



**46°**  
**Congresso Nazionale**  
Associazione Nazionale  
Medici Cardiologi Ospedalieri

**ANMCO**  
**2015**

Il follow up del paziente adulto con cardiopatia congenita operata (GUCH)

## **La chirurgia extracardiaca: valutazione del rischio pre-operatorio**



**Giorgio Faganello**



**Ambulatorio delle Cardiopatie Congenite dell'Adulto**

Centro Cardiovascolare

Dir. Andrea Di Lenarda, Trieste





European Heart Journal (2014) **35**, 2383–2431  
doi:10.1093/eurheartj/ehu282

**ESC/ESA GUIDELINES**

European  
Society of  
Anaesthesiology **ESA**

## 2014 ESC/ESA Guidelines on non-cardiac surgery: cardiovascular assessment and management

- **Annually:**
  - 5.7 million procedures in European patients with increased risk of cardiovascular complications
  - For EU countries: at least 167,000 cardiac complications due to non-cardiac surgical procedures, of which 19,000 are life-threatening

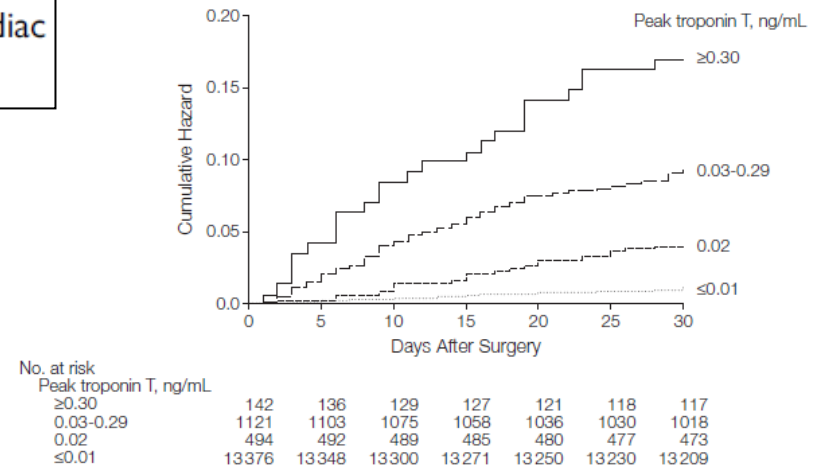


## Association Between Postoperative Troponin Levels and 30-Day Mortality Among Patients Undergoing Noncardiac Surgery

JAMA. 2012;307(21):2295-2304

Worldwide, non-cardiac surgery is associated with an average overall complication rate of 7–11% and a mortality rate of 0.8–1.5%, depending on safety precautions.<sup>2</sup> Up to 42% of these are caused by cardiac complications.<sup>3</sup> When applied to the population in the European Union member states, these figures translate into at least 167 000 cardiac complications annually due to non-cardiac surgical procedures, of which 19 000 are life-threatening.

Kaplan-Meier Estimates of 30-Day Mortality Based on Peak Troponin T Values





## LINEE GUIDA

### Linee guida per la valutazione preoperatoria del rischio cardiaco e la gestione perioperatoria del paziente cardiopatico nella chirurgia non cardiaca

Tabella 3. Numero di dimissioni per intervento chirurgico nei periodi 1994-1995 e 2004-2005 per fascia di età riportato dalla US National Hospital Discharge Survey (ospedali a breve degenza, non federali)<sup>15</sup>.

Età (anni)	N. procedure (migliaia)		Variazione %
	1994-1995	2004-2005	
18-44	7311	7326	+2.1
45-64	4111	5210	+26.7
65-74	3069	3036	-1.1
≥75	3479	4317	+24.1
≥18	17 969	19 889	+10.7

#### Impatto dell'invecchiamento della popolazione

Nell'arco dei prossimi 20 anni, il progressivo aumento dell'età media della popolazione si tradurrà in un forte impatto sulla gestione perioperatoria dei pazienti. Si stima che la popolazione anziana necessiti di interventi chirurgici in misura 4 volte superiore rispetto al resto della popolazione<sup>11</sup> e, anche se non sono disponibili dati sul totale dei pazienti sottoposti ad intervento chirurgico in Europa, si prevede che entro il 2020 il numero aumenterà del 25% e che durante tale periodo la popolazione anziana incrementerà di oltre il 50%. Il numero complessivo di procedure chirurgiche aumenterà ancora più velocemente in ragione della crescente frequenza di interventi correlati con l'età<sup>12</sup>. I risultati della US National Hospital Discharge Survey evi-

(G Ital Cardiol 2010; 11 (10 Suppl 2): e136-e181)

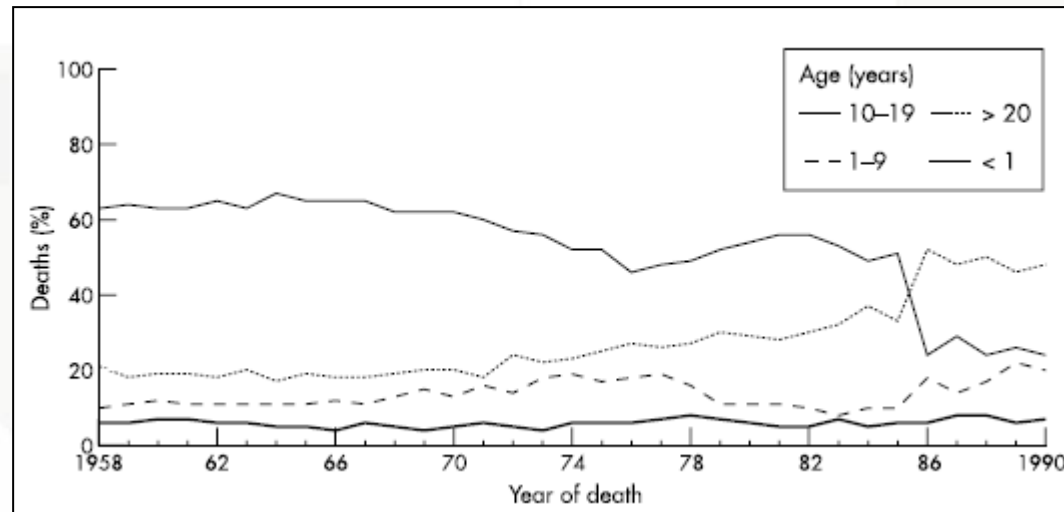


## BRITISH CARDIAC SOCIETY

Grown-up congenital heart (GUCH) disease:  
current needs and provision of service for adolescents  
and adults with congenital heart disease in the UK

