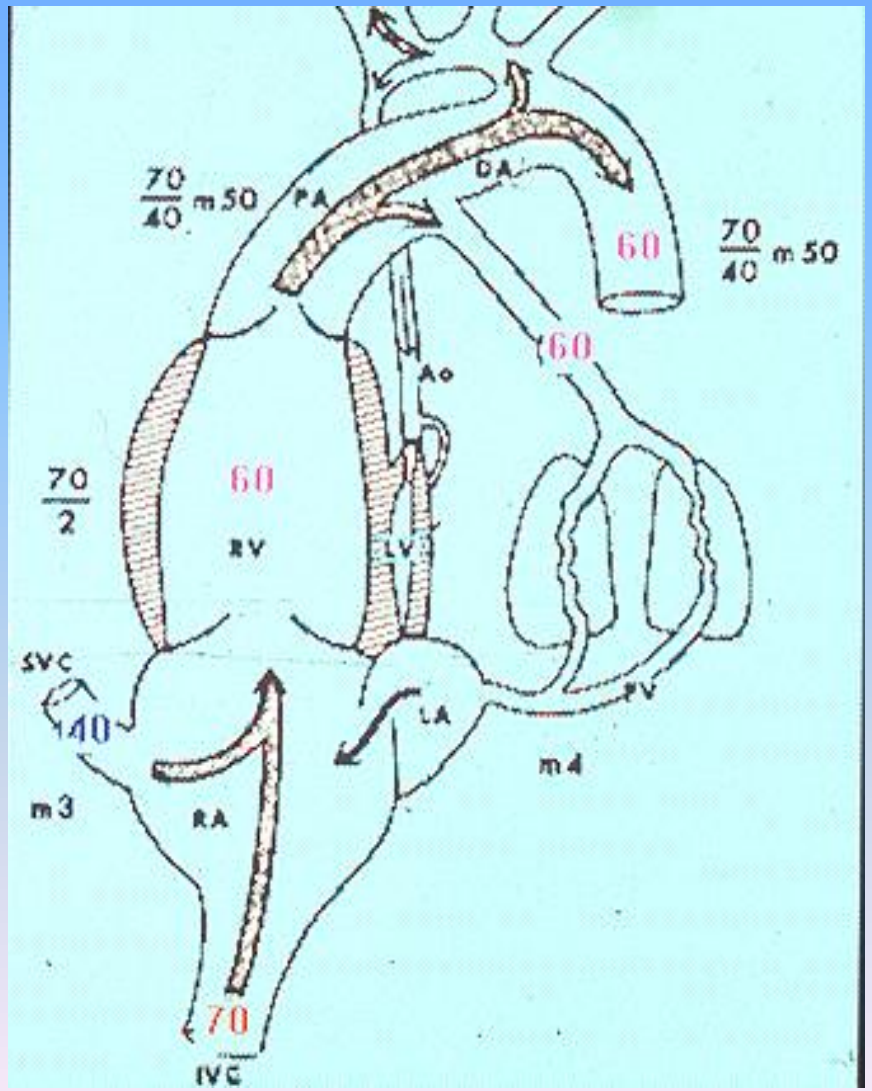


1) Mitral valve atresia
2) Severe aortic stenosis
3) Hypoplastic left ventricle
4) Hypoplastic asc. aorta
5) Patent ductus arteriosus
6) Patent foramen ovale



Sindrome del cuore sinistro ipoplasico Fisiopatologia

- **Ventricolo destro sistemico**
- **Dotto – dipendenza del circolo coronarico**




Stabilizzare preoperatoriamente il neonato
Trasporto ad un centro di cardiochirurgia
PGE1
0.05-0.1
mcg/kg/min
Sostenere il neonato dopo l'intervento chirurgico
Posticipare l'intervento chirurgico ad un momento più favorevole (impiego a lungo termine)

Grazie!