FONDAZIONE CENTRO CARDIOLOGIA
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Le cardiopatie congenite complesse dell'adulto con ostruzione all'efflusso sinistro e destro. Come gestirle e cosa fare

Gabriele Vignati

6° Convengo di Cardiorisonanza

Milano 17 ottobre 2012

Cosa s'intende per cardiopatia congenita (CC) complessa?

Cardiopatia nella quale coesistono più anomalie strutturali.

Il 45% delle CC hanno moderata complessità e il 15% elevata complessità.

Peculiarità pz adulto con CC complessa

- Rari i pz con cardiopatia in storia naturale:
  - -non indicazione chirurgica
  - -cardiopatie misconosciute (immigrazione)

- Pz con cardiopatia operata:
  - -follow-up irregolari
  - -frequenti residui, sequele complicanze
  - -poco nota prognosi a lughissimo termine
  - -ripercussioni sulla funzione ventricolare

Caratteristiche CC complesse con ostruzione agli efflussi

-CC in storia naturale: relativamente rare (prevalente ostruzione efflusso sistemico)

-CC operate: forme più comuni (prevalente ostruzione efflusso polmonare)

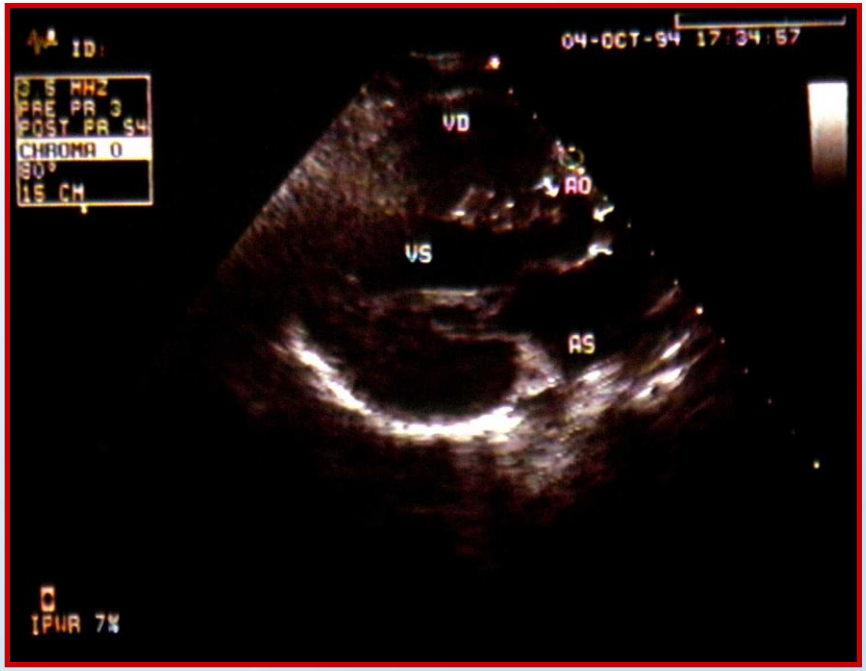
Cardiopatie con ostruzione efflusso sinistro

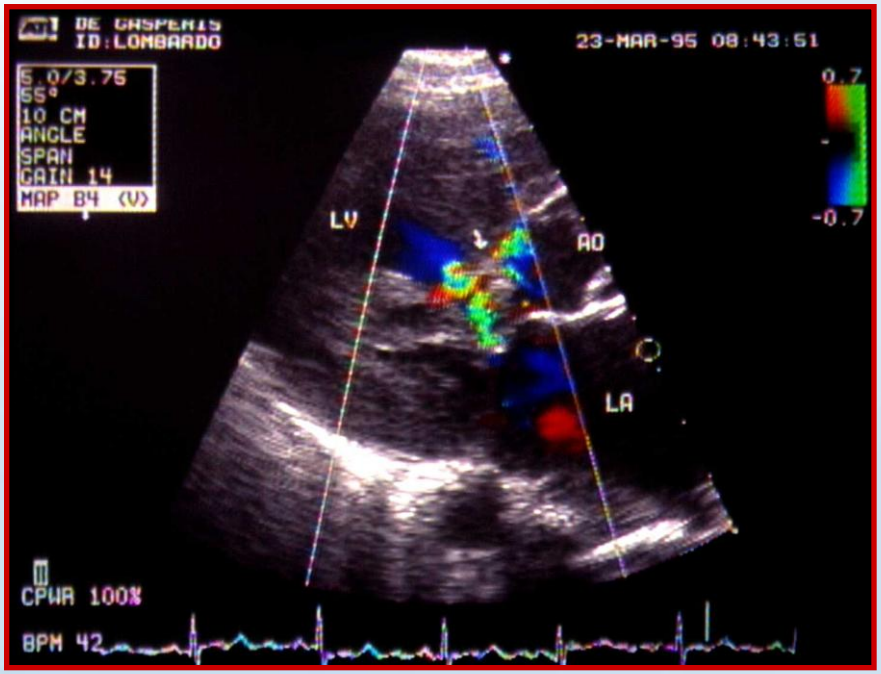
In storia naturale

- stenosi sottovalvolare aortica a tunnel
- stenosi valvolare con ipoplasia anulare
- ipoplasia aorta ascendete/arco
- forme complesse di coartazione