Report of the British Cardiac Society Working Party\*

*Heart 2002;88(Suppl I):i1-i14*







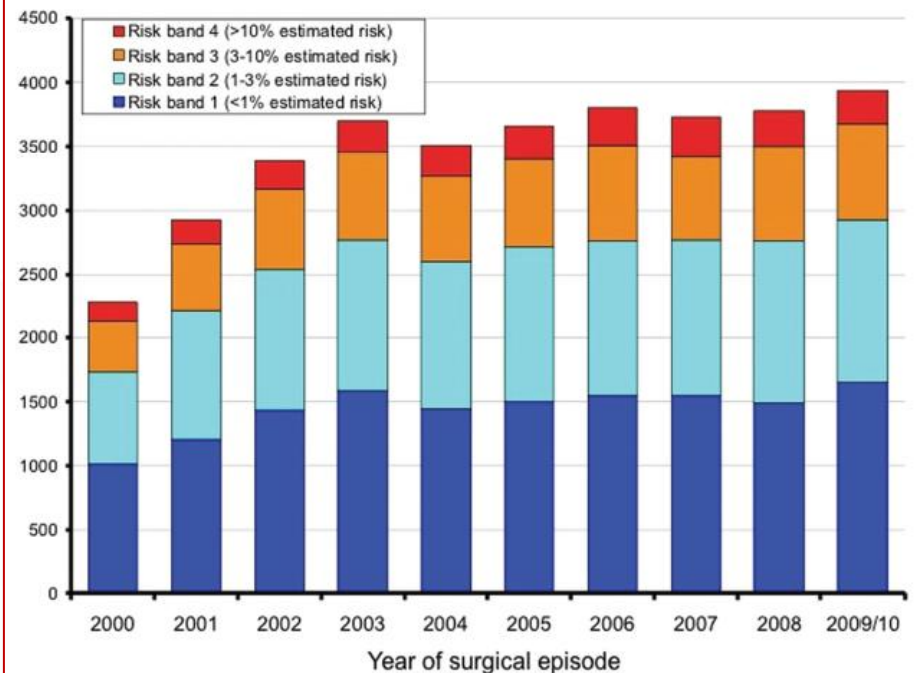
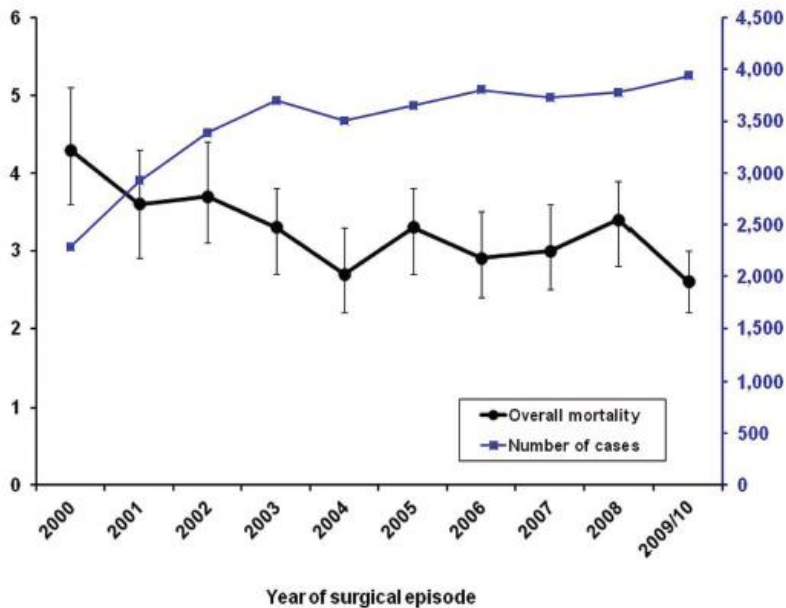
## Congenital heart disease

### Trends in 30-day mortality rate and case mix for paediatric cardiac surgery in the UK between 2000 and 2010

2015

Mortality rate (%)

Number of 30-day surgical episodes

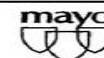




# La chirurgia extracardiaca: valutazione del rischio pre-operatorio



## Original Article



### Outcomes of Noncardiac Surgical Procedures in Children and Adults With Congenital Heart Disease

MARK A. WARNER, M.D., ROBERT J. LUNN, M.D.,\* PATRICK W. O'LEARY, M.D., AND DARRELL R. SCHROEDER, M.S., FOR THE MAYO PERIOPERATIVE OUTCOMES GROUP†

Mayo Clin Proc 1998;73:728-734

Table 1.—Primary Pathophysiologic Finding or Diagnosis in Study Patients With Congenital Heart Disease (N = 276)\*

Primary diagnosis†	No. of patients
<b>Shunt</b>	<b>155</b>
Isolated VSD	58
Patent ductus arteriosus	36
ASD‡	35
Complete AV septal defect	10
Double-outlet right ventricle	7
Primum ASD and cleft left AV valve	4
Truncus arteriosus	3
Other	2
<b>Cyanotic CHD</b>	<b>68</b>
Single ventricle, excluding heterotaxia	25
Tetralogy of Fallot	15
Pulmonary valve atresia and VSD	10
Complex complete TGA	8
Ebstein anomaly of the tricuspid valve	5
Simple complete TGA	4
Heterotaxia syndromes	1
<b>Obstructive CHD</b>	<b>48</b>
Pulmonary stenosis§	21
Coarctation syndrome, including interrupted arch	18
Complex TGA	6
Vascular ring	3
<b>Other</b>	<b>5</b>
Congenital aortic regurgitation	1
Congenitally corrected TGA	1
Other anomalies	3

Table 2.—Characteristics Associated With Complications\* of Noncardiac Surgical Procedures in Patients With Congenital Heart Disease†

Characteristic	First procedure‡ (N = 276)			All procedures§ (N = 480)	
	No. of patients	Complications (%)	P§	No. of procedures	Complications (%)
Overall	276	5.4	...	480	5.8
Gender			NS		
Male	134	4.5		230	4.8
Female	142	6.3		250	6.8
Age (yr)¶			0.009		
<2	62	12.9		106	10.4
2-5	58	1.7		104	1.9
6-10	49	2.0		84	2.4
11-17	38	10.5		71	7.0
≥18	69	1.4		115	7.0
Primary physiologic feature of CHD			NS		
Shunt	155	5.8		273	6.6
Cyanotic	68	5.9		118	6.8
Obstructive	48	4.2		83	2.4
Other	5	0.0		6	0.0
Scheduled as inpatient procedure			0.024		
No	93	1.1		175	1.7
Yes	183	7.7		305	8.2
ASA classification#			<0.001		
1	37	2.7		58	3.5
2	114	0.9		188	2.1
3	105	6.7		197	7.1
4	19	36.3		36	19.4
5	1	100.0		1	100.0
Emergency procedure			NS		
Yes	31	3.2		57	8.8
No	245	5.7		423	5.4
Previous operation for CHD			NS		
No	116	4.3		202	5.0
Yes, palliative	19	15.8		30	13.3
Yes, definitive	141	5.0		248	5.7
Cyanotic status at time of procedure			0.002		
Cyanotic	30	20.0		46	21.3
Acyanotic	246	4.5		434	4.2
Pulmonary hypertension at time of procedure			NS		
Yes	20	15.0		32	21.9
No	256	4.7		448	4.7
Treated for CHF at time of procedure			<0.001		
Yes	29	24.1		47	21.3
No	247	3.2		433	4.2
Type of anesthesia**			NS		
General	232	6.5		409	6.1
Regional	24	0.0		41	4.9
Local	14	0.0		19	0.0
MAC	2	0.0		6	0.0
Combination	3	0.0		4	25.0
None	1	0.0		1	0.0



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VP shunt placement  
Craniotomy for AV fistula  
VP shunt placement  
Inguinal hernia repair,  
bronchoscopy  
Tunneled CVC placement,  
tracheostomy  
Tracheostomy  
BMT, cleft palate repair  
Bronchoscopy  
Bronchoscopy, tracheal  
dilation  
Tracheostomy  
AS of lumbar scoliosis  
Craniotomy for tumor  
resection  
PS of lumbar scoliosis  
Tonsillectomy  
Dental extractions

**5.8% complicanze  
1 decesso**

Age (yr)¶			0.009
<2	62	12.9	
Cyanotic status at time of procedure			0.002
Cyanotic	30	20.0	
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4	19	36.3	
5	1	100.0	

CHF, prolonged ventilation  
Seizures  
Seizures, prolonged intubation  
RI, prolonged ventilation  
CHF  
CHF, hepatic failure, cardiac arrest  
Pneumonia  
IO pneumothorax  
RI, bronchospasm  
Atrial flutter  
RI, reintubated in PACU  
CHF  
CHF and bronchospasm in PACU  
Cardiac arrest and death after induction of anesthesia  
Severe bradycardia during induction of anesthesia



