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DE LYON

Anesthésie en chirurgie Non Cardiaque en cardiopathie congénitale Considérations cliniques

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Préambule

Progrés majeur (diagnostic, imagerie, interventionnel, anesthésie réanimation)

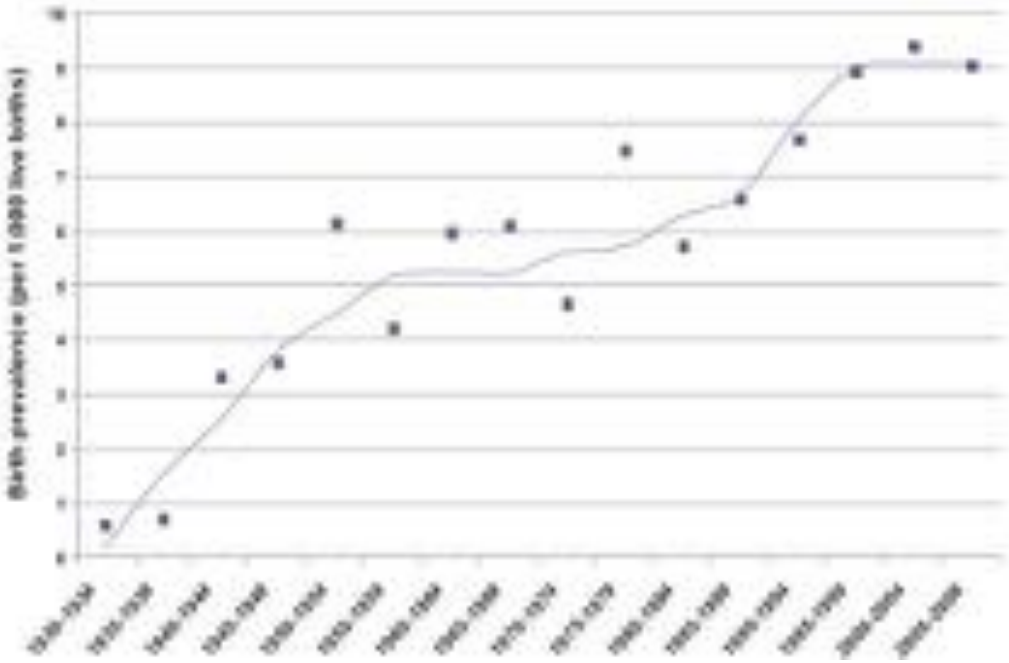
Epidémiologie en changement

Compléxité de plus en plus grande > essor d’une spécialité

La vie n’est pas un long fleuve tranquille

Birth Prevalence of Congenital Heart Disease Worldwide

A Systematic Review and Meta-Analysis



van der Linde *et al.*

JACC Vol. 58, No. 21, 2011

Birth Prevalence of Congenital Heart Disease Worldwide

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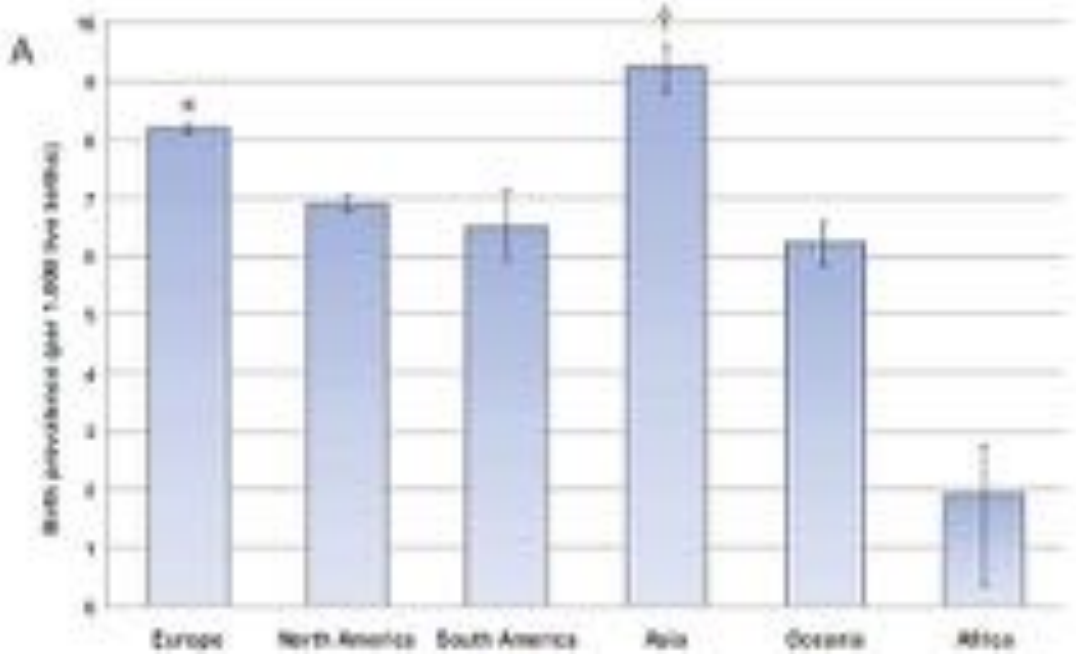
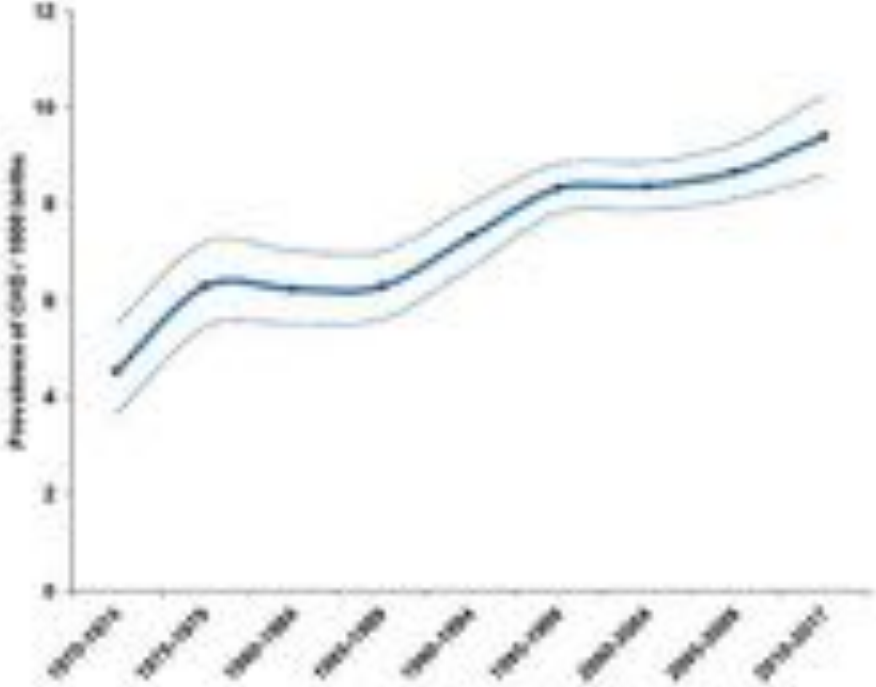


van der Linde *et al.*

JACC Vol. 58, No. 21, 2011

Early Childhood

Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies



Yingjuan Liu,

International Journal of Epidemiology, 2019, 455–463

Global birth prevalence of congenital heart defects 1970–2017: updated systematic review and meta-analysis of 260 studies

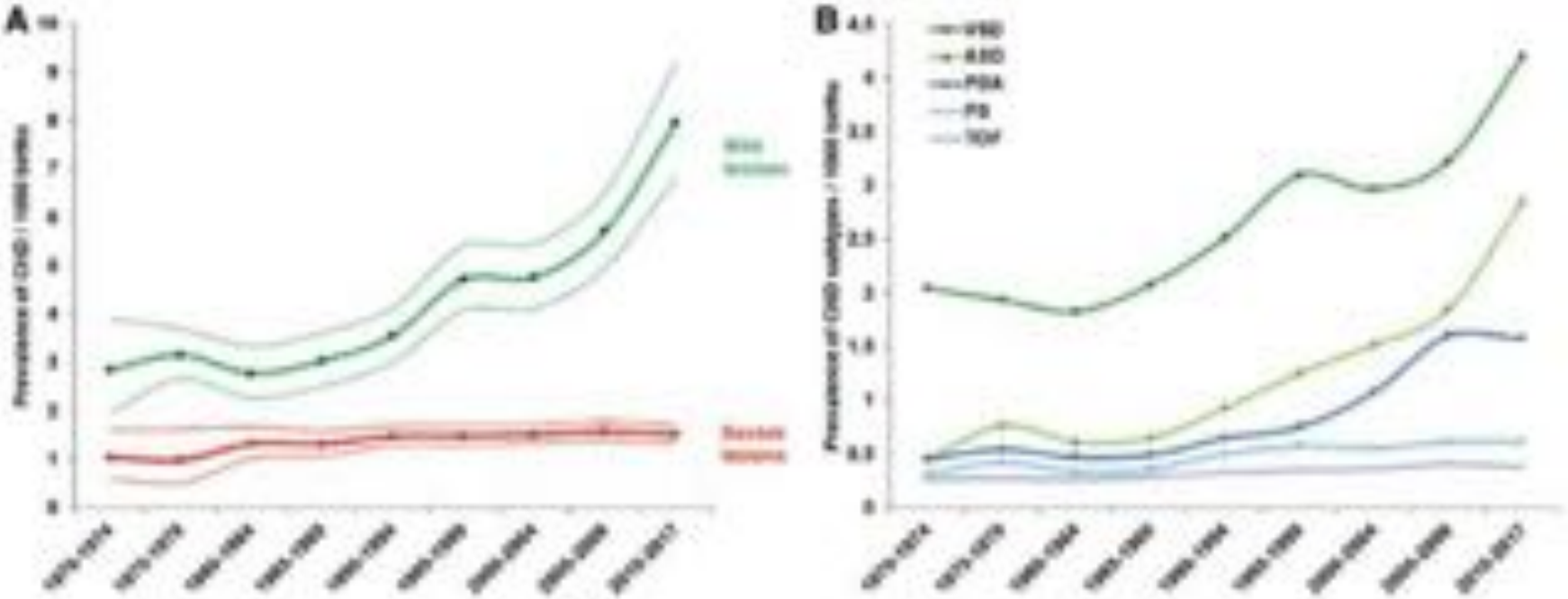
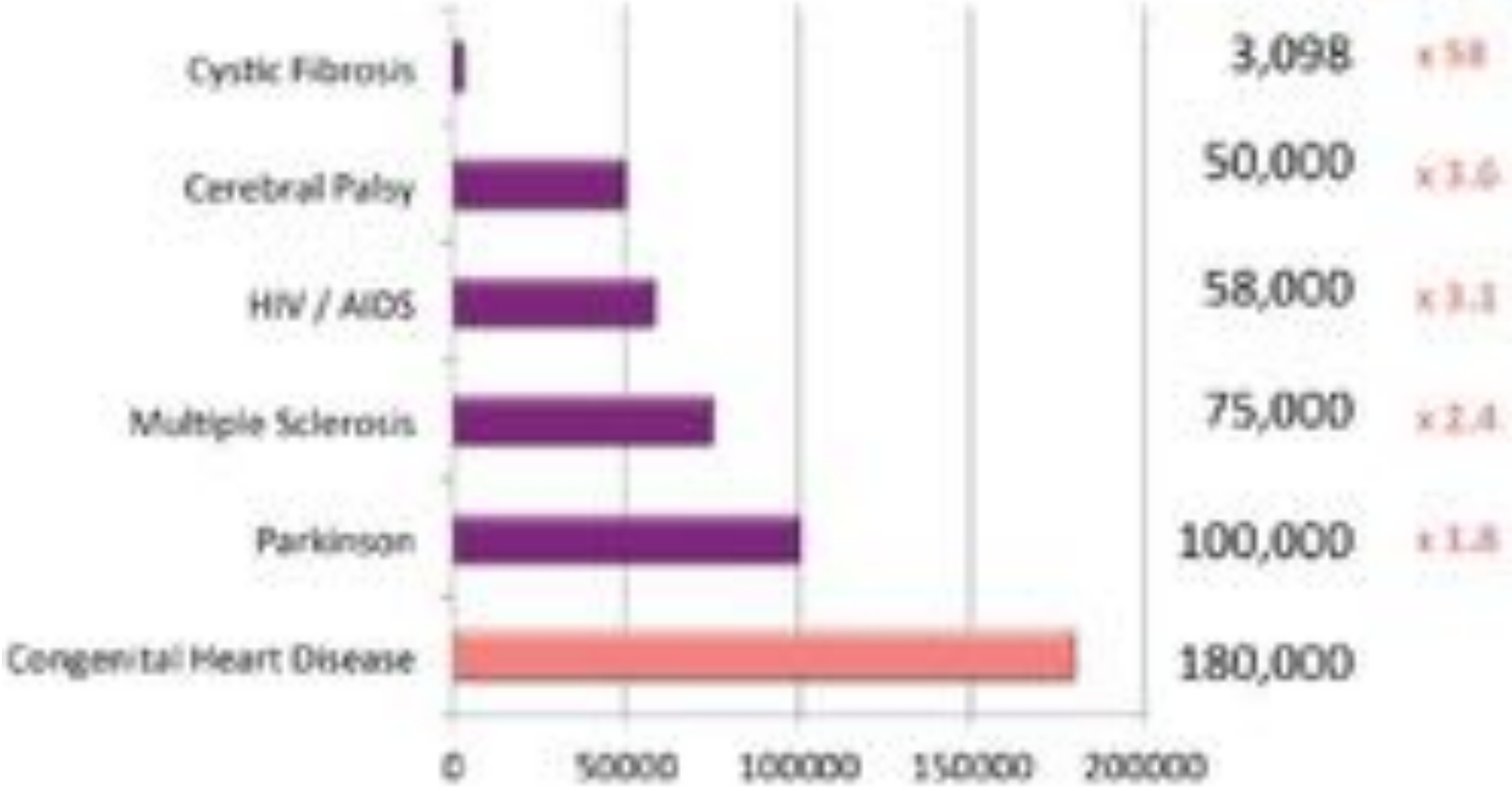


Figure 4. The birth prevalence of CHD subtypes and their changes over time. (A) The prevalence of mild and severe CHD lesions during 1970–2017. Thick lines are the estimated prevalence of CHD lesions, thin lines represent the 95% CI. (B) The birth prevalence of the five most frequent CHD subtypes during 1970–2017. PS, pulmonary stenosis; TOF, tetralogy of fallot.

Yingjuan Liu,

International Journal of Epidemiology, 2019, 455–463



Sources: Cerebral Palsy Canada, Cystic Fibrosis Association, Public Health Agency of Canada; Canadian Congenital Heart Alliance

Genetic syndromes and extracardiac malformations in CHD

-rétrospective

-monocentrique

-période de 10 ans

-2036 foetus atteints de CC

-Sd Génétique 9,8%

-Malfo extracardiaque majeure 11,7 %

-Grèvent le pronositc++++

Table 1 – Distribution of extracardiac anomalies in congenital heart disease

	Total	Overall ECAs	Isolated CAs	ECAs	ECAs without CA	ECAs with CA	Isolated CHD
Heterotaxy, including isomerism and mirror imagery	69	22 (31.9)	1	22	21	1	38 (55.1)
Anomalies of the venous return	27	5 (18.5)	2	3	3	0	20 (74.1)
Anomalies of the atria and interatrial communications	5	1 (20)	1	1	2	1	3 (60)
Anomalies of the atrioventricular junctions and valves	156	60 (38.5)	40	30	20	10	89 (57)
Complex anomalies of atrioventricular connections	52	2 (3.8)	1	1	1	0	46 (88.5)
Functionally univentricular hearts	467	39 (8.4)	18	25	21	4	415 (88.9)
Ventricular septal defects	35	22 (63)	14	12	8	6	20 (57.1)
Anomalies of the ventricular outflow tracts	829	126 (15.4)	82	69	44	25	650 (80.3)
Anomalies of the extrapericardial arterial trunks	243	43 (25.5)	36	40	34	14	172 (70.8)
Congenital anomalies of the coronary arteries	2	0 (0)	0	0	0	0	2 (100)
Cardiomyopathy	31	12 (38.7)	3	10	9	1	17 (54.8)
Atrioventricular block	39	1 (2.6)	0	1	1	0	38 (97.4)
Arrhythmia	31	1 (3.2)	0	1	1	0	30 (96.8)
Tumor	45	25 (55.6)	2	24	23	1	18 (40)
Ventricular diverticulum or aneurysm	5	0 (0)	0	0	0	0	5 (100)
Total	2036	378 (18.6)	200 (9.8)	239 (11.7)	178 (8.7)	61 (3)	1572 (77.2)

Data are expressed as number or number (%). CAs: cytogenetic anomaly; CHD: congenital heart disease; ECAs: extracardiac anomaly (CAs excluded); ECMs: extracardiac malformation.

M. Bensemlali et al.

Archives of Cardiovascular Disease (2016) **109**, 330–336

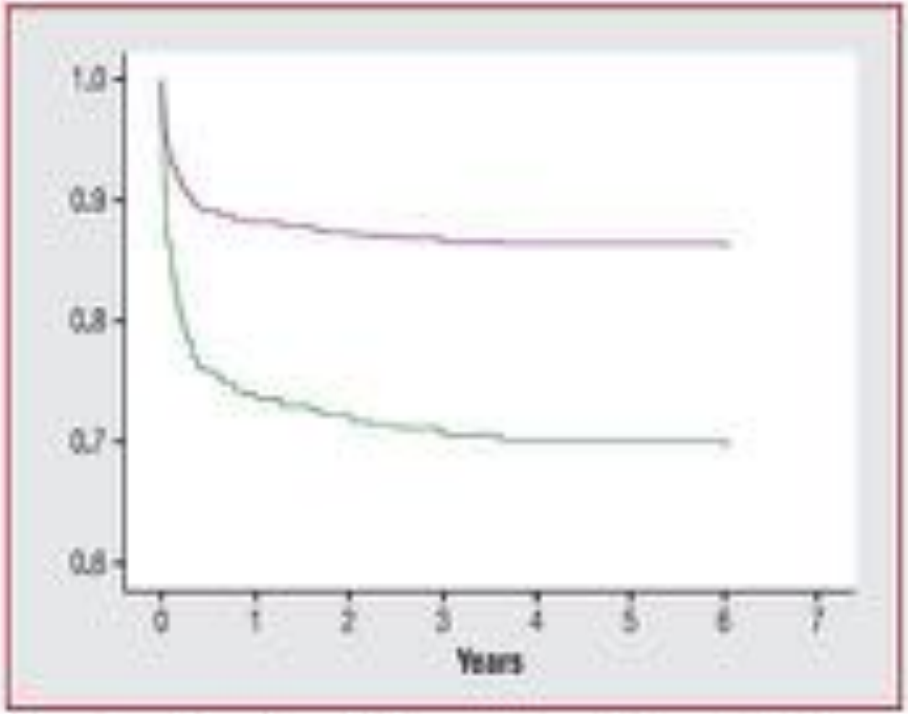


Figure 2. Actuarial survival curve of associated and isolated congenital heart disease (CHD). Purple line: survival of isolated CHD; Green line: survival of associated CHD with a major extracardiac anomaly.

Effect of Concomitant Birth Defects and Genetic Anomalies on Infant Mortality in Tetralogy of Fallot

-étude monocentrique rétrospective base de données Caroline du Nord 2003 2012

-496 T4F / Evaluation taux mortalité à 1 an

-Association entre Anomalies extra cardiaques et TOF

-16% si 1 anomalie
-19% si 2 anomalies
-36% si > 3 anomalies

- Fonction type Atteintes extra cardiaques
-Yeux OR 2,83 (95% CI 1,08-7,32)
-Digestive OR 4,43 (95% CI 1,57-12,45)

Eric G. Jernigan

BIRTH DEFECTS RESEARCH 00:00–00 (2017)

Atteintes extra cardiaque

Timing de prise en charge

Collision avec cure CC

Autres atteintes (IOT difficile,..)

Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery

TABLE 1 ACS NSQIP Classification of CHD Based on Residual Lesion Burden and Functional Status

Classification	Definition and Criteria
Minor CHD	• Cardiac condition with or without medication and maintenance (e.g., atrial septal defect, small-to-moderate ventricular septal defect with no symptoms) • Repair of congenital heart defect with normal cardiovascular function and no medication
Major CHD	• Repair of congenital heart defect with residual hemodynamic abnormality with or without medications (e.g., tetralogy of Fallot with wide open pulmonary insufficiency, hypoplastic left heart syndrome including stage 1 repair)
Severe CHD	• Uncorrected cyanotic heart disease • Patients with any documented pulmonary hypertension • Patients with ventricular dysfunction requiring medications • Listed for heart transplant

ACS NSQIP = American College of Surgeons National Surgical Quality Improvement Program; CHD = congenital heart disease.

Faraoni *et al.*

Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery

TABLE 1 Propensity Matching for Controls and CHD Patients

Matching Criteria	Before Matching			After Matching		
	CHD (n = [illegible])	Controls (n = [illegible])	p Value	CHD (n = [illegible])	Controls (n = [illegible])	p Value
Male	[illegible] (56)	[illegible] (57)	[illegible]	[illegible] (56)	[illegible] (56)	1.00
Age groups			[illegible]			1.00
<6 months	[illegible] (37)	[illegible] (13)		[illegible] (36)	[illegible] (36)	
6–12 months	[illegible] (11)	[illegible] (8)		[illegible] (12)	[illegible] (12)	
1–6 yrs	[illegible] (26)	[illegible] (27)		[illegible] (26)	[illegible] (26)	
6–12 yrs	[illegible] (14)	[illegible] (26)		[illegible] (14)	[illegible] (14)	
>12 yrs	[illegible] (12)	[illegible] (27)		[illegible] (12)	[illegible] (12)	
ASA class 3 or 4	[illegible] (72)	[illegible] (79)	[illegible]	[illegible] (72)	[illegible] (72)	1.00
Elective surgery	[illegible] (80)	[illegible] (73)	[illegible]	[illegible] (80)	[illegible] (80)	1.00
Operative complexity	[illegible] (10–23)	[illegible] (9–16)	[illegible]	[illegible] (9–23)	[illegible] (9–23)	1.00

Values are n (%) or median (interquartile range). Operative complexity was defined for each procedure using the relative value unit based on current procedural terminology codes. p values in **bold** are statistically significant imbalance between the 2 groups, chi-square test.

ASA = American Society of Anesthesiologists; CHD = congenital heart disease.

Post-Operative Outcomes in Children With and Without Congenital Heart Disease Undergoing Noncardiac Surgery

Faraoni *et al.*

OPEN

Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

Figure 1. Number of noncardiac procedures per participating center in the United States. The map was created using mapchart.net (https://mapchart.net/usa.html).

Viviane G. Nasr[1]✉

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Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

2015		2016		2017		2018		2019	
Diagnosis	(%)	Diagnosis	(%)	Diagnosis	(%)	Diagnosis	(%)	Diagnosis	(%)
Atrial septal defect	38.99	Atrial septal defect	32.28	Atrial septal defect	32.00	Atrial septal defect	31.77	Atrial septal defect	32.47
Patent ductus arteriosus	15.03	Patent ductus arteriosus	14.86	Patent ductus arteriosus	14.75	Patent ductus arteriosus	13.90	Patent ductus arteriosus	13.99
Ventricular septal defect	5.55	Ventricular septal defect	12.54	Ventricular septal defect	11.94	Ventricular septal defect	11.78	Ventricular septal defect	11.49
Congenital insufficiency of aortic valve	4.95	Congenital insufficiency of aortic valve	4.65	Congenital insufficiency of aortic valve	4.75	Congenital insufficiency of aortic valve	5.02	Congenital insufficiency of aortic valve	4.80
Stenosis of pulmonary artery	3.53	Stenosis of pulmonary artery	3.55	Stenosis of pulmonary artery	3.36	Stenosis of pulmonary artery	3.54	Stenosis of pulmonary artery	3.47
Hypoplastic left heart syndrome	3.24	Tetralogy of fallot	2.33	Hypoplastic left heart syndrome	2.25	Hypoplastic left heart syndrome	2.52	Hypoplastic left heart syndrome	2.52

Table 3. Most common cardiac diagnoses codes in patients with congenital heart disease presenting for a surgical procedure.