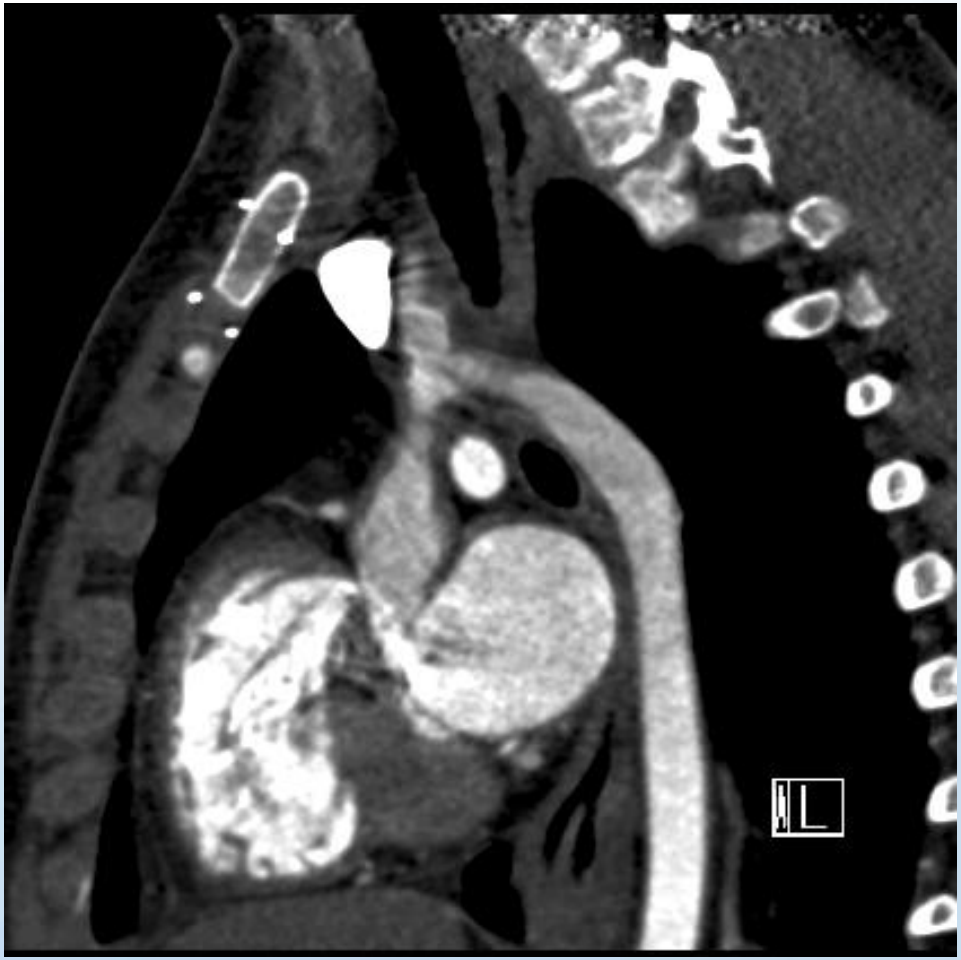
Patologie efflusso sinistro

Stenosi sottovalvolare aortica a tunnel





Ipoplasia aorta ascendente e arco



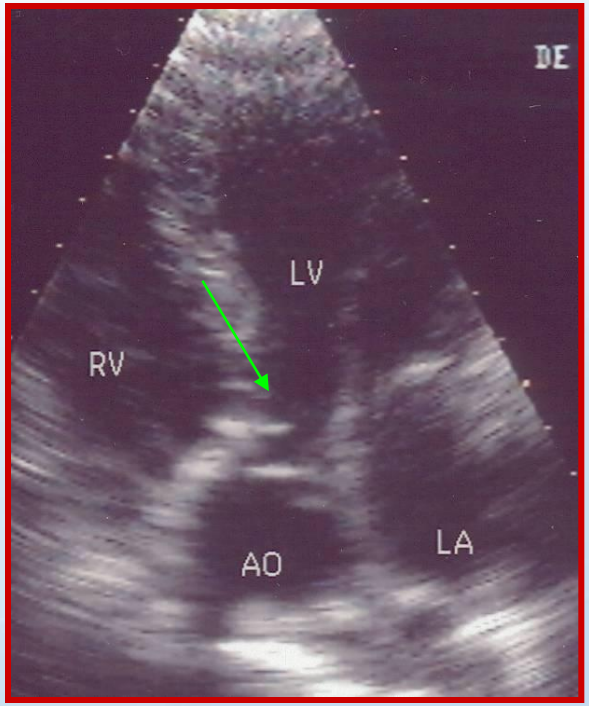
Cardiopatie con ostruzione efflusso sinistro

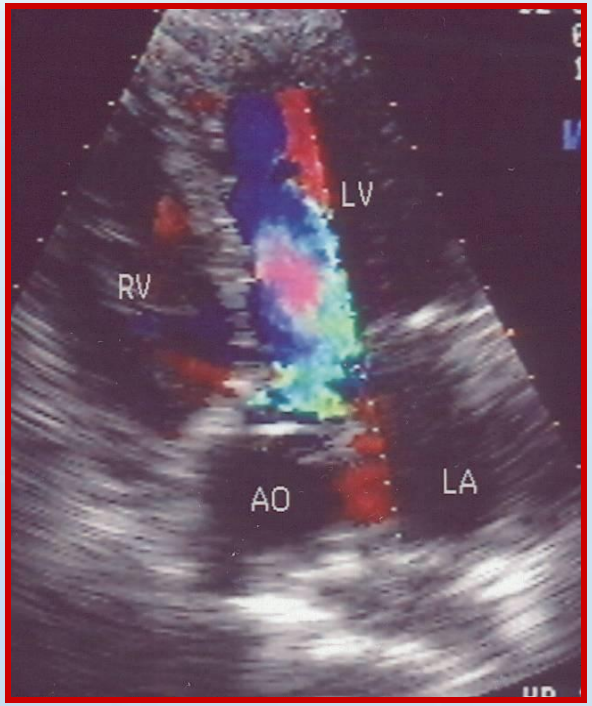
Forme post-operatorie

- recidive stenosi sottovalvolare
- stenosi sottovalvolare in cuore univentricolare
- recidive stenosi sopravalvolare aortica
- malfunzionamenti protesi/autograft valvolari
- ricoartazione aortica

