

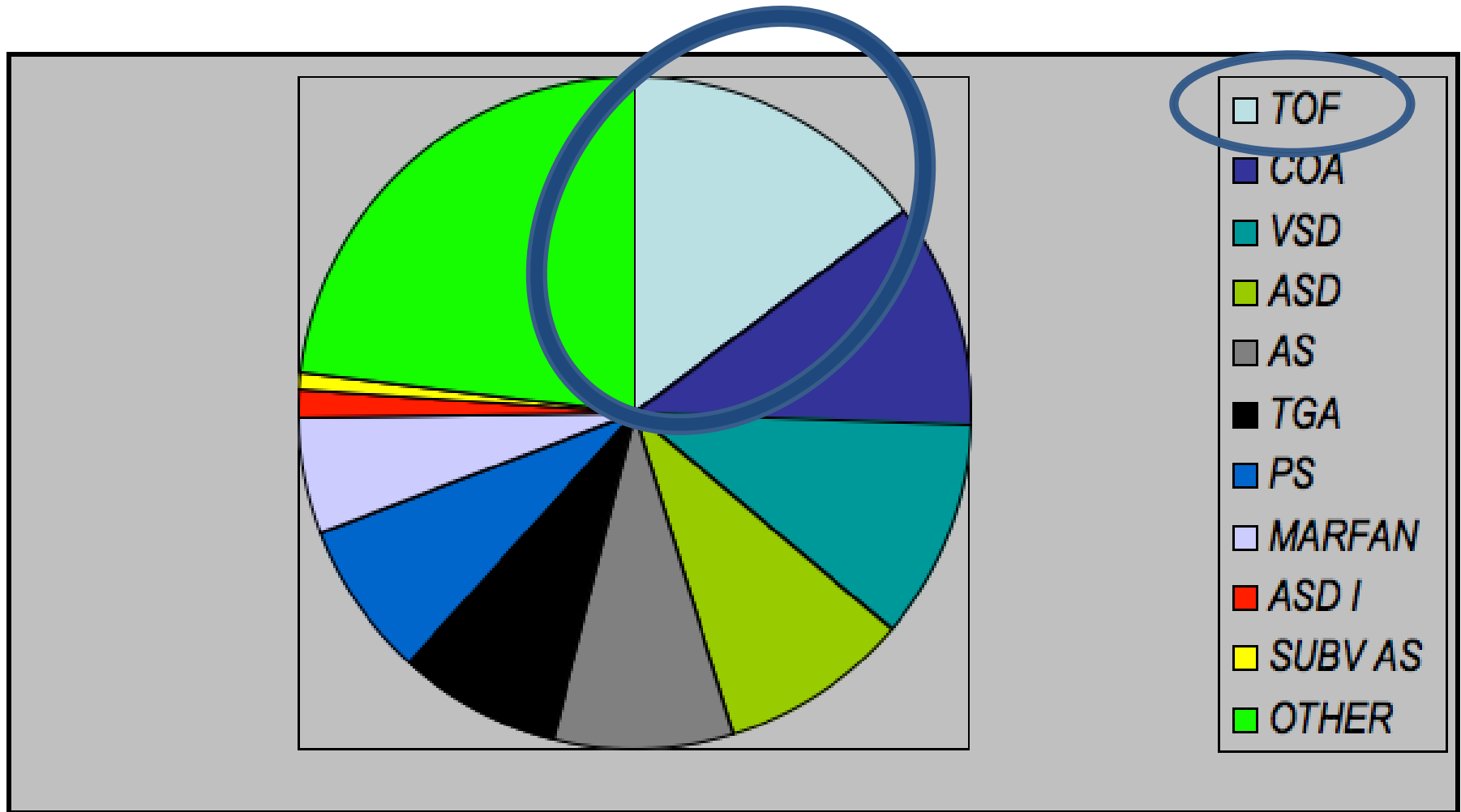


# EMODINAMICA NEL PAZIENTE OPERATO DI TETRALOGIA DI FALLOT

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Cardiologia Pediatrica  
AOU Città della Salute e della Scienza  
Torino

**Il paziente adulto  
con cardiopatia  
congenita**

# PREVALENZA DELLE CARDIOPATIE CONGENITE NELLA POPOLAZIONE ACHD



# POSSIBILI SCENARI NEL FALLOT ADULTO

1. **Pazienti in storia naturale:** molto raro
2. **Pazienti con palliazione definitiva (SHUNT):** raro
3. **Patologia dei rami polmonari:** frequente
4. **Patologia valvolare** stenosi/insufficienza o forma combinata
  - a. 80% pz correzione con **patch transanulare-patch tronco polmonare/valve sparing**
  - b. 20% pz correzione con **condotto**. (atresia polmonare/anomalie coronariche ecc)

# FISIOPATOLOGIA FALLOT OPERATO

IPOAFFLUSSO **POLMONARE**

SOVRACCARICO DI **PRESSIONE** VD

SOVRACCARICO DI **VOLUME** VD

PROBLEMI VASCOLARI (AORTA, CORONARIE)

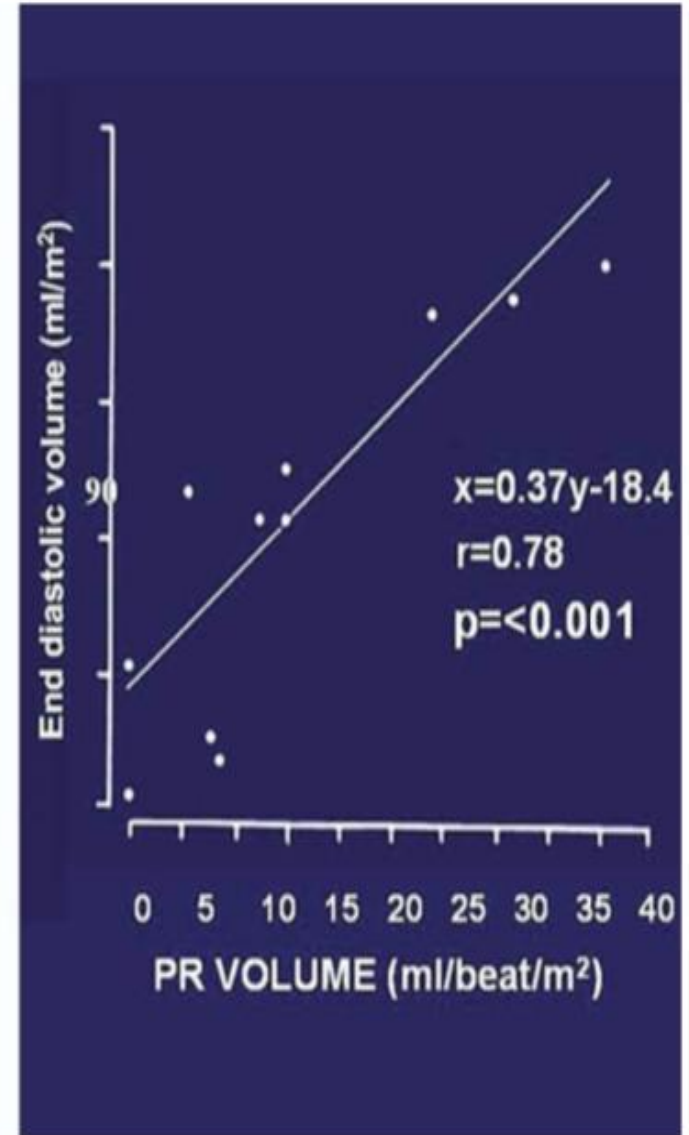
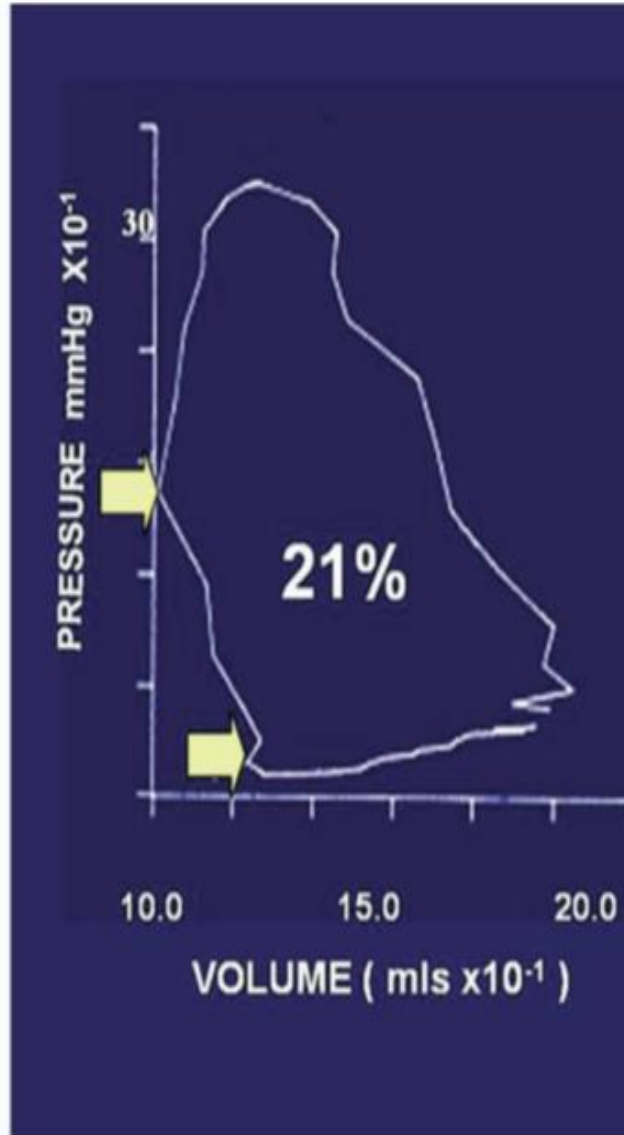
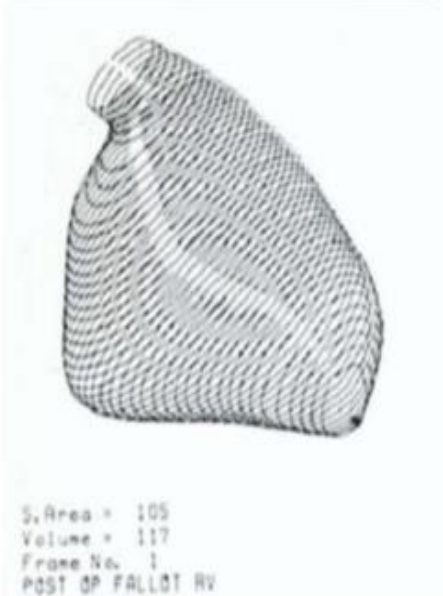


**DISFUNZIONE  
VENTRICOLARE**

# INDICAZIONI AL CATETERISMO DG-TP

1. *SEMPRE **SUCCESSIVO** A DIAGNOSTICA PER IMMAGINI COMPLETA (ECO→RM/TC)*
2. *COMPLETA CON **INFORMAZIONI FUNZIONALI** (PRESSIONI-SATURAZIONI, PORTATE, RESISTENZE, CALCOLO SHUNT)*
3. *TRATTAMENTO STENOSI RAMI POLMONARI*
4. *TRATTAMENTO STENOSI EFFLUSSO DESTRO*
5. *TRATTAMENTO INSUFFICIENZA POLMONARE*
6. *VALUTAZIONE PRE CCH/TRAPIANTO*

