Ambulatorio
Cardiopatie Congenite
dell'Adulto

IL LABORATORIO DI CARDIAC IMAGING NELLO SCOMPENSO CARDIACO

30-31 OTTOBRE 2015

Trieste

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Giorgio Faganello

Ambulatorio delle Cardiopatie Congenite dell'Adulto
Centro Cardiovascolare
Trieste

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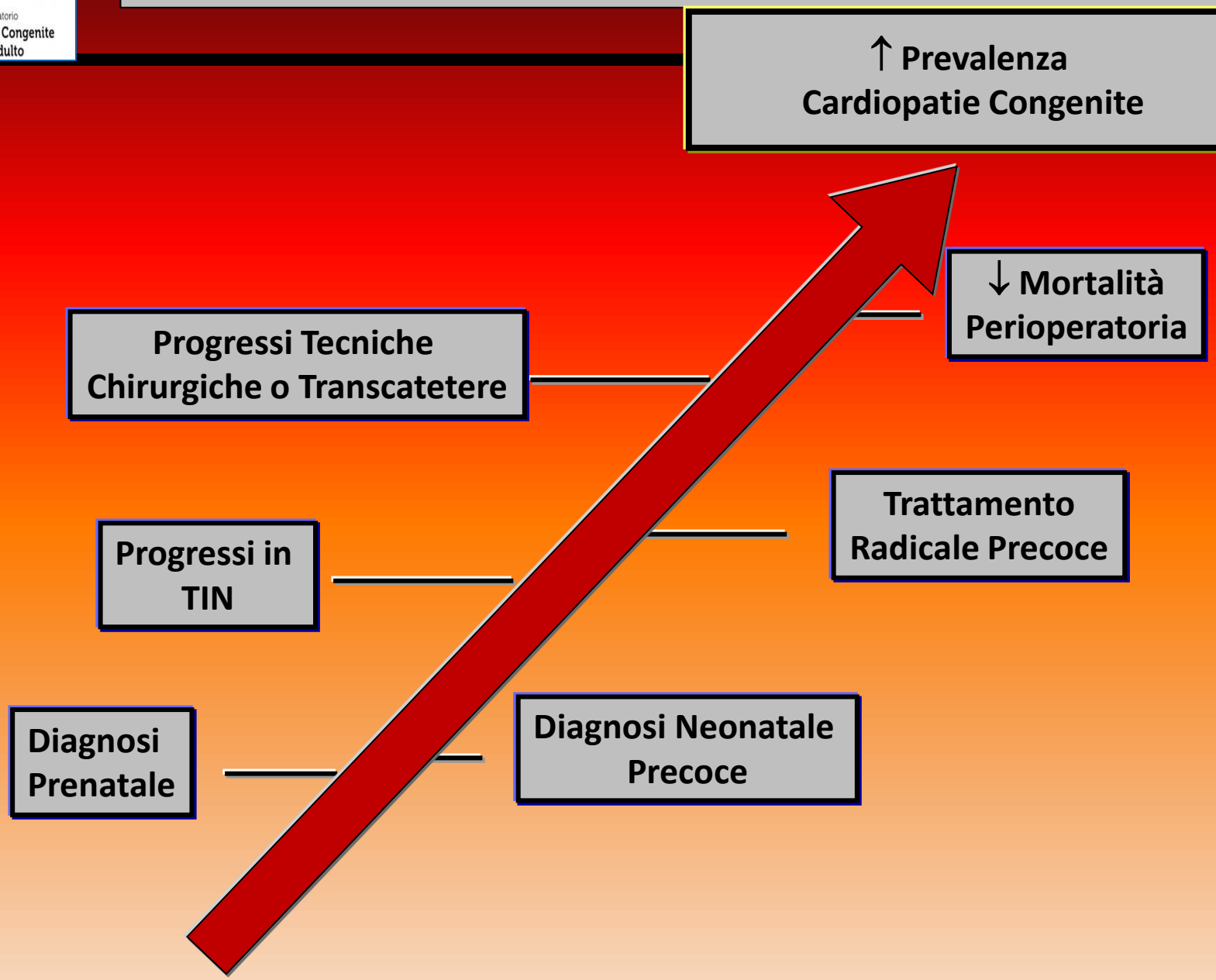
Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto



Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

CONGENITAL HEART DISEASE

Changing Mortality in Congenital Heart Disease

Paul Khairy, MD, PHD,* Raluca Ionescu-Ittu, MSC,†§ Andrew S. Mackie, MD, SM,†
Michal Abrahamowicz, PHD,§ Louise Pilote, MD, MPH, PHD,‡§ Ariane J. Marelli, MD†

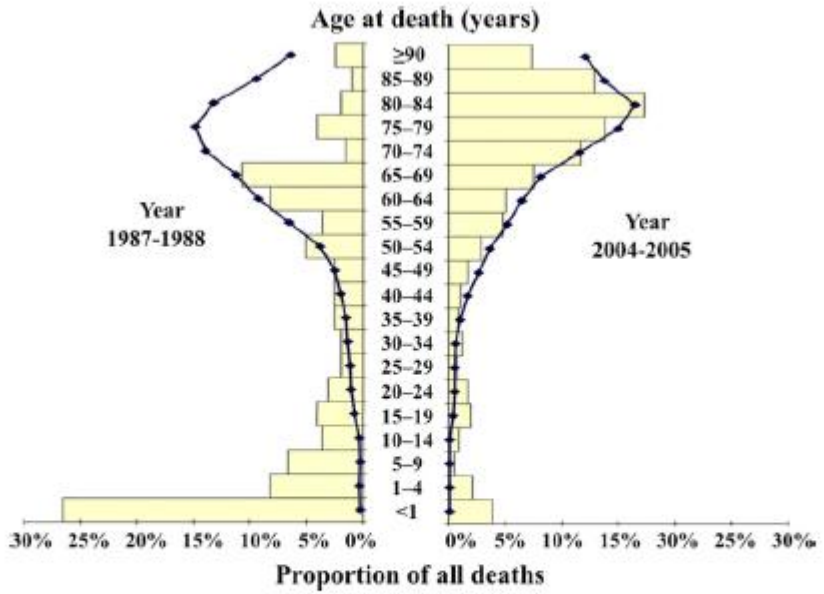


Figure 1 Distribution of Age at Death in Patients With Congenital Heart Disease in 1987 to 1988 and 2004 to 2005

Histogram bars depict the proportion of all deaths (x-axis) according to age at death (y-axis) in our cohort of patients with congenital heart disease in the first (1987 to 1988; **left**) and final (2004 to 2005; **right**) years of observation. **Bold black curves with diamonds** represent the corresponding age at death distribution in the general Quebec population during the same periods of observation.

J Am Coll Cardiol 2010;56:1149–57

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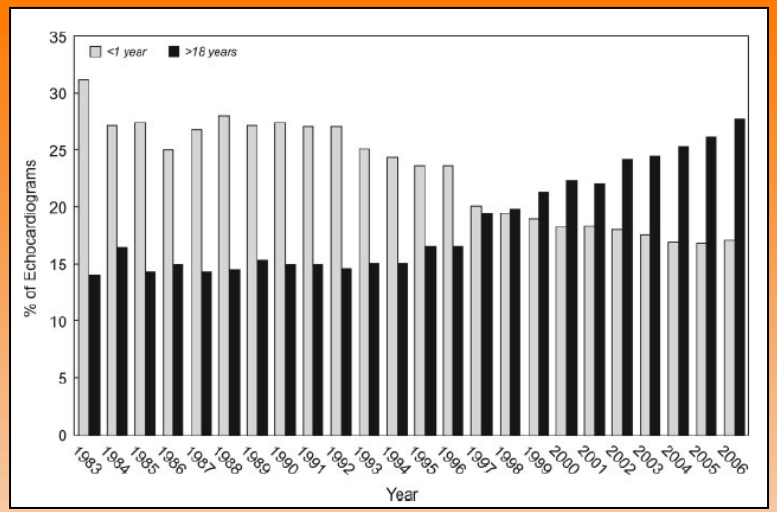
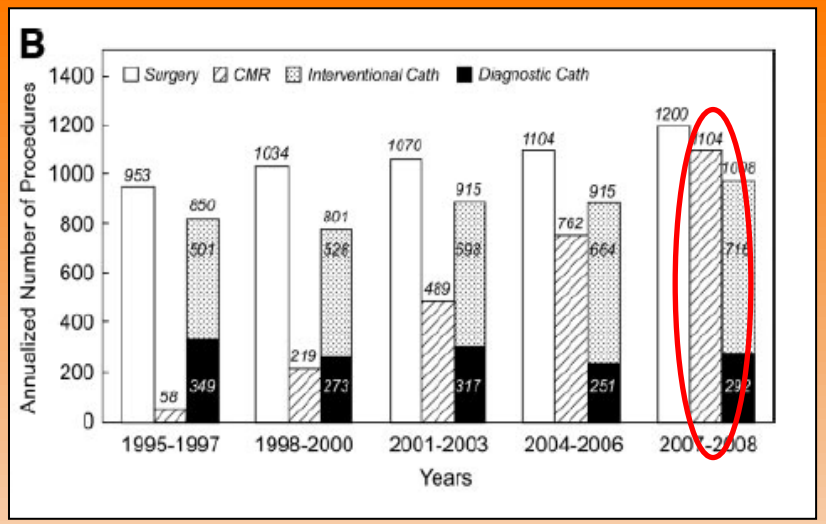
Advances in Cardiovascular Imaging

