厚生労働科学研究「成人に達した先天性心疾患の診療体制の確立に向けた総合的研究」　平成２２年度第３回班会議議事録

平成２３年２月２６日ホテル新大阪東口ステーションビル　会議室４０１
午後１時―午後３時３０分

参加者（１６名、敬称略）：中西敏雄、丹羽公一郎、八尾厚史、賀藤　均、堀端洋子、水野芳子、落合 亮太、市田 蕗子、松崎多千代、中井久仁夫、市川　肇、神谷千津子、城戸 佐知子、赤木 禎治、森崎隆之、白石 公

1.事務連絡

2.班会議の前回までの経過、厚生労働省への報告状況（白石）
厚生労働省への報告会の内容（平成２３年２月８日）

分担研究者からの研究報告
1.赤木先生（岡山大学）
岡山大学における成人先天性心疾患診療の現状と問題点
岡山大学と倉敷中央病院、岡大小児科と岡大外科にデータが分散
データベースの構築が困難
エコー技師の養成が必要で教育プログラムの作成を現在手がけている
循環器内科医２名が成人先天性心疾患診療に参加

2.落合先生（東京大学）、賀藤先生（成育医療センター）
成人先天性心疾患患者の社会的自立に関する研究報告

3.落合先生（東京大学）、八尾先生（東京大学）
循環器内科における成人先天性心疾患診療に関する全国調査

4.堀端先生、丹羽先生（千葉県循環器病センター）
千葉県循環器病センターにおける成人先天性心疾患外来の現状

5.市田先生（富山大学）

成人先天性心疾患専門医制度に向けた提言

Level 1, Level 2 を主体に制定考える

6.森崎先生（国立循環器病研究センター）

診療体制への提言

7.城戸先生（兵庫こども病院）

ファロー四徴患者の突然死に関して

8.松崎先生（富山大学）

成人先天性心疾患患者の社会心理的背景に関する調査報告

分担研究報告

平成２２年度厚生労働科学研究費、循環器疾患等生活習慣病対策
総合研究事業　成人に達した先天性心疾患の診療体制の確立に向けた総合的研究
研究成果抄録

研究課題：我が国における成人先天性心疾患の診療実態調査、成人先天性心疾患患者数調査と遠隔医療支援システムの確立に向けた総合的研究

主任研究者：国立循環器病研究センター小児循環器診療部
白石　公

分担研究者:　千葉県循環器病センター診療部長
丹羽公一郎

研究協力者：千葉県循環器病センター成人先天性心疾患診療部　医師　堀端洋子、同看護師　水野芳子

１．研究目的

この四半世紀、先天性心疾患の救命率が向上し患者の多くが成人期を迎えるようになった。我が国における成人先天性心疾患患者数は急激に増加しており、その診療は非常に専門性が高い。このため診療体制の構築が必須の分野であり、その基礎データである患者総数や診療実態について昨年度に調査を行った。その結果、成人先天性心疾患患者数は1997年には小児患者と成人患者の両者がほぼ同数となり、さらに2007年には成人患者数は409,101人となり、小児患者数をこえたと考えられる。今後10-20年はこの急激な増加傾向が続くと考えられる。また診療実態については、成人先天性心疾患を診療している施設数は多いが、一施設で多数の患者を診ている専門施設は非常に少なく、また専門のトレーニングを受けている医師も少ないことがわかった。このことからも患者数の増加に見合う専門の医療従事者の育成と診療施設の構築が急務であるが、そのために有効な手段として遠隔医療支援システムがある。これは専門外の医師が、患者データを専門医師と共有し、的確な診断と治療を行うために有用な方法である。特に、成人先天性心疾患の様に専門性が高く、専門医の少ない分野では、遠隔医療支援システムは望ましい診療方法であるが、このことに関する検討は行われていない。

本研究では、1)　千葉県内で成人先天性心疾患に関する遠隔医療支援システムの構築に関する検討を行い、それを全国ネットとして広げていく可能性を検討する、2)成人先天性心疾患専門外来を受診する患者の特徴を明らかにし、専門外来におけるニー

