Passer Du Consensus D'experts À L'evidence Based Medicine en Cardiologie Congénitale Adulte

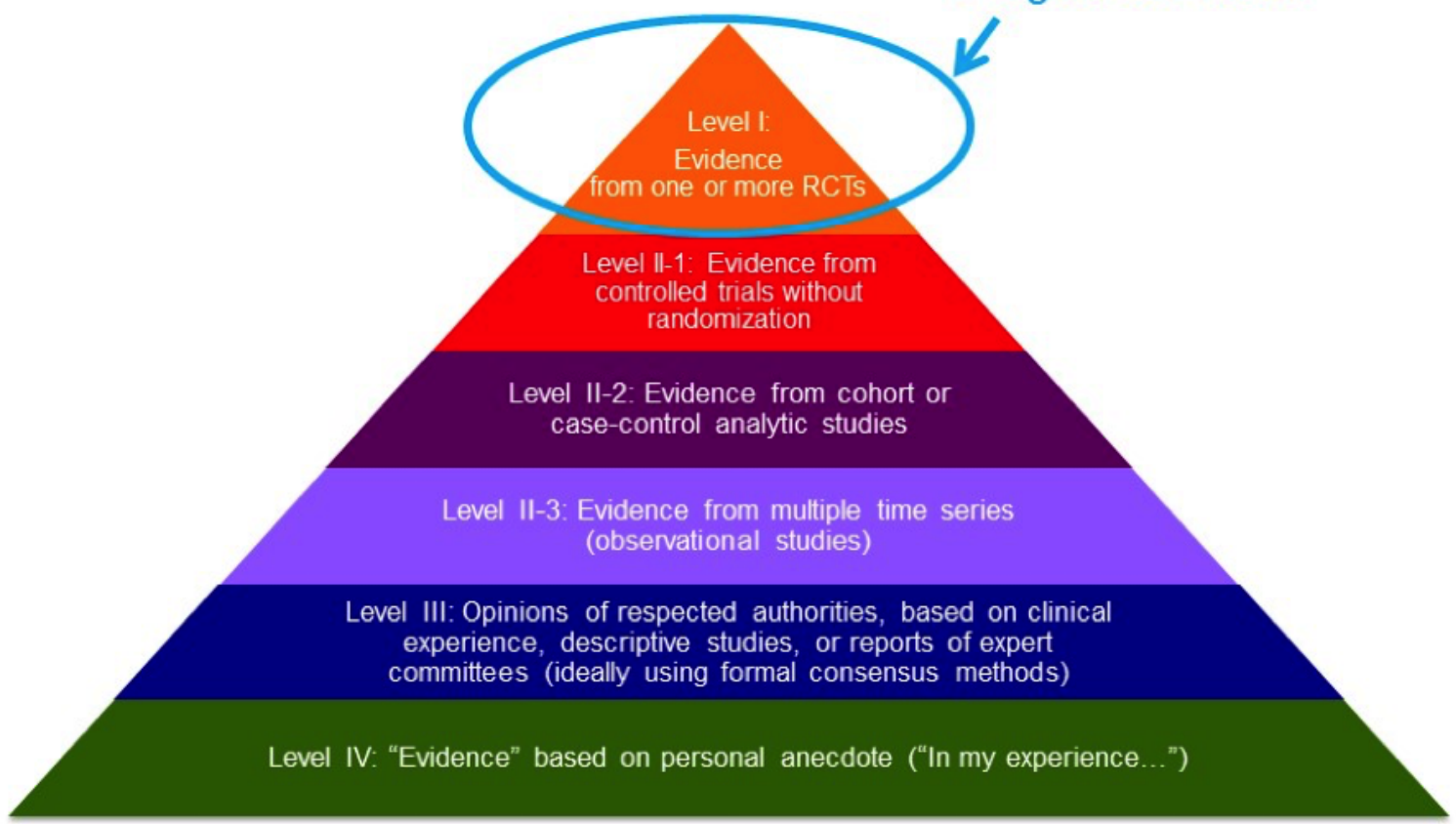
L. Iserin; Necker, HEGP

Archibald Leman Cochrane (1909 –1988)

prisonnier de guerre, medecin à Salonique , auteur de
Effectiveness and Efficiency: Random Reflections on Health Services

the hero of the book is the randomized control trial, and the villains are the clinicians in the "care" part of the National Health Service (NHS) who either fail to carry out such trials or succeed in ignoring the results if they do not fit in with their own preconceived ideas"

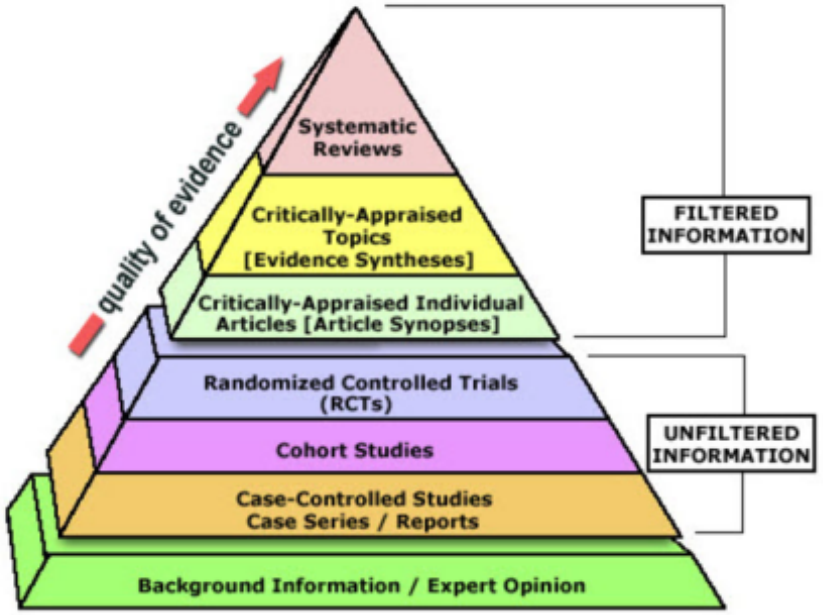
Levels of Evidence



EBM

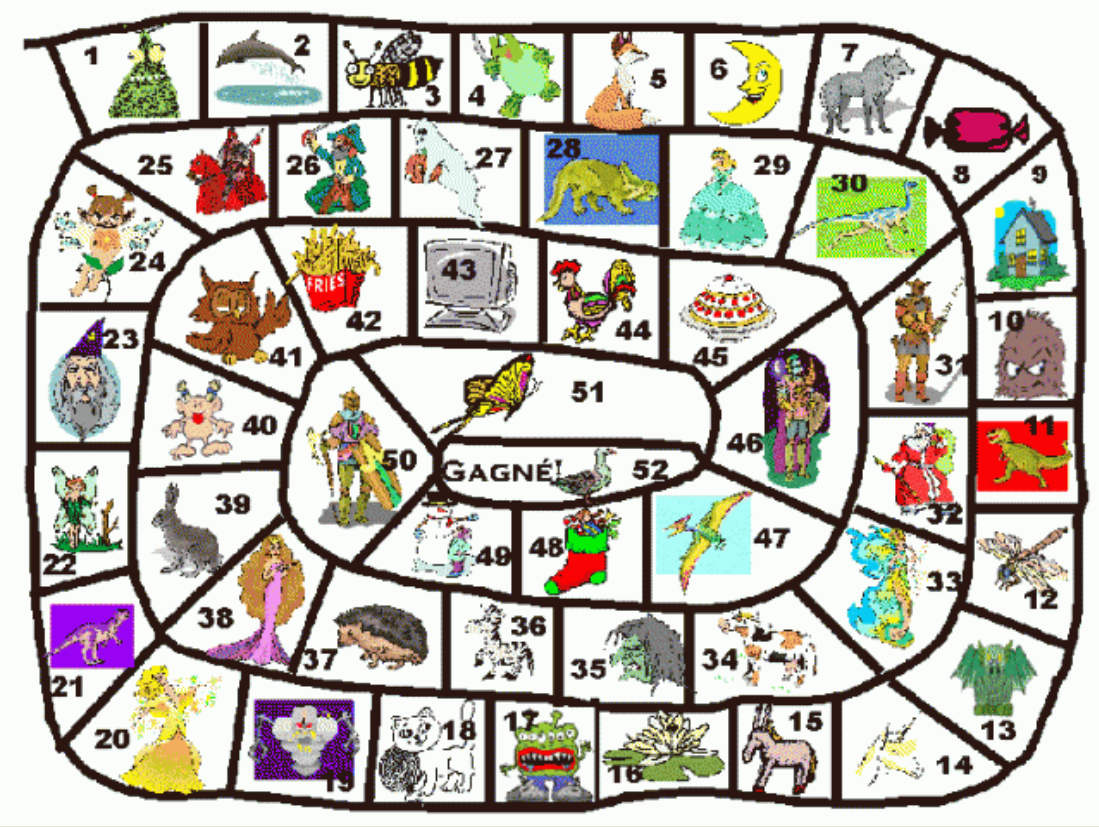
Levels of evidence pyramid

The levels of evidence pyramid provides a way to visualize both the quality of evidence and the amount of evidence available. For example, systematic reviews are at the top of the pyramid, meaning they are both the highest level of evidence and the least common. As you go down the pyramid, the amount of evidence will increase as the quality of the evidence decreases.



Produced by Jan Glover, David Izzo, Karen Odato and Lei Wang.

