UNIUNEA EUROPEANĂ | GUVERNUL ROMÂNIEI | Fondul Social European POSDRU 2007-2013 | Instrumente Structurale 2007-2013 | GUVERNUL ROMÂNIEI MINISTERUL MUNCII, FAMILIEI, PROTECȚIEI SOCIALE ȘI PERSOANELOR VÂRSTNICE OIRPOSDRU REGIUNEA CENTRU | UNIVERSITATEA DE MEDICINĂ ȘI FARMACIE "CAROL DAVILA" BUCUREȘTI

«AD-COR Program inovativ de formare in domeniul cardiologiei pediatrice POSDRU/179/3.2/S/152012"

Iulie 2015

MODUL TEORETIC

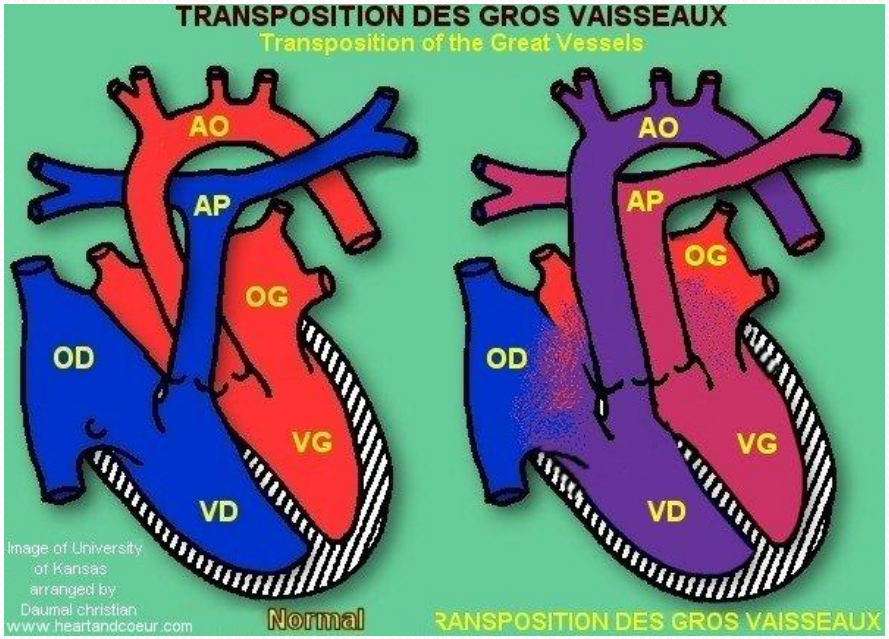
MCC CIANOGENE CU DEBIT PULMONAR CRESCUT

Continut documentat/ validat/ prezentat de:

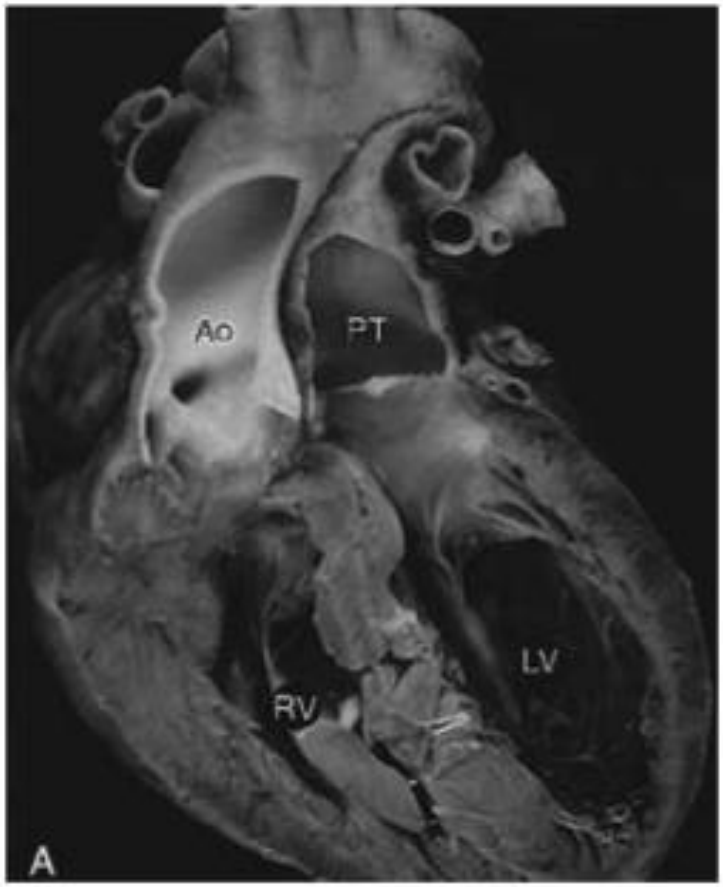
- Expert formare medici: NICOLESCU Alin
- Expert formare medici: VEDUTA Alina

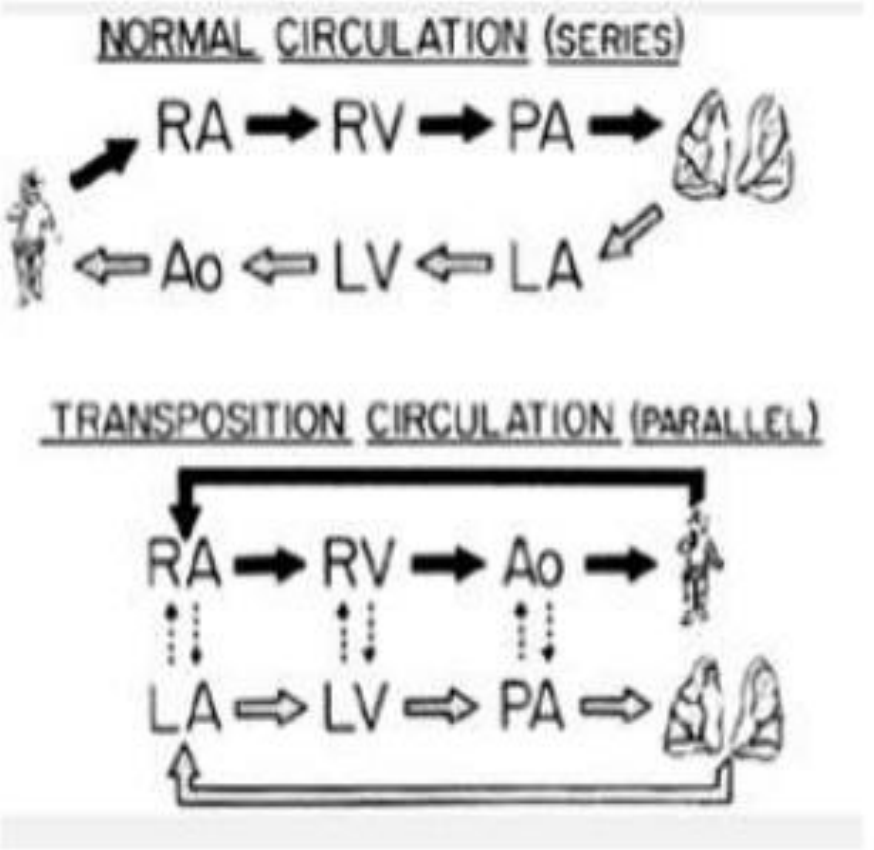
- TRANSPOZITIA DE MARI VASE
- TRUNCHI ARTERIAL COMUN
- VENTRICUL UNIC

TRANSPOZITIA DE MARI VASE



- 5% din MCC
  - Ao are origine in VD
  - AP are origine in VS
  - PFO/DSA
  - DSV -30%
  - DSV+obstructie semnificativa CEVS: 10%
  - Asociere alte MCC: CoAo,Arc aortic intrerupt
  - Forma clasica D transpozitie,Ao este situata anterior si la dreapta fata de AP