ズを検討する、3) 成人先天性心疾患患者の生活の質（QOL）と小児科から成人医療施設への移行期の問題を明らかにする、ことを目的とした。

2．研究方法

1）成人先天性心疾患に関する遠隔医療支援システムの構築：千葉県内で、画像転送ネットワークを構築するため、千葉県内で、先天性心疾患、成人先天性心疾患を診療している基幹病院（千葉県循環器病センター、千葉県こども病院。千葉大学病院、東京女子医科大学八千代医療センター）で、画像転送ネットワークを構築する。すでに千葉県こども病院に、サーバーが設置されており、千葉県循環器病センターなどの病院では、computerでアクセスすることにより、リアルタイム画像を共有する。心エコー装置は、共通の既存の装置を用いる必要があるが、これらの病院では、同機種の心エコー装置がすでに納入されているので、これを利用することが可能である。CT, MRI,血管造影データなどは、Dicom形式による video画像とする。共通の出力画面とレポートを採用する。このための適切な回線の調査、コンピューターソフト検索などを行う。今後、この方法を全国ネットとして広げていく可能性について検討する。画像転送ネットワークでの個人情報は、個人が特定されない形をとることに十分な配慮をする。各病院の資料持ち出し許可を取り、患者名、生年月日、ID番号などは、消去した画像を共有する。

2）成人先天性心疾患専門外来初診患者の特徴：2009年4月から2010年12月までに千葉県循環器病センター成人先天性心疾患外来を初診した15歳以上の先天性心疾患患者113名を対象に、年齢、性別、病名、受診理由をカルテから後方視的に調査した。

3）18才以上のCHD外来患者に対し自己記入式QOL尺度SF-36日本語版及び疾患に関する不安や心配などについての半構成面接を行った。

3．研究結果及び考察

1）成人先天性心疾患に関する遠隔医療支援システムの構築： 7 月に開催された小児循環器病学会でライブデモを行い、会場と千葉県こども病院をインターネット(NTT 光ネクスト回線、VPN ネクスト)を用いて接続し、画質(エコー、CT、心電図モニターなど)が良好であること、3 点(演者、座長、こども病院)での画像を供覧しながらのディスカッションが可能であることを確認した。また、共通のソフトウエアを使用することにより、適切な画像処理、計測、データ解析が行えた。施設間で共通して用いるレポートシステムは は現在作成中である。

2）受診時平均年齢は 28±14 歳。管理移行 57 名(50%)、治療およびセカンドオピニオン 42 名(37%)、職場または学校健診 4 名(4%)であった。治療およびセカンドオピニオン目的で紹介受診した 42 名中 18 名(43%)は県外からの紹介であった。

3）QOL の尺度 SF-36 について対象 73 名

（男性 41、女性 32）の結果を国民標準値と比較した。全体に精神的健康度が低く、特に 20 才代女性は低かった。また、79.2%が疾患に関係する不安・心配をもち、22.4%は不整脈・突然死に関する不安だった。これら精神的健康度は疾患重症度とは関連なかった。

4．結論

ACHD 専門外来を受診する患者の半数は成人期に達したための管理移行であった。成人先天性の専門的な知識を必要とする複雑心疾患、あるいは姑息術のみしか施行されていない患者など成人期での治療方針決定が困難な例も多い。精神的健康度が低く、身体的社会的問題を多く抱える患者に対して相談や心理的支援も重要といえる。遠方からの紹介も多く、今後 ACHD を専門とする医師の養成と日本各地での ACHD 診療施設の構築が必要である。今後、新たな成人患者数が増加するとともに、診療体制の確立、専門医療従事者の育成が急務である。これと同時に、数少ない専門施設、専門医師との情報共有と診断、治療方針の決定を行うために、今後、遠隔医療支援システムを有効に用いることが望まれる。また欧米では小児科から成人診療施設への移行期に CHD 患者の 1/2～3/4 が受診しなくなっているという調査結果があり、今後、受診中断の実態調査や医療チームの支援内容の明確化なども必要と考えられる。