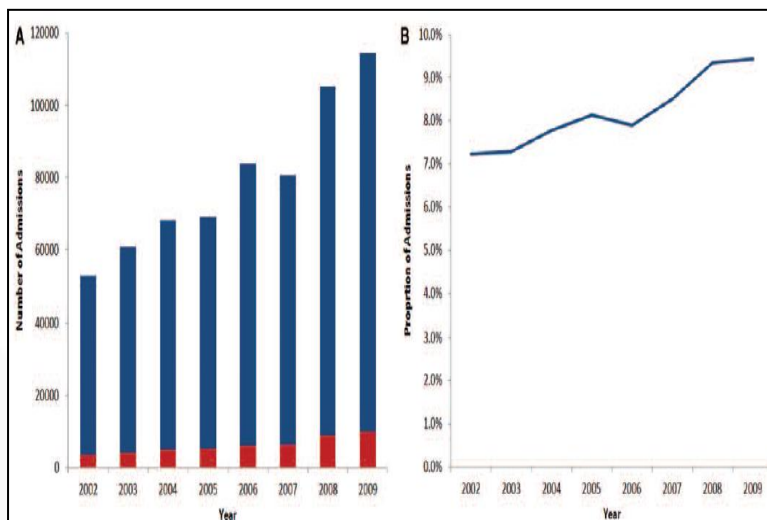


# La chirurgia extracardiaca: valutazione del rischio pre-operatorio



## PERIOPERATIVE MEDICINE

### Perioperative Outcomes of Major Noncardiac Surgery in Adults with Congenital Heart Disease



10004 GUCH  
VS  
37.581 non GUCH

**Table 1.** Demographic and Baseline Characteristics of ACHD and Comparison Cohorts

	ACHD Cohort	Comparison Cohort	
	n = 10,004	n = 37,581	
	n (%)	n (%)	P Value
Age (yr, mean $\pm$ SE)	57.6 ( $\pm$ 0.3)	58.1 ( $\pm$ 0.2)	0.0192
Female	5,520 (55.2)	21,005 (56.0)	0.18
Race			0.06
White	6,140 (61.8)	23,374 (62.7)	
Black	534 (5.3)	1,885 (5.0)	
Hispanic	469 (4.7)	1,547 (4.1)	
Asian	136 (1.4)	559 (1.5)	
Native American	39 (0.4)	150 (0.4)	
Others	185 (1.9)	754 (2.0)	
Nonelective status	4,717 (47.2)	17,659 (46.8)	0.67
Payer			<0.001
Medicare	4,313 (43.2)	15,900 (42.4)	
Medicaid	781 (7.8)	2,988 (7.9)	
Private	4,296 (43.0)	15,527 (41.4)	
Self-Pay	254 (2.5)	1,384 (3.7)	
Others	342 (3.4)	1,728 (4.6)	
Operation by surgical service			<0.001
ENT	225 (2.3)	969 (2.6)	
General surgery	3,514 (35.1)	13,329 (35.5)	
Gynecologic	770 (7.7)	3,078 (8.2)	
Neurosurgery	1,201 (12.0)	4,881 (13.0)	
Orthopedic	2,991 (29.9)	12,061 (32.2)	
Thoracic	1,596 (15.9)	4,563 (12.3)	
Urologic	597 (6.0)	2,445 (6.5)	
Hospital size			<0.001
Small	6,812 (68.1)	24,916 (66.3)	
Medium	2,153 (21.5)	8,553 (22.8)	
Large	1,038 (10.4)	4,112 (10.9)	
Hospital teaching status			<0.001
Nonteaching	5,901 (58.9)	19,493 (51.9)	
Teaching	4,103 (41.1)	18,088 (48.1)	
van Walraven score (mean $\pm$ SE)	3.4 $\pm$ 0.8	3.1 ( $\pm$ 0.7)	0.68



# La chirurgia extracardiaca: valutazione del rischio pre-operatorio



## PERIOPERATIVE MEDICINE

### Perioperative Outcomes of Major Noncardiac Surgery in Adults with Congenital Heart Disease

**Table 2.** Outcomes

Outcome	ACHD Cohort	Comparison Cohort		
	n = 10,004	n = 37,581		
	n (%)	n (%)	P Value	OR (95% CI)
Death	407 (4.1)	1,355 (3.6)	0.031	.13 (1.01-1.27)
LOS (median [IQR])	4.8 (2.4-10.4)	2.9 (1.5-5.6)	<0.001	
Total charges (median [IQR])	\$42,171 (\$22,918-\$93,847)	\$26,982 (\$15,814-\$46,784)	<0.001	
ARF	620 (6.2)	1,826 (4.9)	<0.001	.29 (1.18-1.42)
Pneumonia	942 (9.4)	2,998 (8.0)	<0.001	.20 (1.11-1.29)
Respiratory failure	916 (9.2)	2,933 (7.8)	<0.001	.19 (1.10-1.29)
DVT/PE	405 (4.1)	773 (2.1)	<0.001	2.01 (1.78-2.27)
Stroke	607 (6.1)	1,168 (3.1)	<0.001	2.01 (1.82-2.23)
MI/cardiac arrest	431 (4.3)	1,307 (3.5)	<0.001	.25 (1.12-1.40)
Composite	2,145 (21.4)	6,003 (16.0)	<0.001	.44 (1.36-1.52)

Values are reported as number (percentage) unless otherwise denoted as median (IQR). Composite = ARF, pneumonia, respiratory failure, DVT/PE, stroke, MI, and cardiac arrest.

ACHD = adult congenital heart disease; ARF = acute renal failure; DVT = deep venous thrombosis; IQR = interquartile range; LOS = length of stay; MI = myocardial infarction; OR = odds ratio; PE = pulmonary embolus.

**Table 3.** Mortality by Lesion Type within the ACHD Cohort

	Died	
	n	n (%)
Atrial septal defect	4,068	155 (3.8)
Congenital aortic stenosis/aortic insufficiency	1,789	53 (3.0)
Congenital mitral stenosis/regurgitation	85	3 (3.5)
Congenital conduction defect*	469	10 (2.1)
Congenital coronary anomaly	248	9 (3.6)
Pulmonic stenosis	239	13 (5.4)
Tetralogy of Fallot	121	7 (5.8)
Ventricular septal defect	831	52 (6.3)
Ebstein anomaly	65	4 (6.2)
Others	1,745	76 (4.4)
Combined complex†	344	25 (7.3)

**Conclusions:** Compared with a matched control cohort, ACHD patients undergoing noncardiac surgery experienced increased perioperative morbidity and mortality. Within the limitations of a retrospective analysis of a large administrative dataset, this finding demonstrates that this is a vulnerable population and suggests that better efforts are needed to understand and improve the perioperative care they receive.



## La chirurgia extracardiaca: valutazione del rischio pre-operatorio

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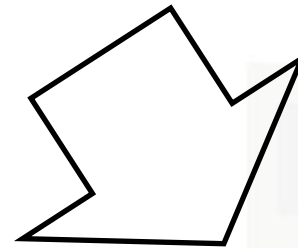
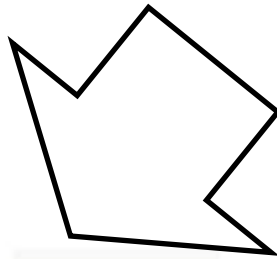
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**PROBLEMA  
TRASVERSALE**



## La chirurgia extracardiaca: valutazione del rischio pre-operatorio



 European Heart Journal (2014) 35, 2383–2431 **ESC/ESA GUIDELINES** European Society of Anaesthesiology **ESA**  
doi:10.1093/eurheartj/ehu282

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**2014 ESC/ESA Guidelines on non-cardiac surgery:  
cardiovascular assessment and management**

**PRACTICE GUIDELINE: FULL TEXT**

**ACC/AHA 2008 Guidelines for the  
Management of Adults With Congenital Heart Disease**

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)



# La chirurgia extracardiaca: valutazione del rischio pre-operatorio



European Heart Journal (2014) 35, 2383–2431  
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ESC/ESA GUIDELINES