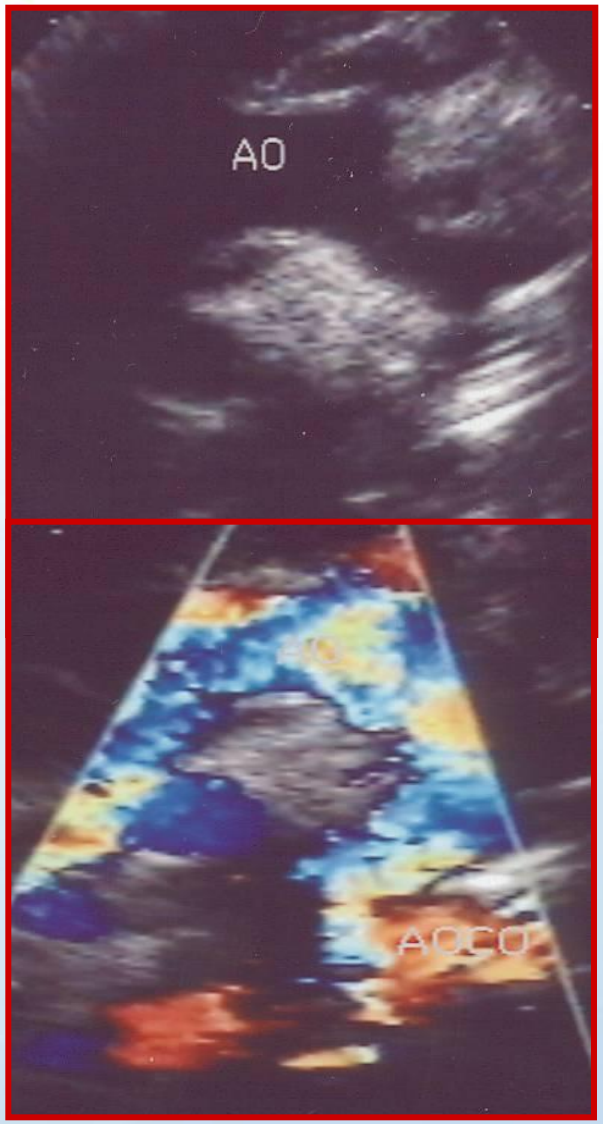
Stenosi sottovalvolare aortica

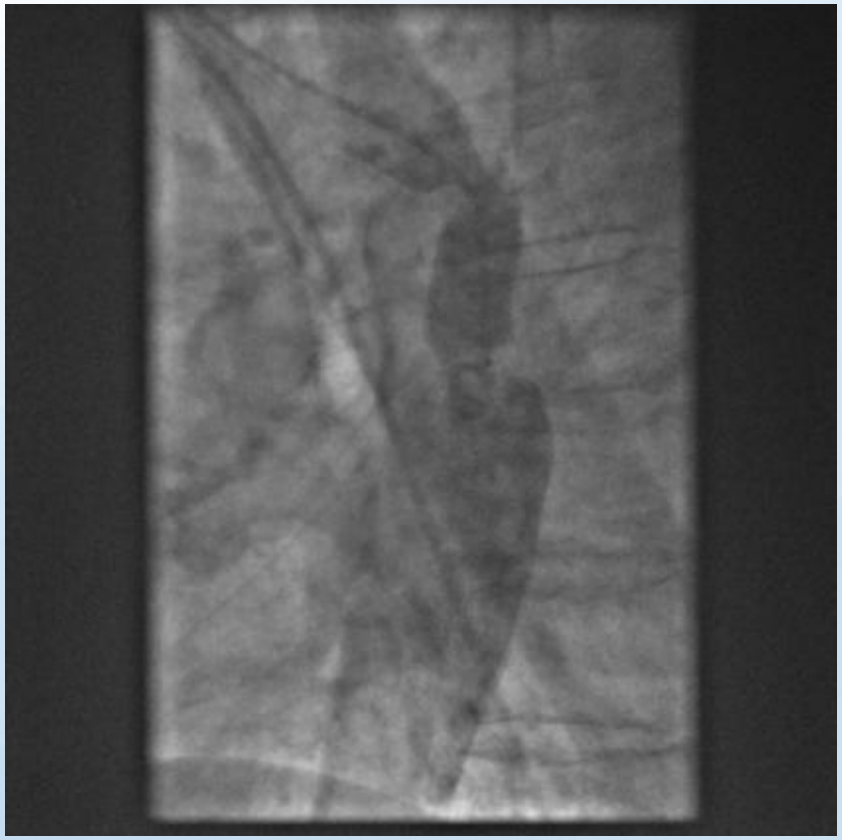
- Recidive non rare
- Può comparire dopo correzione CAV e DIV





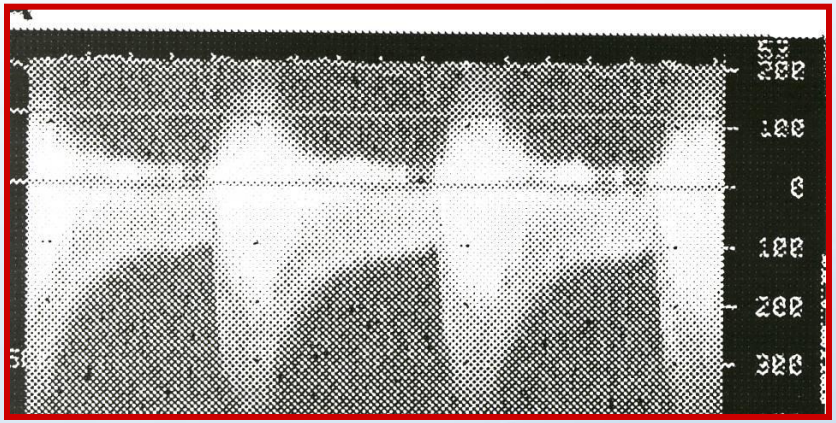
Ricoartazione Aortica



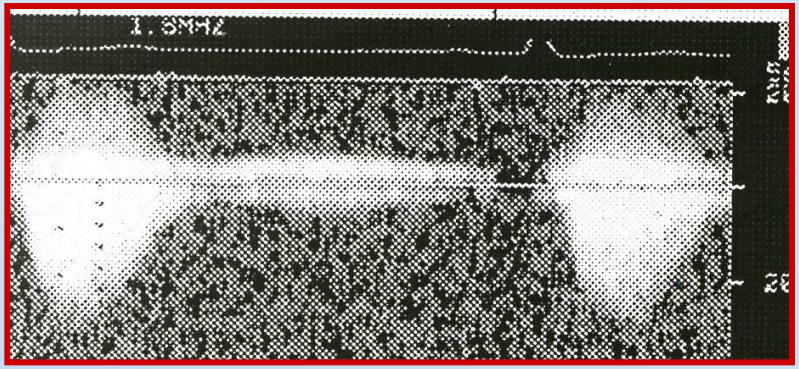


Gradienti residui dopo correzione CoAo

Ricoartazione



Gradiente “fisiologico”