5．研究成果

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11. 知的所有権の出願・取得状況

なし。

研究成果の刊行に関する一覧表

書籍

著者氏名	論文タイトル名	書籍全体の編集者名	書　籍　名	出版社名	出版地	出版年	ページ
丹羽公一郎	成人先天性心疾患	五十嵐隆	小児科学レビュー2010	総合医学社	東京	2010	42-48
丹羽公一郎、立野滋	成人における先天性心疾患	川名正敏、北風政史、小室一成、	カラー版循環器病学　基礎と臨床	西村書店	東京	2010	935-957
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雑誌

発表者氏名	論文タイトル名	発表誌名	巻号	ページ	出版年
丹羽公一郎	成人先天性心疾患の最新動向、頻度、今後の	医学のあゆみ	232	775-778	2010
丹羽公一郎、川副泰隆	心疾患の妊娠出産の適応、管理に関するガイ	周産期医学	40	495-498	2010
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Silversides CK, Kiess M, Beauchesne L, Bradley T, Connelly M, Niwa K, Mulder B, Webb G, Colman J, Therrien J	Canadian cardiovascular society 2009 consensus conference on the management of adults with congenital heart disease: outflow tract obstruction, coarctation of the aorta, tetralogy of Fallot, Ebstein anomaly and Marfan's syndrome.	Can J Cardiol	26	e80-97	2010
Silversides CK, Marelli A, Beauchesne L, Dore A, Kiess M, Salehian O, Bradley T, Colman J, Connelly M, Harris L, Khairy P, Mital S, Niwa K, Oechslin E, Poirier N, Schwerzmann M, Taylor D, Vonder Muhll I, Baumgartner H, Benson L, Celermajer D, Greutmann M, Horlick E, Lanzberg M, Meijboom F, Mulder B, Warnes C, Webb G, Therrien J.	Canadian cardiovascular society 2009 consensus conference on the management of adults with congenital heart disease: executive summary.	Can J Cardiol.	26	143-150	2010
Shiina Y, Toyoda T, Kawasoe Y, Tateno S, Shirai T, Wakisaka Y, Matsuo K, Mizuno	Prevalence of adult patients with congenital heart disease in Japan.	Int J Cardiol.	146	13-6	2011

Kaemmerer H, Mebus S, Schulze-Neick I, Eicken A, Trindade PT, Hager A, Oechslin E, Niwa K, Lang I, Hess J.	The adult patient with Eisenmenger Syndrome: A medical update after Dana Point. Part I: Epidemiology, Clinical Aspects and Diagnostic Options.	Current Cardiology Reviews.	6	343-355	2010
Mebus S, Schulze-Neick I, Oechslin E, Niwa K, Trindade PT, Eicken A, Hager A, Lang I, Hess J, Kaemmerer H.	The adult patient with Eisenmenger Syndrome: A medical update after Dana Point. Part II: Medical treatment-study results.	Current Cardiology Reviews.	6	356-362	2010
Oechslin E, Mebus S, Schulze-Neick I, Niwa K, Trindade PT, Eicken A, Hager A, Lang I, Hess J, Kaemmerer H.	The adult patient with Eisenmenger Syndrome: A medical update after Dana Point. Part III: Specific management and surgical aspects.	Current Cardiology Reviews.	6	363-372.	2010

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Prevalence of adult patients with congenital heart disease in Japan

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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 the1970s

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.

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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.

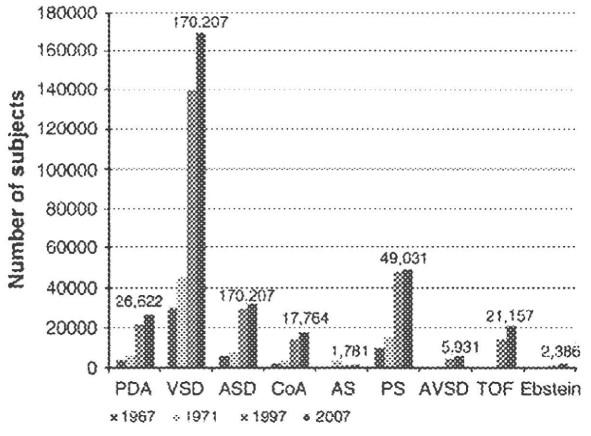

Fig. 1. The number of adults with major CHD from 1967 to 2007. This figure shows that adults with major CHD in 1967, in 1971, in 1997 and in 2007, consecutively. Number of adults with major CHD is increasing explosively.

