Epidemiology of congenital heart diseases

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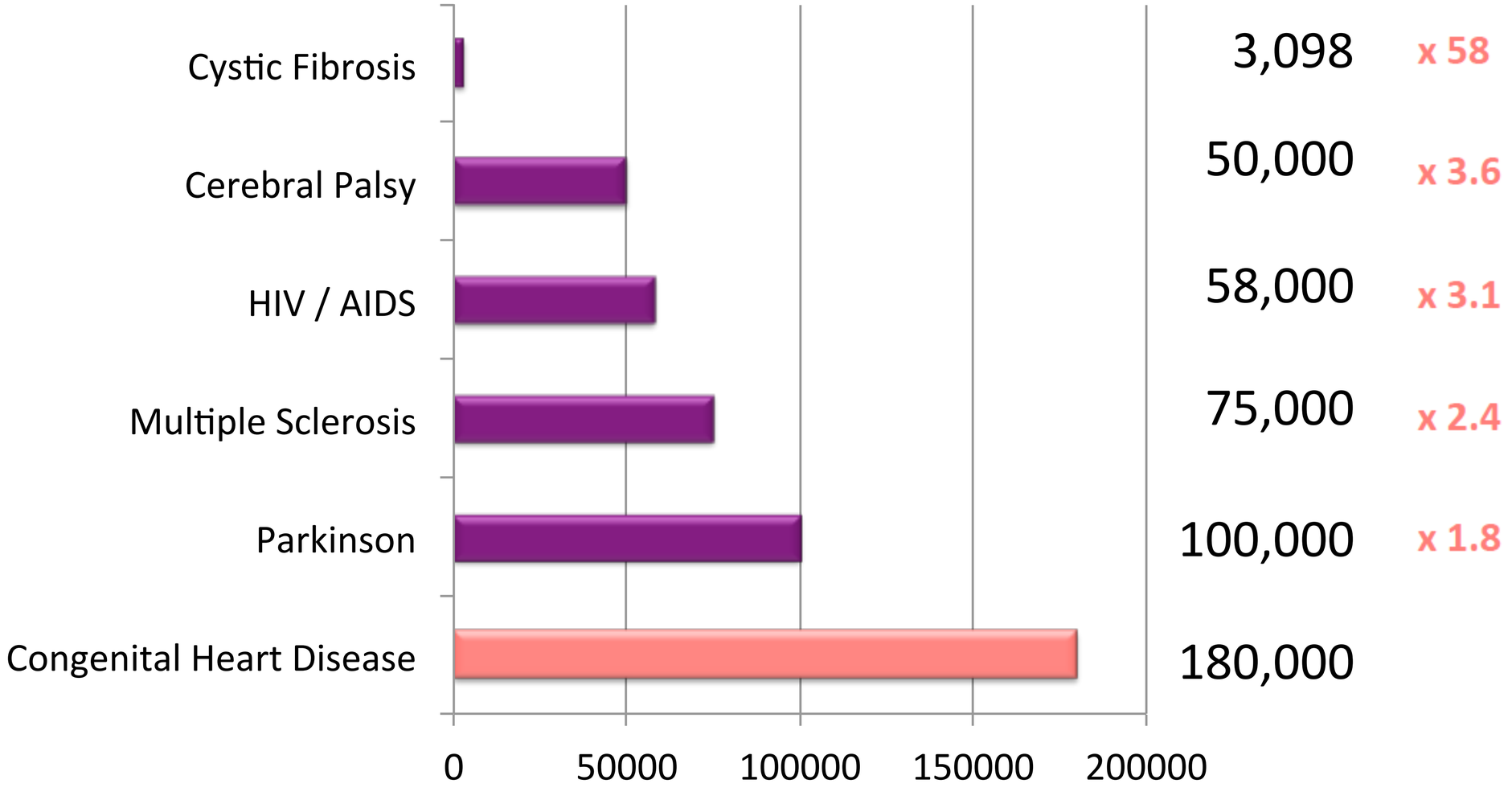
Maladies Cardiaques Héréditaires- CARDIOGEN

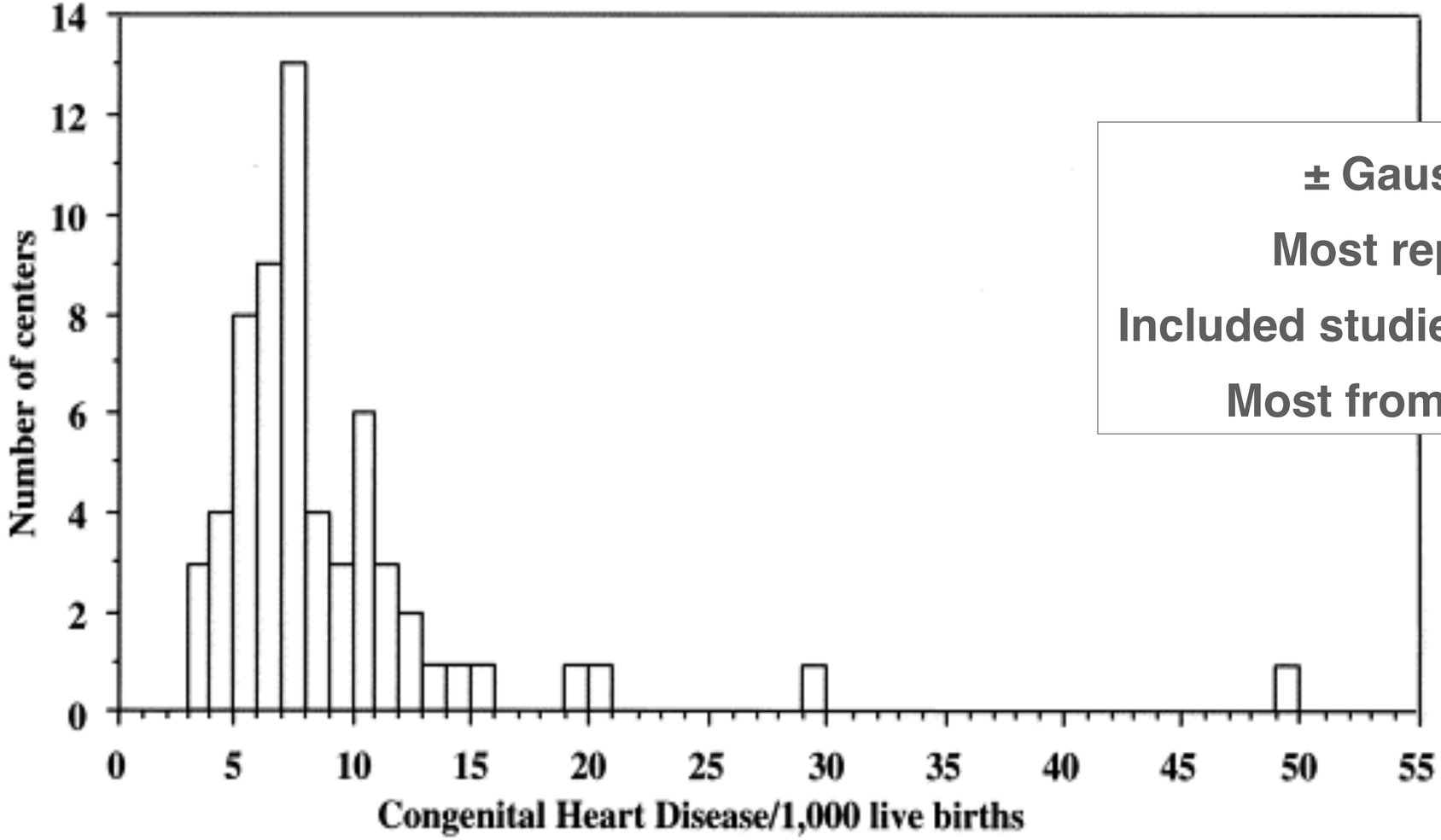
Epidemiology of congenital heart diseases

Comparison with other common diseases



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

Histogram of the incidence of congenital heart diseases per 1,000 live births in 62 reports



± Gaussian curve;

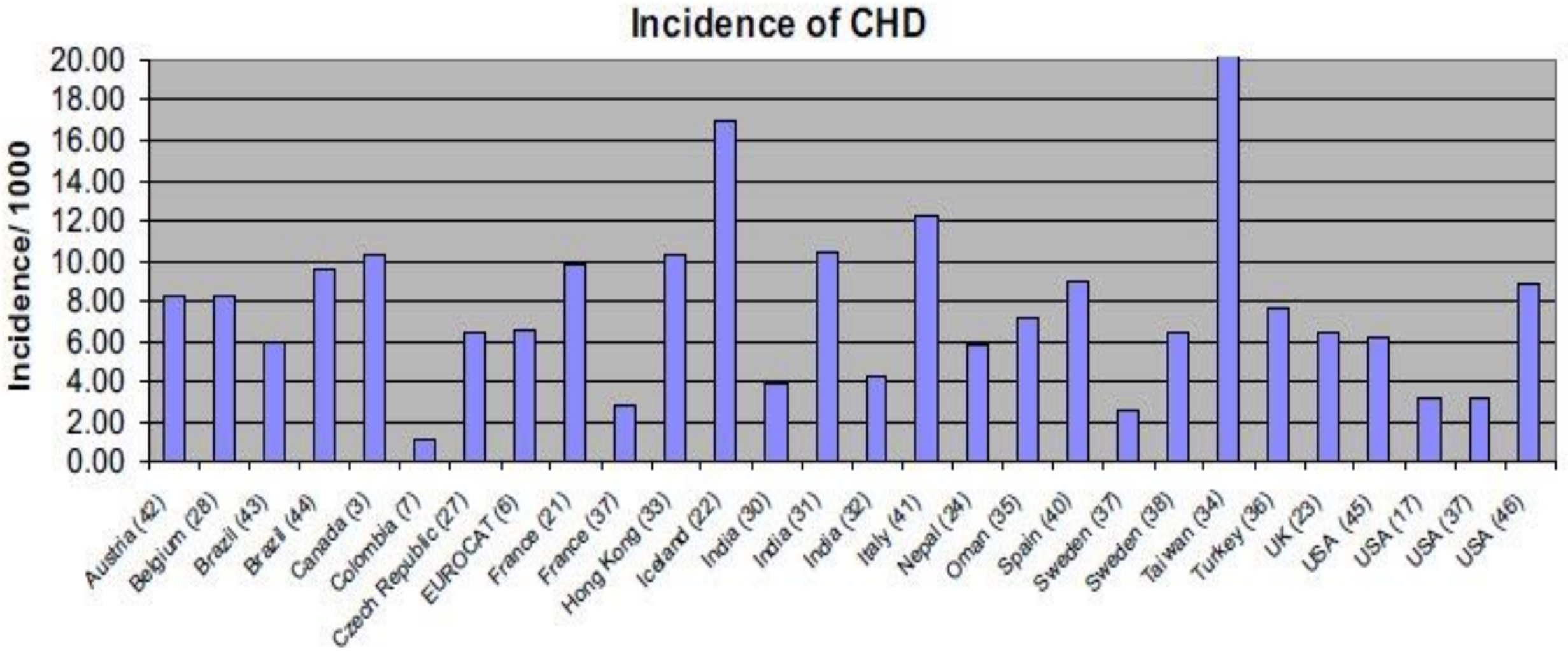
Most reported 8/1000

Included studies < 1985 and > 1985

Most from Western world

Hoffman and Kaplan. JACC 2002;19:1890-1900

Incidence of CHD: only studies > 1985
(during the echocardiography era)