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Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

Variable	2015	2016	2017	2018	2019
Number of cases	38,272	40,313	41,454	43,834	45,993
Age (years)	2.8 (0.9, 6.5)	2.9 (0.9, 6.7)	3 (1, 7)	3.2 (1, 7.2)	3.4 (1.1, 7.4)
Neonates (< 30 days)	3246 (8.5%)	3393 (8.4%)	3459 (8.3%)	3492 (8%)	3520 (7.7%)
Infants (30 days to < 1 year)	6958 (18.2%)	7073 (17.6%)	7082 (17.1%)	7377 (16.8%)	7287 (15.8%)
1 to < 5 years	15,437 (40.3%)	16,092 (39.9%)	16,329 (39.4%)	17,094 (39%)	17,608 (38.3%)
5 to < 12 years	8574 (22.4%)	9399 (23.3%)	10,031 (24.2%)	10,837 (24.7%)	12,092 (26.3%)
12 years or older	4055 (10.6%)	4356 (10.8%)	4553 (11%)	5054 (11.5%)	5486 (11.9%)
Gender					
Female	16,179 (42.3%)	17,287 (42.9%)	17,481 (42.2%)	18,626 (42.5%)	19,726 (42.9%)
Male	22,089 (57.7%)	23,017 (57.1%)	23,965 (57.8%)	25,229 (57.5%)	26,263 (57.1%)
Unknown	4 (0.01%)	9 (0.02%)	8 (0.02%)	3 (0.01%)	4 (0.01%)
Birth weight (kg)	2944 (1390, 3210)	2637 (1580, 3210)	2620 (1510, 3201)	2630 (1500, 3203)	2630 (1535, 3210)

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Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

Variable	2015	2016	2017	2018	2019
Comorbidities					
Vent dependent	5894 (16.4%)	5837 (16.5%)	6001 (16.5%)	6198 (16.1%)	6175 (15.8%)
Any complex chronic condition	28,389 (74.2%)	28,963 (71.9%)	30,09 (72.4%)	32,396 (73.9%)	34,232 (74.4%)
Gastrointestinal	11,274 (29.5%)	12,099 (30%)	12,683 (30.6%)	13,600 (31%)	14,525 (31.6%)
Hematologic or immunologic	1896 (5%)	1910 (4.7%)	1958 (4.7%)	2133 (4.9%)	2207 (4.8%)
Malignancy	1552 (4.1%)	1573 (3.9%)	1615 (3.9%)	1731 (4%)	1884 (4.1%)
Metabolic	1887 (4.9%)	1891 (4.7%)	1982 (4.8%)	2118 (4.8%)	2182 (4.7%)
Neurologic and neuromuscular	5807 (15.2%)	5867 (14.6%)	6170 (14.9%)	6798 (15.5%)	7023 (15.3%)
Renal and urologic	3743 (9.8%)	3852 (9.6%)	3980 (9.6%)	4528 (10.3%)	4740 (10.3%)
Respiratory	6522 (17%)	6749 (16.7%)	7088 (17.1%)	7765 (10.3%)	8030 (17.5%)
Premature and neonatal	3237 (8.5%)	3624 (9%)	4042 (9.8%)	4406 (10.1%)	4538 (9.9%)
Technology dependent	13,670 (35.7%)	14,187 (35.2%)	14,749 (35.6%)	16,005 (36.5%)	16,780 (36.5%)

Viviane G. Nasr[1]✉

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Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

Variable	2015	2016	2017	2018	2019
Syndromes					
22q11 deletion syndrome	771 (2%)	763 (1.9%)	789 (1.9%)	842 (1.9%)	937 (2%)
Down syndrome	3889 (10.2%)	4262 (10.6%)	4503 (10.9%)	4596 (10.5%)	4868 (10.6%)
Trisomy 13, 18	204 (0.5%)	213 (0.5%)	243 (0.6%)	258 (0.6%)	291 (0.6%)
Prematurity (weeks)					
< 24	480 (1.3%)	504 (1.3%)	467 (1.1%)	489 (1.1%)	499 (1.1%)
24–36	7877 (20.6%)	7987 (19.8%)	8547 (20.6%)	9328 (21.3%)	9888 (21.5%)
37–38	4140 (10.8%)	4668 (11.6%)	4913 (11.9%)	5174 (11.8%)	5861 (12.7%)

Viviane G. Nasr[1]✉

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Trends in mortality rate in patients with congenital heart disease undergoing noncardiac surgical procedures at children's hospitals

Variable	2013	2014	2015	2016	2017	P-value for trend
Number of total encounters	38,272	40,313	41,454	43,814	45,995	<0.001*
Number of unique patients	28,239	30,102	31,242	32,889	34,744	
Range of number of encounters per patient	1 to 23	1 to 28	1 to 18	1 to 27	1 to 31	
Mortality rate	445/38,272 (1.16%)	524/40,313 (1.30%)	509/41,454 (1.23%)	475/43,814 (1.09%)	486/45,995 (1.06%)	0.02*
Type of patient						
Inpatient	14,449	14,941	15,646	15,742	15,839	0.004*
Mortality rate	445 (3.08%)	523 (3.50%)	509 (3.30%)	475 (3.02%)	486 (3.07%)	
Outpatient (ambulatory)	19,719	21,189	22,312	23,911	25,890	<0.001*
Mortality rate	0 (0%)	0 (0%)	0 (0%)	0 (0%)	2 (0.02%)	
Observation unit	4094	4163	4096	4211	4264	0.67†
Mortality rate	0 (0%)	1 (0.02%)	0 (0%)	2 (0.05%)	1 (0.02%)	
Length of stay						
Inpatients	7 (2, 27)	7 (3,30)	7 (3,31)	8 (3,30)	8 (3,30)	

Table 2. Description of number of encounters, patients, mortality rate and length of stay. Observation means stay strictly less than 24 h.

Viviane G. Nasr[1]✉

Population-Based Study of Congenital Heart Disease and Revisits After Pediatric Tonsillectomy

Type of heart defect	ICD-9 Code	N (%)
Minor congenital heart defects		507 (59%)
Atrial septal defects	745.5	272 (32%)
Ventricle septal defects	745.4	177 (21%)
Patent ductus arteriosus	747.0	75 (9%)
Pulmonary stenosis	747.3, 747.31, 747.32, 747.39	31 (4%)[†]
Major congenital heart defects		97 (11%)
Coarctation of the aorta	747.10	30 (4%)
Transposition of the great artery	745.10	30 (4%)
Tetralogy of Fallot	745.2	28 (3%)
Hypoplastic left heart syndrome	746.7	13 (2%)
Pulmonary atresia	746.01	24 (3%)[*]
Total anomalous pulmonary venous return	747.41	
Tricuspid Atresia	746.1	
Truncus arteriosus	745.0	
Double-outlet ventricle	745.11	
Single ventricle	745.3	
Ebstein's anomaly	746.2	
Aortic interruption/atresia/stenosis	747.11, 747.22	
Unclassified congenital heart defect		302 (35%)

TABLE I.
Classification of Congenital Heart Defects as Major or Minor, Based on ICD-9 Code (N = 858).

Miller et al.

Laryngoscope Investigative Otolaryngology 4: February 2019

Population-Based Study of Congenital Heart Disease and Revisits After Pediatric Tonsillectomy

TABLE V.
Mixed Effects Logistic Regression Model for Occurrence of Readmission or Emergency Department Visit Following Pediatric Tonsillectomy ± Adenoidectomy (N = 240,774).

Characteristics	OR	95% CI	*P*-value
CHD complexity			
None	Ref.		
Minor	1.08	(0.79, 1.48)	.65
Major	1.17	(0.85, 1.61)	.34
Comorbidities			
Obstructive sleep apnea	1.00	(0.96, 1.04)	.88
Chromosomal anomaly	1.41	(1.21, 1.63)	<.001
Congenital airway anomaly	1.32	(1.03, 1.69)	.03
Neuromuscular impairment	1.40	(1.18, 1.65)	<.001

Miller et al.

Laryngoscope Investigative Otolaryngology 4: February 2019

Laparoscopy Is Safe in Infants and Neonates with Congenital Heart Disease: A National Study of 3684 Patients

Materials and Methods: We identified children <1 year of age with CHD who underwent abdominal surgery in the 2012–2013 American College of Surgeons National Surgical Quality Improvement Project Pediatric database. Patients were stratified by surgical approach: open surgery (OS) versus LS. We then compared postoperative complications, postoperative length of stay, and 30-day mortality by using multivariable regression methods.

Results: In total, 3684 patients met study criteria: 2502 underwent OS while 1182 underwent LS. Infants who underwent LS were older (98 days versus 36 days), larger by weight (4.2 kg versus 3.2 kg), and more likely to require nutritional support preoperatively (74.7% versus 60.5%) (all $P < .001$). After multivariable adjustment, LS was associated with lower overall complication rate (odds ratio [OR] 0.42, 95% confidence interval [CI] 0.34–0.52, $P < .001$) and shorter postoperative length of stay (effect size −1.8 days, 95% CI −1.8–1.2, $P < .001$). LS and OS demonstrated similar 30-day mortality (OR 0.71, 95% CI 0.38–1.32, $P = .28$).

Variables	*OR/effect size*	*Lower 95% CI*	*Upper 95% CI*	P
Complications				
Overall	0.42	0.34	0.52	<.001
Pulmonary	0.56	0.4	0.78	<.001
Bleeding	0.35	0.26	0.46	<.001
Wound	0.6	0.39	0.93	.02
Postoperative length of stay (days)	−1.8	−1.8	−1.2	<.001
30-Day readmission	1.62	1.21	2.17	<.001
30-Day mortality	0.71	0.38	1.32	.28

KIM ET AL.

JOURNAL OF LAPAROENDOSCOPIC & ADVANCED SURGICAL TECHNIQUE
Volume 26, Number 10, 2016

Laparoscopy Is Safe in Infants and Neonates with Congenital Heart Disease: A National Study of 3684 Patients

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Wound	0.6	0.39	0.93	.02
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30-Day mortality	0.71	0.38	1.32	.28

KIM ET AL.

JOURNAL OF LAPAROENDOSCOPIC & ADVANCED SURGICAL TECHNIQUE
Volume 26, Number 10, 2016

Outcomes of laparoscopic and open surgery in children with and without congenital heart disease

Methods—We performed a retrospective cohort study using the 2013–2015 National Surgical Quality Improvement Project-Pediatrics. We matched 45,012 children <18 years old who underwent laparoscopic surgery to 45,012 children who underwent open surgery. We determined

Results—Among children with minor CHD, laparoscopic surgery was associated with lower 30-day mortality (Odds Ratio [OR] 0.34 [95% Confidence Interval 0.15–0.79]), inhospital mortality (OR 0.42 [0.22–0.81]) and 30-day morbidity (OR 0.61 [0.50–0.73]). As CHD severity increased, this benefit of laparoscopic surgery decreased for 30-day morbidity (ptrend=0.01) and in-hospital mortality (ptrend=0.05), but not for 30-day mortality (ptrend=0.27). Length-of-stay was shorter for laparoscopic approaches for children at cost of higher readmissions. On subgroup analysis, laparoscopy was associated with lower odds of postoperative blood transfusion in all children.

Chu et al.

J Pediatr Surg. 2018 October

A Team-Based Approach for Children With Congenital Cardiac Disease Undergoing Antireflux Procedure With Gastrostomy

Parameters	Antireflux procedures on children with CCD (n = 51)
Age, weeks, median (IQR)	3 (2-6.5)
ASA classification, n (%)	
3	26 (51.0)
4	25 (49.0)
Previous noncardiac surgery, n (%)	18 (35.2)
Previous cardiac surgery, n (%)	49 (96.1)
Operative time, min, median (IQR)	68.5 (48-89
Insufflation pressures, mmHg, mean (SD)	9.2 (1.4)
Intraoperative vasopressor use, n (%)	1 (1.9)
Complications, n (%)	
None	48 (94.0)
Reintubation for airway edema	1 (2.0)
Cardiac arrest with return of normal sinus rhythm	1 (2.0)
Conversion to open due to adhesions	1 (2.0)
Immediate extubation postprocedure, n (%)	35 (68.6)
Unplanned readmission, n (%)	5 (9.8)
Still in hospital at 30 days, n (%)	15 (29.4)
Discharge home within 30 days, n (%)	31 (60.8)
Mortality, n (%)	4 (7.8)

CURRENT OPINION

Congenital heart disease in adults (when kids grow up) pediatric geriatric anesthesia

Pablo Motta[a], Ana M. Manrique[b], Sara L. Partington[c], Sana Ullah[d], and Luis M. Zabala[d]

Geriatric Congenital Heart Disease

Burden of Disease and Predictors of Mortality

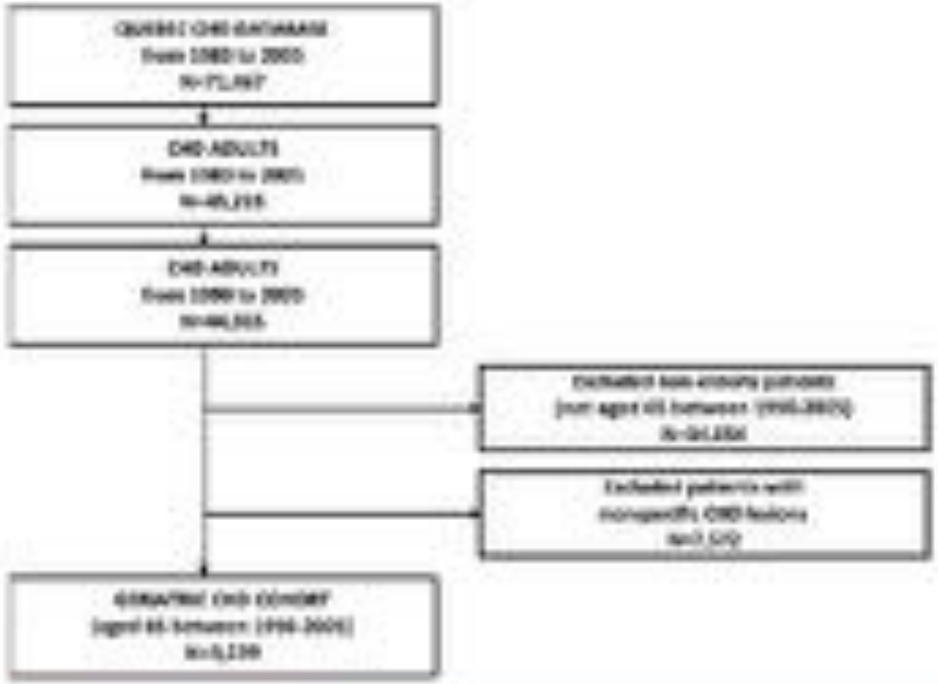


Figure 1 Flow Diagram for Derivation of Study Cohort

The geriatric CHD cohort included patients with a confirmed diagnosis of congenital heart disease (CHD) who were 65 years of age at any point between 1990 and 2005.

Geriatric Congenital Heart Disease

Burden of Disease and Predictors of Mortality

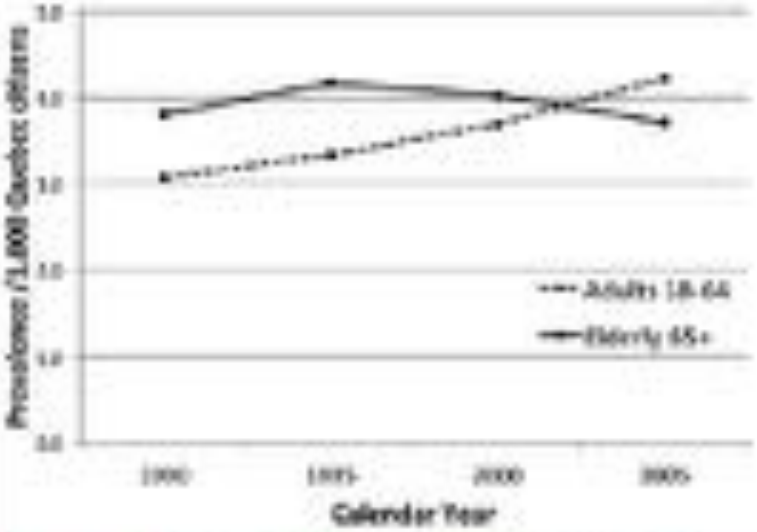


Figure 2 Trends in Prevalence of ADHD in Different Age Groups

In 2005, the prevalence of ADHD was 3.7 per 1,000 in the elderly and 4.2 per 1,000 in nonelderly adults. Using 1996 as a reference, the prevalence remained constant in the elderly (prevalence odds ratio: 0.98; 95% confidence interval [CI]: 0.93 to 1.03), whereas it increased in nonelderly adults (prevalence odds ratio: 1.37; 95% CI: 1.34 to 1.40).

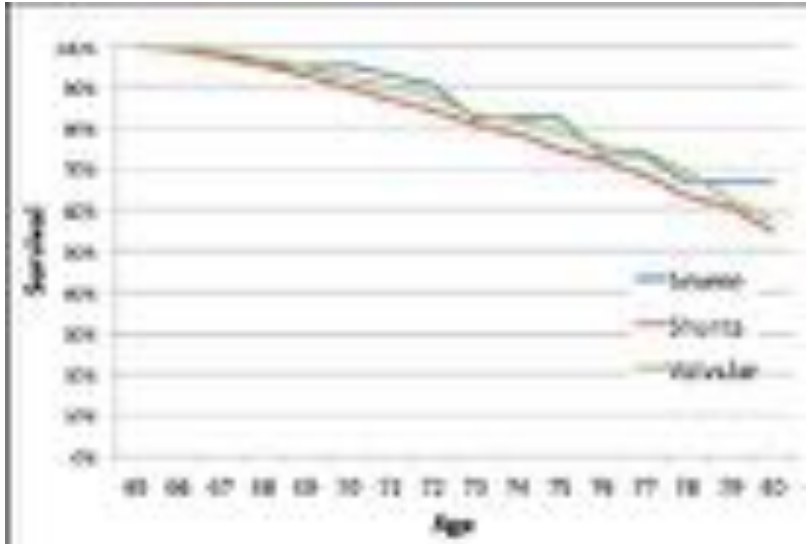
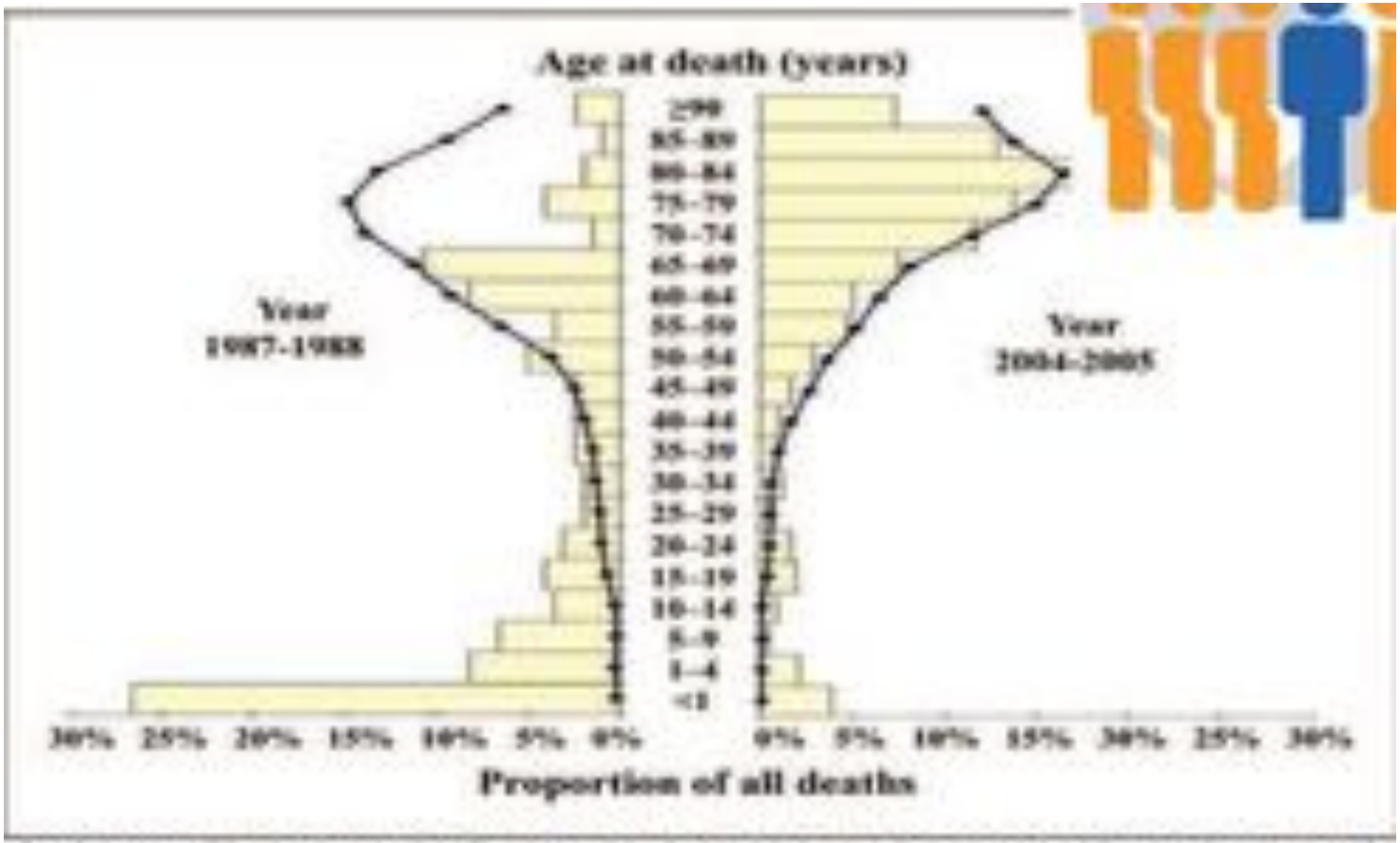


Figure 3 Kaplan-Meier Curves for Unadjusted All-Cause Mortality

The 15-year survival was 90%, 87% for patients with severe lesions, 85% for patients with shunts, and 58% for patients with valvular lesions (log-rank $p<0.001$).

Afilalo *et al.*
Geriatric Congenital Heart Disease



Baehner et Ellerkmann - 2017 - Anesthesia in adults with congenital heart disease.pdf.

Adult Congenital Heart Disease in a Nationwide Population 2000–2014: Epidemiological Trends, Arrhythmia, and Standardized Mortality Ratio

- This nationwide adult congenital heart disease (ACHD) study disclosed an increasing trend of ACHD prevalence and percentage of severe ACHD (217.00 per 100 000 and 11.7% in 2014, respectively).