Multimodality Noninvasive Imaging for Assessment of Congenital Heart Disease

Ashwin Prakash, MD; Andrew J. Powell, MD; Tal Geva, MD

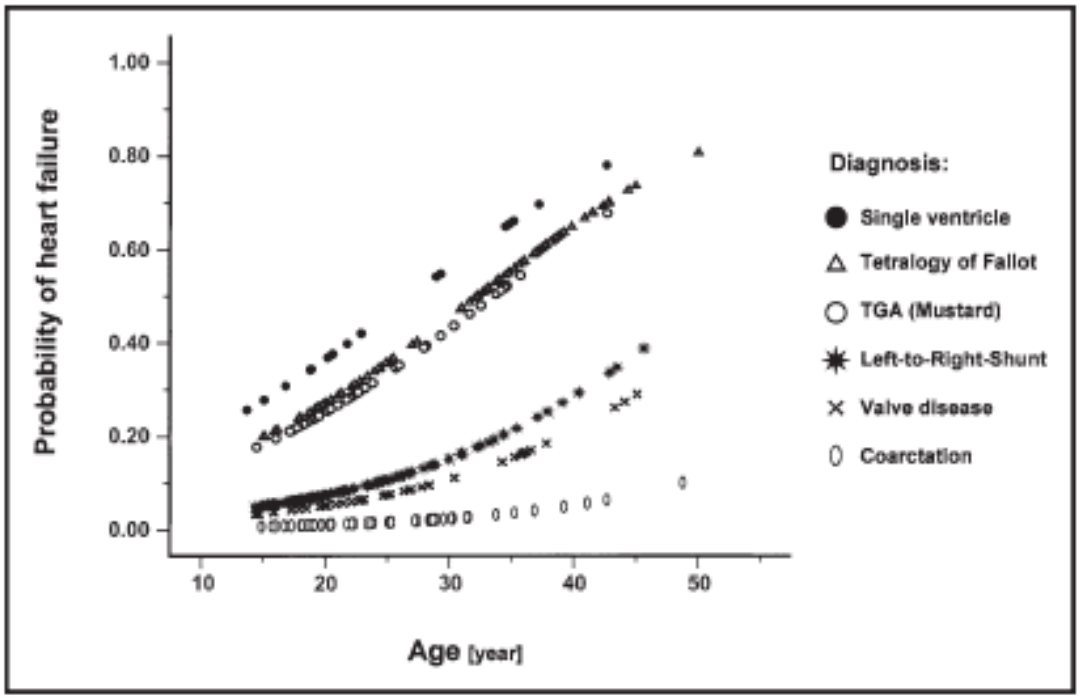




Circ Cardiovasc Imaging. 2010;3:112-125.

Incidence and Risk Distribution of Heart Failure in Adolescents and Adults With Congenital Heart Disease After Cardiac Surgery

Kambiz Norozi, MD[a,*], Armin Wessel, MD, PhD[a], Valentin Alpers[c], Jan Ole Arnhold[c], Siegfried Geyer, PhD[b], Monika Zoege, PhD[b], and Reiner Buchhorn, MD, PhD[c]

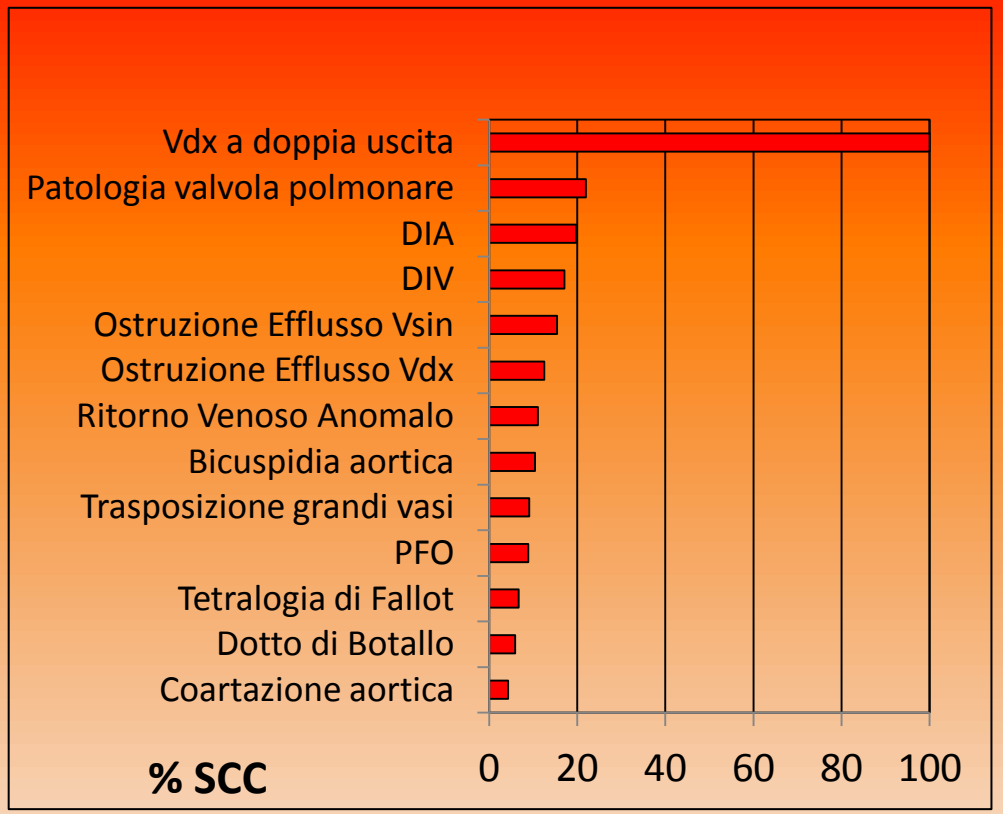


Am J Cardiol 2006;97:1238–1243

L'Osservatorio GUCH di Trieste

(11/2009-01/2015; n=492-2.3%; 13.8 % pz con SCC)

PATOLOGIA	PAZIENTI
	492
DIV-DIA	129
Ritorni vn anomali	14
Fallot	23
CoA	36
Botallo	24
TGA	16
CAV	11
Bicuspidia	150
Pat. Valv. Polmonare	39
St. sub-sopravalvolare Aortica	50



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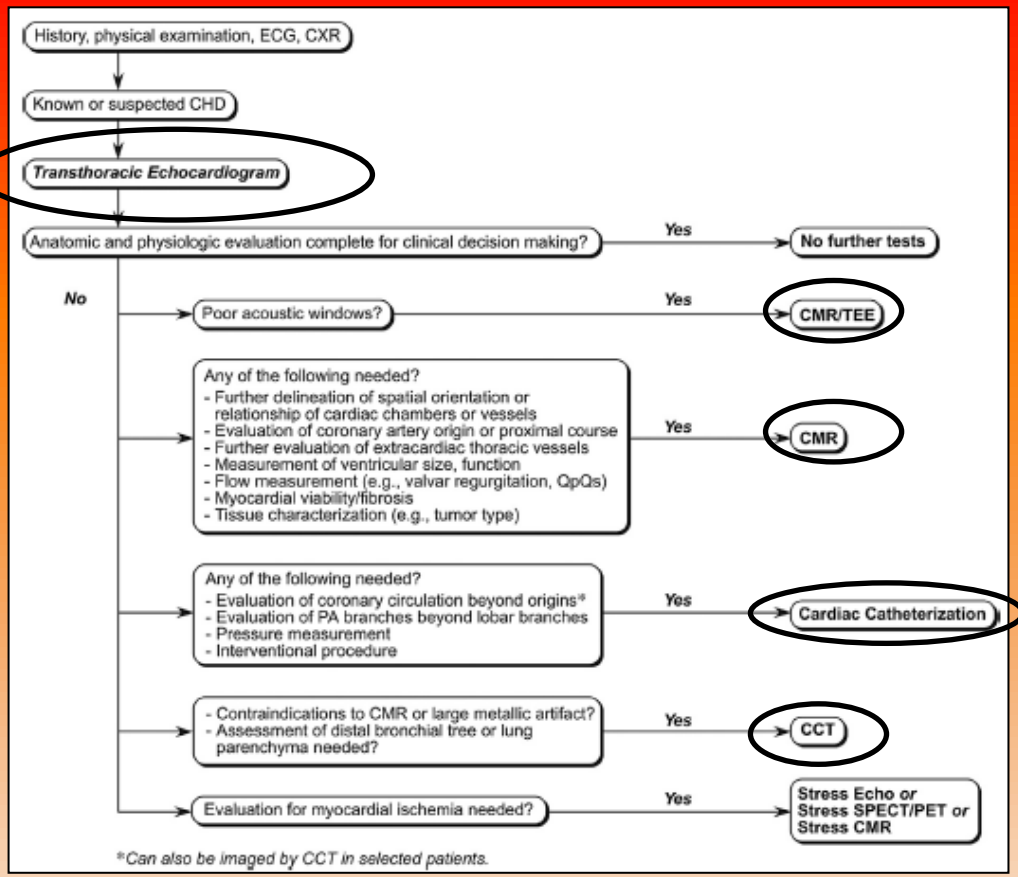
Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Canadian Journal of Cardiology 29 (2013) 830–840