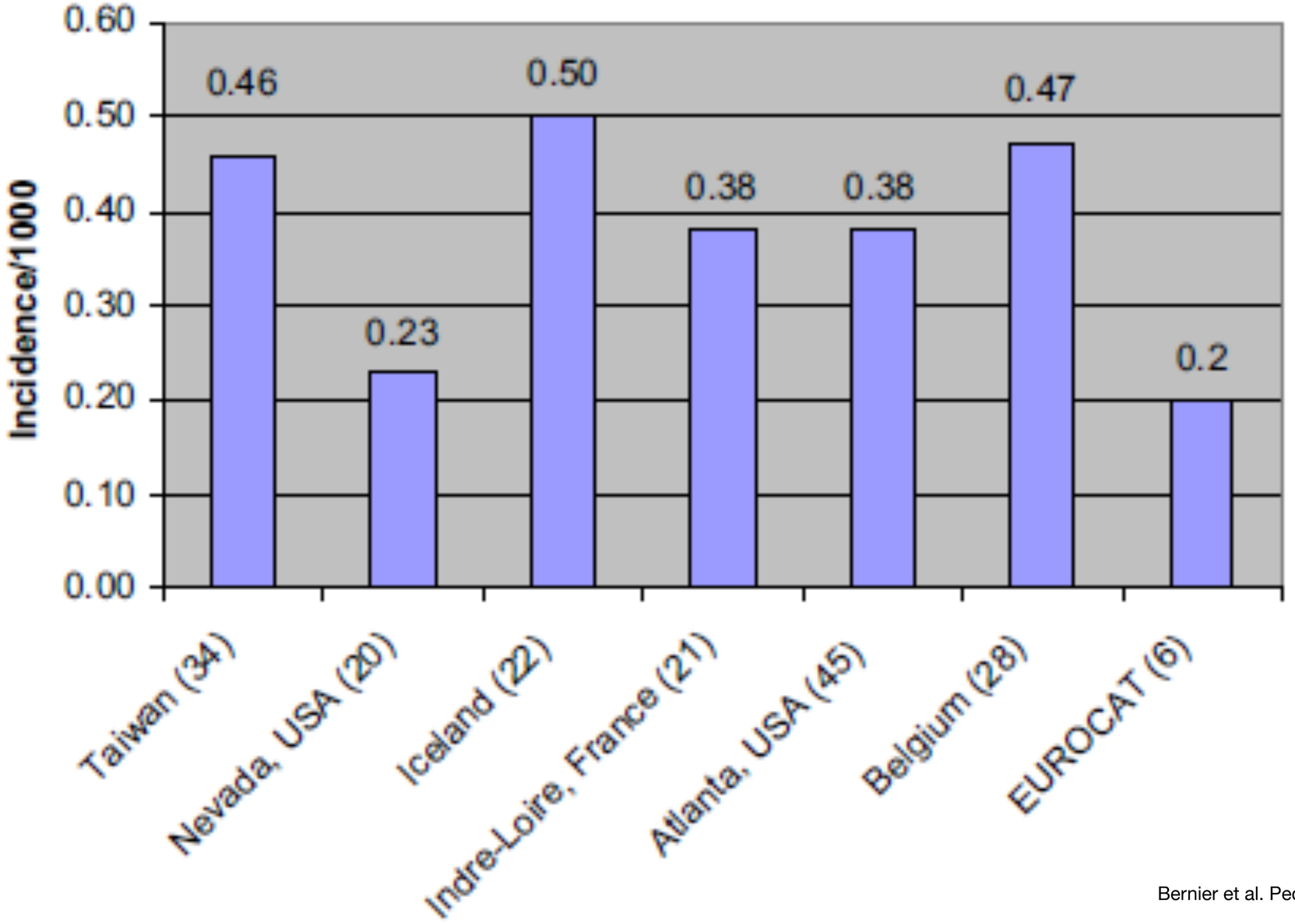
Bernier et al. The Challenge of Congenital Heart Disease Worldwide: Epidemiologic and Demographic Facts. Pediatr Card Surg Ann 2010;13:26-34.

Incidence of VSD: big variation

Country	Sample	Year	Incidence Per 1,000	% of CHD
Egypt (Referrals to a cardiologist)[51]		1995-1996	0.364	36%
India[30]	10,964	1994	1.4	35%
Atlanta, GA, USA[45]	937,195	1995-1997	1.66	30%
Brazil[43]	80,262	1989-1998	1.68	28%
India[32]	11,833		1.93	46%
Oman[35]	139,707	1994-1996	2	28%
Bohemia, Czech Republic[27]	91,823	1980	2.01	31%
Turkey[36]	219,870	1995-2002	2.51	33%
EUROCAT[6]	3,300,300		2.65	33%
Taiwan[34]	45,725	2008	3.13	
Canada[3]	325,000	1999	3.56	34%
Nepal (patients in tertiary hospital)[24]	14,461	2006	4	70%
India (patients admitted to cardiology)[31]	10,641		5.62	54%
France[21]	26,082	1991-1994	6.36	64%
Iceland[22]	44,013	1990-1999	7.68	45%
Saudi Arabia (children referred to hospital)[48]	604	1994-1996		32.5%
Cote d'Ivoire[49]	259	1969-1976		38.6%
Nigeria[50]	260	1965-1970		27%-46%

Bernier et al. Pediatr Card Surg Ann 13:26-34 2010

Tetralogy of Fallot (hard-to-miss diagnosis): big variation



Bernier et al. Pediatr Card Surg Ann 13:26-34 2010

How to explain these differences in birth prevalence of congenital heart diseases ?

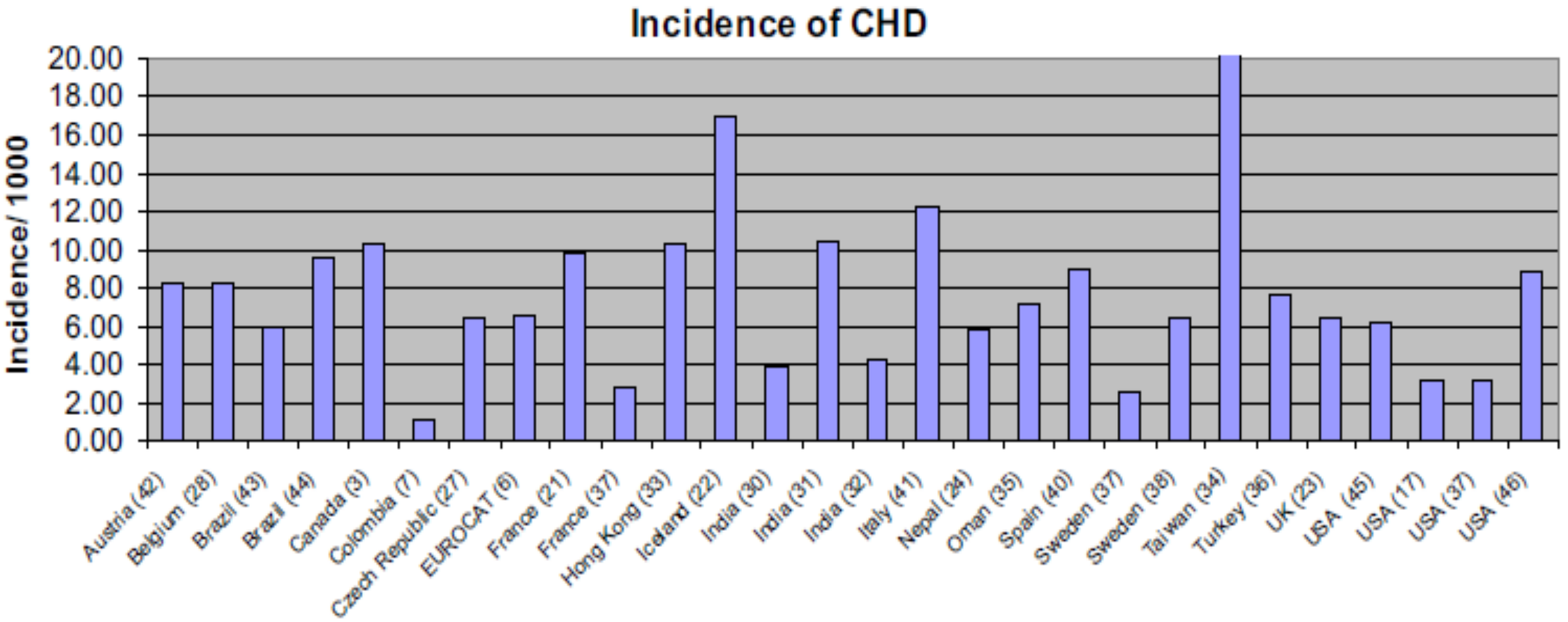
- The differences between studies are substantial

- Explanation of big differences?
  - **Methodological differences?**

    Very likely
  - Ethnic differences?

    Likely

Ethnic differences?



Accessibility of health care, socio-economic standard of a country

Prenatal diagnosis, pregnancy termination, perinatal and early neonatal mortality for congenital heart disease

Paris Registry of Congenital Malformations, 1983-2000

Chromosomal anomalies excluded

	1983-1988 %	1989-1994 %	1995-2000 %	p
Prenatal diagnosis	**23.0**	**31.7**	**47.3**	<0.001
Pregnancy termination	9.9	14.7	15.4	0,037
Still birth	7.0	4.0	3.2	0,028
First day mortality	2.9	1.1	1.1	0,075
First week mortality	10.1	4.9	3.3	<0.001
Perinatal mortality	16.3	8.7	6.4	<0.001

Khosnood B et al. Pediatrics 2005;115:95-101

Prenatal diagnosis, pregnancy termination, perinatal and early neonatal mortality for selected (isolated) congenital heart anomalies

Paris Registry of Congenital Malformations, 1983-2000

HLHS	83-88 %	89-94 %	95-00 %	p
Prenatal diagnosis	**31.8**	**82.8**	**88.9**	<0.001
Pregnancy termination	13.6	72.4	63.0	<0.001
First week mortality	83.3	75.0	50.0	0.12
Perinatal mortality	84.2	75.0	50.0	0.10

TGA	83-88 %	89-94 %	95-00 %	p
Prenatal diagnosis	**12.5**	**48.1**	**72.5**	<0.001
Pregnancy termination	0	7.4	0	0.62
First week mortality	18.8	8.3	2.6	0.04
Perinatal mortality	23.5	12.0	5.0	0.02

Khosnood B et al. Pediatrics 2005;115:95-101

Prenatal diagnosis, pregnancy termination, perinatal and early neonatal mortality for selected (isolated) congenital heart anomalies

Paris Registry of Congenital Malformations, 1983-2000

ToF	83-88 %	89-94 %	95-00 %	p
Prenatal diagnosis	20.0	37.5	69.7	<0.005
Pregnancy termination	10.0	12.5	0	0.07
First week mortality	0	0	0	-
Perinatal mortality	0	7.1	2.9	0.63

Coarctation of the aorta	83-88 %	89-94 %	95-00 %	p
Prenatal diagnosis	0	33.3	42.4	0.03
Pregnancy termination	0	0	0	-
First week mortality	0	0	0	-
Perinatal mortality	0	0	0	-

Khosnood B et al. Pediatrics 2005;115:95-101

The EPICARD study

EPIdémiologie des enfants ou fœtus ayant une **CARD**iopathie congénitale

Methods

Population based study
Recruitment over a period of 3 years (2005-2008)
8 years follow-up
Population in Paris area (n = 317 538)
Patients : children (or fœtuses) with CHD diagnosed prenatally, at birth or during the first year of life (n = 2 867)
Evaluation of cardiac, respiratory and neurodevelopment outcomes

Inclusion criteria

Live-birth with diagnosis of CHD
Confirmation of CHD by echocardiography
TOP / Still-birth with confirmed CHD in Paris area

Data collected

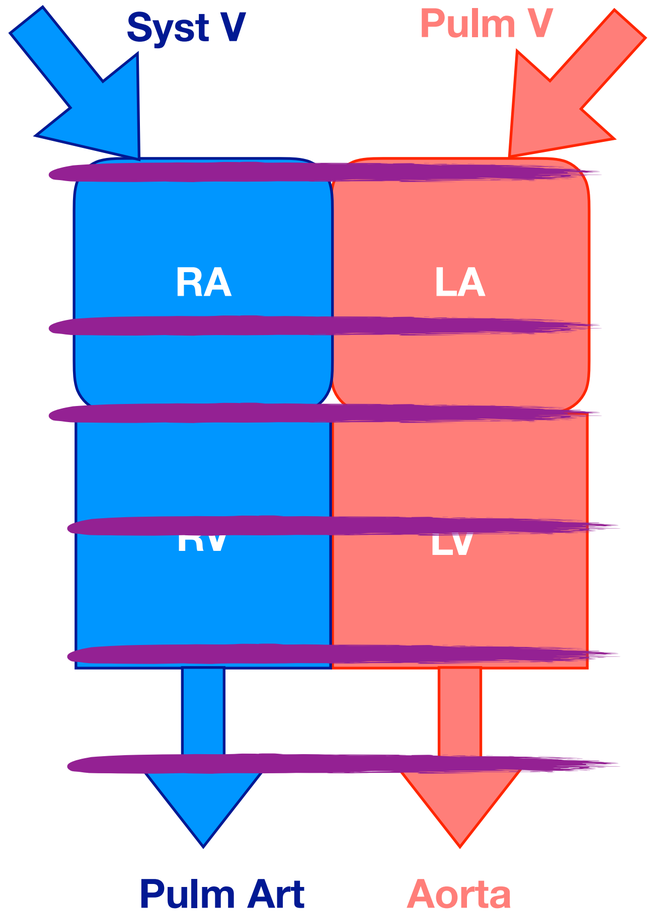
Type of CHD
Extracardiac malformations/ chromosomal anomalies / syndromes
Coding of CHD by two pediatric cardiologists
Confirmation of diagnosis of TOP/still-birth by foetopathologists
Perinatal management/surgical procedures/interventional procedures/

The EPICARD study

EPIdémiologie des enfants ou fœtus ayant une **CARD**iopathie congénitale

Anatomic and Clinical Classification of Congenital Heart Diseases ACC-CHD

ACC-CHD categories	Examples
Heterotaxy	Heterotaxy syndromes
Anomalies of venous connections	Total anomalous pulmonary venous return
Anomalies of atria	Atrial septal defect
Anomalies of AV junction and AV valves	Atrioventricular septal defect
Complex anomalies of AV junction	Double discordance
Functionally univentricular heart	Hypoplastic left heart syndrome
Ventricular septal defects	Perimembranous VSD
Anomalies of ventriculo-arterial connections	Transposition of the great arteries, DORV
Anomalies of extra pericardial trunks	Coarctation of the aorta
Congenital anomalies of coronary arteries	ALCAPA



Houyel L, Bonnet D. Orphanet J 2011

Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects: A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Total number of birth = 317 538

Live births = 314 022

Total 2867 cases

- 2349 Live birth 82.0%
- 465 TOP 16.2%
- 53 IUFD 1.8%

N = 2867

- 1753 (61.1%) Isolated CHD
- 393 (13.7%) Chromosomal anomalies
- 409 (14.3%) Extracardiac anomalies