# CORRELAZIONE GRADO RIGURGITO-VOL VD



# INSUFFICIENZA POLMONARE CRONICA

## Fisiopatologia



Funzione valvolare

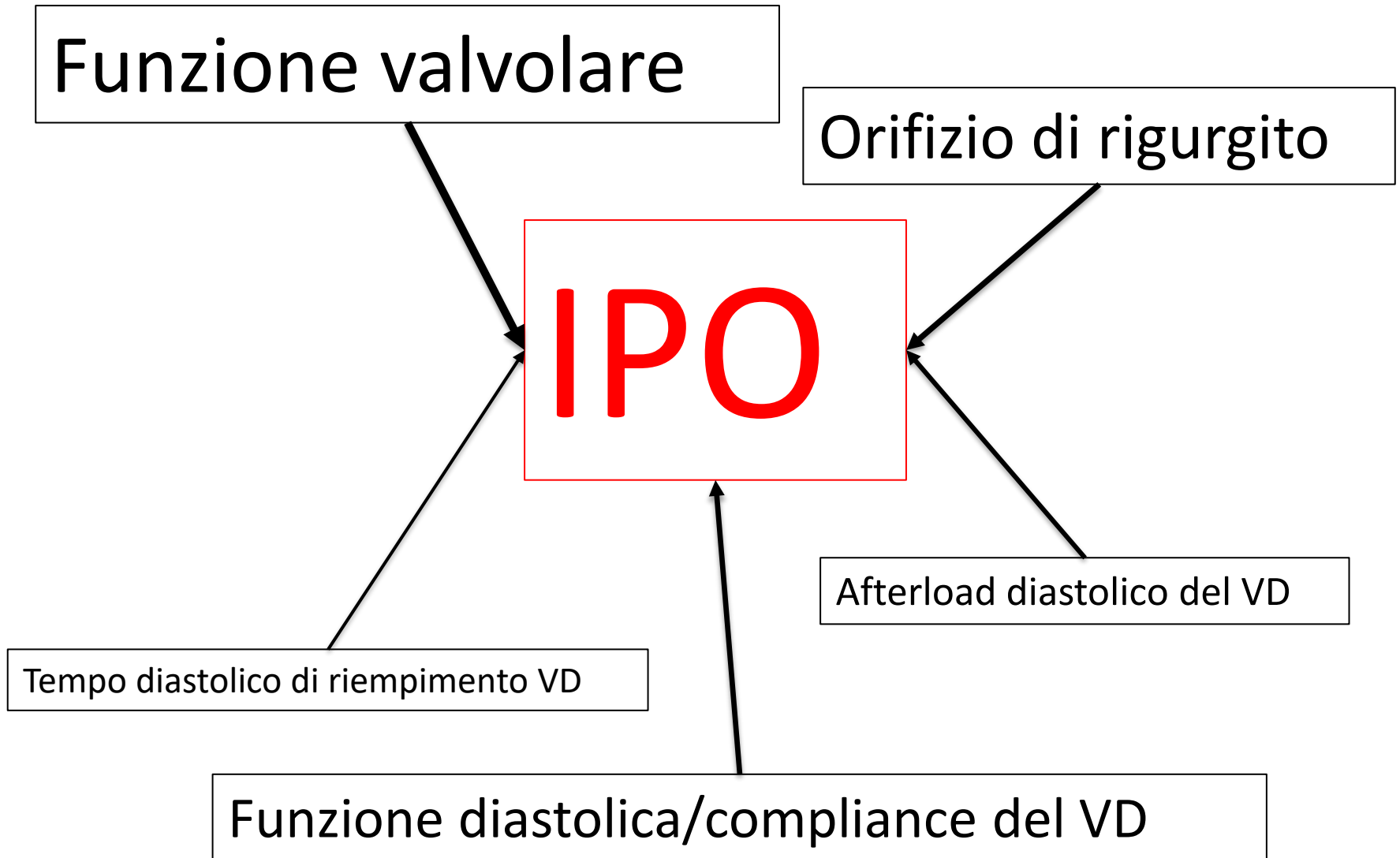
Orifizio di rigurgito

**IPO**

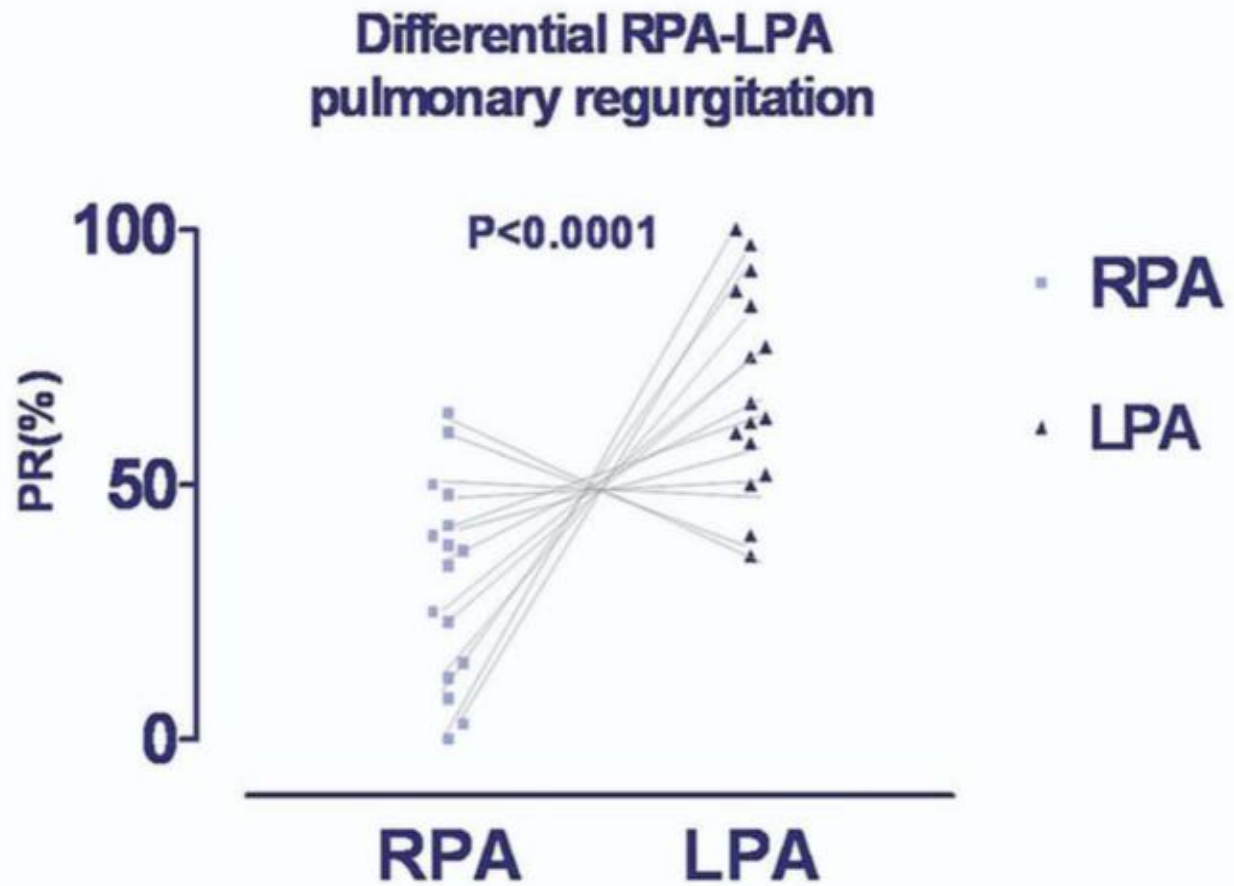
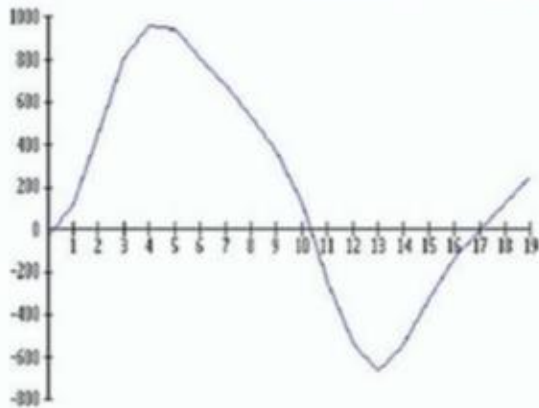
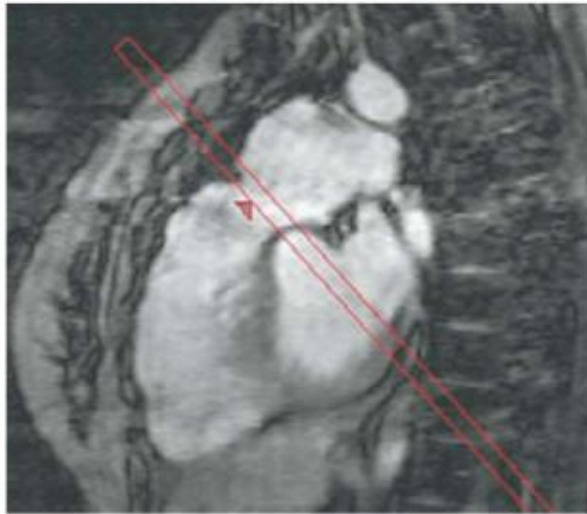
Afterload diastolico del VD

Tempo diastolico di riempimento VD

Funzione diastolica/compliance del VD

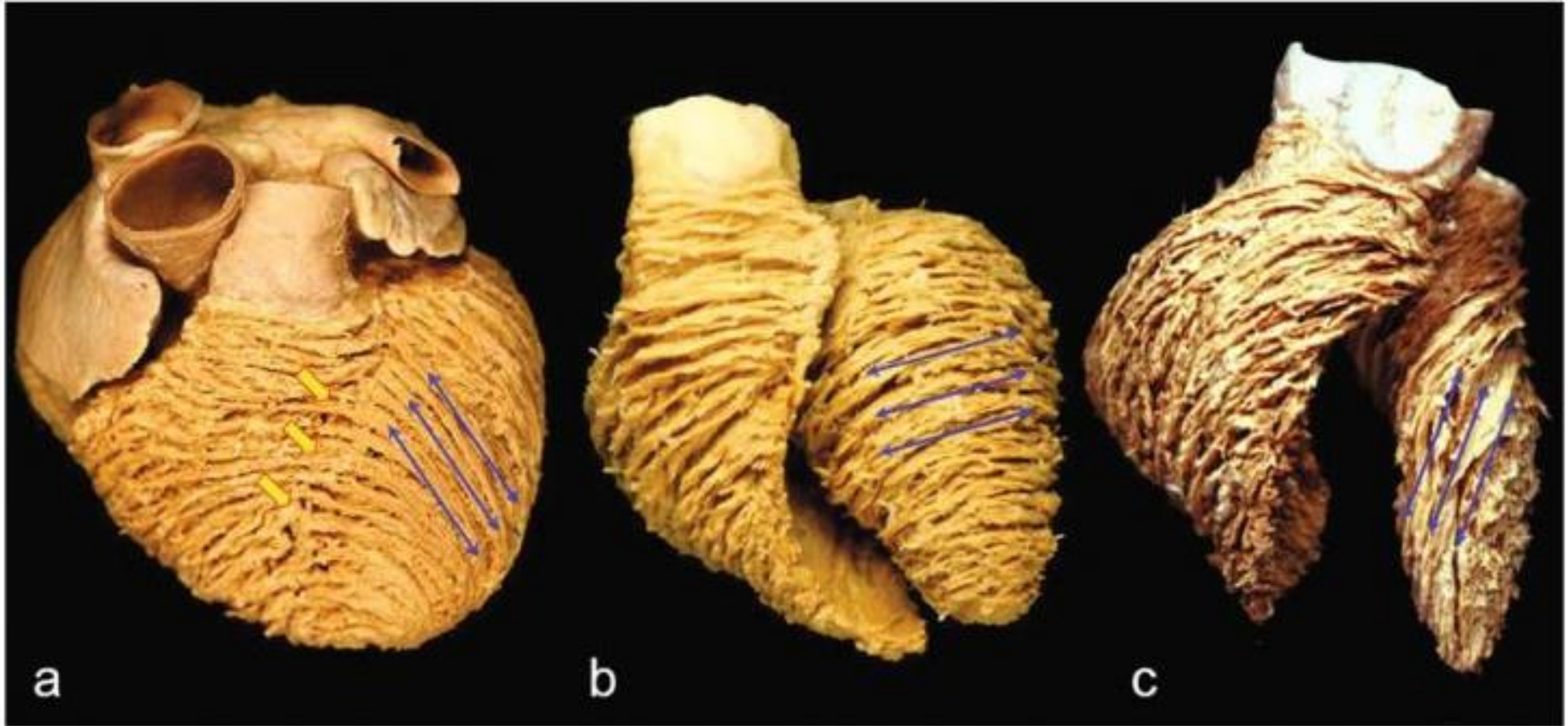


# LE ARTERIE POLMONARI CONTRIBUISCONO ALLO STESSO MODO AL RIGURGITO POLMONARE ?

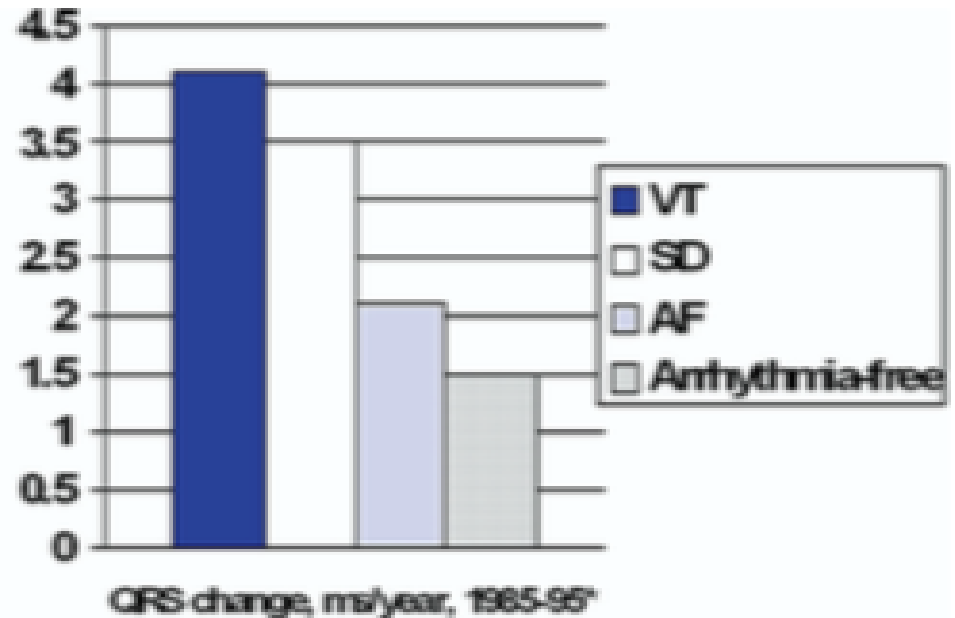
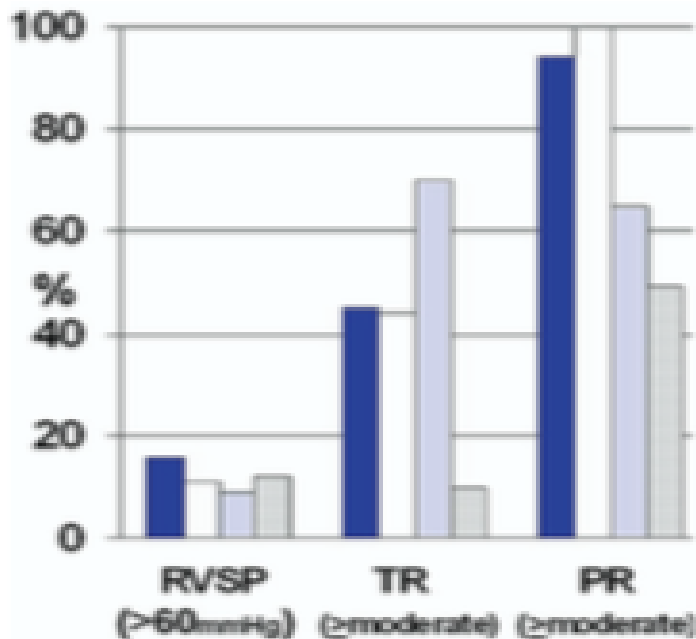


# INTERDIPENDENZA VENTRICOLARE

Interazioni tra ventricoli: (non sono unità singole): condividono pericardio, SIV, strati superficiali delle fibre miocardiche del cuore, coronarie



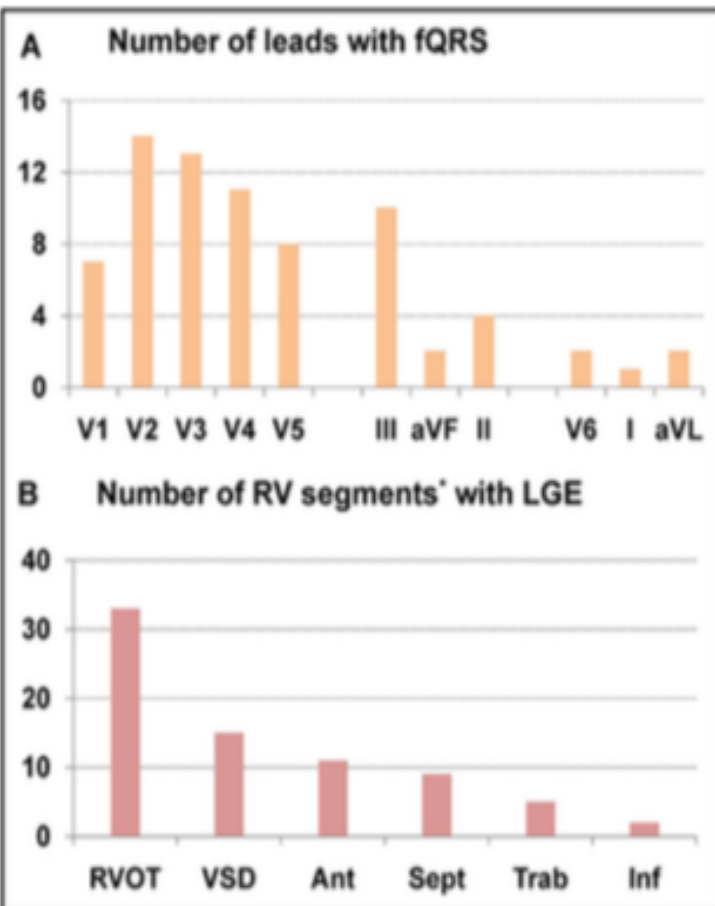
# INTERAZIONI ELETTRO-MECCANICHE



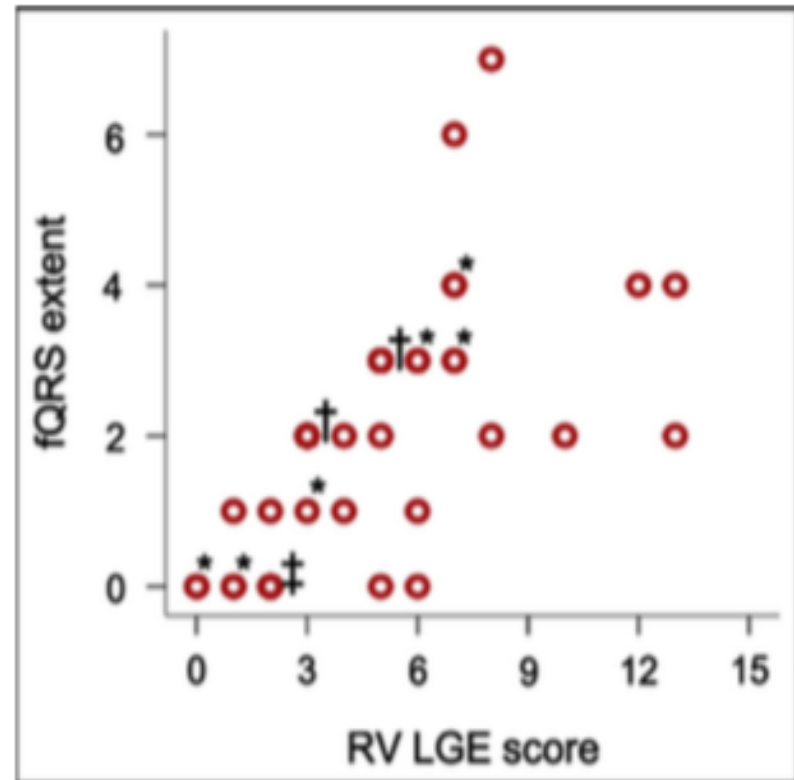
- Correlazione diretta tra dilatazione del VD e durata del QRS(msec)
- Durata QRS > 180msec: associato ad aumentato rischio di aritmie ventricolari e morte improvvisa
- 170msec<QRS<180msec e dispersione QT: aumentato rischio morte improvvisa
- ITR almeno moderata aumenta rischio aritmico (FA/FL)
- IPO almeno moderata aumenta rischio aritmico (TV/MI)
- Aumento del QRS>3.5msec/aa predittore forte MI

# Relation of Fragmented QRS Complex to Right Ventricular Fibrosis Detected by Late Gadolinium Enhancement Cardiac Magnetic Resonance in Adults With Repaired Tetralogy of Fallot

Seung-Jung Park, MD, PhD<sup>a</sup>, Young Keun On, MD, PhD<sup>a</sup>, June Soo Kim, MD, PhD<sup>a</sup>,  
Seung Woo Park, MD, PhD<sup>a</sup>, Ji-Hyuk Yang, MD, PhD<sup>b</sup>, Tae-Gook Jun, MD, PhD<sup>b</sup>,  
I-Seok Kang, MD<sup>d</sup>, Heung Jae Lee, MD, PhD<sup>d</sup>, Yeon Hyeon Choe, MD, PhD<sup>c</sup>, and  
June Huh, MD, PhD<sup>d,\*</sup>