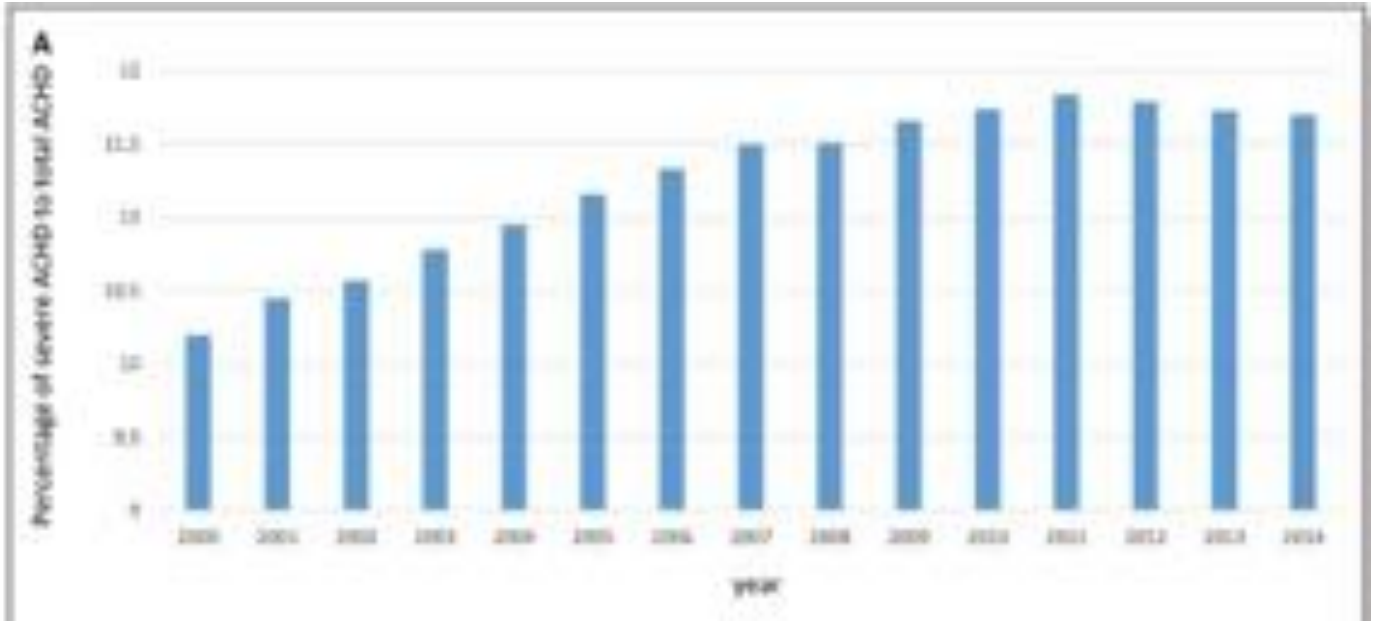
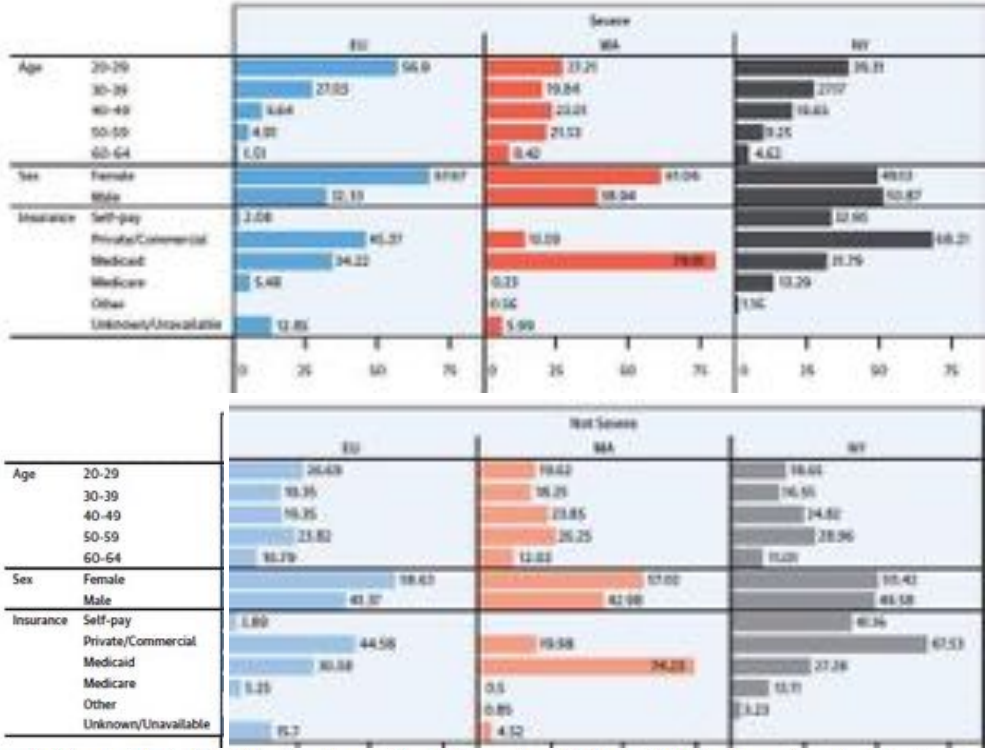
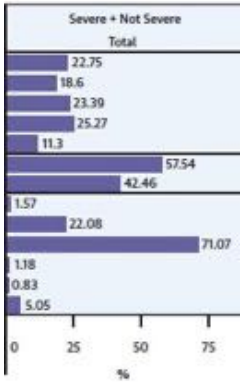


Figure 1. A, Percentage of severe adult congenital heart disease in the whole adult congenital heart disease cohort from 2000 to 2014.

Characteristics of Adults With Congenital Heart Defects in the United States





e, sex, and insurance status of adults with congenital heart defects (CHDs) by location and by CHD severity. *2009 to 2010 for Massachusetts. EU = Emory iversity: Clayton, Cobb, DeKalb, Fulton, and Gwinnett counties, Georgia; MA = Massachusetts: entire state; NY = New York: Allegany, Bronx, Cattaraugus, autauqua, Erie, Genesee, Monroe, Niagara, Orleans, Westchester, and Wyoming counties.

Gurvitz *et al.* JACC VOL. 76, NO. 2, 2020

Population Demographics of Adults With CHD

Adult Congenital Heart Disease in a Nationwide Population 2000–2014: Epidemiological Trends, Arrhythmia, and Standardized Mortality Ratio

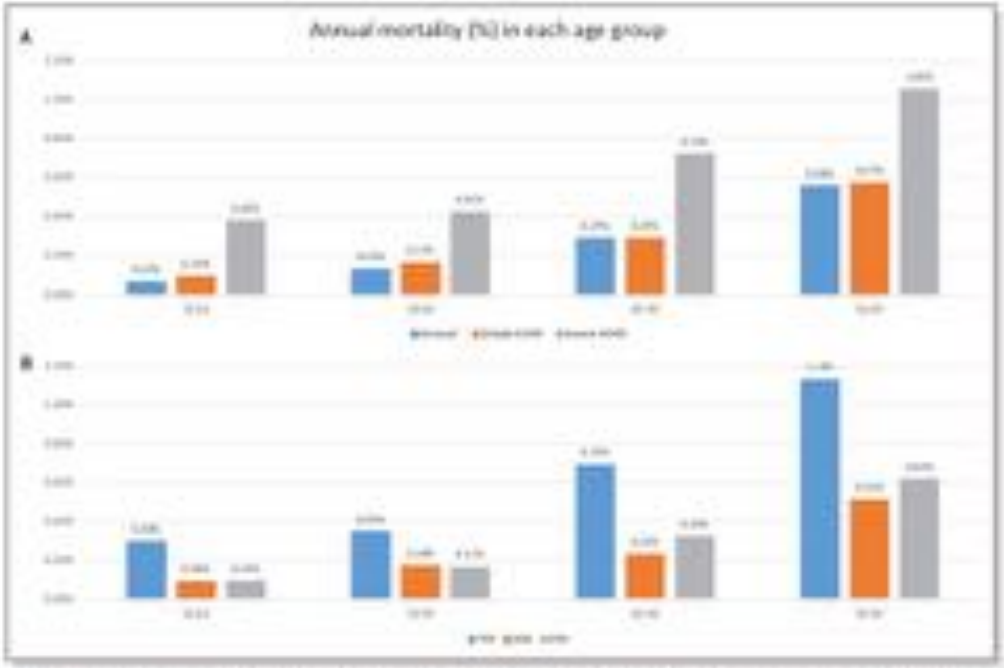


Figure 2. A, Age-group-specific mortality in general population, simple adult congenital heart disease (simple ACHD), and severe adult congenital heart disease (severe ACHD) patients. B, Age-group-specific mortality in adult patients with tetralogy of Fallot (TOF), secundum atrial septal defect (ASD), and ventricular septal defect (VSD).

Adult Congenital Heart Disease in a Nationwide Population 2000–2014: Epidemiological Trends, Arrhythmia, and Standardized Mortality Ratio

Clinical Perspective

What Is New?

- Standardized mortality ratio is higher not only in severe ACHD (3.164), but also in women simple ACHD (1.704), with higher proportions of cardiac, labor, and sudden death as causes of death.

Table 3. Causes of Death in Adult Congenital Heart Disease and General Population Aged 20 to 59 Years of Age During the Study Period 2000–2014

Causes of death	ACHD			General Population
	Severe	Simple	Total	
Cardiac	[illegible]	[illegible]	[illegible]	[illegible]
Malignancy	[illegible]	[illegible]	[illegible]	[illegible]
External causes	[illegible]	[illegible]	[illegible]	[illegible]
CVA	[illegible]	[illegible]	[illegible]	[illegible]
SD	[illegible]	[illegible]	[illegible]	[illegible]*
Labor complication	[illegible]	[illegible]	[illegible]	[illegible]
Out of hospital death	[illegible]	[illegible]	[illegible]	NA
ES CPR	[illegible]	[illegible]	[illegible]	NA
Others	[illegible]	[illegible]	[illegible]	[illegible]

CVA, indicates cerebrovascular accident; ES CPR, cardiopulmonary resuscitation in emergency service; SD, sudden death.
*Data 2007–2014.

Causes of death in a contemporary adult congenital heart disease cohort

CHD type	16y	25y	35y	45y	55y	65y	75y
Simple	1452	1374	1103	848	641	420	199
Moderate	1029	905	554	281	158	76	29
Complex	545	452	226	72	20	5	1

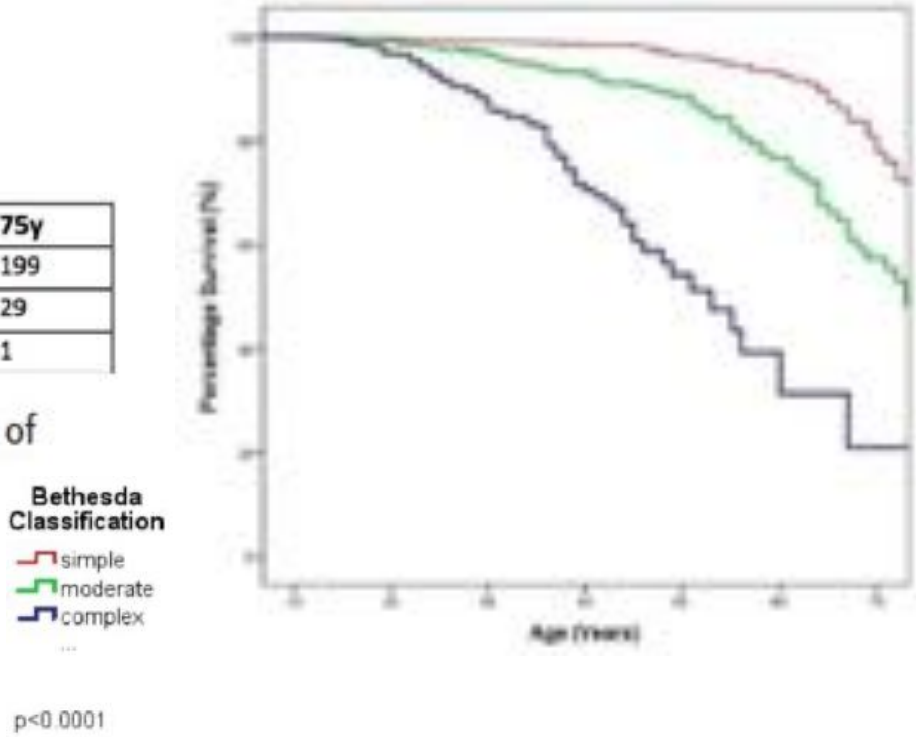


Figure 1 Kaplan-Meier survival curves arranged by severity of underlying adult congenital heart disease (CHD).

Yu C, *et al*. *Heart* 2018;**104**:1678–1682.

Causes of death in a contemporary adult congenital heart disease cohort

Table 2 Cause of death by Bethesda Classification

% Cause of death*	Simple (n=147)	Moderate (n=101)	Complex (n=91)	All patients (n=341)	P values†
Heart failure	14%	15%	25%	17%	0.054
Malignancy	20%	7%	4%	13%	0.0002
Sudden cardiac death	1.4%	13%	22%	10%	<0.0001
Vascular	9%	10%	3%	8%	0.181
Pneumonia	7%	7%	7%	7%	1.000
Ischaemic heart disease	8%	3%	2%	5%	0.121
Endocarditis	7%	2%	6%	5%	0.231
Perioperative	5%	4%	4%	5%	0.853
Acute myocardial infarction	5%	7%	1%	4%	0.140
Sepsis	3%	4%	3%	4%	1.000
Other causes of death	20.6%	28%	26%	26%	

*Percentage of patients who died from stated condition within each 'complexity' category.
†P-values are for χ^2 across the three 'complexity' groups.

Yu C, *et al*. *Heart* 2018;**104**:1678–1682.

Adult Congenital Heart Disease in a Nationwide Population 2000–2014: Epidemiological Trends, Arrhythmia, and Standardized Mortality Ratio

Clinical Perspective

What Is New?

- Freedom from tachyarrhythmia at age 50 years was 0.574 and 0.710 in severe and simple ACHD. Cardiac causes

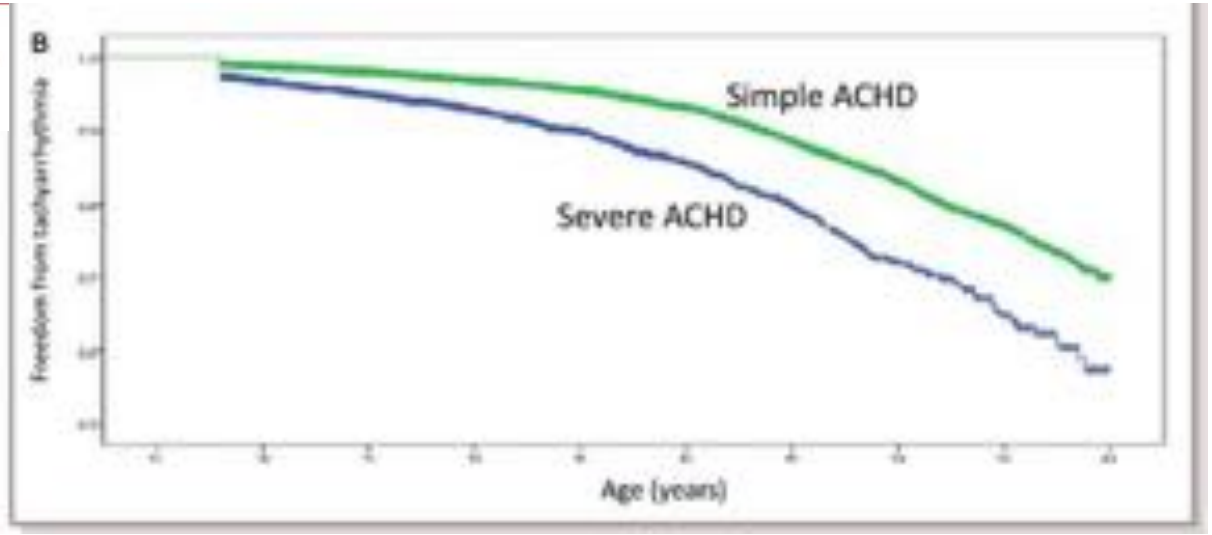


Figure 1. disease col 2014. B, Freedom from tachyarrhythmia in simple adult congenital heart disease (simple ACHD) and severe adult congenital heart disease (severe ACHD) cohorts.

ESC
European Society of Cardiology
European Heart Journal (2021) **42**, 563–645
doi:10.1093/eurheartj/ehaa554

Risk estimates for arrhythmic events and bradycardias in ACHD

Type of CHD	AVRT	SNRT/SVT	AF	Sustained VT	SCD	Bradycardia: SND		Bradycardia: AV block	
						Congenital	Acquired	Congenital	Acquired
Secundum ASD		++	++			(+)	+		(+)
Sinus venosus defect		++	+				+		
AVSD/primum ASD		++	++	(+)		(+)		(+)	++
VSD		+	(+)	+	(++)				+
Ebstein anomaly	+++	++	+	(+)	+++		++		
TOF		++	++	++	++		+		+
TGA									
Atrial switch		+++	+	++	+++		+++		+
Arterial switch		+		(+)	(+)		(+)		
cc-TGA	++	+	+	(+)	+++			+	++
Fontan operation									
Atriopulmonary connection		+++	++		+		++		
Intracardiac atrial baffle		++	+		+		++		
Extracardiac conduit		+	+		+		+		
Fontan-type physiology (univentricular palliated CHD)		++	++		+++				

Empty cells indicate that although not specifically studied, arrhythmic events may occur (for example).

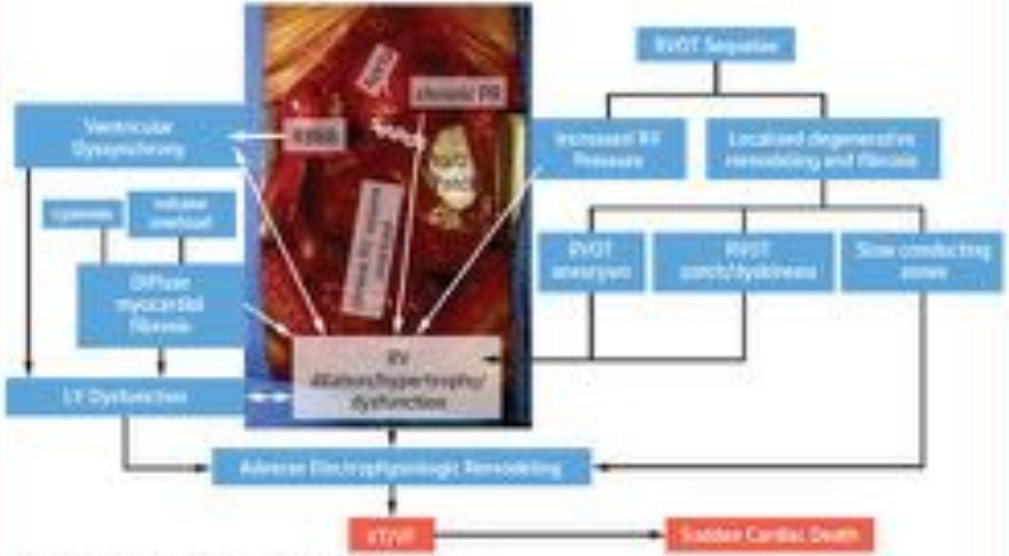
(+) = minimal risk; + = additive risk; ++ = moderate risk; +++ = high risk

Preventing Arrhythmic Death in Patients With Tetralogy of Fallot

JACC Review Topic of the Week

Mitchell I. Cohen, MD,[a] Paul Khairy, MD, PhD,[b] Katja Zeppenfeld, MD, PhD,[c] George F. Van Hare, MD,[d]
Dhanunjaya R. Lakkireddy, MD,[e] John K. Triedman, MD[f]

CENTRAL ILLUSTRATION Midline Sternotomy View of a Patient With Repaired Tetralogy of Fallot



Cohen, M.I. et al. J Am Coll Cardiol. 2020;77(6):769-79.

The potential contributory mechanisms leading to sudden cardiac death are shown. LV = left ventricular; RV = right ventricular; RVOT = right ventricular outflow tract; VF = ventricular fibrillation; VT = ventricular tachycardia.

CLINICAL RESEARCH

Characteristics and outcomes of heart failure-related hospitalization in adults with congenital heart disease

Caractéristiques et résultats des hospitalisations liées à une insuffisance cardiaque chez le patient adulte avec une cardiopathie congénitale

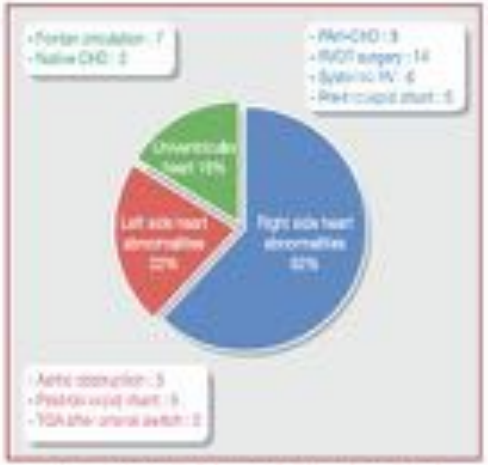


Figure 1. Distribution of congenital heart defects in patients admitted for heart failure. Congenital heart defects were clas-

Objectifs. — Nous avons cherché à caractériser les hospitalisations en rapport avec l'IC du patient ACC et à déterminer les facteurs de risque et le pronostic de l'IC dans cette population.

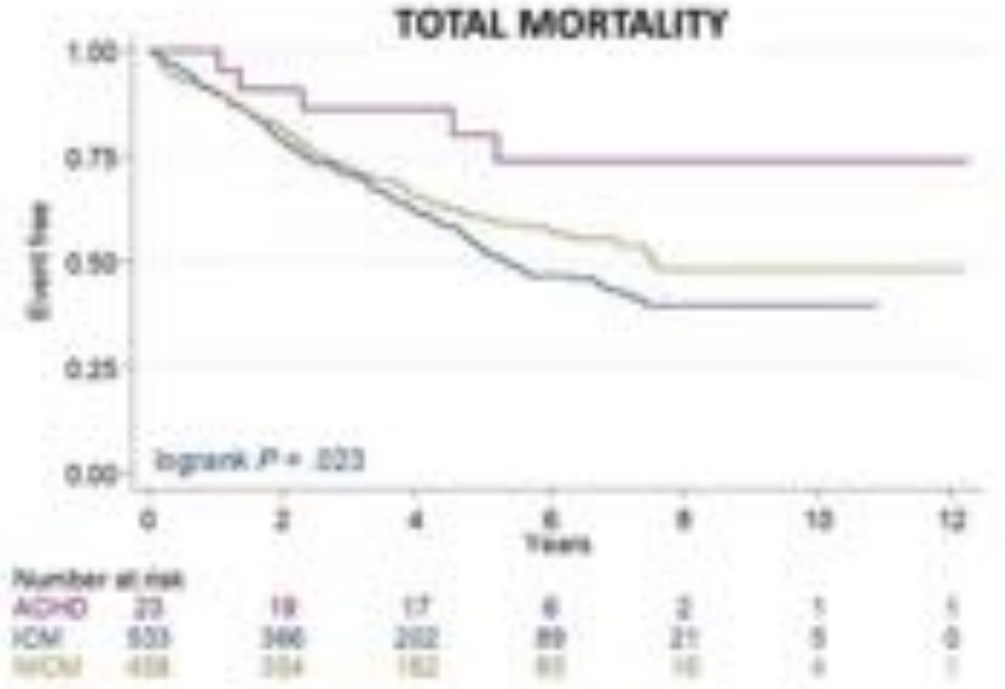
Méthodes. — Au total, 471 patients ACC admis dans notre unité sur une période de 24 mois ont été prospectivement inclus. Les données cliniques et biologiques, ainsi que la prise en charge de l'IC ont été recueillies. La survenue d'événements cardiovasculaires majeurs après IC au cours du suivi a été également relevée.

Résultats. — L'IC était le motif d'hospitalisation principal dans 13 % des cas (76/583 hospitalisations). Les patients IC étaient significativement plus âgés (âge médian 44 ± 14 vs 37 ± 15 ans ; $p = 0,01$), avec des cardiopathies congénitales plus complexes ($p = 0,04$). En analyse multivariée, l'hypertension artérielle pulmonaire (OR 6,2, IC 95 % 3,5–10,7), les antécédents d'insuffisance cardiaque (OR 9,8, IC 95 % 5,7–16,8) et d'arythmie auriculaire (OR 3,6 IC 95 % 2,2–5,9) étaient des facteurs de risque significatifs d'hospitalisation pour IC ($p < 0,001$). La durée moyenne de séjour des patients IC était plus longue (12,2 jours contre 6,9 jours ; $p = 0,01$) et 25 % des patients ont nécessité une prise en charge en soins intensifs. Vingt pourcent (11/55 patients IC) sont décédés, 18 % (10/55) ont été ré-hospitalisés pour IC, et 11 % (6/55) ont eu une transplantation cardiaque sur un suivi médian de 18 mois (IC 95 % 14–20 mois). Le risque d'événements cardiovasculaires était 19 fois plus élevé après l'hospitalisation liée à l'IC.

N.B. Moussa et al.

Archives of Cardiovascular Disease (2017) 110, 283—291

Long-term outcomes of cardiac resynchronization therapy in adult congenital heart disease



LEYVA ET AL.