Distribution of categories of CHD and associated anomalies

ACC-CHD categories	Total		Live births	
	% of chromosomal anomalies	% of extra cardiac anomalies	% of chromosomal anomalies	% of extra cardiac anomalies
Heterotaxy	0	24.3	0	25.0
Anomalies of venous connections	19.4	16.1	7.7	15.4
Anomalies of atria	9.9	19.8	7.5	19.0
Anomalies of AV junction and AV valves	57.3	12.7	43.1	13.8
Complex anomalies of AV junction	0	7.7	0	0
Functionally univentricular heart	15.8	19.6	8.3	20.8
Ventricular septal defects	9.3	11.1	3.9	11.0
Anomalies of ventriculo-arterial connections	10.7	18.8	4.5	14.1
Anomalies of extra pericardial trunks	15.9	31.2	3.2	26.4
Congenital anomalies of coronary arteries	0	0	0	0

Khoshnood B et al. Heart 2012;98:1667-73

ACC-CHD categories	Isolated CHD Tertiary center %	Isolated CHD Population based %	
Heterotaxy	55	75.7	
Anomalies of venous connections	74.1	64.5	
Anomalies of atria	60	70.3	
Anomalies of AV junction and AV valves	57	30	AVSD with chromosomal anomalies are not referred
Complex anomalies of AV junction	88.5	92.3	
Functionally univentricular heart	88.9	64.6	FUVH are referred when isolated
Ventricular septal defects	52.7	79.6	VSD are referred when non isolated
Anomalies of ventriculo-arterial connections	80.3	70.3	
Anomalies of extra pericardial trunks	70.8	52.9	
Congenital anomalies of coronary arteries	100	100	

Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects

A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Proportion of prenatal diagnosis

All CHDs

ACC-CHD categories	% of prenatal diagnosis
All cases excluding chromosomal anomalies	25.6
All cases excluding chromosomal and other extra cardiac anomalies	23
All cases excluding chromosomal, other anomalies and simple VSD	40.2

In categories of CHDs

ACC-CHD categories	% of prenatal diagnosis (n)
Heterotaxy	89.2 (37)
Anomalies of venous connections	16.0 (25)
Anomalies of atria	4.3 (164)
Anomalies of AV junction and AV valves	67.0 (91)
Complex anomalies of AV junction	100.0 (13)
Functionally univentricular heart	92.5 (133)
Ventricular septal defects	9.6 (1353)
Anomalies of ventriculo-arterial connections	39.2 (503)
Anomalies of extra pericardial trunks	44.7 (143)
Congenital anomalies of coronary arteries	0 (9)

Specific CHDs

Type of CHD	% of prenatal diagnosis
Congenitally corrected transposition of the great	100
Functionally univentricular heart	92.5
TGA	84
DORV	98

Khoshnood B et al. Heart 2012;98:1667-73

Prevalence, pre- and post-natal diagnosis, and infant mortality of newborns with congenital heart defects:
A population-based study using the International Paediatric and Congenital Cardiac Code (IPCCC)

The EPICARD Study Group

Proportion of Termination of pregnancy

All CHDs : total 16%

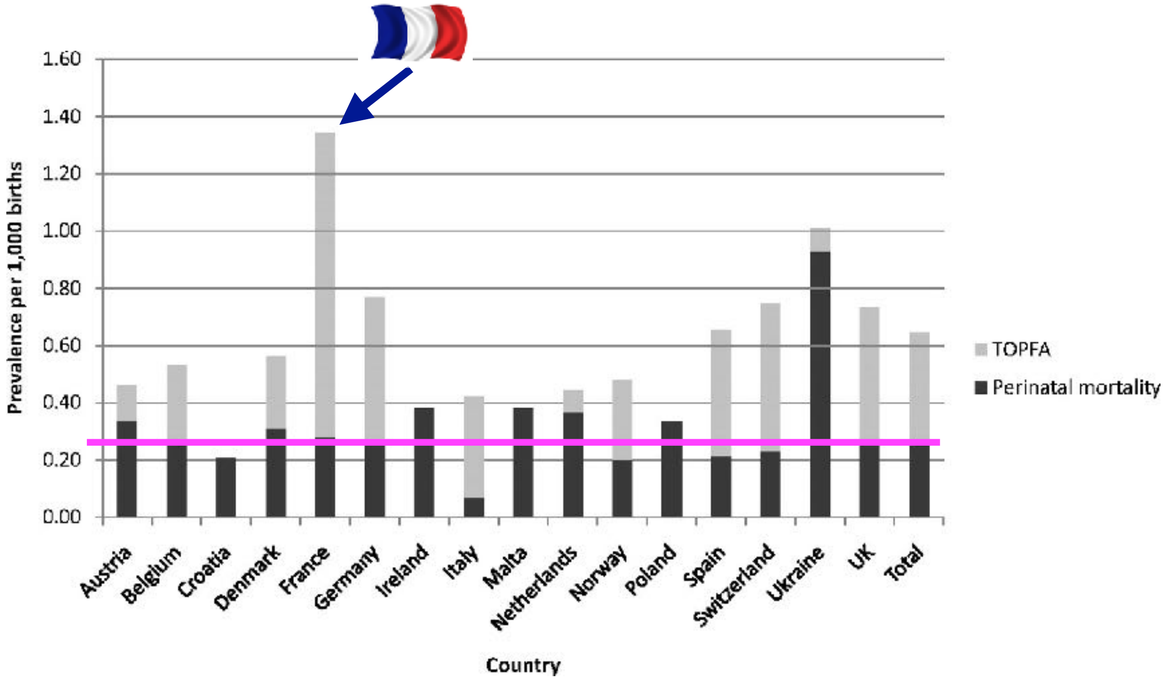
ACC-CHD categories	% TOP
All cases excluding chromosomal anomalies	9.8
All cases excluding chromosomal and other extra cardiac anomalies	6.4
All cases excluding chromosomal, other anomalies and simple VSD	14.0

In categories of CHDs

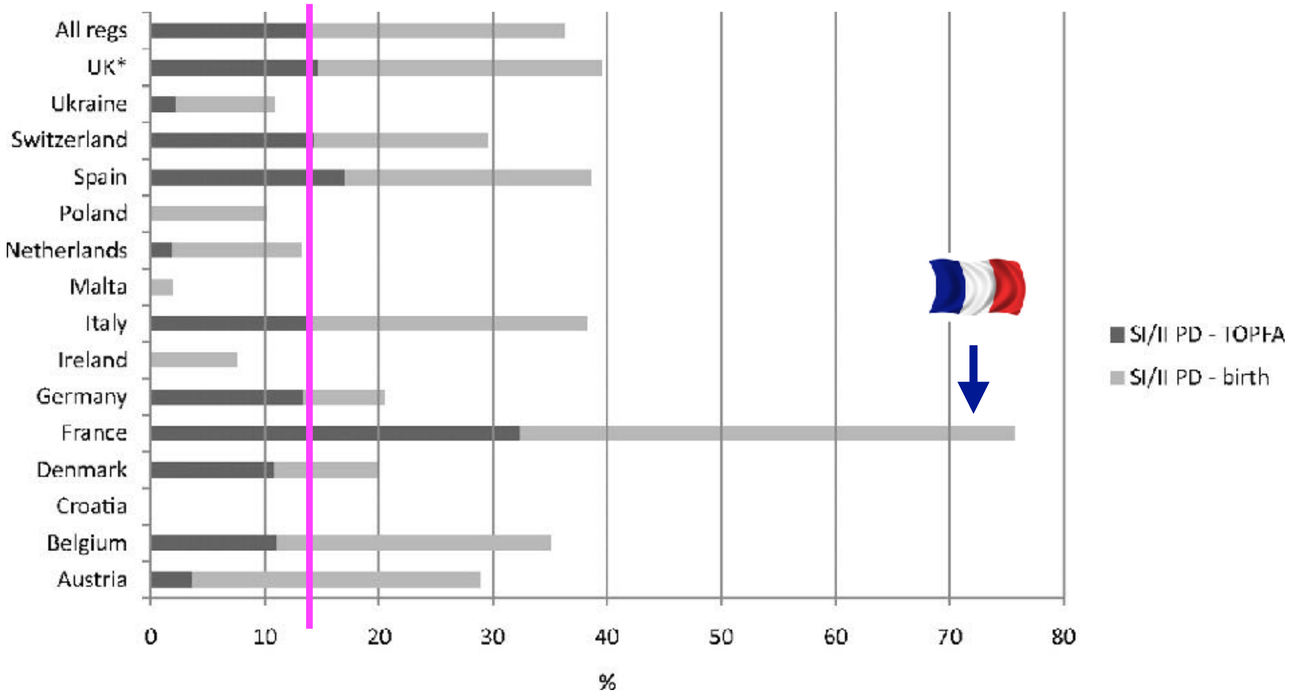
ACC-CHD categories	% TOP
Heterotaxy	75.7
Anomalies of venous connections	16.1
Anomalies of atria	4.4
Anomalies of AV junction and AV valves	42.7
Complex anomalies of AV junction	46.2
Functionally univentricular heart	62.7
Ventricular septal defects	5.7
Anomalies of ventriculo-arterial connections	18.5
Anomalies of extra pericardial trunks	23.5
Congenital anomalies of coronary arteries	0

Khosnood B et al. Heart 2012;98:1667-73

Perinatal mortality and terminations of pregnancy for fetal anomaly (TOPFA) associated with **nonchromosomal** congenital heart defects per 1000 births, by country, 2000 to 2005.



Proportion of nonchromosomal SI/SII congenital heart defect cases prenatally diagnosed (PD)* by pregnancy outcome (terminations of pregnancy for fetal anomaly [TOPFA] or birth), by country, 2000 to 2005.



Severity I (SI): single ventricle, hypoplastic left heart, hypoplastic right heart, Ebstein anomaly, and tricuspid atresia
Severity II (SII): pulmonary valve atresia, common arterial truncus, atrioventricular septal defects, aortic valve atresia/stenosis, transposition of great vessels, tetralogy of Fallot, total anomalous pulmonary venous return, and coarctation of aorta, without additional CHD subgroups classified as severity I.
Severity III (SIII): ventricular septal defect (VSD), atrial septal defect, and pulmonary valve stenosis, without additional CHD subgroups classified as SI or SII.

American Heart Association.
Helen Dolk et al. Circulation. 2011;123:841-849
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Is TOP for CHD more frequently performed in France ?

- TOP is allowed in France in case of CHD not amenable to « repair » and of « a peculiar severity »

- After 14 weeks, TOP is not allowed without a complex process with multidisciplinary approach in a national reference center for prenatal diagnosis (after 22 or 24 weeks in many other European countries)

- TOP for CHD is stable for more than 25 years while the proportion of prenatal diagnosis has increased dramatically

- Finally, the proportion of prenatal diagnosis for severe defects is a crucial issue:

  - if you diagnose 100% of hypoplastic left heart syndromes and 50% of parents chose TOP : perinatal mortality is 50%

  - if you diagnose 10% of hypoplastic left heart syndromes and 100% of parents chose TOP: perinatal mortality of prenatally diagnosed HLHS is 10%

Infant mortality in newborns with congenital heart defects
The EPICARD Study Group

ACC-CHD categories	Prenatal diagnosis N	Postnatal diagnosis <7days %	8-28 days %	29 days-1 year %	Infant mortality %	95%CI
Heterotaxy	8	25.0	0.0	12.5	37.5	8.5-75.5
Anomalies of venous	26	3.9	11.5	11.5	26.9	11.6-47.8
Anomalies of atria	174	0.6	0.6	2.3	3.5	1.3-7.3
Anomalies of AV junction and AV valves	109	8.3	7.3	12.8	28.4	20.2-37.0
Complex anomalies of AV	7	0.0	0.0	14.3	14.3	0.4-57.9
Functionally univentricular	48	41.7	12.5	4.1	58.3	43.2-72.4
Ventricular septal defects	1396	0.2	0.5	0.9	1.6	1.0-2.4
Anomalies of ventriculo-arterial connections	447	2.3	2.0	4.0	8.3	5.9-11.2
Anomalies of extra pericardial trunks	124	3.2	6.5	2.4	12.1	6.9-19.2
Congenital anomalies of coronary arteries	9	0	0	11.1	11.1	0.3-48.2
All	2348	2.1	1.8	2.5	6.4	5.5-7.5
All except chromosomal anomalies and /or anomalies of other systems and IVSD	784	2.9	2.2	3.6	8.7	6.8-10.9

Proportion of preterm births for newborns with CHD (excluding isolated ASD)

Congenital Heart Defect	GESTATIONAL AGE (weeks)									
		<32			32-37			<37		
	N	%	95%CI**	p*	%	95%CI**	p*	%	95%CI**	p*
s	2189	2.4	1.8-3.1	0.008	11.1	9.8-12.5	<0.001	13.5	12.1-15.0	<0.001
vithout chromosomal anomalies	2055	2.3	1.7-3.0	0.02	10.2	8.9-11.6	<0.001	12.5	11.1-14.0	<0.001
vithout chromosomal and/or lies of other systems***	1770	2.1	1.5-2.9	0.04	9.4	8.1-10.8	<0.001	11.5	10.1-13.1	<0.001
vithout chromosomal and/or lies of other systems, excluding l VSD	667	3.9	2.6-5.6	<0.001	13.6	11.1-16.5	<0.001	17.5	14.7-20.6	<0.001
National Perinatal Survey 2003	1815	1.3			5.9			7.2		