Congenital adulte : un long parcours semé d embuches



démarche EBM

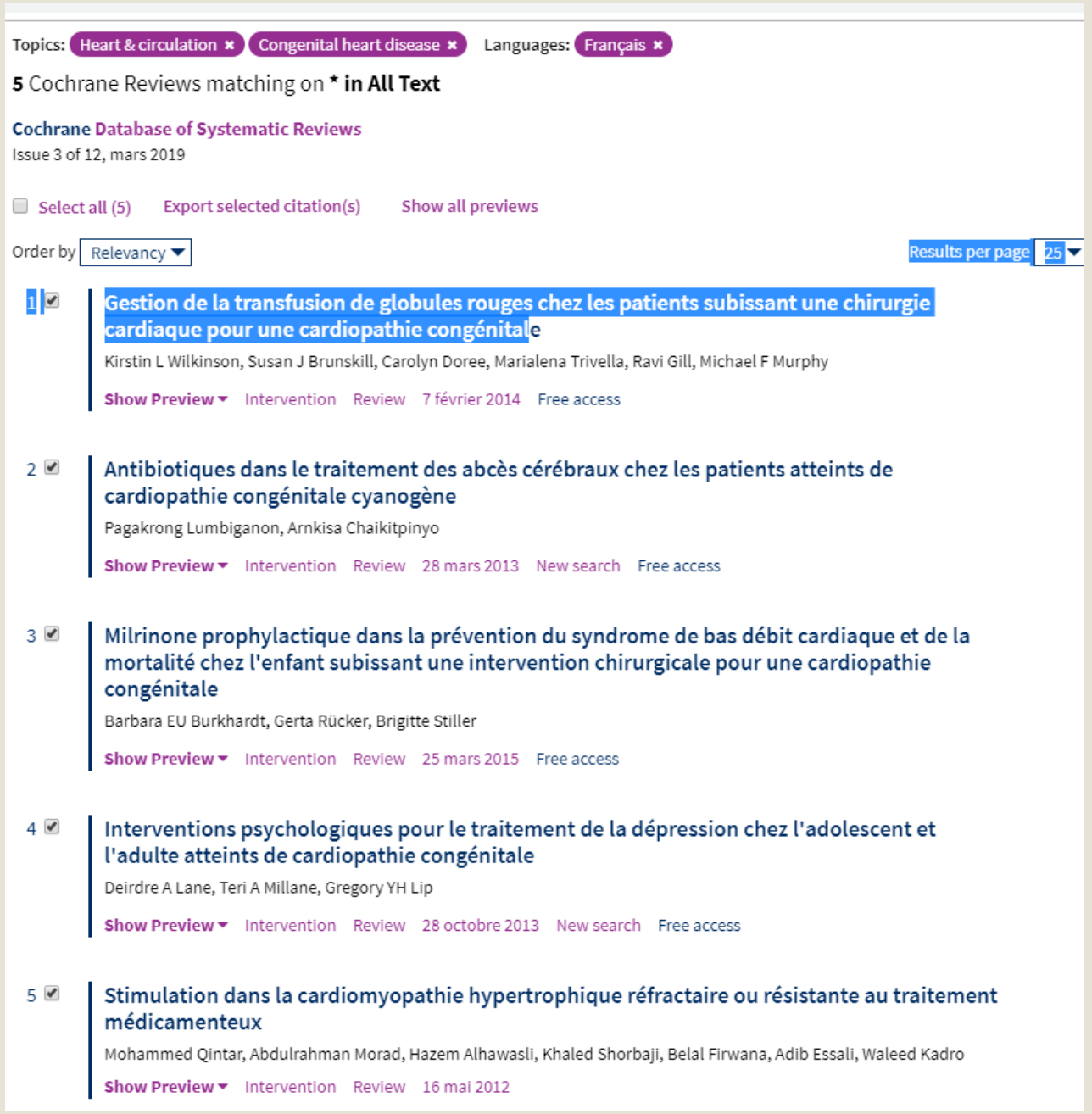
- 3 composantes :

  1. L'expérience clinique du praticien
  2. Les meilleures données actuelles (preuves) de la recherche clinique
  3. Les préférences du patient

- **Étapes d'une démarche EBM**
- Pour résoudre un problème clinique concernant un patient donné, la démarche EBM suit 4 étapes :

  1. Formuler le problème médical en une question claire et précise
  2. Rechercher dans la littérature des articles pertinents (du point de vue méthodologique) pour la question posée
  3. Évaluer la validité et la pertinence des résultats trouvés
  4. Intégrer les résultats retenus à son patient

cochrane



Topics: Heart & circulation ✖ Congenital heart disease ✖ Languages: Français ✖

5 Cochrane Reviews matching on * **in All Text**

Cochrane Database of Systematic Reviews
Issue 3 of 12, mars 2019

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1 **Gestion de la transfusion de globules rouges chez les patients subissant une chirurgie cardiaque pour une cardiopathie congénitale**
Kirstin L Wilkinson, Susan J Brunskill, Carolyn Doree, Marialena Trivella, Ravi Gill, Michael F Murphy
Show Preview Intervention Review 7 février 2014 Free access

2 **Antibiotiques dans le traitement des abcès cérébraux chez les patients atteints de cardiopathie congénitale cyanogène**
Pagakrong Lumbiganon, Arnkisa Chaikitpinyo
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3 **Milrinone prophylactique dans la prévention du syndrome de bas débit cardiaque et de la mortalité chez l'enfant subissant une intervention chirurgicale pour une cardiopathie congénitale**
Barbara EU Burkhardt, Gerta Rücker, Brigitte Stiller
Show Preview Intervention Review 25 mars 2015 Free access

4 **Interventions psychologiques pour le traitement de la dépression chez l'adolescent et l'adulte atteints de cardiopathie congénitale**
Deirdre A Lane, Teri A Millane, Gregory YH Lip
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5 **Stimulation dans la cardiomyopathie hypertrophique réfractaire ou résistante au traitement médicamenteux**
Mohammed Qintar, Abdulrahman Morad, Hazem Alhawasli, Khaled Shorbaji, Belal Firwana, Adib Essali, Waleed Kadro
Show Preview Intervention Review 16 mai 2012

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Topics: Heart & circulation ✖ Congenital heart disease ✖

3 Cochrane Protocols matching on *** in All Text**

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1 **Routine screening by echocardiography to reduce morbidity and mortality from congenital heart disease in neonates with Down syndrome**
William McGuire, Peter W Fowlie
Show Preview Intervention Protocol 20 juillet 2005

2 **Prophylactic corticosteroids for paediatric heart surgery with cardiopulmonary bypass**
Ben Gibbison, José Carlos Villalobos Lizardi, Karla Isis Avilés Martínez, Daniel P Fudulu, Miguel Angel Medina Andrade, Giordano Pérez-Gaxiola, Alvin WL Schadenberg, Serban C Stoica, Stafford L Lightman, Gianni D Angelini, Barnaby C Reeves
Show Preview Intervention Protocol 30 août 2018

3 **Interventions to increase physical activity for people with congenital heart disease**
Susanne H Klausen, Roselien Buys, Lars Louis Andersen, Asle Hirth, Brian W McCrindle, Hanne Kjaergaard, Jørn Wetterslev
Show Preview Intervention Protocol 22 janvier 2018 Withdrawn Free access

Questions

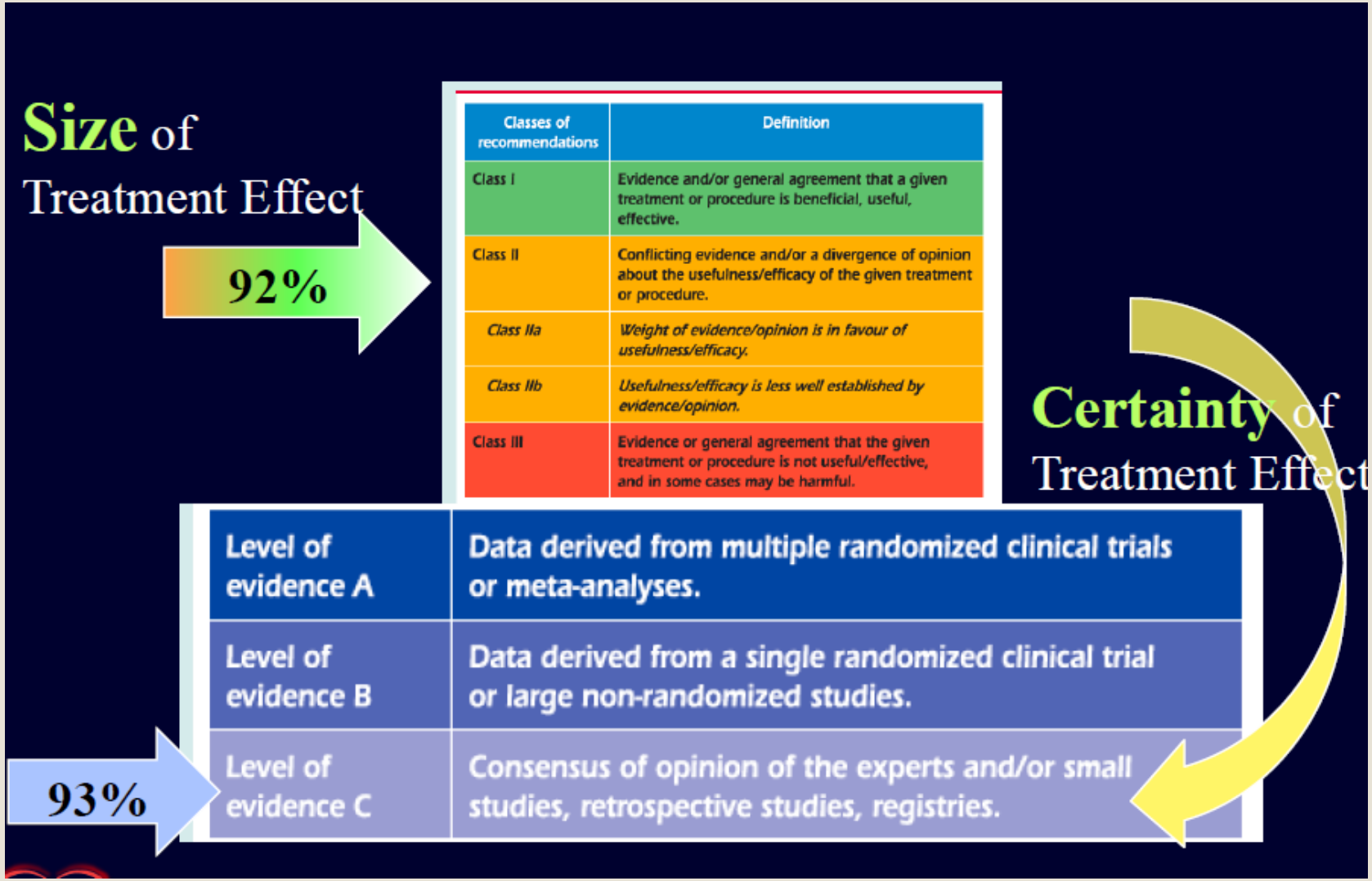
- comment peut on suivre longtemps une cohorte?
- En quoi le suivi des cardiopathies adultes change la pratique des cardiopédiatres?
- Quelle aorte dans les années à venir pour les switch