Ao
PT
LV
RV
A


NORMAL CIRCULATION (SERIES)
RA
RV
PA
Ao
LV
LA
TRANSPOSITION CIRCULATION (PARALLEL)
RA
RV
Ao
LA
LV
PA

Fiziopatologie

- TMV+PFO(DSA restrictiv)

-Nn este cianotic la nastere si are sat O2=30-50%

Rezulta **glicoliza anaeroba** → **acidoza metabolica**

-hipoxie+acidoza → **disfunctie miocardica**

-postnatal

- scad RVP si creste fluxul pulmonar
- suprasolicita AS si VS.Asociat cu disfunctia miocardica=insuf cardiaca

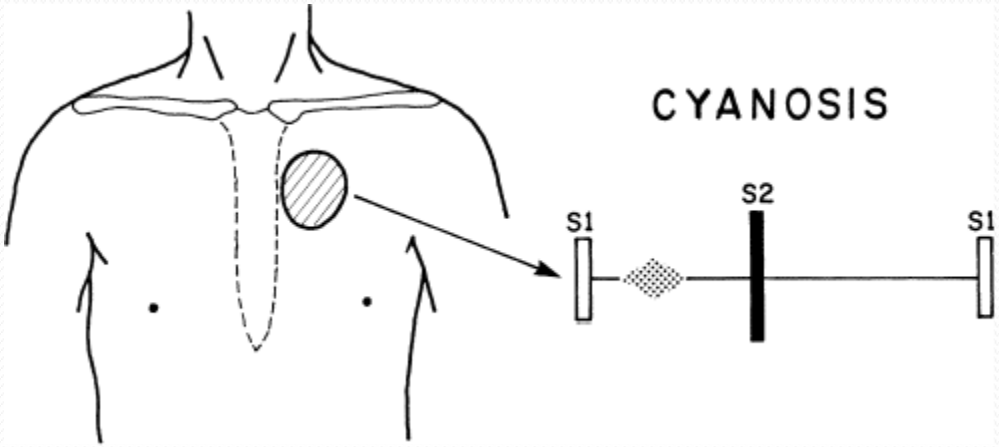
- TMV+ DSA

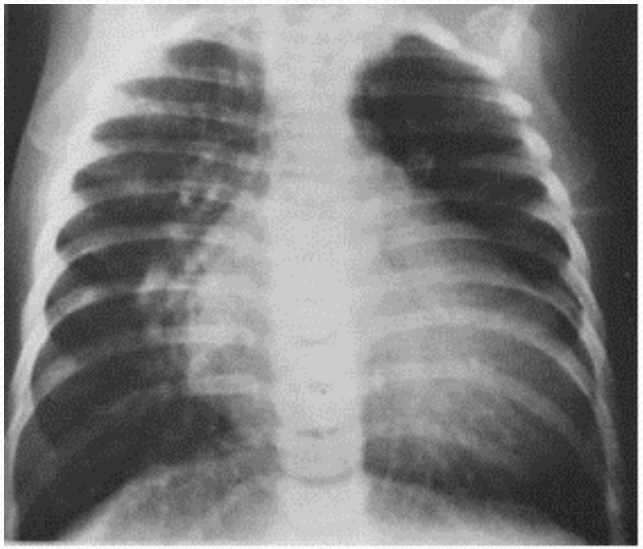
Sat O_2 =80 %

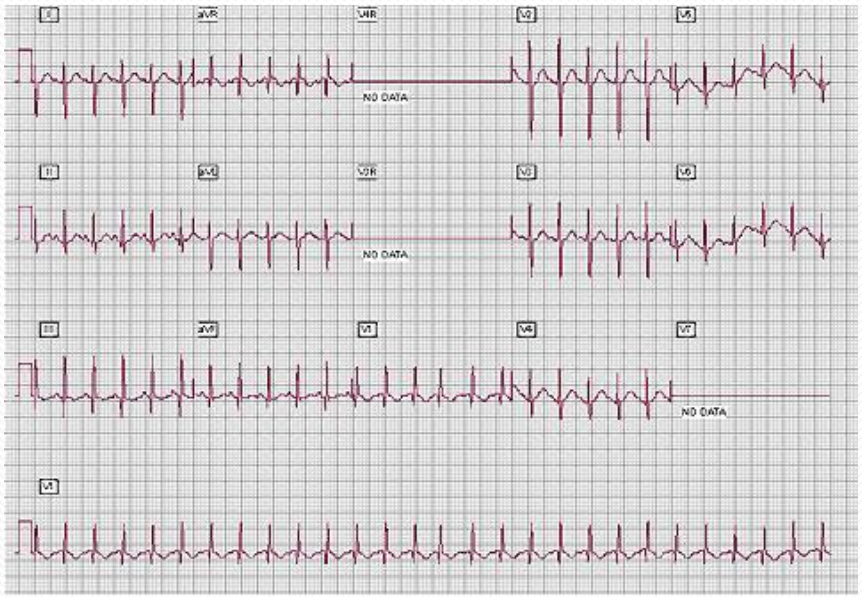
-nu apare acidoza si nici IC in primele 10-15 zile postnatal

