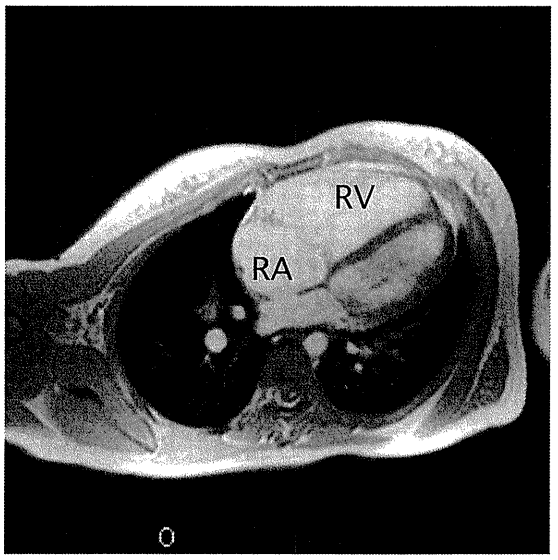
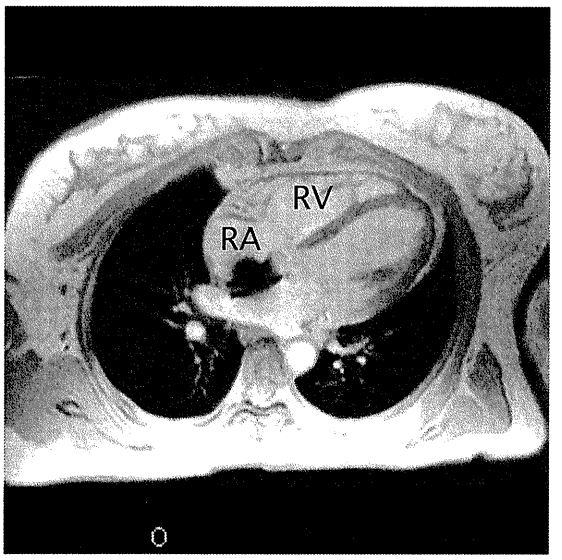
心房中隔欠損、デバイス閉鎖前後

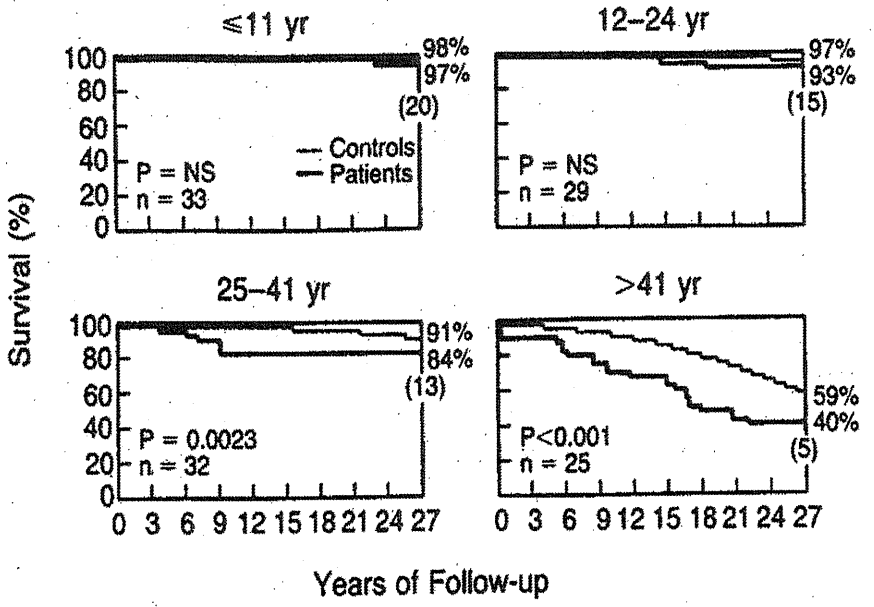


右室、右房拡大



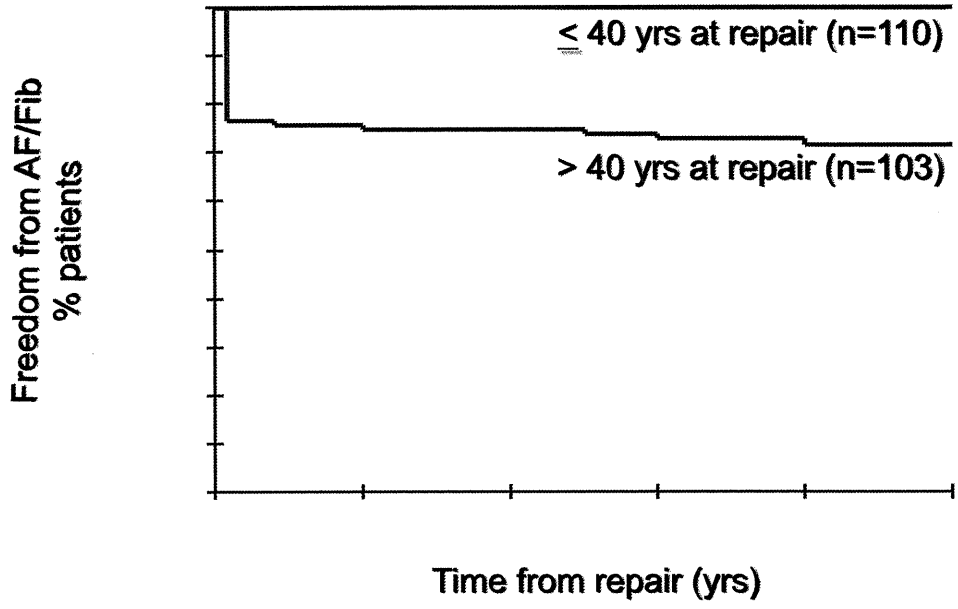
右室、右房拡大の減少

ASD修復術後生存率 (ASD vs 一般)