Review

New Directives in Cardiac Imaging: Imaging the Adult With Congenital Heart Disease



Ventricolo destro in (normale) posizione sottopolmonare

TOF

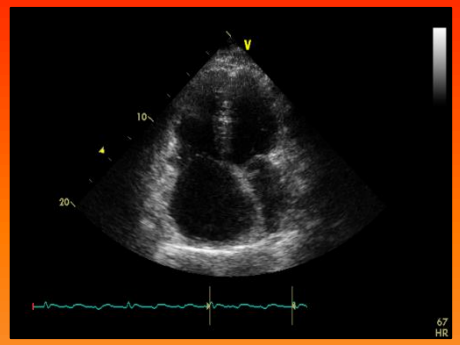
Ventricolo destro sistemico

TGA

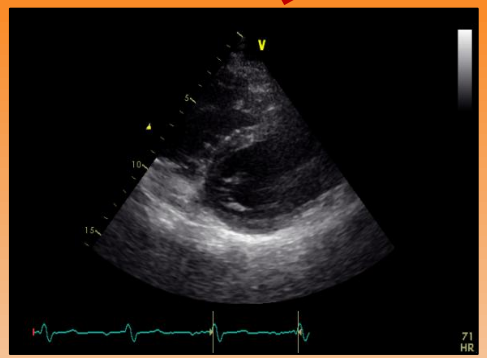
Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Tetralogia di Fallot

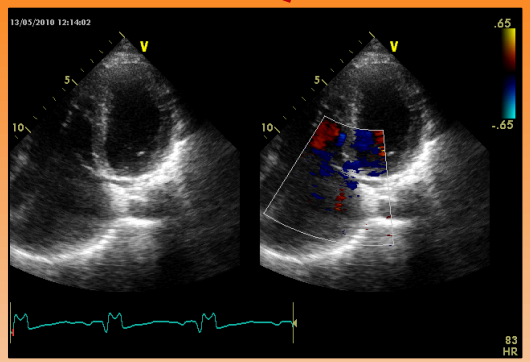
Aspetti ecocardiografici delle complicanze più frequenti



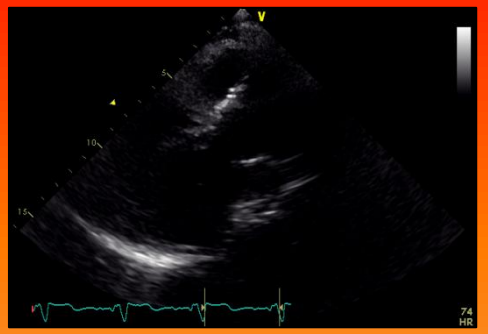
Dilatazione/disfunzione Ventricolo dx



Appiattimento del setto IV da sovraccarico di volume/pressione



Dilatazione anello tricuspide con comparsa di IT



Dilatazione anello aortico ed aorta ascendente

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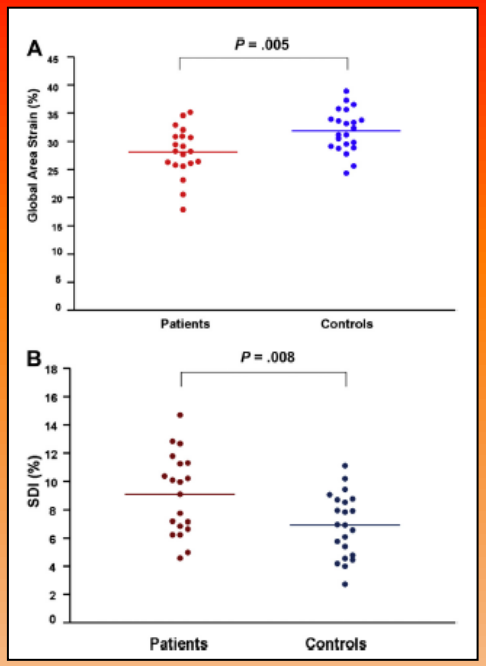
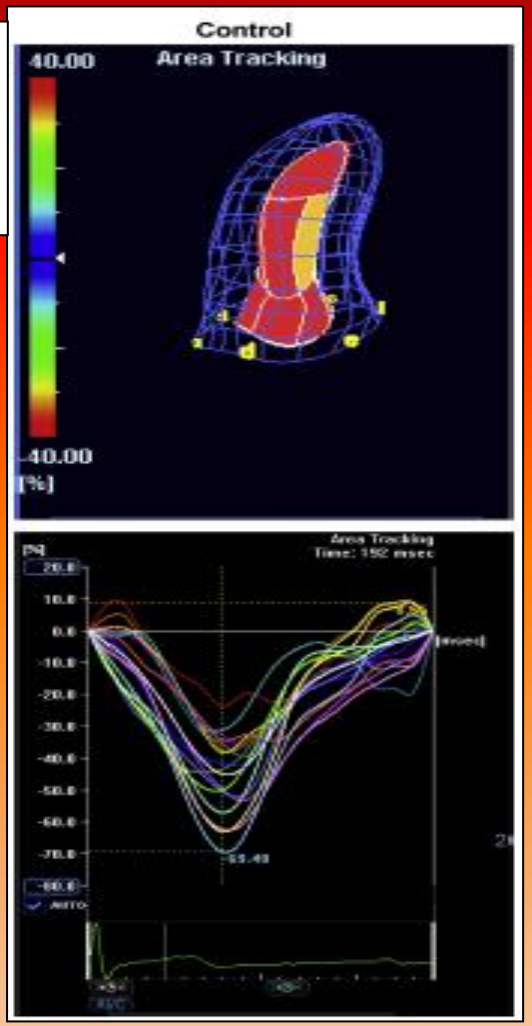
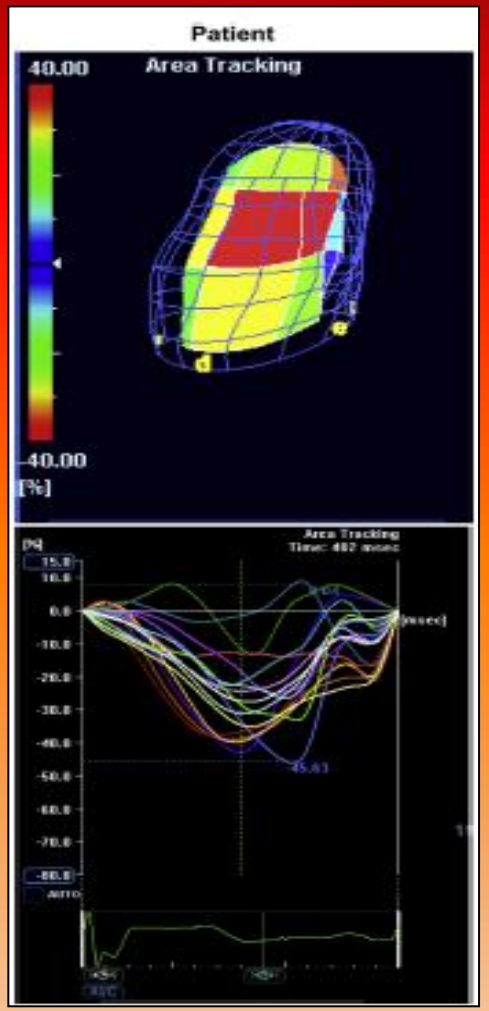
Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Right Ventricular Mechanics in Adults after Surgical Repair of Tetralogy of Fallot: Insights from Three-Dimensional Speckle-Tracking Echocardiography







Conclusions: In adults after tetralogy of Fallot repair, 3D RV deformation is impaired in association with RV dyssynchrony, volume overloading, and reduced EF. (J Am Soc Echocardiogr 2014;27:423-9.)