Pacing Clin Electrophysiol. 2019;42:573–580.

Advanced Heart Failure in Adults With Congenital Heart Disease

TABLE 1 Factors Signaling Clinical Worsening in a Patient With Complex CHD

	Factor	First Author (Ref. #)
Exercise testing	Peak VO_2 is: 1. Reduced in adults with CHD without HF compared with healthy subjects of similar age 2. Varies by anatomic lesion 3. Generally predicts hospitalization or death (more data needed)	Diller et al. [7], Kempny et al. [8]
	• A decline in peak VO_2 during consecutive CPETs in patients with Fontan physiology predicts increased risk of death or HT	Cunningham et al. [9]
	• CPET results should not independently drive decisions to pursue HT evaluation	Menachem et al. [6]
Functional classification	• A strong direct relationship exists between functional class (NYHA) and peak VO_2 • 2.5× and 8.7× increased mortality risk in NYHA functional class II and class III compared with NYHA functional class I, respectively	Brody et al. [10]
	• Patients with CHD overestimate their physical functioning compared with objective exercise test results	Gratz et al. [11]
	• Atypical manifestations of HF may lead to a delay in evaluation for advanced HF therapies and subsequent worse outcomes with HT	Alshawabkeh et al. [12]
Hospitalization	• 5× higher mortality risk in patients when admitted for HF • 1- and 3-yr mortality after first HF admission were 24% and 35%, respectively	Zomer et al. [13]

Menachem *et al.*

JACC: HEART FAILURE VOL. 8, NO. 2, 2020
FEBRUARY 2020:87–99

Advanced Heart Failure in Adults With Congenital Heart Disease

Electrophysiology		
Atrial arrhythmias	• >50% of patients with complex CHD will develop atrial arrhythmias by 65 yrs of age • 2 to 3× increased risk of HF or occurrence of cardiac intervention	Bouchardy et al. (14), Lindsay and Moore (15)
	• Atrial tachycardia is an independent predictor of death and death/hospitalization in patients with Fontan	Giannakoulas et al. (16)
Sudden cardiac death	• Sudden cardiac death is the second most common cause of death in adults with CHD	Lindsay and Moore (15)
Ventricular arrhythmias	• QRS duration ≥180 ms is a predictor of malignant ventricular arrhythmia in patients with tetralogy of Fallot	Gatzoulis et al. (17)
	• ICDs are indicated in patients who survive cardiac arrest secondary to ventricular fibrillation or tachycardia in the absence of a reversible cause	Avila et al. (18) Khairy et al. (19)

Menachem *et al.*

JACC: HEART FAILURE VOL. 8, NO. 2, 2020
FEBRUARY 2020:87-99

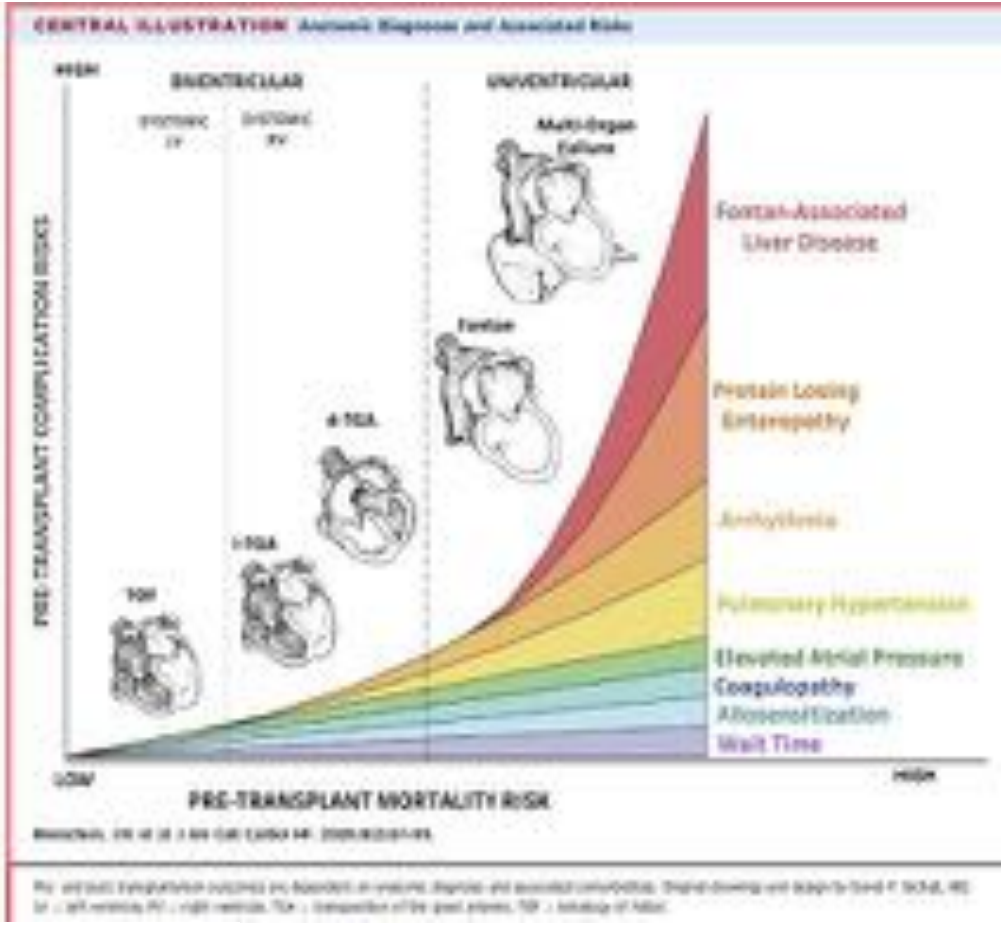
Advanced Heart Failure in Adults With Congenital Heart Disease

Biomarkers		
Anemia	• Patients with anemia are associated with a 3× higher mortality risk than nonanemic patients	Dimopoulos et al. (20)
Natriuretic peptides	• Levels vary by CHD diagnosis, repair, and sequelae. Levels are related to exercise capacity • Risk of death and HF is low in those with normal and consecutively stable values	Eindhoven et al. (21), Baggen et al. (22)
hsCRP	• hsCRP >2.98 mg/L (highest quartile) showed increased risk of all-cause mortality or nonelective cardiovascular hospitalization	Opotowsky and Webb (23)
Liver disease	• MELD-XI is directly related to the incidence of a composite endpoint of sudden death, death from HF, or cardiac transplantation in patients with Fontan	Assenza et al. (24)
Red cell distribution	• Elevated RDW is an independent predictor of all-cause mortality or nonelective cardiovascular hospitalization	Alshawabkeh et al. (25)
Renal function	• Moderate or severe GFR reduction led to 5× higher mortality than those with normal GFR	Dimopoulos et al. (26)
Serum sodium	• Sodium concentration <136 mmol/L demonstrated a 3× increased mortality risk	Dimopoulos et al. (27)

CHD = congenital heart disease; CPET = cardiopulmonary exercise test; GFR = glomerular filtration rate; HF = heart failure; hsCRP = high-sensitivity C-reactive protein; HT = heart transplantation; ICD = implantable cardioverter-defibrillator; MELD-XI = model of end-stage liver disease excluding international normalized ratio; NYHA = New York Heart Association; RDW = red cell distribution width; VO_2 = oxygen consumption.

Menachem *et al.*

JACC: HEART FAILURE VOL. 8, NO. 2, 2020
FEBRUARY 2020:87–99



Menachem *et al.*

Advanced Heart Failure in Adults With Congenital Heart Disease

JACC: HEART FAILURE VOL. 8, NO. 2, 2020
FEBRUARY 2020:87–99

Genetics of congenital heart diseases
Le Hasard et la nécessité
Old textbooks and clinical genetics

REVIEWS

Maternal and environmental risk factors for neonatal AKI and its long-term consequences

Is low birth weight an antecedent of CKD in later life? A systematic review of observational studies.

J Am Soc Nephrol. 2008 Jan; 19(1): 151–157.
doi: 10.1681/ASN.2007020252 10.1681/ASN.2007020252

Low Birth Weight Increases Risk for End-Stage Renal Disease

Acquired Comorbidities in Adults with Congenital Heart Disease: An Analysis of the German National Register for Congenital Heart Defects

ACHD that are representative on a population level are not available. Methods: The German National Register for Congenital Heart Defects was screened for ACHD. Underlying congenital heart disease (CHD), patient demographics, previous interventional/surgical interventions, and comorbidities were retrieved. Patients <40 years of age were compared to those ≥40 years. Results: A total of 4673 patients (mean age 33.6 ± 10.7 years, female 47.7%) was included. At least one comorbidity was present in 2882 patients (61.7%) altogether, and in 56.8% of patients below vs. 77.7% of patients over 40 years of age ($p < 0.001$). Number of comorbidities was higher in patients ≥40 years (2.1 ± 2.1) than in patients <40 years (1.2 ± 1.5, $p < 0.001$). On multivariable regression analysis, age and CHD complexity were significantly associated with the presence and number of comorbidities.

Susanne J. Maurer

J. Clin. Med. **2021**, *10*, 314.

Acquired Comorbidities in Adults with Congenital Heart Disease: An Analysis of the German National Register for Congenital Heart Defects

-Comorbidités Fqtes < 40 ans :

Endocriniens (24,3%) : Obésité, Thyroïde
CardioCirculatoires (21,5%) : HTA
Atteinte SNC (10,4%)

-Comorbidités Fqtes > 40 ans :

CardioCirculatoires (50,5%) : HTA, AVC,...
Endocriniens (50,2%) : Obésité, thyroïde, dyslipidémie
Atteinte SNC (15,2%):

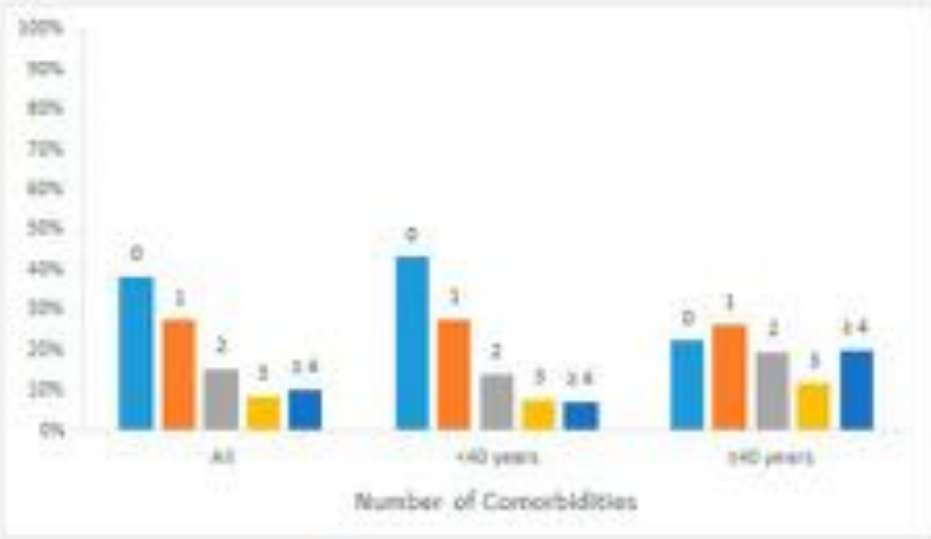


Figure 1. Number of comorbidities according to age group.

Susanne J. Maurer

J. Clin. Med. **2021**, *10*, 314.

Acquired Comorbidities in Adults with Congenital Heart Disease: An Analysis of the German National Register for Congenital Heart Defects

Table 4. Association between age, complexity of CHD, and gender with number of comorbidities.

	Incidence Rate Ratio	95% Confidence Interval	*p*
Age (in years)	1.030	1.026–1.033	<0.001
Male	0.983	0.909–1.062	0.658
Moderate CHD	1.245	1.132–1.370	<0.001
Severe CHD	1.351	1.210–1.509	<0.001
Biventricular CHD	0.793	0.677–0.928	0.004

CHD, congenital heart disease.

Susanne J. Maurer

J. Clin. Med. **2021**, *10*, 314.

Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease

A Scientific Statement From the American Heart Association

Figure 1. Noncardiac complications in adults with congenital heart disease (CHD).

The medical history of adults with complex congenital heart disease affects their social development and professional activity

Figure 1. Distribution of congenital heart disease in the study population (n = 135). AS: aortic stenosis; ASD: atrial septal defect; AVSD: atrioventricular septal defect; CoA: aortic coarctation; Other: other defects (n = 7); PDA: patent ductus arteriosus; PS: pulmonary stenosis; TGA: transposition of the great arteries and congenitally corrected transposition of the great arteries; TOF: tetralogy of Fallot; VSD: ventricular septal defect; UH: univentricular heart.

Méthodes. — Nous avons inclus consécutivement à partir d'une cohorte observationnelle monocentrique, 135 ACC (40 ± 12 ans) qui ont répondu à un questionnaire évaluant les aspects psychosociaux et de la vie quotidienne. La complexité de la cardiopathie était classée selon la conférence de Bethesda.

Résultats. — La complexité de la cardiopathie était simple pour 61 patients (45,2 %), modérée pour 50 (37,0 %) et complexe pour 24 (17,8 %). Une chirurgie cardiaque a été effectuée pour 86,5 % des patients ayant une cardiopathie complexe ou modérée. Les complications (insuffisance cardiaque [n = 15/24], arythmie [n = 14/24], hypertension pulmonaire [n = 6/24]) ont été principalement observées dans le groupe complexe ($p = 0,002$). L'activité physique était moins importante dans le groupe complexe ($p = 0,03$, et aucune activité pour 58,8 % mais 33 % avaient été contre indiqués). Les patients des groupes complexe et modéré ont tendance à avoir un niveau d'étude inférieur aux patients du groupe simple (respectivement 31,2 %, 33,3 % versus 45,7 % ont le Baccalauréat ; $p = 0,47$). L'obtention du Baccalauréat était plus faible pour les patients ayant au moins une complication ($p = 0,038$) et plus d'une chirurgie cardiaque ($p = 0,03$). Dans le groupe complexe, 56,3 % des patients étaient au chômage ($p = 0,048$).

C. Karsenty et al.

Archives of Cardiovascular Disease (2015) 108, 589—597

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of Cardiology

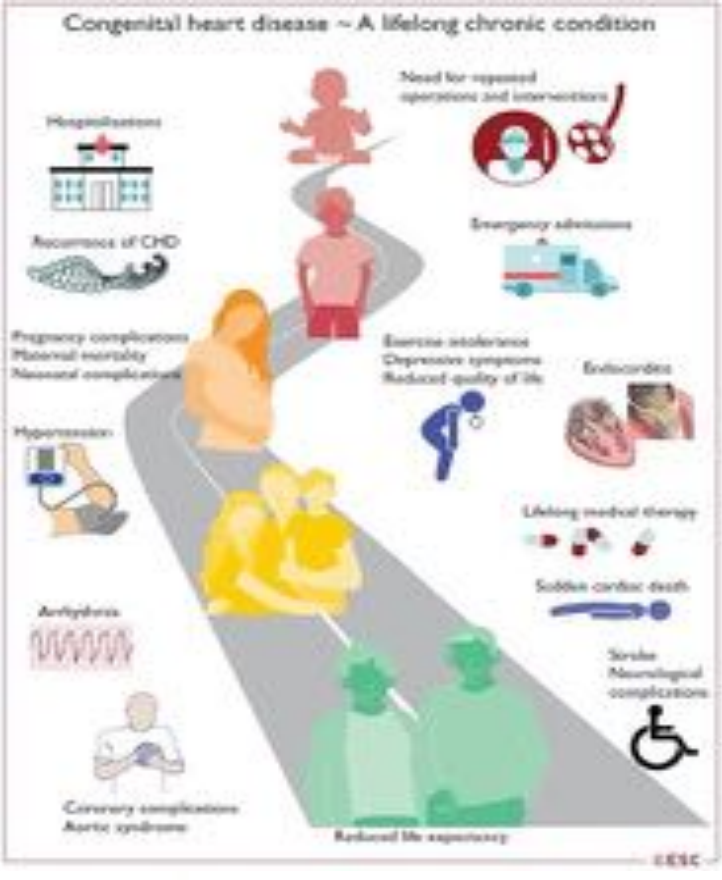

Figure 1 Central illustration. Congenital heart disease is a lifelong chronic condition. CHD = congenital heart disease; SCD = sudden cardiac death.

European Heart Journal (2020)

Decentralization of Surgical Care for ACHD

Bryan G. Maxwell PLOS ONE 2014

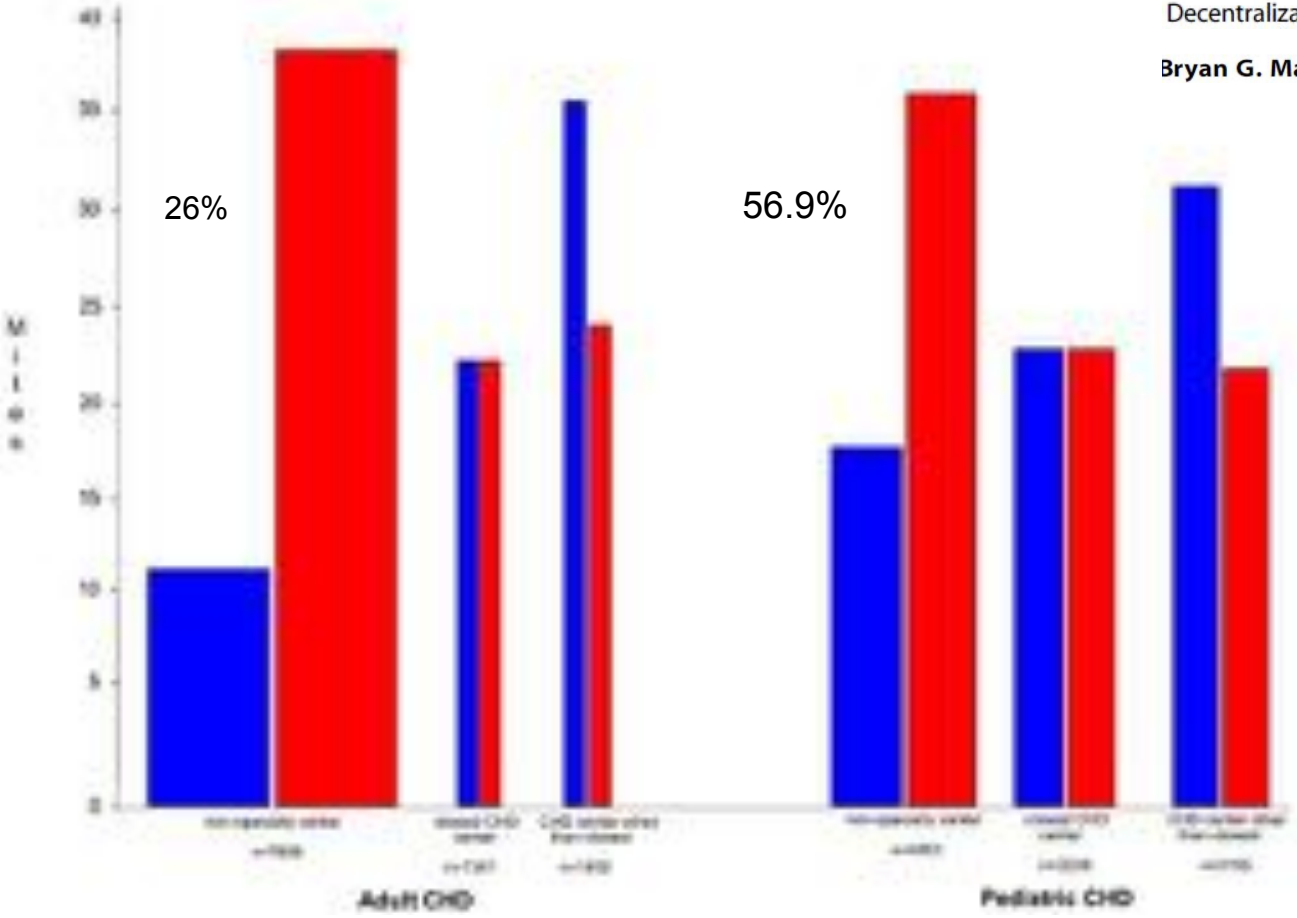

Figure 1. Distance from care. Values are the mean distance to the facility where surgery occurred (blue bars) and to the nearest CHD center of expertise (red bars), by patient subgroup. Width of each pair of columns is proportional to the relative size of the corresponding subgroup.
doi:10.1371/journal.pone.0106730.g001

Transition in Patients with Congenital Heart Disease in Germany: Results of a Nationwide Patient Survey

Helm et al. Frontiers in Pediatrics

-Enquête National Allemande

-1828 ACHD

-Age Moyen 31.7+/-11.7 ans

-66.1% ne savaient pas Si suivi par un spécialiste ACHD

-Enjeu Transition+++

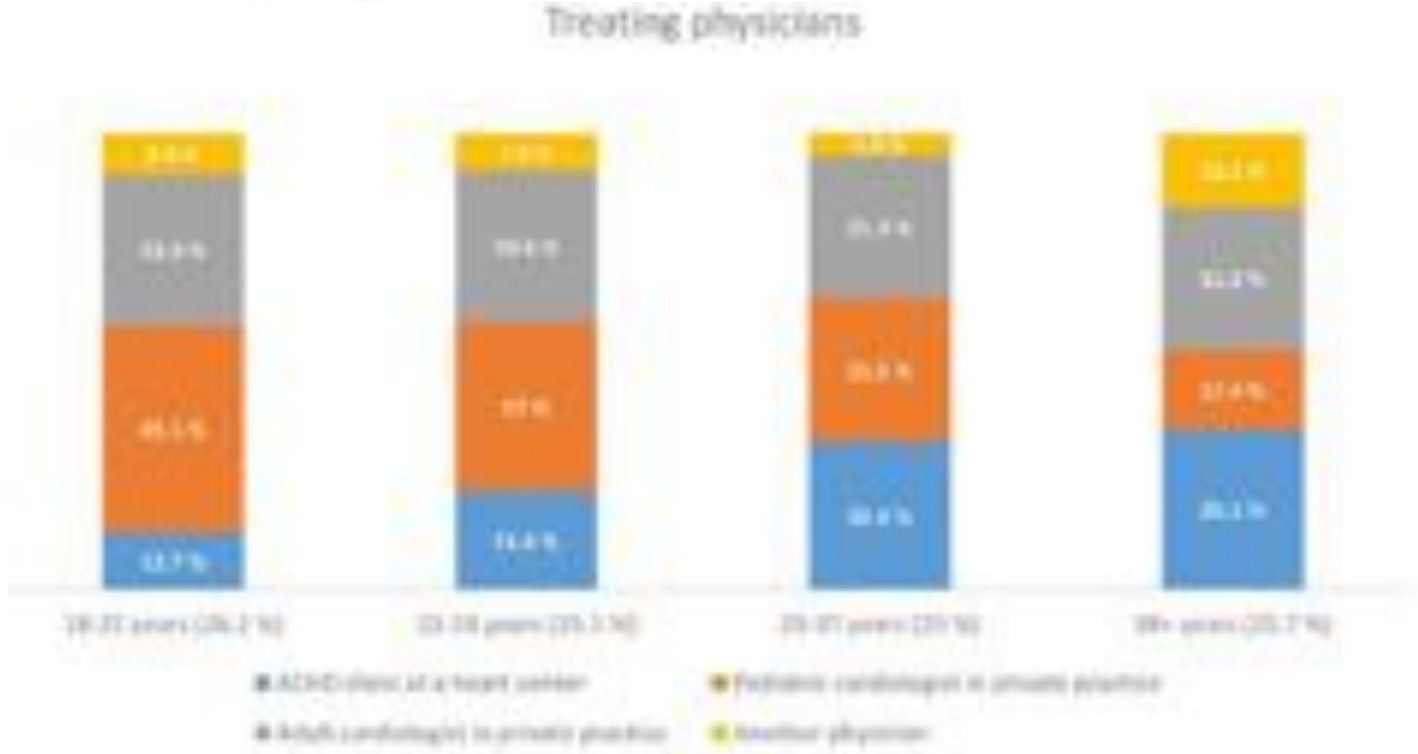

FIGURE 1 | Treating physicians.