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

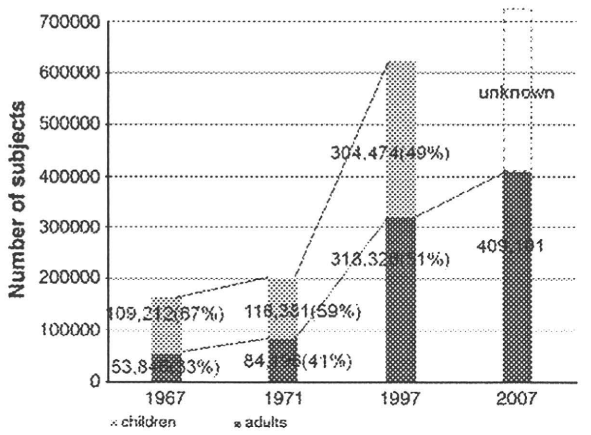

Fig. 2. The total number of patients with CHD from 1967 to 2007. This figure shows that the total number of patients with CHD in 1997 is much larger than that in 1967 with remarkably higher ratio of adults/children.

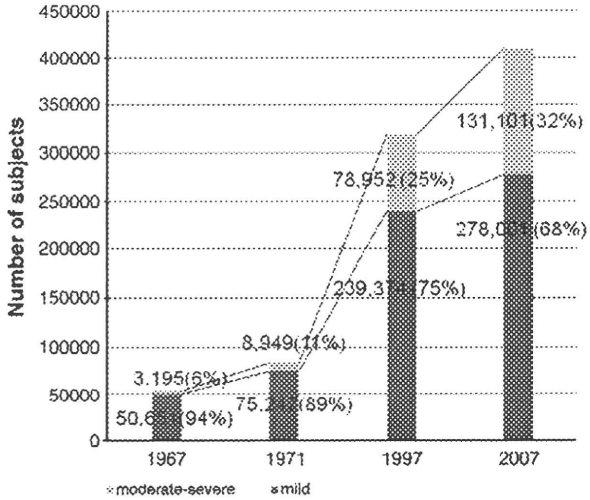

Fig. 3. The number and percentage of adults with CHD according to the severity of CHD from 1967 to 2007. This figure shows that the number and percentage of adults with moderate to severe CHD have been increasing dramatically from 1967 to 2007.

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

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Current Issue in the Management of Adult Congenital Heart Diseases in Asia Pacific

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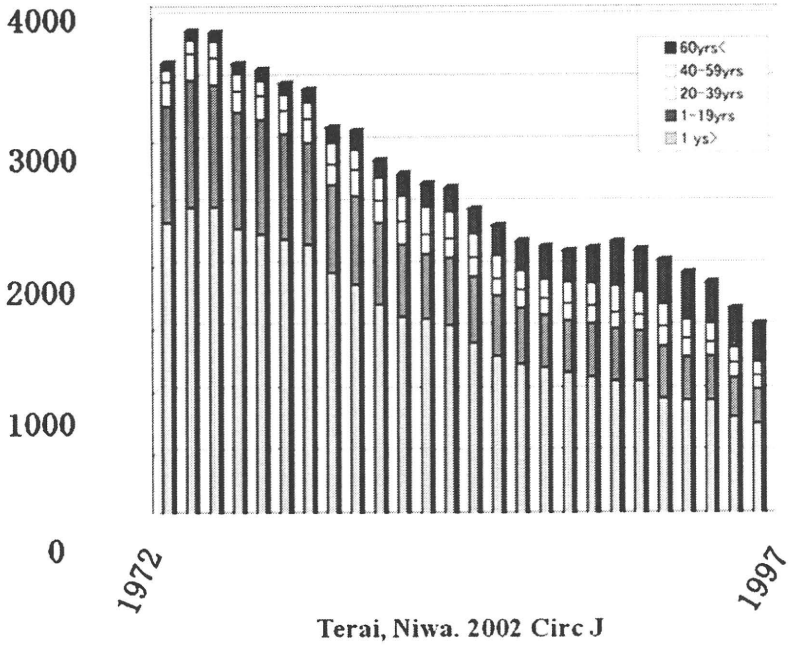
Cumulative mortality from congenital heart disease