European Society of Anaesthesiology

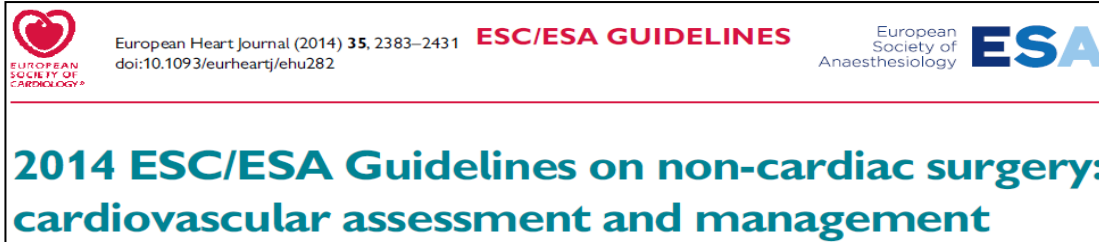
ESA

## 2014 ESC/ESA Guidelines on non-cardiac surgery: cardiovascular assessment and management

**Table 3** Surgical risk estimate according to type of surgery or intervention<sup>a,b</sup>

Low-risk: < 1%	Intermediate-risk: 1–5%	High-risk: > 5%
<ul style="list-style-type: none"><li>• Superficial surgery</li><li>• Breast</li><li>• Dental</li><li>• Endocrine: thyroid</li><li>• Eye</li><li>• Reconstructive</li><li>• Carotid asymptomatic (CEA or CAS)</li><li>• Gynaecology: minor</li><li>• Orthopaedic: minor (meniscectomy)</li><li>• Urological: minor (transurethral resection of the prostate)</li></ul>	<ul style="list-style-type: none"><li>• Intraperitoneal: splenectomy, hiatal hernia repair, cholecystectomy</li><li>• Carotid symptomatic (CEA or CAS)</li><li>• Peripheral arterial angioplasty</li><li>• Endovascular aneurysm repair</li><li>• Head and neck surgery</li><li>• Neurological or orthopaedic: major (hip and spine surgery)</li><li>• Urological or gynaecological: major</li><li>• Renal transplant</li><li>• Intra-thoracic: non-major</li></ul>	<ul style="list-style-type: none"><li>• Aortic and major vascular surgery</li><li>• Open lower limb revascularization or amputation or thromboembolism</li><li>• Duodeno-pancreatic surgery</li><li>• Liver resection, bile duct surgery</li><li>• Oesophagectomy</li><li>• Repair of perforated bowel</li><li>• Adrenal resection</li><li>• Total cystectomy</li><li>• Pneumonectomy</li><li>• Pulmonary or liver transplant</li></ul>





## 3.4 Risk indices

of risk for cardiac events. The Lee index or 'revised cardiac risk' index, a modified version of the original Goldman index, was designed to predict post-operative myocardial infarction, pulmonary oedema, ventricular fibrillation or cardiac arrest, and complete heart block. This risk index comprises six variables: type of surgery, history of IHD, history of heart failure, history of cerebrovascular disease, pre-operative treatment with insulin, and pre-operative creatinine  $> 170 \mu\text{mol/L}$  ( $> 2 \text{ mg/dL}$ ), and used to be considered by many clinicians and researchers to be the best currently available cardiac-risk prediction index in non-cardiac surgery.

of non-cardiac surgical patients. A new predictive model was recently developed to assess the risk of intra-operative/post-operative myocardial infarction or cardiac arrest, using the American College of Surgeons National Surgical Quality Improvement Program (NSQIP) database.<sup>44</sup> This NSQIP MICA model was built on the 2007 data



# Surgical Risk Calculator



*Please enter as much of the following information as you can to receive the best risk estimates.  
A rough estimate will still be generated if you cannot provide all of the information below.*

Age Group

Sex

Functional status

Emergency case

ASA class

Wound class

Steroid use for chronic condition

Ascites within 30 days prior to surgery

Systemic sepsis within 48 hours prior to surgery

Ventilator dependent

Disseminated cancer

Diabetes

Hypertension requiring medication

Previous cardiac event

Congestive heart failure in 30 days prior to surgery

Dyspnea

Current smoker within 1 year

History of severe COPD

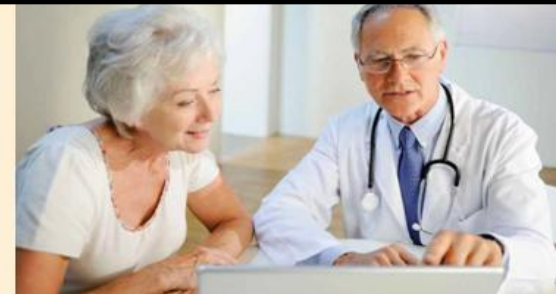
Dialysis

Acute Renal Failure

BMI Calculation: Height (in)

Weight (lbs)

# Surgical Risk Calculator



Outcomes	Estimated Risk	Chance of Outcome
Serious Complication	2%	Average
Any Complication	2%	Above Average
Pneumonia	<1%	Below Average
Cardiac Complication	<1%	Below Average
Surgical Site Infection	<1%	Below Average
Urinary Tract Infection	<1%	Below Average
Venous Thromboembolism	<1%	Below Average
Renal Failure	<1%	Below Average

Il follow up del paziente adulto con cardiopatia congenita operata (GUCH)

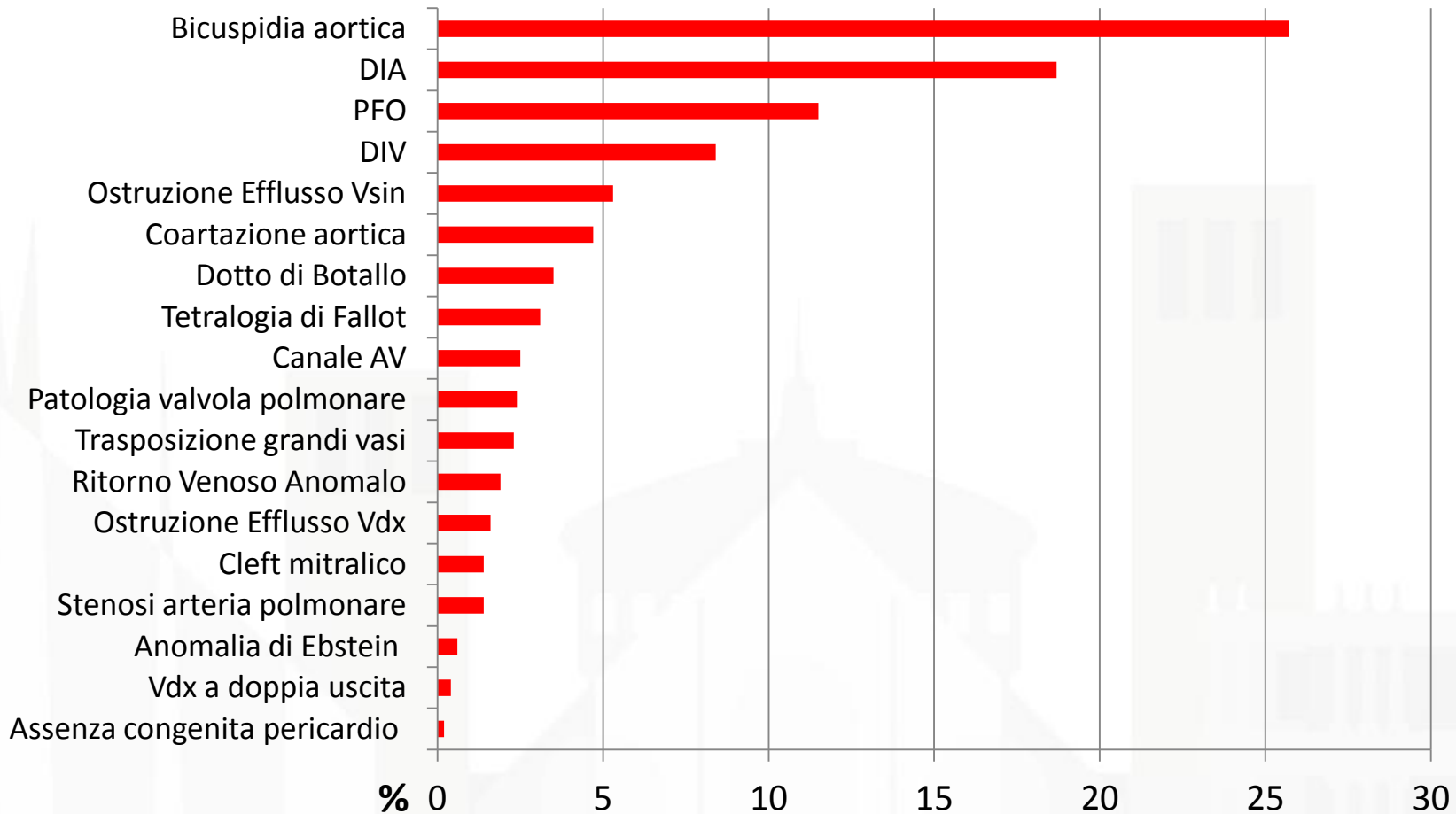


# La chirurgia extracardiaca: valutazione del rischio pre-operatorio



## L'Osservatorio GUCH di Trieste

(11/2009-05/2015; n=486-2.3%; 37% pz con CCH)