Do we have the ACHD physician resources we need to care for the burgeoning ACHD population?

FIGURE 1 States with and without ACHD programs

EZZEDDINE ET AL.

Congenital Heart Disease

Do we have the ACHD physician resources we need to care for the burgeoning ACHD population?

FIGURE 2 Number of patients served divided by number of ACHD physicians (state-by-state)

Conclusions: The ACHD population continues to grow, and the looming national physician shortage is likely to greatly affect the ability to meet the complex needs of this growing population. In order to bring the ACHD patient: physician ratio to 1000:1, a minimum of 170 additional ACHD board-certified physicians are needed now.

EZZEDDINE ET AL.

Congenital Heart Disease

Original article

Characteristics of patients with adult congenital heart disease treated by non-specialized doctors: The potential loss of follow-up

ABSTRACT

Background: In the treatment of adult congenital heart disease (ACHD), the transfer of patients from pediatric cardiologists to ACHD cardiologists is of relevance. However, little is known about the clinical courses of ACHD patients that have been referred by non-CHD-specialized doctors (n-CSDs).

Methods: This retrospective cohort study included 230 patients (average age: 37 ± 15.2 years, male: 97) who were referred to a single specialized ACHD center between April 2016 and July 2019. We compared the characteristics and clinical courses between patients referred by n-CSDs and those referred by CHD-specialized doctors (CSDs).

Results: Overall, 121 (53%) patients were referred by n-CSDs. Among them, 91 (75%) patients were referred by adult cardiologists. Univariate analysis showed that the patients referred by n-CSDs were older than those referred by CSDs (41.6 ± 16.3 vs. 32.0 ± 12.0 years, $p < 0.01$), were more likely to have simple CHD, and less likely to have severe CHD (27.0% vs. 12.8% and 36.5% vs. 40.4%, respectively, $p < 0.01$). Patients referred by n-CSDs were also more likely to have a history of loss of follow-up (16.5% vs. 3.7%, $p < 0.01$) and to require invasive treatments after referral, including cardiac surgeries and transcatheter interventions (47.9% vs. 26.6 %, $p < 0.01$). Notably, unintended invasive treatments that were not designated by the referring doctors were more frequently required in patients with moderate complexity referred by n-CSDs (30.0% vs. 23.3%, $p = 0.02$).

Conclusions: Patients with moderate CHD complexity referred by n-CSDs are more likely to require unintended invasive treatments. Referrals to specialized ACHD centers may be most beneficial for these patients.

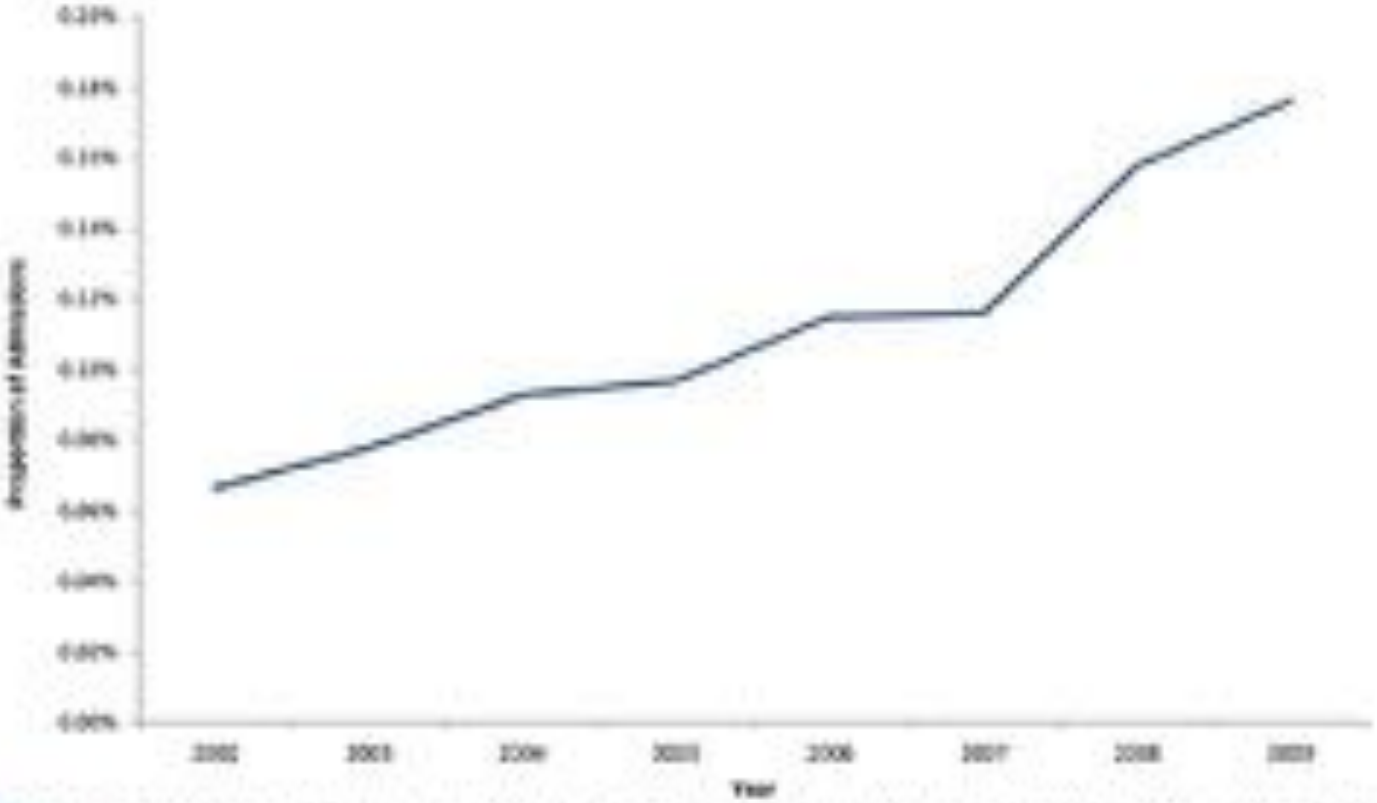

Fig. 1. Adult congenital heart disease admission records with noncardiac surgery as a percentage of all admissions with noncardiac surgery, by year (2002–2008). Discharge sample weights were used to produce national estimates. Trend is increasing over time (*P* value for trend is <0.001).

Table 2. Outcomes

	ACHD Cohort	Comparison Cohort		
	n = 15,004	n = 37,581		
Outcome	n (%)	n (%)	*P* Value	OR (95% CI)
Death	407 (4.1)	1,355 (3.6)	0.031	1.13 (1.01–1.27)
LOS (median [IQR])	4.8 (2.4–10.4)	2.9 (1.5–5.8)	<0.001	
Total charges (median [IQR])	$42,171 ($22,918–$93,847)	$26,962 ($15,814–$46,784)	<0.001	
ARF	620 (6.2)	1,826 (4.9)	<0.001	1.29 (1.18–1.42)
Pneumonia	942 (9.4)	3,998 (8.0)	<0.001	1.20 (1.11–1.29)
Respiratory failure	916 (9.2)	3,933 (7.8)	<0.001	1.19 (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	1.25 (1.13–1.40)
Composite	2,145 (21.4)	6,003 (16.0)	<0.001	1.44 (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.

2020 ESC Guidelines for the management of adult congenital heart disease

3.2 Organization of care

When patients with CHD are approaching adulthood, they require transfer to ACHD care. This transfer should be preceded by a preparatory transition phase, which continues into adulthood according to the needs of the patient. Special healthcare organization and training programmes are required to meet the needs of this patient population.[7] Importantly, the care for ACHD patients is a lifelong process

Table 5 Staff requirements for specialist ACHD centres*

Discipline	Required number
Adult/paediatric cardiologist with ACHD certification	≥2
ACHD imaging specialist (certified in TTE/TOE, CMR, CCT)	≥2
Congenital interventional cardiologist	≥2
CHD surgeon	≥2
Anaesthesiologist with CHD experience and expertise	≥2
Specialist nurse (if national professional nursing bodies allow specialization)	≥2
Invasive electrophysiologist with ACHD experience	≥1
Pulmonary vascular disease expert	≥1
Clinical geneticist	≥1
Psychologist	≥1
Social worker	≥1
Palliative care team	

ACHD = adult congenital heart disease, CHD = congenital heart disease, CMR = cardiovascular magnetic resonance, CCT = cardiovascular computed tomography, TOE = transoesophageal echocardiography, TTE = transthoracic echocardiography.
*Modified from Baumgartner et al.[4]

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3.2. Access to Care

Recommendation for Access to Care Referenced studies that support the recommendation are summarized in Online Data Supplement 3.		
COR	LOE	Recommendation
I	B-NR	1. Physicians caring for patients with ACHD should support access to care by a) assuring smooth transitions for adolescents and young adults from pediatric to adult providers[illegible] (Level of Evidence: B-NR); and b) promoting awareness of the need for lifelong specialized care through outreach and educational programs (Level of Evidence: C-EO).
	C-EO	

Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease

A Scientific Statement From the American Heart Association

Figure 1. Noncardiac complications in adults with congenital heart disease (CHD).

3.12. Noncardiac Surgery

Recommendations for Noncardiac Surgery

Referenced studies that support recommendations are summarized in Online Data Supplement 19.

COR	LOE	Recommendations
I	C-LD	1. Optimization before and close surveillance after invasive procedures, regardless of the complexity of the anatomic defect or type of procedure is beneficial for patients with ACHD. [illegible]
I	B-NR	2. In patients with ACHD AF classification IB-D, IIA-D, and IIIA-D* noncardiac surgical and interventional procedures should be performed in a hospital with or in consultation with experts in ACHD when possible. [illegible]

*See Tables 3 and 4 for details on the ACHD AF classification system.

3.12. Noncardiac Surgery

Recommendations for Noncardiac Surgery

Referenced studies that support recommendations are summarized in Online Data Supplement 18.

Table 6. Continued

Circumstance	Possible Solution	Rationale	Example
Administration of anesthesia for invasive procedures in patients with ACHD AP classification IB-D, IIA-D, and IIIA-D*	1. Performed by, or in collaboration with, an anesthesiologist with expertise in the management of patients with ACHD.	1. ACHD-specific issues need to be addressed when considering anesthesia, including underlying cardiac physiology and hemodynamics, and the effects of anesthetic medications and ventilation strategies.	The application of anesthesia for laparoscopic procedures can be especially challenging in Fontan patients. Significant cardiovascular and respiratory alterations may occur as a result of increased intra-abdominal pressure and decreased venous return. Abdominal insufflation may lead to lower preload and hypotension, while at the same time elevating systemic vascular resistance and compromising cardiac output. Deviations in pulmonary vascular resistance attributable to hypercarbia can be caused by either direct carbon dioxide absorption or hypoventilation.
	2. If clinical urgency precludes transfer, consultation with an anesthesiologist with ACHD expertise would be of benefit to on-site providers who do not have ACHD expertise.	2. Many patients with ACHD have had surgeries in the past, which may have created or identified airway or vascular access concerns. Patients with ACHD can also have underlying restrictive and/or obstructive lung disease that should be considered.	

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2018 ACHD Guideline

Table 4. ACHD AP Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

II Moderate Complexity
Repaired or unrepaired conditions
Aorto-left ventricular fistula
Anomalous pulmonary venous connection, partial or total
Anomalous coronary artery arising from the pulmonary artery
Anomalous aortic origin of a coronary artery from the opposite sinus
AVSD (partial or complete, including primum ASD)
Congenital aortic valve disease
Congenital mitral valve disease
Coarctation of the aorta
Ebstein anomaly (disease spectrum includes mild, moderate, and severe variations)
Infundibular right ventricular outflow obstruction
Ostium primum ASD
Moderate and large unrepaired secundum ASD
Moderate and large persistently patent ductus arteriosus
Pulmonary valve regurgitation (moderate or greater)
Pulmonary valve stenosis (moderate or greater)
Peripheral pulmonary stenosis
Sinus of Valsalva fistula/aneurysm
Sinus venosus defect
Subvalvar aortic stenosis (excluding HCM; HCM not addressed in these guidelines)
Supravalvar aortic stenosis
Straddling atrioventricular valve
Repaired tetralogy of Fallot
VSD with associated abnormality and/or moderate or greater shunt

Table 4. ACHD AP Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

III: Great Complexity (or Complex)
Cyanotic congenital heart defect (unrepaired or palliated, all forms)
Double-outlet ventricle
Fontan procedure
Interrupted aortic arch
Mitral atresia
Single ventricle (including double inlet left ventricle, tricuspid atresia, hypoplastic left heart, any other anatomic abnormality with a functionally single ventricle)

III: Great Complexity (or Complex) (Continued)
Pulmonary atresia (all forms)
TGA (classic or d-TGA; CCTGA or l-TGA)
Truncus arteriosus
Other abnormalities of atrioventricular and ventriculoarterial connection (ie, crisscross heart, isomerism, heterotaxy syndromes, ventricular inversion)

Table 4. ACHD AP Classification (CHD Anatomy + Physiological Stage = ACHD AP Classification)

Physiological Stage	
A	C
NYHA FC I symptoms	NYHA FC III symptoms
No hemodynamic or anatomic sequelae	Significant (moderate or greater) valvular disease, moderate or greater ventricular dysfunction (systemic, pulmonic, or both)
No arrhythmias	Moderate aortic enlargement
Normal exercise capacity	Stenosis of arterial stenosis
Normal renal/hepatic/pulmonary function	Mild or moderate hypoxemia/cyanosis
B	Hemodynamically significant shunt
NYHA FC II symptoms	Arrhythmias controlled with treatment
Mild hemodynamic sequelae (mild aortic enlargement, mild ventricular enlargement, mild ventricular dysfunction)	Pulmonary hypertension (less than severe)
Mild valvular disease	End-organ dysfunction responsive to therapy
Trivial or small shunt (not hemodynamically significant)	D
Arrhythmia not requiring treatment	NYHA FC IV symptoms
Abnormal objective cardiac limitation to exercise	Severe aortic enlargement
	Arrhythmias refractory to treatment
	Severe hypoxemia (almost always associated with cyanosis)
	Severe pulmonary hypertension
	Eisenmenger syndrome
	Refractory end-organ dysfunction

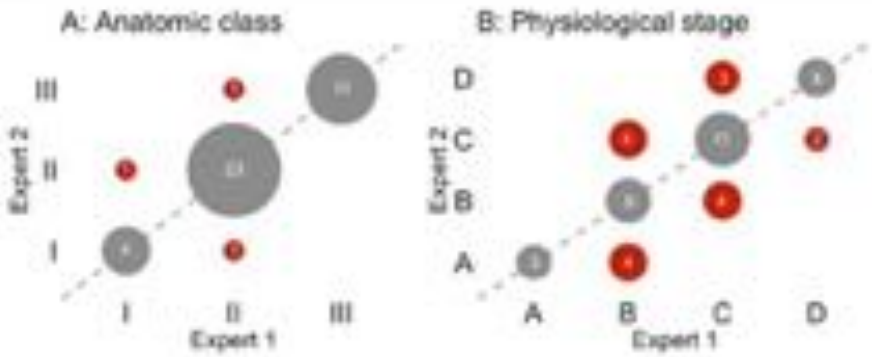
Interobserver agreement of the anatomic and physiological classification system for adult congenital heart disease

Table II. Interrater reliability of AnatC and PhyS for patients (n = 41) reviewed by a pair of experts

	Agreement (%)	κ_{LD} (95% CI)
AnatC	92.7	0.88 (0.75-1.00)
PhyS	58.5	0.57 (0.39-0.75)

Agreement is presented as percent agreement and κ_{LD}.

Figure 1



Disagreements between experts regarding AnatC and PhyS classification for 41 patients. Bubble matrix of expert agreement for AnatC (**A**, left) and PhyS (**B**, right). Bubble size corresponds to count. The dashed line indicates perfect agreement. Gray bubbles indicate cases of agreement, and red bubbles indicate cases where there was disagreement; all disagreements varied by only 1 level.

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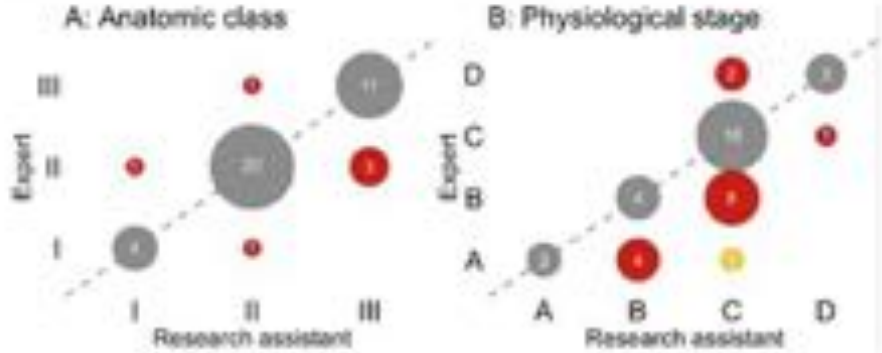
Interobserver agreement of the anatomic and physiological classification system for adult congenital heart disease

Table III. Interrater reliability of AnatC and PhyS between RAs and ACHD experts for 41 patients

	Agreement (%)	κ_{ω} (95% CI)
AnatC	85.4	0.77 (0.60-0.95)
PhyS	61.0	0.53 (0.34-0.72)

Agreement is reported as percent agreement and κ_{ω}.

Figure 2



Disagreements between RAs and experts regarding AnatC and PhyS classification for 41 patients. Bubble matrix of agreement between RAs and experts for AnatC (**A**, left) and PhyS (**B**, right). Bubble area corresponds to count. The dashed line indicates perfect agreement. Gray bubbles indicate cases of agreement, red bubbles indicate cases where there was disagreement by 1 level, and yellow indicates disagreement by 2 levels.

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Interobserver agreement of the anatomic and physiological classification system for adult congenital heart disease

Table IV. Three examples of disagreement between experts assigning AnatC and PhysS classifications

Case #	R1	R2	Clinical summary	Rater comments & notes	Disagreement source(s)	Disagreement category
1	IC	ID	CHD: subaortic stenosis Comorbidities: atrial fibrillation, currently on flecainide and metoprolol for rate and rhythm control, no ablation in prior 24 m	R2: Is the arrhythmia refractory to treatment (PhyS: D)? Patient had arrhythmia that recurred within 24 m but only started medication 10 d prior	Arrhythmia status	2, 3
2	IC	IB	CHD: aortic coarctation Comorbidities: HTN and decreased PVC	R1: PVC 60%-70% predicted. Also, would HTN in a patient with aortic coarctation be considered end-organ dysfunction? R2: Patient reports persistent fatigue	End-organ dysfunction, cardiopulmonary exercise testing	2
3	IB	IC	CHD: repaired tetralogy of Fallot Comorbidities: moderate pulmonary stenosis and severe pulmonary regurgitation, moderate right ventricle enlargement and good systolic function	No comment	Data oversight, valve disease	1

Raters were given the opportunity to make comments on a given case. Disagreement categories are (1) data oversight, (2) unclear definitions, and (3) disagreement about interpretation of the guidelines.
R1, rater 1; R2, rater 2; HTN, hypertension; PVC, forced vital capacity

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2018 AHA/ACC Guideline for the Management of Adults With Congenital Heart Disease

A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines

Classification :
ACHD AP = Anatomie + Physiologie

- ANATOMIE : Complexité
  - pathologie initiale
  - geste chirurgical
- PHYSIOLOGIE :
  - Shunt
  - Pathologie aortique
  - HTAP
  - Hypoxémie
  - Arythmie
  - Dysfonction d'organe

Questions à considérer :

- Prophylaxie de l'endocardite infectieuse
- Monoventricule sous jacent
- Anomalie anatomique modifiant les accès veineux et artériels
- Shunt persistant
- Valvulopathie
- Trouble du rythme
- Polyglobulie
- HTAP (oui? réversible?)
- Abord veineux (accès facile (portabilité gaine))
- Anticoagulation (curative?)
- Monitorage de la fonction rénale et hépatique
- Handicap

Facteurs de risque associé à une morbi-mortalité péri-opératoire :

- Cyanose
- Insuffisance cardiaque congestive
- Mauvais état général
- Jeune âge
- Hypertension pulmonaire
- Opération sur le système respiratoire ou neurologique
- Cardiopathie complexe
- Opération urgente

Analyse de la situation

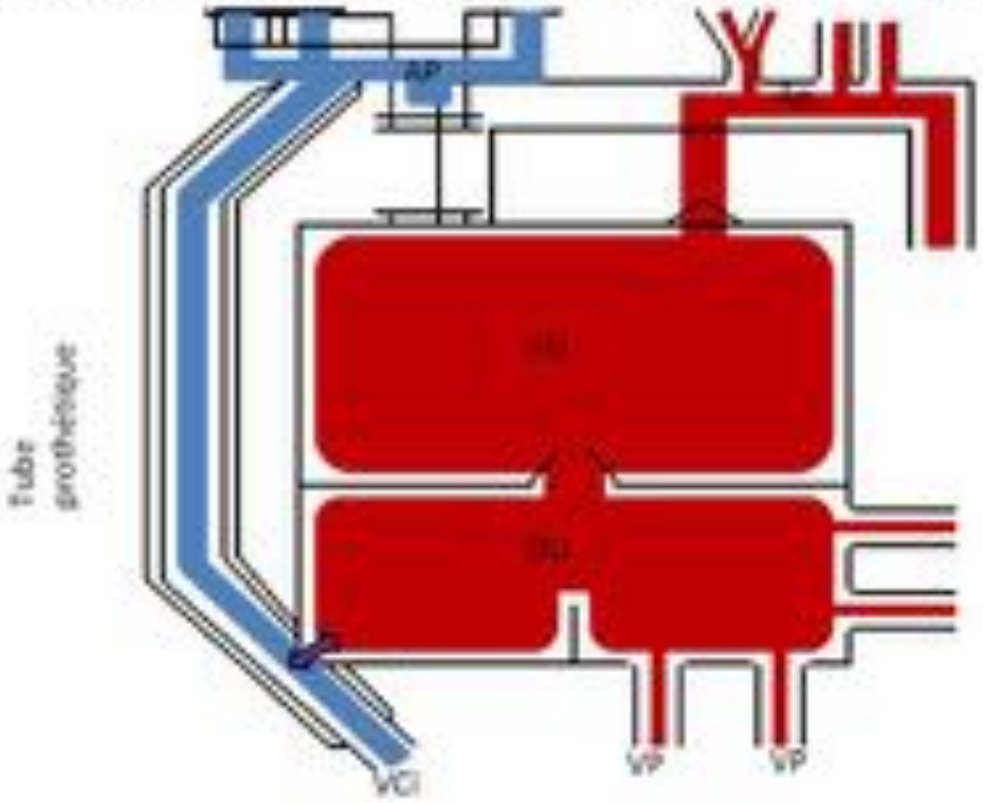
- Type de pathologie initiale
- Type de chirurgie (curative ou palliative)
- Lésions résiduelles+++
- Statut Fonctionnel+++
- Ne pas oublier les comorbidités++++

Table 12. Specific Management Practices for Cyanotic CHD

Recording clinical oxygen saturation at rest (>5 min) rather than immediately after effort (eg, walking into a clinic examination room).
Meticulous intravenous care to avoid air or particulate matter, which may include use of airparticulate filters on all intravenous access lines, when feasible, and careful de-airing of all lines.
Cerebral imaging for any new headache or neurologic sign to assess for possible cerebral abscess, hemorrhage, or stroke.
Measurement of serum uric acid and treatment with allopurinol in a patient with a history of gout.
Supplemental oxygen as needed for symptom relief but not to a target oxygen saturation level and not if there is no demonstrable symptomatic benefit.
Avoidance of or cautious use of therapies that may reduce the patient's hypoxia-mediated drive to ventilation, such as opioids or, in rare circumstances, excess supplemental oxygen.[illegible]
Anesthesia by providers with expertise in anesthesia for patients with ACHD for any noncardiac surgery.