- quel traitement médical (ou chirurgical) pour les vd systémiques?
- comment prévenir la mort subite?
- Quel traitement pour l'hta séquellaire des coarctations?
- Cardiopathie congénitale et cardiopathie acquises en fait on assez en terme de prévention cardio vasculaire

« Les grands absents pour l'instant…»

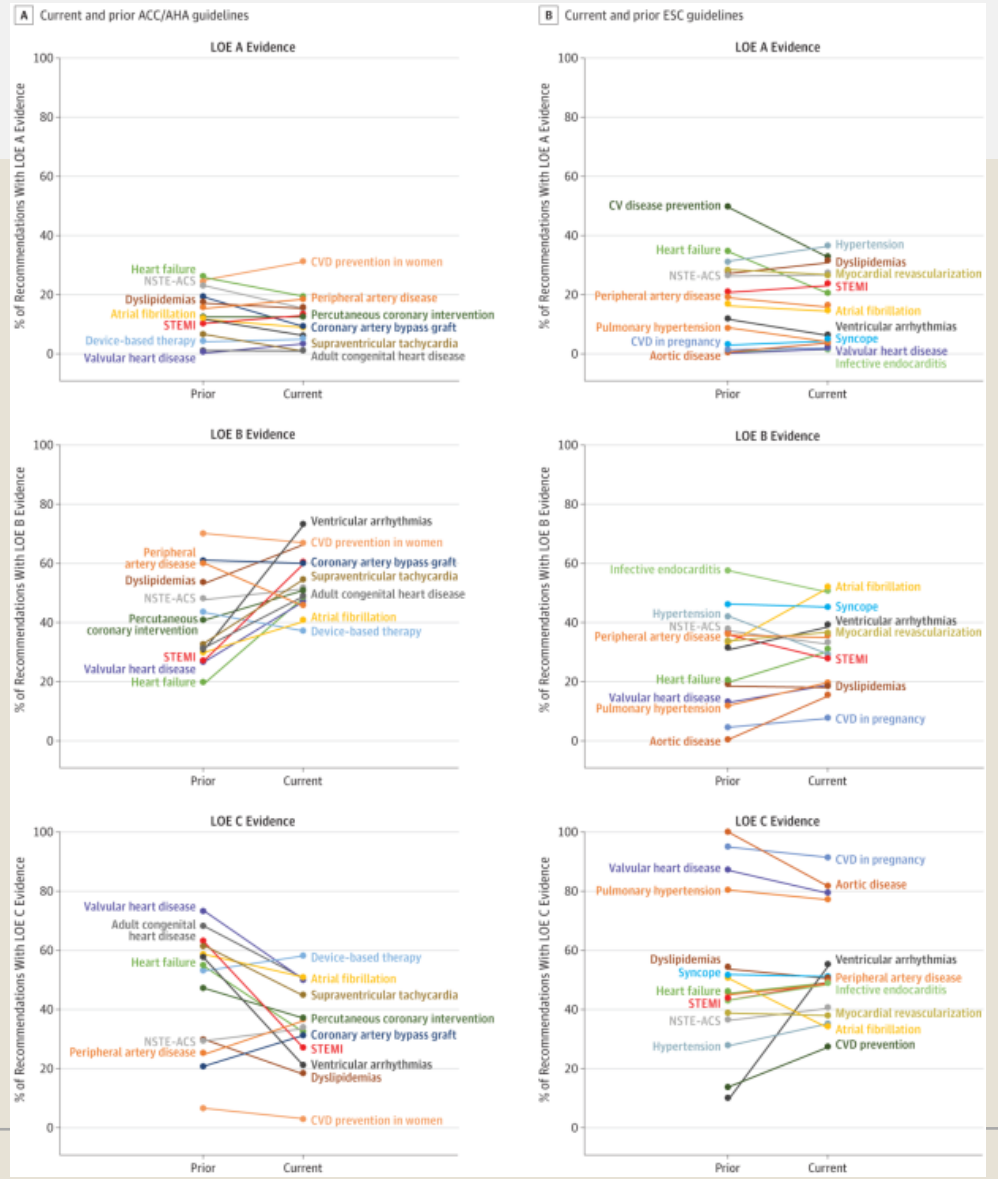
- Hypo VG
- tronc artériel commun

Consensus d experts, ESC guch recommandations , From Marelli, JESFC2019



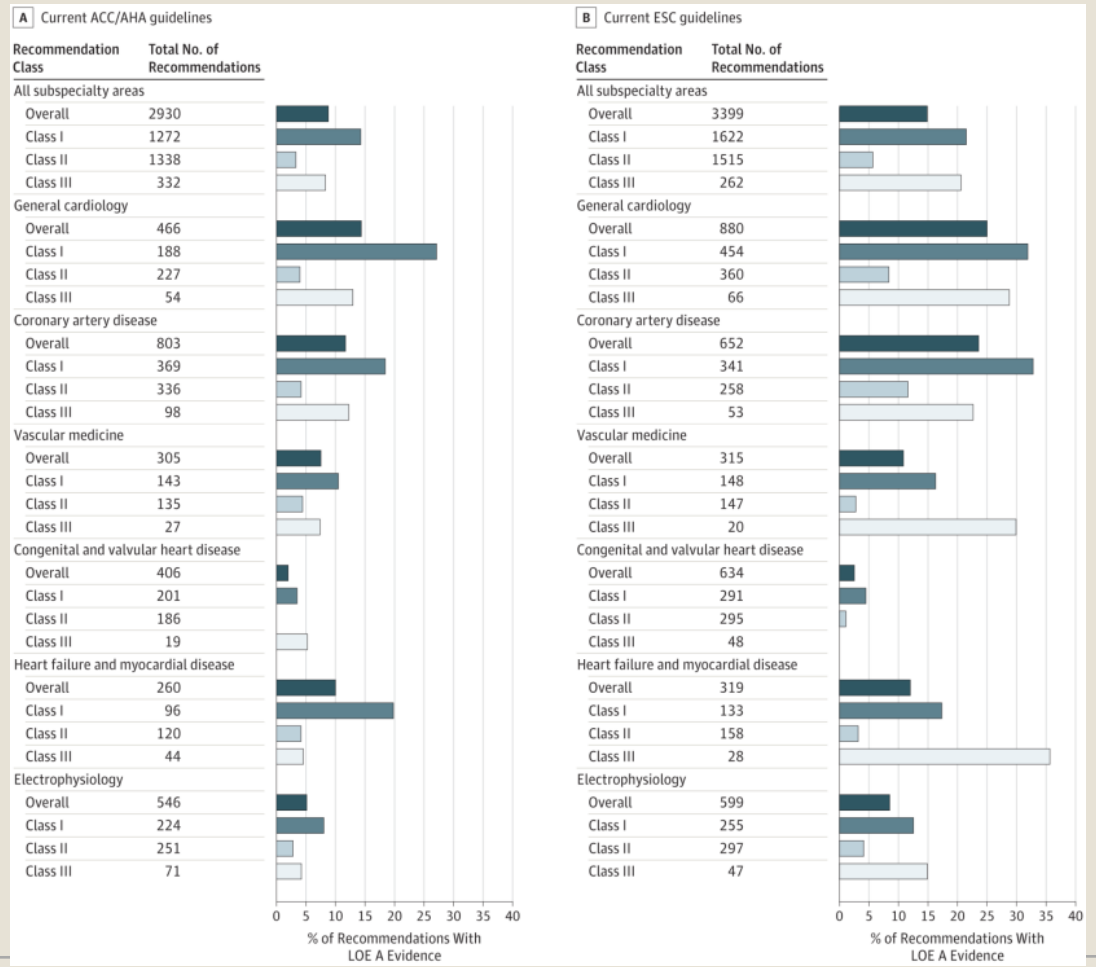
Levels of Evidence Supporting American College of Cardiology/American Heart Association and European Society of Cardiology Guidelines, 2008-2018

JAMA. 2019



From: **Levels of Evidence Supporting American College of Cardiology/American Heart Association and European Society of Cardiology Guidelines, 2008-2018**