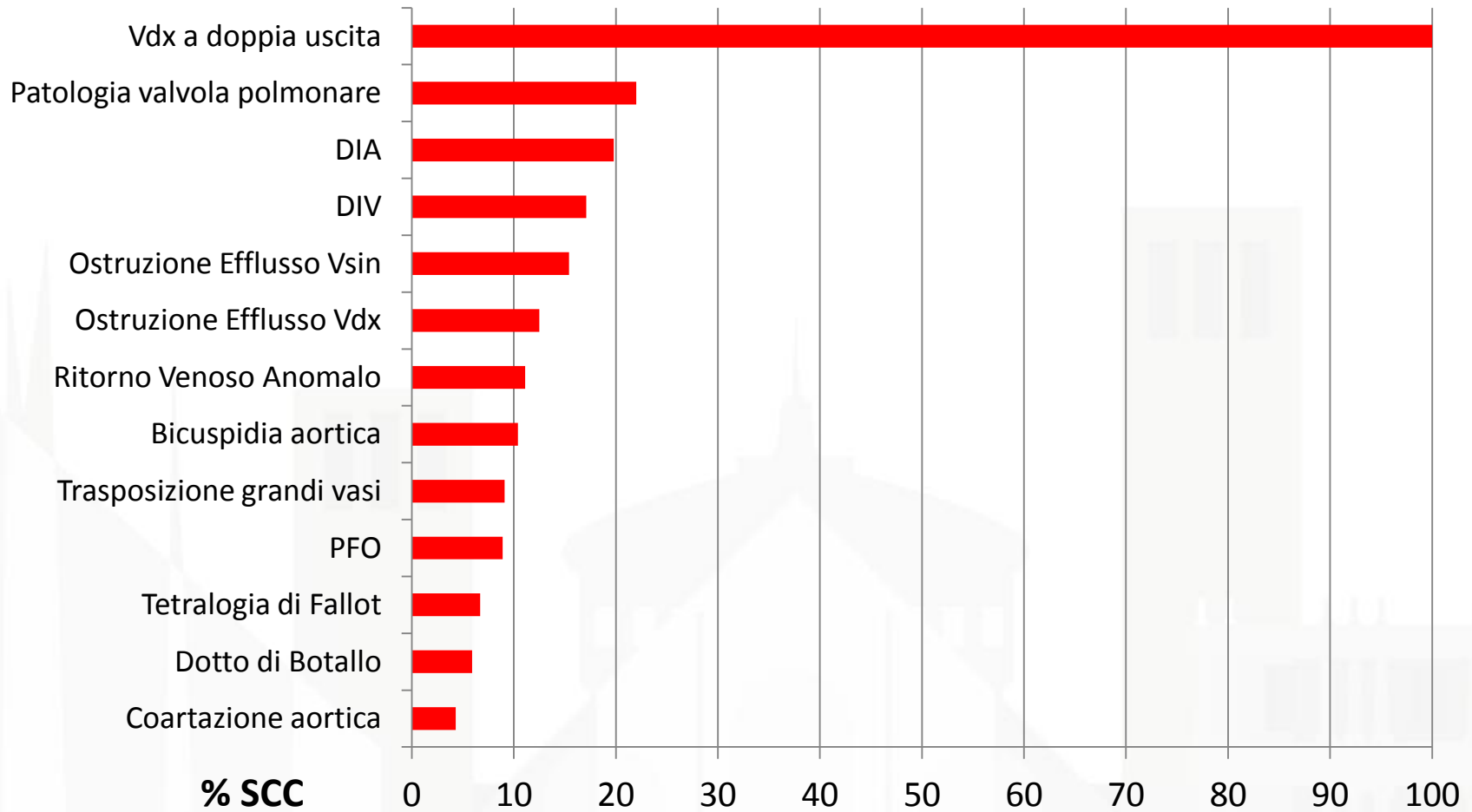


La chirurgia extracardiaca: valutazione del rischio pre-operatorio



**L'Osservatorio GUCH di Trieste**

**(11/2009-05/2015; n=486-2.3%; 13.8 % pz con SCC)**







European Heart Journal (2014) 35, 2383–2431  
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## 2014 ESC/ESA Guidelines on non-cardiac surgery: cardiovascular assessment and management

### 5.9 Congenital heart disease

Children, adolescents and adults with congenital heart disease are generally regarded as being at increased risk when undergoing non-cardiac surgery but this risk will vary enormously, according to the degree of associated heart failure, pulmonary hypertension, arrhythmias, and shunting of blood—with or without associated oxygen desaturation and by the complexity of the underlying condition.<sup>222</sup> A thorough understanding of the underlying congenital heart disease, including anatomy, physiology, and identification of risk factors, is vital before surgery. When the defect is simple, the circulation physiologically normal and the patient well compensated, the risk may be quite low; however, complicated patients with congenital heart disease should only undergo non-cardiac surgery after thorough evaluation by a multidisciplinary team in a specialized centre. Prophylaxis for endocarditis should be initiated according to the ESC Guidelines on congenital heart disease and infective endocarditis.<sup>190,222</sup>

#### Recommendation on patients with congenital heart disease

Recommendation	Class <sup>a</sup>	Level <sup>b</sup>
It is recommended that, patients with complex congenital heart disease be referred for additional specialist investigation before undergoing elective non-cardiac surgery, if feasible.	I	C

<sup>a</sup>Class of recommendation.

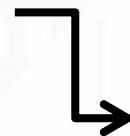
<sup>b</sup>Level of evidence.



## Anaesthetic management and outcomes in patients with surgically corrected D-transposition of the great arteries **BJA** undergoing non-cardiac surgery

*British Journal of Anaesthesia* **104** (1): 12–15 (2010)

Event	Age (yr)	ASA	Procedure	Pertinent medical history	Intervention and outcomes
Severe bradycardia during insufflation of abdomen	15.6	III	Laparoscopic gynaecologic surgery	Sinus bradycardia and bundle branch block, pulmonary stenosis	Glycopyrrolate; sent to general care
Failed extubation	14.6	III	Anterior spine fusion	Mild pulmonary and tricuspid regurgitation, pulmonary stenosis; severe restrictive lung disease; dyspnoea on exertion and fatigue	Tracheotomy several days after surgery; remains ventilator-dependent 3 yr after operation
Failed extubation	0.25	III	Supraglottoplasty	Kabuki syndrome; Pierre Robin sequence, floppy epiglottis, laryngomalacia; mild aortic regurgitation and mild pulmonary artery stenosis	Tracheotomy; decannulated 1 yr later
Bleeding/haematoma	1.9	III	Circumcision	Mild pulmonary regurgitation and aortic insufficiency	Re-operation; discharged home after



Importantly, 8% of these patients had an adverse event during or immediately after surgery. However, only one of these events was likely related to the underlying residual cardiac condition (i.e. bradycardia). Two were related to significant pulmonary or airway comorbidities. The first of



## Anaesthetic management and outcomes after noncardiac surgery in patients with hypoplastic left heart syndrome: a retrospective review.