Non-estrogen–containing birth control for women of child-bearing potential (intrauterine device may be a preferred option). Avoidance of birth control entirely is not a safe, acceptable option.
Patients can travel safely on commercial airlines without undue risk.[illegible] Preflight simulation testing or mandated supplemental oxygen are not usually indicated, although adequate hydration and movement during the flight are appropriate.
Measurement of coagulation parameters (eg, activated partial thromboplastin time, international normalized ratio, thrombin time) in a patient with an elevated hematocrit >55% requires adjustment of anticoagulant volume in the blood collection vials to account for reduced plasma volume in the draw.[illegible]

Montage univentriculaire / anastomose systémicopulmonaire



2020 ESC Guidelines for the management of adult congenital heart disease

Evaluation and Management of the Child and Adult With Fontan Circulation

A Scientific Statement From the American Heart Association

Table 4. Key Points: Arrhythmias

Atrial tachycardia develops in >50% of patients late after Fontan, although current surgical techniques may reduce this incidence.
Development of atrial tachycardia in patients with Fontan repairs is associated with significant morbidity, including heart failure and atrial thrombosis, and requires urgent and aggressive therapy.
Ventricular pacing may negatively affect cardiac output and lower quality-of-life scores.

Table 3. Prevalence of Arrhythmias and Pacemaker Placement

Arrhythmia	Atriopulmonary	Lateral Tunnel	Extracardiac Connection
Sinus node dysfunction, %	30–42	18–44	15–44
Supraventricular tachycardia, %	60–80	7–33	5–15
Pacemaker, %	11–23	5–55	3–27
Automated implantable cardioverter-defibrillator, %	1.4		
Ventricular tachycardia or sudden death, % (%/y)	2–10 (0.15–0.2)		

AHA SCIENTIFIC STATEMENT

Diagnosis and Management of Noncardiac Complications in Adults With Congenital Heart Disease

A Scientific Statement From the American Heart Association

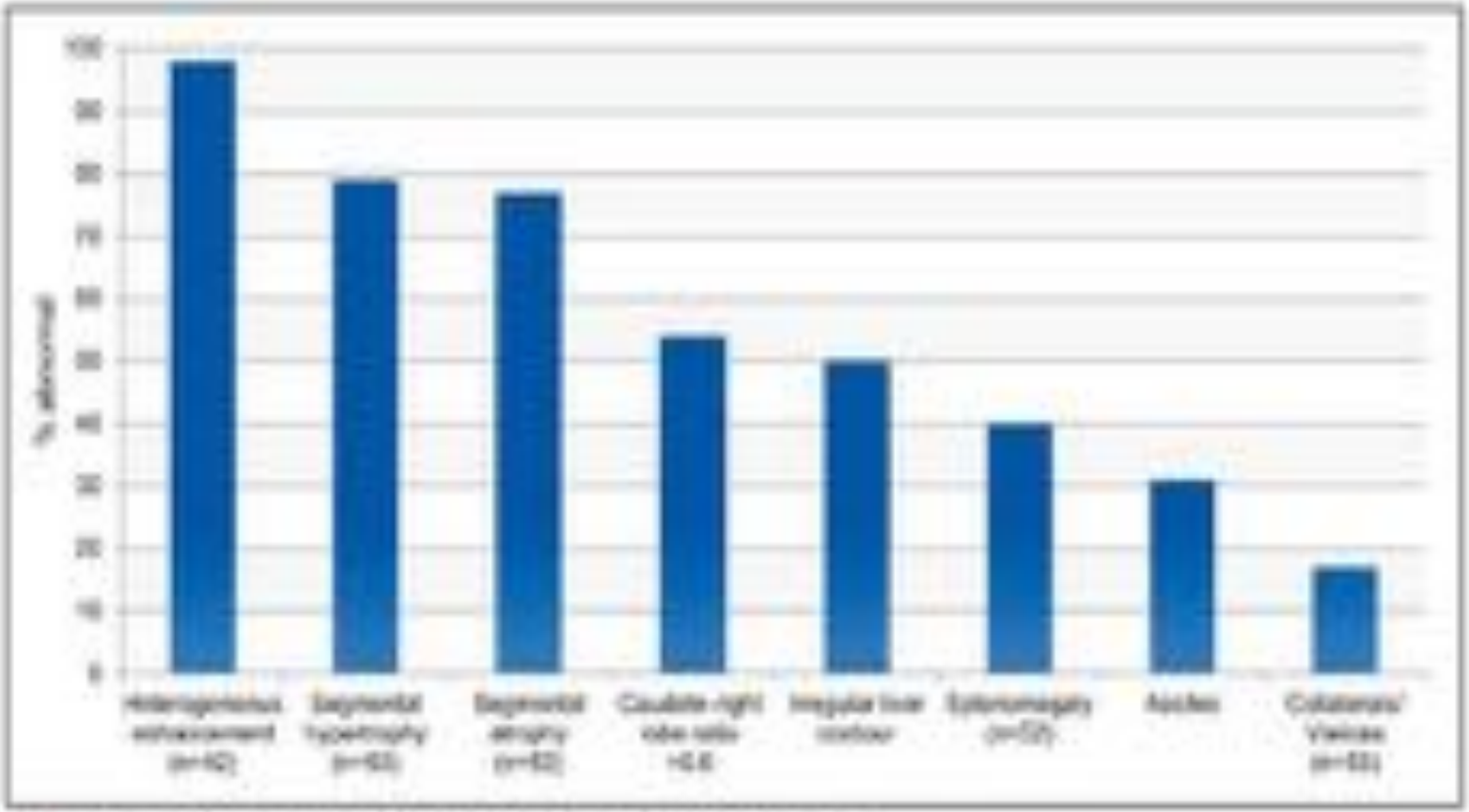
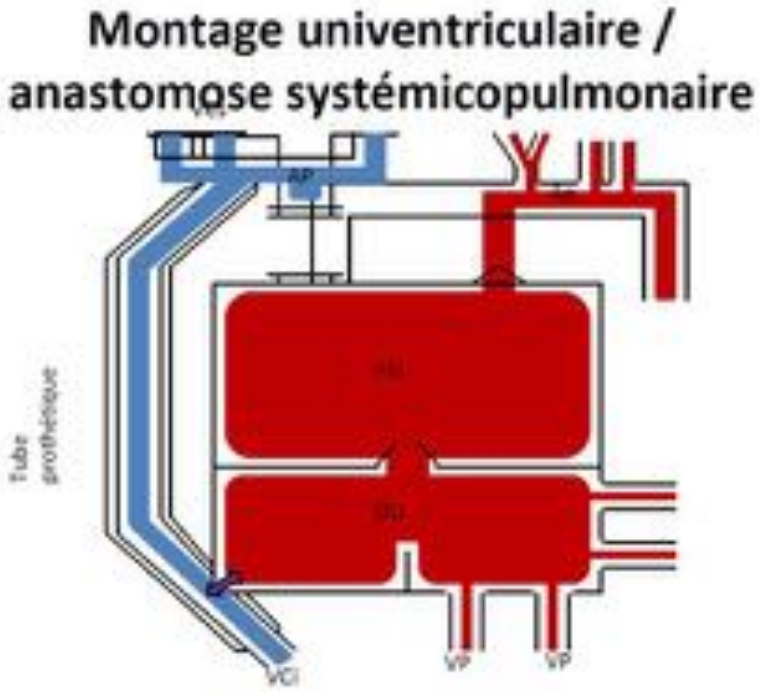


Figure 1. Findings on abdominal imaging (n=140) in the adult with Fontan circulation.

Reprinted from Wu et al[17] with permission from The American Association for Thoracic Surgery. Copyright © 2010, The American Association for Thoracic Surgery.

Laparoscopic Surgery in a Patient with Fontan Physiology



Craig D. McClain, MD

(Anesth Analg 2006;103:856-8)

CASE REPORT Open Access

A case report of Fontan procedure-related hepatocellular carcinoma: pure laparoscopic approach by low and stable pneumoperitoneum

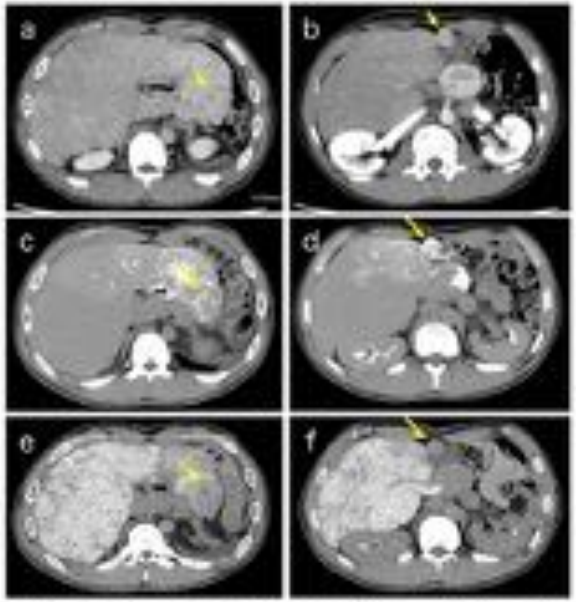


Fig. 1 Abdominal contrast-enhanced CT and CT arteriography and arterial portography. Abdominal contrast-enhanced CT examination revealed hypovascular mass in segment 2 (**a**), and a hypervascular mass in segment 4 on arterial phase, followed by delayed washout (**b**). CT arteriography showed that both masses were hypervascular tumors (**c**: segment 2, **d**: segment 4), and CT during arterial portography showed that both were low-density masses (**e**: segment 2, **f**: segment 4)

CASE REPORT Open Access

A case report of Fontan procedure-related hepatocellular carcinoma: pure laparoscopic approach by low and stable pneumoperitoneum

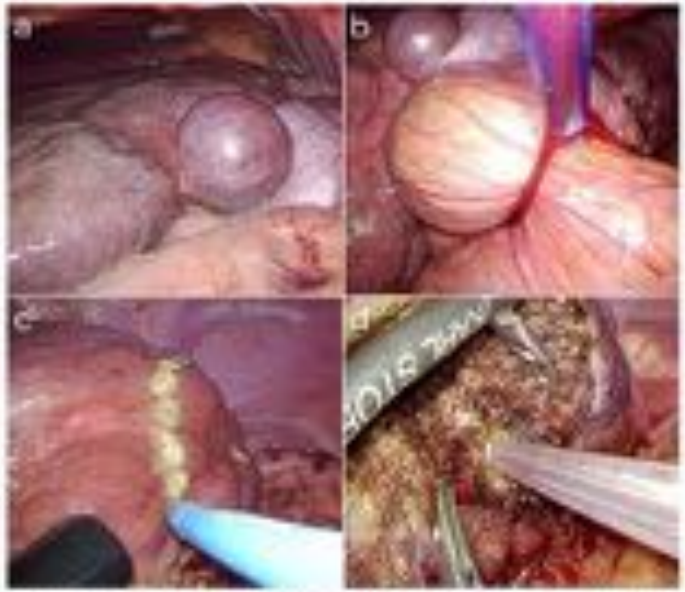
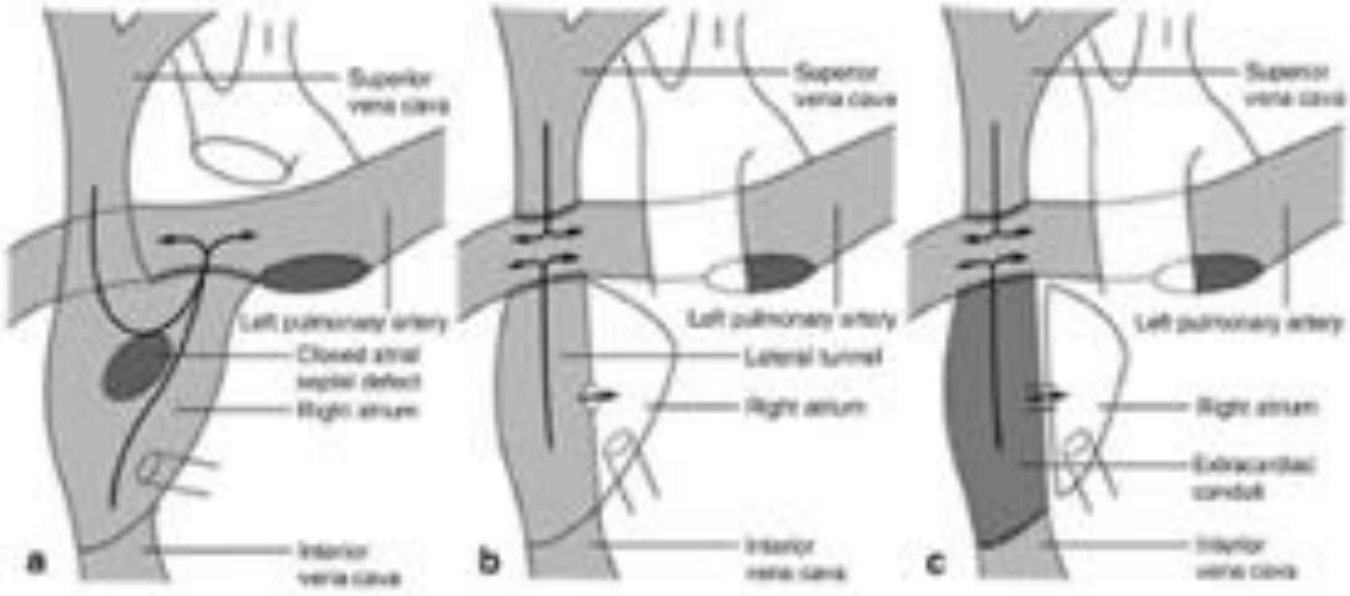


Fig. 2 Intraoperative findings. The liver revealed hepatic congestion due to Fontan circulation (a). The Pringle maneuver was applied during hepatic resection (b). The laparoscopic non-anatomical resections were performed (c, d)

Haemodynamics in a patient with Fontan physiology undergoing laparoscopic cholecystectomy

Fig. 1 Different types of Fontan circulation. **a** Atriopulmonary connection. **b** Intracardiac total cavopulmonary connection (lateral tunnel). Situation of the patient described in this case report. **c** Extracardiac total cavopulmonary connection. Reprinted by permission from Macmillan Publishers Ltd. [11]



S.J.A. Pans

Neth Heart J (2015) 23:383–385

Haemodynamics in a patient with Fontan physiology undergoing laparoscopic cholecystectomy

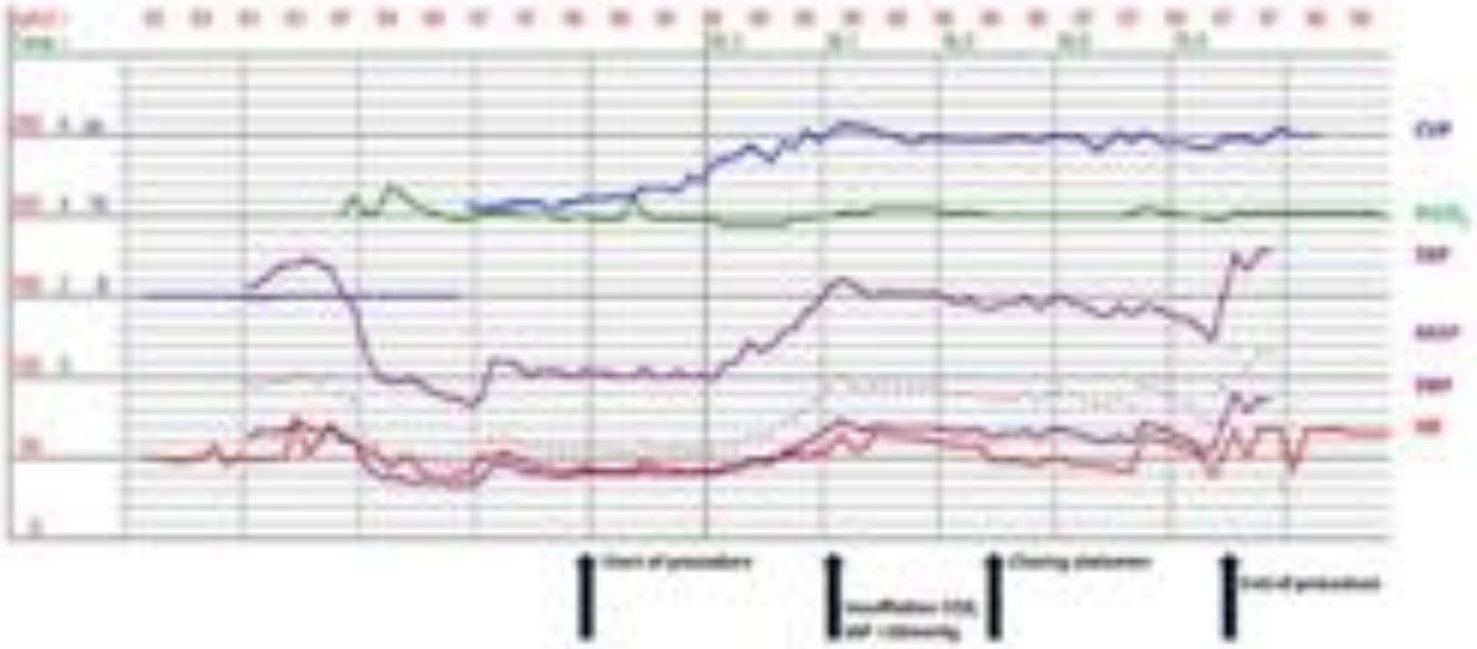


Fig. 2 Tracing of haemodynamic and ventilatory parameters. CVP central venous pressure, PetCO2 end-tidal carbon dioxide, SBP systolic blood pressure, MAP mean arterial pressure, DBP diastolic blood pressure, HR heart rate

S.J.A. Pans

Neth Heart J (2015) 23:383–385

Stratégie ?

-Type de pathologie initiale

-Type de correction BiV, UniV, palliative,.

-Lésions résiduelles+++

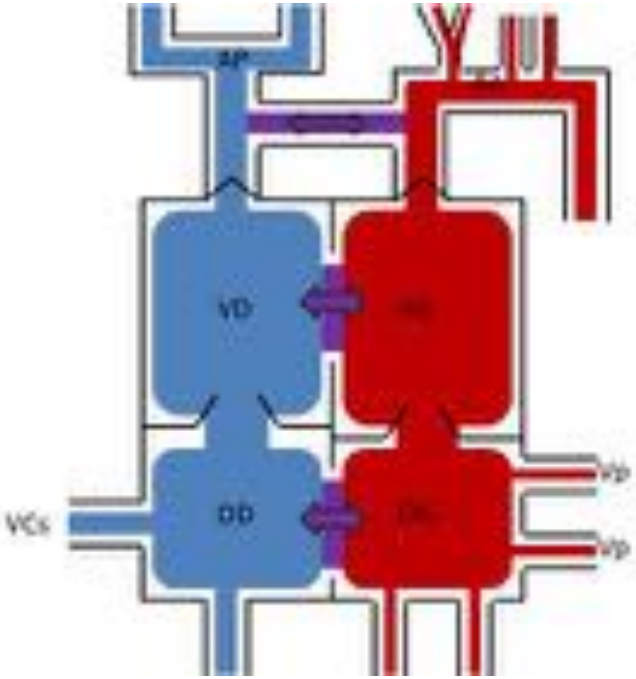
-Type physiologie (UniV, Cyanose,....)

-Accès veineux/artériels+++

-Comorbidités++/Atteintes Organes

-Stratégie a priori (anesthésie/monito/…)

-Surveillance post op+++



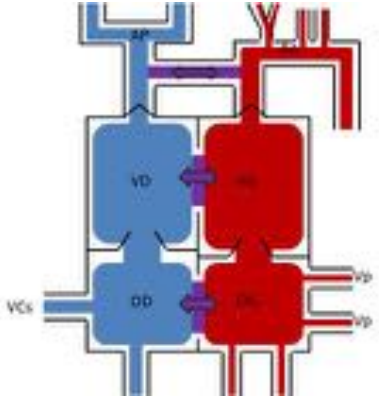
Stratégie ?

-Nourrisson 6 mois de vie chirurgie atrésie duodénale

Type 1, trisomie 21, à 2 mois cure complète CAVc,

Hémoptysies post opératoires > coelio ? timing Chir ?

Fibroscopie Bronchiques ?



-Nrs 2 mois de vie APSO Type 4 Di George,

Hirschprung, chirurgie abaissement colique avant 1e temps opératoire

Et Maintenant….

Ce n’est pas de la chirurgie non cardiaque…

Mais Nous allons parler de quelque chose de capital….

ESC
European Society of Cardiology
European Heart Journal (2018) 00, 1–41
doi:10.1093/eurheartj/ehy340

ESC GUIDELINES

2018 ESC Guidelines for the management of cardiovascular diseases during pregnancy

The Task Force for the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

Endorsed by: the International Society of Gender Medicine (IGM), the German Institute of Gender in Medicine (DGesGM), the European Society of Anaesthesiology (ESA), and the European Society of Gynecology (ESG)

ESC
European Society
of Cardiology

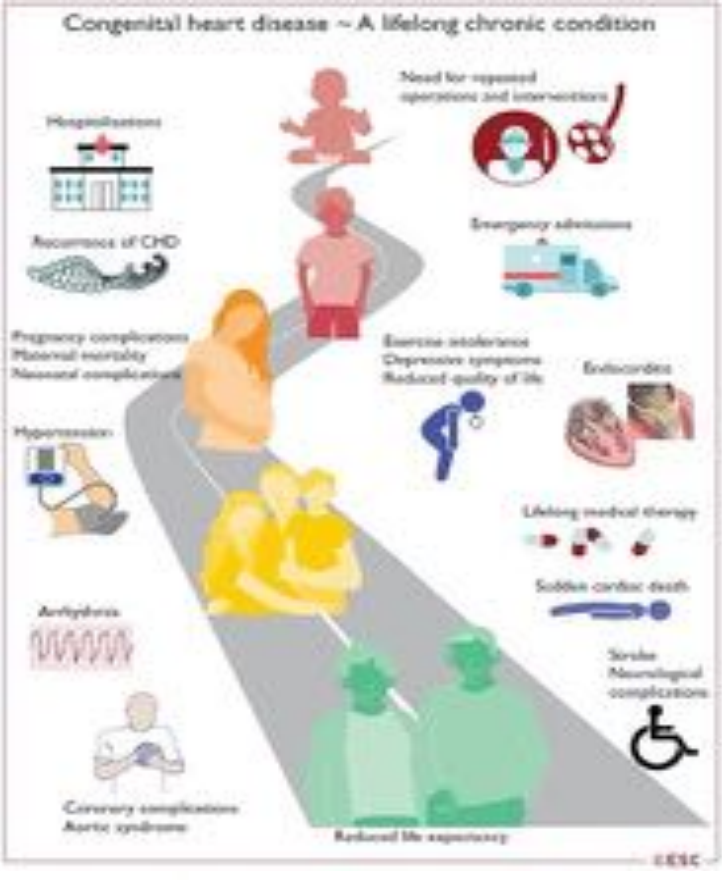


Figure 1 Central illustration. Congenital heart disease is a lifelong chronic condition. CHD = congenital heart disease; SCD = sudden cardiac death.

European Heart Journal (2020)

*NDLR. Ceci n'est pas une « recommandation pour la pratique clinique » élaborée selon la méthodologie complète recommandée par la HAS mais une **note technique** conçue en collaboration avec l'Agence de la biomédecine en attendant un document plus complet.*

Grossesses chez les femmes porteuses d'un syndrome de Turner : Attention danger !

31 août 2008 - *A. Lansac (Président du CNGOF).*

L'Agence de la biomédecine (ABM) signale deux cas de décès survenus en France en fin de grossesse chez des femmes porteuses d'un syndrome de Turner et vacciniées après un don d'ovocytes.

Chez les femmes porteuses d'un syndrome de Turner, les grossesses spontanées sont très rares (1 %)[1]. La don d'ovocytes constitue pour la cible auparavant d'autre alternative possible à obtenir une grossesse.