Framm  
V<sub>1</sub>-V<sub>5</sub>,  
fQRS c  
fQRS c  
fQRS p



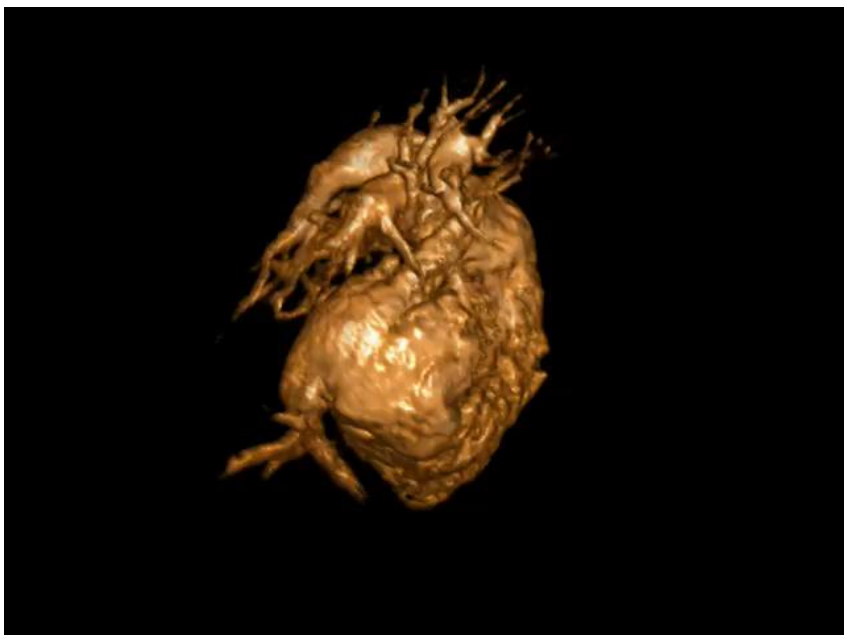
ntigue:

# INDICAZIONE- QUANDO?

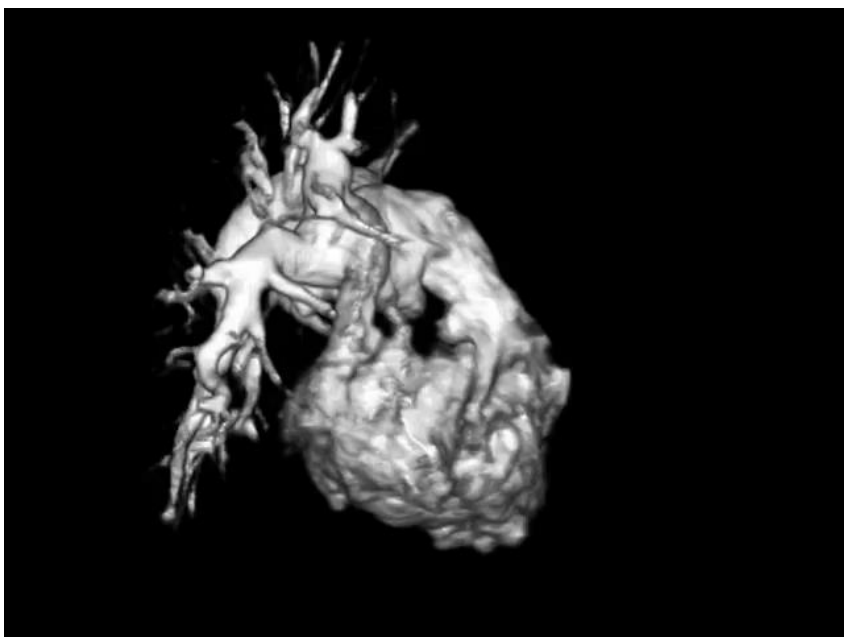
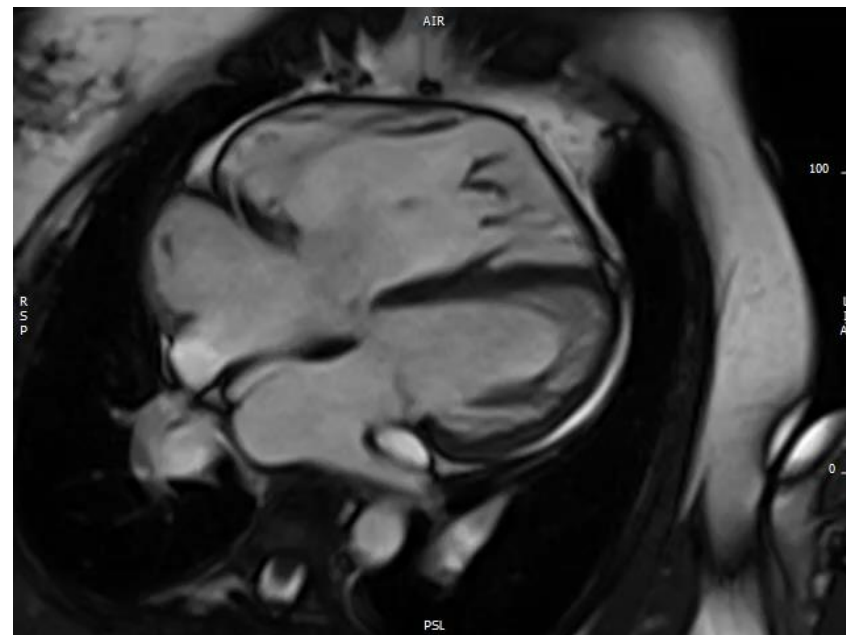
*Arriviamo troppo **presto o tardi**??*

*Con dati RM operiamo prima di quello che avremmo fatto seguendo linee guida (dati registro USA dal 2004-2012: triplicati PVR)*

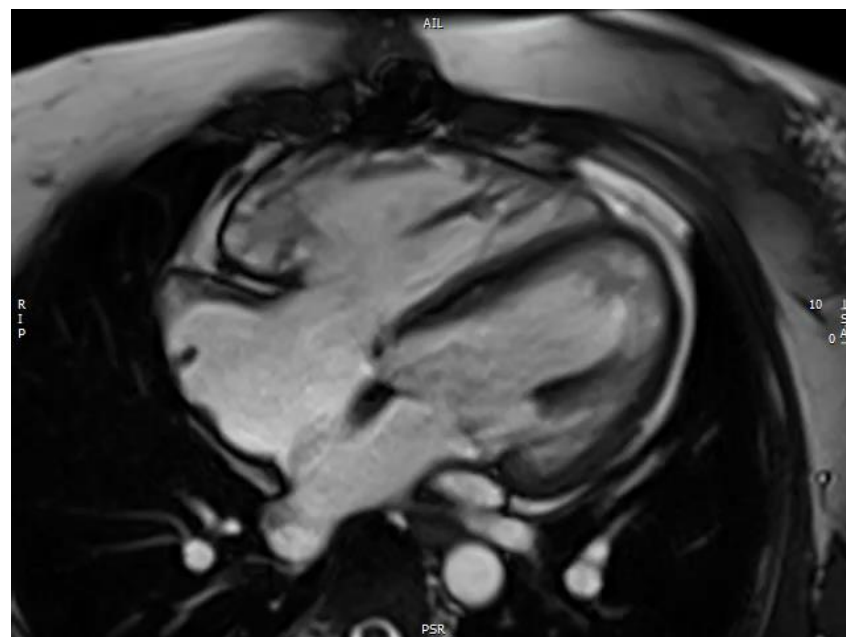
- Insufficienza valvolare polmonare severa ( $FR \geq 35\%$  alla RMN)
- **NO reverse remodeling del VD** se intervento con  $VTDVD > 150-170\text{ml/m}^2$ ,  $VTsVD > 80-85\text{ml/m}^2$
- **FE VD < 45%: bassa probabilità di miglioramento funzione VD**
- **$VTDVD/VTDVS \geq 1,5$**  associato a sintomatologia oppure  **$\geq 2,0$**  in assenza di sintomi)
- **Pressione sistolica VD  $\geq 2/3$**  della pressione sistemica in presenza di sintomi oppure  **$> 3/4$**  in assenza di sintomi
- **Ridotta tolleranza allo sforzo** (Picco di consumo di ossigeno  $\leq 65\%$  del valore predittivo)
- **Aritmie ventricolari sostenute**



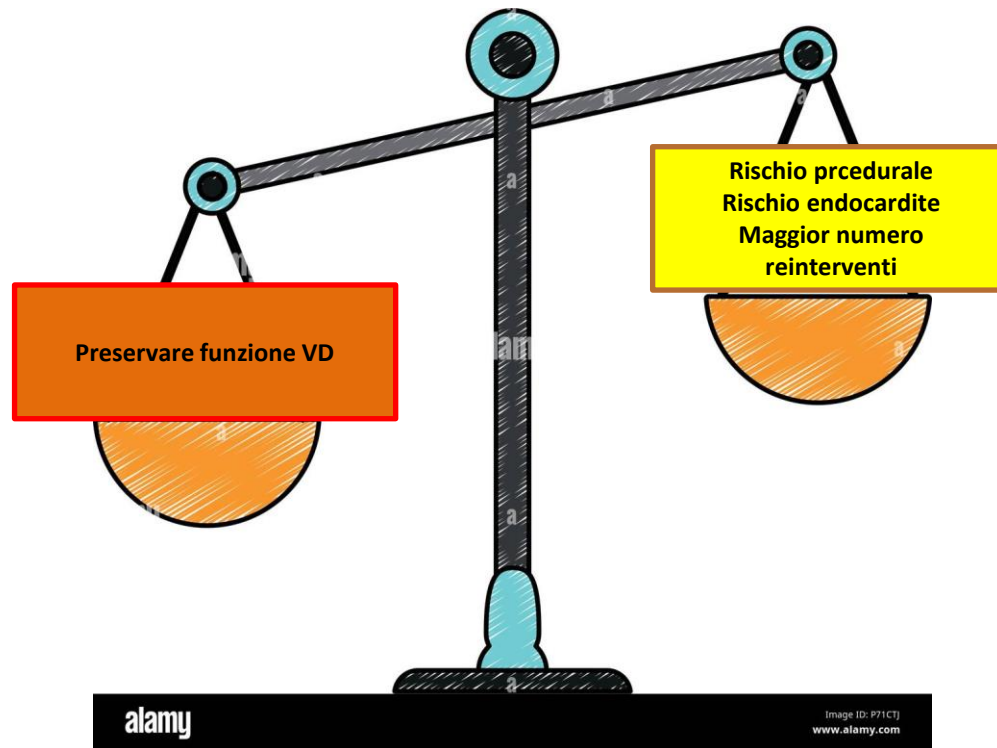
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# VALVOLAZIONE PRECOCE?



ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). Eur Heart J 2010; 31(23): 2915-57.

AHA/ACC guideline for the management of adults with congenital heart disease: Executive summary: A report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines. J Am Coll Cardiol 2019; 139(14): e637-97.

## Systematic Review/Meta-analysis

# **Pulmonary Valve Replacement for Pulmonary Regurgitation in Adults With Tetralogy of Fallot: A Meta-analysis—A Report for the Writing Committee of the 2019 Update of the Canadian Cardiovascular Society Guidelines for the Management of Adults With Congenital Heart Disease**

- Incidence rate of **death** was 1% per year
- Incidence rate of **sustained ventricular arrhythmias** was 1% per year
- **PVR improved symptoms**
- Indexed RV end-diastolic (61.29 mL/m<sup>2</sup>, 43.64 to 78.94 mL/m<sup>2</sup>) and end-systolic (37.20 mL/m<sup>2</sup>, 25.58 to 48.82 mL/m<sup>2</sup>) **volumes decreased** after PVR
- **RV EF did not change**

**Fino al 2000 Chirurgia era l'unica possibilità...**

**Medtronic Melody:** marchio CE nel 2006: **condotti** / **valvole** disfunzionanti  
**Edwards Sapien XT:** FDA approv. 2016 per **condotti** disfunzionanti

- 229
- USA,
- 58%
- Dege
- 42%

## ON A 1962 VESPA SCOOTER TOUR DE HIMALAYAS



FOLLOW US ON @northernareasofpakistan

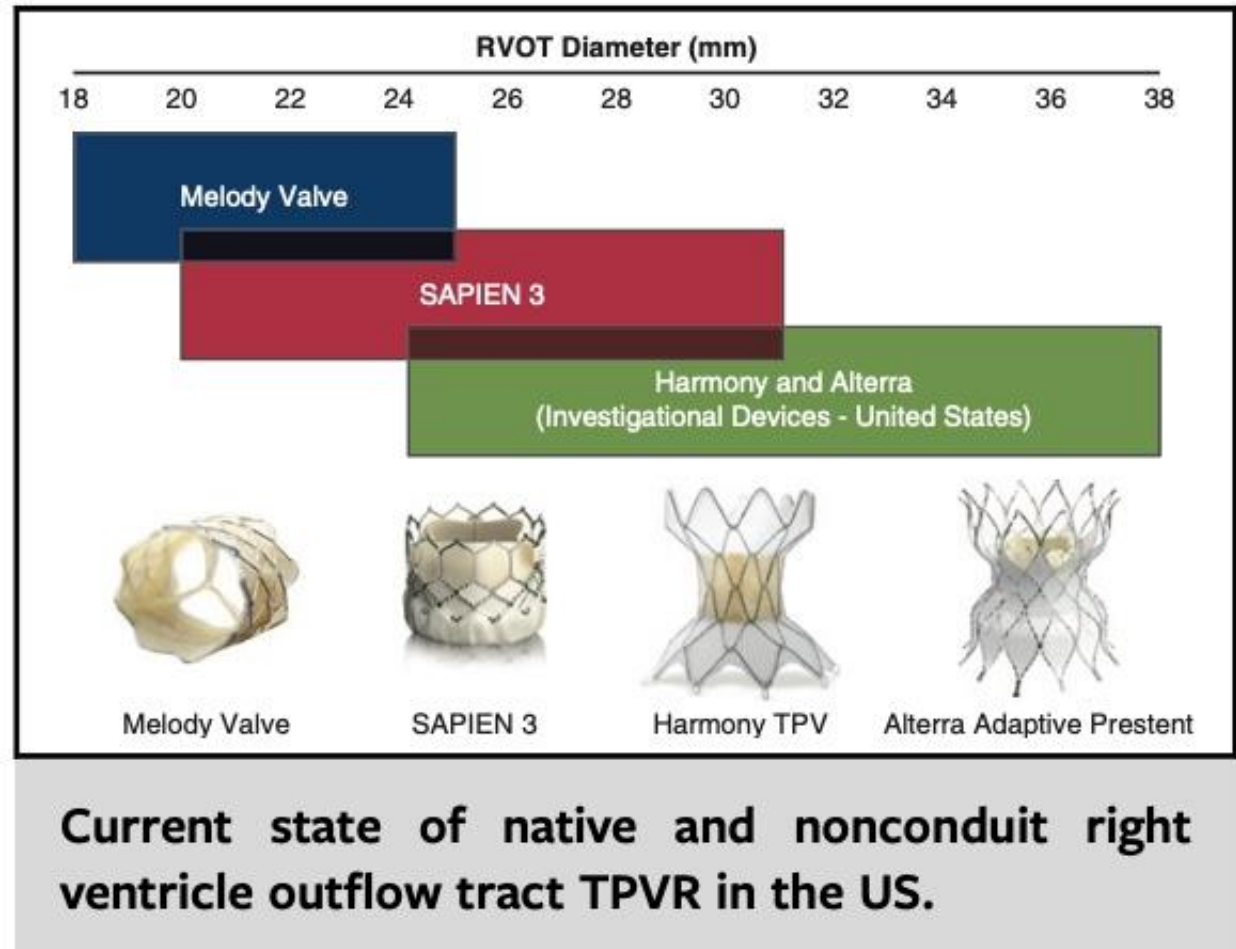
Tour de Himalayas On A 1962 Vespa Scooter

Guarda

Fino al 2018 solo il 58% degli impianti aveva successo nei nativi !!

Martin MH, et al. Safety and feasibility of Melody transcatheter pulmonary valve replacement in the native right ventricular outflow tract. JACC Cardiovasc Interv. 2018;11:1642-50.