Deci DSA nerestrictiv este esential in evolutia clinica a TMV

Rashkind

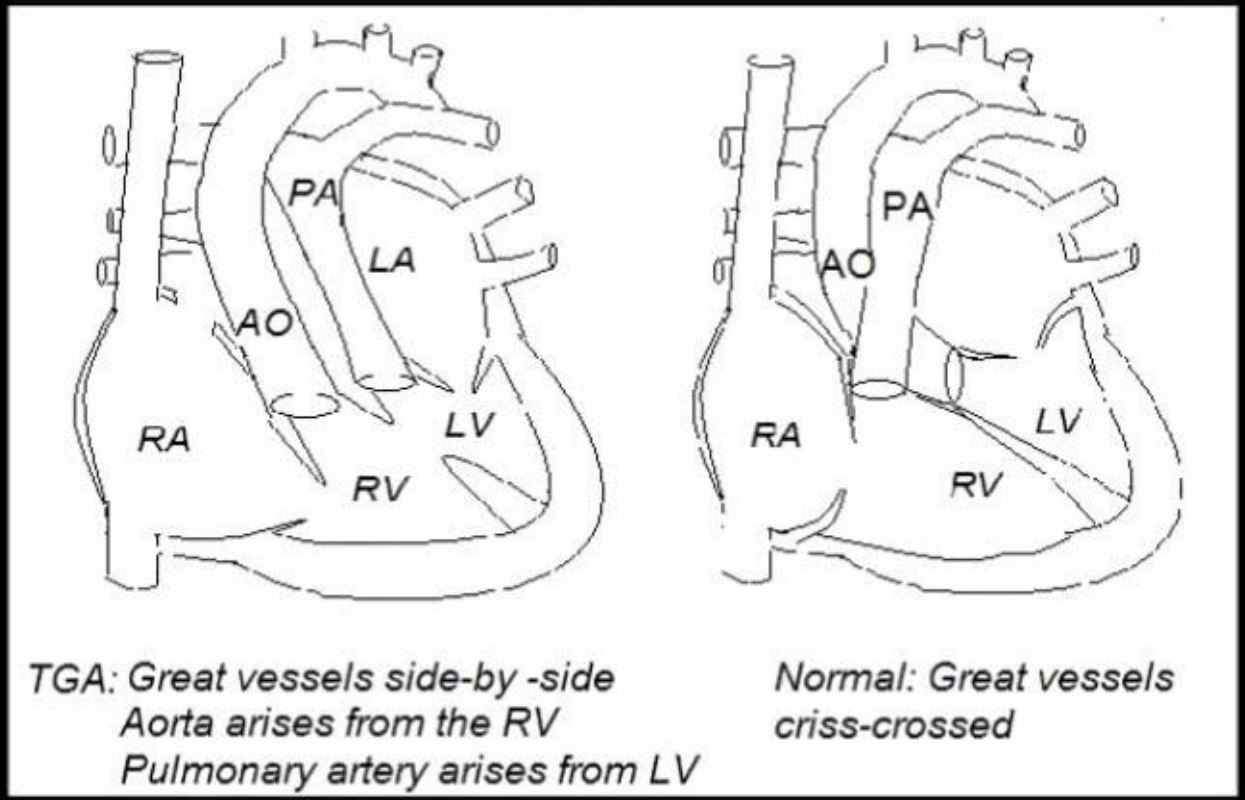






Children's Hospital Boston

ECO



TGA: Great vessels side-by -side
Aorta arises from the RV
Pulmonary artery arises from LV

Normal: Great vessels criss-crossed

Evolutie naturala

- Hipoxie,acidoza,ICC-90% deces in primele 6 l
- TMV cu SIV intact este grupul cel mai afectat ,cu imbunatatire spectaculoasa dupa Rashkind
- TMV+CAP evolutie ca TMV+DSV
- TMV+DSV+St pulm=plaman protejat,evolutia cea mai buna

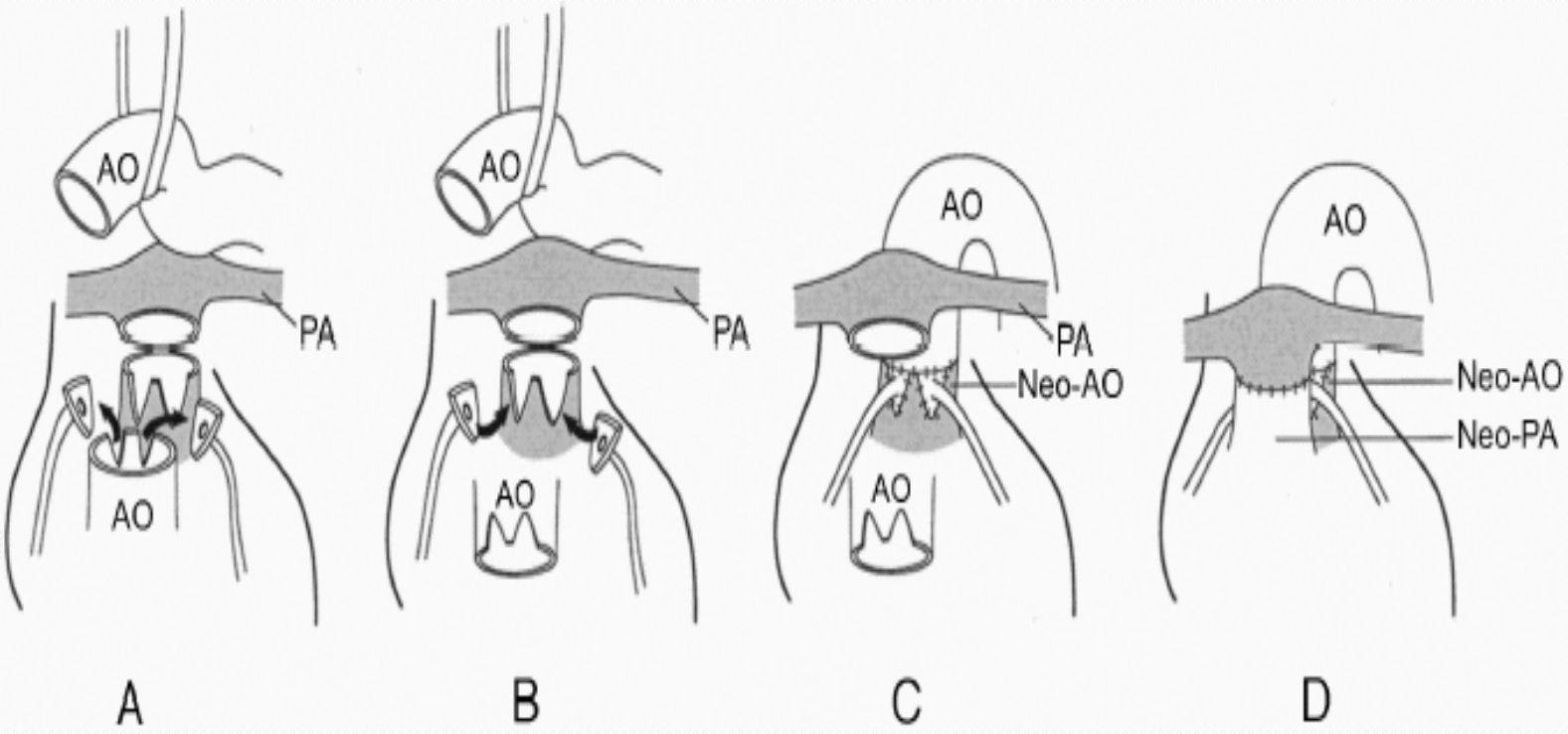
Management

- Medical
- -corectia acidozei
- -corectia hipoglicemiei
- -corectia hipocalcemiei
- Prostaglandina E1-deschiderea si mentinerea CAP
- Tratamentul IC: diuretic ,suport inotrop