Ces grossesses sont dangereuses car 5 à 10 % des femmes atteintes d'un syndrome de Turner ont une malformation cardiovasculaire sévère[1,2,3] : coarctation de l'aorte (10 % des cas), valve aortique bicuspide chez 1/3 des patientes.[3]
Les complications obstétricales les plus sévères sont donc cardiovasculaires, telles que l'extrémisation d'une hypertension préexistante et la dissection aortique pouvant, comme dans le syndrome de Marfan, entraîner le décès[4]. Ce risque de décès par dissection, dissection ou rupture artérielle est estimé à dix fois chez des femmes porteuses du syndrome et est augmentée le risque de décès 100 fois supérieur à la normale et à 50 % des dissections artérielles qui peuvent se produire en fin de grossesse[5]. Une revue de la littérature entre 1961 et 2004 a répertorié 15 cas de dissection de l'aorte chez des femmes porteuses d'un syndrome de Turner. Sur les 7 cas survenus après PMA, 4 femmes sont décédées.[6,7]. Parfois des cas de rupture hépatique ont été observés.[8,9]

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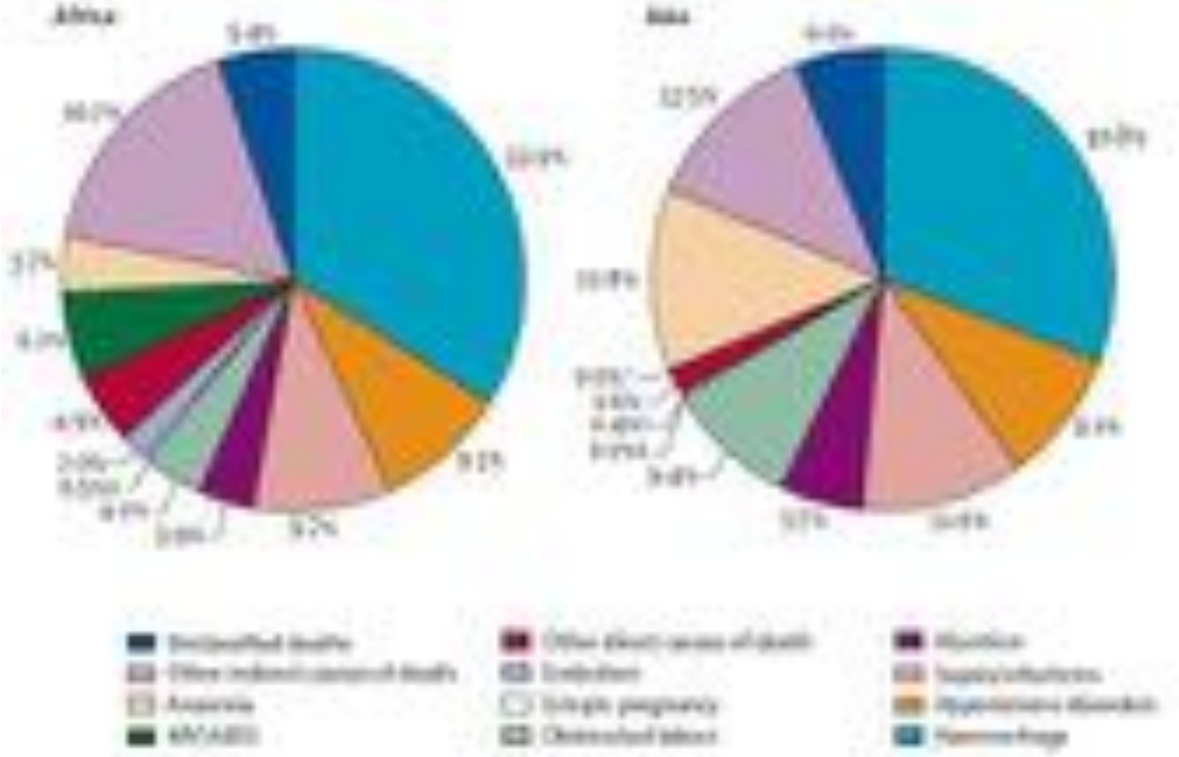
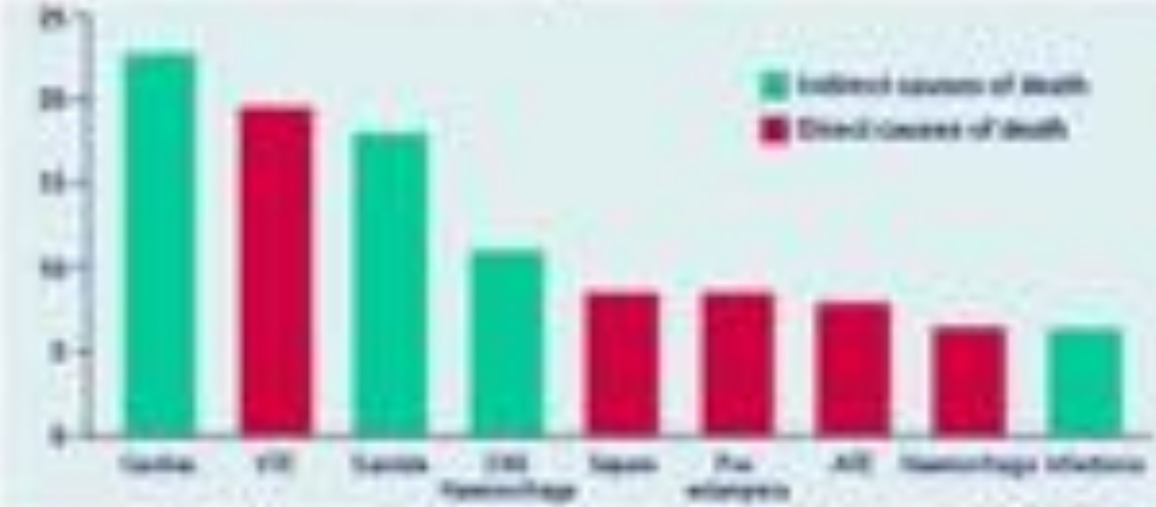


Figure 3: Geographical variation in distribution of causes of maternal deaths
*Represents HIV/AIDS. †Represents embolism. ‡Represents ectopic pregnancy. §Represents anaemia

Major Causes of Maternal Death (UK 2003-2005)

Overall death rates per million maternities



ENQUÊTE NATIONALE CONFIDENTIELLE SUR LES MORTS MATERNELLES (ENCMM) 2013-2015.

Synthèse

JANVIER 2021

POINTS CLÉS

- Entre 2013 et 2015, 262 morts maternelles ont été enregistrées en France, soit un ratio de mortalité maternelle jusqu'à 1 an de la fin de la grossesse de 10,8 décès pour 100 000 naissances vivantes.
- Malgré leur rareté, une amélioration est encore possible, car plus de la moitié des décès maternels sont considérés comme probablement ou possiblement évitables et dans deux tiers des cas, les soins dispensés n'ont pas été optimaux.
- La mortalité par hémorragie obstétricale poursuit sa diminution, divisée par deux en quinze ans
- Les maladies cardiovasculaires et les suicides deviennent les deux premières causes de mortalité maternelle.
- Le risque de mortalité maternelle augmente avec l'âge des femmes à partir de 30 ans
- La mortalité des femmes migrantes est plus élevée que celle des femmes nées en France.
- Territorialement, les DOM et l'Île-de-France se distinguent par un niveau de mortalité maternelle plus élevé.

Les maladies cardiovasculaires

- Une dyspnée récente, s'aggravant, en particulier en fin de grossesse et en post-partum, n'est pas banale et doit faire évoquer une complication cardiaque.
- Devant une douleur thoracique chez la femme enceinte, la dissection aortique doit être envisagée avec le même degré d'urgence que l'infarctus du myocarde ou l'embolie pulmonaire, même en l'absence de maladie du tissu conjonctif connue.

Outcome of pregnancy in patients with structural or ischaemic heart disease: results of a registry of the European Society of Cardiology

-Registre international

-**R**egistry **O**f **P**regnancy **A**nd **C**ardiac disease

-multicentrique / Prospective

-depuis 2007

-Europe/Nord Afrique/Etats Unis/

-Sous section en fonction pathologie

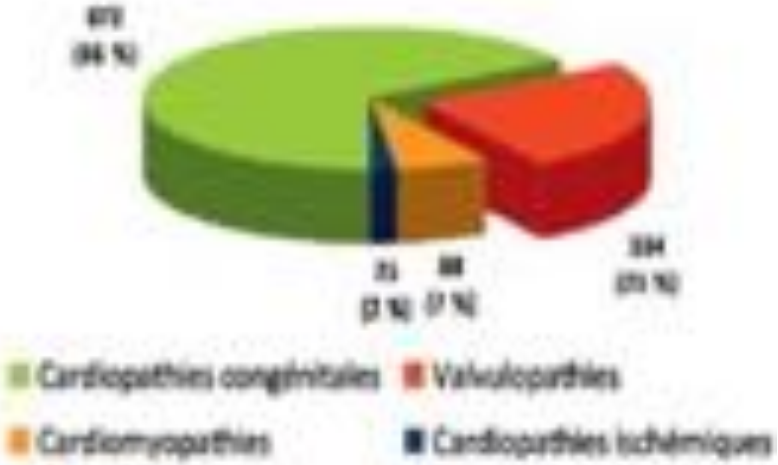
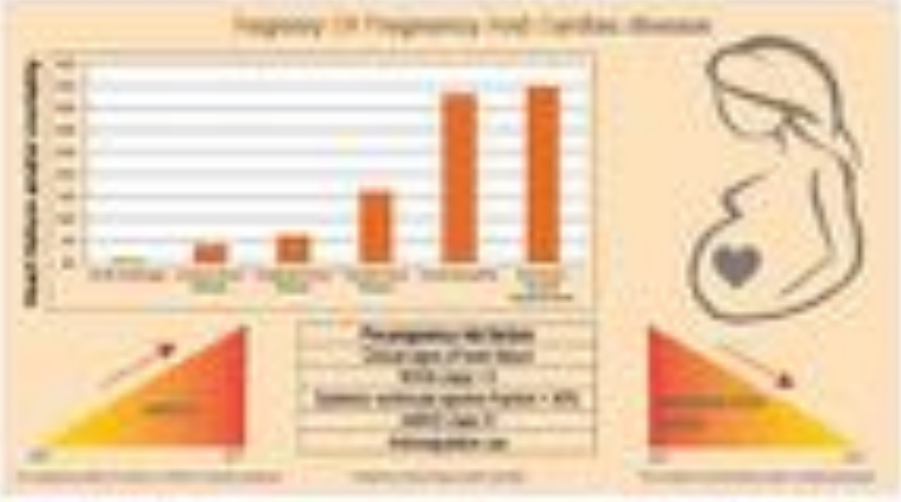


Figure 9.2. Étiologies des cardiopathies présentes durant la grossesse entre 2007–2011 dans le registre de la Société Européenne de Cardiologie [4].

Pregnancy outcomes in women with cardiovascular disease: evolving trends over 10 years in the ESC Registry Of Pregnancy And Cardiac disease (ROPAC)



Take home figure From 2007 to 2016, pregnant women with heart disease were included in the Registry Of Pregnancy And Cardiac disease (ROPAC). During that time, the incidence of 'maternal death or heart failure' stagnated, despite the inclusion of more seriously ill women (mWHO II). Especially women with cardiomyopathies or pulmonary arterial hypertension were at risk of 'maternal death or heart failure'. Pre-pregnancy risk factors were signs of heart failure prior to pregnancy (NYHA class > II, systemic ventricular ejection fraction of below 40%, mWHO class IV conditions) and anticoagulation use.

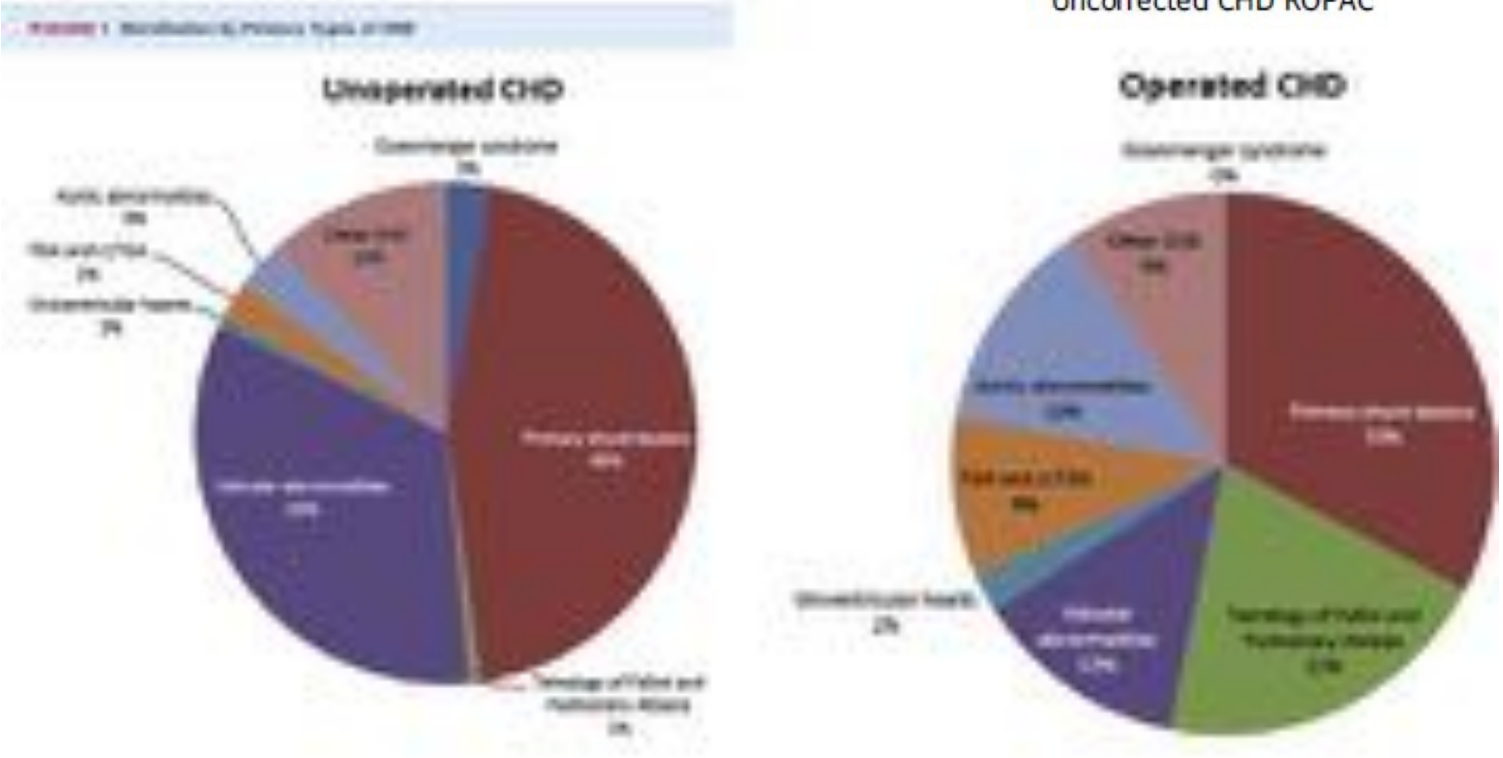
Pregnant Women With Uncorrected Congenital Heart Disease

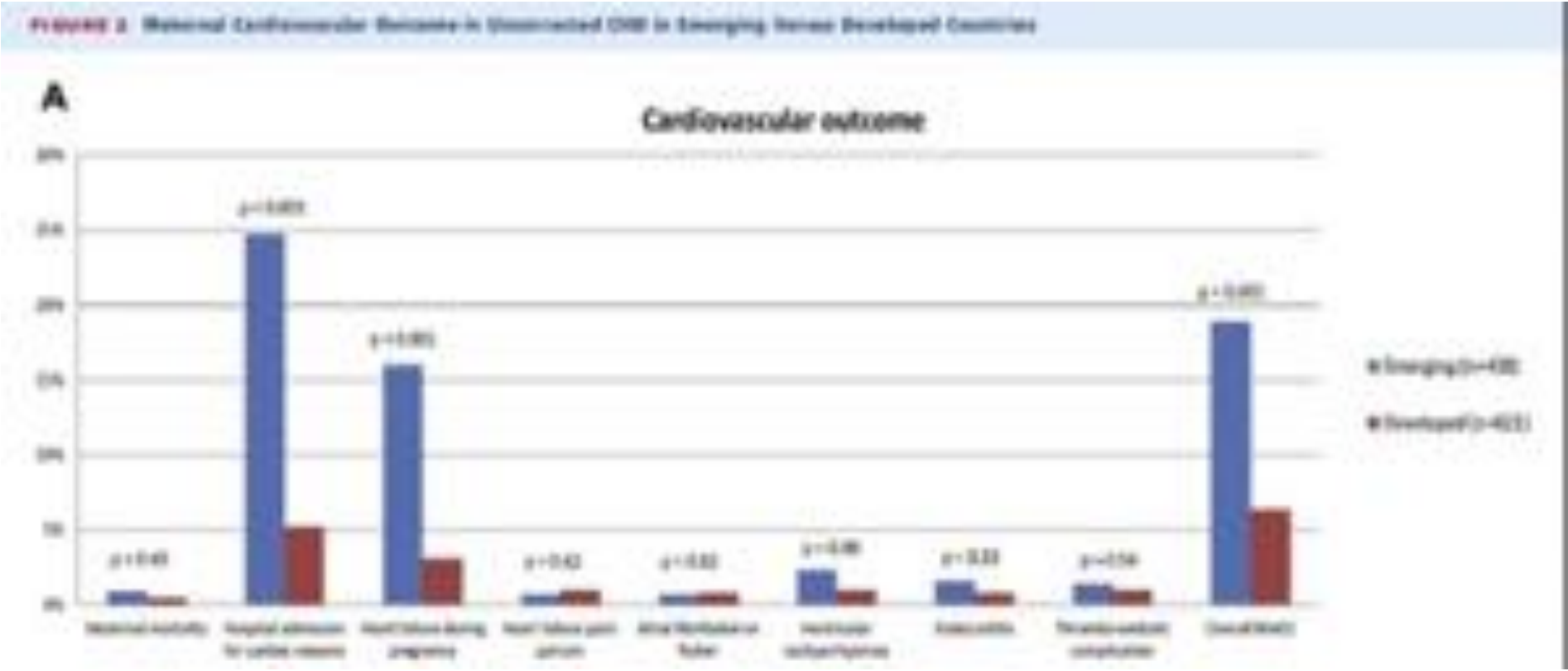
Heart Failure and Mortality

JACC: HEART FAILURE VOL. 8, NO. 2, 2020

Sliwa *et al.*

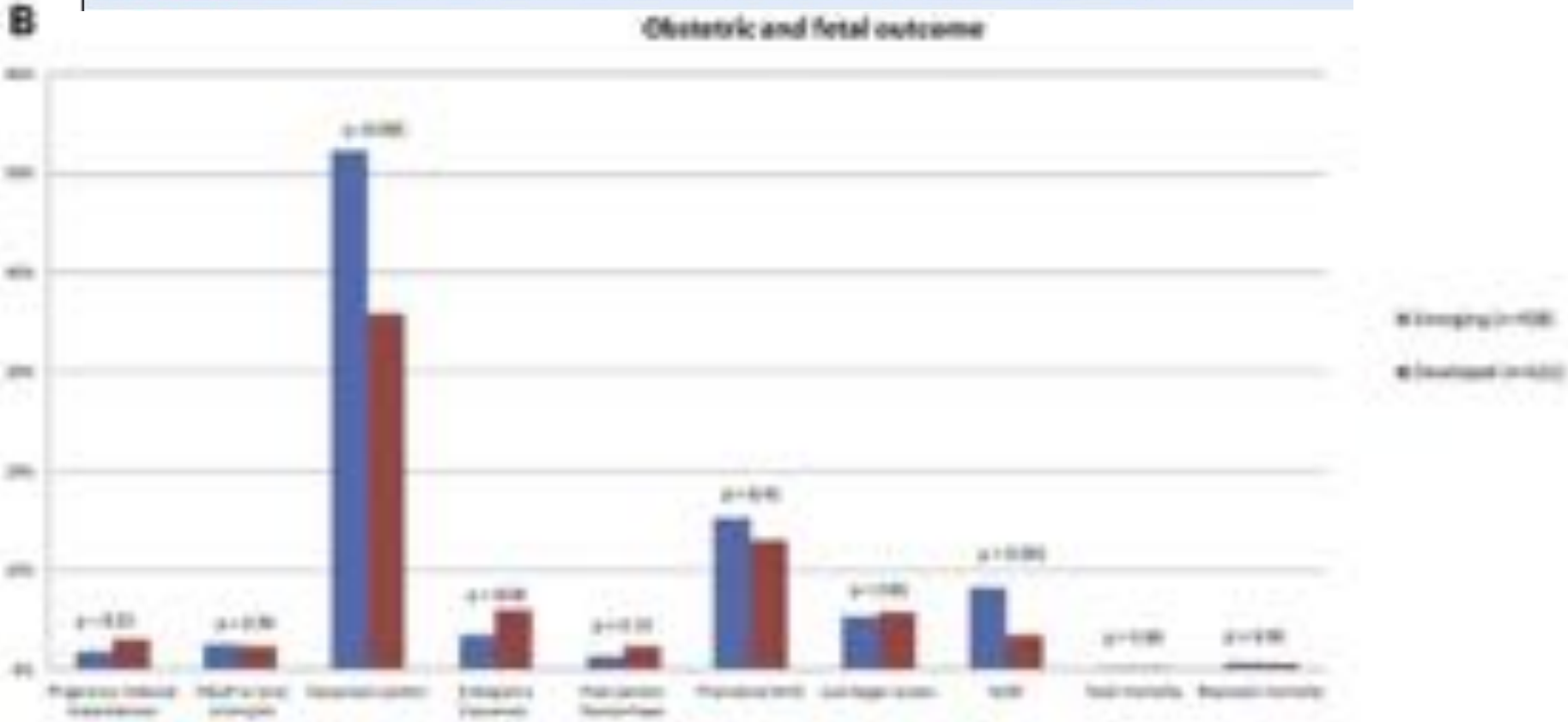
Uncorrected CHD ROPAC





(A) Maternal cardiovascular outcome in women with uncorrected CHD emerging versus developed countries.

FIGURE 2 Maternal Cardiovascular Outcome in Uncorrected CHD in Emerging Versus Developed Countries



(B) Obstetric and fetal outcomes in women with uncorrected CHD emerging versus developed countries.

European Heart Journal (2011) 32, 3147–3197
doi:10.1093/eurheartj/ehr218

ESC GUIDELINES

ESC Guidelines on the management of cardiovascular diseases during pregnancy

The Task Force on the Management of Cardiovascular Diseases during Pregnancy of the European Society of Cardiology (ESC)

Endorsed by the European Society of Gynecology (ESG), the Association for European Paediatric Cardiology (AEPC), and the German Society for Gender Medicine (DGesGM) et autour l'année de l'ESH !