# EVOLUZIONE DELLA TPVR



- Successo procedurale 96.2%
- Libertà da reintervento 93.7% a 3aa

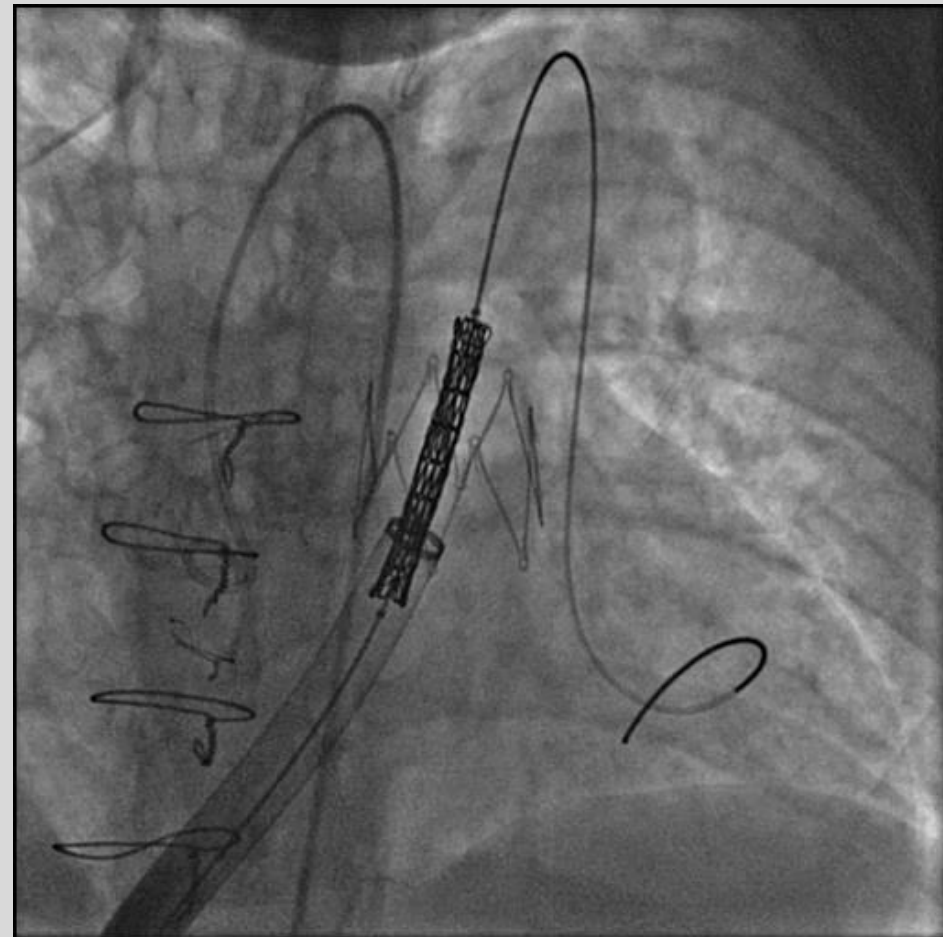
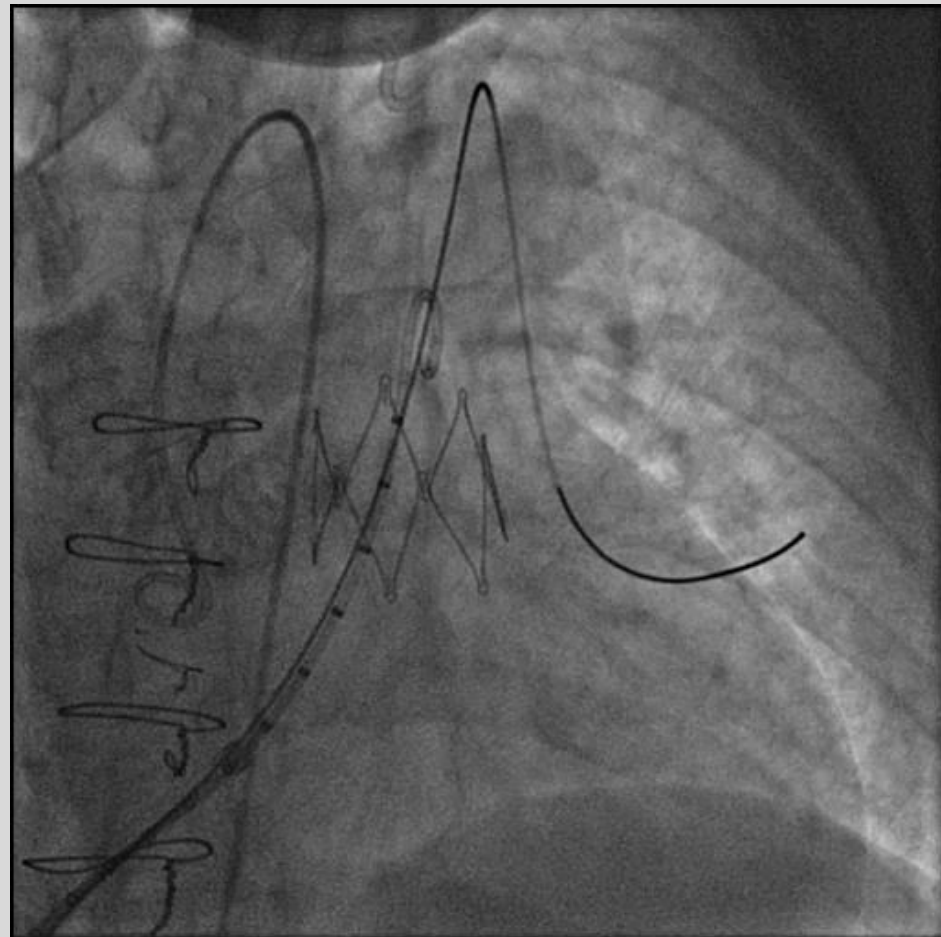
1. Transcatheter pulmonary valve replacement in native and nonconduit right ventricle outflow tracts. **J Thoracic and Cardiovascular Surgery** 2020 (162), 3
2. Kenny D et al. 3-Year Outcomes of the Edwards SAPIEN Transcatheter Heart Valve for Conduit Failure in the Pulmonary Position From the COMPASSION Multicenter Clinical Trial. **JACC Cardiovasc Interv** 2018; 11: 1920-1929
3. Fraisse A et al. Melody® transcatheter pulmonary valve implantation: results from a French registry. **Arch Cardiovasc Dis** 2014; 107: 607- 614

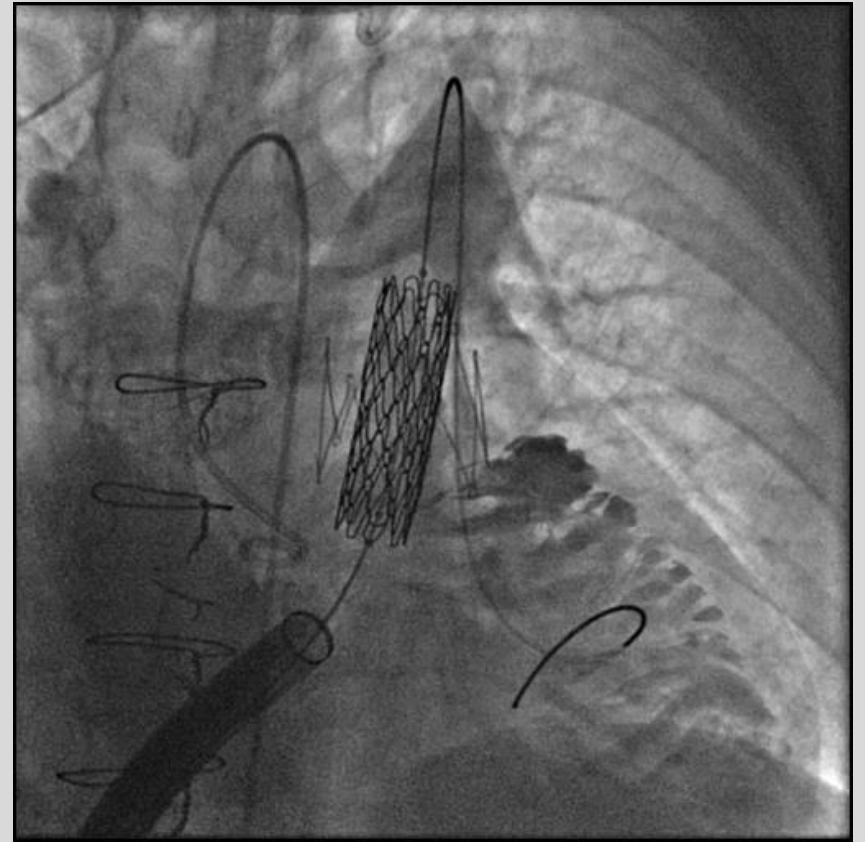
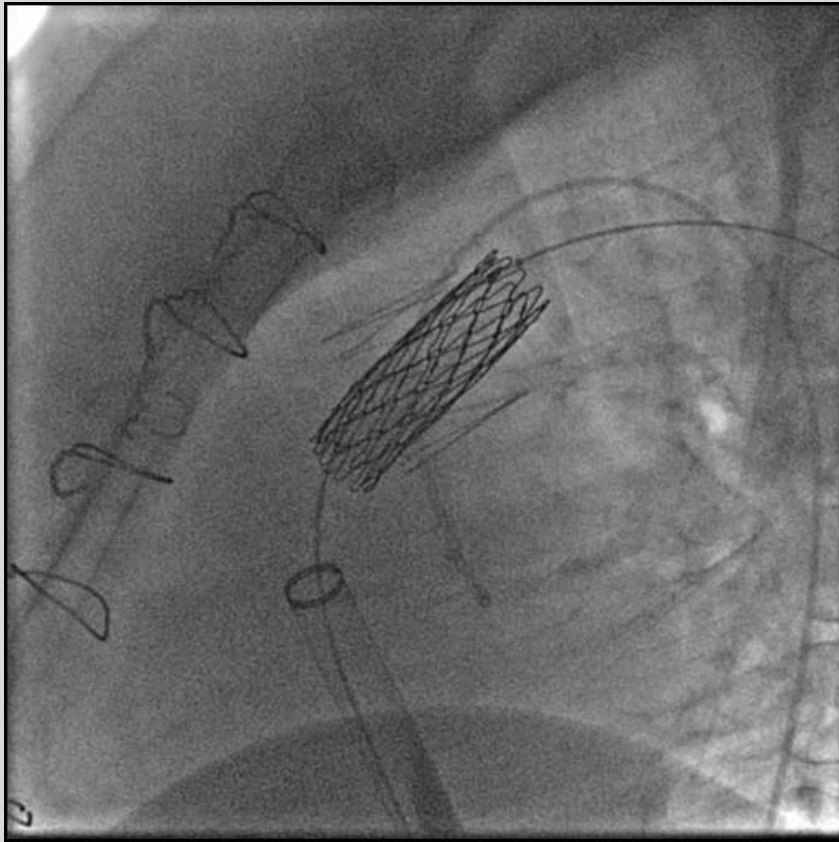
# DIVERSI SCENARI PPVI- DIVERSE TECNICHE

1. IN CONDOTTI VALVOLATI (Angioplastica->stent->valvolazione)
2. IN RVOT NATIVI O PATCHED ( $\pm$ stent->valvolazione diretta)
3. IN PREGRESSE VALVOLAZIONI ( $\pm$ stent->valvolazione diretta)

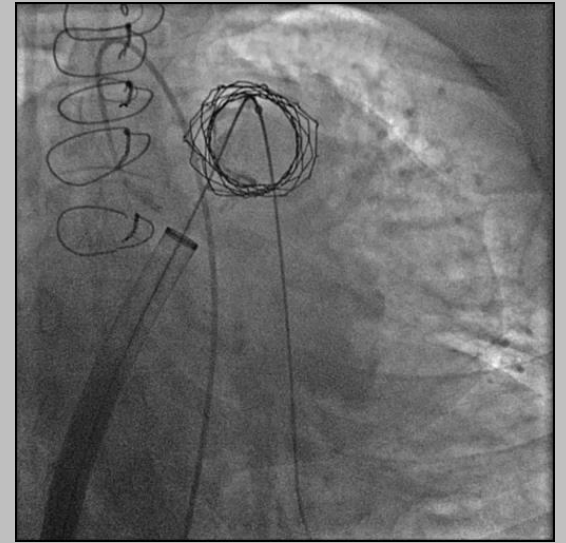
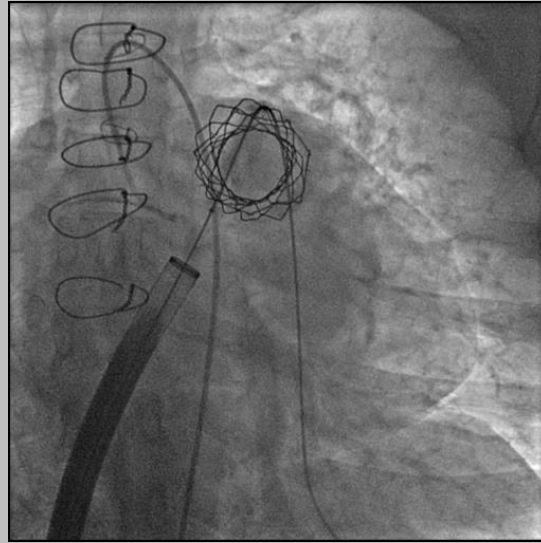
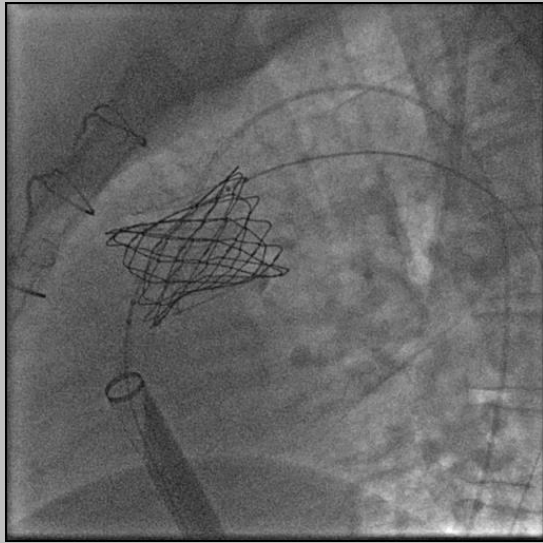
# VALVULOPATIA RESIDUA

42aa. S/P TF. S/P Valvola biologica Biointegral 27 in sede polmonare. Stenosi severa. Grad picco 44mmHg.



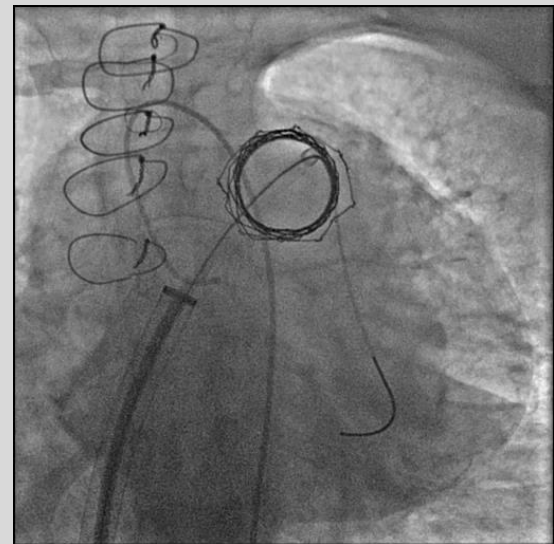
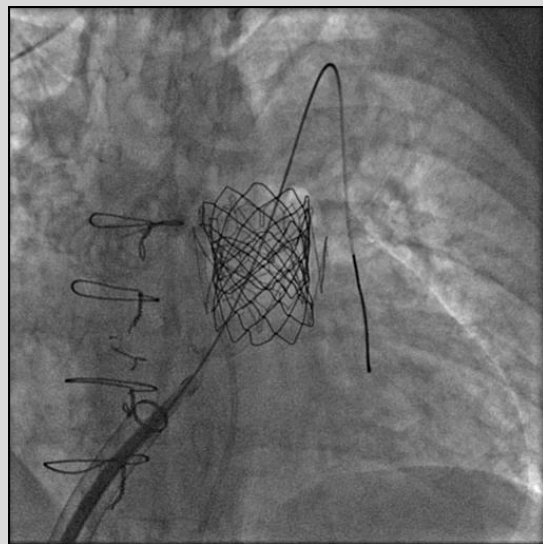
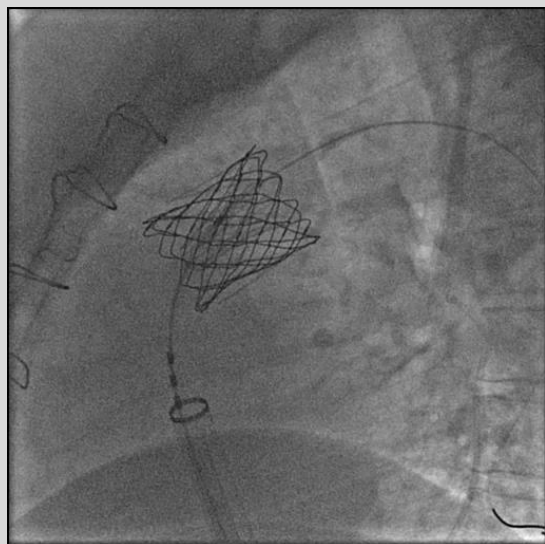