Management

- Chirurgical

- Timing:
- TMV /+DSV/+CAP --2 sapt
- TMV-st pulm: 6-9l
- Mortalitate <5 %

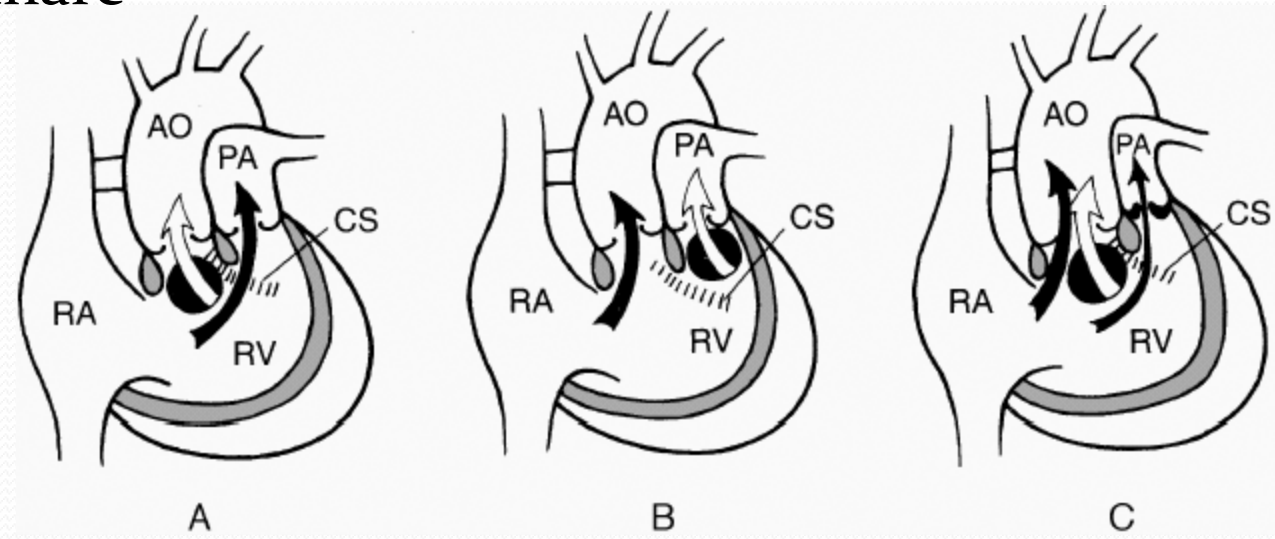


Dubla cale de iesire VD

< 1% din MCC

Ao dextropusa >50%.Ambele vase mari au originea in VD
-vase mari normopuse/transpuse
-DSV: subaortic sau subpulmonar(Taussig-Bing)

-Discontinuitate intre inelul mitral si inelul vavelor semilunare

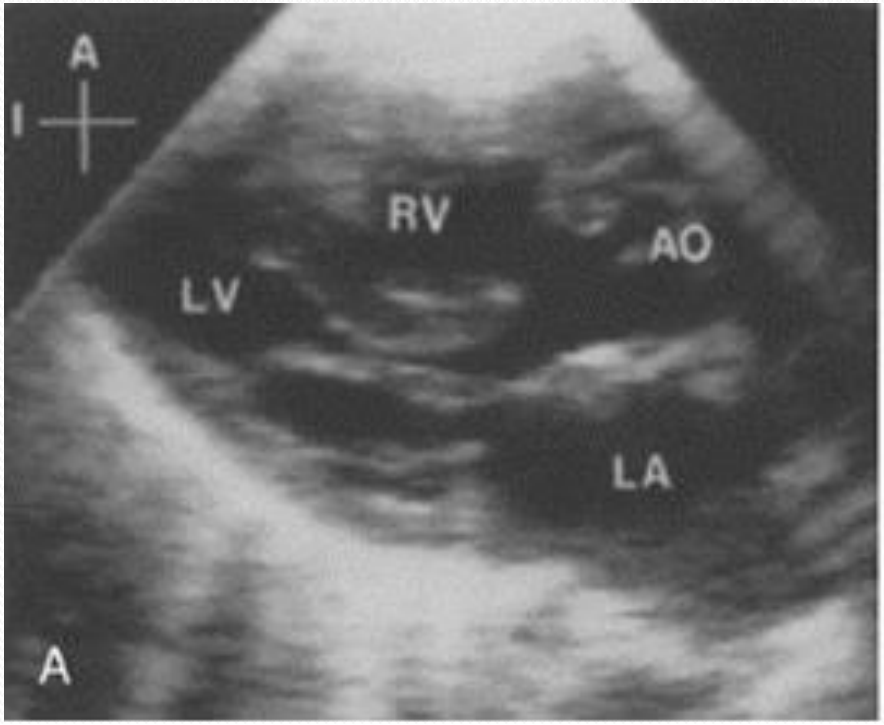


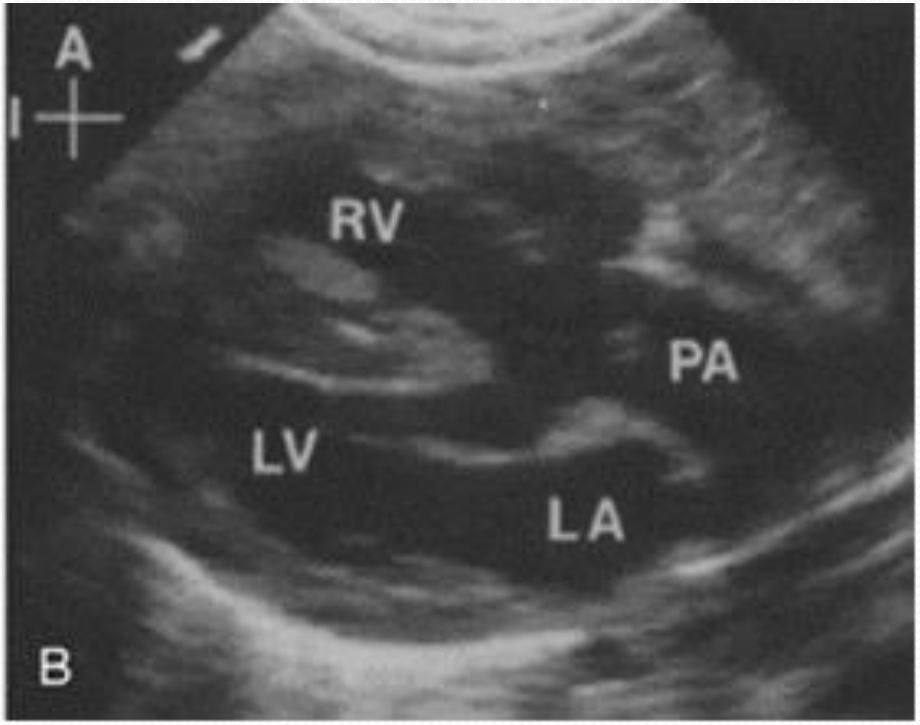
Manifestari clinice

- Fara stenoza pulmonare

-evolutie tip DSV larg nerestrictiv → ICC

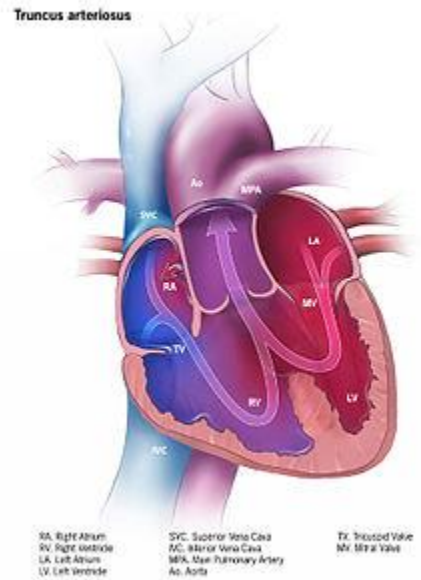
- Cu stenoza pulmonara
- -evolutie tip Tetralogie Fallot