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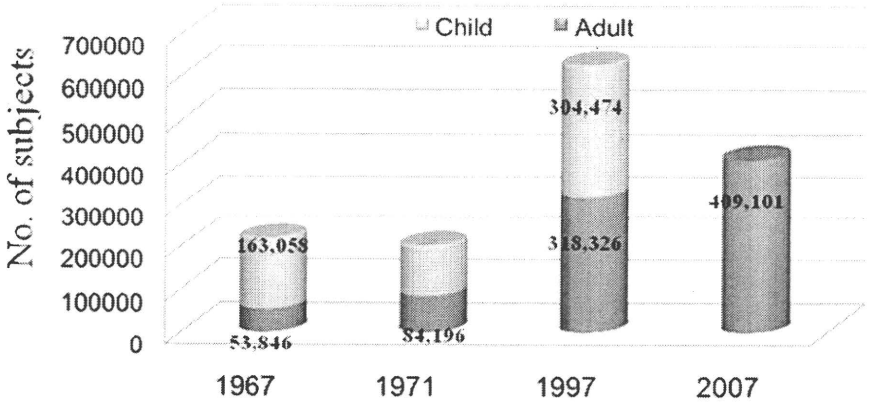
No of ACHD patients in Japan

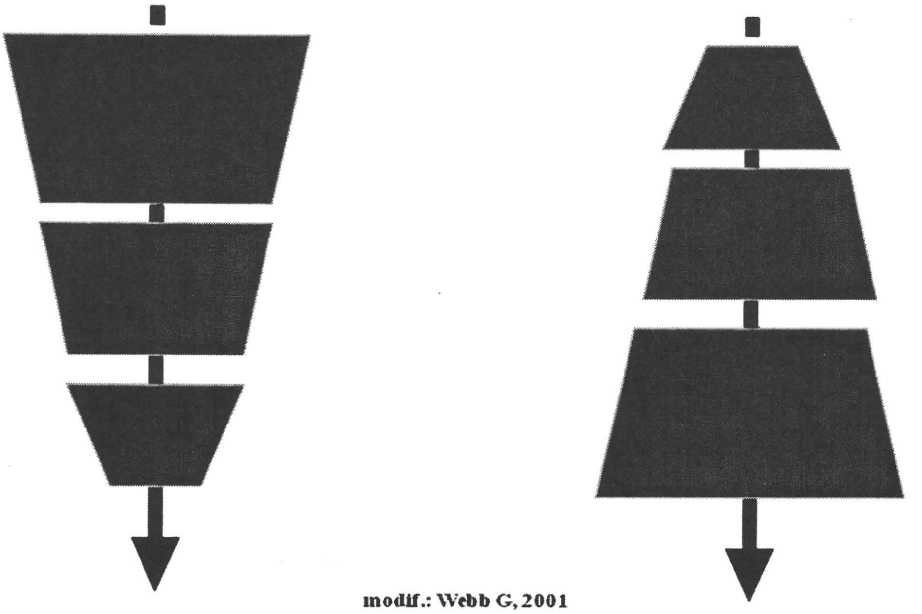


(Shiina Y, Niwa K. IJC 2009)

Changing profile of congenital heart disease

Children with CHD

Adults with CHD



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Prevalence of ACHD in the general population

Country	Group	Population (year 2000)	Prevalence/ 1000 population		Total ACHD (year 2000)		
			Severe	Others	Severe	Others	Total
US	Children	72,293,812	1.45	10.44	104,826	754,747	859,573
	Adults	209,128,094	0.38	3.71	79,469	775,865	855,334
Canada	Children	7,137,778	1.45	10.44	10,350	74,518	84,868
	Adults	23,551,257	0.38	3.71	8,949	87,375	96,324

