VI Congresso Nazionale di Ecocardiochirurgia

MILANO, 15-17/10/2012

Le cardiopatie nell'adulto con shunt destro e sinistro. E' tutto così semplice?

Adele Borghi

Cardiologia pediatrica e delle cardiopatie congenite

Ospedale dei Bambini

Spedali Civili - Brescia

Milano, 17/10/2012

LO SHUNT

- In condizioni normali il circolo sistemico e polmonare sono completamente separati ed in serie
- La presenza di una comunicazione anomala, a qualsiasi livello, comporta uno shunt
- Lo shunt può essere
  - sinistro-destro
  - bilanciato o bidirezionale
  - destro sinistro

  secondo la direzione del flusso tra i due circuiti
- Uno shunt può essere isolato o essere parte di un complesso malformativo

FISIOPATOLOGIA

- Iperafflusso senza ipertensione polmonare (QP/QS>1 – PAP=)
- Iperafflusso con ipertensione polmonare (QP/QS>1 – PAP⬆)

  >>> (S.di Eisenmenger)
- Ipoafflusso polmonare con ipertensione polmonare (QP/QS<1 – PAP⬆)
- Ipoafflusso polmonare senza ipertensione polmonare (QP/QS<1 – PAP=)

ANATOMIA

- Atriale
  - PFO
  - DIA
  - RVAP
- Ventricolare
  - DIV
  - CAV
- Vascolare
  - Dotto arterioso
  - Fistole AV
  - Collaterali veno-venosi

Le Linee Guida ESC 2010

Table 1 Classes of recommendations

Classes of recommendations	Definition
Class I	Evidence and/or general agreement that a given treatment or procedure is beneficial, useful, effective.
Class II	Conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of the given treatment or procedure.
Class IIa	*Weight of evidence/opinion is in favour of usefulness/efficacy.*
Class IIb	*Usefulness/efficacy is less well established by evidence/opinion.*
Class III	Evidence or general agreement that the given treatment or procedure is not useful/effective, and in some cases may be harmful.

Table 2 Levels of evidence

Level of evidence A	Data derived from multiple randomized clinical trials or meta-analyses.
Level of evidence B	Data derived from a single randomized clinical trial or large non-randomized studies.
Level of evidence C	Consensus of opinion of the experts and/or small studies, retrospective studies, registries.

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PERVIETA' DEL FORAME OVALE

Variante fisiologica presente nel 20-25% della popolazione adulta

Profilassi delle recidive emboliche (ictus criptogenico) con terapia interventistica indicata in casi selezionati

Non indicata profilassi primaria

DIFETTO INTERATRIALE

- **Frequente in prima diagnosi**
- **Tipologia**
  - Ostium secundum 80%
  - Ostium primum 15%
  - Seno venoso c.sup. 5%
  - Seno venoso c.inf. <1%
  - Seno cor. fenestrato <1%

DIFETTO INTERATRIALE

- **Presentazione clinica e storia naturale**
  - Asintomatici fin oltre i 40 anni
  - Ridotta tolleranza allo sforzo
  - Aritmie
  - Scompenso destro
  - Infezioni polmonari
  - Embolia paradossa (rara)

DIFETTO INTERATRIALE

- **Evoluzione e complicanze**
  - Scompenso dx (tardivo)
  - Aritmie (SV)
  - Ipertensione polmonare (5%)
  - Embolia paradossa (rara)
- **Indicazioni terapeutiche**
  - Chiusura interventistica
  - Chiusura chirurgica

DIA

Table 3 Indications for intervention in atrial septal defect

Indications	Class[a]	Level[b]
Patients with significant shunt (signs of RV volume overload) and PVR <5 WU should undergo ASD closure regardless of symptoms	I	B[26]
Device closure is the method of choice for secundum ASD closure when applicable	I	C
All ASDs regardless of size in patients with suspicion of paradoxical embolism (exclusion of other causes) should be considered for intervention	IIa	C
Patients with PVR ≥5 WU but <2/3 SVR or PAP <2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L–R shunt (Qp:Qs >1.5) may be considered for intervention	IIb	C
ASD closure must be avoided in patients with Eisenmenger physiology	III	C

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DIFETTO INTERVENTRICOLARE

- **Tipologia**
  - Perimembranoso 80%
    - ✓ Tipo CAV
    - ✓ Malallineato
    - ✓ Esteso
  - Muscolare 15-20%
  - Tratto efflusso 5%
    - ✓ Sopracrestale
    - ✓ Sottoarterioso
  - Talora multipli
  - Spesso presenti nelle c. complesse
  - Postchirurgici