A
I
RV
AO
LV
LA
A


A
I
RV
PA
LV
LA
B

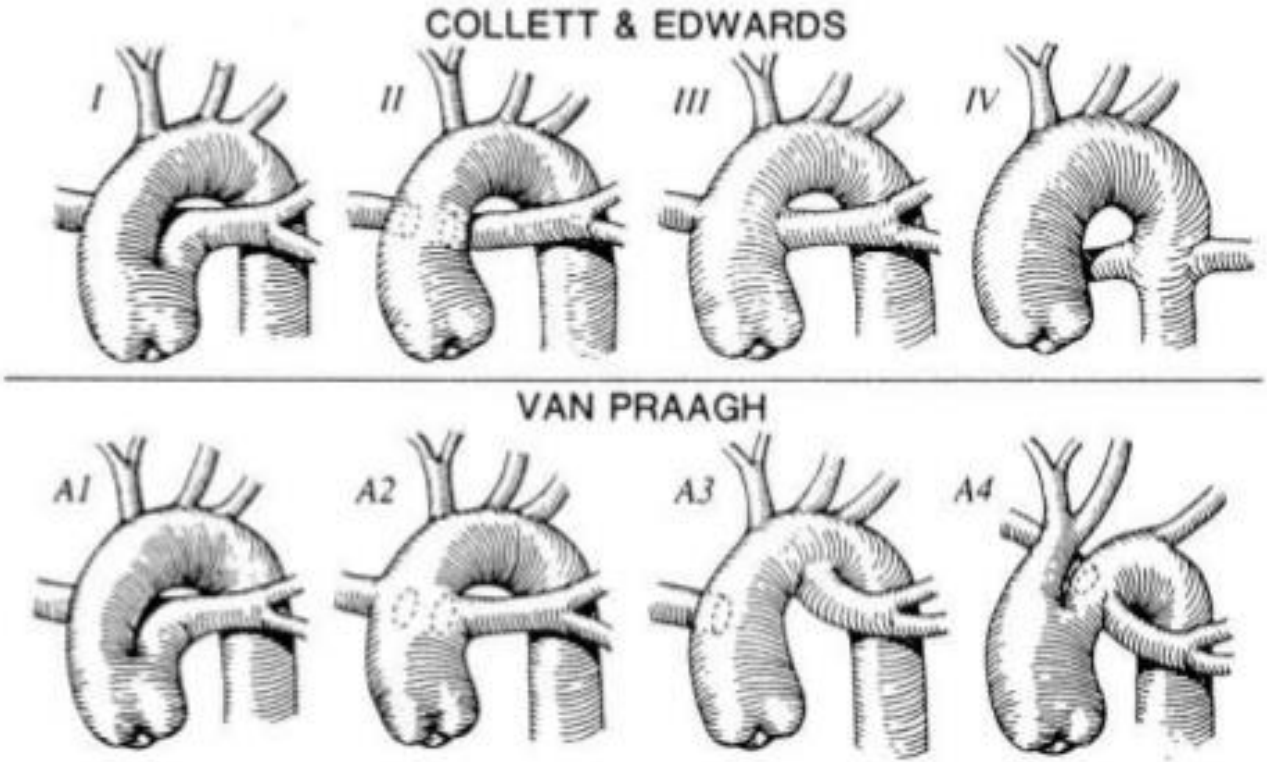

FR 55Hz
13cm
2D
72%
C 50
P Low
HGen
M3
G
P
R
1.7
3.4
JPEG
*** bpm

TRUNCHI ARTERIAL COMUN



- <1 %
- -exista un singur trunchi arterial ce furnizeaza debit pentru circulatia sistemica,pulmonara,coronariana
- DSV larg
- Valva truncala: bicuspa,tricuspa,quadricuspa,de cele mai multe ori cu grad de incompetenta
- F rar stenotica
- Asocierea cu Sd Di George 30%

Classification & Anatomy



- Frecvent tipul I si II cu flux crescut pulmonar,evolutie asemanatoare cu transpozitia de mari vase

- Tipul III si IV frecvent cu debit scazut pulmonar-evolutie asemanatoare T.Fallot

Manifestari clinice

- Cianoza care poate apare imediat dupa nastere
- ICC la zile sau saptamani dupa nastere
- Istoric de dispnee la alimentare,curba ponderala deficitara,infectii respiratorii frecvente

Evaluare clinica

- Ex fizica:

-tahicardie,tahipnee

-Suflu sistolic,suflu diastolic daca exista incompetenta de valva truncala

- Rx

-Cardiomegalie

-Cresterea vascularizatiei pulmonare/scaderea vascularizatiei pulmonare

- ECG

-HBV


15cm
2D
73%
C 50
P Low
HGen
G
P
R
1.7
3.4
JPEG
*** bpm


B27°/V40°
HR 141
SRI II 4
STIC
1 D 0.77cm


DGW 1
Har-high
Pwr 89 %
On -4
C0 / M0
P1 / E3

Management

- Medical

-trat ICC

- Chirurgical

-abordare biventriculara

-mortalitate <10%

-varsta optima<3l

Operative technique for type I truncus arteriosus.

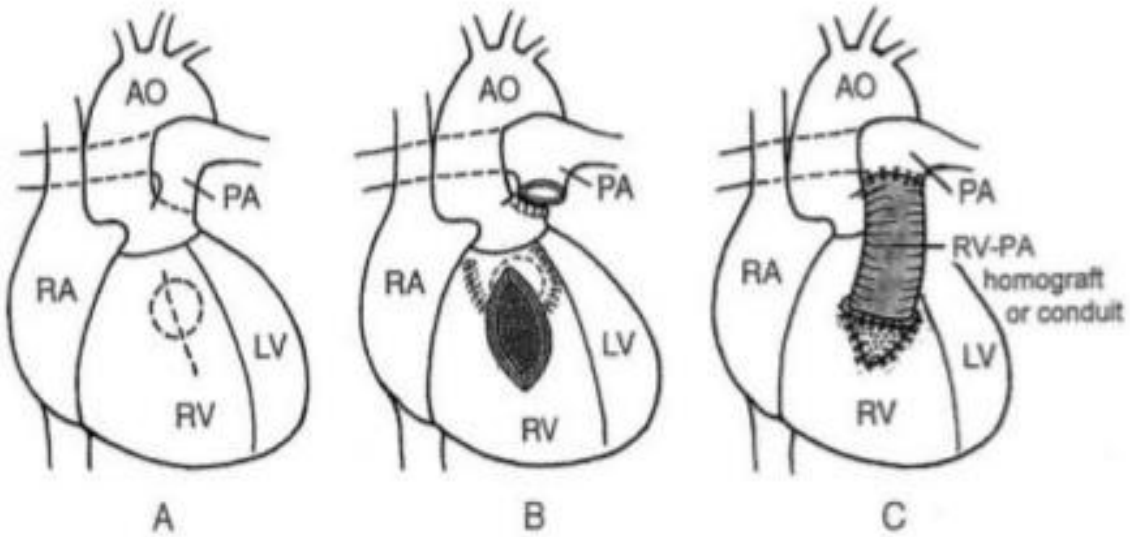
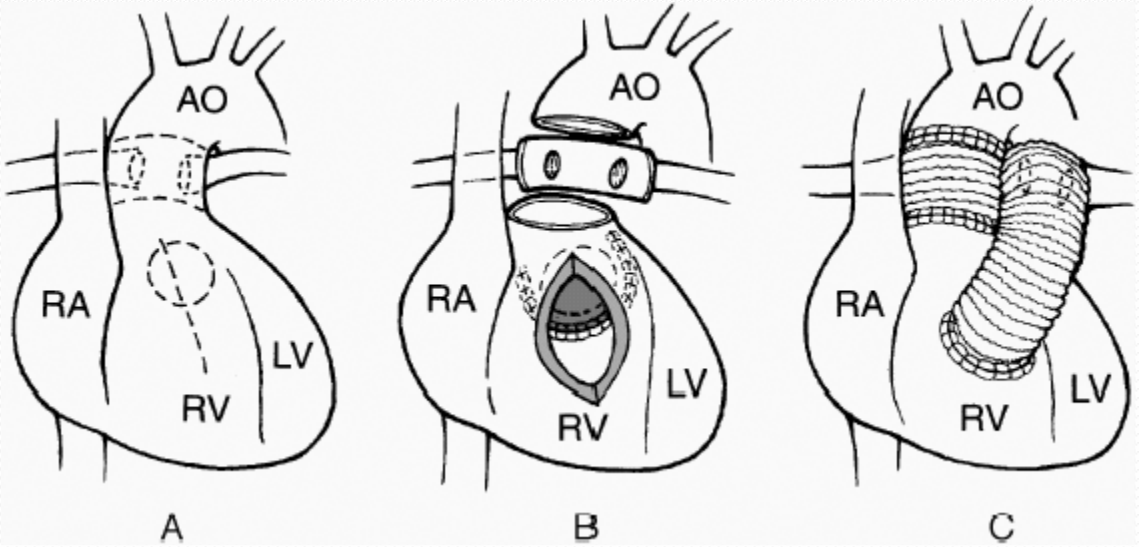
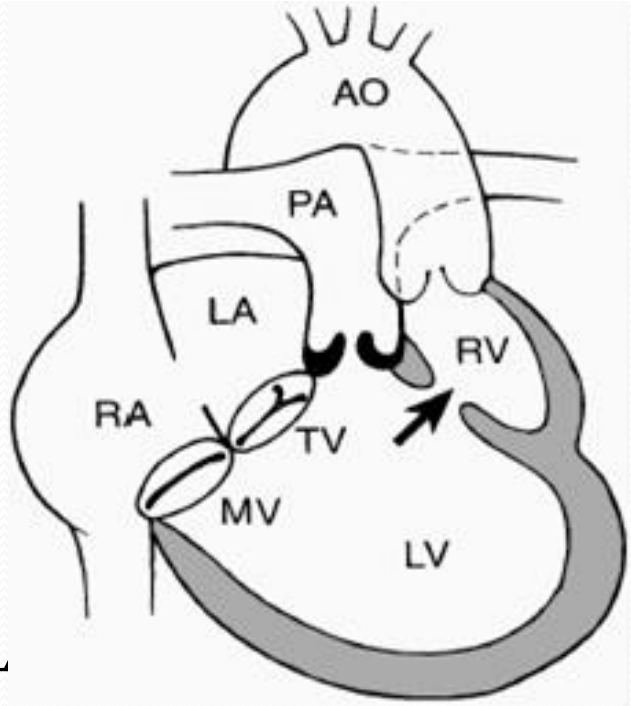