Murphy et al NEJM 1990

ASD修復術後の心房細動の合併率

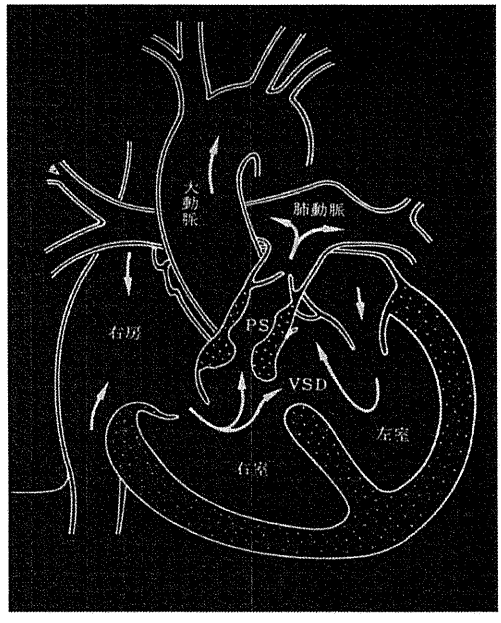


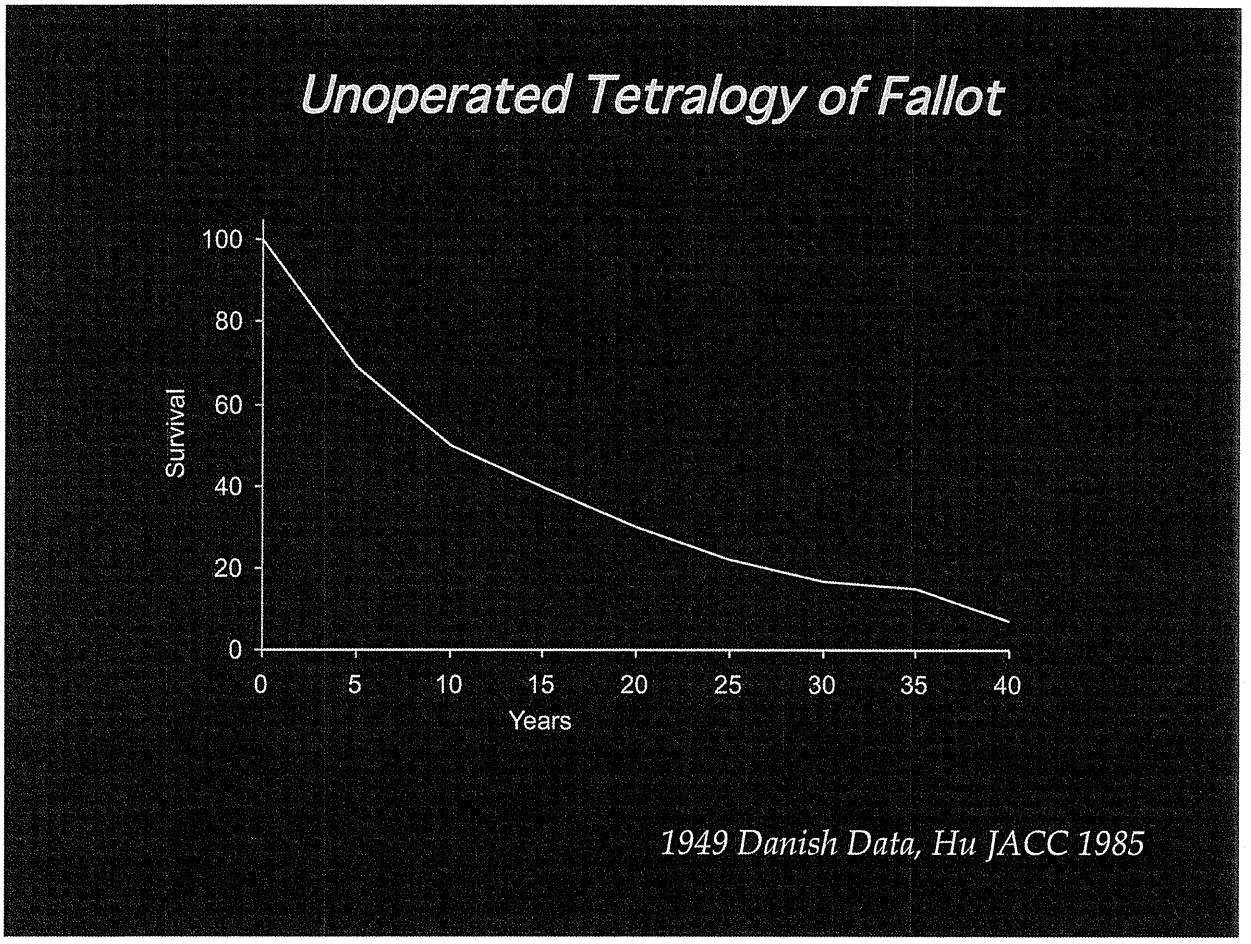
Gatzoulis et al NEJM 1999

ファロー四徴症
全先天性心疾患の10%

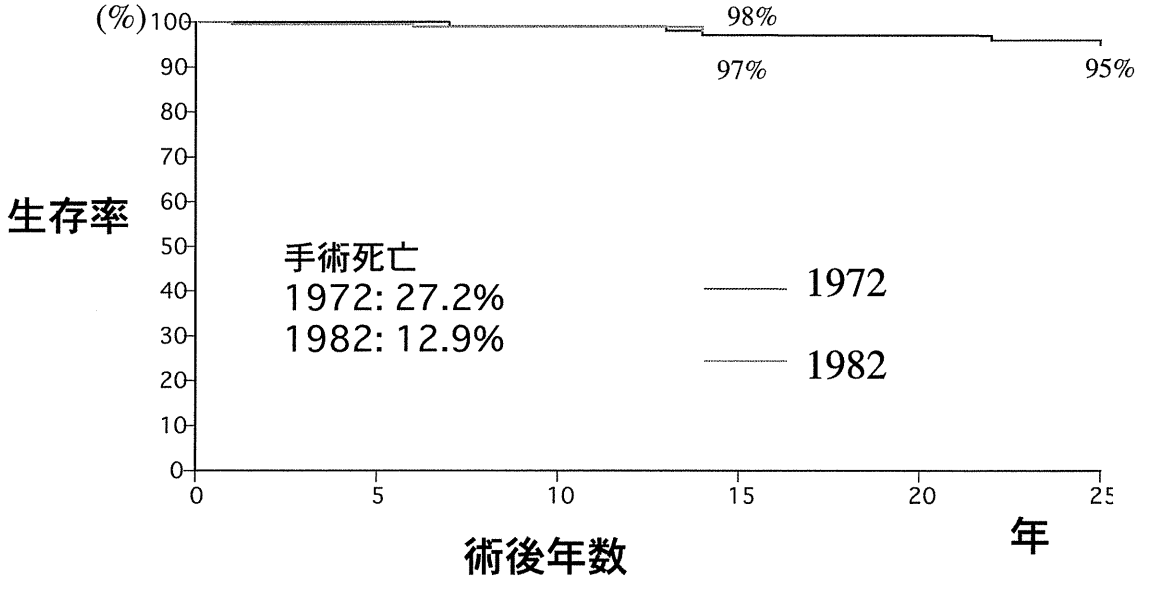
1) 肺動脈狭窄
2) 心室中隔欠損
3) 大動脈騎乗
4) 右室肥大

手術
肺動脈形成(狭窄解除)
右室流出路拡大(パッチ)
心室中隔欠損パッチ閉鎖

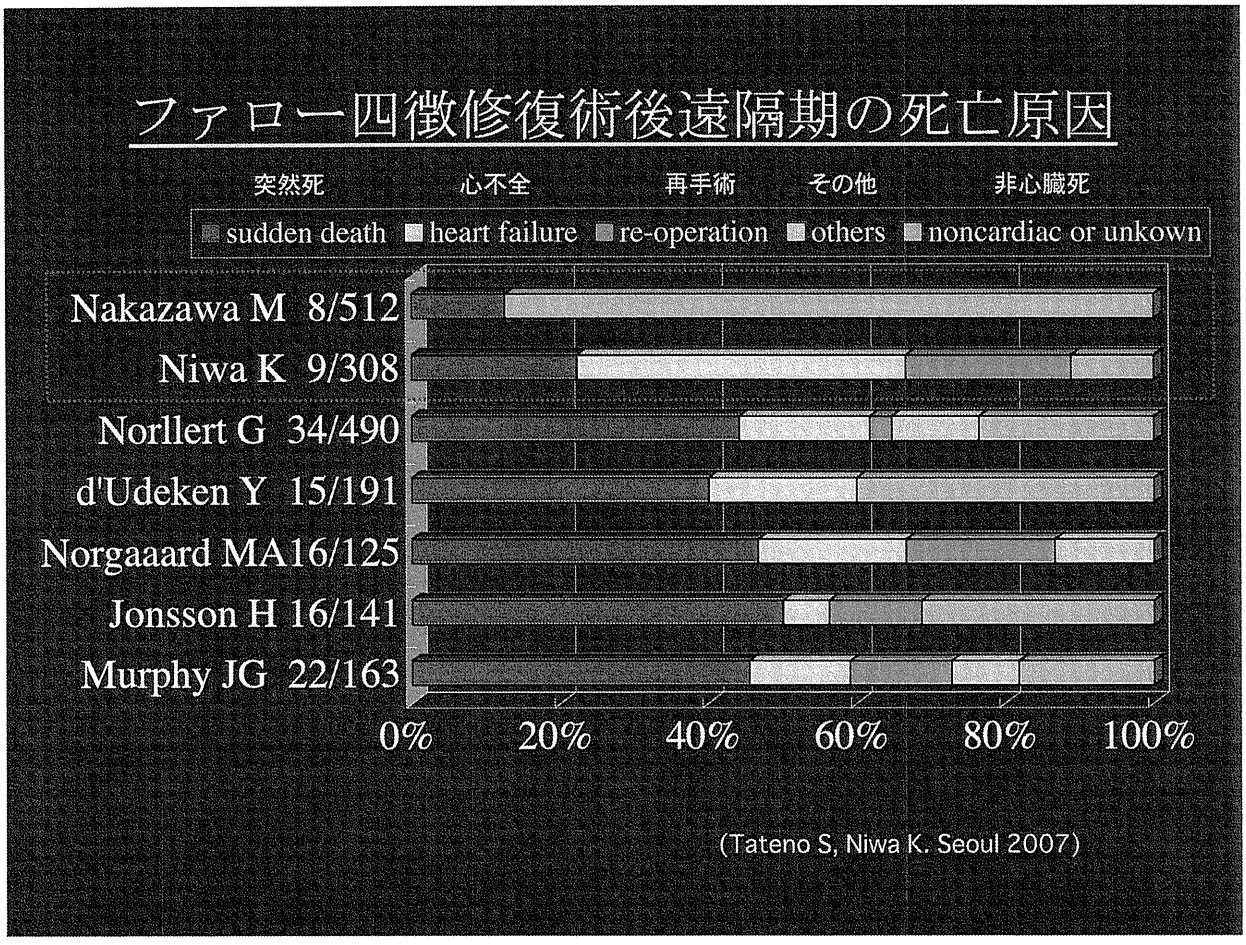


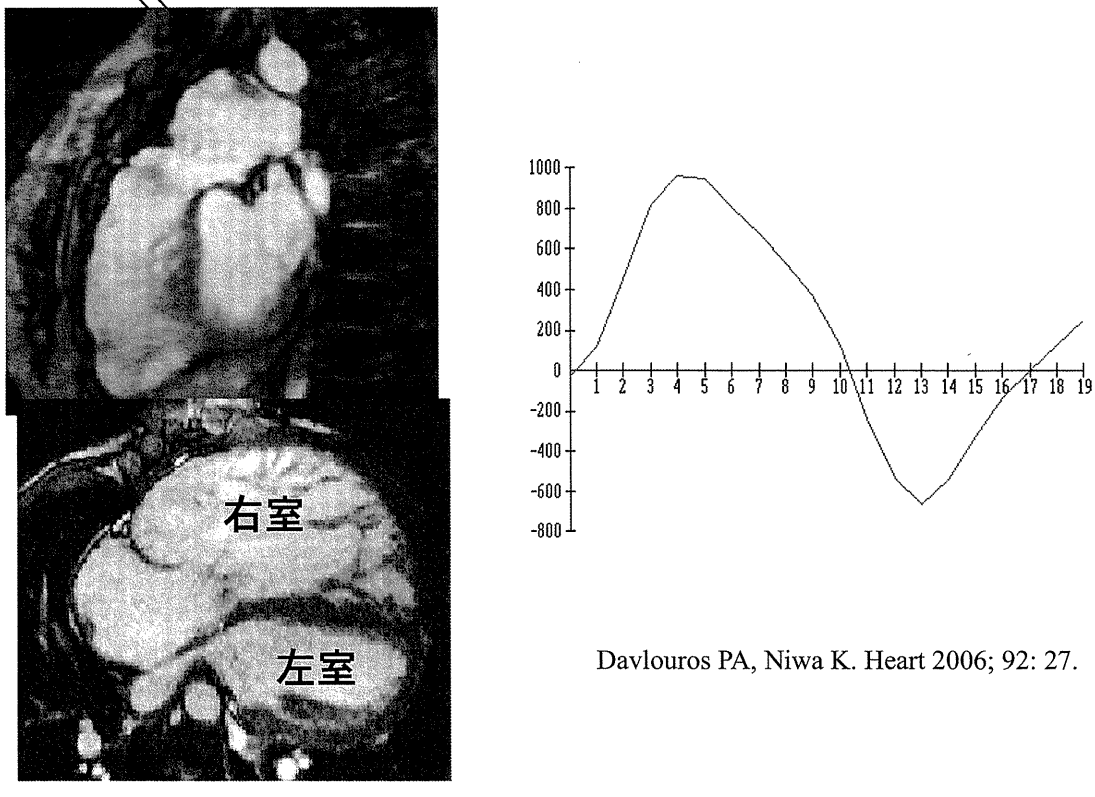


ファロー四徴症手術生存後の生命曲線



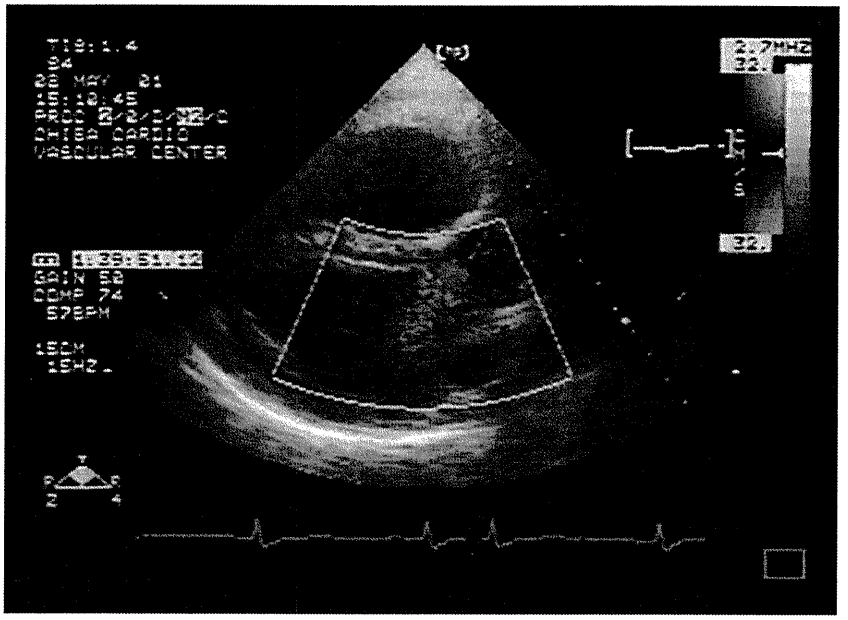
(Niwa K, Hamada H. Cardiol Young 2002;12)



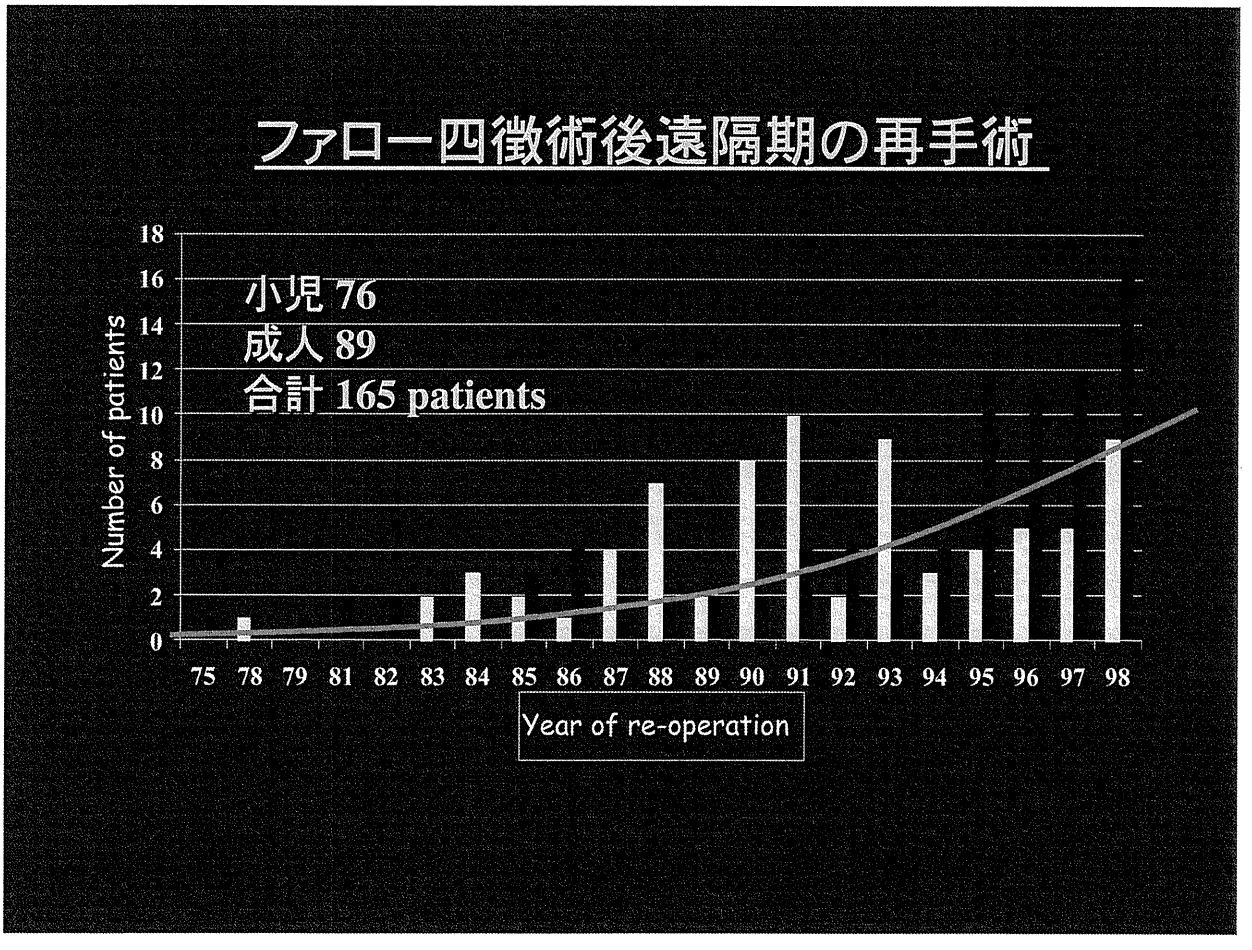


Davlouros PA, Niwa K. Heart 2006; 92: 27.