*CP stent ricoperto 45mm su BIB 26mm*



*Ctr angiografico + CGF. Post dilatazione con Pallone  
Atlas Gold 26 a 10atm. Ricontrollo CGF*

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*Impianto Valvola Sapien 3 26. Ricontrollo CGF*

# CRITICITÀ TPVR

- Dimensioni RVOT
- Eterogeneità forma RVOT
- RVOT è uno «spazio dinamico»
- Compliance Landing-zone

**BALLOON INTERROGATION**

**ACCURATO IMAGING TC/RM**

Balloon Expandable Valve

Self- expanding Valve

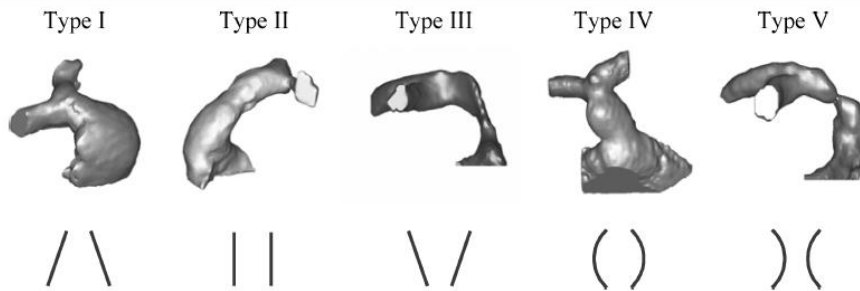
Dal 2021 Sapien 3 marchio CE per RVOT nativi

2022 P-Venus Valve marchio CE per RVOT nativi fino a 36mm

# RVOT

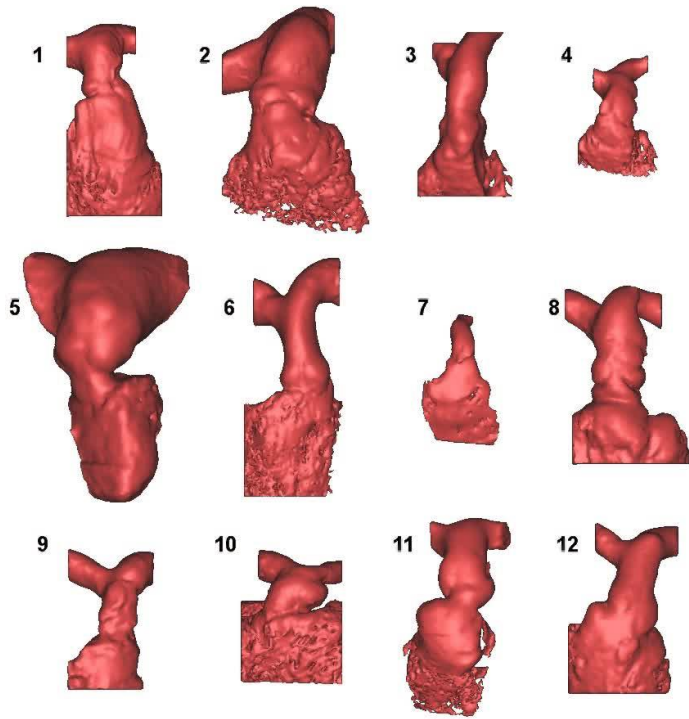


Valente et al 2014



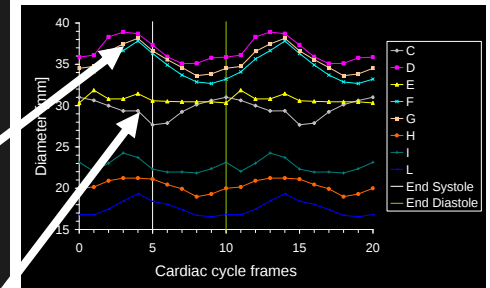
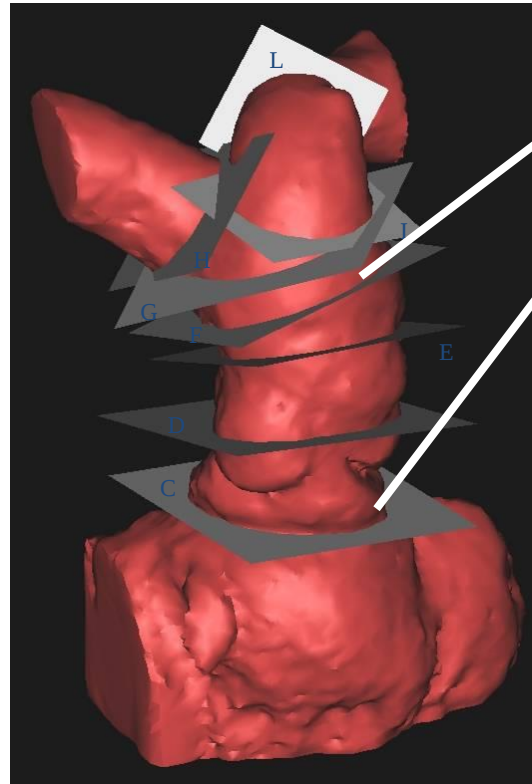
Schievano et al JCMR 2007

# Dynamic RVOT dimensions



**4D CT**

*Mimics, Materialise*

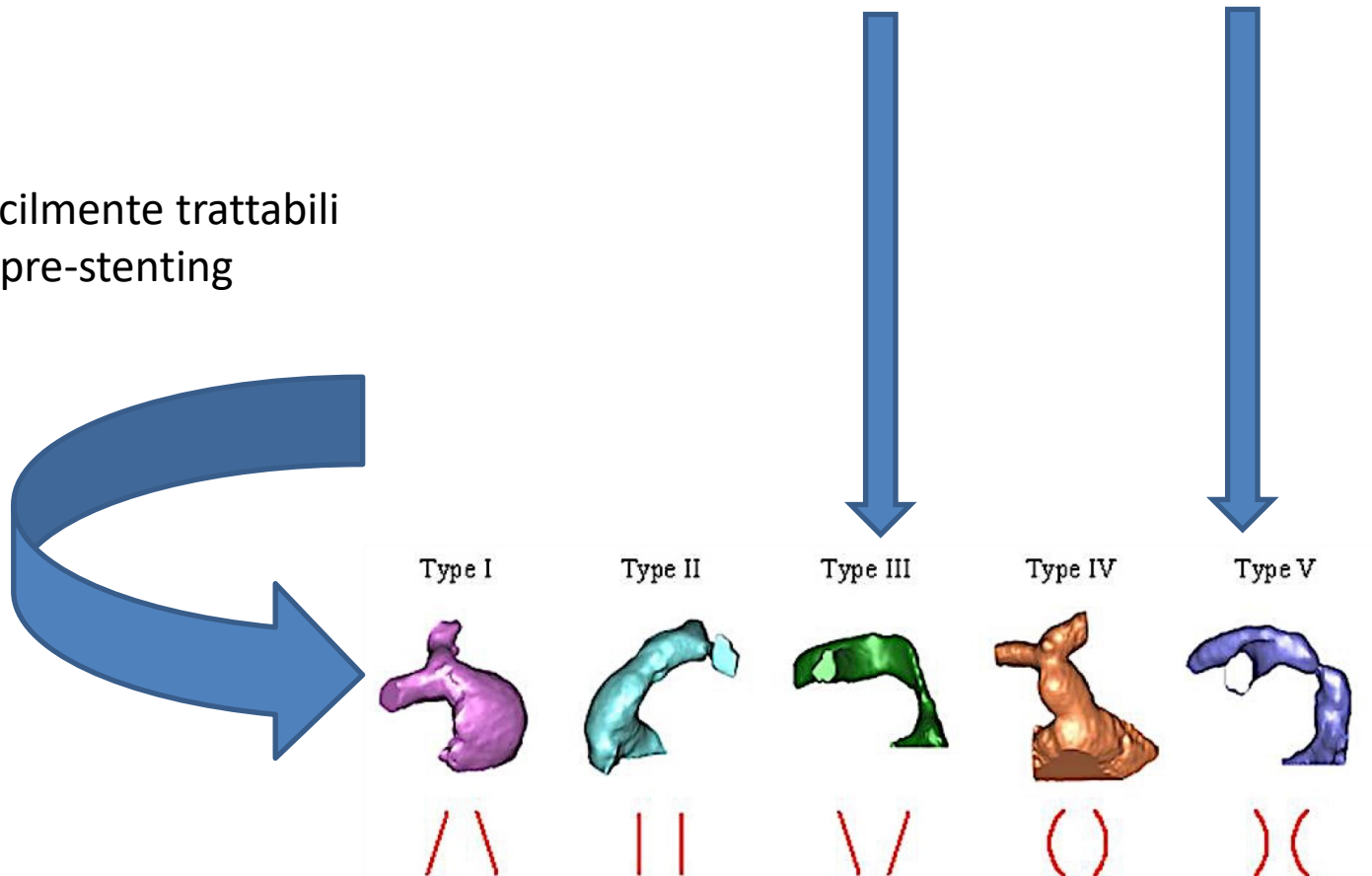


*Schievano, EuroIntervention 2010*

# Morfologia RVOT

*Ideali per pre-stenting e balloon-expandable*

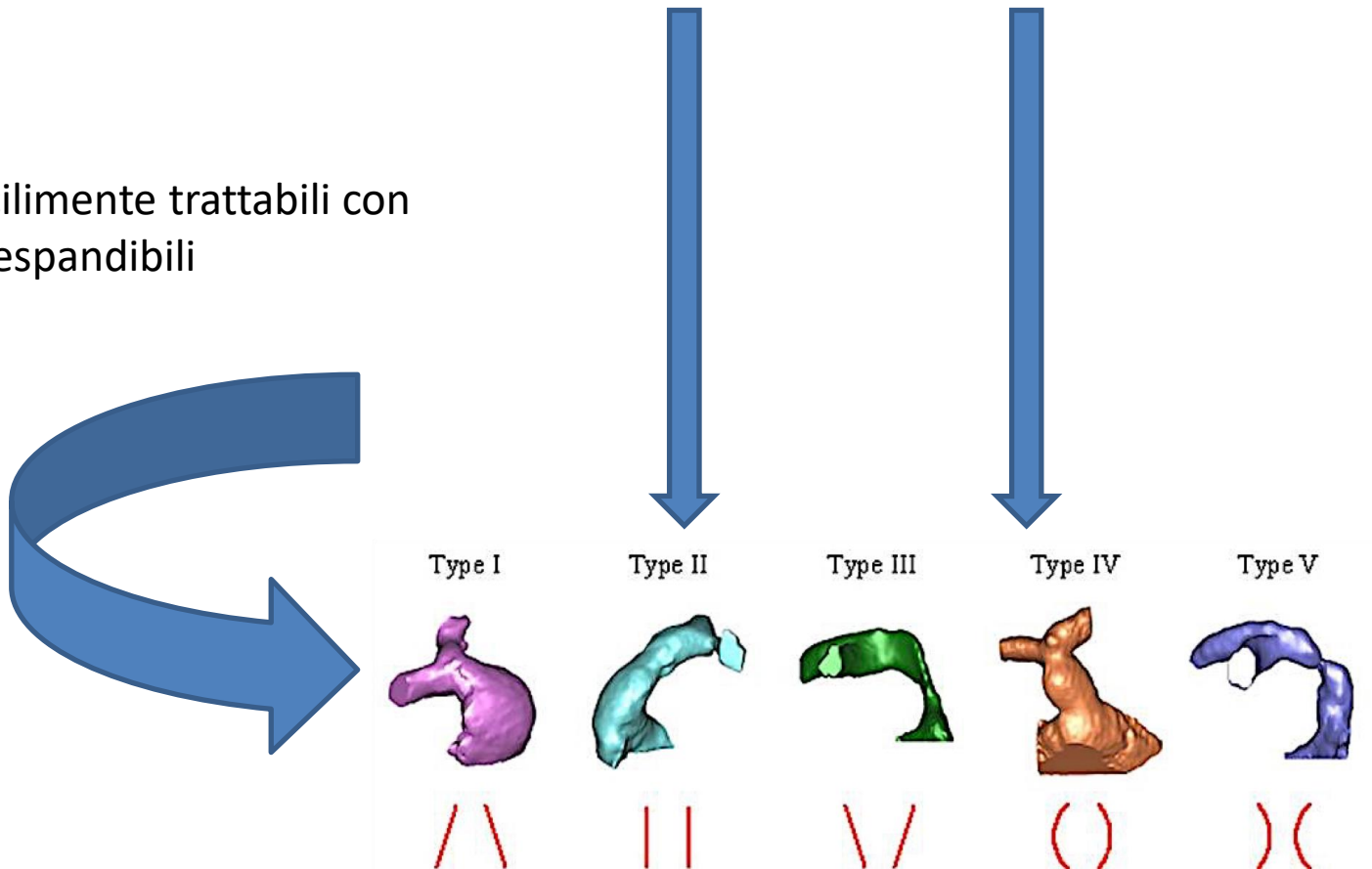
Difficilmente trattabili  
con pre-stenting



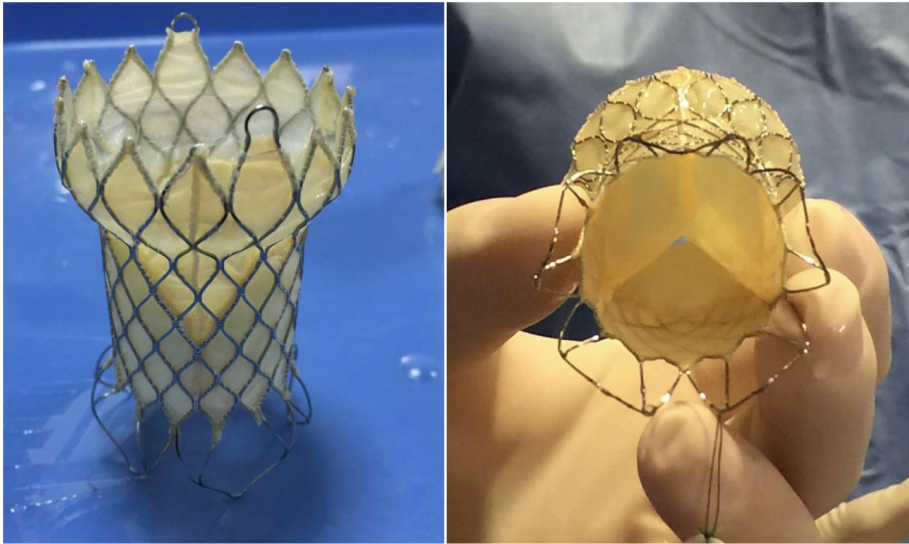
# Morfologia RVOT

*Ideali per valvole auto-espandibili*

Difficilmente trattabili con  
autoespandibili



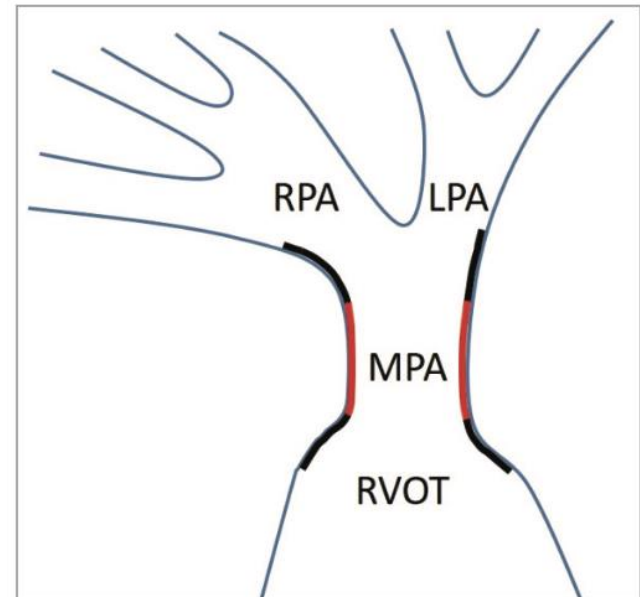
# P-venus



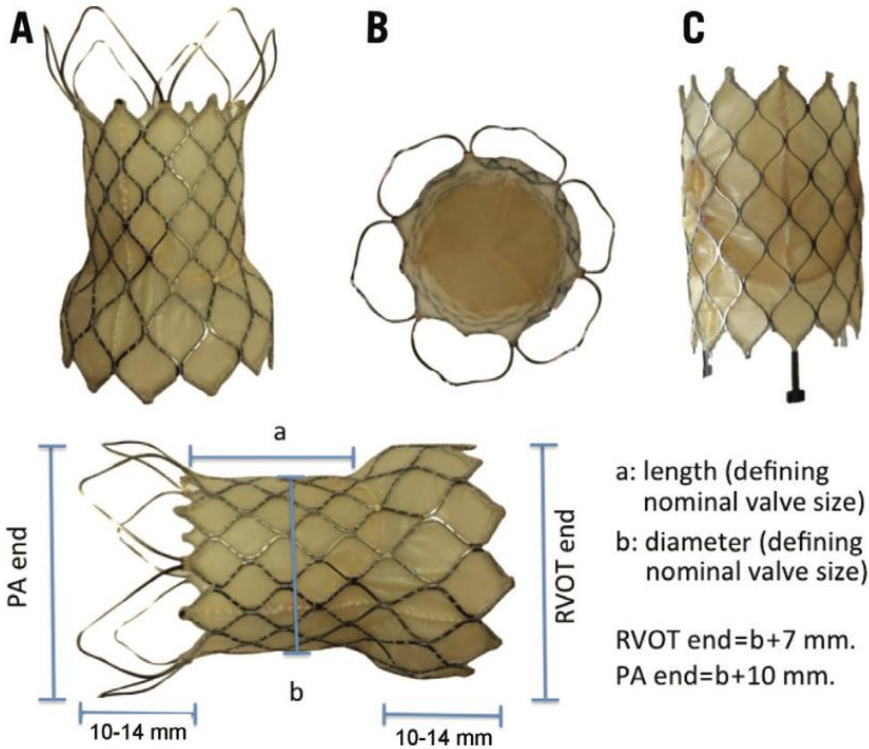
- Valvola autoespandibile
- Per RVOT nativi-patched
- Svasata ai lati
- Non coperta sul versante polmonare
- Max diametro 36mm (efflusso max 30-32mm)