Figure 14-59 Operative technique for type I truncus arteriosus. **A,** Truncus arteriosus type I is shown with a large ventricular septal defect (VSD; broken circle) directly under the truncal valve. The vertical broken line on the right ventricle (RV) is the site of the right ventriculotomy. **B,** The pulmonary artery (PA) trunk has been cut away from the truncal artery, and the opening in the truncal artery is sutured to the truncal artery. Patch closure of the VSD (which is visible through the ventriculotomy) is completed in such a way that only left ventricular blood goes out to the truncal artery (creating the left ventricle [LV]–to–truncal artery pathway). **C,** A valved conduit or homograft is anastomosed to the pulmonary trunk. The posterior half of the proximal conduit is anastomosed to the upper end of the ventriculotomy. A small pericardial patch is trimmed and sutured into place to fill the defect between the allograft and the lower end of the right ventriculotomy. AO, aorta; RA, right atrium.


AO
RA
LV
RV
A
AO
RA
LV
RV
B
AO
RA
LV
RV
C

Dubla cale de intrare VS

- <1%
- -ambele VAV sunt conectate la un ventricul predominant ,de obicei VS (80%)
- -exista un ventricul rudimentar
- -un vas arterial are originea in ventriculul predominant,celalalt in ventriculul rudimentar
- -D-TGA sau L-TGA in 85% din cazuri
- **-forma cea mai frecventa : DCIVS/L TGA ,aorta cu origine din ventriculul rudimentar**
- 50% din cazuri ,stenoza pulmonara
- Frecvent asociere cu CoAo sau intrerupere de arc aortic

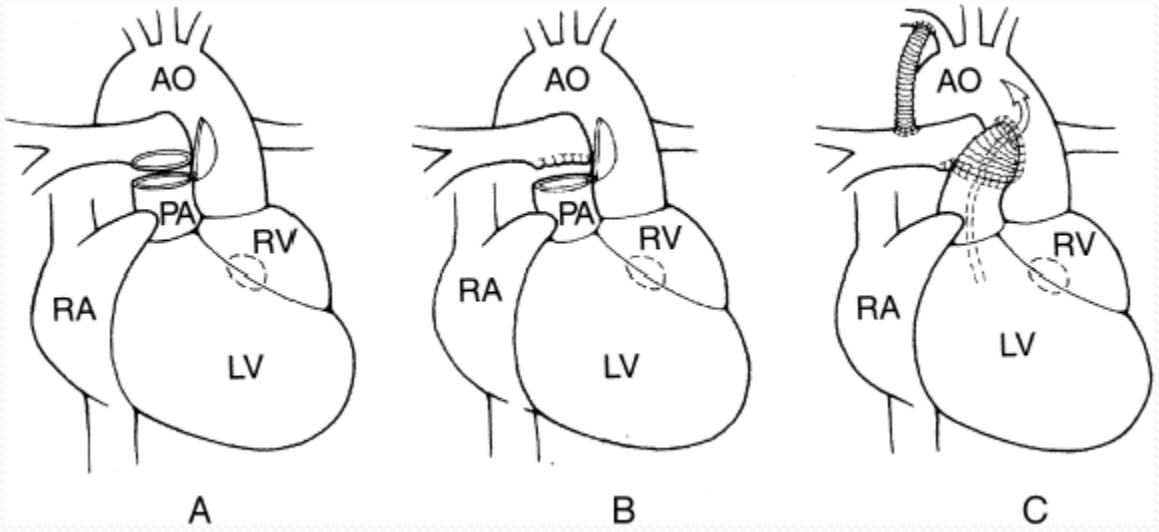



P
G
P
R
1,9
3,8

- Evolutia depinde de fluxul pulmonar
- Flux pulmonar crescut
- -evolutia asemanatoare TMV sau DSV larg
- Flux pulmonar scazut
- -evolutie aemanatoare TF