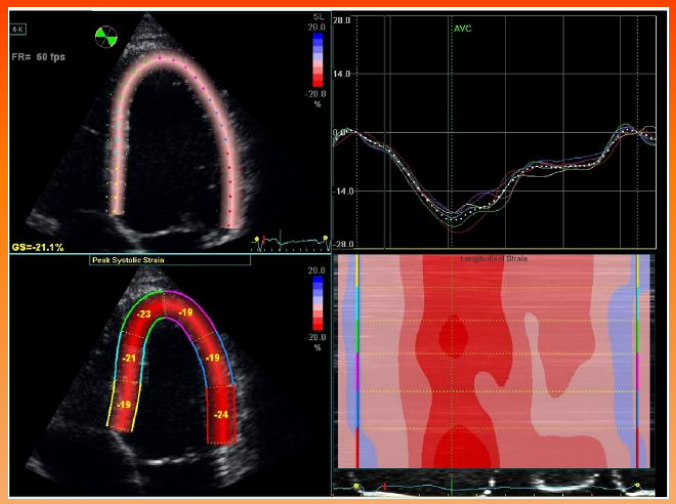
Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Right ventricular–left ventricular interaction in adults with Tetralogy of Fallot: A combined cardiac magnetic resonance and echocardiographic speckle tracking study☆



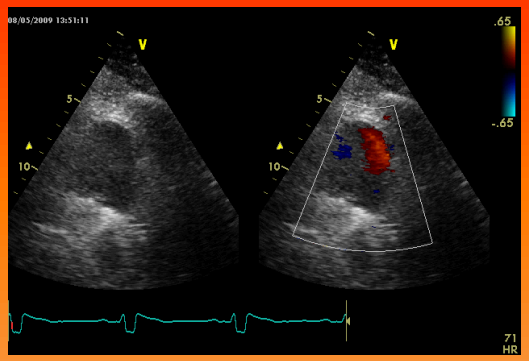
TOF.
LVEF normale….. tuttavia
LV longitudinal strain ridotto
-> SUBCLINICAL LV MYOCARDIAL DAMAGE

Conclusion: Left and right ventricular function both by CMR and speckle-tracking is interrelated in adults with repaired ToF. Despite normal LV ejection fraction, 2D longitudinal strain is significantly reduced in ToF patients, suggesting subclinical LV myocardial damage. Considering the potential prognostic value of LV dysfunction in ToF, this measurement may gain importance and should be included in future outcome studies.