JAMA. 2019;321(11):1069-1080. doi:10.1001/jama.2019.1122



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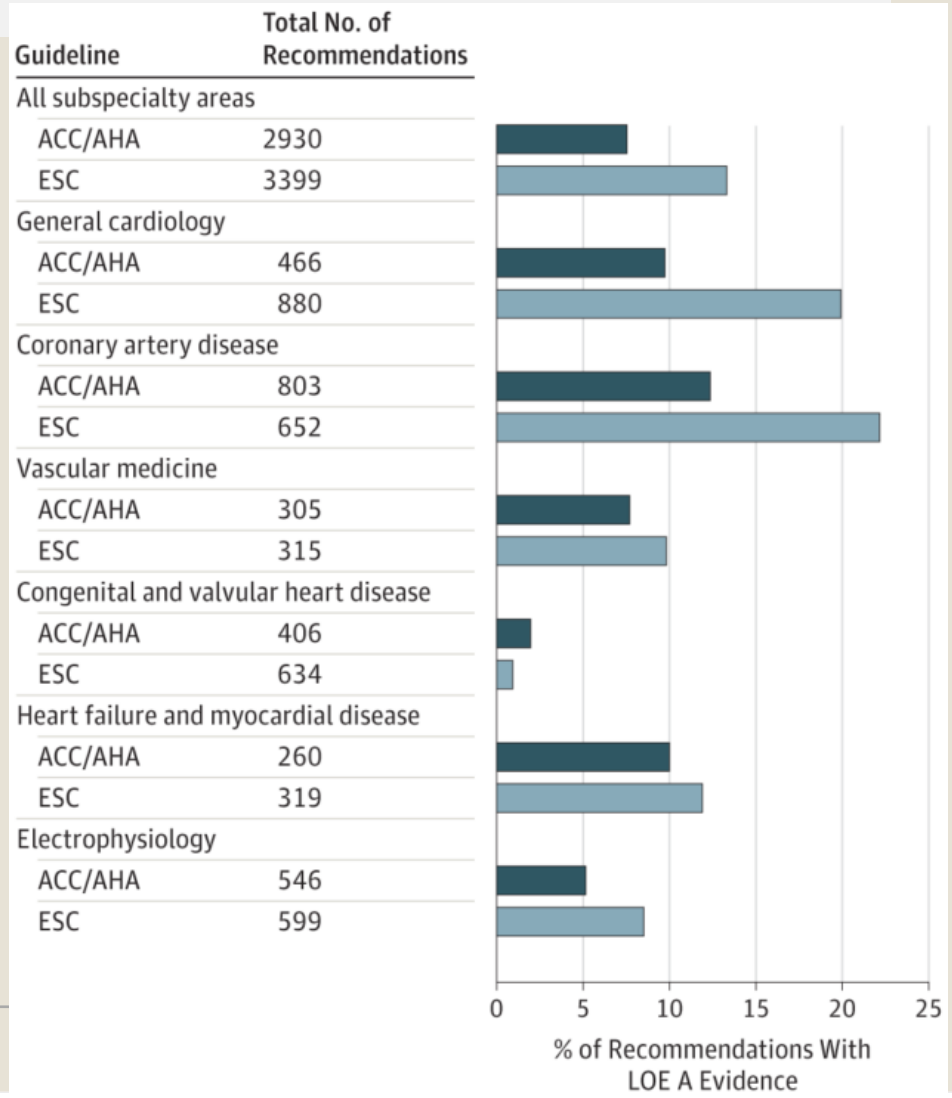
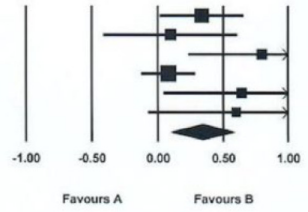
JAMA. 2019;321(11):1069-1080. doi:10.1001/jama.2019.1122



Patient outcomes after transcatheter and surgical pulmonary valve replacement for pulmonary regurgitation in patients with repaired tetralogy of Fallot: A quasi-meta-analysis

Meta Analysis VO2 Max TC PVR

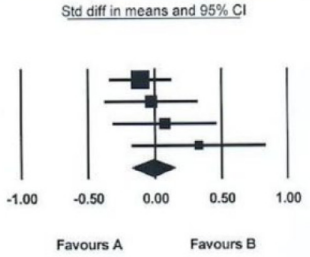
Study name	Std diff in means	Standard error	Variance	Lower limit	Upper limit	Z-Value	p-Value
Borik et al 2015	0.336	0.163	0.026	0.017	0.654	2.067	0.039
Coats et al 2007	0.098	0.259	0.067	-0.409	0.605	0.379	0.705
Khambadkone et al 2005	0.799	0.287	0.082	0.236	1.362	2.783	0.005
McElhinney et al 2010	0.080	0.104	0.011	-0.123	0.284	0.774	0.439
Muller et al 2014	0.643	0.305	0.093	0.045	1.240	2.109	0.035
Wilson et al 2015	0.601	0.344	0.118	-0.073	1.274	1.748	0.080
	0.343	0.123	0.015	0.103	0.584	2.797	0.005



Pooled pre-PVR VO2 max = 31.69 mL/kg/min
Pooled post-PVR VO2 max = 34.76 mL/kg/min

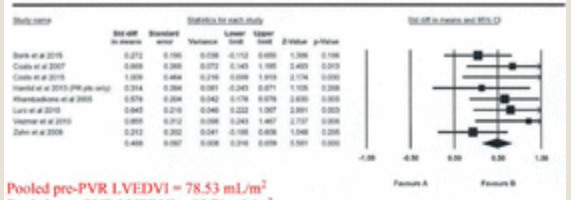
Meta Analysis VO2 Max Surgical PVR

Study name	Std diff in means	Standard error	Variance	Lower limit	Upper limit	Z-Value	p-Value
Frigiola 2012	-0.107	0.117	0.014	-0.337	0.123	-0.913	0.361
Gorter 2014	-0.027	0.177	0.031	-0.374	0.319	-0.153	0.878
Ho 2015	0.074	0.196	0.039	-0.311	0.459	0.376	0.707
Tsang 2010	0.329	0.257	0.066	-0.174	0.832	1.282	0.200
	-0.012	0.083	0.007	-0.174	0.150	-0.144	0.885



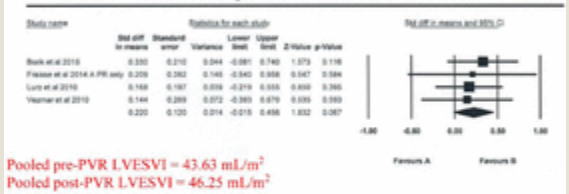
Pooled pre-PVR VO2 max = 26.75 mL/kg/min
Pooled post-PVR VO2 max = 27.15 mL/kg/min

Meta Analysis LVEDVI TC PVR



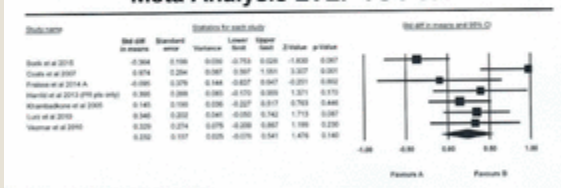
Pooled pre-PVR LVEDVI = 78.53 mL/m²
Pooled post-PVR LVEDVI = 85.71 mL/m²

Meta Analysis LVESVI TC PVR



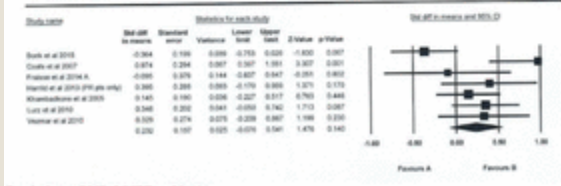
Pooled pre-PVR LVESVI = 43.63 mL/m²
Pooled post-PVR LVESVI = 46.25 mL/m²

Meta Analysis LVEF TC PVR



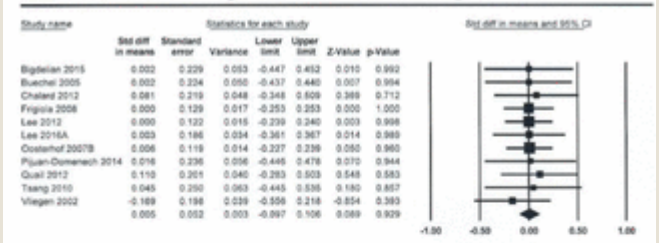
Pooled pre-PVR LVEF = 58.4%
Pooled pre-PVR LVEF = 58.35%

Meta Analysis LVEF TC PVR



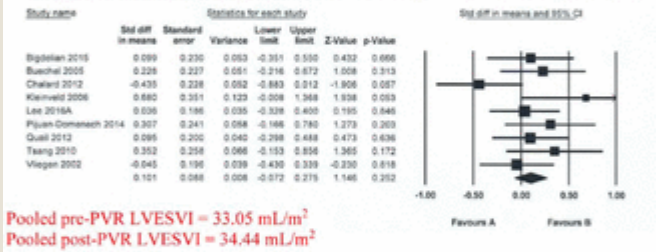
Pooled pre-PVR LVEF = 58.4%
Pooled pre-PVR LVEF = 58.35%

Meta Analysis LVEDVI Surgical PVR



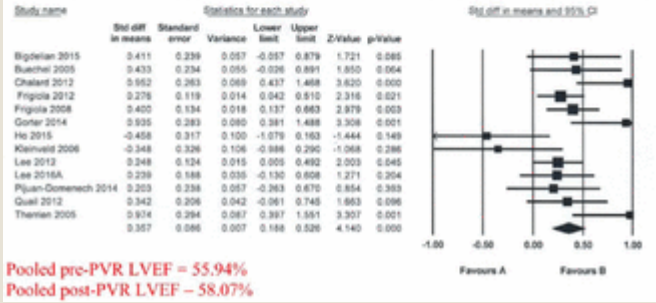
Pooled pre-PVR LVEDVI = 76.26 mL/m²
Pooled post-PVR LVEDVI = 82.03 mL/m²