## Selection criteria for percutaneous pulmonary valve implantation

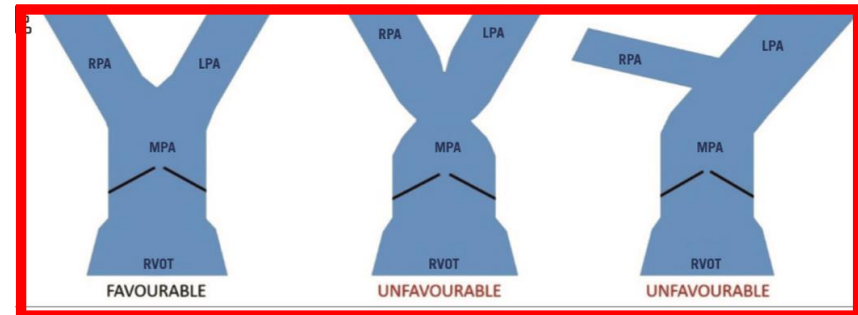
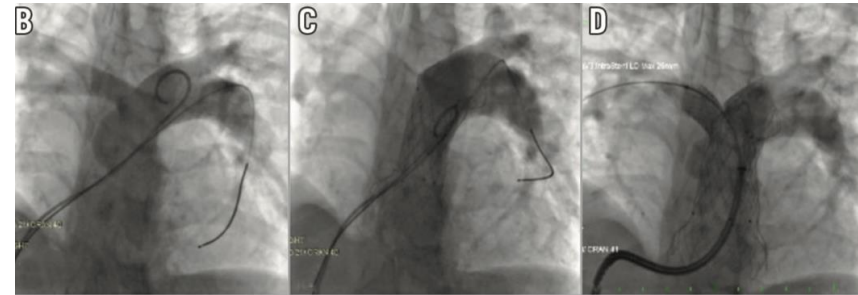
1. Significant increase in right ventricular size (RVEDVi >150 ml/m<sup>2</sup> by CMRI or less if RVEF <45%).
2. Significant pulmonary regurgitation (PRF >30% by CMRI or severe by TTE).
3. Symptomatic deterioration either subjective or objective by exercise testing.
4. Favourable anatomy (no RVOTO, no branch pulmonary stenosis, no potential coronary artery obstruction).
5. Size of the RVOT not suitable for available valves by angiographic assessment (larger than 22 to 26 mm but smaller than 32 mm).
6. Patent central veins and weight >25 kg.



# P-venus



- Diametro centrale (b) valvola 28 (+2mm)->36mm
- Lunghezza (a) 25-30-35mm
- Scelta diametro: MPA balloon interrogation +2-4mm

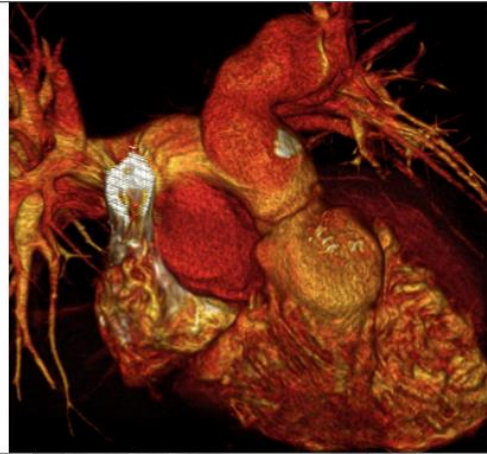


TF RVOT PATCHED – INDAGINE TC

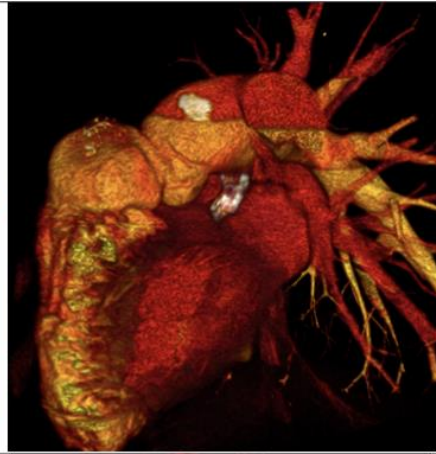


# IMAGING AVANZATO PRE PROCEDURA

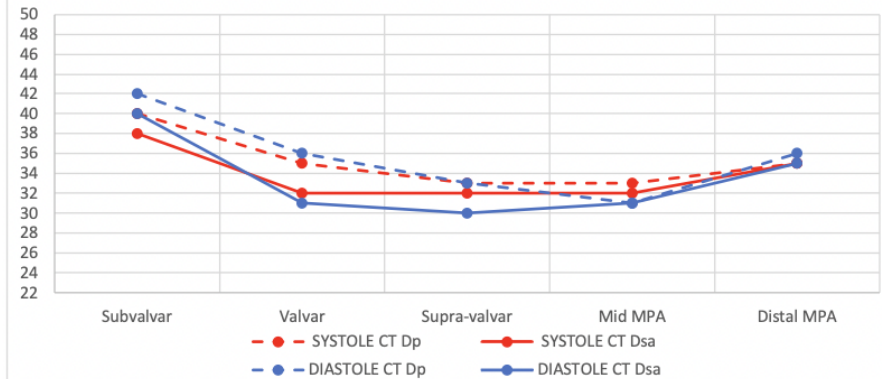
CRANIAL CAUDAL VIEW



LATERAL VIEW

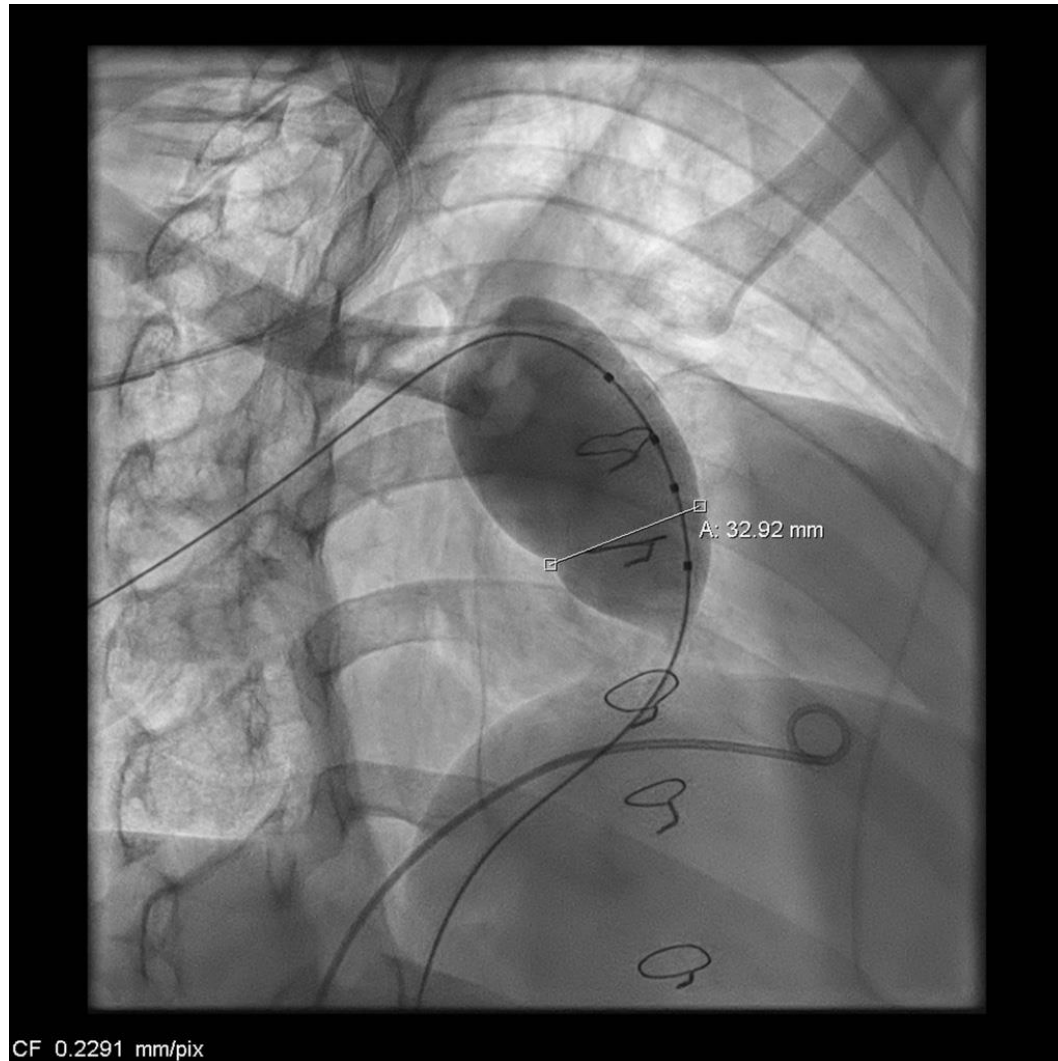


RVOT MPA DIMENSION (CT)

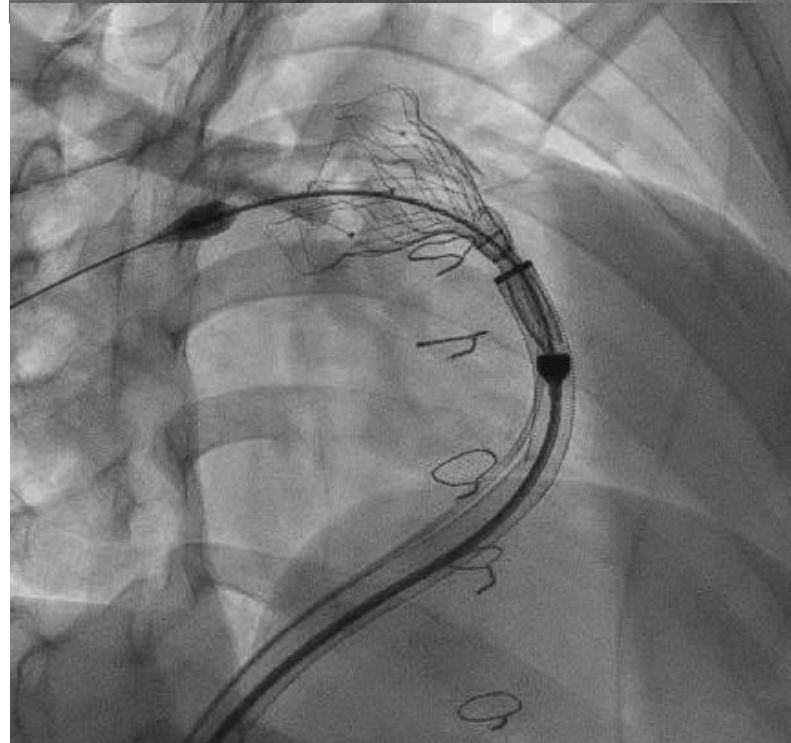
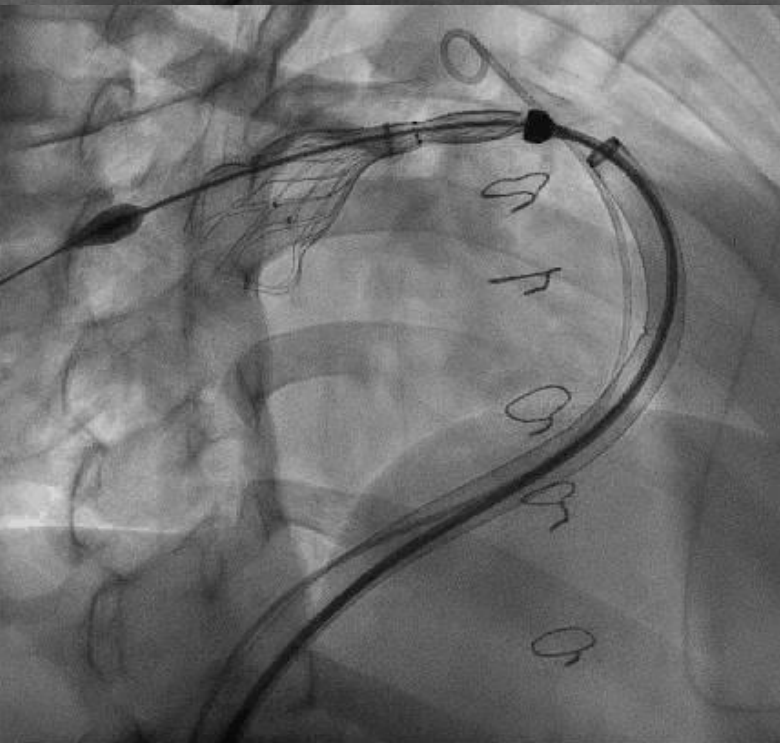
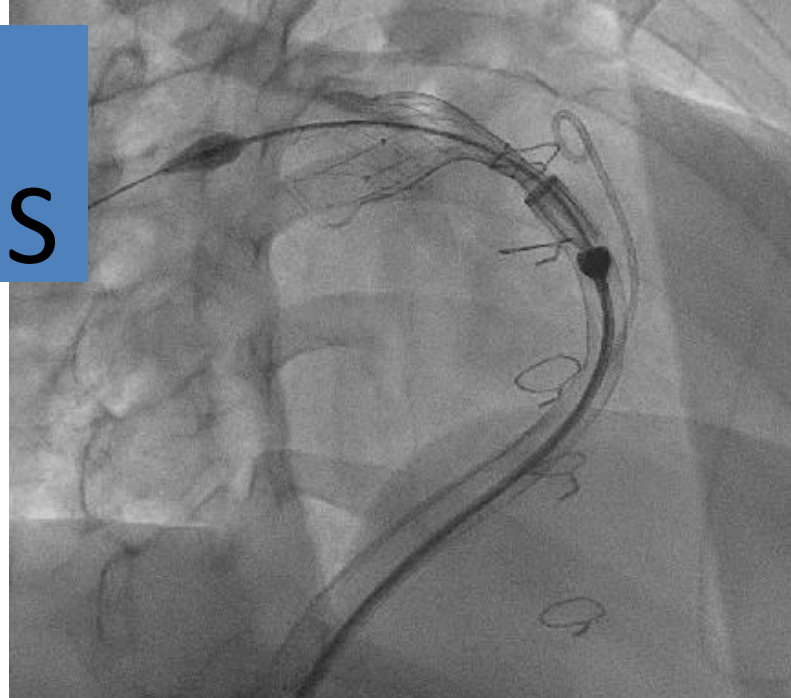
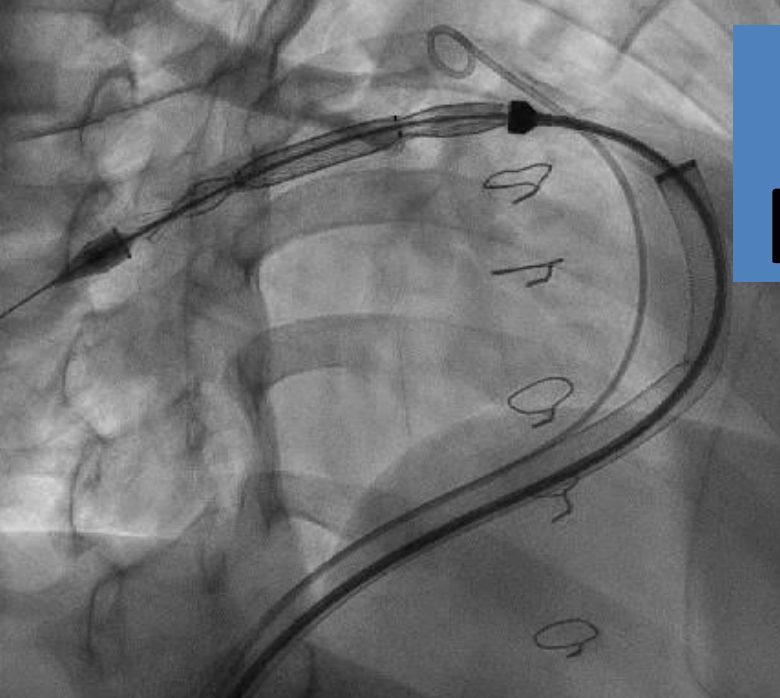


| DIAMETER (mm) systole          | MPR   | PERIMETER | SURFACE AREA |
|--------------------------------|-------|-----------|--------------|
| SUBVALVAR                      | 33x44 | 40        | 38           |
| VALVAR                         | 24x40 | 35        | 32           |
| SUPRAVALVAR                    | 26x40 | 33        | 32           |
| MID MPA                        | 26x37 | 33        | 32           |
| DISTAL MPA                     | 32x38 | 35        | 35           |
| LPA ORIGIN                     | 19x23 |           |              |
| RPA ORIGIN                     | 25x34 |           |              |
| LENGTH (VALVAR-PA BIFURCATION) | 40-45 |           |              |