心エコー 所見、大動脈弁逆流



右室圧: 45mmHg
中等度三尖弁閉鎖不全
高度肺動脈閉鎖不全
中等度大動脈閉鎖不全
左室容積 : 160%N
右室収縮末期径: 50mm



修復術後TOFの妊娠 -日本多施設研究-　(10施設)

98 人, 143 妊娠, (年齢 27 歳)
母体死亡: 0,
流産: 6%,人工流産: 3%, 未熟児産: 10%

	NYHA I	NYHA II	
不整脈	8 %	11 %	ns
心不全	5 %	11 %	ns
出産後不整脈	2 %	0 %	ns
出産後心不全	2 %	43 %	<0.01

(Akagi T,Niwa K. Circulation 2005;112: Supple II-682. AHA 2005)

先天性心疾患の妊娠、出産に関して多く聞かれる質問

1,妊娠は安全か,子どもを普通に産めるのか
2,普通の子どもが生まれるのか
3,どの疾患、どの状態だと出産が難しいか
　避妊すべきか、妊娠継続が可能か
4,妊娠中に気をつける点は何か
5,帝王切開になるのか
6,服用中の薬剤はどうするのか、授乳時は大丈夫か
7，子どもを普通に□ てられるか

(丹羽公一郎. 先天性心疾患の方のための妊娠・出産ガイドブック。
中央法規出版、東京、2006. 11. 1.)