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

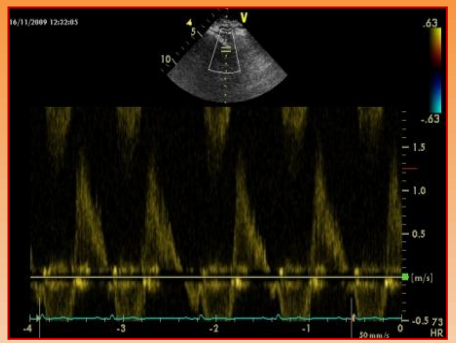
Tetralogia di Fallot

Aspetti ecocardiografici delle complicanze più frequenti

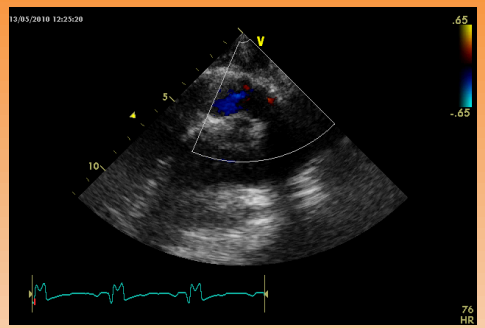


Aumentato rischio di
Endocardite Infettiva

Stenosi polmonare



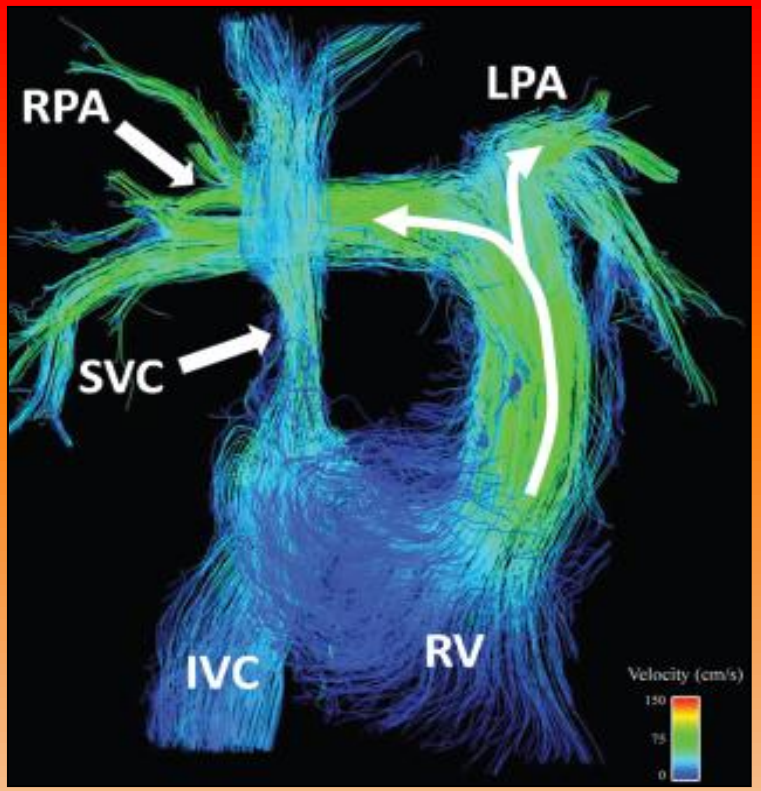
Insufficienza della valvola polmonare

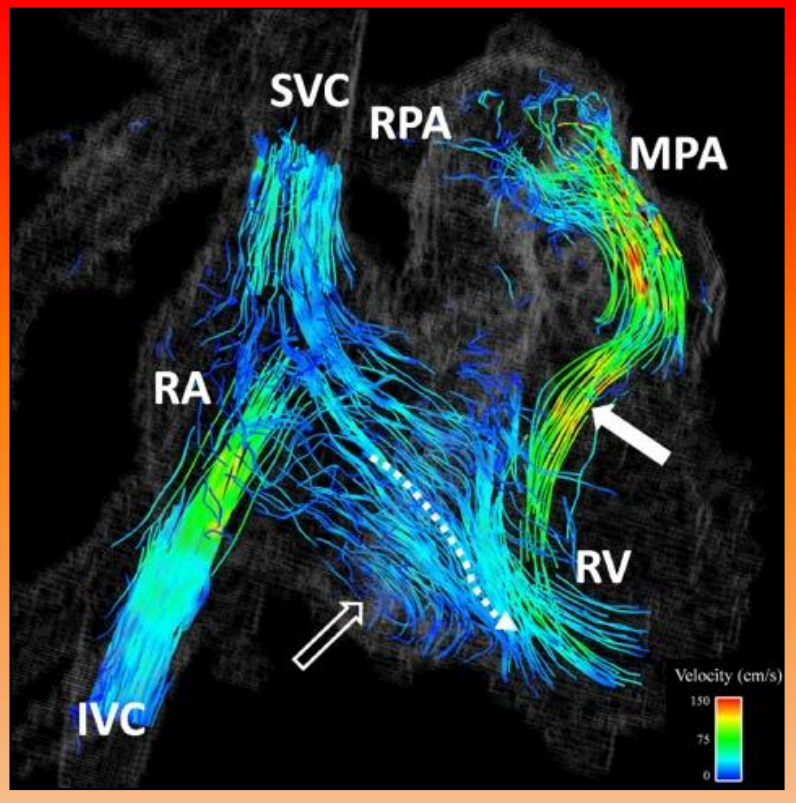


Progressione della stenosi polmonare

Tetralogia di Fallot

INSUFFICIENZA POLMONARE





MRI – FUNZIONE VENTRICOLARE E FLUSSIMETRICA

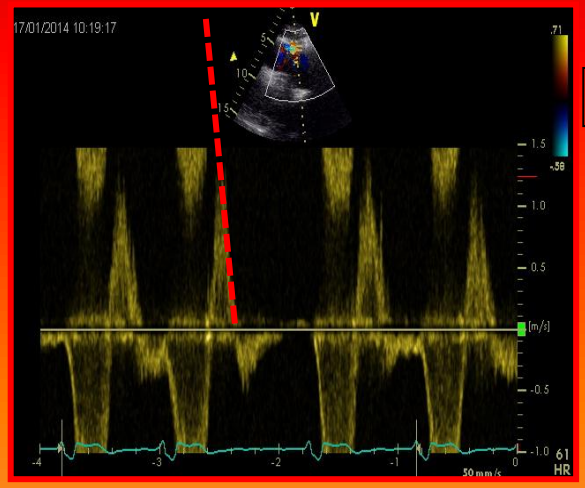
TOF e 4D CMR

JCMR 2012

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

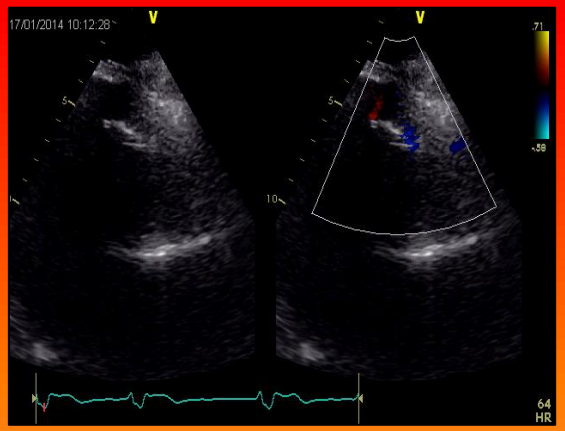
Tetralogia di Fallot

INSUFFICIENZA POLMONARE



PHT <100 msec

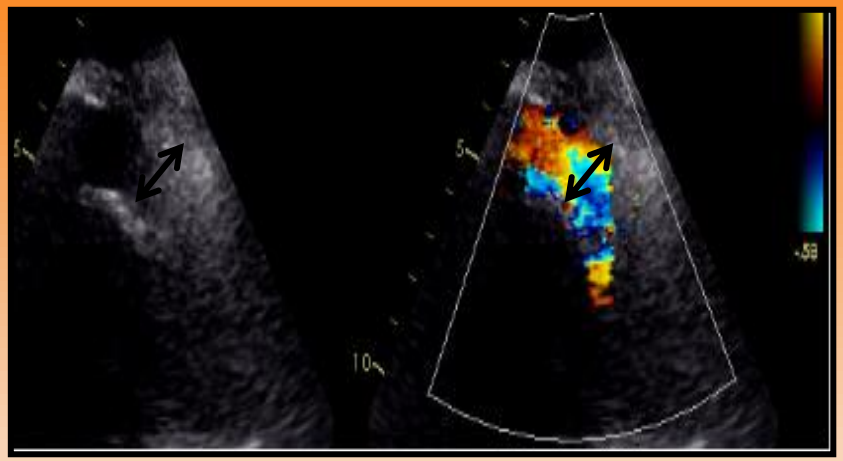
JASE 2003; 16:1057-62



JET PR >50-65% RVOT

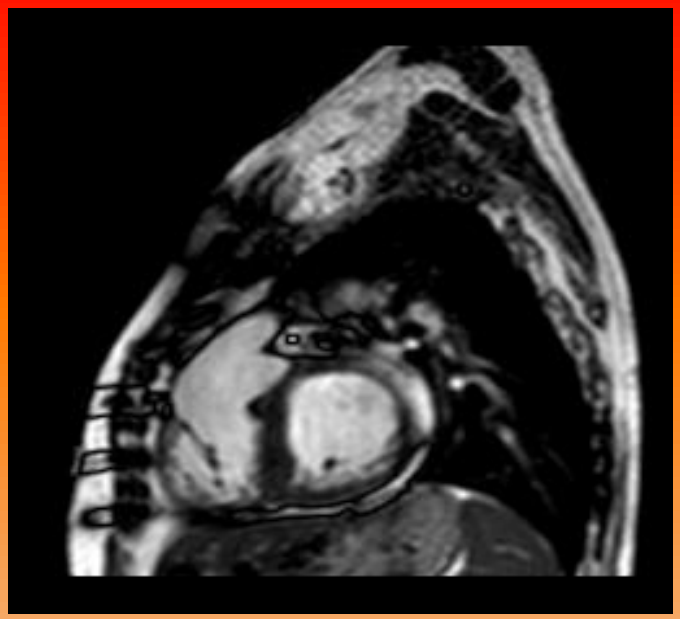
PR < 50% DIASTOLE

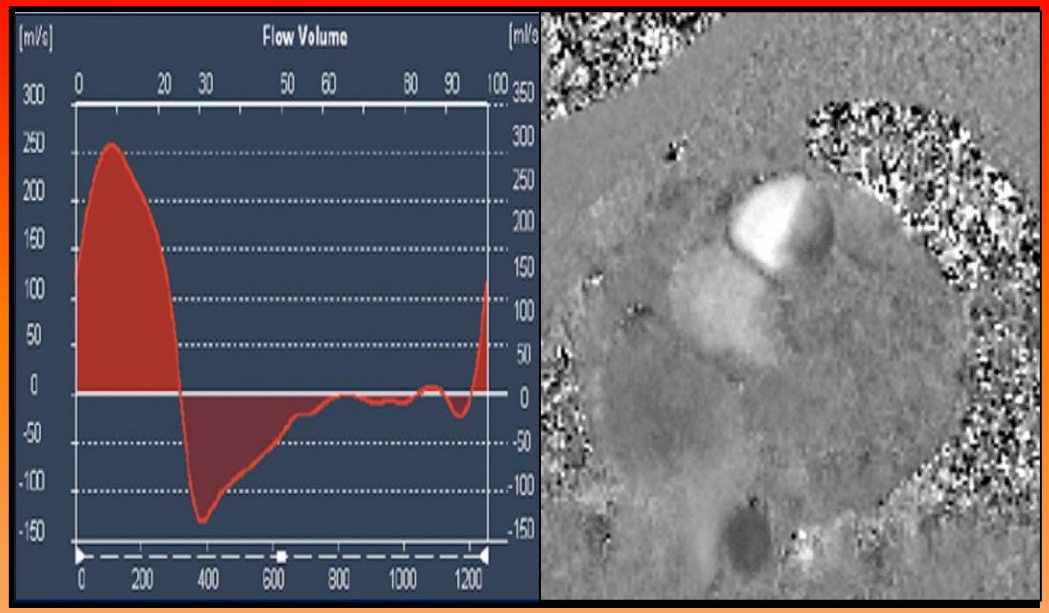
Am Heart J. 2004



Tetralogia di Fallot

INSUFFICIENZA POLMONARE





Sequenze SSFP

Sequenze phase contrast

Frazione rigurgitante 40%

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

Ventricular size and function assessed by cardiac MRI predict major adverse clinical outcomes late after tetralogy of Fallot repair

A L Knauth,[1,2] K Gauvreau,[1] A J Powell,[1] M J Landzberg,[1,2] E P Walsh,[1] J E Lock,[1] P J del Nido,[3] T Geva[1]

Heart 2008;**94**:211–216.

88 TOF
Età media alla riparazione: 3 yrs
FuP 4.2 yrs

Outcomes:
morte, TVNS, incremento classe funzionale a NYHA III-IV

Eventi maggiori: 18 pts (20.5%)

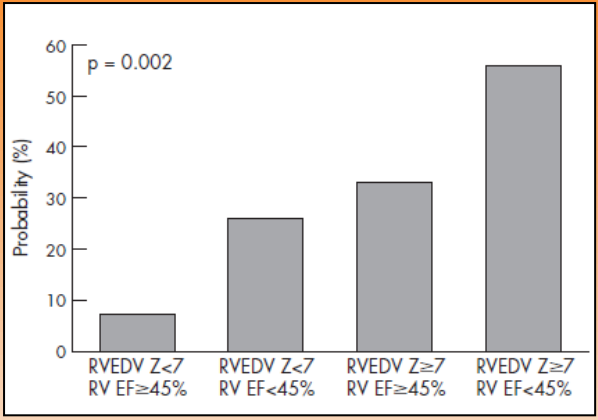


Table 2 Predictors of major adverse outcomes

Predictors	Odds ratio (95% CI)	Area under ROC curve	p Value
Univariate analysis (controlling for time from baseline evaluation to most recent follow-up)			
Age at TOF repair ⩾6 years	7.78 (2.26 to 26.7)	0.748	0.001
Era of repair before 1970	4.05 (1.11 to 14.8)	0.591	0.035
RV end-diastolic volume Z ⩾7	4.98 (1.47 to 16.9)	0.688	0.01
RV end-systolic volume index >50 ml/m²	5.82 (1.22 to 27.7)	0.708	0.027
RV ejection fraction <45%	5.31 (1.59 to 17.8)	0.750	0.007
LV ejection fraction <55%	7.13 (2.11 to 24.0)	0.760	0.002
LV mass/volume ratio >1.5	4.56 (1.27 to 16.4)	0.688	0.02
QRS ⩾180 ms	6.27 (1.86 to 21.1)	0.673	0.003
Diuretics at baseline evaluation	7.68 (2.03 to 29.1)	0.646	0.003
Digoxin at baseline evaluation	4.67 (1.30 to 16.8)	0.689	0.018
β-Blocker at baseline evaluation	6.88 (1.12 to 42.3)	0.654	0.037
NYHA class II or III at baseline evaluation	5.27 (1.55 to 17.9)	0.724	0.008
Multivariate analysis (controlling for time from baseline evaluation to most recent follow-up)			
Model 1			
LV ejection fraction <55%	8.05 (2.14 to 30.2)		0.002
RV end-diastolic volume Z ⩾7	4.55 (1.10 to 18.8)		0.037
	Area under ROC curve for the model: 0.850		
Model 2			
RV ejection fraction <45%	5.60 (1.47 to 21.2)		0.011
RV end-diastolic volume Z ⩾7	4.00 (1.10 to 14.6)		0.036
	Area under ROC curve for the model: 0.807		