Laas E et al. Pediatrics 2013

Proportion of spontaneous and medically-induced preterm birth for newborns with CHD for all live births

Congenital Heart Defect		Spontaneous preterm birth (<37 weeks)		Medically-induced* preterm birth (<37 weeks)	
	n	%	95%CI**	%	95% CI**
es	2189	9.7	8.5-11.0	3.7	2.9-4.6
vithout chromosomal anomalies	2055	8.8	7.6-10.1	3.6	2.8-4.5
vithout chromosomal and/or anomalies of other s***	1770	7.8	6.6-9.2	3.5	2.7-4.5
vithout chromosomal and/or anomalies of other s, excluding isolated VSD	667	12.0	9.6-14.7	5.4	3.8-7.4
National Perinatal Survey of 2003	1815	3.9		3.3	

Laas E et al. Pediatrics 2013

Proportion of preterm birth according to different categories of CHD

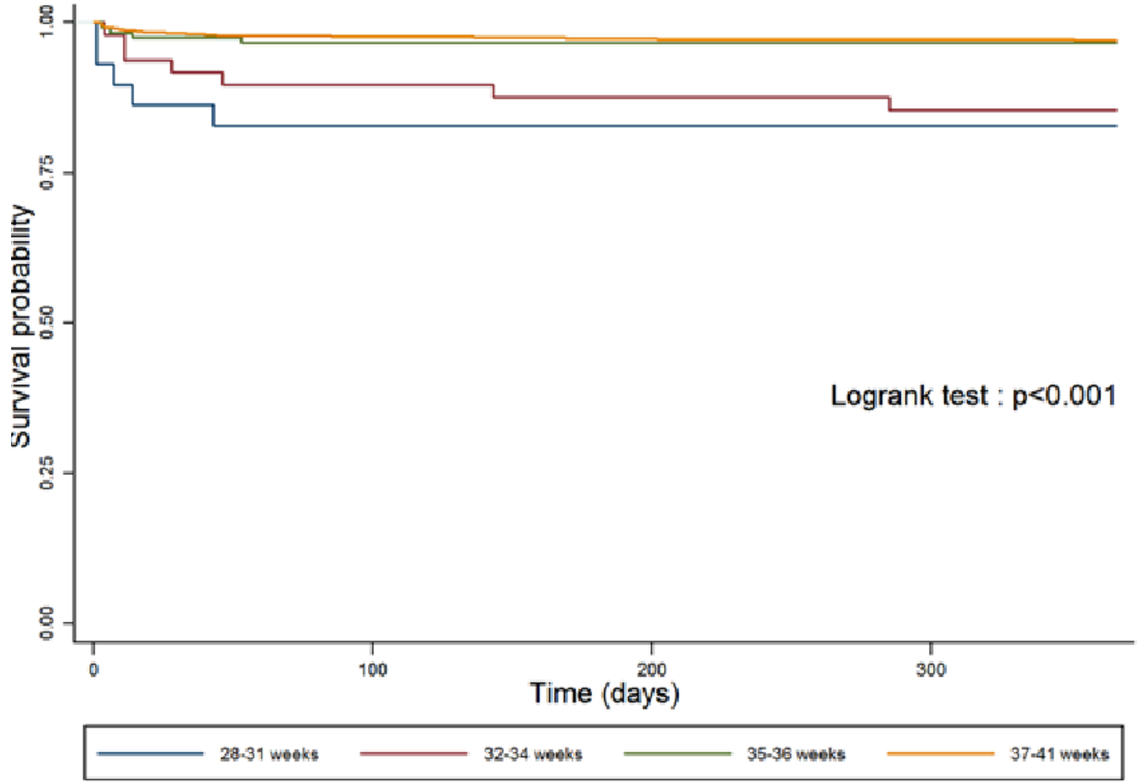
CHD CATEGORIES	GESTATIONAL AGE (weeks)					
	<32		32-37		<37	
						95%CI*
Anomalies of the venous return	0	0-13.2**	3.8	0.1-19.6	3.8	0.1-19.6
Anomalies of the atrioventricular valves	4.6	1.5-10.4	19.2	12.3-27.9	23.8	16.2-33.0
Functionally univentricular hearts	2.1	0.1-11.1	18.7	8.9-32.6	20.8	10.5-35.0
VSD	1.7	1.1-2.5	8.5	7.1-10.0	10.2	8.6-11.9
Anomalies of the ventricular outflow tract	3.8	2.2-6.0	14.8	11.7-18.5	18.6	15.1-22.6
Anomalies of the great arteries	0.8	0,02-4.4	18.5	12.1-26.5	19.3	12.8-27.4
French National Perinatal Survey of 2003	1.3		5.9		7.2	

*95% binomial exact confidence interval

* *97.5% one-sided confidence interval

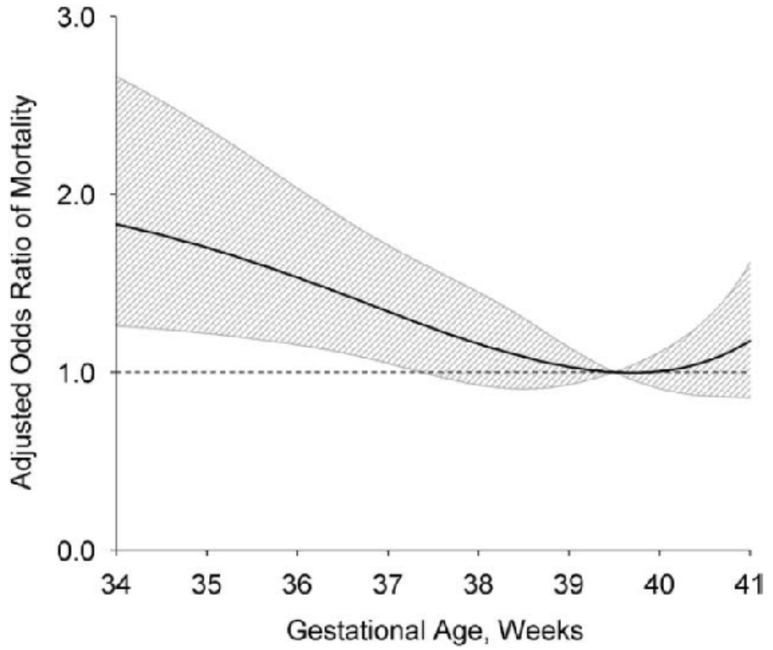
Laas E et al. Pediatrics 2013

Impact of preterm birth on infant mortality for newborns with congenital heart defects
The EPICARD Study Group



- Preterm birth is associated with an approximately **four-fold higher risk** of infant mortality for newborns with CHD.
- This excess risk appears to be mostly limited to newborns **< 35 weeks of gestation** and is disproportionately **due to early deaths**.

Laas E et al. in press



In hospital mortality according to term in CHDs

Gestational Age, wk	No. of Patients	Observed Mortality, % (95% CI*)	Unadjusted OR (95% CI)	*P* Value	Adjusted OR (95% CI)	*P* Value
34	132	15.2 (9.5-22.4)	2.81 (2.11-3.73)	<0.001	1.83 (1.26-2.66)	0.002
35	177	15.3 (10.3-21.4)	2.50 (1.92-3.25)	<0.001	1.70 (1.22-2.37)	0.002
36	357	16.2 (12.6-20.5)	2.15 (1.70-2.71)	<0.001	1.53 (1.15-2.03)	0.003
37	524	13.2 (10.4-16.4)	1.79 (1.44-2.22)	<0.001	1.34 (1.05-1.71)	0.02
38	949	9.0 (7.2-11.0)	1.43 (1.16-1.76)	<0.001	1.16 (0.93-1.45)	0.19
39.5	2321	7.3 (6.3-8.4)	Reference level	-		-
41	156	7.7 (4.0-13.1)	0.99 (0.73-1.36)	0.96	1.18 (0.86-1.62)	0.32

Costello JM et al. Circulation. 2014;12:2511-2517.

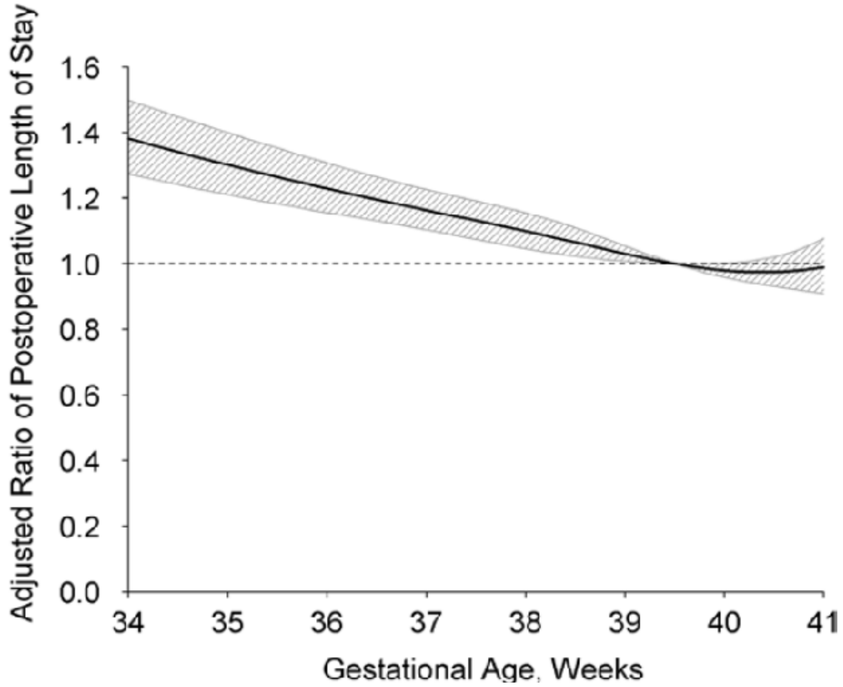
Mortality rates in low birth weight infants

Cardiac surgery for CHD

	Mortality rate, 1–2.5 kg (n = 517)	Mortality rate, 2.5–4 kg (n = 2505)	Risk ratio (95% CI)	*P* value
RACHS-1				
2	5.0 (139)	2.4 (709)	2.1 (0.89–4.97)	.03
3	14.2 (183)	5.6 (840)	2.5 (1.62–3.99)	<.01
4	21.9 (105)	9.7 (462)	2.3 (1.43–3.55)	<.01
6	30.0 (90)	21.1 (494)	1.4 (1.00–2.04)	.03
Aristotle				
1	80.0 (5)	26.7 (15)	3.0 (1.16–7.73)	.32
2	11.7 (222)	5.5 (876)	2.1 (1.36–3.37)	<.01
3	13.6 (88)	5.2 (544)	2.7 (1.40–5.01)	<.01
4	20.78 (178)	13.6 (965)	1.5 (1.10–2.13)	<.01
Risk level missing	16.7 (24)	1.9 (105)	8.75 (1.7–45.04)	.08

Curzon CL et al. JTCS 2008

Duration of hospital stay according to term in CHDs



Gestational Age, wk	Observed Postoperative LOS, d*	Unadjusted Extra Days (95% CI)†	Unadjusted Ratio (95% CI)	*P* Value	Adjusted Extra Days (95% CI)†	Adjusted ratio (95% CI)	*P* Value
34.0	19.0 (10.0-42.5)	7.7 (5.7-9.8)	1.48 (1.36-1.61)	<0.001	6.1 (4.4-8.0)	1.38 (1.27-1.50)	<0.001
35.0	24.0 (12.0-44.0)	6.2 (4.6-8.0)	1.39 (1.29-1.50)	<0.001	4.8 (3.4-6.4)	1.30 (1.21-1.40)	<0.001
36.0	18.0 (10.0-37.0)	5.0 (3.7-6.4)	1.31 (1.23-1.40)	<0.001	3.7 (2.5-4.9)	1.23 (1.16-1.31)	<0.001
37.0	18.5 (10.0-35.0)	3.9 (2.8-5.1)	1.24 (1.18-1.32)	<0.001	2.6 (1.7-3.6)	1.16 (1.10-1.23)	<0.001
38.0	18.0 (9.0-35.0)	2.7 (1.7-3.7)	1.17 (1.11-1.23)	<0.001	1.6 (0.7-2.5)	1.10 (1.05-1.15)	<0.001
39.5	15.0 (8.0-29.0)	Reference level	-	-	-	-	-
41.0	13.5 (8.0-25.3)	–0.9 (–2.3 to –0.5)	0.94 (0.86-1.03)	0.20	–0.2 (–1.5 to –1.2)	0.99 (0.91-1.08)	0.80

Unadjusted values were estimated by models that included a center variable but did not include other patient level risk factors. CI indicates confidence interval; and LOS, length of stay.

*Data were reported as median (interquartile range).

†Unadjusted extra days were calculated by multiplying the population median postoperative length of stay (16 days) by the ratio of medians – 1.

Costello JM et al. Circulation. 2014;12:2511-2517.