DIFETTO INTERVENTRICOLARE

- **Presentazione clinica e storia naturale**
  - Shunt insignificante, VS e PAP normali (m. di Roger)
  - Shunt significativo, VS poco dilatato, PAP normale o variamente aumentata (rari)
  - Ampi, PAP e RVP elevate (Eisenmenger)
  - Operati con shunt residuo

DIFETTO INTERVENTRICOLARE

- **Evoluzione e complicanze**
  - Endocardite batterica
  - Insufficienza aortica
  - SPI (rara)
  - Stenosi subaortica (rara)
  - Aritmie
- **Indicazioni terapeutiche**
  - Follow-up
  - Chiusura chirurgica
  - Chiusura interventistica (?)

DIFETTO INTERVENTRICOLARE

Table 4 Indications for intervention in ventricular septal defect

Indications	Class[a]	Level[b]
Patients with symptoms that can be attributed to L–R shunting through the (residual) VSD and who have no severe pulmonary vascular disease (see below) should undergo surgical VSD closure	I	C
Asymptomatic patients with evidence of LV volume overload attributable to the VSD should undergo surgical VSD closure	I	C
Patients with a history of IE should be considered for surgical VSD closure	IIa	C
Patients with VSD-associated prolapse of an aortic valve cusp causing progressive AR should be considered for surgery	IIa	C
Patients with VSD and PAH should be considered for surgery when there is still net L–R shunt (Qp:Qs >1.5) present and PAP or PVR are <2/3 of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy)	IIa	C
Surgery must be avoided in Eisenmenger VSD and when exercise-induced desaturation is present	III	C
If the VSD is small, not subarterial, does not lead to LV volume overload or pulmonary hypertension, and if there is no history of IE, surgery should be avoided	III	C

DIFETTO INTERVENTRICOLARE

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DOTTO DI BOTALLO

- **Presentazione clinica e storia naturale**
  - Generalmente isolato
  - Piccoli, VS e PAP normali
  - Moderati, VS dilatato e PAP normale
  - Moderati, VS normale e PAP elevata
  - Ampi, VS normale, PAP elevata (Eisenmenger)

DOTTO DI BOTALLO

- **Evoluzione e complicanze**
  - Disfunzione VS
  - Scompenso
  - Angina (anziano)
  - Ipertensione polmonare
  - Endoarterite batterica (??)
- **Indicazioni terapeutiche**
  - Chiusura interventistica
  - Chiusura chirurgica

DOTTO DI BOTALLO

Table 6 Indications for intervention in patent ductus arteriosus

Indications	Class[a]	Level[b]
PDA should be closed in patients with signs of LV volume overload	I	C
PDA should be closed in patients with PAH but PAP <2/3 of systemic pressure or PVR <2/3 of SVR	I	C
Device closure is the method of choice where technically suitable	I	C
PDA closure should be considered in patients with PAH and PAP >2/3 of systemic pressure or PVR >2/3 of SVR but still net L–R shunt (Qp:Qs >1.5) or when testing (preferably with nitric oxide) or treatment demonstrates pulmonary vascular reactivity	IIa	C
Device closure should be considered in small PDAs with continuous murmur (normal LV and PAP)	IIa	C
PDA closure should be avoided in silent duct (very small, no murmur)	III	C
PDA closure must be avoided in PDA Eisenmenger and patients with exercise-induced lower limb desaturation	III	C