Sintomatologia

- Comparsa spesso tardiva dei sintomi
- Ostruzioni valvolari, sotto e sopravalvolari
  - -dispnea, angina, sincopi, scompenso
- Coartazione, ricoartazione
  - -ipertensione arteriosa, dispnea, claudicatio
    scompenso

Indications for Intervention in Valvular Aortic Stenosis (1)

	Class[a]	Level[b]
• Patients with severe AS and any valve related symptoms (AP, dyspnoea, syncope) should undergo valve replacement.	I	B
• Asymptomatic patients with severe AS should undergo surgery when they develop symptoms during exercise testing.	I	C
• Regardless of symptoms, surgery should be performed when systolic LV dysfunction is present in severe AS (LVEF < 50%), unless it is due to other causes.	I	C
• Regardless of symptoms, surgery should be performed when patients with severe AS undergo surgery of the ascending aorta or of another valve or coronary artery bypass grafting.	I	C
• Regardless of symptoms, aortic surgery should be considered if the ascending aorta is greater than 50 mm (27.5 mm/m^2 BSA) and no other indications for cardiac surgery are present.	IIa	C
• Asymptomatic patients with severe AS should be considered for surgery when they present with a fall in blood pressure below baseline during exercice testing.	IIa	C

a = class of recommendation. b = level of evidence.
AP = angor pectoris; AS = aortic stenosis; BSA = body surface area; LV = left ventricle; LVEF = ventricular ejection fraction.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Indications for Intervention in Valvular Aortic Stenosis (2)

	Class[a]	Level[b]
• Asymptomatic patients with severe AS and moderate-to-severe calcification and a rate of peak velocity progression of ≥ 0.3 m/sec/year should be considered for surgery.	IIa	C
• Patients with moderate AS undergoing coronary artery bypass surgery or surgery of the ascending aorta or another valve should be considered for additional valve replacement.	IIa	C
• Severe AS with low gradient (< 40 mmHg) and LV dysfunction with contractile reserve should be considered for surgery.	IIa	C
• Severe AS with low gradient (< 40 mmHg) and LV dysfunction without contractile reserve may be considered for surgery.	IIb	C
• Asymptomatic patients with severe AS and excessive LV hypertrophy (≥ 15 mm), unless this is due to hypertension, may be considered for surgery.	IIb	C

a = class of recommendation. b = level of evidence.
AP = angor pectoris; AS = aortic stenosis; BSA = body surface area; LV = left ventricle; LVEF = ventricular ejection fraction.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Indications for Intervention in Supravalvular Aortic Stenosis

	Class[a]	Level[b]
• Patients with symptoms (spontaneous or on exercise test) and mean Doppler gradient ≥ 50 mmHg should undergo surgery.	I	C
• Patients with mean Doppler gradient < 50 mmHg should undergo surgery when they have:		
– symptoms attributable to obstruction (exertional dyspnoea, angina, syncope) and/or,	I	C
– LV systolic dysfunction (without other explanation),	I	C
– severe LVH, attributable to obstruction (not related to hypertension),	I	C
– when surgery for significant CAD is required.	I	C
• Patients with mean Doppler gradient ≥ 50 mmHg* but without symptoms, LV systolic dysfunction, LVH or abnormal exercise test may be considered for repair when the surgical risk is low.	IIb	C

a = class of recommendation. b = level of evidence.
*Doppler derived gradients may overestimate the obstruction and may need confirmation by left heart catheterization.
CAD = coronary artery disease; LV = left ventricle; LVH = ventricular hypertrophy.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Indications for Intervention in Coarctation of the Aorta

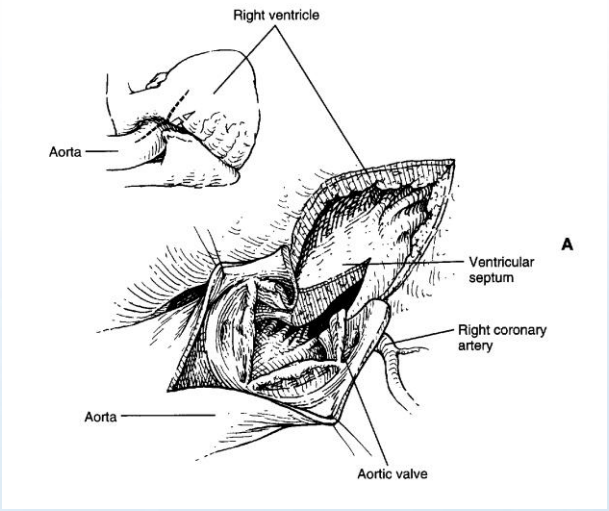
- All patients with a non-invasive pressure difference > 20 mmHg between upper and lower limbs, regardless of symptoms but with upper limb hypertension (> 140/90 mmHg in adults), pathologic blood pressure response during exercice, or significant LVH should have intervention.
- Independent of the pressure gradient, hypertensive patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) should be considered for intervention.
- Independent of the pressure gradient and presence of hypertension, patients with ≥ 50% aortic narrowing relative to the aortic diameter at the diaphragm level (on CMR, CT or invasive angiography) may be considered for intervention.

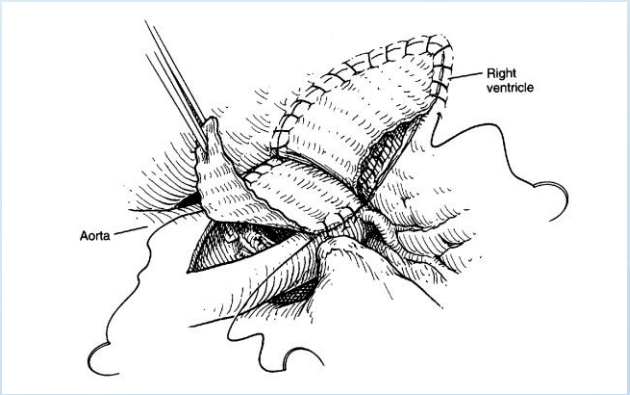
Class[a]	Level[b]
I	C
IIa	C
IIb	C

a = class of recommendation. b = level of evidence.
CMR = cardiac magnetic resonance; CoA = coarctation of the aorta; CT = computed tomography; LVH = left ventricular hypertrophy.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