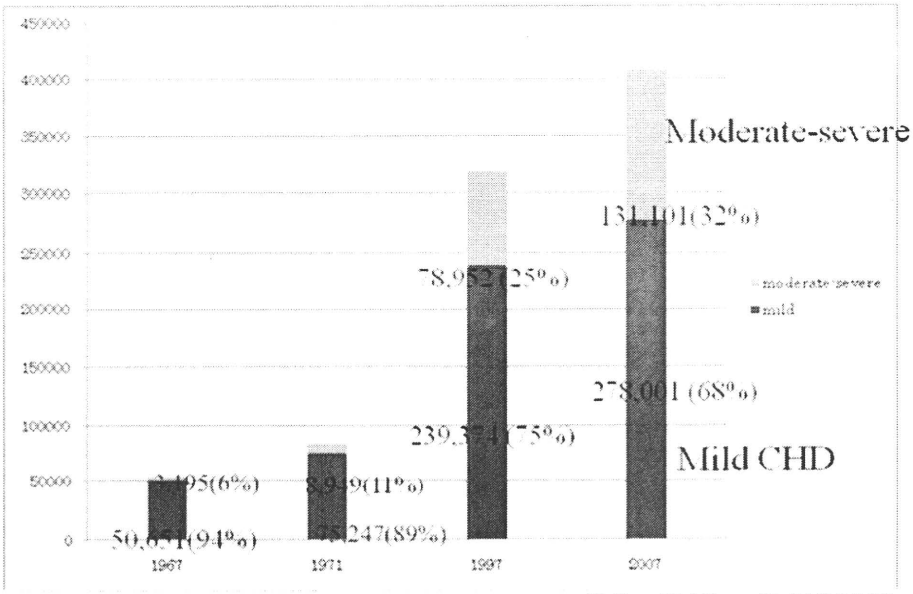
Marelli AJ et al.AHJ2009;157(1):1-7

Japan Adults 104,479,000 409,101

Proportion of ACHD in Asian Countries Survey Asia Pacific Society of ACHD

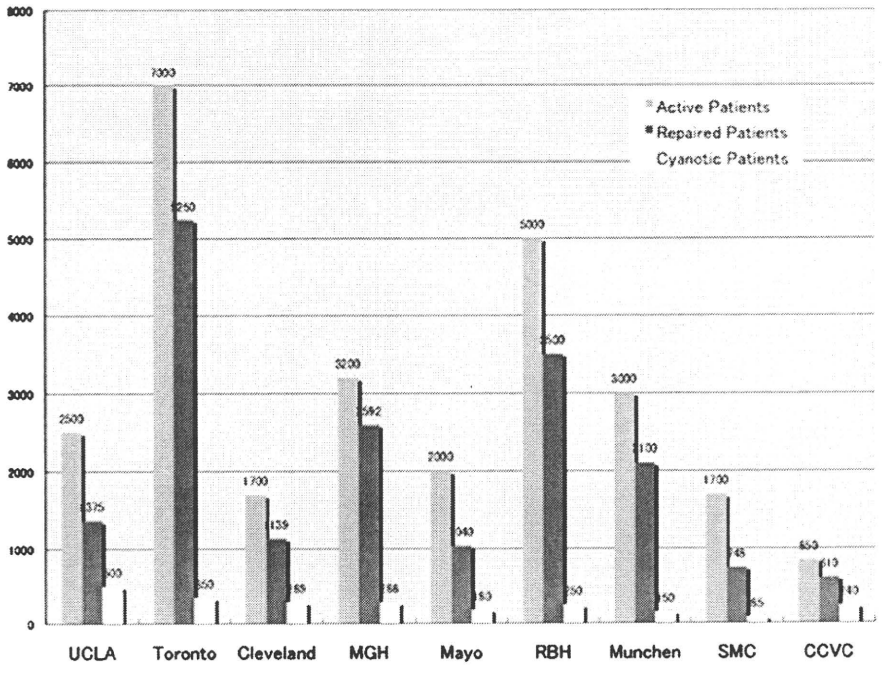
Heung Jae Lee. 1st APACHD 2008, Jeju)

ACHD according to the severity from 1967 to 2007



Shiina Y, Niwa K. IJC 2009

Patients in Adult CHD Facilities



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