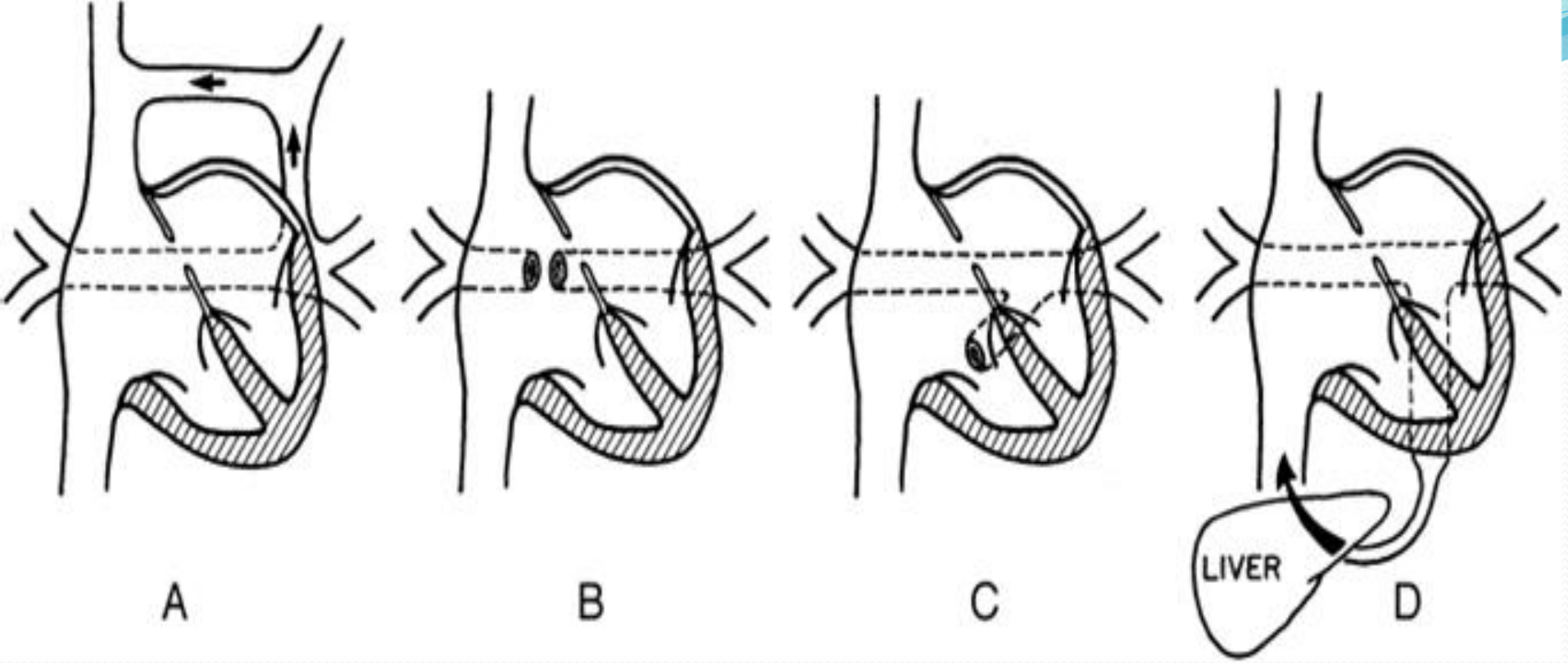
Management

- Medical
- -trat ICC
- -mentinerea CAP deschis
- Chirurgical
- -proceduri paleative catre procedura Fontan



DRENAJ VENOS PULMONAR TOTAL ABERANT

- 1% din MCC
- Nu exista comunicare directa intre venele pulmonare si AS;venele pulmonare se varsa in sistemul venos tributar AD sau direct AD
- Exista DSA
- Cavitatile stangi sunt reduse de volum
- Poate exista obstructie in intoarcerea venoasa asociata cu HTP (frecvent cele infracardiace)



- Supracardiac 50%

-dreneaza in SVC printr-o vena verical si vena inominata stg

- Cardiac 20 %: fie in sinusul coronar sau direct in AD
- Infracardiac 20% : in VCI,vena hepatica,vena porta
- Mixt 10 %

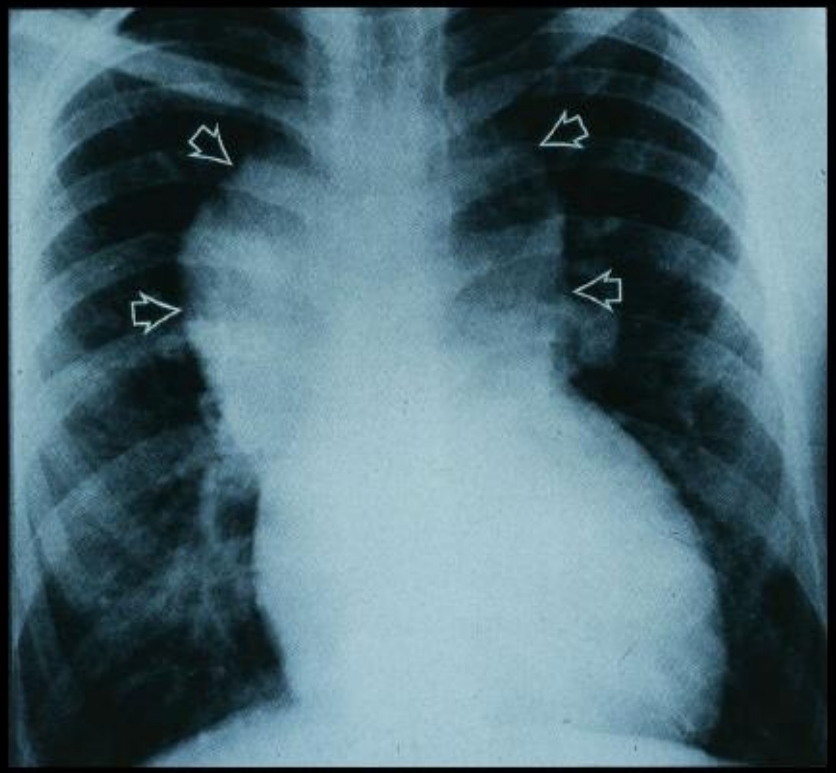
Manifestari clinice

- Depind de prezenta sau absenta obstructiei in intoarcerea venoasa
- Fara obstructie

-ICC(tahicardie,tahipnee,dispnee,hepatomegalie)

-cianoza usoara

Supracardiac TAPVC - CXR



"Snowman" appearance secondary to dilated vertical vein, innominate vein and right superior vena cava draining all the pulmonary venous blood