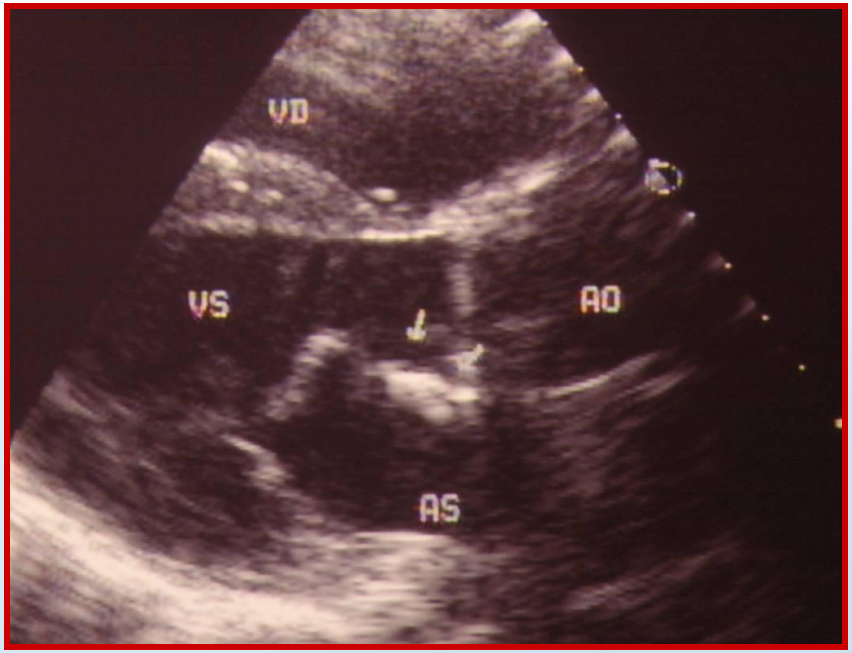
Intervento di Konno



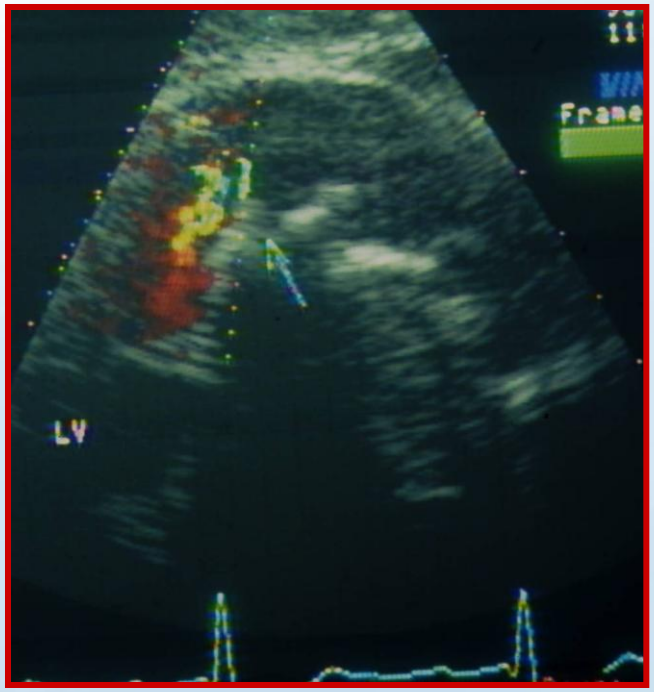


- Correzione stenosi a tunnel
- Correzione pat.valvola Ao con ipoplasia anulare
- Valvola Ao può essere conservata

Correzioni atipiche efflusso sin.

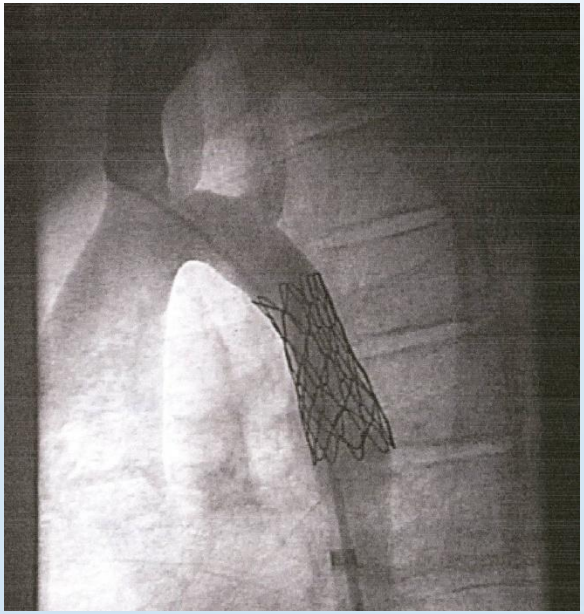


Ampliamento anulus sul lembo anteriore mitrale



Condotto apico-aortico

Ampliamento con stent di ricoartazione



Trattamento percutaneo terapia di prima scelta:

- **-bassa invasività**
- **-rischi contenuti (stent ricoperti)**
- **-non utilizzabile se segmenti molto angolati o correzione con condotti di piccolo calibro**