New concepts
Enhancing WHO classification of maternal risk
Introduction of the pregnancy heart team
More attention for assisted reproductive therapy
Discussion of the use of bromocriptine in PPCM
Introduction of specific levels of surveillance based on low/medium/high risk for arrhythmia with haemodynamic compromise at delivery
New information on pharmacokinetics in pregnancy, more detailed information on pharmacodynamics in animal experiments on all drugs (Supplementary Data)
Peripartum caesarean section is discussed
Advice on contraception and the termination of pregnancy in women with cardiac disease is now provided

Table 3 Modified World Health Organization classification of maternal cardiovascular risk

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Diagnosis (if either wise well and uncomplicated)	Small or mild – pulmonary stenosis – patent ductus arteriosus – mitral valve prolapse Successfully repaired simple lesions (atrial or ventricular septal defect, patent ductus arteriosus, anomalous pulmonary venous drainage) Atrial or ventricular ectopic beats isolated	Unoperated atrial or ventricular septal defect Repaired tetralogy of Fallot Mild arrhythmias (supraventricular arrhythmias) Turner syndrome without aortic dilatation	Mild left ventricular impairment (EF 45–50%) Hypertrophic cardiomyopathy Marfan or related connective tissue disease not considered WHO I or IV (mild mitral valvopathy, moderate aortic stenosis) Marfan or other HCAD syndrome without aortic dilatation Aorta >45 mm in bicuspid aortic valve pathology Repaired coarctation Atrioventricular septal defect	Moderate left ventricular impairment (EF 30–45%) Previous peripartum cardiomyopathy without any residual left ventricular impairment Mechanical valve Systemic right ventricle with good or mildly decreased ventricular function Fontan circulation If otherwise the patient is well and the cardiac condition uncomplicated Unrepaired cyanotic heart disease Other complex heart disease Moderate mitral stenosis Severe asymptomatic aortic stenosis Moderate aortic dilatation (45–49 mm in Marfan syndrome or other HCAD), 40–50 mm in bicuspid aortic valve, Turner syndrome 40–45 mm/m², tetralogy of Fallot >50 mm	Pulmonary arterial hypertension Severe systemic ventricular dysfunction (EF <30% or NYHA class III–IV) Previous peripartum cardiomyopathy with any residual left ventricular impairment Severe mitral stenosis Severe symptomatic aortic stenosis Systemic right ventricle with moderate or severely decreased ventricular function Severe aortic dilatation (>49 mm in Marfan syndrome or other HCAD), >50 mm in bicuspid aortic valve, Turner syndrome >45 mm/m², tetralogy of Fallot >50 mm Vascular Ehlers-Danlos Severe coarctation Fontan with any complication

Table 3 **Modified World Health Organization classification of maternal cardiovascular risk**

	mWHO I	mWHO II	mWHO II–III	mWHO III	mWHO IV
Risk	No detectable increased risk of maternal mortality and minimal increased risk in morbidity	Small increased risk of maternal mortality or moderate increase in morbidity	Intermediate increased risk of maternal mortality or moderate to severe increase in morbidity	Significantly increased risk of maternal mortality or severe morbidity	Extremely high risk of maternal mortality or severe morbidity
Maternal cardiac event rate	2.5–5%	5.7–10.5%	10–19%	19–27%	40–100%
Counselling	Yes	Yes	Yes	Two expert counselling required	Two pregnancy contraindicated. If pregnancy occurs, termination should be discussed
Care during pregnancy	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease
Minimal follow-up visits during pregnancy	Once or twice	Once per trimester	Bimonthly	Monthly or bimonthly	Monthly
Location of delivery	Local hospital	Local hospital	Referral hospital	Expert centre for pregnancy and cardiac disease	Expert centre for pregnancy and cardiac disease

General recommendations

Recommendations	Class[a]	Level[b]
Pre-pregnancy risk assessment and counselling is indicated in all women with known or suspected congenital or acquired cardiovascular and aortic disease.[39]	I	C
It is recommended to perform risk assessment in all women with cardiac diseases of childbearing age before and after conception, using the mWHO classification of maternal risk.[11]	I	C
It is recommended that high-risk patients are treated in specialized centres by a multidisciplinary pregnancy heart team.[39]	I	C
Vaginal delivery is recommended as the first choice in most patients; for most important exceptions see below.[96]	I	C
Induction of labour should be considered at 40 weeks of gestation in all women with cardiac disease.	IIa	C
Caesarean delivery should be considered for obstetrical indications or for patients with dilatation of the ascending aorta >45 mm, severe aortic stenosis, pre-term labour while on oral anticoagulants, Eisenmenger's syndrome, or severe heart failure.	IIa	C

3.8.7 Anaesthesia/analgesia

Epidural analgesia reduces labour pain and can be used to provide anaesthesia for caesarean section if necessary. However, it can cause systemic hypotension (10%) and must be carefully titrated, especially in patients with obstructive valve lesions or diminished ventricular function who may benefit from invasive BP monitoring. All i.v. fluids need to be used carefully.[101]

Why is post-partum haemorrhage more common in women with congenital heart disease?

Matthew Cauldwell[a,*], Kate Von Klemperer[b], Anselm Uebing[b], Lorna Swan[b], Philip J. Steer[a], Michael Gatzoulis[b], Mark R. Johnson[a]

ORIGINAL ARTICLE

Is a planned caesarean section in women with cardiac disease beneficial?

Titia P E Ruys,[1] Jolien W Roos-Hesselink,[1] Antonia Pijuan-Domènech,[2] Elena Vasario,[3] Ishat R Gaisin,[4] Bernard Iung,[5] Leisa J Freeman,[6] Elaine P Gordon,[7] Petronella G Pieper,[8] Roger Hall,[6] Eric Boersma,[1] Mark R Johnson,[9] on behalf of the ROPAC investigators

Vaginal delivery is recommended as the first choice in most patients; for most important exceptions see below.[96]	I	C
Induction of labour should be considered at 40 weeks of gestation in all women with cardiac disease.	IIa	C

Caesarean delivery should be considered for obstetrical indications or for patients with dilatation of the ascending aorta >45 mm, severe aortic stenosis, pre-term labour while on oral anticoagulants, Eisenmenger's syndrome, or severe heart failure.	IIa	C

Complications liées à la grossesse chez les patientes présentant une cardiopathie congénitale : étude rétrospective au sein d'un centre de compétence M3C.

DEVILLE THOMAS

LE 18 OCTOBRE 2018

SOUS LA DIRECTION DE PASCAL AVIERINOS

Avec l'aimable autorisation du Dr DEVILLE THOMAS

Incidence des complications

28 % cardiovasculaires maternelles

27% per partum
à 28SA en moyenne

12% post partum

Agglomérats pavimenteux (FT)
+ oreillettes

E + AIT + dilatation AO + D

AUCUN DECES

60% obstétricales

à 29 SA en moyenne

Dont : 10% lésions vasculaires

15% IMG

MAP> HSG PE> DG > Saignements

30% périnatales

25% fœtales
à 28SA en moyenne

RCIU > Int. Th.Hydro > ADCF

62% néonatales

Prématurité 39%
RNS 24%
Neurologie 12%
Morbidité adaptation 9%

1 DECES NEONATAL !!

Avec l'aimable autorisation du Dr DEVILLE THOMAS

Scores prédictifs des risques ?

CARPREG	Oui	Non	Non	Oui (6 mois)	Oui
Mamo	Non précisé	Non précisé	Non précisé	Non précisé	Non précisé
mORWG	Oui	Non	Non	Oui (2 semaines)	Non précisé
CARDIA	Oui	Non	Non	Oui (6 mois)	Oui
CARPREG II	Oui	Non	Non	Oui (6 mois)	Oui

Avec l'aimable autorisation du Dr DEVILLE THOMAS

Score prédictif des risques ?

Cinq scores de risque de complication cardiaque pendant la grossesse et leurs sous-groupes de sévérité

	CARPREG I	CARPREG II	Karnis	mSWG	ZAHARA
Sévérité 1	0 point	0 à 1 point	Risque faible	Classe I	1,3% de risque
Sévérité 2	1 point	2 points	Risque modéré	Classe II	7,1 % de risque
Sévérité 3	> 1 point	3 points	Risque élevé	Classe III	17,5% de risque
Sévérité 4	-	4 points		Classe IV	41,1% de risque
Sévérité 5	-	> 4 points	-	-	70% de risque

Avec l'aimable autorisation du Dr DEVILLE THOMAS

Take home message

Incidence des complications :

- [illegible]
- [illegible]

Application des 3 scores de risque dans notre population :

- [illegible]

Parcours de soins :

- [illegible]
- [illegible]
- [illegible]

Encourager les patients à informer une prescription au plus tôt suivi en centre de compétence MOC, suivi, dépistage et gestion à prescrire, créer des scores de risque

Avec l'aimable
autorisation du
Dr DEVILLE THOMAS

S C 1993 :
cardiopathie/montage chirurgical

- Cardiopathie congénitale : ventricule unique sur atrésie tricuspide.

- Antécédents cardiovasculaires :

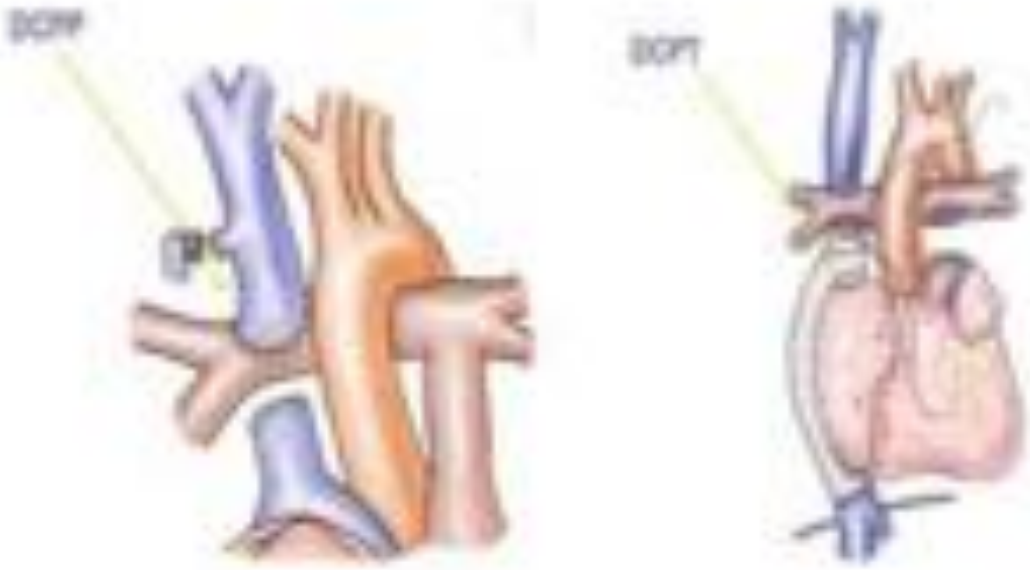
– 26/07/93 : Blalock droit (mise position tube en Goretex n°5).

– 3/06/93 : DCPP avec élargissement de l'artère pulmonaire droite par patch de Goretex

– 26/06/2001 : DCPT (tube extra cardiaque de Gonetex N°20)

Atrésie tricuspide> Blalock

Circulation de Fontan



AHA SCIENTIFIC STATEMENT

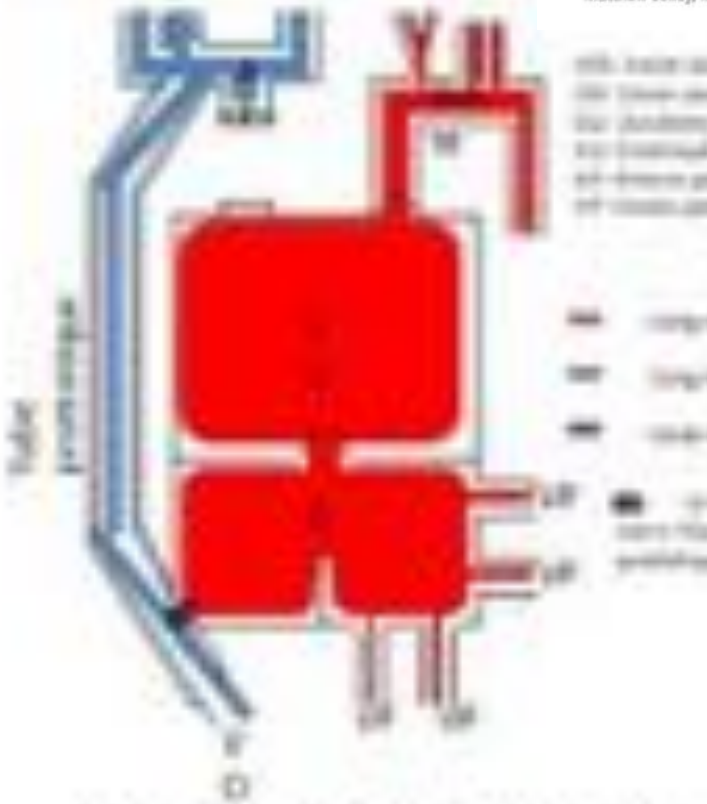
Evaluation and Management of the Child and Adult With Fontan Circulation

A Scientific Statement From the American Heart Association

REVIEW ARTICLE

Fontan Physiology Revisited

Matthew Jolley, MD,*† Steven D. Colan, MD,† Jonathan Rhodes, MD,† and James DiNardo, MD*



S C 16/07/1993 : situation clinique

- Antécédents : [illegible]
- Clinique : [illegible]
- Traitement : [illegible]
- ECG : [illegible]
- Holter ECG : [illegible]
- Biologie : [illegible]
- ETT : [illegible]

COMPTE-RENDU DE CATHETERISME CARDIAQUE

CHU Arnaud de Villeneuve, Montpellier

[illegible]

Procédure: Évaluation hémodynamique dans le cadre d'une atrésie tricuspide totalisée avec désir de grossesse

Trajet des sondes: [illegible]

Site	Condition 1	
	Pression (mmHg)	Sat(%)
VCI		
VCS	[illegible]	
OD		
VD		
TAP		
WPC diast	[illegible]	
PC moyenne		
OG		
VG	[illegible]	
Ao Asc	[illegible]	
Ao thorax		
Ao Desc	[illegible]	

Intervention(s)

Embolisation de l'artère mammaire int g avec un AVP 4 de 5 mm + 2 coils Embolium

Angiographies

- 1.0 2 Angiographie descendante de face normale (pas de collatérales visibles) puis injection sélective dans l'artère mammaire g montrant un retour veineux pulmonaire modérée. Embolisation de celle-ci puis injection sélective de la mammaire droite normale
- 0.0 Angiographie de face dans la VCS montrant les veines pulmonaires correctement bloquées, sans dérivée. Retour veineux pulmonaire normal (pas de malformations artério-veineuses visibles)

Conclusion

- Hémodynamique normale voire même vasculaire
- Embolisation artère mammaire g

Alain Franco, Jean-Christophe Bouza

Maternal and fetal outcomes of pregnancy with Fontan circulation: A multicentric observational study

Marielle Gouton[a,b], Jacky Nizard[c], Michal Patel[d], François Savoldo[e], Maria Jaworska[f], Jelena Radojevic[g], Aneel Mathurin[h], Pascal Amedro[i], Elise Bayre[j], Fabien Labombarda[k], Guy Valéryanès[l], Alain Dumesnil[a,m], Laurianne Le Clerc[n], Magalie Ladouceur[a,o,p]

ABSTRACT

Background: Despite serious long-term sequel, women with Fontan palliation have reached childbearing age. However there's paucity of data on the pregnancy outcomes and management of this condition. We aimed to determine the maternal and fetal outcomes of pregnancy in women with Fontan palliation.

Methods: The multicentric, retrospective study included women with Fontan circulation followed in 13 French specialized centers from January 1990 to June 2016. All pregnancies were reviewed, including miscarriages, abortions, terminations and term births. We reviewed maternal and fetal outcomes.

Results: Thirty-seven patients had 99 pregnancies. Mean age was 27 ± 5 years at first pregnancy. There were 18 miscarriages (27%) and 36 live births with 1 twin pregnancy. Cardiac events occurred in 6 (16%) pregnancies, with no maternal death. The most common cardiac complication was atrial arrhythmia, which occurred in 3 pregnancies. Hematological complications including thromboembolic hemorrhagic events (n = 17) occurred in 5 women antepartum (n = 2 [7%] and 4 women postpartum (n = 14 [4]). Two of the 2 thromboembolic events occurred in patients without anticoagulation. There was a high incidence of prematurity (n = 25 [70, 95%]). Anticoagulation was associated with adverse neonatal outcome (OR = 10.8, 95% CI [1.3–91.4], p = 0.01). After a median follow-up of 24 months, there was no significant worsening of clinical status and thromboembolic disease noted.

Conclusions: Pre-selected women can successfully complete pregnancy with Fontan circulation. There is an increase in cardiac and neonatal morbidity during pregnancy. Because thromboembolism could have a severe consequence on Fontan circulation, anticoagulation should be indicated during pregnancy and postpartum period.

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Table 3
Maternal baseline characteristics, TOF, total cohort and subgroups, overview.

Characteristics	
Age at first pregnancy, mean ± SD, 4 years	27 ± 5
Functional class	
I/II/III	21 (13%)
IV/NYHA	16 (10%)
I/II/III, I and II	8
Hypertension, Lancaster – 1000	1
Past medical history	
Atrial arrhythmia	7
Permanent pacemaker implant	7
Internal cardioverter defibrillator	1
Heart failure	8
Thromboembolism	7
Hemodynamics	
Normal systemic ventricular function	21 (14%)
Mild systemic ventricle dysfunction	41 (11%)
Severe systemic ventricle dysfunction	8
Atrio-ventricular valve regurgitation grade I–II/III–IV	34 I–II/4
TOF, pulmonary flow	8

Table 4
Obstetrical complications and neonatal outcomes. CHD: congenital heart disease, PPC: premature gestation, LVAC: left superior vena cava, SV: single ventricle.

Obstetrical complications	n
Miscarriage	14 (7%)
Chorioamnionitis	1
Tachycardia (>3)	1
Preeclampsia	1
Premature rupture of membranes	7
Premature labor	4
Postpartum hemorrhage	1
Fetal/neonatal complications	**n**
Live births	18.7 (term pregnancy)
Birth weight (g), mean ± SD	2988 ± 78
Fetal/neonatal complications	
Intrauterine fetal death	1
Stillbirth	1
Neonatal death	1
Fetal distress	4
SGA (<10th percentile)	4
Prematurity (term – 37 WG)	23 (10%)
33–36 + 6 WG	23
28–32 + 6 WG	7
22–27 + 6 WG	1
Recurrence of CHD	1/1 (CVC, 1%)

Retrospective UK multicentre study of the pregnancy outcomes of women with a Fontan repair

Matthew Cauldwell,[1] Philip J Steer,[1] Samantha Bonner,[2] Omar Asghar,[3] Lorna Swan,[4] Kenneth Hudson,[5] Catherine G O'Head,[6] Adam Daniel Jones,[7] Nicola Ridler,[8] Margaret Simpson,[9] Aidan P Bridge,[10] Tarun Siddiqui,[10] Katherine W English,[11] Lucy Mawdsley,[12] Dilip Abraham,[13] Andrew J Sands,[14] Aarthi R Nikkar,[15] Stephanie L Curtis,[16] Louise Coats,[17-19] Mark R Johnson[1]

Cauldwell M, et al. Heart 2018;104:401–406. doi:10.1136/heartjnl-2017-311783

Methods A retrospective study of women with a Fontan circulation delivering between January 2005 and November 2016 in 10 specialist adult congenital heart disease centres in the UK.

Results 50 women had 126 pregnancies, resulting in 68 (54.0%) miscarriages, 2 terminations of pregnancy, 3 intrauterine deaths (at 30 weeks), 53 (42.1%) live births and 6 neonatal deaths. Cardiac complications in pregnancies with a live birth included heart failure (n=7, 13.3%), arrhythmia (n=6, 11.3%) and pulmonary embolism (n=1, 1.9%). Very low baseline maternal saturations (less than 85% measured, compared with 60 of 116 (51.7%) who had baseline saturations of ≥85% (p=0.008). Obstetric and neonatal complications were common: preterm delivery (n=38, 72.2%), small for gestational age (<10th percentile, n=30, 55.6%, <5th centile, n=19, 35.2%) and postpartum haemorrhage (n=23, 42.6%). There were no maternal deaths in the study period.

Table 2 Baseline maternal characteristics at first pregnancy

Demographics (n=50 unless stated otherwise)	N (%)
Age (years, median [IQR])	27 [24–29]
(minimum and maximum)	19–36
Body mass index (median and IQR)	21 [19–26]
(minimum and maximum)	18–39
Functional status	
NYHA class I	40 (80)
NYHA class II	10 (20)
NYHA class III and IV	0 (0)
Smoking status	1 (2)
Baseline arrhythmia (n=52)	10 (20)
Prior atrial arrhythmia	10 (20)
Prior ablation	10 (20)
Permanent pacemaker	7 (14)
Prior thrombosis	2 (4)
Prior thromboembolism	0 (0)
Cardiotoxicities	
Anthracycline	2 (4)
Hypertension	7 (14)
Ventricular function echocardiography	
Normal	40 (80)
Mildly impaired	10 (20)

Ventricular size (apical four chamber echocardiography)	
Mild	10 (20)
Moderate	10 (20)
Severe	10 (20)
Severe	0 (0)
CARPEG score	
0	16 (32)
1	26 (52)
2	8 (16)
3 and 4	0 (0)

CARPEG, Cardiac Disease in Pregnancy Risk; NYHA, New York Heart Association.

Table 3 Cardiac medication prescribed prior to pregnancy

Live births (n=53)	N (%)
No antiplatelet agents/anticoagulants	1 (1.9)
Aspirin	9 (17.0)*
Clopidogrel	1 (1.9)
Warfarin	32 (60.4)*
Aspirin and low molecular weight heparin – prophylactic dose	2 (3.8)
Beta blockers	6 (11.3)
ACE inhibitors	3 (5.7)
Diuretics	2 (3.8)
No medication	3 (5.7)

*One patient discontinued prior pregnancy

Table 4 Maternal cardiac complications during pregnancy

Total live births (n=53)	N (%)
Heart failure	7 (13.2)
Arrhythmia	6 (11.3)
Venous thromboembolism	1 (1.9)

Table 5 Obstetric and neonatal complications

Obstetric complications (total pregnancies n=1243)	N (%)*
Miscarriage	
First trimester	[illegible]
Second trimester	[illegible]
Obstetric complications (total live births n=1[illegible])	
Blood loss >1000 ml	[illegible]
Pre-eclampsia	[illegible]
Pregnancy-induced hypertension	[illegible]
Gross postpartum clots	[illegible]
Spontaneous preterm birth	[illegible]
Preterm delivery—fetal indication only	[illegible]
Preterm delivery—fetal and maternal indication	[illegible]
Preterm premature rupture of membranes	[illegible]
Neonatal complications (total births n=1[illegible])	
Intrauterine death	[illegible]
Neonatal death	[illegible]
SGA (<10th centile)	[illegible]
Preterm delivery	
<37 weeks	[illegible]
<33 weeks	[illegible]
<28 weeks	[illegible]

*Only eight women in our cohort had a pregnancy without a documented obstetric complication.

SGA, small for gestational age.

Cauldwell M, et al. Heart 2018;104:401–406. doi:10.1136/heartjnl-2017-311793

BMJ

S C 16/07/1993
Au total

- [illegible]
- [illegible]
  - [illegible]
  - [illegible]
  - [illegible]
- [illegible]
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  - [illegible]
  - [illegible]
- [illegible]

Mais….. Fausse couche….

ORIGINAL ARTICLE

Pregnancy in Women With a Fontan Circulation

A Systematic Review of the Literature

WHAT THE STUDY ADDS

- Arrhythmia and heart failure are the most frequent cardiac complications among women after Fontan palliation during pregnancy.
- Miscarriage are common in these patients, and successful pregnancies can be frequently complicated with premature deliveries and intrauterine growth retardation.
- Postpartum hemorrhage is common among these patients.

METHODS AND RESULTS: We conducted a systematic review to evaluate maternal and fetal outcomes of pregnancy in women with a Fontan circulation. Among 1150 studies that were screened, 6 studies had sufficient longitudinal data points to qualify for meaningful inclusion, yielding 255 pregnancies in 133 women after Fontan procedure resulting in 115 live births (45%, including reports from 1986 to 2015). There was a total of 137 pregnancy losses (89%), with 115 miscarriages (45%), 19 elective terminations of pregnancy (7%), 2 stillbirths (1%), and 1 ectopic pregnancy (1%). The most common cardiovascular adverse events were supraventricular arrhythmia affecting 8.4% (range, 3%–37%) and heart failure affecting 3.9% (range, 3%–11%) of pregnancies. These complications were successfully managed with conventional approaches. No maternal deaths were reported. Postpartum hemorrhage was the predominant obstetric complication affecting 14% of the patients. Most patients were on antiplatelet agents (27%) or anticoagulants (50%) whereas only a minority (11%) were on neither. Among the 115 live births, 68 were premature (59%), 37 were small for gestational age (20%), and neonatal death occurred in 6 patients (5%).

Take Home Messages

- Population en augmentation
- Risque perdu de vue importante+++
- Collision pathologie initiale

    Syndrome génétique / période néonatale

    Type de réparation / Lésions résiduelles++++

    Comorbidités Acquises+++

- Stratégie globale / Equipe entraînée multidisciplinaire+++
- ”Le Hasard ne favorise que les esprits préparés”