*Eur J Anaesthesiol. 2012*

Adverse events occurred in 11 (15%) cases, including cardiovascular and respiratory instability, airway obstruction and postoperative stridor, with 13 (18%) patients admitted to the ICU postoperatively

## Outcomes of general anesthesia for noncardiac surgery in a series of patients with Fontan palliation.

*Paediatr Anaesth. 2013*

Thirty-nine general anesthetics were administered to 31 patients for noncardiac surgery after Fontan palliation. **Perioperative complications occurred in 12 of the 39 (31%)** noncardiac surgeries, and there was one postoperative death that occurred on day 13 after ventral hernia repair. The two patients who had complications that did not resolve (long-term dialysis and death) had ejection fractions well below the mean for the group (22% and 28%).



**PRACTICE GUIDELINE: FULL TEXT**

## **ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease**

A Report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease)

### **Table 7. Congenital Cardiac Lesions and Perioperative Risk for Noncardiac Surgery**

#### **High risk**

- Pulmonary hypertension, primary or secondary
- Cyanotic congenital heart disease
- New York Heart Association class III or IV
- Severe systemic ventricular dysfunction (ejection fraction less than 35%)
- Severe left-sided heart obstructive lesions

#### **Moderate risk**

- Prosthetic valve or conduit
- Intracardiac shunt
- Moderate left-sided heart obstruction
- Moderate systemic ventricular dysfunction



# La chirurgia extracardiaca: valutazione del rischio pre-operatorio

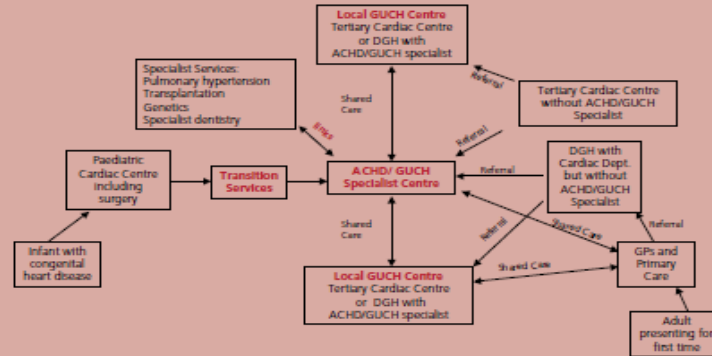


## Adult Congenital Heart Disease

A commissioning guide for services for young people and Grown Ups with Congenital Heart Disease (GUCH)



### Adults with Congenital Heart Disease (ACHD) or Grown up Congenital Heart Disease (GUCH) Service Model



### A multi disciplinary approach

#### 27. Indicators of high quality care

- All young people and adults with congenital heart disease receive care delivered by a multi disciplinary, multi speciality team. Within the NHS, the implications of their cardiac condition are taken into account within other specialties and there is expertise available in other relevant specialties such as obstetrics, anaesthetics and dentistry.
- Patients have access to ongoing support and advice on living with their condition and in particular follow up after a period of acute care. They also have ready access to information on understanding their condition and on accessing services which is appropriate to their level of understanding.
- For every congenital heart disease patient, GPs and the identified local GUCH centre receive communications from specialist centres on diagnosis, prognosis, the care plan and information to support the patient.
- Patients receive ongoing support and advice from a specialist GUCH nurse.
- Specialist centres provide a 24-hour point of reference for patients on any aspect of care including, for example, contraception, pregnancy and dental care.
- Specialist centres and local GUCH centres promote patient support groups such as the GUCH Patients Association and establish local support groups including support for carers.





## Linee Guida sul Follow-up del Cardiopatico Congenito Operato

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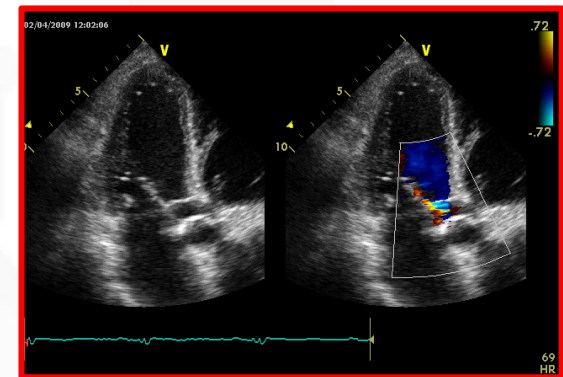
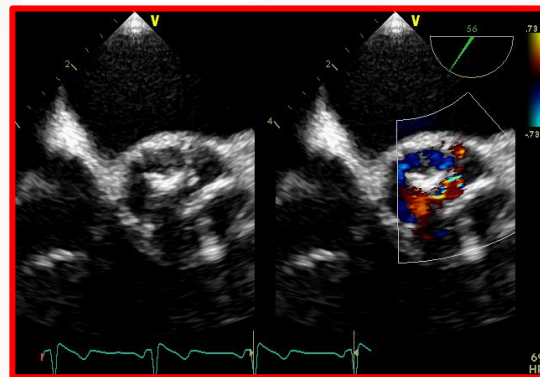
Luciano Daliento, Coordinatore

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Fernando Maria Picchio, Patrizia Presbitero,  
Vello Sperandio, Gabriele Vignati

### OSTRUZIONE DELL'EFFLUSSO VENTRICOLARE SINISTRO

Stenosi aortica valvolare / sottovalvolare / sopravalvolare

Chirurgia extracardiaca: non vi è un rischio aumentato a meno che non vi sia una disfunzione contrattile del ventricolo sinistro, generalmente conseguente ad un' insufficienza aortica oppure ad una stenosi residua importante nel qual caso sarebbe preferibile prima correggere il problema cardiaco e quindi affrontare, se possibile in un secondo momento o, se necessario, contestualmente, il problema extracardiaco. Anche in questa occasione si raccomanda una scrupolosa profilassi antibiotica dell'endocardite batterica.





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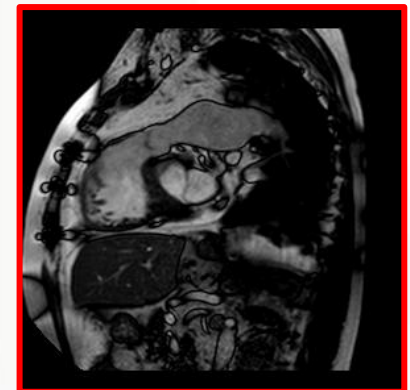
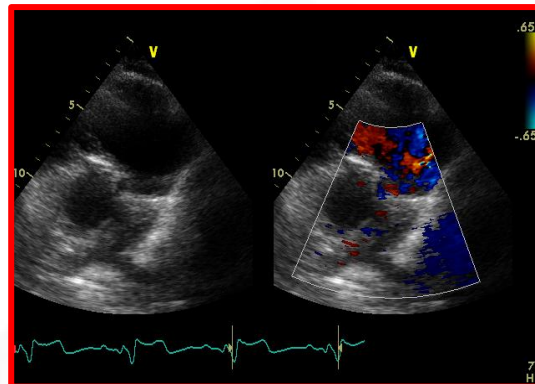
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## STENOSI POLMONARE VALVOLARE ISOLATA

*Chirurgia extracardiaca: i rischi sono legati alla eventuale presenza di gradienti residui significativi e soprattutto allo stato funzionale del ventricolo destro e all'eventuale presenza di aritmie.*





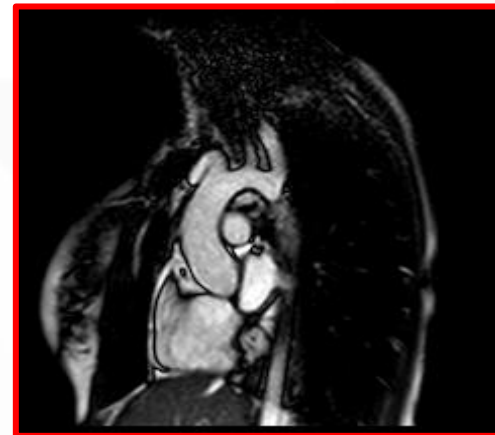
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## Coartazione istmo aorta