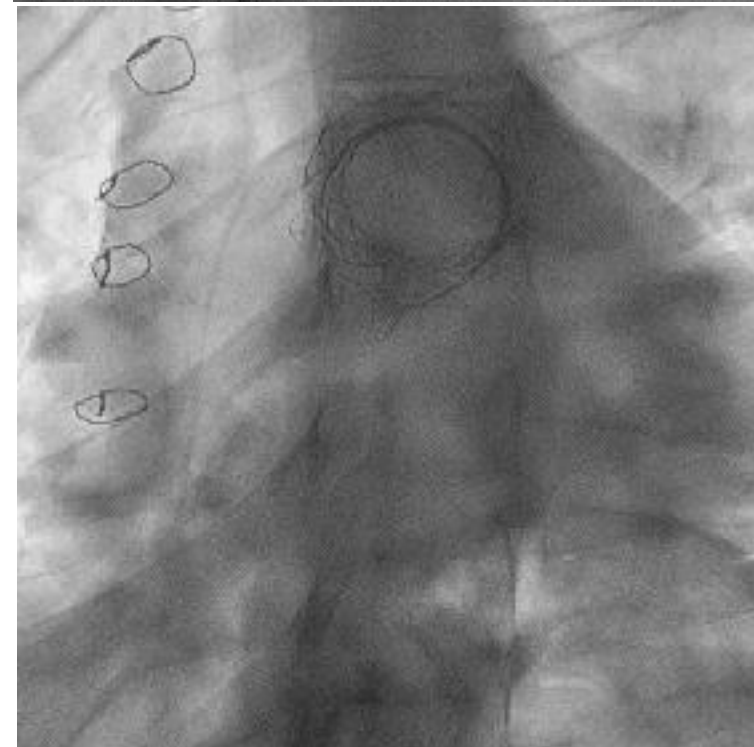
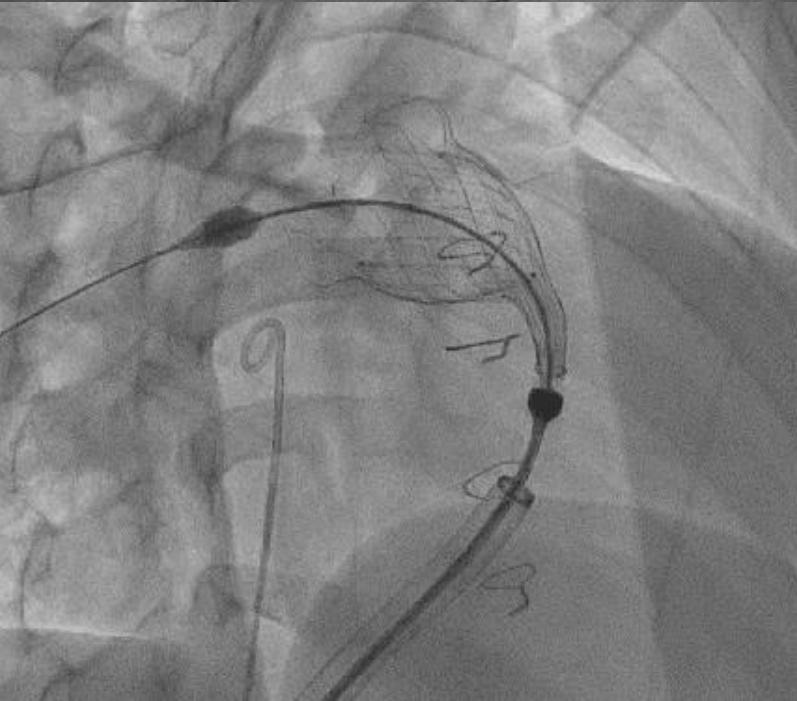
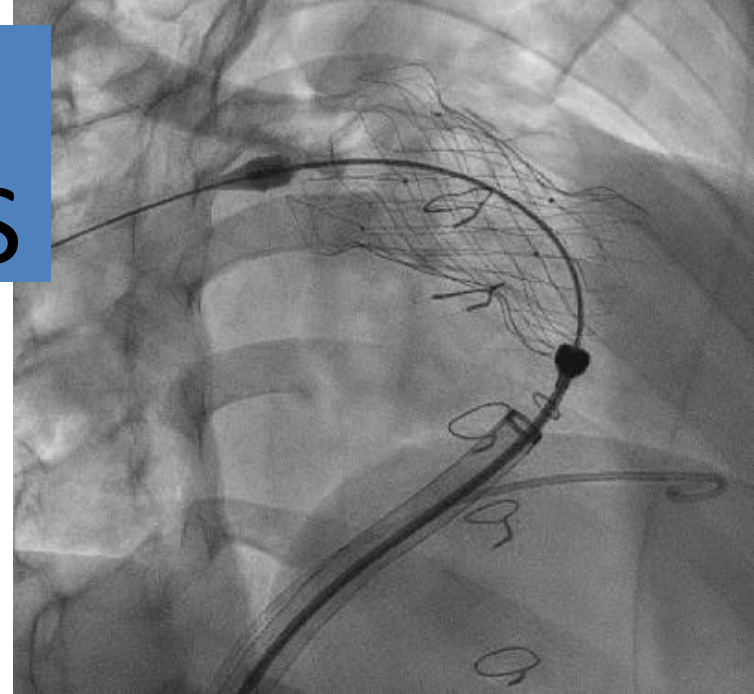
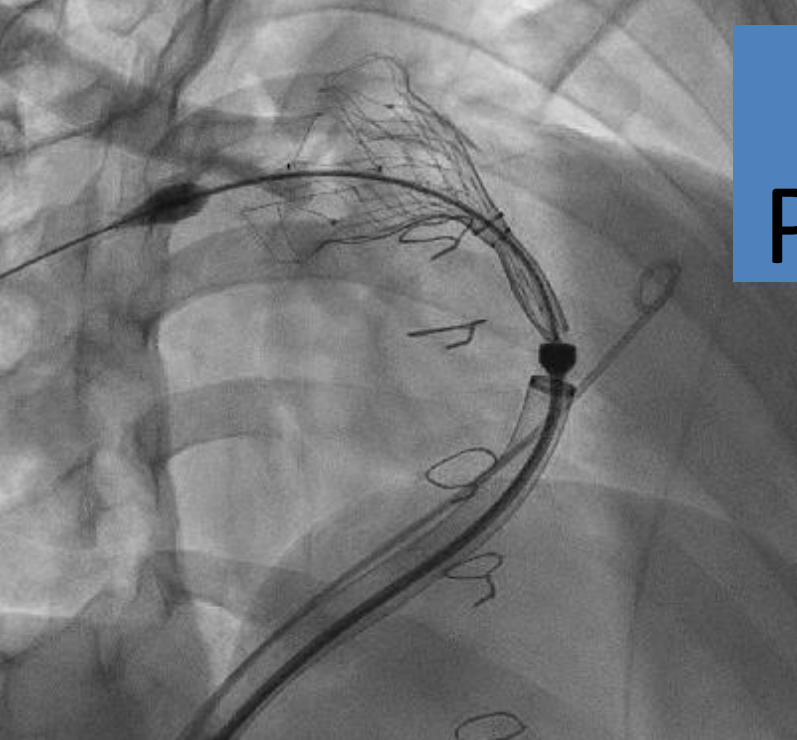
# BALLOON INTERROGATION