Meta Analysis LVESVI Surgical PVR



Pooled pre-PVR LVESVI = 33.05 mL/m²
Pooled post-PVR LVESVI = 34.44 mL/m²

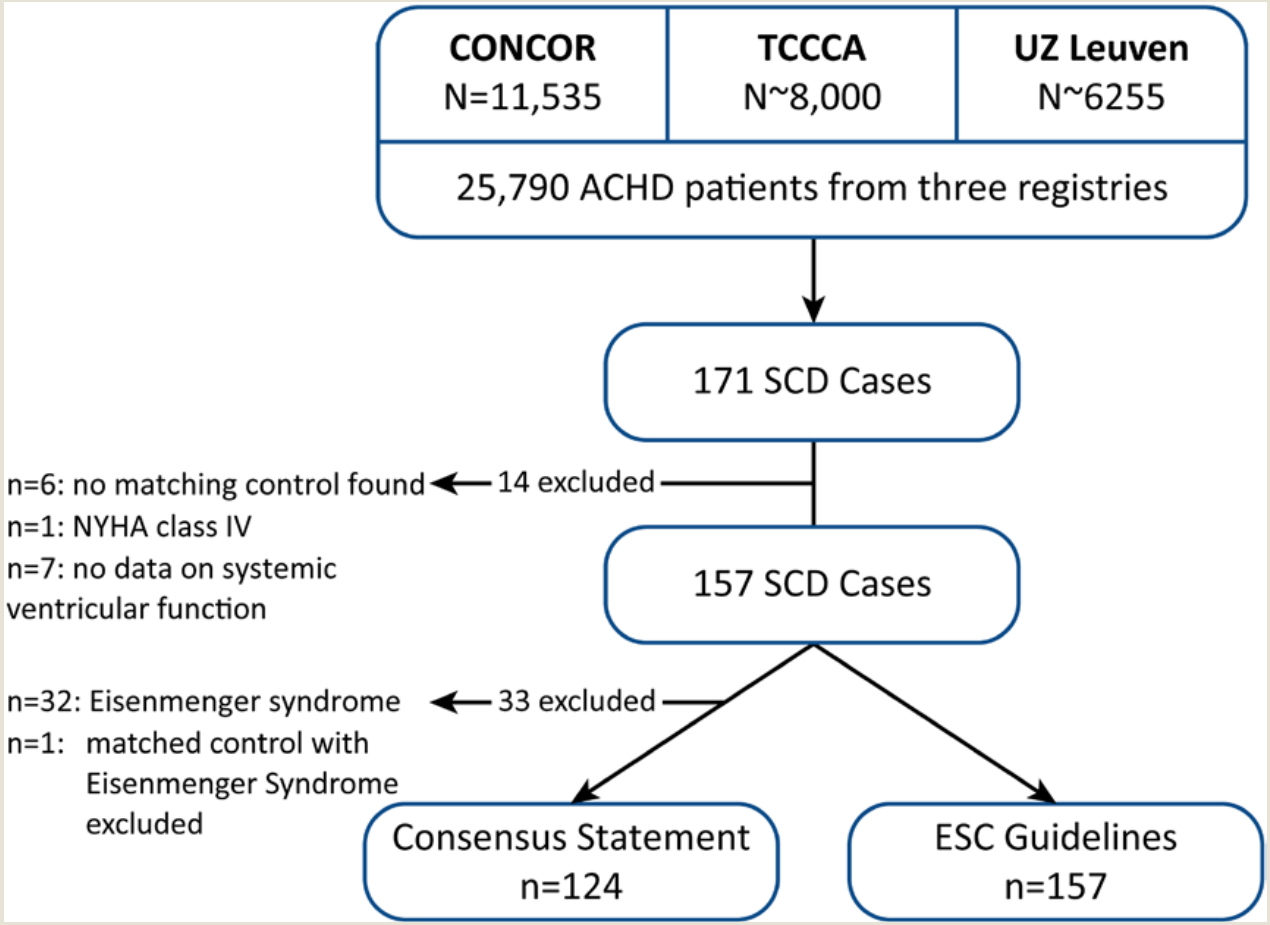
Meta Analysis LVEF Surgical PVR



Pooled pre-PVR LVEF = 55.94%
Pooled post-PVR LVEF – 58.07%

European Journal of Cardiovascular Nursing, vol. 16, 2017.

Prevention of Sudden Cardiac Death in Adults With Congenital Heart Disease Do the Guidelines Fall Short?

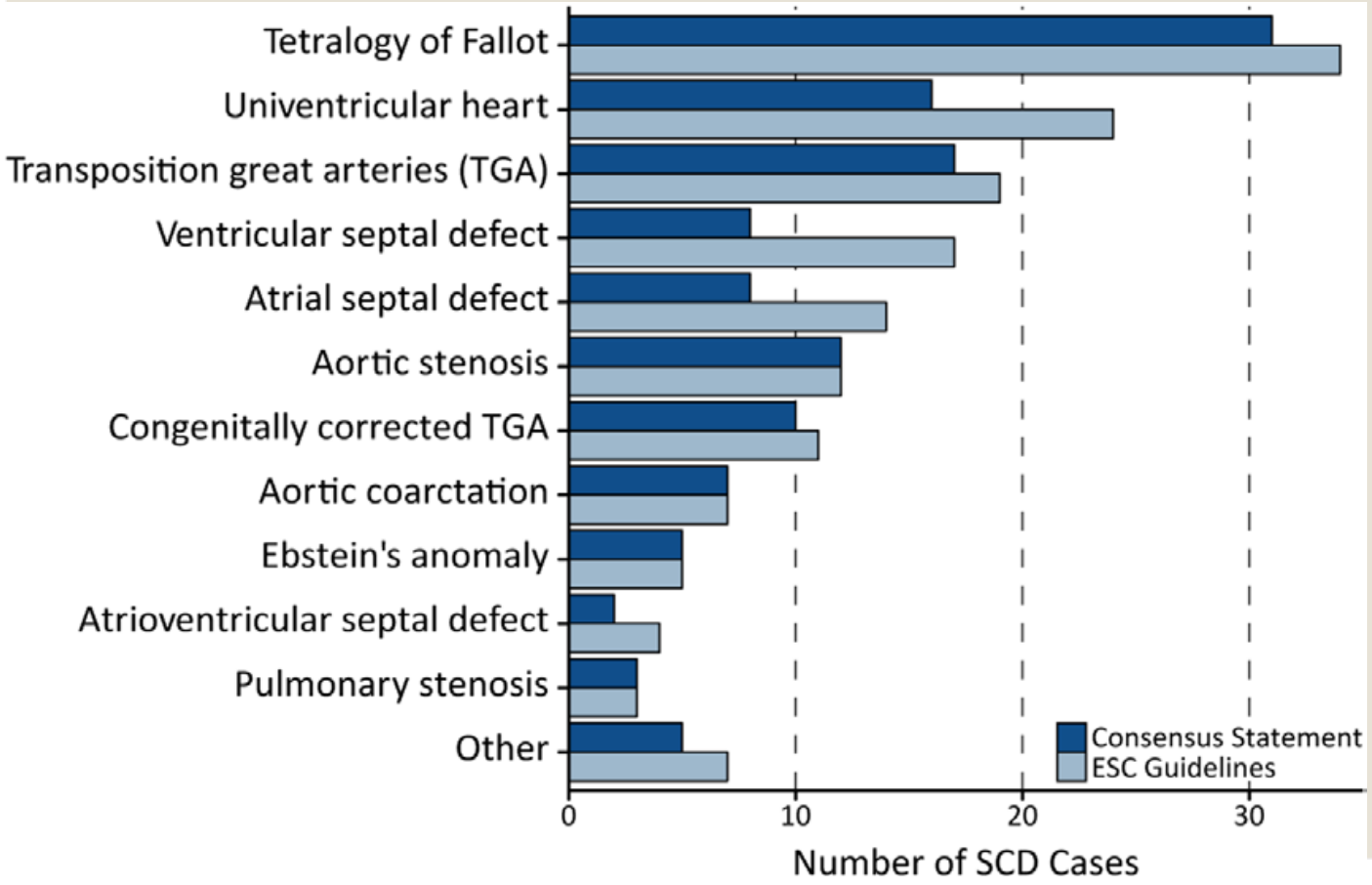


Vehmeijer; ***Circ Arrhythm Electrophysiol*. 2017**

Prevention of Sudden Cardiac Death in Adults With Congenital Heart Disease Do the Guidelines Fall Short?

- primary prevention
- class I, systemic left ventricular ejection fraction
- ≤35%, biventricular physiology, and NYHAII or III symptoms;
- class IIa, adults with ToF and multiplerisk factors for SCD, such as left ventricular systolic or diastolic dysfunction, nonsustained ventricular tachycardia, QRS duration ≥180 ms, extensive right ventricular scarring, or inducible sustained ventricular tachycardia at electrophysiological study;
- class IIb adults with a single or systemic right ventricular ejection fraction <35%, particularly in the presence of additional risk factors, such as complex ventricular arrhythmias (defined as nonsustained ventricular tachycardia), unexplained syncope, NYHA II or III
- symptoms, QRS duration ≥140 ms, or severe systemic AV-valve
- regurgitation.

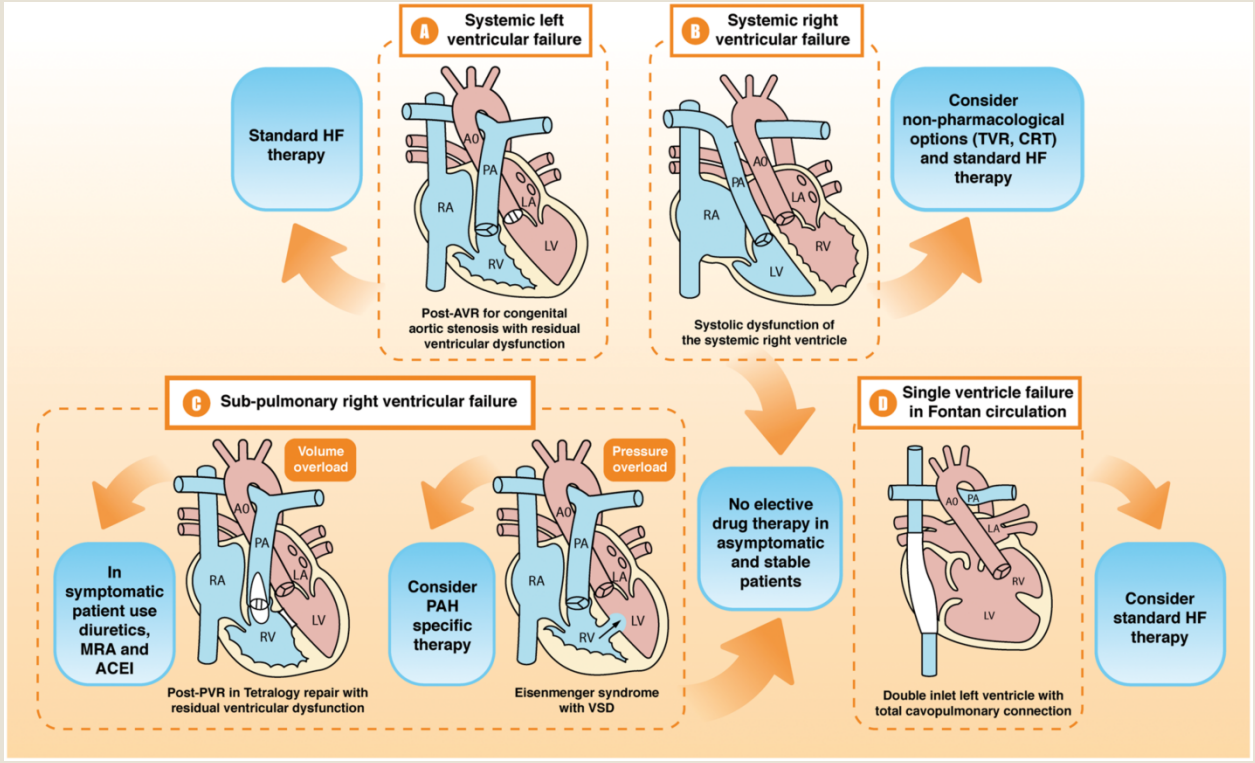
Prevention of Sudden Cardiac Death in Adults With Congenital Heart Disease
Do the Guidelines Fall Short?