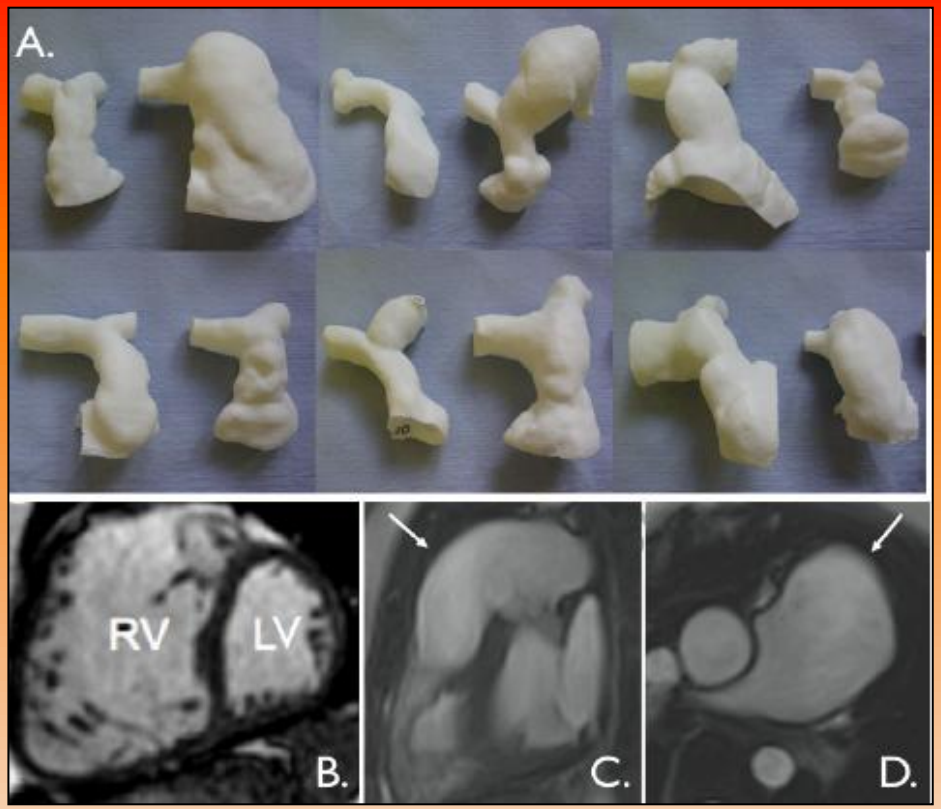
CI, confidence interval; LV, left ventricular; ROC, receiver operator characteristic; RV, right ventricular; TOF tetralogy of Fallot.

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

REVIEW Open Access

The Role of Cardiovascular Magnetic Resonance in Pediatric Congenital Heart Disease

Hopewell N Ntsinjana, Marina L Hughes and Andrew M Taylor*



Journal of Cardiovascular Magnetic Resonance 2011, **13**:51

Ventricolo destro in (normale) posizione sottopolmonare

TOF

Ventricolo destro sistemico

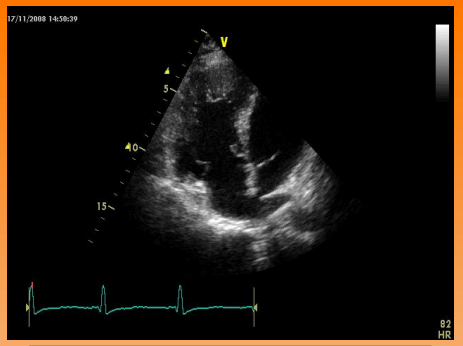
TGA (Mustard/Senning)

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

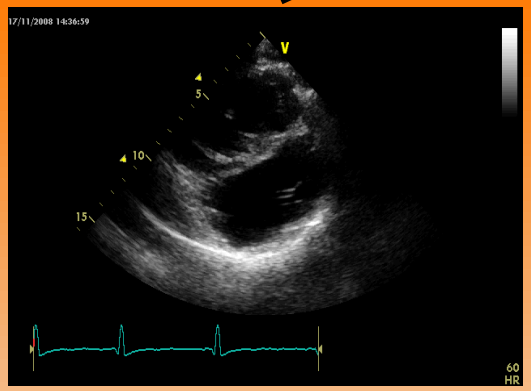
Trasposizione dei grandi vasi

Operata con tecnica dello switch atriale sec Mustard o Senning

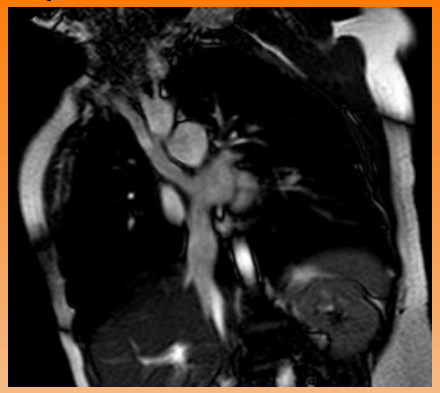
Aspetti ecocardiografici delle complicanze più frequenti



Disfunzione del ventricolo sistemico



Appiattimento SIV da sovraccarico di pressione



Stenosi dei ritorni venosi sistemici o polmonari

Rigurgito valvola atrio-ventricolare sistemica

Ambulatorio Cardiopatie Congenite dell'Adulto

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

European Heart Journal (2014) **35**, 1666–1674
doi:10.1093/eurheartj/ehu102

CLINICAL RESEARCH
Congenital heart disease

The natural and unnatural history of the Mustard procedure: long-term outcome up to 40 years

Judith A.A.E. Cuypers[1]*, Jannet A. Eindhoven[1], Maarten A. Slager[1], Petra Opić[1], Elisabeth M.W.J. Utens[2], Willem A. Helbing[3], Maarten Witsenburg[1], Annemien E. van den Bosch[1], Mohamed Ouhlous[4], Ron T. van Domburg[1], Dimitris Rizopoulos[5], Folkert J. Meijboom[6], Ad J.J.C. Bogers[7], and Jolien W. Roos-Hesselink[1]

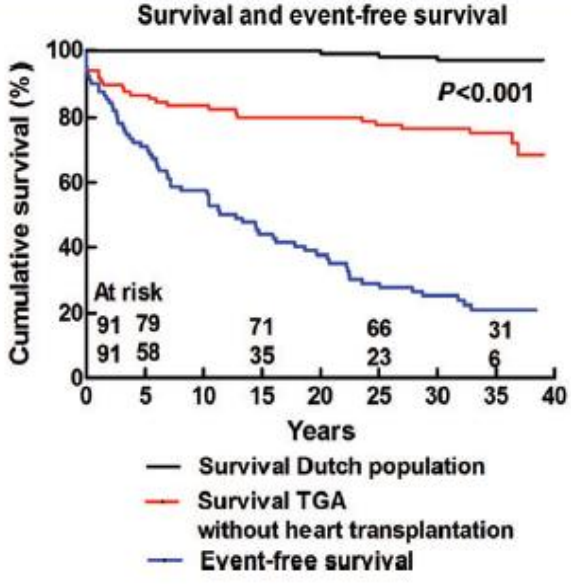


Table 4 Predictors for clinical endpoints; all-cause mortality, reinterventions, heart failure, and arrhythmias