Trattamento chirurgico ricoartazione

-Rischi maggiori

-Necessità di cec

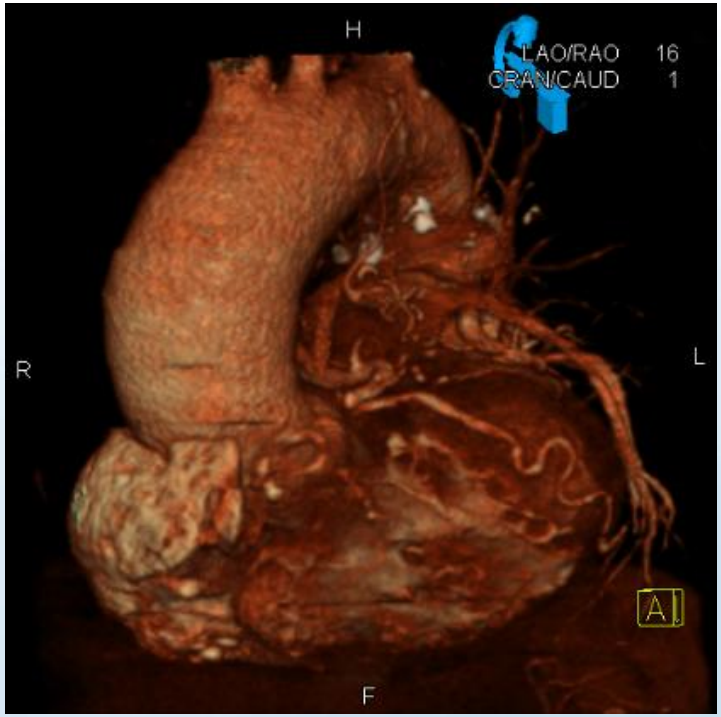
-Impiego condotti extranatomici

Cardiopatie con ostruzione efflusso destro

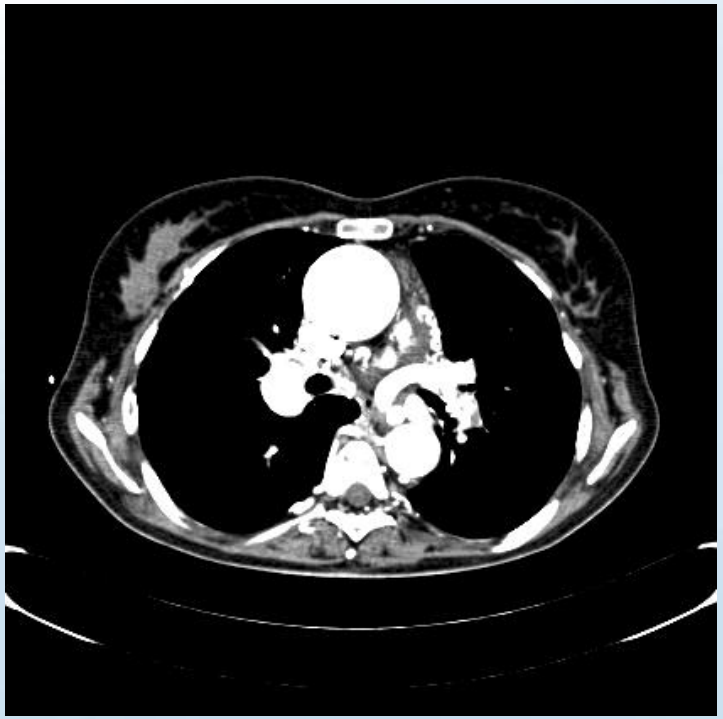
In storia naturale

- T.Fallot
- Atresia polmonare con DIV e collaterali
- TCGA/TGA con DIV e SPo
- Cuore univentricolare con SPo

Atresia polmonare con perfusione multifocale



Rami polmonari ipoplasici confluenti deconnessi dal VD



Collaterale a perfondere polmone dx

Cardiopatie con ostruzione efflusso destro

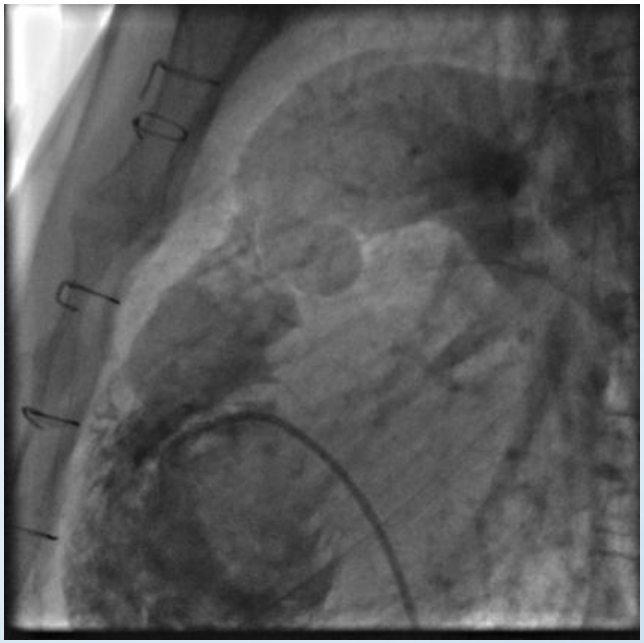
Forme post-operatorie

- cardiopatie corrette con condotto VD-AP
  - atresia PO con DIV, Truncus, TGA con DIV e Spo

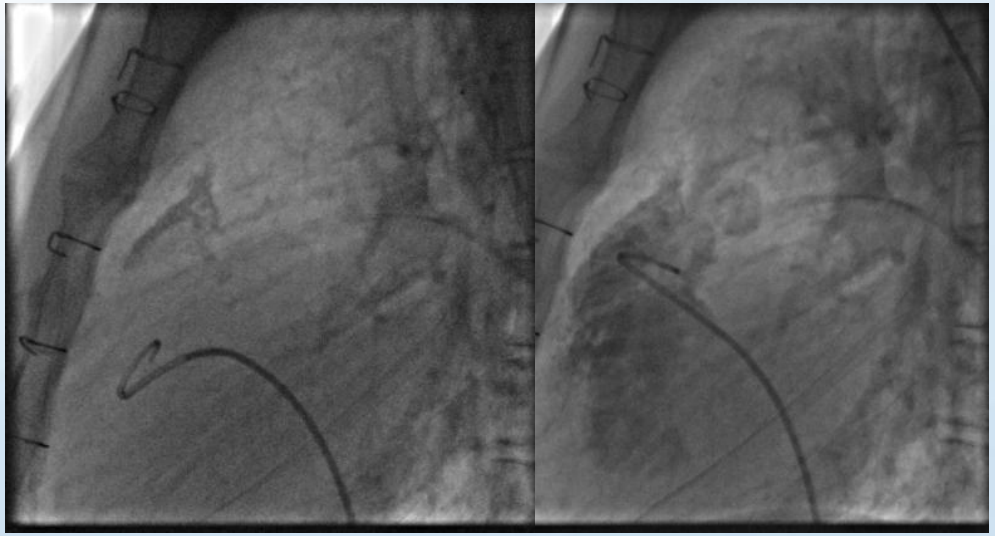
- cardiopatie con patologie rami polmonari
  - T.Fallot, Atresia Po con DIV, TGA corretta con switch