結婚

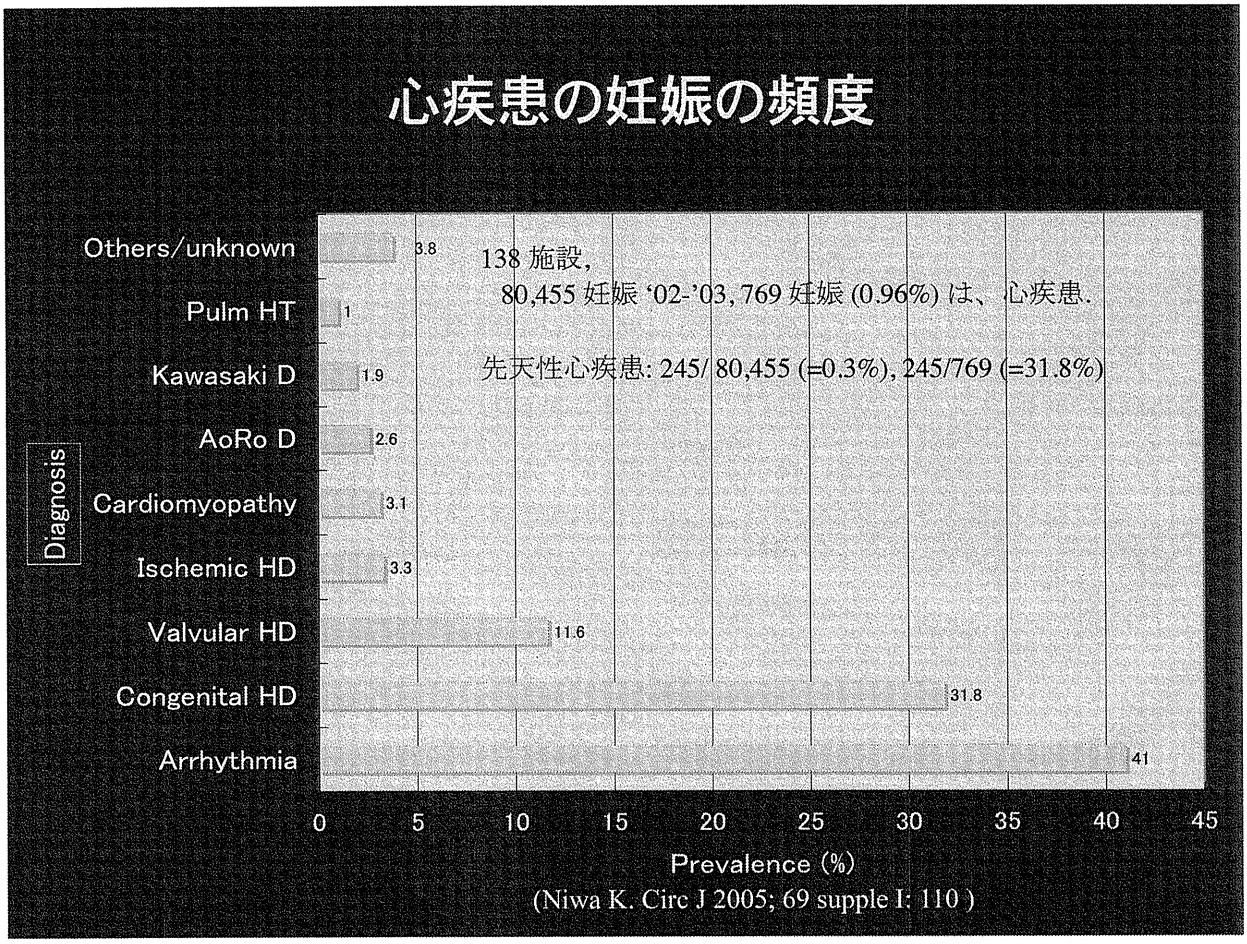
	チアノーゼ型 (n=13)	非チアノーゼ型 (n=102)	全体 (n=115)	一般
婚姻状況 (既婚/未婚/離婚)	2/11/0 (15%)	34/68/0 (33%)	36/79/0 (31 %)	31.6% (25-29y)
婚姻状況—男女差— (既婚/未婚)			男:10/52(19%) 女:26/63(41%)	
子供	1 (8%)	24 (23%)	25 (22%)	14% (<29y) 23% (<34y)
子供—男女差— (既婚/未婚)			男:6/52(12%) 女:19/63(30%)	
子供の心臓病	0 (0%)	1 (3%)	1 (3%)	

(Niwa K, et al. J Cardiol 2002; 39: 259-66)

妊娠出産時の循環生理

1,血行動態的変化(運動時と類似した変化)
　全血液量増加:正常時の140-150%
　心拍出量増加: 正常時の140-150%
　末梢血管拡張、静脈血圧上昇
　出産時出血:500-900ml
　出産後4週間で妊娠前の状態に復帰
2,血液学的変化(凝固能亢進、貧血)
3,呼吸機能の変化(分時換気量増加)
4,内分泌学的変化(コルチゾール,エストロゲン, アルドステロンなどの増加)
5,自律神経学的変化(心拍数増加)
6,大動脈壁変化(弾性線□ 断裂)

(丹羽公一郎. 先天性心疾患の方のための妊娠・出産ガイドブック. 中央法規出版、東京、2006. 11. 1.)



妊娠出産ー左右短絡疾患

1，126人の309妊娠、出産.

自然流産:17%、母体死亡:0、心奇形再発危険率:2.5%.

2，NYHAが良好で、心機能の良い例での出産は安全.

3,心房中隔欠損症心室中隔欠損症：心内修復術後で、遺残症や肺高血圧症が無く、心機能分類が良好であれば、母体と胎児の予後は良好.

(Zuber M,. Heart 1999;81:861-7)

厳重な注意を要する、妊娠前に修復しておくべき、妊娠は控えた方が良いと考えられる先天性心疾患

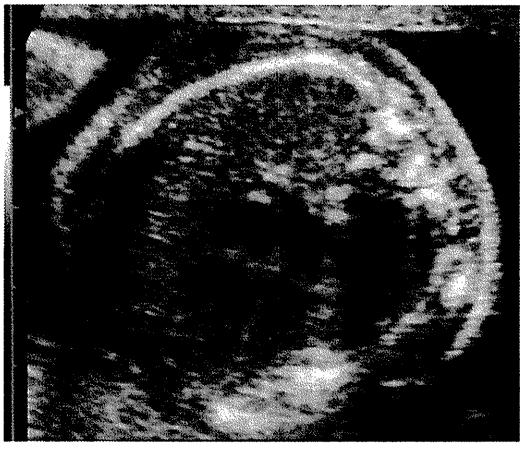
1, 肺高血圧 (アイゼンメンゲル症候群)
2, 流出路狭窄(大動脈弁高度狭窄、＞50mmHg)
3, 心不全(NYHA3度以上、左室駆出率 <35%)
4, 大動脈拡張疾患(大動脈拡張期径>45mm)
5, 機械弁
6, チアノーゼ型疾患(酸素飽和度:＜85%)
7, フォンタン手術後

(中澤誠. 心疾患患者の妊娠・出産の適応、管理に関するガイドライン。Circ J 2005)

先天性心疾患女性に対する胎児超音波検査

先天性心疾患の女性から
約5%の心疾患が出生

本人、夫や前児が心疾患の場合



28yr ファロー四徴症術後
妊娠 21wk 胎児エコー
(心室中隔欠損)

社会的自立

1,社会的自立とは、先天性心疾患患者が、成人となって、□ 般社会に参加し、広い意味で社会的貢献をしていくことが出来ること。

2,具体的には、教□ 、就業、結婚、出産、保険、年金、レクリエーション、社会活動などがあげられます。

3,これらの社会的自立を妨げる因子は、

□ 療面、患者側、社会側

の三つの側面に分けて考えられます。

(Niwa K, et al. J Cardiol 2002; 39: 259-66)

教□ 、就業

	チアノーゼ型 (n=13)	非チアノーゼ型 (n=102)	全体 (n=115)	一般
高校卒業	9 (69%)	90 (88%)	99 (86%)	94% (25-29y)
短大専門学校卒業	0 (0%)	43 (42%)	43 (37%)	27%(男)
大学卒業	0 (0%)	19 (19%)	19 (17%)	34%(女)
就業 (yes/no)	4/6 (40%)	68/10 (87%)	72/16 (82%)	80.3% (25-34y)
就職の有利さ一経験 (有利/不利/無関係)	2/1/1 (50%)	4/8/56 (6%)	6/9/57 (8%)	
就職の有利さ一推測 (有利/不利/無関係)	0/9/4	0/49/53	0/58/57	

(Niwa K, et al. J Cardiol 2002; 39: 259-66)

保険

	チアノーゼ型 (n=13)	非チアノーゼ型 (n=102)	総計 (n=115)	一般
身体障害者認定	12 (92%)	23 (23%)	35 (30%)	
身体障害者等級(1/3/4)	10/2/0	12/8/3	22/10/3	
障害者基礎年金	8	6	14	
生命保険 (yes/no/(denied)	2/8(1) (18%) (不明3)	52/37(5) (55%) (不明13)	54/45(6) (51%) (不明16)	60.8% (20-29y)
病名告知 (yes/no)	0 (不明2)	15/7 (不明30)	15/7 (不明32)	

(Niwa K, et al. J Cardiol 2002; 39: 259-66)

社会生活におよぼす心理社会的影響因子

1, 心理的要因
- 情動の変動が大きい
- 認知機能発達の遅れ
- 身体的な□ 象 (外科手術創部, からだが小さい)
- 劣等意識

3, 社会的要因
- 精神的成熟の遅れ(親の過保護)
- 友人関係をうまく築けない

内的障害についての周囲の無理解

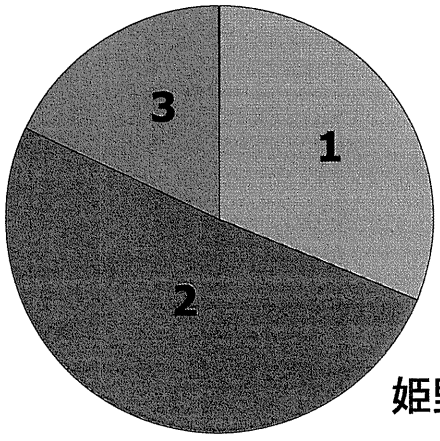
社会保障体制の不備

(Kovacs AH. AHJ 2005;150:193.)