Endpoint	Univariable model			Multivariable model		
	HR	CI	P-value	HR	CI	P-value
All-cause mortality						
Early postoperative arrhythmias	2.04	(0.76–5.50)	0.158	1.50	(0.55–4.10)	0.432
Temperature during surgery	1.15	(1.04–1.26)	**0.004**	1.05	(0.95–1.17)	0.328
Complex or simple TGA	0.99	(0.43–2.25)	0.972	1.00	(0.44–2.30)	0.995
Age at operation	1.11	(0.95–1.28)	0.182	0.97	(0.80–1.17)	0.738
Operated before 1977	4.26	(1.44–12.60)	**0.009**	2.89	(1.00–8.33)	**0.049**
Reintervention						
Early postoperative arrhythmias	0.50	(0.12–2.09)	0.343	1.23	(0.62–0.36)	0.739
Temperature during surgery	0.97	(0.88–1.08)	0.589	1.03	(0.90–1.18)	0.622
Complex or simple TGA	1.35	(0.64–2.86)	0.428	1.33	(0.63–2.81)	0.450
Age at operation	0.88	(0.73–1.08)	0.239	0.99	(0.77–1.28)	0.944
Operated before 1977	0.37	(0.18–0.78)	**0.008**	0.34	(0.14–0.84)	**0.020**
Heart failure						
Early postoperative arrhythmias	3.00	(0.81–11.12)	0.100	8.13	(2.15–30.72)	**0.002**
Temperature during surgery	1.02	(0.89–1.18)	0.760	0.92	(0.77–1.11)	0.393
Complex or simple TGA	0.70	(0.22–2.19)	0.695	1.26	(0.38–4.19)	0.706
Age at operation	1.23	(1.01–1.50)	**0.038**	1.26	(1.01–1.56)	**0.041**
Operated before 1977	1.22	(0.39–3.86)	0.730	0.57	(0.15–2.07)	0.391
Arrhythmias						
Early postoperative arrhythmias	4.62	(1.96–10.90)	**<0.001**	3.82	(1.54–9.49)	**0.004**
Temperature during surgery	1.11	(1.02–1.21)	**0.012**	1.05	(0.95–1.17)	0.331
Complex or simple TGA	1.12	(0.53–2.36)	0.765	1.42	(0.65–3.09)	0.378
Age at operation	1.18	(1.03–1.35)	**0.019**	1.09	(0.91–1.29)	0.356
Operated before 1977	1.95	(0.96–3.98)	0.065	1.10	(0.48–2.51)	0.829

Conclusion Long-term survival after Mustard repair is clearly diminished and morbidity is substantial. Early postoperative arrhythmias are a predictor for heart failure and late arrhythmias.

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

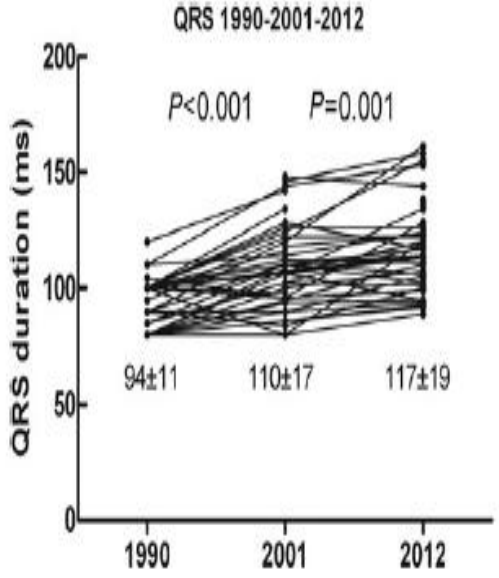
European Heart Journal (2014) **35**, 1666–1674
doi:10.1093/eurheartj/ehu102

CLINICAL RESEARCH
Congenital heart disease

The natural and unnatural history of the Mustard procedure: long-term outcome up to 40 years

Judith A.A.E. Cuypers[1]*, Jannet A. Eindhoven[1], Maarten A. Slager[1], Petra Opić[1], Elisabeth M.W.J. Utens[2], Willem A. Helbing[3], Maarten Witsenburg[1], Annemien E. van den Bosch[1], Mohamed Ouhlous[4], Ron T. van Domburg[1], Dimitris Rizopoulos[5], Folkert J. Meijboom[6], Ad J.J.C. Bogers[7], and Jolien W. Roos-Hesselink[1]

	1990[10]	2001[3]	2012	*P*-value*	
	n = 58	*n* = 54	*n* = 50	2012 vs. 1990	2012 vs. 2001
Echocardiogram	*n* = 58	*n* = 53	*n* = 47		
RV systolic function normal	40 (69%)	3 (6%)	1 (2%)	**<0.001**	0.3
Valve regurgitation (>trace)					
Aortic regurgitation	4 (7%)	5 (9%)	7 (15%)	**0.03**	0.2
Mitral regurgitation	8 (14%)	12 (23%)	11 (23%)	0.3	0.7
Pulmonary regurgitation	15 (26%)	27 (51%)	27 (57%)	**<0.001**	0.3
Tricuspid regurgitation	36 (62%)	45 (85%)	43 (92%)	**0.001**	0.2
Severe tricuspid regurgitation	1 (2%)	10 (19%)	17 (38%)	**<0.001**	0.1
Mitral regurgitation vmax (m/s)	2.9	2.5	2.8	0.7	0.4
Pulmonary regurgitation vmax (m/s)	1.9	1.9	2.0	0.1	0.9



	Mean ± SD	IQR	Abnormal (%)[a]
Echocardiography			
MR vmax (m/s)	2.9 ± 0.8	(0.2–3.4)	50
PR vmax (m/s)	1.9 ± 0.7	(1.4–2.50)	26
RV annulus (mm)	50 ± 7	(45–55)	94
RV apex-base (mm)	81 ± 10	(74–87)	35
TAPSE (mm)	13 ± 3	(11–15)	89
RV FAC (%)	25 ± 10	(19–30)	79
TDI S′ tricuspid annulus	7.5 ± 1.5	(6.3–8.5)	97
IVC collapse >50% (*n*, %)	33 (77%)		
Cardiac magnetic resonance imaging			
	n = 24		
LV EDV/BSA (mL/m^2)	60 (37–89)		
LV ESV/BSA (mL/m^2)	21 (10–46)		
LV EF (%)	63 (43–76)		
RV EDV/BSA (mL/m^2)	93 (50–145)		
RV ESV/BSA (mL/m^2)	41 (17–78)		
RV EF (%)	47 (28–71)		

Lo Scompenso Cardiaco nella cardiopatia congenita dell'adulto

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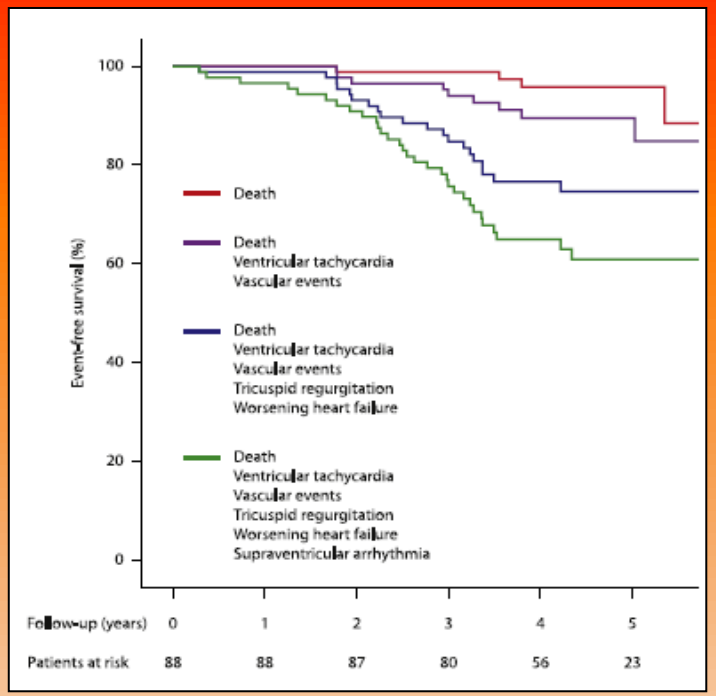
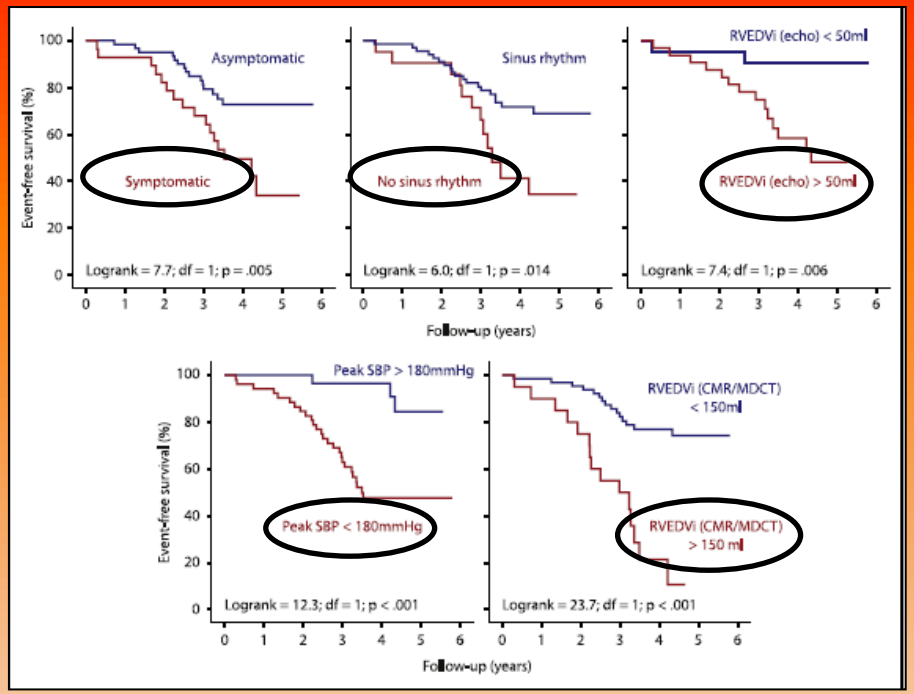
Adult Congenital Heart Disease