- malfunzionamenti protesi/autograft valvolari

Ostruzioni efflusso destro



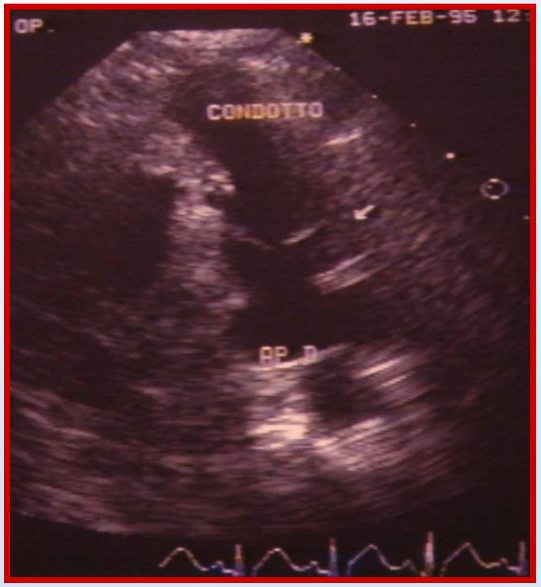
Esiti correzione T.Fallot
Stenosi infundibolare residua

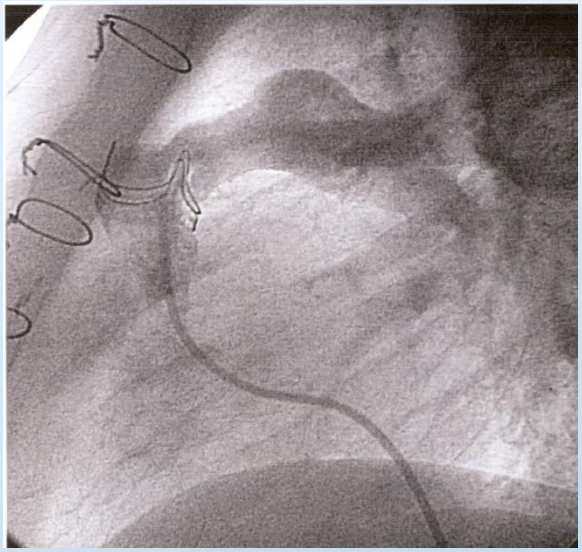


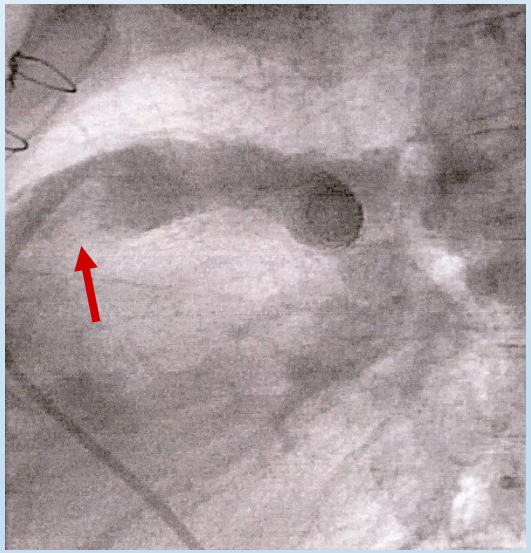
Esiti correzione T.Fallot
Calcificazioni patch e stenosi residua

Patologie condotti

A distanza di 20 anni più del 60% dei condotti sono mal funzionanti per degenerazione valvolare e/o per ostruzione da proliferazione intimale e da lesioni calcifiche



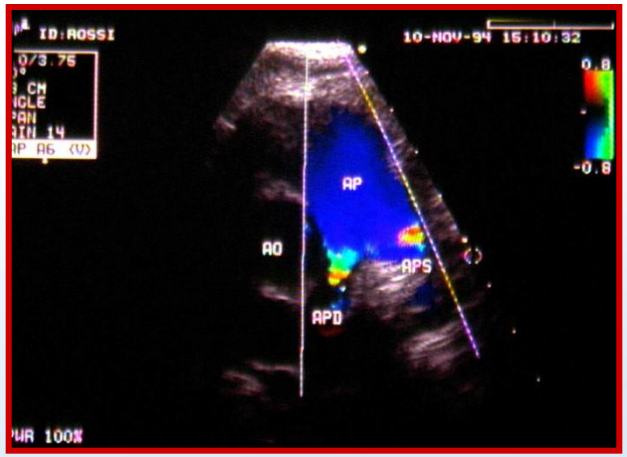




Sovraccarichi volume e/o pressione

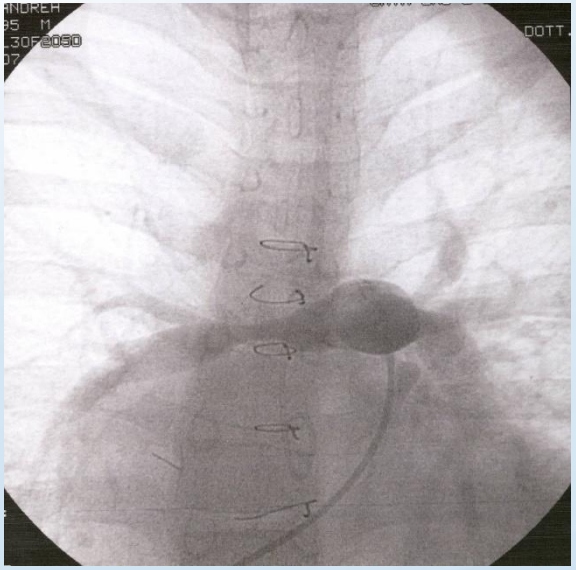
Disfunzione ventricolare (scompenso destro)

Patologie dei rami polmonari



T.Fallot operata
Stenosi biforcazione

Esiti switch per TGA
Ipoplasia AP sinistra



T.Fallot operata
Stenosi AP destra

Conseguenze patologie dei rami polmonari

- Missmatch ventilazione/perfusione
- Ipertensione sul polmone controlaterale
- Sovraccarico pressione VD