60% des Morts subites n'avaient pas eu d indication a un défibrillateur selon les recommandations

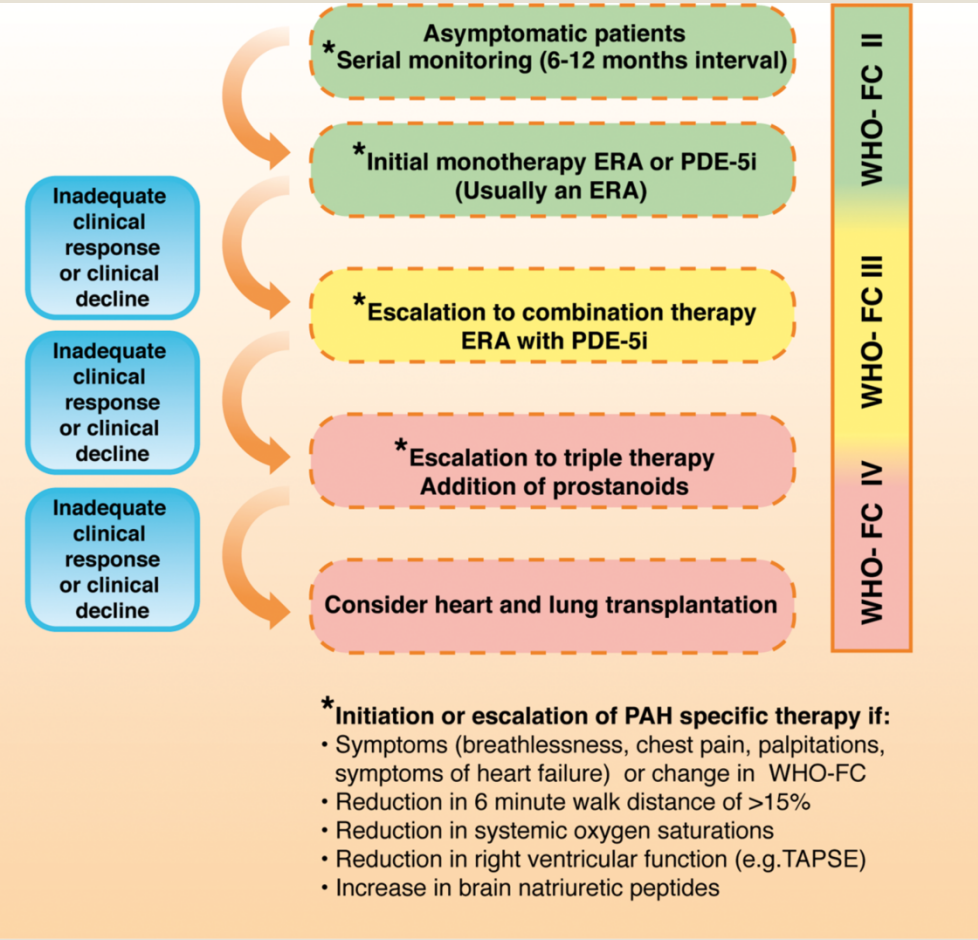
Vehmeijer; ***Circ Arrhythm Electrophysiol*. 2017**

Heart failure therapy for different adult congenital heart disease subgroups



European Heart Journal, 2018,

Suggested treatment algorithm for patients with Eisenmenger syndrome.

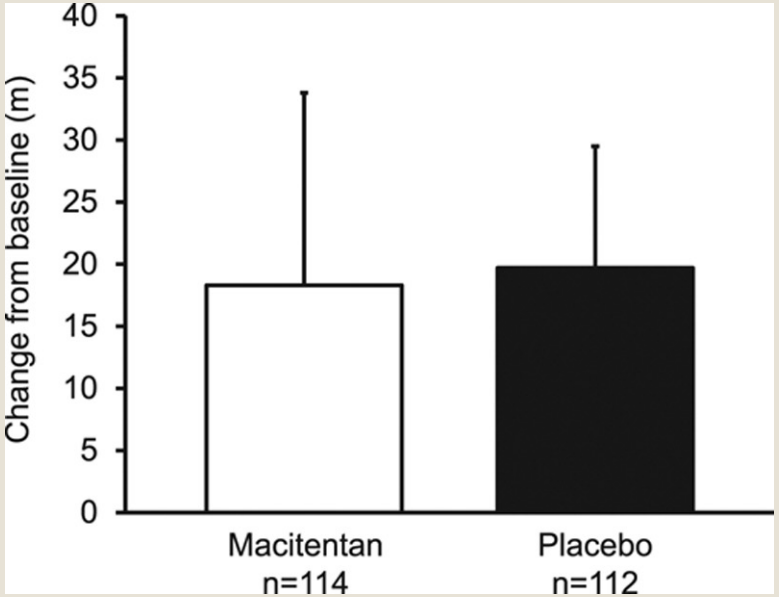


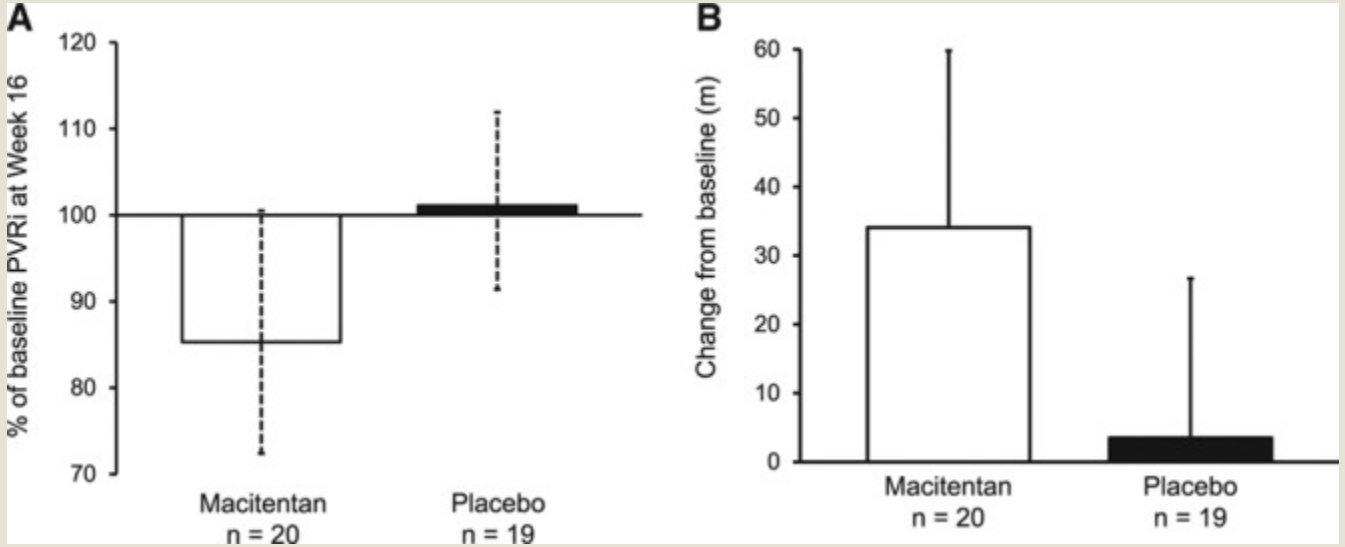
European Heart Journal, 21 August 2018, ehy480, https://doi.org/10.1093/eurheartj/ehy480

Medical Therapy for Systemic Right Ventricles: A Systematic Review for the 2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

First Author	Sample (N) Exp/Obs	Male (N)	Age Mean or Median (y) (SD or Range) Exp/Obs	Intervention	Diagnosis	Mean Follow-Up Time (mo)
RCT						
Dos et al. (19) 23972966	14*/12	8 (T) 8 (C)	24.9±4.3 (T) 28.3±6.1 (C)	Eplerenone	Abff	12
Therrien et al. (20) 18672299	8/9	3 (T) 8 (C)	27±5.7 (T) 26±5.2 (C)	ACE	Abff	12
Van der Bom et al. (17) 23247302	44/44	29 (T) 28 (C)	33±10 (T) 33±10 (C)	ARB	CCTGA/ Abff	38
Other						
Dore et al. (18)† 16216961	29	24	30.3±10.9	ARB	CCTGA/ Abff	4
Hechter et al. (31) 11230861	14	12	31 (26, 42)	ACE	Abff	24
Tutarel et al. (21) 20843567	14/14	11 (T) 10 (C)	25.2±3.5 (T) 24.6±2.3 (C)	ARB	Abff	13
Giardini et al. (33)‡ 21882492	8	5	26 (18, 31)	BB	CCTGA/ Abff	12
Doughan et al. (35)‡ 17317376	31/29	20 (T) 18 (C)	29±6 (T) 27±6 (C)	BB	Abff	10
Bouallal et al. (32)‡ 20519056	14	7	35 (24, 57)	BB	SV	13
Josephson et al. (34)‡ 16835671	8	5	29 (22, 37)	BB	Abff	36

Exemple de RCT: Maestro trial





Gatzoulis, Circulation. 2019

Challenges to multicenter research in adults with congenital heart disease.