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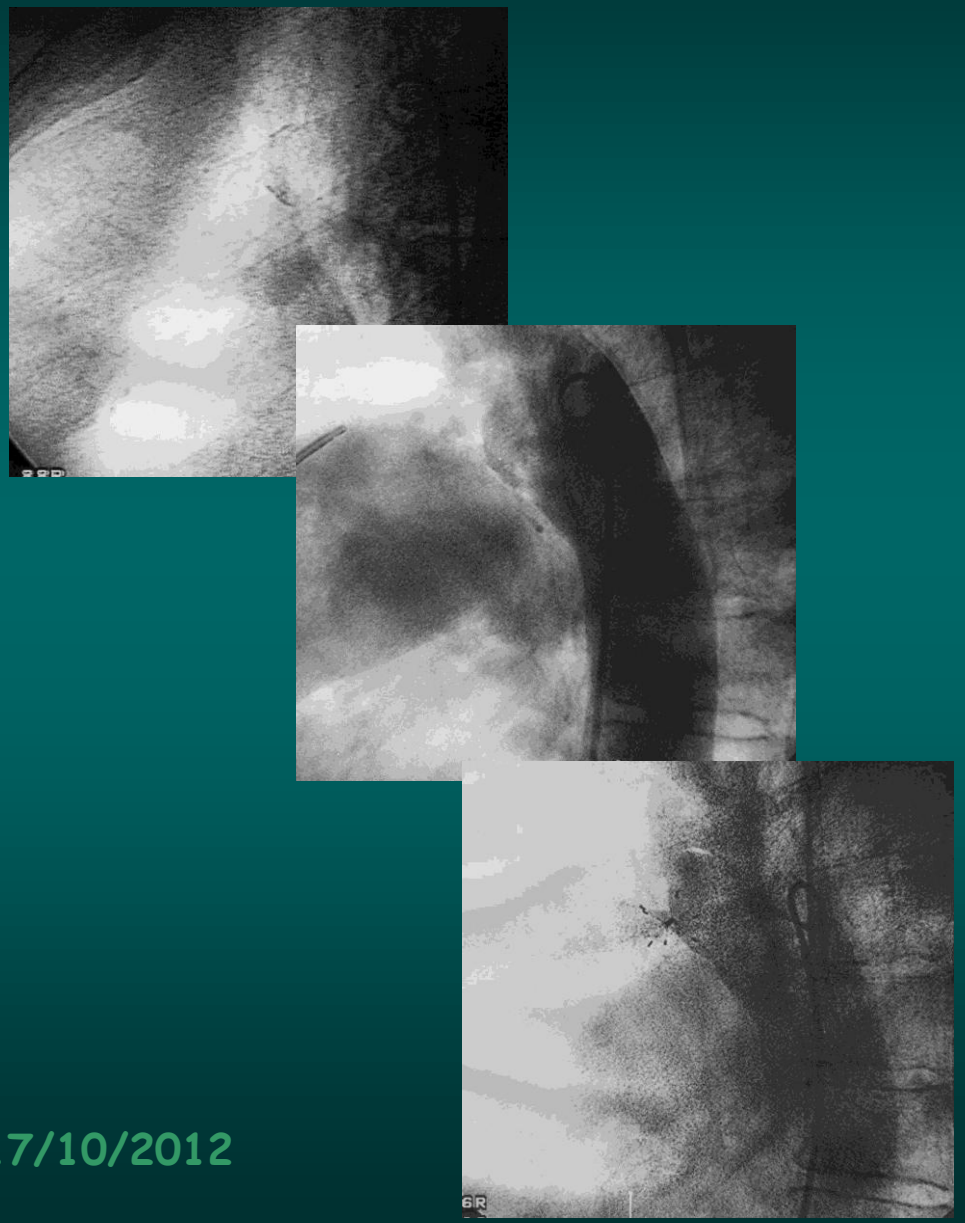

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Conclusioni

- Nell'adulto la causa più comune di shunt è il DIA (compreso il CAV parziale)
- La presenza di shunt insignificante, con PAP normali non è un'indicazione all'intervento **(III)**
- La presenza di shunt S-D significativo con PAP normale indica la correzione, con o senza sintomi **(Ia/b)**
- La presenza di complicanze o eventi clinici correlati allo shunt indica la correzione, anche se lo shunt non è significativo **(IIa)**

Conclusioni

- L'ipertensione polmonare con fisiologia tipo Eisenmenger controindica l'intervento, indipendentemente dall'entità dello shunt **(III)**
- La presenza di shunt S-D significativo con PAP aumentate, ma RVP reversibili, mantiene l'indicazione selettiva **(IIa)**
- La recente introduzione di farmaci efficaci per l'ipertensione polmonare porterà verosimilmente all'adozione di protocolli diagnostico-terapeutici condivisi

Shunt e PAP elevata - Ipotesi di protocollo

QP/QS >1.5, RVPI <6 UWm^2, RVP/RVS <0.3

>>> INTERVENTO

QP/QS >1.5, RVPI tra 6 e 9 UWm^2, RVP/RVS tra 0.3 e 0.5

- Test NO - >>> Farmaci per 12 mesi e rivalutazione

- Test NO + >>> Farmaci per 6 mesi e rivalutazione

✓ Efficace >>> INTERVENTO

✓ Non efficace >>> Bosentan per 6 mesi e rivalutazione

GRAZIE

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