Risk of congenital heart defects associated with assisted reproductive technologies: a population-based evaluation

Table 2
Number of cases and controls and proportions of children/foetuses conceived after assisted reproductive technologies

Category[a]	Subjects with complete data on ART (*n*)	Exposure to ART (%)	*P*-value[b]
Controls	3847	3.6	
All CHD	5493	4.7	0.008
CHD without chromosomal abnormalities	4459	4.9	0.003
CHD without chromosomal abnormalities and excluding VSD	3104	5.0	0.005
Malformations of the outflow tracts and ventriculoarterial connections	1088	5.6	0.003
Malformations of the atrioventricular valves and atrioventricular connections	608	2.6	0.231
Functionally univentricular CHD	402	2.5	0.253
Anomalies of the great arteries	371	5.6	0.331
Ventricular septal defects	2248	5.0	0.006
Anomalies of the atria and interatrial communications	124	4.8	0.463
TGA, heterotaxy syndrome, and discordant atrioventricular connections	475	1.3	0.362
Cardiac neural crest defects and double outlet right ventricle without ventricular hypoplasia	537	1.8	0.014

[a]Subcategories of CHD with less than 100 cases are not shown.

[b]Comparison of the proportion of children/foetuses conceived after ART between the subcategory of CHD and the controls.

Assisted Reproductive Technologies, and particularly Intracytoplasmic Sperm Injection, are specifically associated with a higher risk of Tetralogy of Fallot.

Tararbit et al. 2012

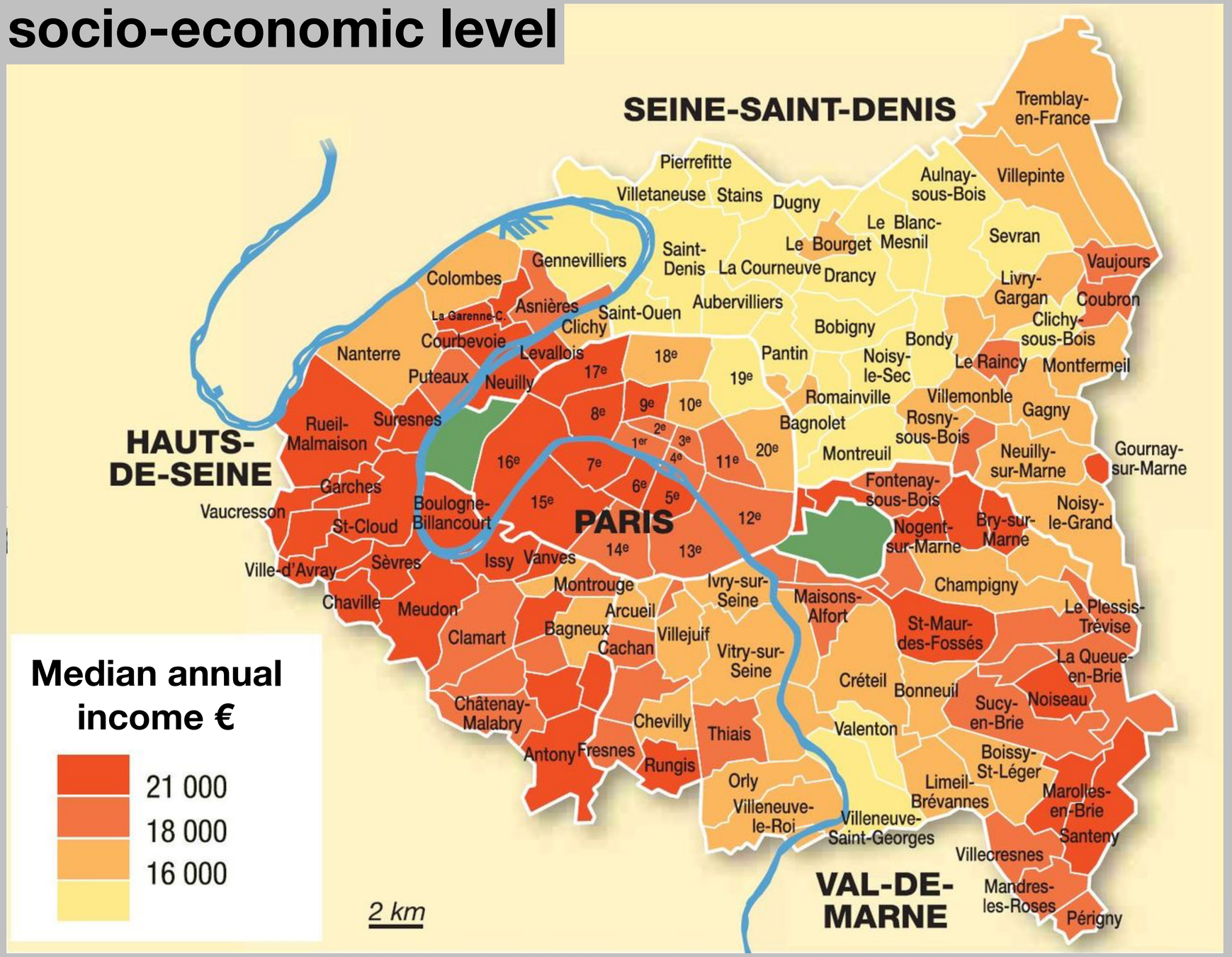
Table 1. Numbers of cases and controls and proportions of fetuses conceived after Assisted Reproductive Technologies (ART).

		N	% exposed to ART	p†
Controls *		4 009	3.5	
All cases	Tetralogy of Fallot	380	6.8	0.001
	Coarctation of the aorta	356	3.7	0.895
	Transposition of the great arteries	420	2.9	0.481
	Hypoplastic left heart syndrome	346	3.2	0.742
	Heterotaxy syndrome	115	4.4	0.635
Cases without chromosomal abnormalities	Tetralogy of Fallot	301	7.6	<0.001
	Coarctation of the aorta	323	4.0	0.635
	Transposition of the great arteries	406	3.0	0.556
	Hypoplastic left heart syndrome	295	3.1	0.673
	Heterotaxy syndrome	114	4.4	0.621

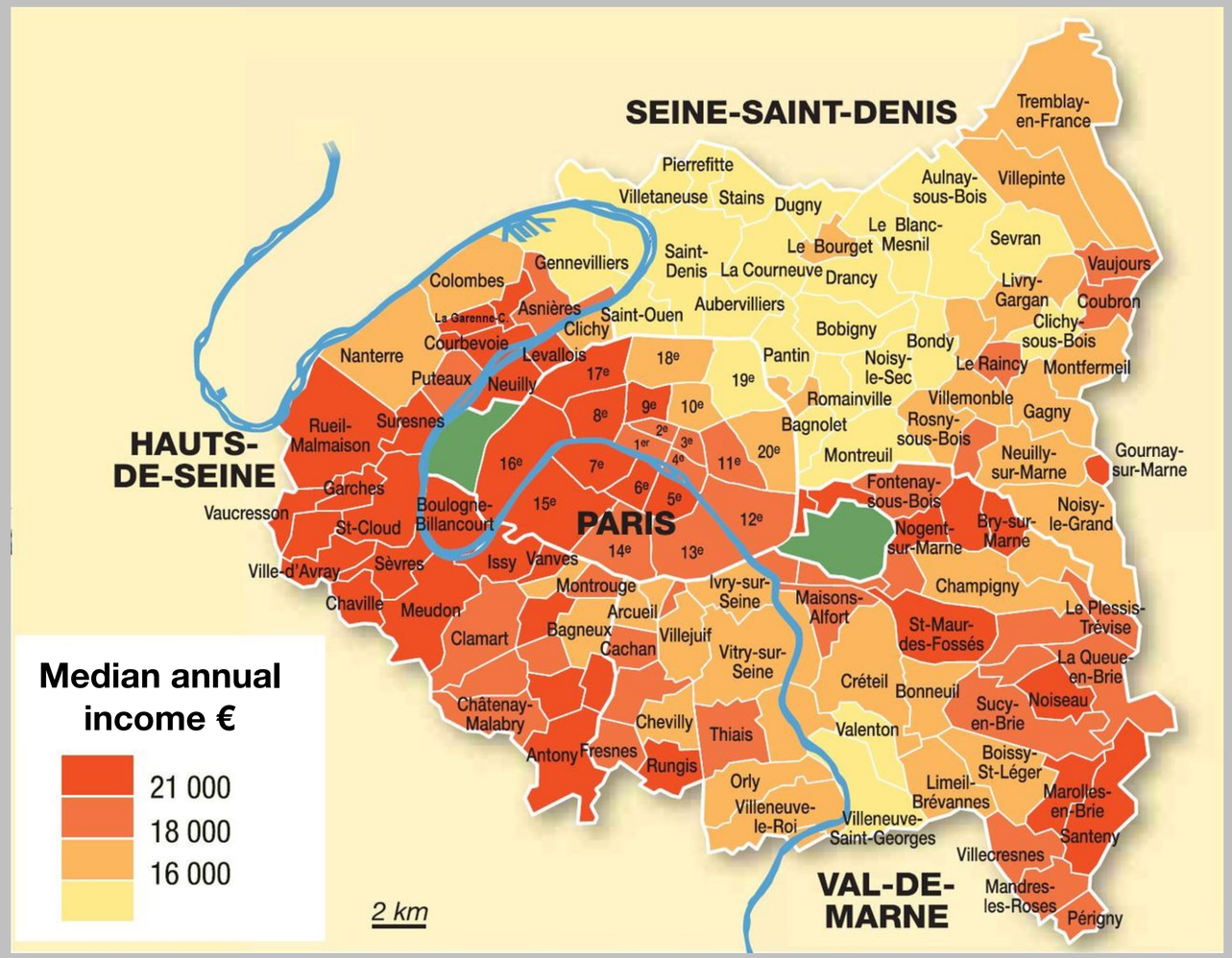
* The following malformations were used as controls: club-foot, angioma, skin abnormality, polydactyly, syndactyly and congenital hip dislocation.

† Comparison of the proportion of children/fetuses conceived after ART between each specific CHD and the malformed controls.