- ..Complexity and heterogeneity of the patient population
- ..Divergences in nomenclature and classification
- ..Institutional variations in treatment strategies
- ..Identification of appropriate clinically relevant end points
- ..Paucity of preliminary data for sample size estimates
- ..Recruitment of a sufficiently large number of patients
- ..Issues regarding patient safety
- ..Research infrastructure for efficient implementation
- ..Limited funding opportunities

Khairy , future cardiology 2012

Répartition des patients

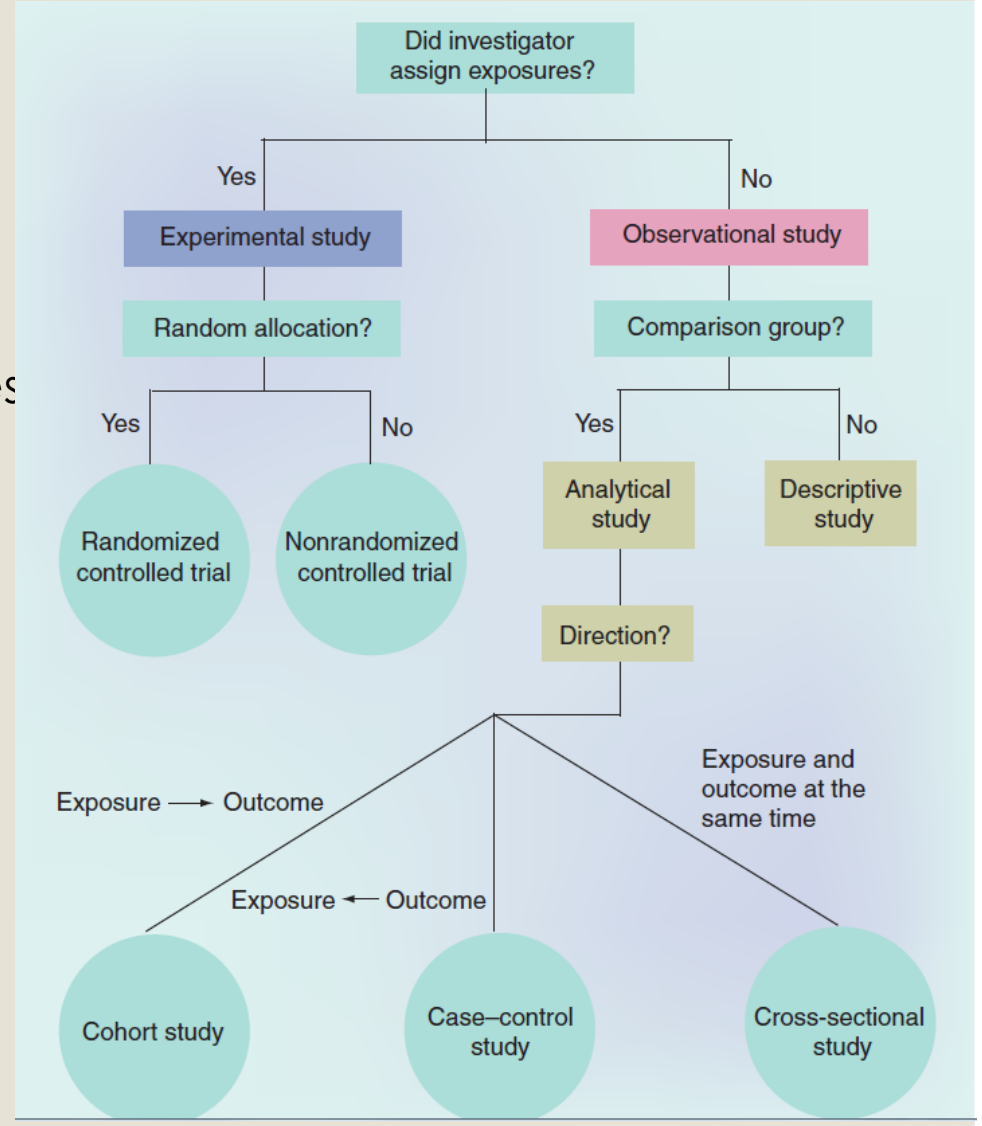
Table 1. Prevalence of selected moderate and severe forms of congenital heart disease in the Quebec population in 2000.

Congenital defect	n	Prevalence ratio†
Aortic coarctation	783	1:9500
Endocardial cushion defect	1748	1:4000
Ebstein's anomaly	79	1:93,000
Tetralogy of Fallot/truncus arteriosis	1779	1:4000
Transposition of the great arteries	659	1:11,000
Univentricular hearts	363	1:20,000

Quebec population in 2000 = 7,357,029.
†Rounded to the nearest 500.

comment progresser?

- centraliser la prise en charge des patients
- Travailler en réseau
- Trouver des financements pour des études randomisées
- Trouver des jeunes avec des idées
- qui aient du temps….
- sans laisser les cliniciens à la mine



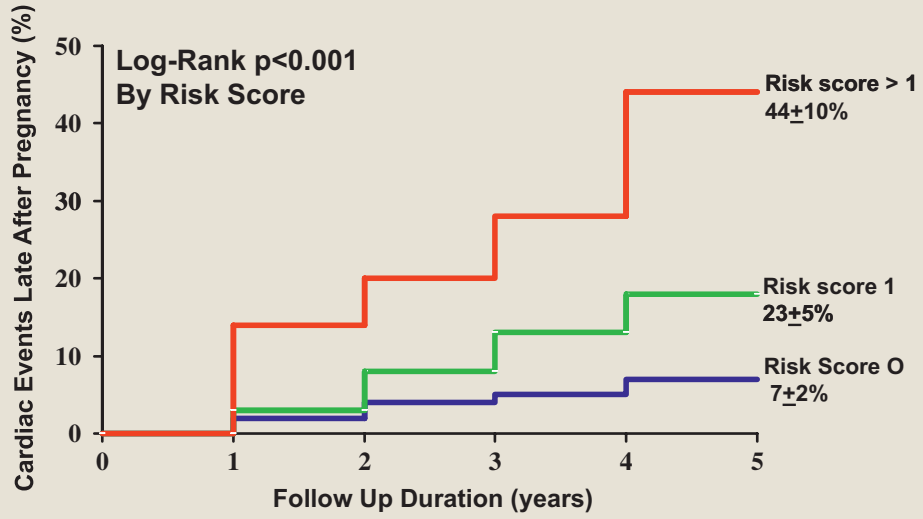
Richard Horton, directeur de la publication du Lancet , 2015

- A lot of what is published is incorrect."
- "
- *Contenant des études avec de petits échantillons, aux effets minimes, aux analyses exploratoires invalides, et avec des conflits d'intérêts évidents, avec l'obsession de suivre des tendances à la mode d'importance douteuse, la science a pris un virage vers l'obscurantisme. En pratique 'de mauvaises méthodes donnent des résultats'*

- ***Those who have the power to act seem to think somebody else should act first.***
- ***The good news is that science is beginning to take some of its worst failings very seriously.***
- ***The bad news is that nobody is ready to take the first step to clean up the system.***

318 women, 405 pregnancies, mean follow up 2.6 years

12% of late cardiac events



Risk score:
NYHA>2 or resting cyanosis

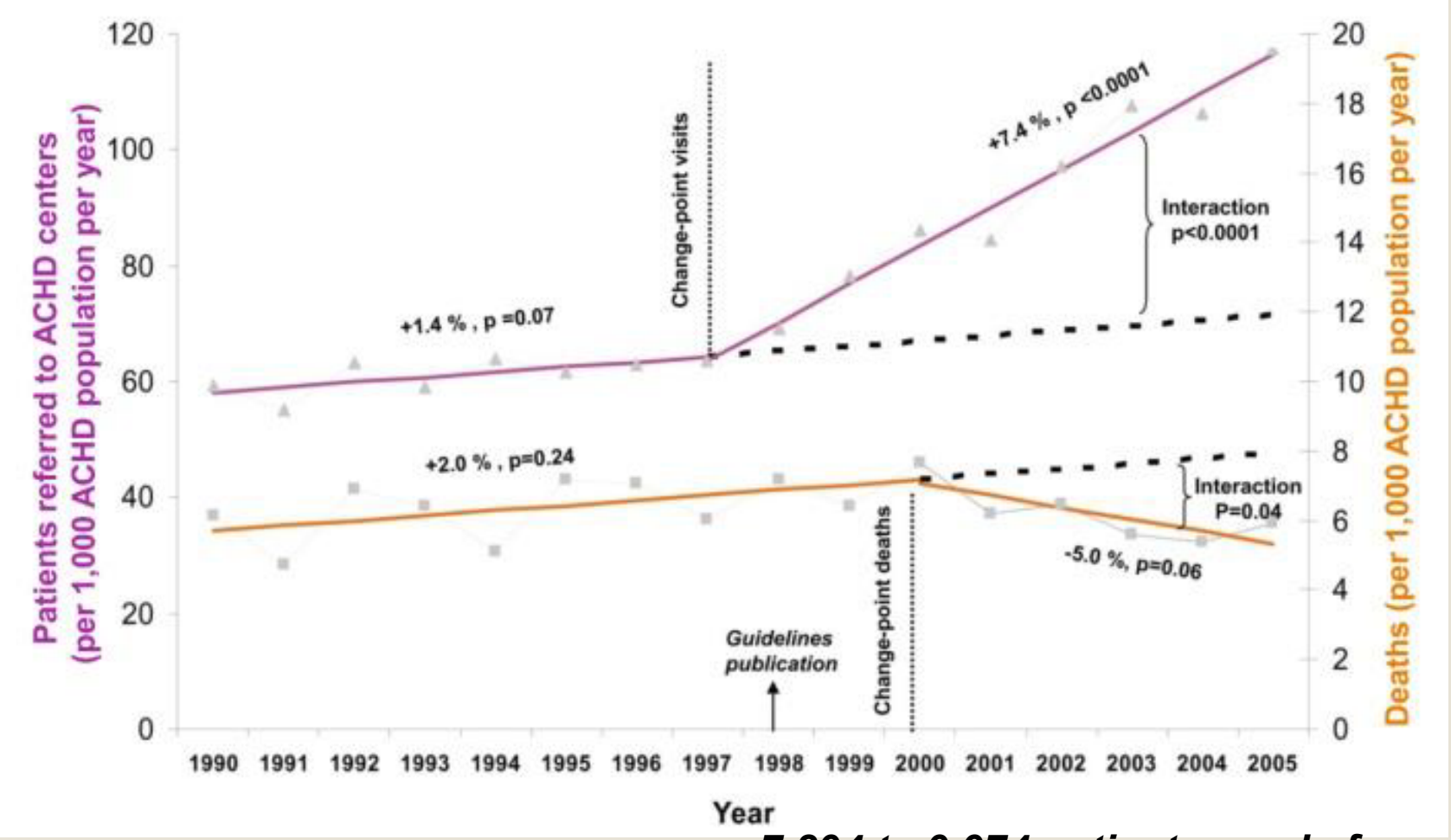
Dysfonction du ventricule sous-aortique or significative PR