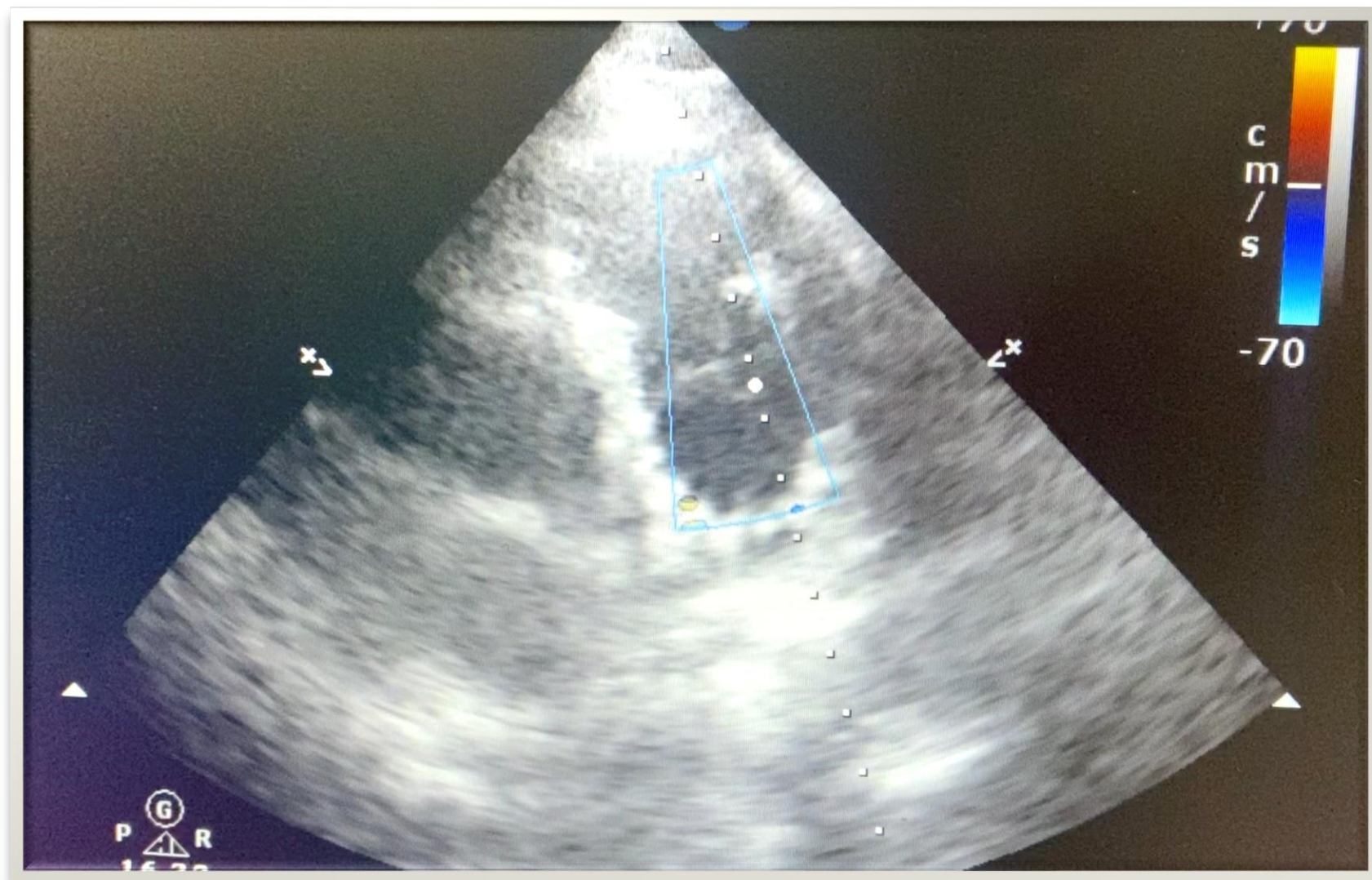


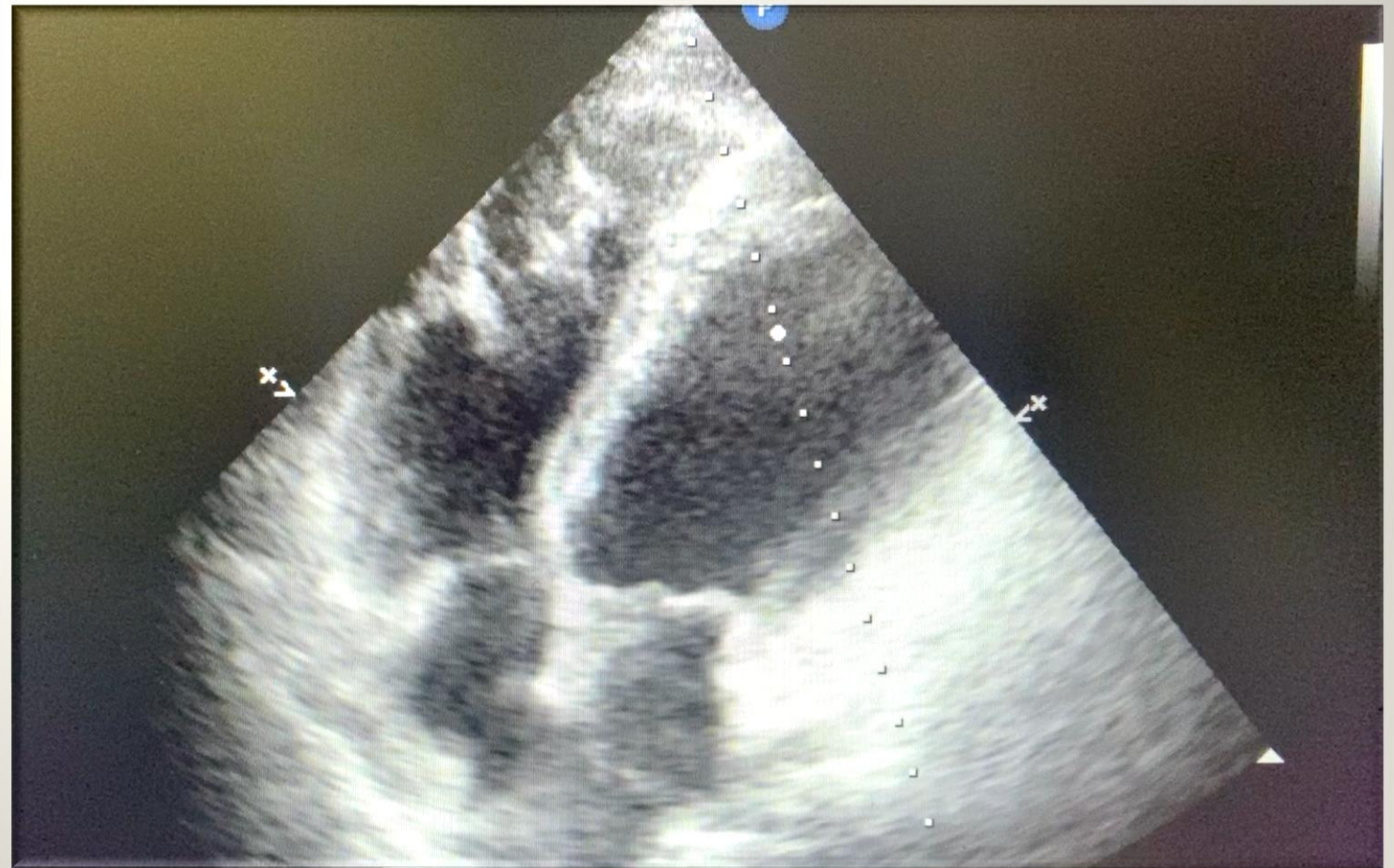
# PPVI P-VENUS



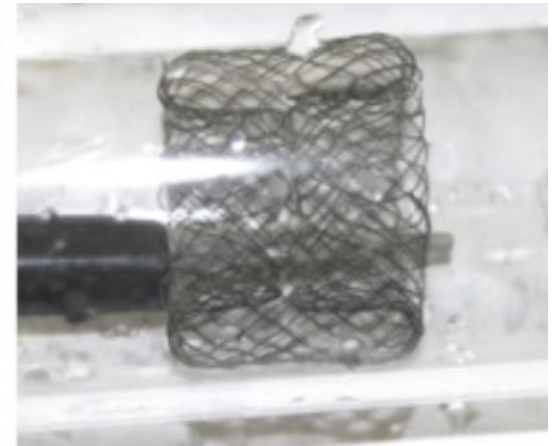
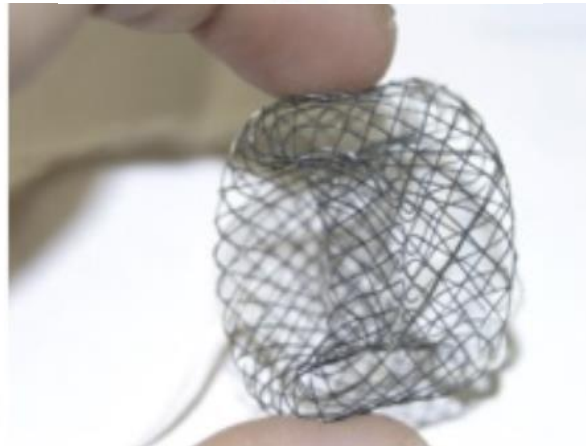
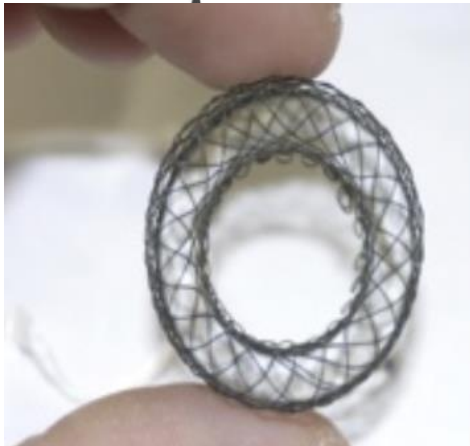
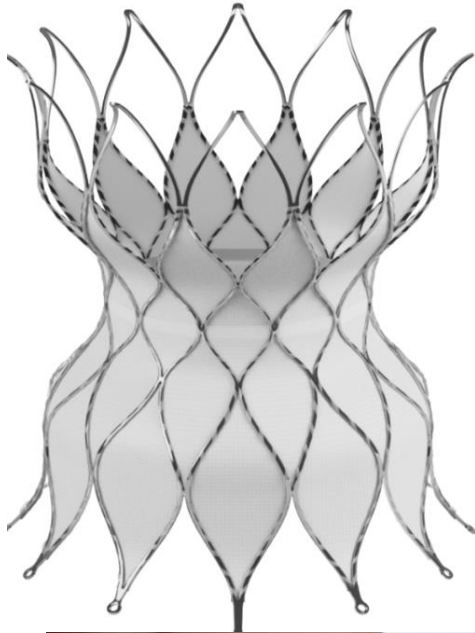
# PPVI P-VENUS







# SISTEMI IN STUDIO PER RVOT NATIVI



# COMPLICANZE

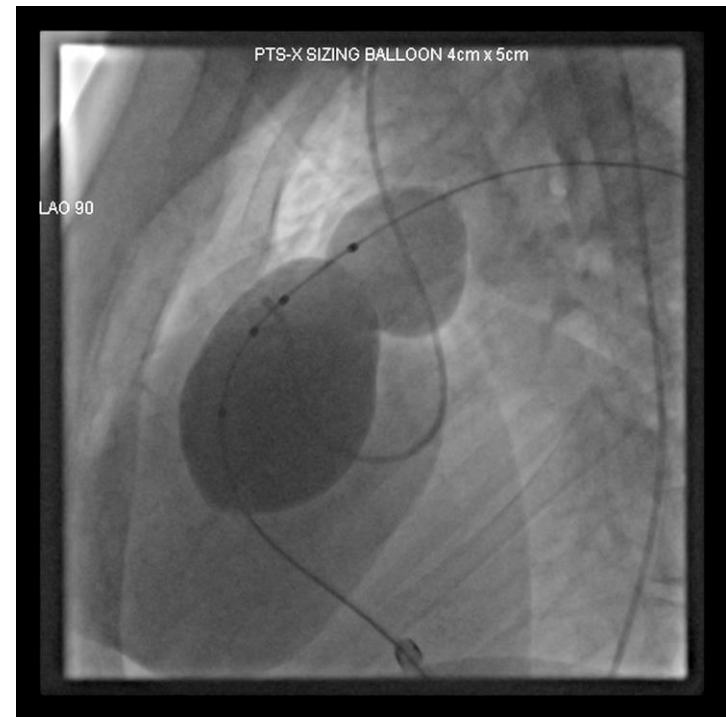
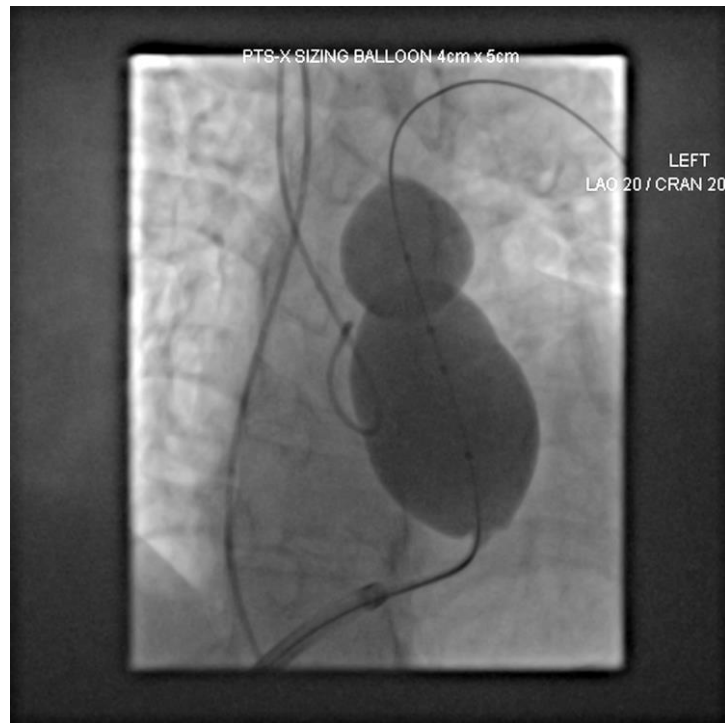
- Rottura condotti non complianti/calcificati (4->19% senza impatto emodinamico, 1% rischio vita, più frequente per homograft; più freq per  $\varnothing < 16\text{mm}$  16-50%)
- Compressione coronarica (5->2%). Attualmente più raro (CS da seno dx!!)
- Errato posizionamento/embolizzazione valvola
- Frattura stent (32% Melody senza prestant in efflusso nativo)-> Sapien non presenta questa problematica
- Danneggiamento valvola tricuspidale
- Mortalità 0.2-0.8%
- **Endocardite**

Hascoet S, et al. Infective endocarditis risk after percutaneous pulmonary valve implantation with the Melody and Sapien valves. **JACC Cardiovasc Interv.** 2017;10:510-517.

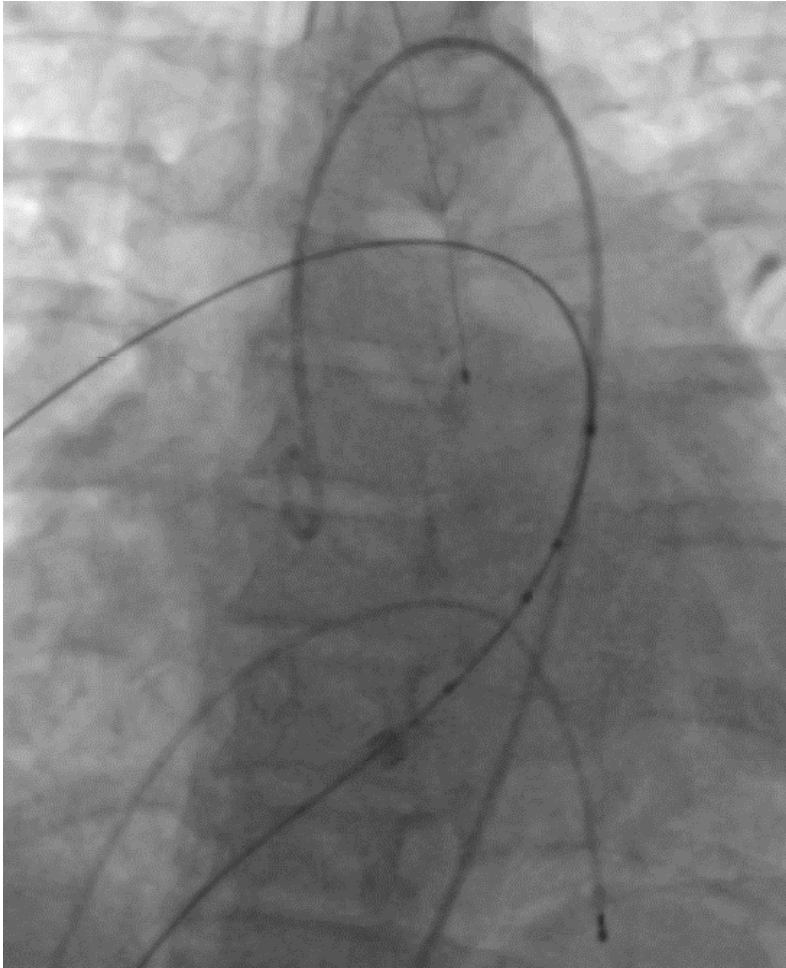
Lehner A et al. The risk of infective endocarditis following interventional pulmonary valve implantation: a meta-analysis. **J Cardiol.** 2019;74:197-205.

Murray BH et al. Risk of coronary artery compression among patients referred for transcatheter pulmonary valve implantation: a multicenter experience. **Circ Cardiovasc Interv** 2013; 6: 535-542

# COMPRESSIONE CORONARICA



# COMPRESSIONE RADICE AORTICA- INSUFFICIENZA AORTICA



# ENDOCARDITE

- Rischio più alto con **Melody** rispetto a Sapien
- **Alta incidenza endocardite**, soprattutto con Melody (3.1% per pz/anno, 7.5% a 2aa)
- **Fattori di rischio:** grad post TPVR>15mmHg ed età <12aa
- **Meccanismo** che causa endocardite: non noto con certezza!
- Rischio correlato al **tipo di valvola** ( valvola bovina vena giugulare vs pericardio porcino)
- Maggiore turbolenza sangue. Maggior capacità adesione batterica? su tessuto/valvola bovina?
- **Manipolazione** durante l'impianto? Danno cuspidi?
- **Condotto degenerato** aumenta rischio rispetto a efflusso nativo?

Hascoet S, et al. Infective endocarditis risk after percutaneous pulmonary valve implantation with the Melody and Sapien valves. **JACC Cardiovasc Interv.** 2017;**10**:510-517.

Lehner A et al. The risk of infective endocarditis following interventional pulmonary valve im- plantation: a meta-analysis. **J Cardiol.** 2019;**74**:197-205.



# 2020 ESC Guidelines for the management of adult congenital heart disease

## The Task Force for the management of adult congenital heart disease of the European Society of Cardiology (ESC)

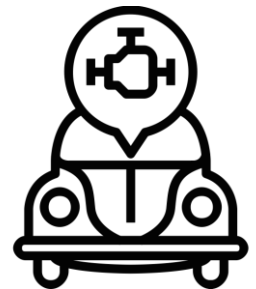
### Recommendations for intervention after repair of tetralogy of Fallot

| Recommendations                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                    | Class <sup>a</sup> | Level <sup>b</sup> |
|------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|--------------------|--------------------|
| PVRep is recommended in symptomatic patients with severe PR <sup>c</sup> and/or at least moderate RVOTO. <sup>d</sup>                                                                                                                                                                                                                                                                                                                                                                                                              | <b>I</b>           | <b>C</b>           |
| In patients with no native outflow tract, <sup>e</sup> catheter intervention (TPVI) should be preferred if anatomically feasible.                                                                                                                                                                                                                                                                                                                                                                                                  | <b>I</b>           | <b>C</b>           |
| PVRep should be considered in asymptomatic patients with severe PR and/or RVOTO when one of the following criteria is present. <ul style="list-style-type: none"><li>● Decrease in objective exercise capacity.</li><li>● Progressive RV dilation to RVESVi <math>\geq 80</math> mL/m<sup>2</sup>, and/or RVEDVi <math>\geq 160</math> mL/m<sup>2</sup> <sup>f</sup>, and/or progression of TR to at least moderate.</li><li>● Progressive RV systolic dysfunction.</li><li>● RVOTO with RVSP <math>&gt; 80</math> mmHg.</li></ul> | <b>IIa</b>         | <b>C</b>           |

# REVERSE REMODELING DEL VD ?

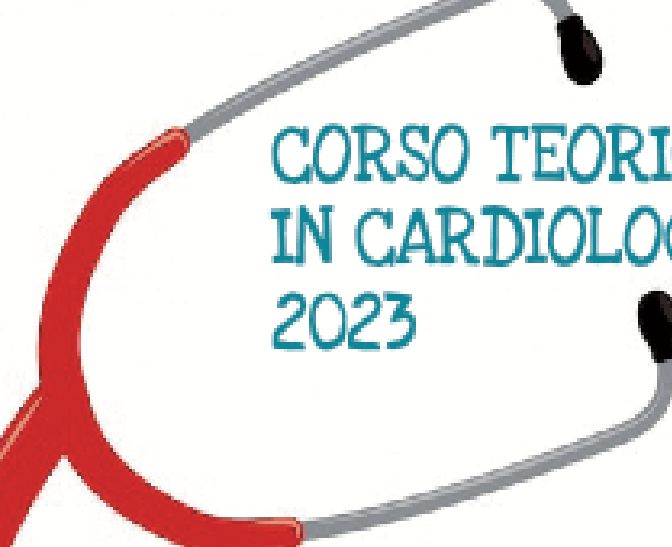
Concetto ancora attuale *cercare* la **normalizzazione FE** o  
realisticamente *sperare* “solo” nella **normalizzazione del volume**  
del VD?

- a) Difficile oltre **RVEDV**>160ml/m<sup>2</sup>
- b) Difficile oltre **RVESV**>80ml/m<sup>2</sup>
- c) **RVEF** <30% associato a prognosi negative



# CONCLUSIONI

- ❖ A 40 aa di follow-up mediamente il 35% dei soggetti è stato sottoposto a PVR, questo numero arriva al 58% se il paziente è attivamente seguito in centro GUCH
- ❖ Non esiste PVR ideale
- ❖ Ogni procedura porta con se un rischio di morbidità e mortalità che va aggregato alla mortalità della correzione iniziale
- ❖ Minimizzare il rischio di PVR è estremamente importante
- ❖ Un grado lieve ( $34 \pm 10$  mmHg) di ostruzione residua RVOT è protettiva per IPO tardiva e riduce volume RV e rischio di disfunzione RV



# CORSO TEORICO PRATICO IN CARDIOLOGIA PEDIATRICA 2023

I EDIZIONE  
Aprile-Giugno

II EDIZIONE  
Ottobre-Dicembre

RESPONSABILE SCIENTIFICO  
**Giuseppe Annoni**

COMITATO SCIENTIFICO  
Gabriella Agnoletti  
Roberto Bordesè  
Giuseppe Mazza  
Anna Maria Villar

SEDE

OSPEDALE INFANTILE REGINA MARGHERITA  
Piazza Polonia 94 - TORINO

*Corso di ecocardiografia clinica di II livello indirizzato a cardiologi dell'adulto, medici dello sport, pediatri, neonatologi e anestesisti che vogliono approfondire le loro competenze nelle cardiopatie congenite e nella gestione del neonato, del lattante e del bambino con problematiche cardiologiche congenite o acquisite. È richiesta una conoscenza dell'ecocardiografia di base e delle proiezioni ecocardiografiche standard.*

*Il corso, riservato a massimo 20 partecipanti per edizione, prevede un primo incontro teorico di una giornata in cui verranno trattate tutte le cardiopatie congenite semplici e una parte pratica di 40 ore complessive di frequenza del laboratorio di ecocardiografia, del reparto di cardiologia e neonatologia; un ultimo incontro teorico di mezza giornata chiuderà il corso.*

*Quota di iscrizione: 500 Euro + IVA comprensiva di materiale didattico, ticket lunch e conseguimento dei crediti ECM in seguito al superamento del test finale.*

*Accreditamento ECM: 50 crediti.*



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