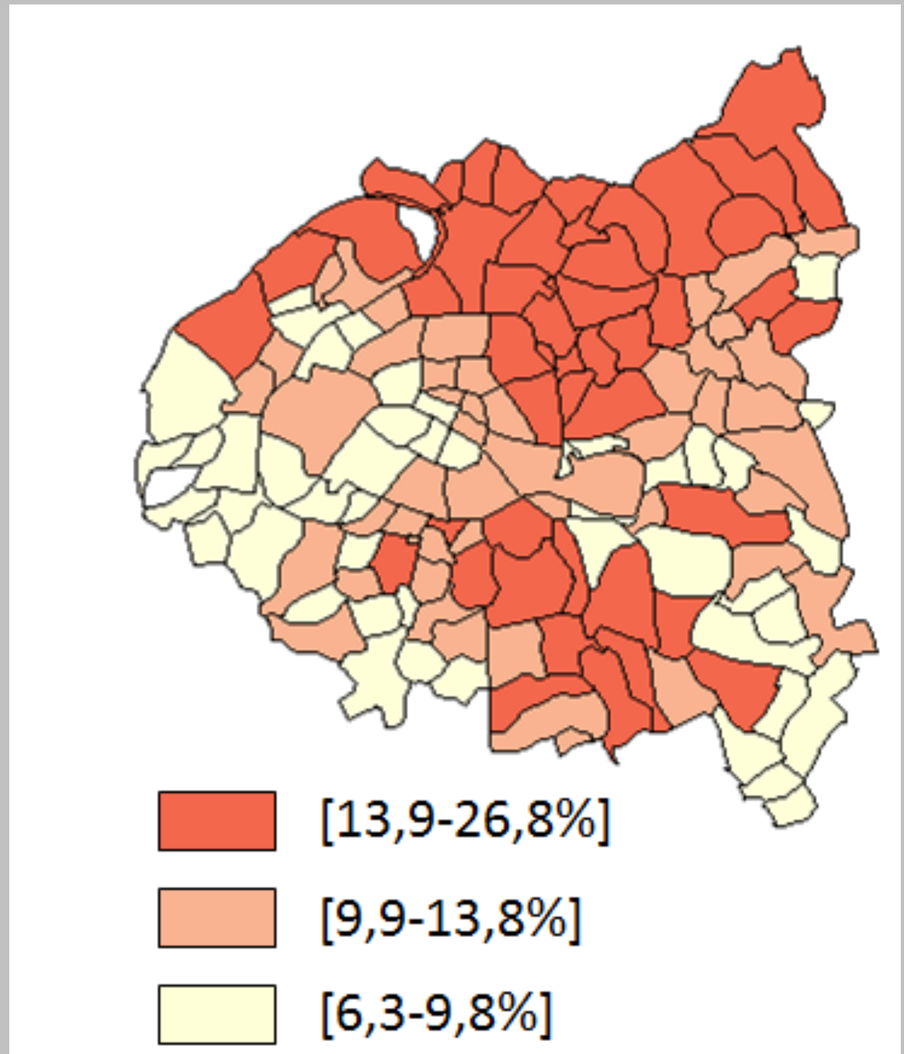
The role of socio-economic level



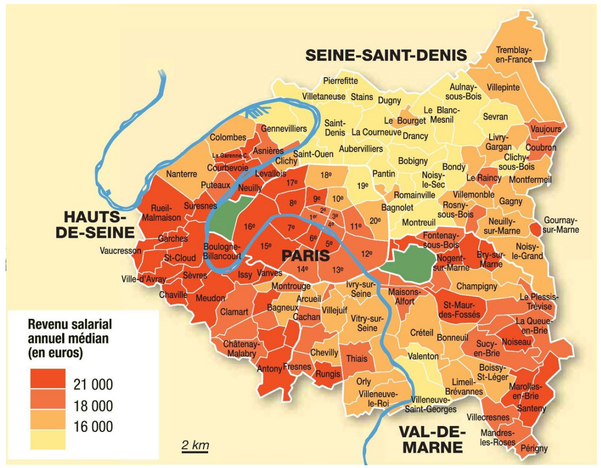
Annual income



Proportion of unemployed

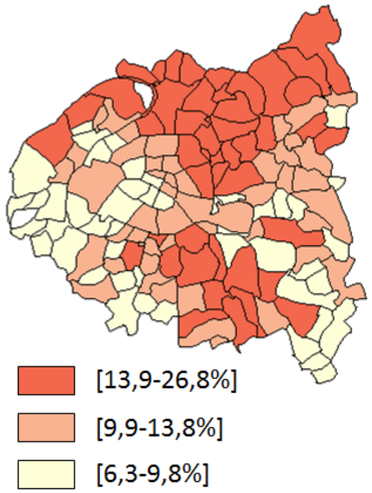


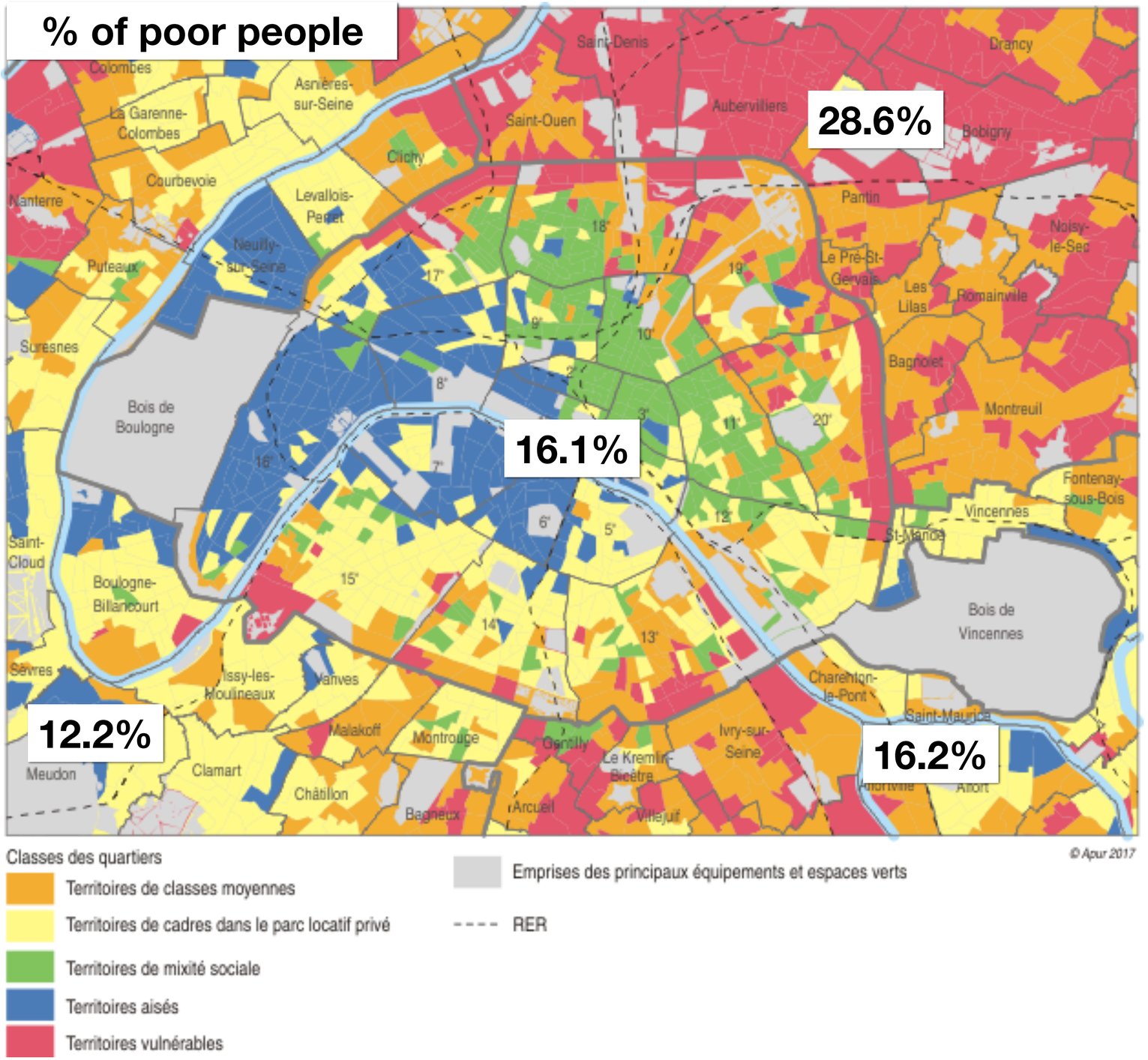
Unemployed % of active population



Vulnerable areas

Schools, security, housing…





Socioeconomic disparities in healthcare are not inevitable
The EPICARD Study Group

Differences in prenatal diagnosis by maternal occupation, geographic origin and place of residence

The proportion of PND of CHD were similar across categories

- The health system organization allowed high availability of reimbursed specialized services that can provide similar access to PND for all socioeconomic groups

	All CHD				
	n 2867	% 29.1	Adj-OR	95%CI	p
Geographic origin					0.62
France	1370	27.7	Ref	Ref	
North African	526	28.1	1.0	0.8-1.3	
African	393	33.6	1.3	1.0-1.7	
Other	562	30.6	1.1	0.9-1.4	
Occupation					0.39
Professional	2139	68.9	Ref	Ref	
None	728	31.6	1.3	0.8-1.5	
Department of residence					0.31
Paris	972	28.8	Ref	Ref	
Hauts de Seine	702	29.5	1.1	0.8-1.4	
Val de Marne	509	26.3	0.7	0.5-1.0	
Seine-Saint Denis	684	25.6	0.9	0.7-1.2	

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Khosnood B et al. 2017

Socioeconomic disparities in healthcare are not inevitable
The EPICARD Study Group

The association between TOP and maternal characteristics in fetuses with prenatal diagnosis of CHD are related to maternal geographic origin

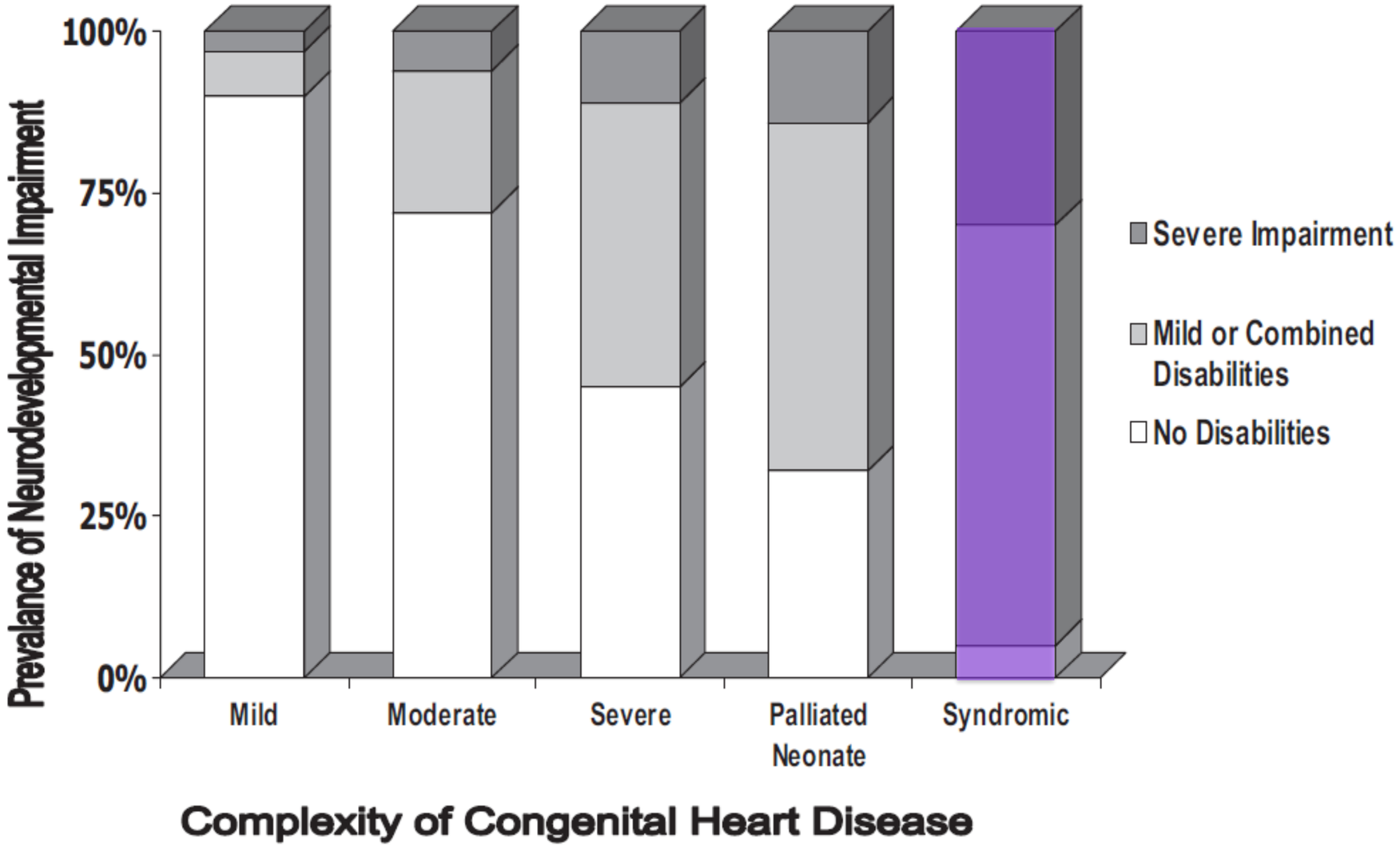
	All CHD				
	n 835	% 41.4	Adj-OR	95%CI	p
Geographic origin					<0.0004
France	380	46.1	Ref	Ref	
North African	148	26.4	0.4	0.2-0.6	
African	132	34.9	0.6	0.3-1.0	
Other	172	48.8	1.1	0.7-1.8	
Occupation					0.09
Professional	165	43.0	Ref	Ref	
None	230	30.4	0.8	0.4-1.3	
Department of residence					0.25
Paris	280	42.1	Ref	Ref	
Seine-Saint Denis	214	38.8	0.9	0.6-1.5	

Khosnood B et al. 2017

How socioeconomic differences in prenatal decision for TOP may influence outcomes ?

- The probability of TOPFA may represent women's preferences that should of course be respected.
- These differences in TOPFA can result in disparities in the spectrum of severity of CHD at birth and thereby, all else equal, in the risk of mortality, morbidity and long-term adverse developmental outcomes for newborns with CHD.
- In addition, families with fewer resources may become disproportionately responsible for the care of newborns with more severe types of CHD.
- The extent to which post-natal management can modify any such disparities needs to be examined.

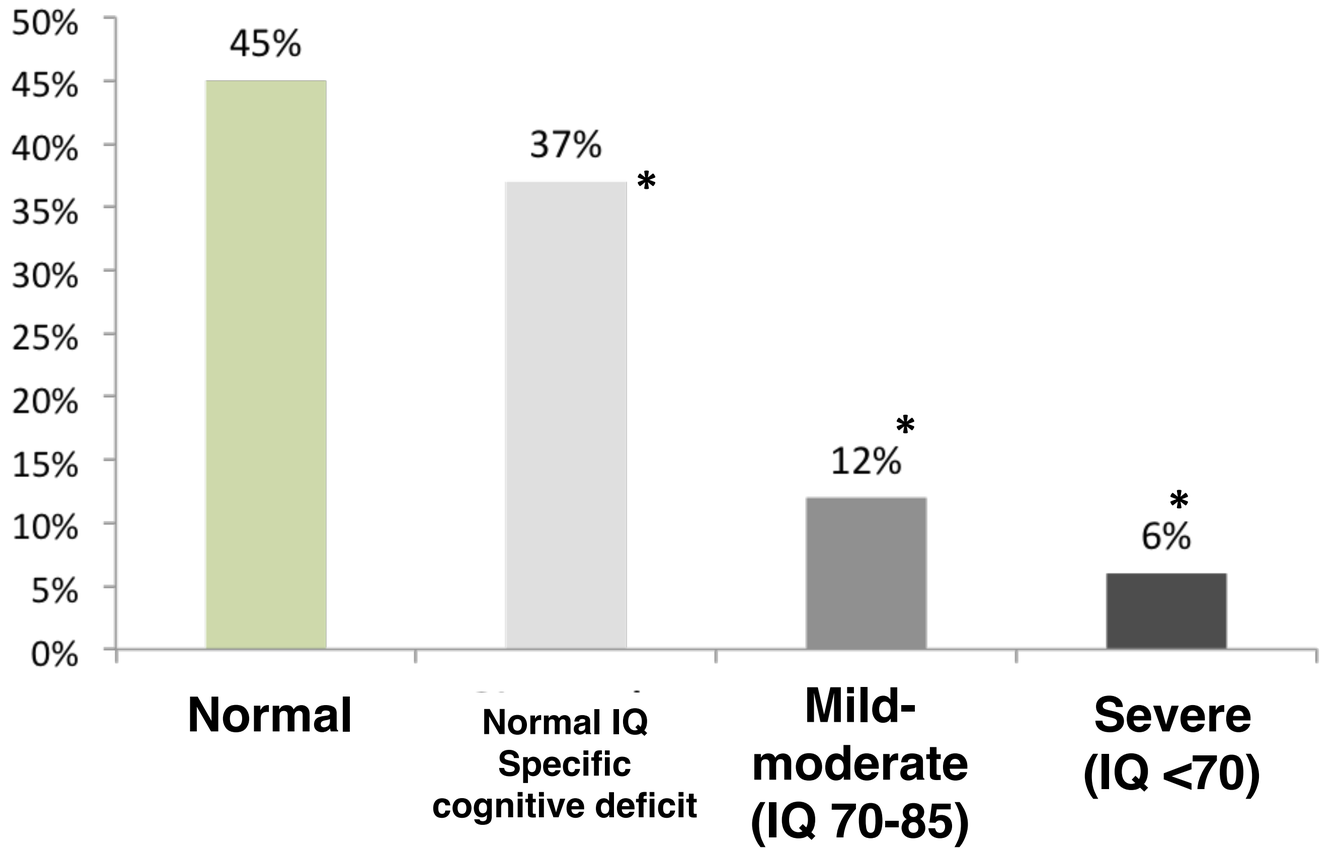
Prevalence of neurodevelopmental anomalies in CHD (0-12 years)