LV Obstruction

Evénements cardiaques avant ou pendant la grossesse

Balint et al. *Heart 2010*

L EBM pour les structures médicales



7,894 to 9,974 patients yearly from 1990-2005

Mylotte et al Circulation 2014;129:1804

Improving heart disease knowledge and research participation in adults with congenital heart disease (The Health, Education and Access Research Trial: HEART-ACHD)☆,☆☆

Anne Marie Valente [a,*], Michael J. Landzberg [a], Ann Gianola [b], Amy J. Harmon [a], Stephen Cook [c], Jennifer G. Ting [d], Karen Stout [e], Karen Kuehl [f], Paul Khairy [g], Joseph D. Kay [h], Michael Earing [i], Linda Houser [j], Craig Broberg [k], Carly Milliren [a], Alexander R. Opotowsky [a], Gary Webb [l], Amy Verstappen [b], Michelle Gurvitz [a], for the Alliance for Adult Research in Congenital Cardiology (AARCC) Investigators and the Adult Congenital Heart Association (ACHA)

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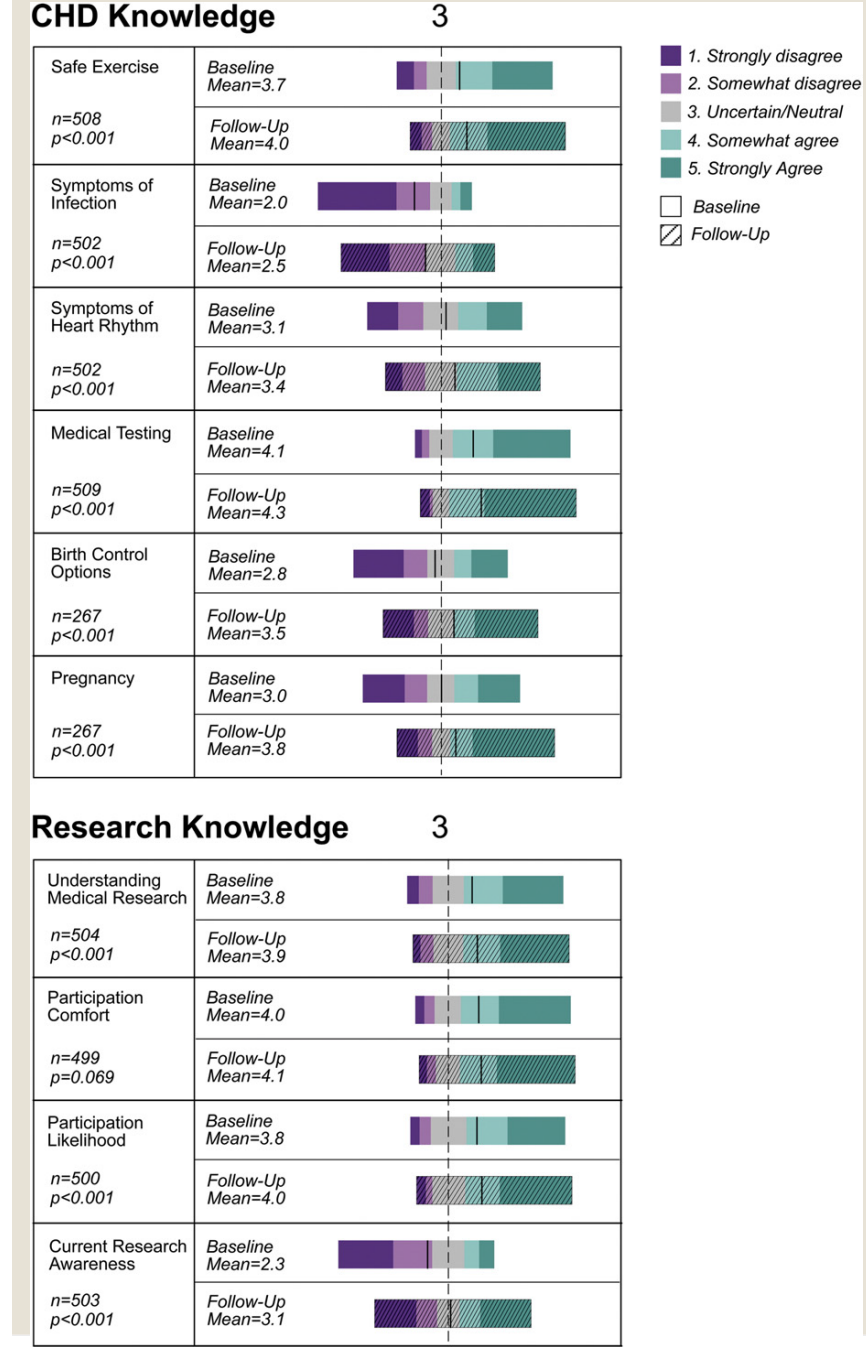
ABSTRACT

Objective: The objective of this prospective multi-center study was to evaluate heart disease knowledge within the adult congenital heart disease (ACHD) population, pilot an educational intervention and assess interest in research participation among new patients at ACHD clinics.
Background: Many adults with congenital heart disease lack knowledge about their heart condition that may contribute to undesirable outcomes.
Methods: Patients ≥ 18 years of age were recruited upon their first presentation to an ACHD clinic and underwent an educational intervention consisting of creation of a personal health information 'passport' and an introduction to web-based resources. Subjects were asked to complete initial and follow-up surveys documenting their perceived knowledge.
Results: Nine hundred twenty-two subjects were recruited from 12 ACHD centers, and 520 (57%) completed follow-up surveys. Patients who completed the follow-up survey were more likely to be women, have more education, and have mild heart disease. At follow-up, the ability of the subjects to name their heart condition improved (78% to 83%, $p = 0.002$). Improvements were seen in mean Likert items regarding perceived knowledge of appropriate exercise ($p < 0.0001$), symptoms of heart rhythm problems or endocarditis ($p < 0.0001$), reasons for cardiac tests ($p < 0.007$), and birth control options and pregnancy safety ($p < 0.0001$). On follow-up, subjects reported a better understanding of medical research ($p < 0.01$), and higher interest in research participation ($p < 0.003$).
Conclusion: This joint clinician–patient pilot program will help inform future efforts toward patient education and participation in research with a focus on standardization of protocols for life-long longitudinal follow-up and continued multi-center collaboration in the ACHD population.



Predicting deterioration of ventricular function in patients with repaired tetralogy of Fallot using machine learning.

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Abstract

AIMS: Previous studies using regression analyses have failed to identify which patients with repaired tetralogy of Fallot (rTOF) are at risk for deterioration in ventricular size and function despite using common clinical and cardiac function parameters as well as cardiac mechanics (strain and dyssynchrony). This study used a machine learning pipeline to comprehensively investigate the predictive value of the baseline variables derived from cardiac magnetic resonance (CMR) imaging and provide models for identifying patients at risk for deterioration.

METHODS AND RESULTS: Longitudinal deterioration for 153 patients with rTOF was categorized as 'none', 'minor', or 'major' based on changes in ventricular size and ejection fraction between two CMR scans at least 6 months apart (median 2.7 years). Baseline variables were measured at the time of the first CMR. An exhaustive variable search with a support vector machine classifier and five-fold cross-validation was used to predict deterioration and identify the most useful variables. For predicting any deterioration (minor or major) vs. no deterioration, the mean area under the curve (AUC) was 0.82 ± 0.06. For predicting major deterioration vs. minor or no deterioration, the AUC was 0.77 ± 0.07. Baseline left ventricular (LV) ejection fraction, LV circumferential strain, and pulmonary regurgitation were most useful for achieving accurate predictions.

CONCLUSION: For the prediction of deterioration in patients with rTOF, a machine learning pipeline uncovered the utility of baseline variables that was previously lost to regression analyses. The predictive models may be useful for planning early interventions in patients with high risk.

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rapport entre quotient intellectuel et utilisation de la pensée rationnelle

Certaines personnes qui sont intellectuellement en mesure ne se donnent pas tellement la peine de s'engager

dans une pensée analytique et sont portées à se fier à leurs intuitions

D'autres ont davantage tendance à vérifier leurs intuitions et raisonner pour s'assurer que ce qu'elles font est justifié
Partager les competences, et non accumuler des clones

Ne pas tomber amoureux de ses hypotheses

Chérir les exceptions