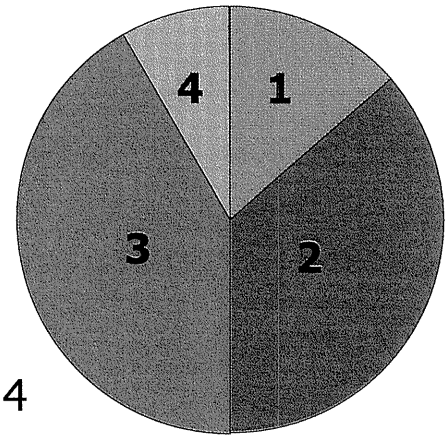
あなたは,手術の時についた傷に対し,現在どのように感じていますか?

1.まったく気にならない
2.ほとんど気にならない
3.少し気になる
4.大変気になる

男性(46人)

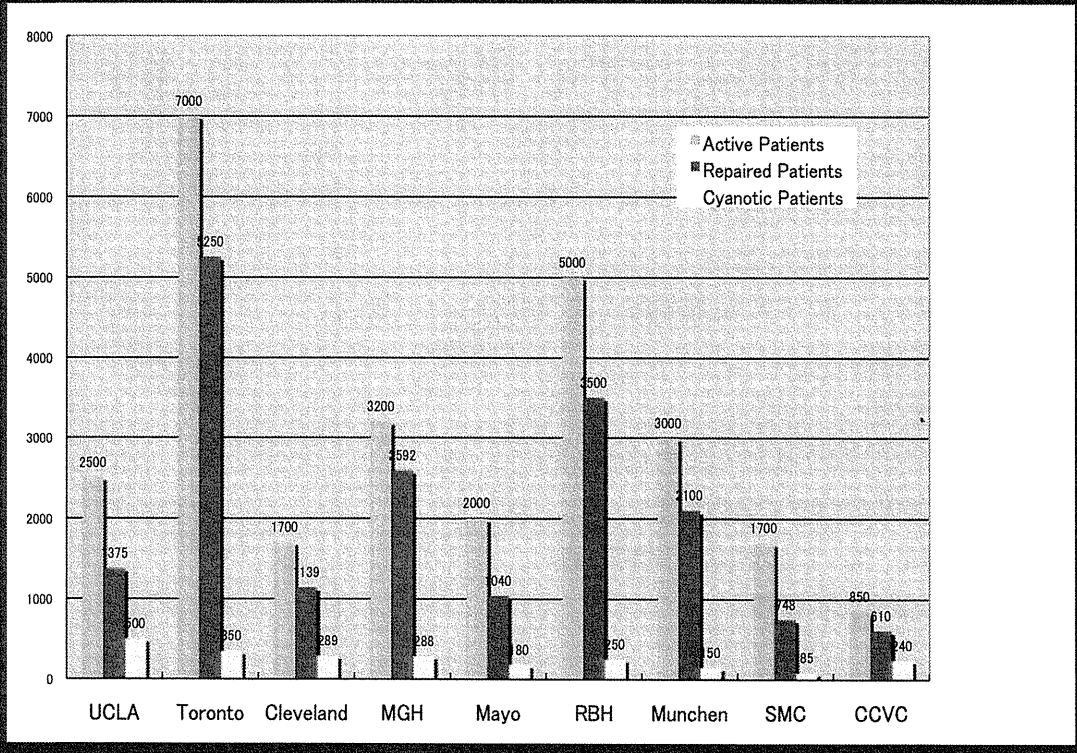


女性(92人)



姫野.2004

成人先天性診療部の患者数



Niwa K. ISACCD 2005

成人先天性心疾患診療部のスタッフ

□□	UCLA	Toronto	Clevelnd	MGH	Mayo	RBH	Munch	SMC	CCVC
□立□□	1978	1959	1980	1976	1987	1975	1974	1995	1998
□□科	内科、小児科	内科	小児科	内科、小児科	内科	内科	内科、小児科	内科、小児科	成人先天性診療部
スタッフ									
循環器科医	3	7	0	1	4	3	2	2	3
小児循環器科医	1	3	4	2	0	2	4	3	4
心臓外科医	2	4	2	2	4	3	3	3	2
専任看護師	1	1	3	0	1	0	0	1	1

(Niwa K. IJC 2005)

臨床的活動内容

施設	UCLA	Toronto	Clevelnd	MGH	Mayo	RBH	Munch	SMC	CCVC
外来									
患者数/週	18-25	40-50	6-30	40-50	12-22	50	40-50	15-20	50
病棟									
	成人	成人	成人,小児	成人,小児	成人	成人	成人,小児	成人,小児	成人,小児
入院/年	350	660	100	180	300	450	500	299	80
心臓外科									
手術数/年	170	150	50	70	100	100	90	85	30
再手術	80%	35%	25%	29%	56%	60%	45%	11%	20%
手術死亡	2 %	1.5 %	2 %	1.5 %	3 %	1.5 %	3%	1.1%	2%

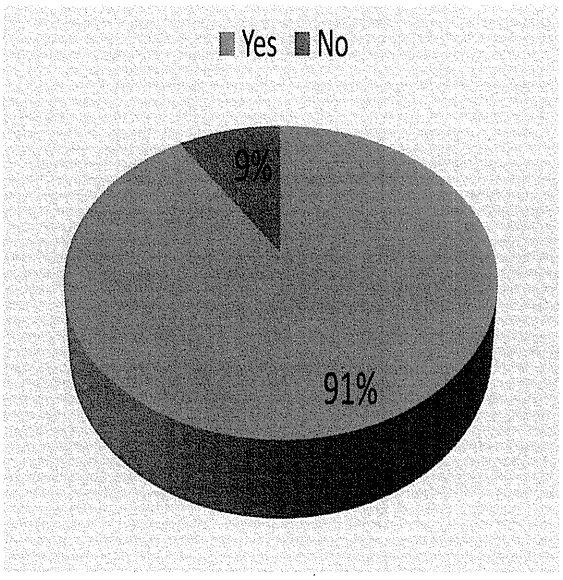
(Niwa K. IJC 2005)

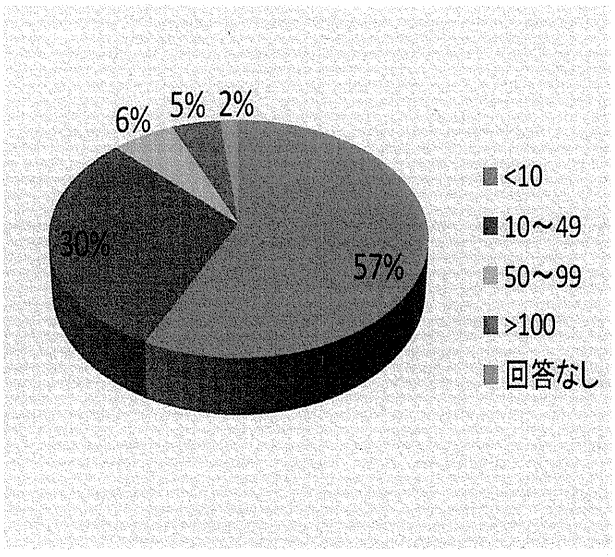
循環器小児科 VS 循環器科

	□ 環□ □ 児□	□ 環□ □
CHDに対する知識、興味	十分	乏しい
成人の病気の知識	乏しい	十分
外来	小児科	内科
病棟	多くは小児病棟	内科病棟
標榜科に対する抵抗感	大きい	殆どない
医者の絶対数	少ない（こどもを診るだけで多忙）	多い（成人疾患で多忙）

小児科、循環器科を問わず、
成人先天性心疾患を専門とする□ 師、□ 療スタッフが不可欠

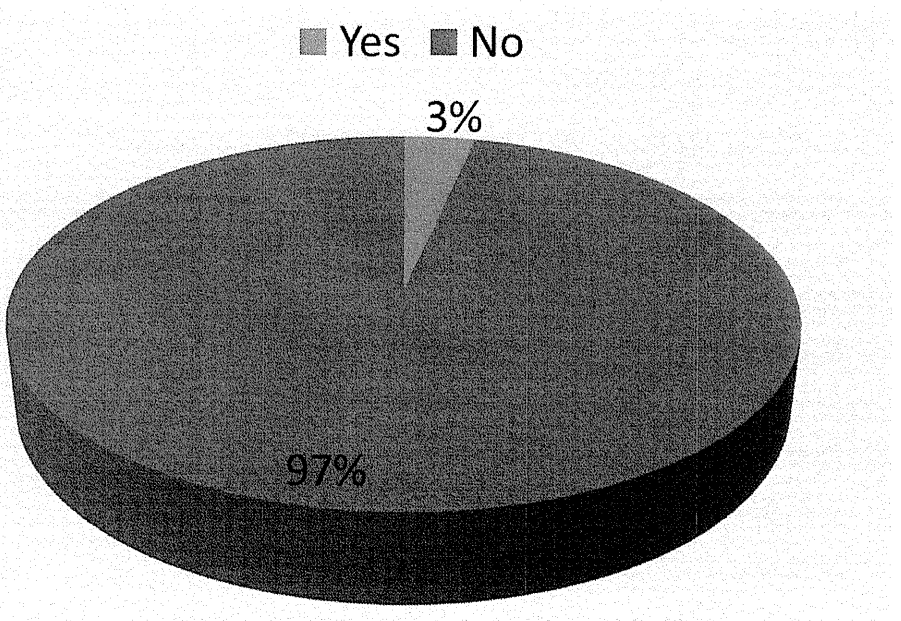
Adult CHD patients in outpatient Clinic





(豊田2008)

Special Clinic for Adult CHD



(豊田2008)