Wernovsky, 2006; Marino et al., 2012

Neurodevelopmental outcomes in CHDs
The EPICARD study group

- K-ABC II at **3 years**
- **415 patients with isolated CHD**



Risk factors:

➢**Complex CHD** and/or
➢Surgery before 1 year
➢GA <39 weeks
➢Socioeconomic factors

* p<0.05

Calderon et al., EPICARD Study Group 2017 Arch Dis Child

Risk factors associated with cognitive outcomes (K-ABC mean global score) in children with CHD

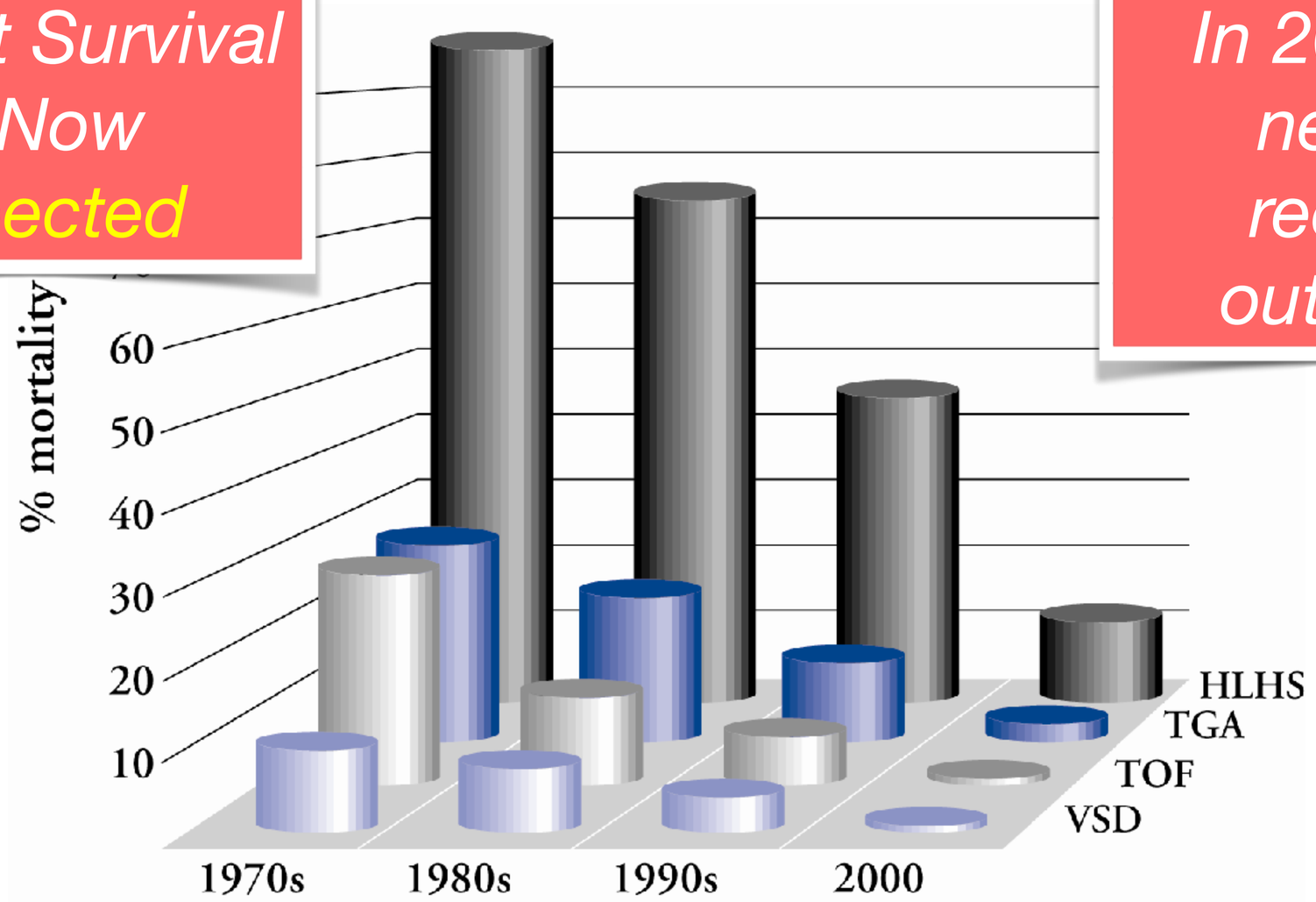
		Unadjusted		Adjusted	
		β	IC 95%	β	IC 95%
Gender	*Female*	ref	–	ref	–
	Male	2.5	[- 3.3 – 8.3]	1.8	[- 3.8 – 7.5]
Maternal education	*Low*	**-12.9**	**[- 20.6 – - 5.2]**	**-14.3**	**[- 22.1 – - 6.4]**
	Intermediate	-5.9	[- 12.0 – 0.2]	**-6.6**	**[- 12.7 – - 0.5]**
	High	ref	–	ref	–
SGA	*No*	ref	–	ref	–
	Yes	**-10.4**	[- 18.4 – - 2.5]	**-14.2**	**[- 22.3 – - 6.2]**
Prematurity	*<37*	**-11.3**	[- 20.7 – - 1.8]	-4.3	[- 14.0 – 5.5]
	≥37	ref	–	ref	–
Complexity of the CHD	Simple	ref	–	ref	–
	Moderate/	-0.5	[- 7.6 – 6.6]	0.3	[- 6.9 – 7.4]
Duration of first ICU stay (d)	–	0.0	[- 0.5 – 0.4]		