- ECG

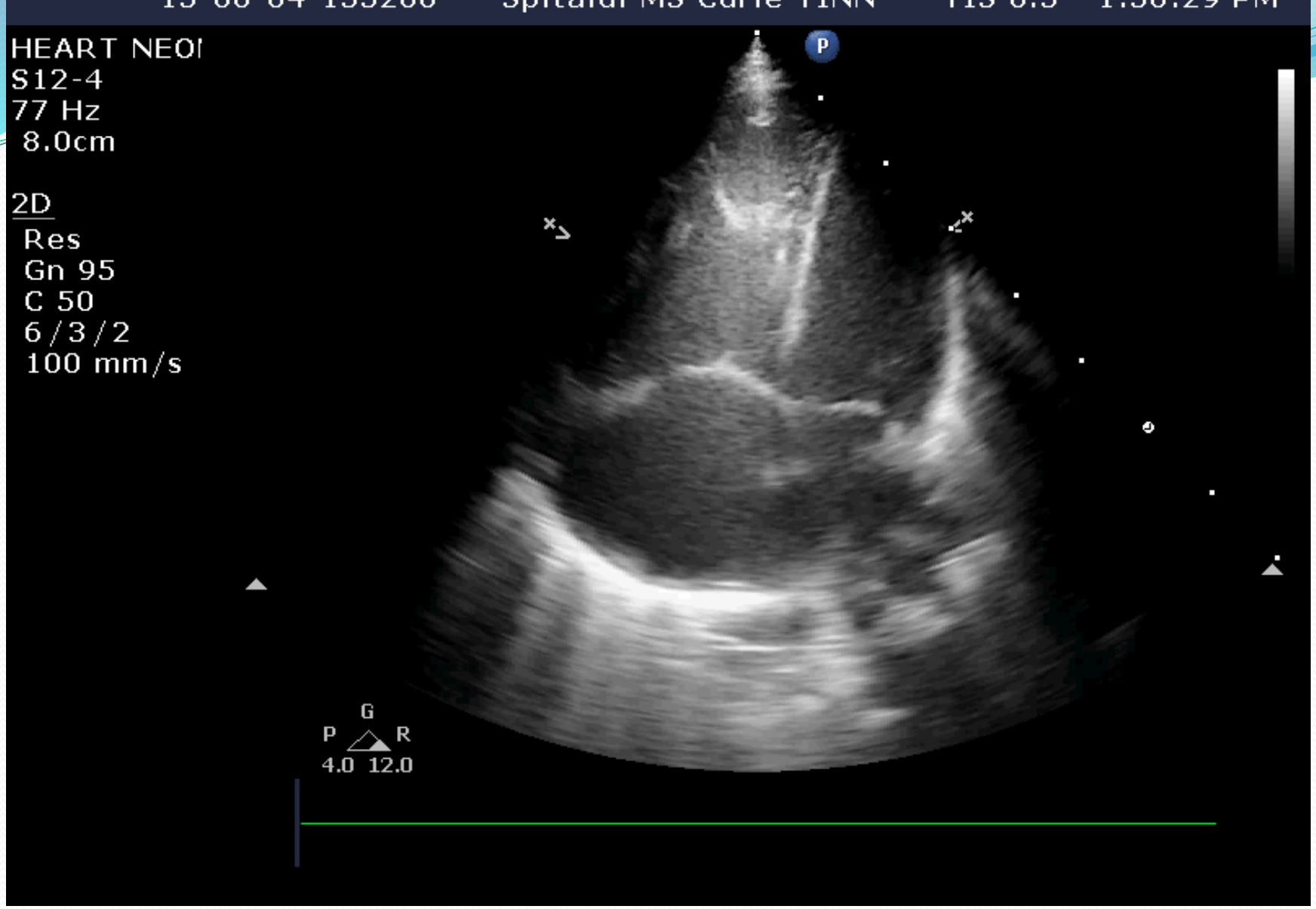
-semne de HVD ,rsR in V1

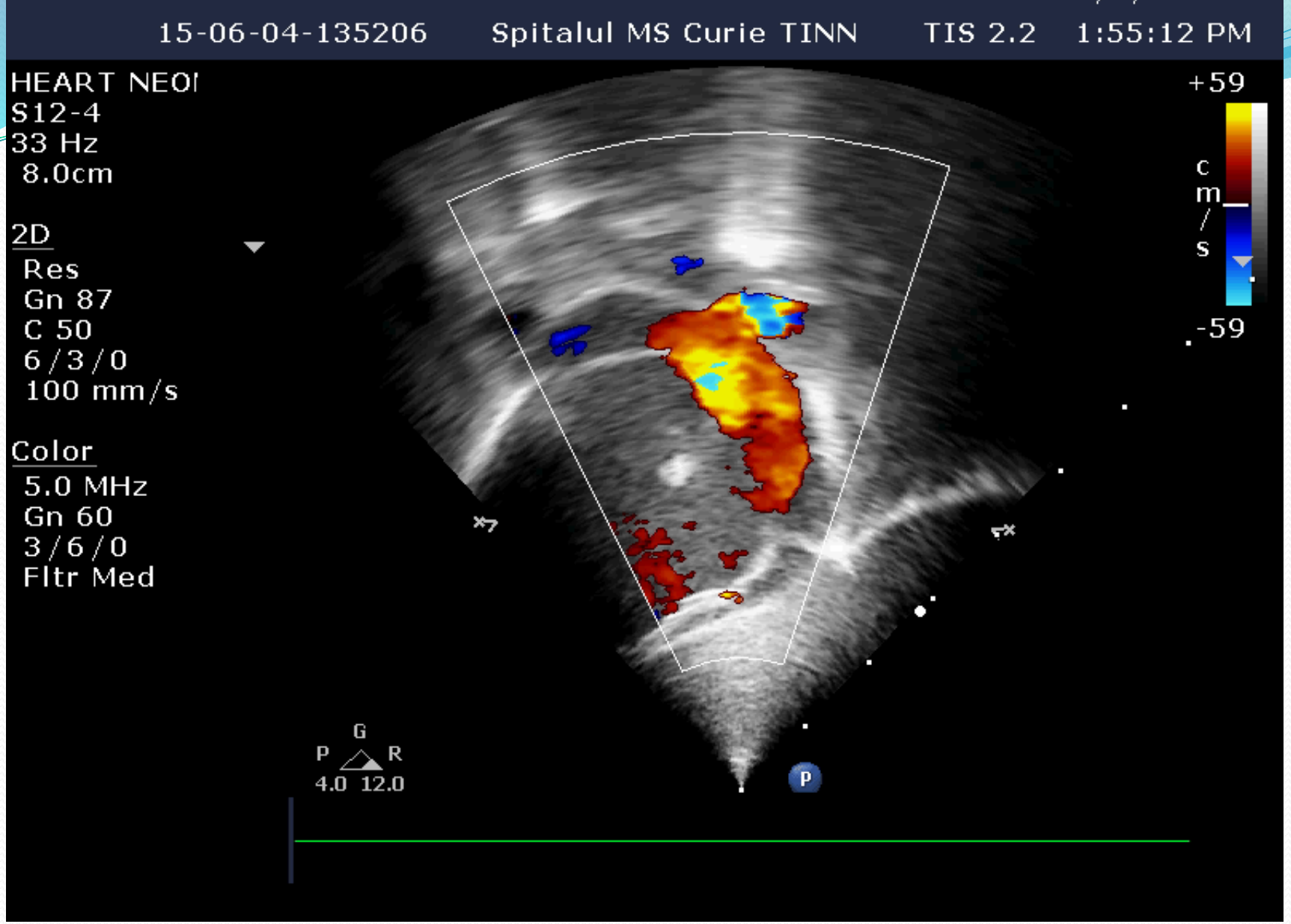
Manifestari clinice

- Cu obstructie in intoarcerea venoasa pulmonara

-cianoza severa

-stress respirator sever


HEART NEOI
S12-4
77 Hz
8.0cm
2D
Res
Gn 95
C 50
6/3/2
100 mm/s
P
G
P
R
4.0
12.0


15-06-04-135206 Spitalul MS Curie TINN TIS 2.2 1:55:12 PM
HEART NEO
S12-4
33 Hz
8.0cm
2D
Res
Gn 87
C 50
6/3/0
100 mm/s
Color
5.0 MHz
Gn 60
3/6/0
Fltr Med
+59
cm/s
-59
G
P
R
4.0 12.0
P

Management

- Medical

-tratamentul ICC;diuretic,suport inotrop

-trat acidozei metabolice

-inducerea unei respiratii alcalotice care sa scada RVP

-PGE1?

- Chirurgical

➢ timing:

-cu obstructie: cat mai repede ,chiar in perioada neonatala

-fara obstructie: 4-6 l,in functie de evolutia DSA

➢ Mortalitate 5 %

Complicatii :

- Criza de HTP
- Obstructie la locul anastomozei sau pe venele pulmonare
- Aritmii frecvent supraventriculare