*Chirurgia extracardiaca: presentano un maggior rischio i pazienti con*  
*ipertensione arteriosa mal controllata dalla terapia o con ricoartazione significativa.*





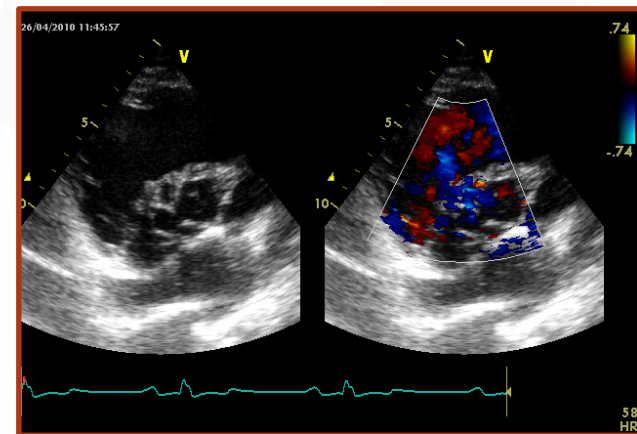
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## DIFETTI DEL SETTO ATRIOVENTRICOLARE

Chirurgia extracardiaca: il rischio è direttamente proporzionale al grado di insufficienza mitralica ed all'entità di un eventuale ipertensione polmonare residua.





# La chirurgia extracardiaca: valutazione del rischio pre-operatorio

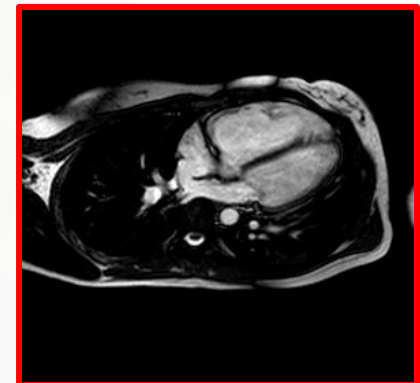
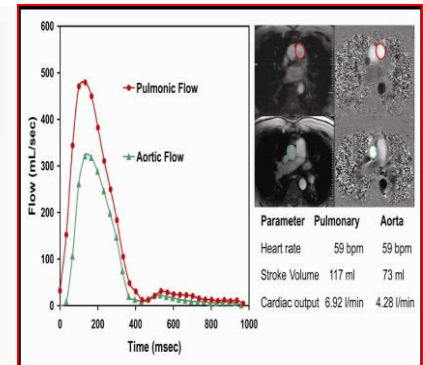
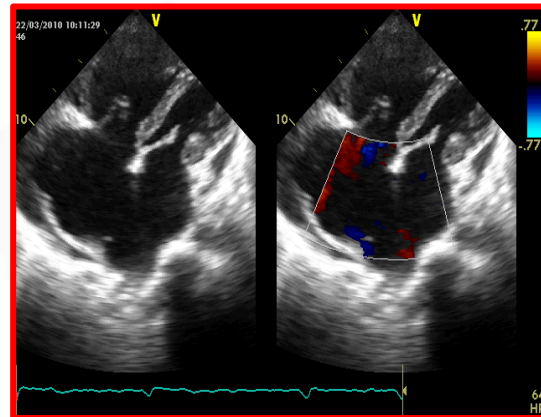
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## SHUNTS PRETRICUSPIDALI

*Chirurgia extracardiaca: non particolari rischi in assenza di dilatazione delle cavità, shunts residui, normale pressione polmonare e ritmo sinusale.*







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## SHUNT'S POST-TRICUSPIDALI

*Chirurgia extracardiaca:* in assenza di aritmie, di ipertensione polmonare e in presenza di una conservata funzione ventricolare non vi sono rischi aggiuntivi. Le aritmie non rappresentano un rischio aggiuntivo se sono controllate dalla terapia medica. La persistenza di ipertensione polmonare è la situazione a maggior rischio: soprattutto al momento dell'anestesia devono essere evitate situazioni di brusca ipotensione sistemica.



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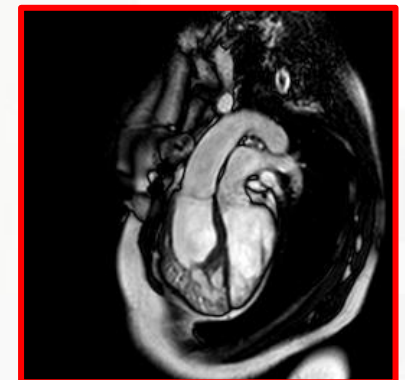
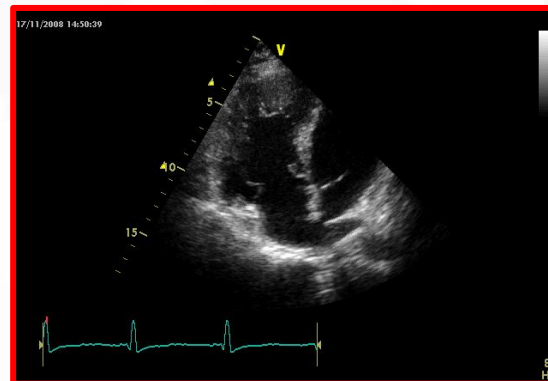
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## TRASPOSIZIONE DELLE GRANDI ARTERIE

### Correzione fisiologica secondo Mustard o Senning

**Rischio operatorio per chirurgia extracardiaca:** ogni intervento deve essere preceduto da un'attenta ed approfondita valutazione cardiologica che preveda l'esecuzione anche di un ecocardiogramma e di un Holter. In assenza di disfunzione contrattile e/o aritmie significative, il rischio operatorio non è dissimile da quello che correrebbe un soggetto senza cardiopatia, viceversa il rischio aumenta proporzionalmente al grado di compromissione del ventricolo destro. In ogni caso la chirurgia extracardiaca di questi pazienti deve essere eseguita in ambienti protetti in cui vi sia la possibilità di ricorrere a tutti quei presidi terapeutici atti a trattare qualsiasi urgenza-emergenza cardiologica.





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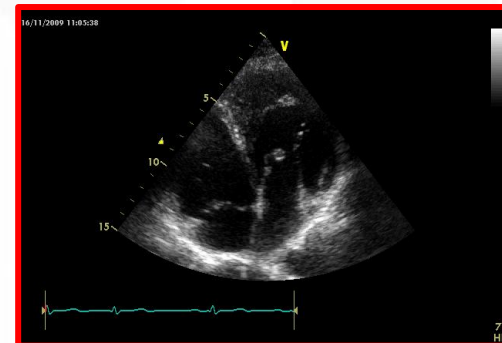
### TRASPOSIZIONE DELLE GRANDI ARTERIE

Correzione anatomica "switch arterioso"

o polmonare. Non si hanno dati sul lungo periodo, perché è troppo recente l'introduzione di questa tecnica ed i neonati non sono ancora divenuti adulti, tuttavia

### TRASPOSIZIONE CONGENTAMENTE CORRETTA DELLE GRANDI ARTERIE

Rischio operatorio per chirurgia extracardiaca: in assenza di alterata performance ventricolare e/o di disturbi del ritmo, le precauzioni sono limitate alla profilassi antibiotica per l'endocardite batterica. In presenza di reliquati particolare attenzione deve essere posta nella conduzione dell'anestesia e del periodo postoperatorio.