Indications for Intervention in Right Ventricular Outflow Tract Obstruction

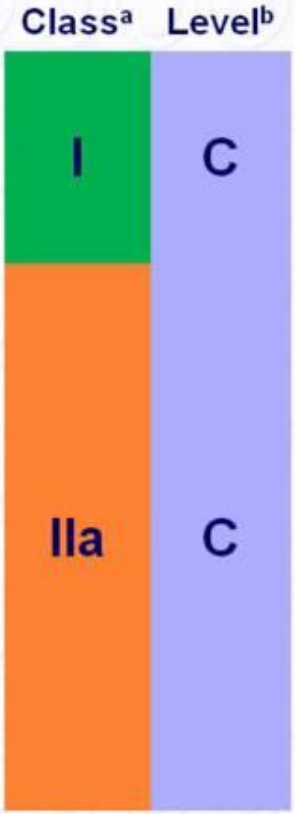
	Class[a]	Level[b]
• RVOTO at any level should be repaired regardless of symptoms when Doppler peak gradient is > 64 mmHg (peak velocity > 4 m/s), provided that RV function is normal and no valve substitute is required.	I	C
• In valvular PS, balloon valvotomy should be the intervention of choice.	I	C
• 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/sec).	I	C
• Intervention in patients with gradient < 64 mmHg should be considered in the presence of 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

a = class of recommendation. b = level 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.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

Indications for Intervention in Patients With Right Ventricular to Pulmonary Artery Conduits

- Symptomatic patients with RV systolic pressure > 60 mmHg (TR velocity > 3.5 m/sec; may be lower in case of reduced flow) and/or moderate/severe PR should undergo surgery.
- Asymptomatic patients with severe RVOTO and/or severe PR should be considered for surgery when at least one of the following criteria is present:
  - decrease in exercise capacity (CPET),
  - progressive RV dilation,
  - progressive RV systolic dysfunction,
  - progressive TR (at least moderate),
  - RV systolic pressure > 80 mmHg (TR velocity > 4.3 m/sec),
  - sustained atrial/ventricular arrhythmias.



a = class of recommendation. b = level of evidence.
CPET = cardiopulmonary exercise testing; PR = pulmonary regurgitation; RV = right ventricle; RVOTO = right ventricular outflow tract obstruction; TR = tricuspid regurgitation.

European Heart Journal 2010; doi:10.1093/eurhearj/ehq249

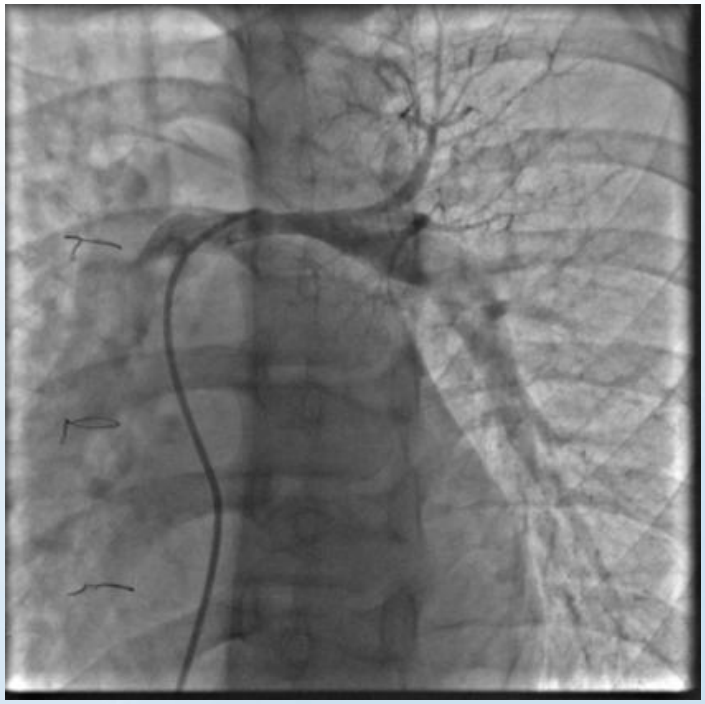
Trattamento ostruzioni efflusso dx

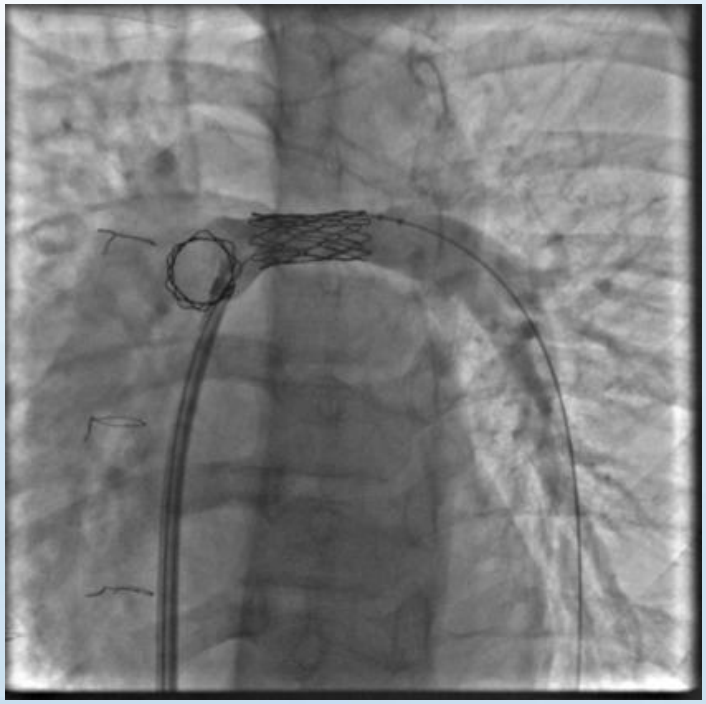
- Patologie condotti
  - -sostituzione chirurgica
  - -impianto protesi percutanea

- Patologia efflusso nativo
  - -ampliamento con patch
  - -recentazione fasci muscolari

- Patologia rami polmonari
  - -ampliamento con stent
  - -ampliamento chirurgico

Trattamento ostruzioni efflusso dx





Considerazioni conclusive

- Le cardiopatie note in storia naturale richiedono regolari controlli ma anche rivalutazione ai fini di ridiscutere eventuali opzioni chirurgiche

- Le cardiopatie complesse operate richiedono regolari controlli per identificare eventuali anomalie e decidere le terapie più appropriate

- La gestione di questi pazienti deve essere fatta da personale con specifiche competenze nell'ambito delle cardiopatie congenite

Grazie per l'attenzione

Dipartimento Cardiologico A. De Gasperis
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