Right Ventricular End-Diastolic Volume Combined With Peak Systolic Blood Pressure During Exercise Identifies Patients at Risk for Complications in Adults With a Systemic Right Ventricle





Conclusions Patients with a right ventricular end-diastolic volume index above 150 ml/m² and peak exercise systolic blood pressure below 180 mm Hg had a 20-fold higher annual event rate than patients without these risk factors. Regular cardiovascular magnetic resonance imaging and exercise testing are important in the risk assessment of these patients. (J Am Coll Cardiol 2013;62:926–36) © 2013 by the American College of Cardiology Foundation

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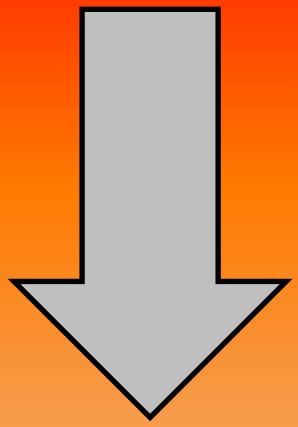

Perché l'imaging?

Latest insights in therapeutic options for systemic right ventricular failure: a comparison with left ventricular failure

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Table 1 Summary of trials with β-blockers, ACE inhibitors and ATII antagonists in patients with a systemic right ventricle

	Agent	TGA/ccTGA	No	Follow-up (months)	MRI*	VO_2max	NYHA	Pro/retro
β-blocker								
Lindenfeld *et al*[17]	Carvedilol	ccTGA	1	7	↑	ND	ND	pro
Giardini *et al*[18]	Carvedilol	both	8	12	↑	=	↑	pro
Josephson *et al*[16]	Various	TGA	8	36	ND	ND†	↑	retro
Doughan *et al*[15]	Various	TGA	31	4	ND	ND	↑	retro
ACE inhibitor								
Hechter *et al*[21]	Various	TGA	14	24	=	=	ND	retro
Robinson *et al*[22]	Enalapril	TGA	9	12	ND	=	ND	pro
Therrien *et al*[23]	Ramipril	TGA	17	12	=	=	ND	pro
ATII antagonist								
Dore *et al*[26]	Losartan	both	29	3.5	ND	=	ND	pro
Lester *et al*[24]	Losartan	TGA	7	2	↑ ‡	ND	ND	pro

CONCLUSIONS

Progressive dysfunction of the systemic right ventricle is an inevitable complication in patients with corrected TGA. The exact pathophysiology remains unclear, but seems multifactorial and heterogeneous. Although treatment regimens for left ventricular failure are well established, convincing data on the treatment of patients with a systemic right ventricle remain sparse. Combined efforts to perform prospective, randomised trials could overcome this shortcoming and provide these patients with an optimal long-term treatment and a better future.

Heart 2009;**95**:960–963.

Effect of Valsartan on Systemic Right Ventricular Function

A Double-Blind, Randomized, Placebo-Controlled Pilot Trial

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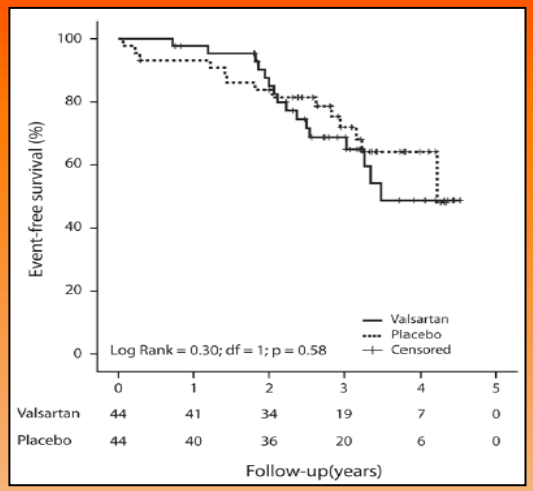

Events	n (%) Valsartan (n=44)	Placebo (n=44)	Hazard Ratio (95% CI)	*P*
Heart failure	6 (14)	5 (11)	1.2 (0.4–4.0)	0.73
Supraventricular arrhythmia	8 (18)	8 (18)	1.0 (0.4–2.7)	0.96
Ventricular arrhythmia	3 (7)	1 (2)	3.1 (0.3–30.3)	0.32
Death	1 (2)	1 (2)	1.0 (0.1–15.7)	0.99
Reoperation	2 (5)	0 (0)	67 (0.01–5.8×10^6)	0.47
Composite end point	16 (36)	14 (32)	1.2 (0.6–2.5)	0.58

Conclusions—There was no significant treatment effect of valsartan on right ventricular ejection fraction, exercise capacity, or quality of life. Valsartan was associated with a similar frequency of significant clinical events as placebo. Small but significant differences between valsartan and placebo were present for change in right ventricular volumes and mass.

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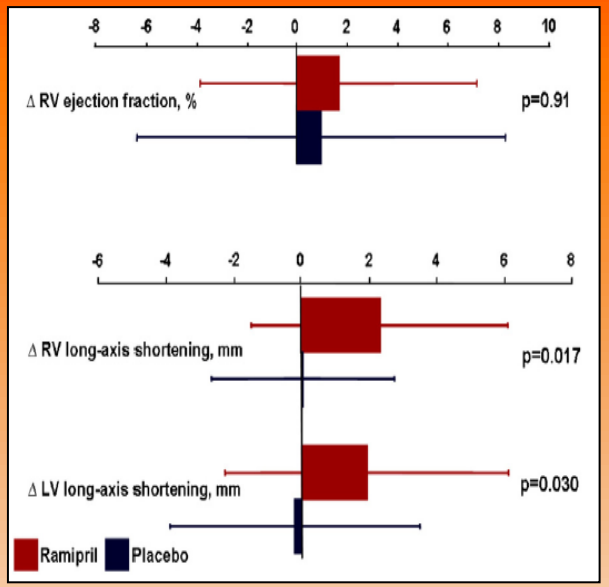
Randomised trial of ramipril in repaired tetralogy of Fallot and pulmonary regurgitation

The *APPROPRIATE* study (*A*ce inhibitors for *P*otential *PR*evention *O*f the deleterious effects of *P*ulmonary *R*egurgitation *I*n *A*dults with repaired *TE*tralogy of Fallot)



There was no difference in the primary endpoint RV ejection fraction. RV long-axis shortening significantly improved in the ramipril group compared to placebo (RV: 2.3 ± 3.8 vs 0.02 ± 2.7 mm; P = 0.017) as did LV long-axis shortening (1.9 ± 4.5 vs −0.2 ± 3.7 mm respectively; P = 0.030). No clear differences were detected between ramipril and placebo for other measures. In a subgroup of patients with restrictive RV physiology, ramipril resulted in decrease in LV end-systolic volume index and increase in LVEF (−2.4 ± 5.0 vs 2.7 ± 3.6 mL/m^2; P = 0.005,

Conclusions: Ramipril is a well tolerated therapy, improves biventricular function in patients with rTOF and may have a particular role in patients with restrictive RV physiology. Larger, longer-term studies are needed to determine if ACE inhibitors can improve both ventricular remodelling and clinical outcomes. (ISRCTN: 97515585)

International Journal of Cardiology 154 (2012) 299–305

CONCLUSIONI

1) **L'imaging integrato è fondamentale nel follow up dei GUCH soprattutto nello spirito dell'*INNOVAZIONE* e del *COWORKING***

2) **La terapia medica non ha dimostrato di essere una valida alternativa nei GUCH alla terapia interventistica/chirurgia**