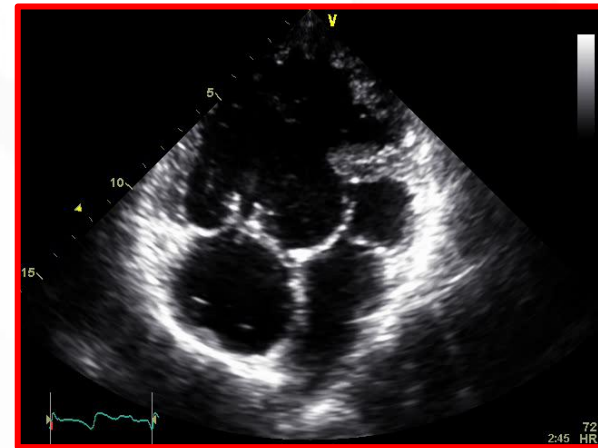
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## INTERVENTO DI FONTAN E SIMILI

**Rischio operatorio per chirurgia extracardiaca:** sono in ogni caso pazienti a rischio aumentato, anche quelli con un risultato ottimale. Si raccomanda monitoraggio del ritmo, profilassi accurata per eventuali tromboembolie polmonari (anche una piccola tromboembolia si rivelerebbe disastrosa per il tipo di emodinamica presente), monitoraggio del carico emodinamico, controllo ematochimico della crasi ematica e degli indici di funzione epatica. Utile la presenza di un cardiologo in sala operatoria.





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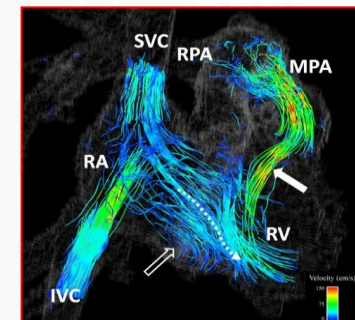
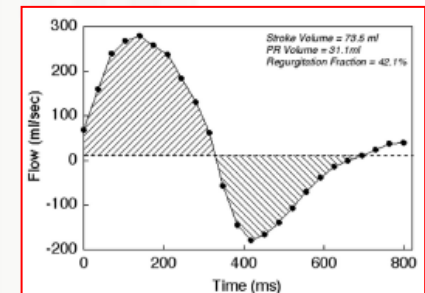
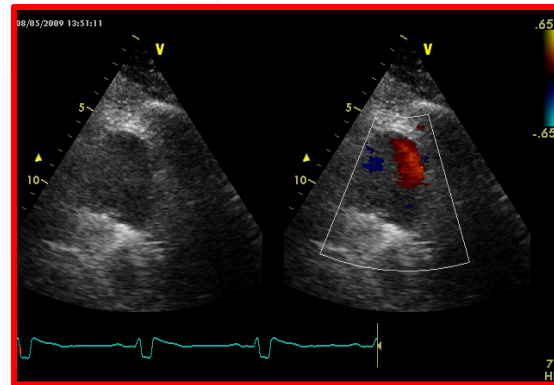
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## TETRALOGIA DI FALLOT

**Rischio operatorio per chirurgia extracardiaca:** in assenza di alterata performance ventricolare e/o di disturbi del ritmo, le precauzioni sono limitate all'antibiotico-profilassi onde minimizzare il potenziale rischio di infezione sistemica e di endocardite batterica specialmente sul condotto protesico. In presenza di alterata performance ventricolare e/o di disturbi del ritmo la condotta anestesiológica durante l'intervento dovrà tenere conto adeguatamente di questi elementi con vari accorgimenti: utilizzo di anestetici a minore effetto inotropo negativo, stretto controllo dell'apporto idrico post-chirurgico onde evitare bruschi incrementi della volemia, ridotto utilizzo di farmaci ad effetto vasocostrittore polmonare o sistemico.







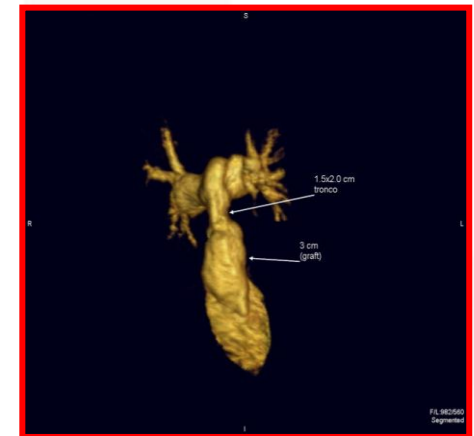
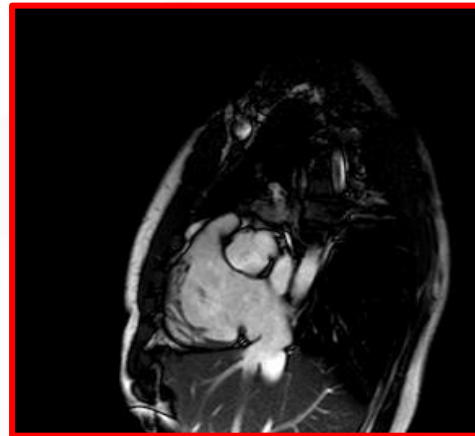
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### ATRESIA POLMONARE CON DIFETTO INTERVENTRICOLARE

*Gravidanza e Rischio operatorio per chirurgia extracardiaca:*  
anche per queste voci sono assimilabili agli operati di tetralogia di Fallot con ampio patch transanulare: lo stato del ventricolo destro e le stenosi residue dei rami polmonari giuocano un ruolo decisivo nella determinazione del rischio.







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## TRONCO ARTERIOSO

***Rischio operatorio per chirurgia extracardiaca:** anche in questo caso profilassi antibiotica per l'endocardite batterica e particolare attenzione alle modalità di anestesia e all'assistenza post-operatoria.*



# ACC/AHA 2008 Guidelines for the Management of Adults With Congenital Heart Disease

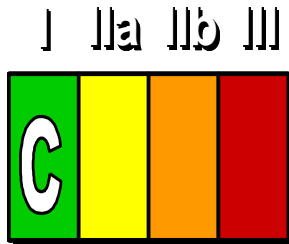
*Developed in Collaboration With:*

*The American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons*

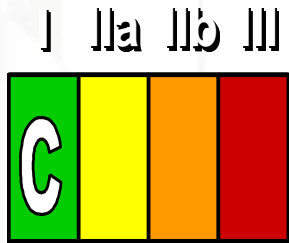
## Recommendations for Noncardiac Surgery



## Basic Preoperative Assessment



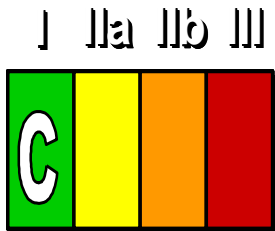
Basic preoperative assessment for ACHD patients should include systemic **arterial oximetry**, an **ECG**, **chest x-ray**, **TTE**, and **blood tests** for full blood count and coagulation screen.



It is recommended that when possible, the preoperative evaluation and surgery for ACHD patients be performed in a **regional center specializing in congenital cardiology**, with experienced surgeons and cardiac anesthesiologists.



## High Risk Populations and ACHD Patients

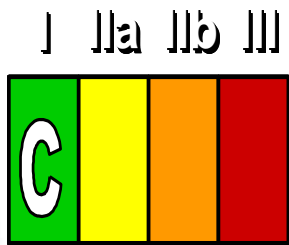


Certain high-risk patient populations should be managed at centers for the care of ACHD patients under all circumstances, unless the operative intervention is an absolute emergency. High-risk categories include patients with the following:

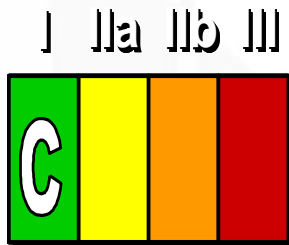
- a) **Prior Fontan procedure.**
- b) **Severe pulmonary arterial hypertension (PAH).**
- c) **Cyanotic CHD.**
- d) **Complex CHD with residua such as heart failure, valve disease, or the need for anticoagulation.**
- e) **Patients with CHD and malignant arrhythmias.**



## Consultation and Assessment of Risk



Consultation with ACHD experts regarding the assessment of risk is recommended for patients with CHD who will undergo noncardiac surgery.



Consultation with a cardiac anesthesiologist is recommended for moderate- and high-risk patients.



## Grazie