Calderon et al. Arch Dis Child 2017

Improving Surgical Results

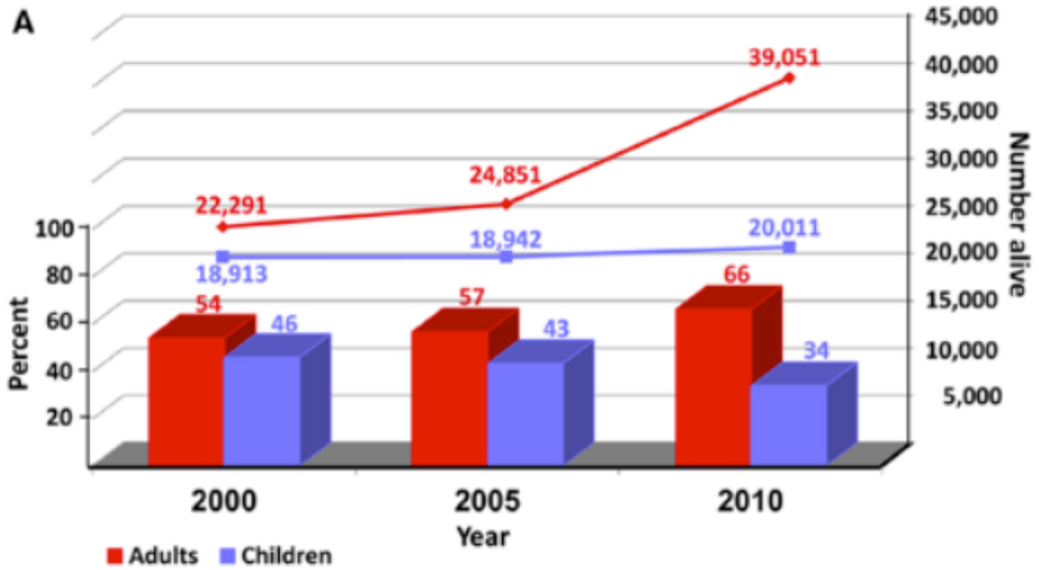
Patient Survival is Now Expected

In 2017, we need to redefine outcomes

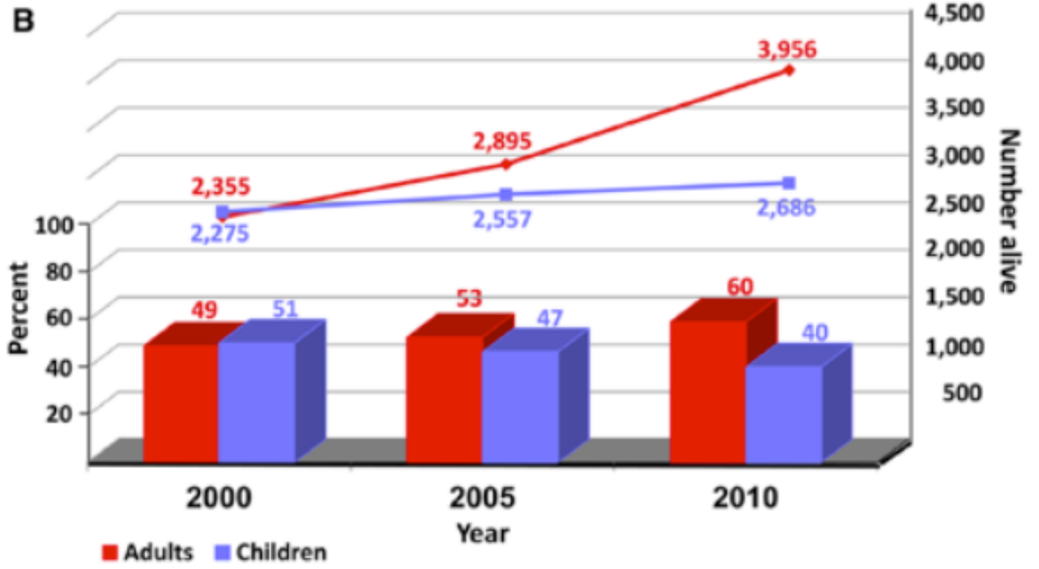


Evolution of mortality in CHD

Increased survival and increased complexity

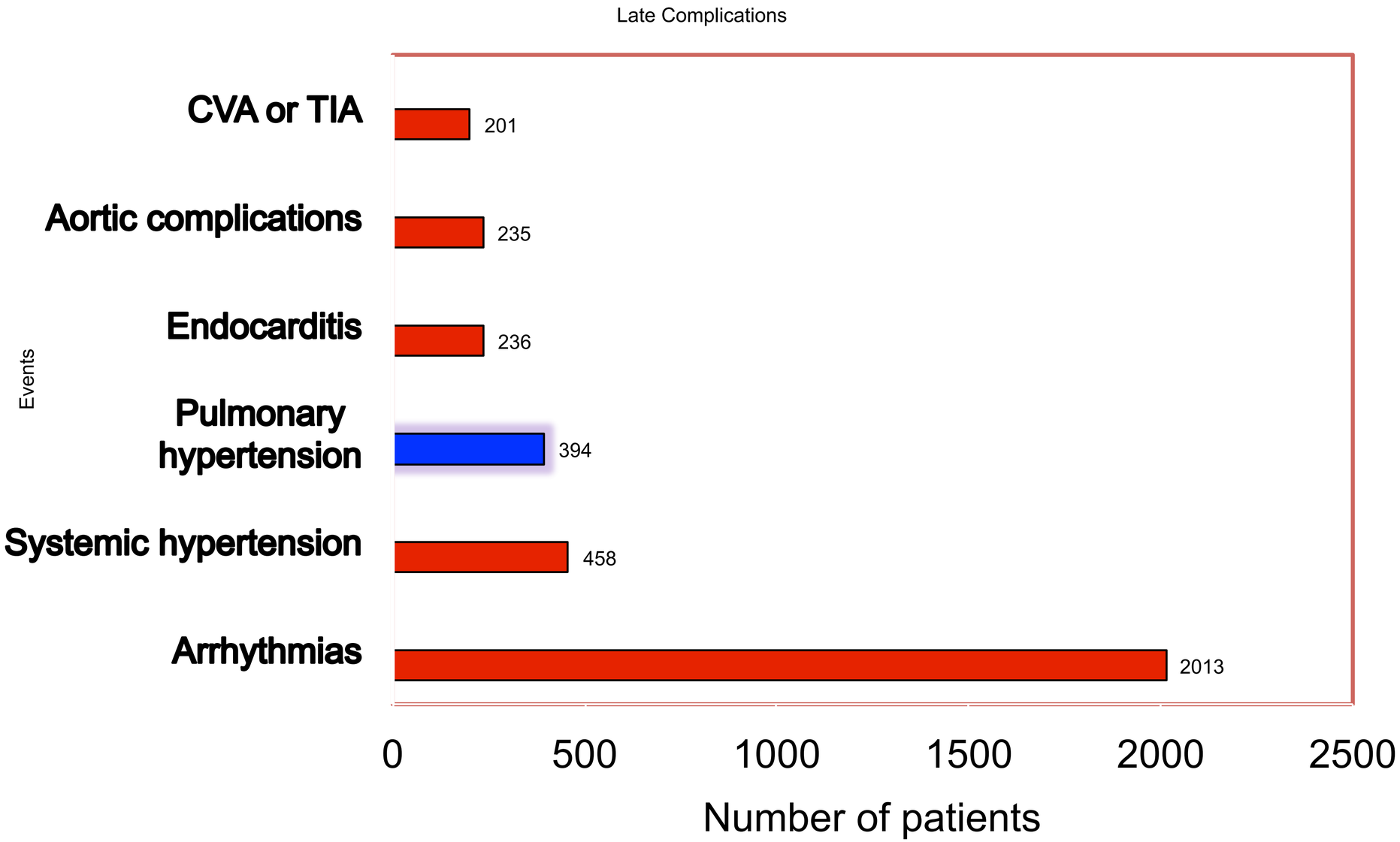


Simple CHD

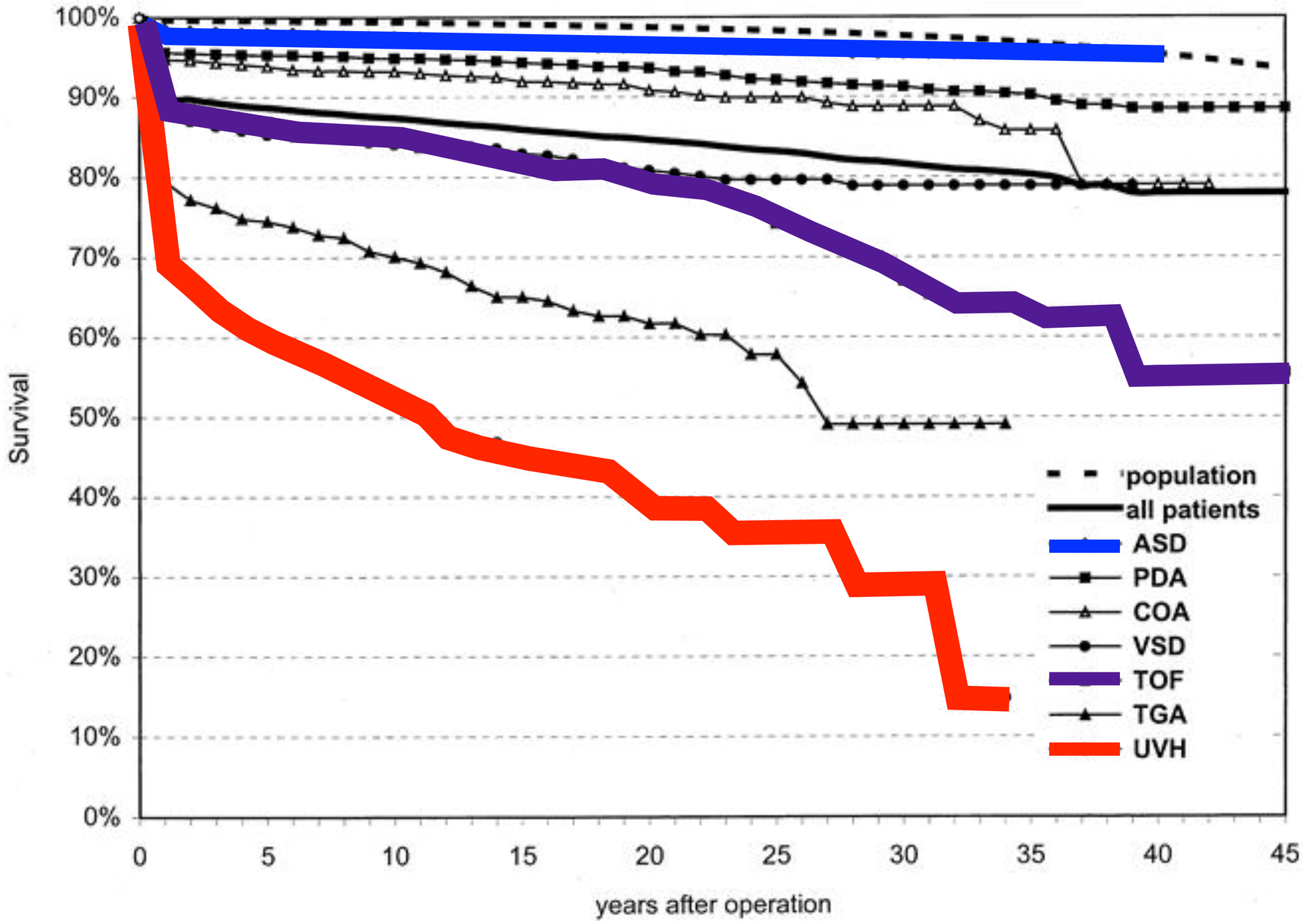


Complex CHD

What are the complications faced by GUCHD patients ?

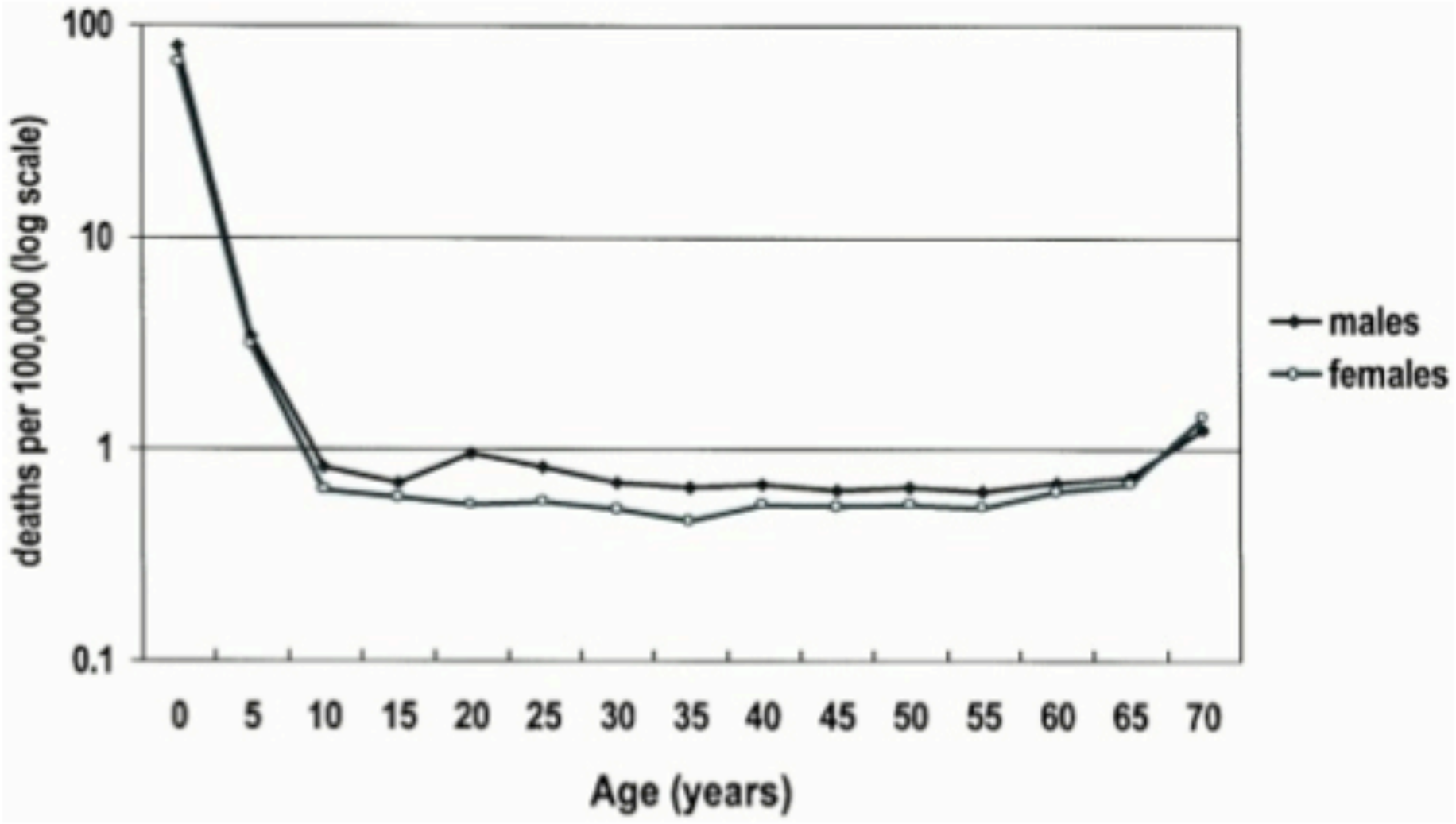


Mortality in GUCHD according to the type of defect



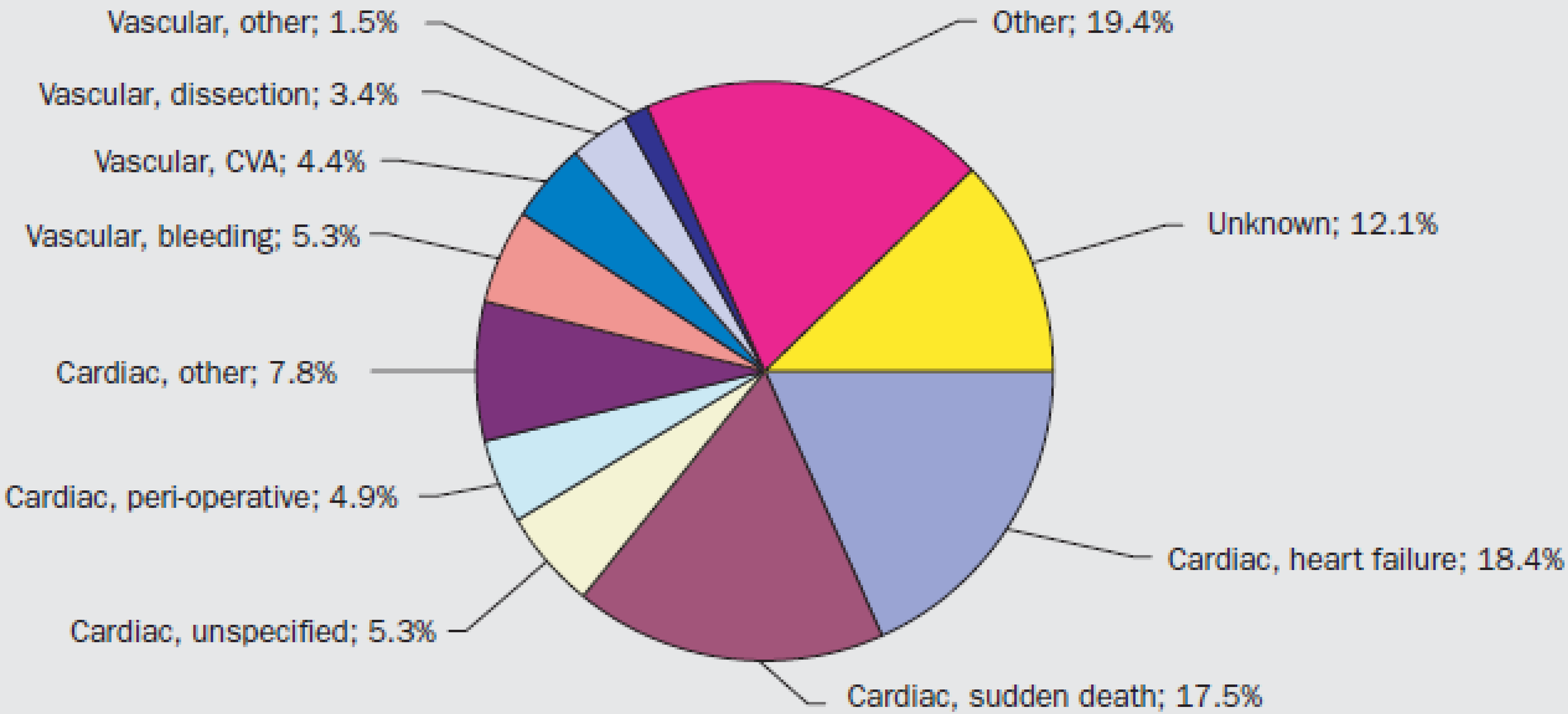
Nieminen et al, Circulation 2001

Death rate due to CHD per 100.000 population by age and sex (USA 1977:1997)



Boneva, R. S. et al. Circulation 2001;103:2376-2381

Causes of death in adult CHD



Epidémiologie des cardiopathies congénitales

1 naissance / 100

6 à 8000 nouveau-nés par an

Plus de 90% de survie à l'âge adulte

Plus de 300.000 personnes ayant une cardiopathie congénitale vivantes en France

Emergences du XXIème siècle

- Nouvelle population de cardiopathies complexes
- Nouvelles morbidités
- Croissance de la consommation de soins
- Questions sociales et sociétales