成人先天性心疾患ネットワーク

日本小児循環器学会分科会
日本成人先天性心疾患研究会
Japanese Society for Adult Congenital Heart Disease

更新日 2001/10/17
4806

HOME・新着情報
このホームページについて
成人先天性心疾患研究会
疾患について
リンク集
その他の情報
患者の方からのQ&A
症例検討

Contactas:
ADULTCHDJPN@aol.com

情報利用に際してのご注意とお願い

「医療情報の利用の手引き」

NEWS

10/17 患者の方よりの質問とお答え 修正大血管転位症とチアノーゼについて

学会情報
The 13th Annual International Symposium
on Congenital Heart Disease in the Adult Symposium を追加

10/11 患者の方よりの質問とお答え 心房中隔欠損症の手術法

学会情報
第四回 成人先天性心疾患研究会を追加

日本成人先天性心疾患研究会ホームページ協力に
千葉県循環器病センターを追加

症例についての内容を更新

9/28 患者の方よりの質問とお答え 心房中隔欠損症の長期管理
心室中隔欠損症術後の胸痛について

8/24 患者の方よりの質問とお答え 先天性心疾患術後の妊娠

結論

1. 外科治療、内科治療の恩恵を受け、先天性心疾患の多くが成人を迎えるようになりました。
2. 先天性心疾患を扱う施設ではその半数以上は成人となっています。
3. 成人先天性心疾患は、小児期と異なる問題点を抱えます。
4. 根治手術と考えられる先天性心疾患は少なく、多くは経過観察を必要とします。
5. 重症先天性心疾患では、加齢ともに、不整脈、心不全、再手術など□ 善すべき問題を抱えます。可能です。
6. 社会的自立を確立するために、移行期(思春期)に患者さん本人が、自分の病気を理解することが大切です。
7. チーム診療体制、社会保障体制を確立する必要があります。
8. この研究会の会員数は、年々増加しています。
9. さらに、この分野にたずさわる□ 師、□ 療関係者が徐々に、しかし、確実に増えてきています。全国に診療体制が広がる事も、それほど先の事ではありません。

International Journal of Cardiology 146 (2011) 13–16

Contents lists available at ScienceDirect

International Journal of Cardiology

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

Prevalence of adult patients with congenital heart disease in Japan

Yumi Shiina [a], Tomohiko Toyoda [a], Yasutaka Kawasoe [a], Shigeru Tateno [a], Takeaki Shirai [a], Yuko Wakisaka [a], Kozo Matsuo [a], Yoshiko Mizuno [a], Masaru Terai [b], Hiromichi Hamada [b], Koichiro Niwa [a,*]

[a] Chiba Cardiovascular Center Department of Adult Congenital Heart Disease and Pediatrics, Japan
[b] Tokyo Women's University Yachiyo Medical Center Department of Pediatrics, Japan

ARTICLE INFO

Article history:
Received 15 April 2009
Accepted 11 May 2009
Available online 2 June 2009

Keywords:
Prevalence
Adults with congenital heart disease
Death certificate
Annual increase

ABSTRACT

Background: Today most patients with congenital heart disease (CHD) can be expected to survive into adulthood. Reports regarding the number of adults with CHD in Japan are scarce. Our study aims to define the number of these adults.
Material and methods: The estimated number of infants born in Japan with major CHDs since 1947 was calculated together with mortality rates. We estimated the number of CHD survivors from data on survival rates of unoperated and postoperative patients. The number of deaths from 1968 to 1997 was analyzed using individual death certificates held by the Japanese Government.
Results: In 1967, 163,058 patients with CHD including 53,846 adults were assumed to be alive. From 1968 to 1997, 548,360 patients with CHD were born and 82,919 died. A total of 622,800 patients, including 304,474 children (49%) and 318,326 adults (51%) were estimated to be alive in 1997. From 1997 to 2007, there has been an estimated increase of 9000 adults every year, and in 2007, 409,101 adults are estimated to be alive.
Conclusions: The prevalence in adults with CHD in Japan has explosively increased from 1967 to 2007. There were 409,101 adults with CHD in 2007 with an annual increase of 9000. These data are crucial for planning the establishment in Japan of special facilities and resources necessary for the care of these patients.

1. Introduction

Owing to advances of surgical and medical management, most patients with congenital heart disease (CHD), even complex CHD, can be expected to reach adulthood. There have been several excellent reports about the estimated number of adults with CHD (ACHD) in Canada, UK, and US [1–3], but there are few reports on the number of ACHD patients from Asian countries including Japan. These data regarding the prevalence of ACHD are crucial in determining the resources and special facilities required for their care.

Our study sought to determine the number of adults with CHD based on the death certificates of CHD registered with the Japanese Government.

2. Material and methods

2.1. Sources of mortality data in Japan

In Japan, death certificates have been registered with the Government within 7 days of demise, and must include name, sex, date of birth, date of death, age of death, place of death, and cause of death. The Japanese Government publishes the number and causes of death, coded according to the International Classification of Disease (ICD) in the annual national vital statistics. For 1968 to 1971, mortality data were obtained from the national vital statistics and the cause of death were coded into ICD. For 1972 to 1997, the data were obtained from individual death certificates registered with the Japanese Government [4]. During the 30-year study period, the causes of death from CHD were classified according to ICD-8 (1968–1978), ICD-9 (1979–1994), and ICD-10 (1995–1997). The years before 1968 were excluded because CHD was not specifically classified in the ICD. The patients with endocardial fibroelastosis and chromosomal anomalies were also excluded. In Japan, the death registration began to be computerized in 1972, and the data from Okinawa prefecture were not available until 1972. ACHD is defined as patients with CHD over the age 15 years.

2.2. Mortality for severe CHD until the 1970s

Because of poor unoperated survival and high surgical death rates in Japan before the 1970s, most patients with complex CHD, except those with tetralogy of Fallot (TOF), died before their first year of life [4,5]. Even in TOF, the operative mortality rate was as high as 27% in 1972 according to a Japanese multicenter study [5]. We therefore excluded patients with severe CHD, except for those with TOF, born before 1972.

2.3. Calculation of the number of adults with CHD

2.3.1. Total number of patients with CHD in 1967

Estimated number of patients with CHD born from 1947 to 1967 was calculated by national birth rates and incidence of CHD in live born babies. The incidence of CHD was 10.6/1000 live births from the Japan Welfare Facilities Survey in 1986 [6]. Then, as in other reports [7], we used data on unoperated patients and death rates of cardiovascular surgery for calculation of survivors with CHD. Patients with CHD alive before the year 1947 were excluded, because official birth rates were not available due to World War II.

* Corresponding author. Department of Adult Congenital Heart Disease and Pediatrics, Chiba Cardiovascular Center, 575 Tsurumai, Ichihara, Chiba 290-0512, Japan. Tel.: +81 436 88 3111; fax: +81 436 88 3032.
E-mail address: KNIWA@aol.com (K. Niwa).

0167-5273/$ – see front matter © 2009 Elsevier Ireland Ltd. All rights reserved.
doi:10.1016/j.ijcard.2009.05.032

2.3.2. Total number of patients with CHD born from 1968 to 1997

These were estimated from the number of patients born with CHD from 1968 to 1997 minus national mortality rate in patients with CHD from 1968 to 1997. Patients with spontaneous closure of ventricular septal defect (VSD), estimated at 15% of CHD, were also excluded from the calculation of the number of ACHD.

2.3.3. Number of ACHD

The number of ACHD patients born before 1971 was calculated from survival rates of unoperated patients, mortality rates of cardiac surgery, and long-term survival rates after surgery. Patients with CHD born from 1972 to 1982 would have been over 15 years old in 1997, and the number was estimated from annual number of live births with CHD and annual number of deaths with CHD. The number of patients with ACHD born from 1972 to 1992 was estimated from annual live births with CHD and total number of deaths with CHD under 5 years of age.

2.3.4. Number of adults with CHD according to severity

Severity of CHD was classified following the Task Force 1 of the 32nd Bethesda conference of the American College of Cardiology [1]. We estimated the number of patients with major CHD in 1967 using previous reported data [Table 1].

3. Results

3.1. Number of deaths due to CHD from1968 to 1997

According to our previous study, during 30 year periods from 1968 to 1997, the total number of deaths due to CHD was 82,919, and number of deaths in different age ranges was 48,672 (59%) in the first year of life, 19,446 (23%) between 1 and 19 years of age and 14,801 (18%) in adults, respectively.

3.2. Number of patients with each major CHD in 1967 [Fig. 1]

Number of patients with each major CHD in 1967 was calculated based on the data of birth rate and unoperated history of CHD as follows; Patent ductus arteriosus: 12,286, VSD: 87,906, Atrial septal defect: 16,363, Coarctation of the aorta: 8315, Aortic stenosis and Pulmonary stenosis: 855 and 30,521 each, Atrioventricular septal defect,: 2254, TOF: 3524, Ebstein's anomaly: 1032, respectively (Fig. 1).

3.3. Number of patients with CHD [Fig. 2]

From data on the number of adults with the various malformations mentioned above, at least 163,058 patients with total CHD were alive in 1967, and 157,359 born from 1947 to 1967 were alive as adults in 1997. From 1968 to 1997, at least 548,360 patients with CHD were born and 82,919 patients with CHD were dead. These data suggest that at least 622,800 patients were alive in 1997 (Fig. 2).

3.4. Number of adults with CHD from 1967 to1997

In 1967, only 53,846 adult patients were alive. In 1971, the number of ACHD patients slightly increased to 84,196. 177,457 born from 1947 to 1971 were alive as adults in 1997. Estimating from the death records, 165,729 born from 1972 to 1982 were adults in 1997.

Table 1
The prevalence, unoperated survival, survival rate after cardiac surgery and long-term survival of major CHD.

	Prevalence in Japan [6]	Survival rate without cardiac surgery	Survival rate after cardiac surgery	Long-term survival after cardiac surgery
PDA[a] [15–17]	3.6%	Large PDAs 20 year-survival;50%	1940–1960;80% 1961– ;98%	Very well
VSD[b] [18–31]	56.6%	Large VSDs[c] 20 year-survival; 60% 40 year-survival; 20% For all degree of VSDs[d] 20 year-survival; 73% 40 year-survival; 47% 60 year-survival; 31%	1960–1980; 87%	Very well (above 95%)
ASD [17–19,33]	5.3%	For all degrees of ASDs 30 year-survival; 75% 50 year-survival; 25%	1955–1970; 80% 1971–;98%	Very well
CoA [32–34]	2.7%	Critical CoA (33%); few survive without repair 20 year-survival; 75% 45 year-survival; 25%	97% from 1950s	10 year-survival; 95% 40 year-survival; 87%
AS[e] [35,36]	0.4%	Critical AS (10%); Few survive without repair	1940–1959; 50% 1960–1979; 65% 1980–1984; 80% 1985–1989; 85% 1990–1994; well	10 year-survival; 96% 25 year-survival; 83%
PS [37,38]	9.6%	Critical PS; Few survive without repair	1940–1959; 75% 1960–1979; 90% 1980– , well	25 year-survival; 95%
AVSD [39,40]	1.8%	Complete AVSD (75%); Few survive without surgery Partial AVSD (25%) 27 year-survival; 75% 50 year-survival; 25%	1960–1980; 40% 1981– ;75%	10 year-survival; 78% 20 year-survival; 65% 5 year-survival; 95% 20 year-survival; 94%
TOF [5]	4.5%	Until 1959; few survive without surgery	1966–1980; 73%	1960–1965 palliative surgery (aorto-pulmonary shunt or BT shunt) 25 year-survival; 50% 40 year-survival; 25% 1966-intracardiac repair 14 year-survival; 97% 25 year-survival; 95%
Ebstein [41–43]	0.4%	3 year-survival; 75% 8 year-survival; 65%		20 year-survival; 95% 30 year-survival; 85%

[a] Silent PDAs, those are found accidentally during echocardiography, are excluded in this calculation.
[b] The incidence of subpulmonary VSDs has been reported to be as high as 28–35% of total VSDs in Japan. Spontaneous closure rate of subpulmonary VSDs has been reported to be only 3.8%, but that of perimembranous VSDs 74%. About 90% of small muscular VSDs close spontaneously.
[c] Large VSDs lead to death or cause Eisenmenger syndrome if unoperated, and this type is 5–11% of all VSDs [44].
[d] In the case of small defects without repair, the long-term mortality rate is very low.
[e] Bicuspid aortic valves are excluded.

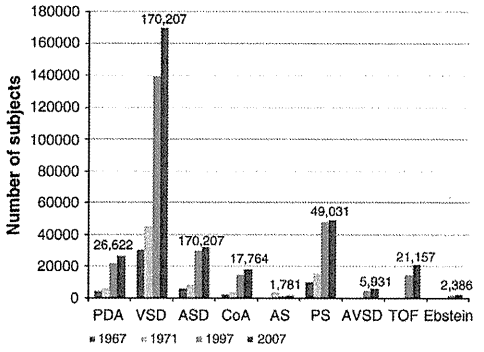


Fig. 1. The number of adults with major CHD from 1967 to 2007. This figure shows that adults with major CHD in 1967, in 1971, in 1997 and in 2007, consecutively. Number of adults with major CHD is increasing explosively.

Excluding patients with spontaneously closed VSD, 140,869 born from 1972 to 1982 were adults in 1997.

Therefore, in total, there were 318,326 adults (51%) and 304,474 children (49%) with CHD in 1997.

3.5. Number of adults with CHD in 2007

In 2007, there were 167,445 adults who were born from 1947 to 1971. After excluding the patients with spontaneously closed VSD, there were 241,656 adults born from 1972 to 1992 (85% of 284,302 adults born in that period). As a result, in 2007, there were a total of 409,101 ACHD patients. From 1997 to 2007, the annual increase in Japan has been about 9000.

3.6. Number of adults with CHD according to the severity of CHD [Fig. 3]

In 1967, 50,651 (94% of ACHD) were mild. In 1997, 239,374 (75%) were mild and 78,952 (25%) were moderate to severe. In 2007, 278,001 (68%) were mild and, as many as 131,101 (32%) were moderate to severe (Fig. 3).

4. Discussions

This study indicates that the prevalence of adults with CHD in Japan has dramatically increased from 1967 to 2007. Of 409,101 adults with CHD, 32% with moderate to severe CHD were alive in 2007 with

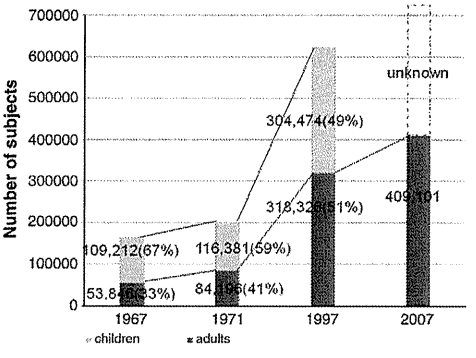


Fig. 2. The total number of patients with CHD from 1967 to 2007. This figure shows that the total number of patients with CHD in 1997 is much larger than that in 1967 with remarkably higher ratio of adults/children.

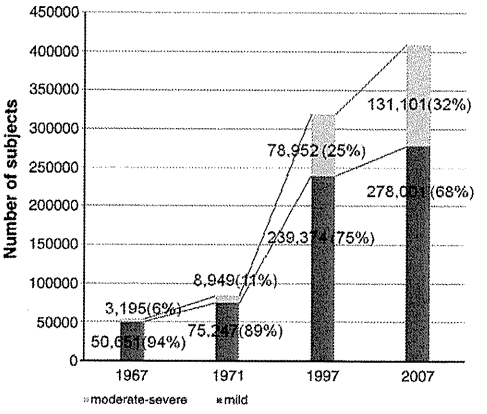


Fig. 3. The number and percentage of adults with CHD according to the severity of CHD from 1967 to 2007. This figure shows that the number and percentage of adults with moderate to severe CHD have been increasing dramatically from 1967 to 2007.

an annual increase of 9000. These data are crucial for judging the establishment of special facilities and staffs caring for adults with CHD in Japan.

Approximately 85% of babies born with CHD can now be expected to reach adulthood owing to improved surgical techniques and medical management [8,9,10]. The incidence of ACHD can be anticipated to increase dramatically over the next decade.

From a recent study, the prevalence of ACHD in Quebec was 4.09 per 1000 live births, and in Canada, an estimated 96,000 ACHD patients in 2000 [3]. Importantly, the prevalence of adults with severe CHD has been increasing, and a half (49%) of patients with severe CHD were adults in 2000 [3].

There were 104,479,000 people aged over 15 in Japan in 2007 [11], and at least 409,101 with ACHD. The prevalence of ACHD was 3.92 per 1000 adults in 2007, a bit smaller than Canadian data (4.09 per 1000 adults). Hoffman reported that in the US if all patients were treated, there would be 750,000 survivors with mild CHD and 580,000 survivors with moderate to severe CHD in 2002. If not treated, there would be 400,000 survivors with mild CHD and 250,000 survivors with moderate to severe CHD [7]. In fact, the actual numbers of surviving adults will be between these two estimates. Wren has also reported that the adult population would consist of 28% complex, 54% significant, and 18% minor CHD [2]. The definition of severity of CHD differs slightly but over 40% of all CHDs in adults can be considered as moderate to severe CHD. Similar to these data is our assumption that at least 32% of all patients with ACHD in Japan have moderate to severe CHD.

In this study, we analyzed the number of ACHD patients, but are also concerned about children with chronic pediatric cardiac disorders such as cardiac tumor, Marfan syndrome, cardiomyopathy, primary arrhythmia, and other cardiac problems that need continuous follow-up and are now getting into adulthood. Also the number of patients with a history of Kawasaki disease is now as high as 200,000, and those with coronary artery lesions need to be continuously followed-up as adults in Japan [12,13]. In this study, we excluded these patients from calculation of ACHD, and can easily underestimate the number of patients graduating from the pediatric clinic and moving to the adult congenital cardiology clinic. Gatzoulis et al. reported a 2695 expansion in outpatients' workload in 10 years (1987 to 1997) in Toronto [14]. Wren has reported a 400% growth in the number of clinics for ACHD in the past 10 years [2].

Only a few adult cardiologists in Japan are aware of this surprising phenomenon and their responsibility for ACHD, but it is important that appropriate arrangements be made for the transfer care of ACHD

from the pediatric to the adult service. Our data will be useful for future establishment of care facilities and resources for this expanding population.

5. Limitations

First, co-existing CHD was not identified from individual death certificates and may result in ICD cording errors. Next, estimating the number of adults with mild CHD, especially perimembranous VSD, is difficult because many of them will have spontaneously closed during early childhood with no need for follow-up. Secondum atrial septal defect, Ebstein's anomaly, and congenitally corrected transposition may not be diagnosed until adulthood resulting in an underestimate in the number of ACHD. Also, the number of patients with bicuspid aortic valve is hard to establish because most of them are without symptoms during childhood and are not diagnosed until adulthood. Most patients with cyanotic CHD and palliative surgery may not reach adulthood, however, some are alive as adults. We may underestimate this population, but the number is very small and would have little effects on our data. Except for several CHDs such as TOF and VSD, Japanese data including unoperated patients, mortality rates, and long-term survival after repair are scarce, so we applied data from North America and Europe. It was impossible to calculate the number of children with CHD in 2007 because there were no official registration systems of the number of CHD patients in Japan, and no data on individual death certificates held by the Japanese Government from 1997 to 2007. So we show only the number of ACHD in 2007 in Fig. 2.

6. Conclusions

The prevalence of adults with CHD has been explosively increasing from 1967 to 2007. Total 409,101 adults with CHD, at least one-third of them thought to have moderate to severe CHD, are alive in 2007 with an annual increase of 9000. These data are important for the establishment of the special facilities and resources for adult CHD patients in Japan, and for the establishment of education and training systems for adult cardiologists in this evolving field.

Acknowledgement

We gratefully acknowledge JK Perloff MD for reviewing the manuscript. The authors of this manuscript have certified that they comply with the Principles of Ethical Publishing in the International Journal of Cardiology [45].

References

[1] Warnes CA, Liberthson R, Danielson GK, et al. Task force 1: the changing profile of congenital heart disease in adult life. J Am Coll Cardiol 2001;37(5):1170–5.
[2] Wren C, O'Sullivan JJ. Survival with congenital heart disease and need for follow up in adult life. Heart 2001;85(4):438–43.
[3] Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation 2007;115(2):163–72.
[4] Terai M, Niwa K, Nakazawa M, et al. Mortality from congenital cardiovascular malformations in Japan, 1968 through 1997. Circ J 2002;66(5):484–8.
[5] Niwa K, Hamada H, Nakazawa M, et al. Mortality and risk factors for late deaths in tetralogy of Fallot: the Japanese Nationwide Multicentric Survey. Cardiol Young 2002;12(5):453–60.
[6] Nakazawa M, Seguchi M. Birth rates and CHD incidence in live born babies from Japan Welfare Facilities survey in 1986. Jpn Pediatr 1986;90:2578–87 [in Japanese].
[7] Hoffman JI, Kaplan S, Liberthson RR. Prevalence of congenital heart disease. Am Heart J 2004;147(3):425–39.
[8] Moller JH, Taubert KA, Allen HD, Clark EB, Lauer RM. Cardiovascular health and disease in children: current status. A Special Writing Group from the Task Force on Children and Youth, American Heart Association. Circulation 1994;89(2):923–30.
[9] Matsuoka R, Mori K, Ando M. Epidemiological research of CHD. Pediatr Cardiol Card Surg 2003;19(6):606–21 [in Japanese].
[10] Kitada M, Ueda K, Nakagawa T, Yamaguchi Y. Follow-up study into early adulthood of patients with congenital heart diseases. Jpn Circ J 1987;51(12):1409–14.
[11] Olley PM, coceani F, Bodach R. E-type prostaglandins: a new emergency therapy for certain cyanotic congenital heart malformation. Circulation 1976;53:728–31.
[12] Annual Summary of Vistal Statistics. The ministry of health and welfare, Japan; 2007.
[13] Yanagawa H, Yashiro M, Nakamura Y, Kawasaki T, Kato H. Results of 12 nation-wide epidemiological incidence surveys of Kawasaki disease in Japan. Arch Pediatr Adolesc Med 1995;149:779–83.
[14] Gatzoulis MA, Hechter S, Siu SC, Webb GD. Outpatient clinics for adults with congenital heart disease: increasing workload and evolving patterns of referral. Heart 1999;81(1):57–61.
[15] Campbell M. Natural history of persistent ductus arteriosus. Br Heart J 1968;30(1):4–13.
[16] Talner CN. In: Donald C, Fyler MD, editors. Report of the New England Regional Infant Cardiac Program, vol. 65(suppl). Pediatrics; 1980. p. 375–461.
[17] Morris CD, Menashe VD. 25-year mortality after surgical repair of congenital heart defect in childhood. A population-based cohort study. JAMA 1991;25;266(24):3447–52.
[18] Laursen HB. Some epidemiological aspects of congenital heart disease in Denmark. Acta Paediatr Scand 1980;69(5):619–24.
[19] Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. Am J Cardiol 1978;42(4):641–7.
[20] Feldt RH, Avasthey P, Yoshimasu F, Kurland LT, Titus JL. Incidence of congenital heart disease in children born to residents of Olmsted County, Minnesota, 1950–1969. Mayo Clin Proc 1971;46(12):794–9.
[21] Ando M, Takao A. Pathological anatomy of ventricular septal defect associated with aortic valve prolapse and regurgitation. Heart Vessels 1986;2(2):117–26.
[22] Anzai T, Iijima T, Yoshida I, Sakata Y, Obayashi T, Ishikawa S. The natural history and timing of the radical operation for subpulmonic ventricular septal defects. Jpn J Surg 1991;21(5):487–93.
[23] Tomita H, Arakaki Y, Yagihara T, Echigo S. Incidence of spontaneous closure of outlet ventricular septal defect. Jpn Circ J 2001;65(5):364–6.
[24] Miyake T, Shinohara T, Nakamura Y, et al. Spontaneous closure of ventricular septal defects followed up from < 3 months of age. Pediatr Int 2004;46(2):135–40.
[25] Hiraishi S, Agata Y, Nowatari M, et al. Incidence and natural course of trabecular ventricular septal defect: two-dimensional echocardiography and color Doppler flow imaging study. J Pediatr 1992;120(3):409–15.
[26] Ooshima A, Fukushige J, Ueda K. Incidence of structural cardiac disorders in neonates: an evaluation by color Doppler echocardiography and the results of a 1-year follow-up. Cardiology 1995;86(5):402–6.
[27] Hoffman JI, Rudolph AM. The natural history of ventricular septal defects in infancy. Am J Cardiol 1965;16(5):634–53.
[28] Campbell M. Natural history of ventricular septal defect. Br Heart J 1971;33(2):246–57.
[29] Roos-Hesselink JW, Meijboom FJ, Spitaels SE, et al. Outcome of patients after surgical closure of ventricular septal defect at young age: longitudinal follow-up of 22–34 years. Eur Heart J 2004;25(12):1057–62.
[30] Otterstad JE, Erikssen J, Frøysaker T, et al. Long term results after operative treatment of isolated ventricular septal defect in adolescents and adults. Acta Med Scand, Suppl 1986;708:1–39.
[31] Gabriel HM, Heger M, Innerhofer P, et al. Long-term outcome of patients with ventricular septal defect considered not to require surgical closure during childhood. J Am Coll Cardiol 2002;39(6):1066–71.
[32] Kirklin JW, barratt-Boyes BG. Coarctation of the aorta and interrupted aortic arch. In: Kirklin JW, Barratt-Boyes BG, editors. Cardiac surgery. New York: Churchill Livingstone; 1993. p. 1263–325.
[33] Hoffman JIE. Reflections on the past, present, and future of pediatric cardiology. Cardiol Young 1994;4:208–23.
[34] Høimyr H, Christensen TD, Emmertsen K, et al. Surgical repair of coarctation of the aorta: up to 40 years of follow-up. Eur J Cardio-thorac Surg 2006;30(6):910–6.
[35] Detter C, Fischlein T, Feldmeier C, Nollert G, Reichart B. Aortic valvotomy for congenital valvular aortic stenosis: a 37-year experience. Ann Thorac Surg 2001;71(5):1564–71.
[36] Perloff JK, Plunkett MD. Survival patterns after cardiac surgery or interventional catherterization: a broadening base. Congenital heart disease in adults. 3nd Edition. Philadelphia: W.B.Saunders; 2008. p. 69–71.
[37] Diller GP, Gatzoulis MA. Pulmonary vascular disease in adults with congenital heart disease. Circulation 2007;115(8):1039–50.
[38] Roos-Hesselink JW, Meijboom FJ, Spitaels SE, et al. Long-term outcome after surgery for pulmonary stenosis (a longitudinal study of 22–33 years). Eur Heart J 2006;27(4):482–8.
[39] Wren C, Richmond S, Donaldson L. Temporal variability in birth prevalence of cardiovascular malformations. Heart 2000;83(4):414–9.
[40] Mitchell SC, Korones SB, Berendes HW. Congenital heart disease in 56,109 births. Incidence and natural history. Circulation 1971;43(3):323–32.
[41] Watson H. Natural history of Ebstein's anomaly of tricuspid valve in childhood and adolescence. An international co-operative study of 505 cases. Br Heart J 1974;36(5):417–27.
[42] Augustin N, Schmidt-Habelmann P, Wottke M, Meisner H, Sebening F. Results after surgical repair of Ebstein's anomaly. Ann Thorac Surg 1997;63(6):1650–6.
[43] Kiziltan HT, Theodoro DA, Warnes CA, O'Leary PW, Anderson BJ, Danielson GK. Late results of bioprosthetic tricuspid valve replacement in Ebstein's anomaly. Ann Thorac Surg 1998;66(5):1539–45.
[44] Vongpatanasin W, Brickner ME, Hillis LD, Lange RA. The Eisenmenger syndrome in adults. Ann Intern Med 1998;128(9):745–55.
[45] Coats AJ. Ethical authorship and publishing. Int J Cardiol 2009;131:149–50.