

Nationwide Survey of Care Facilities for Adults With Congenital Heart Disease in Japan

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Background: The number of adults with congenital heart disease (CHD) is increasing rapidly, but care programs have not been fully established in Japan.

Methods and Results: Questionnaires regarding current status and resources of outpatient and in-hospital services, and management of pregnancy in patients with adult CHD (ACHD) were sent to 1,033 training hospitals for board-certified cardiologists. Useful replies were obtained from 458 hospitals (44%). In 417 hospitals (91%), at least 1 patient was followed in the outpatient clinic; however, only 14 hospitals (3%) had specialized outpatient clinics; 354 hospitals (77%) had in-hospital patients, but only 6 hospitals (2%) admitted >50 patients per year. Surgery for ACHD was performed in 232 hospitals (51%), but in 135 of these (58%), the number of operations was <5 per year. Pregnant women with CHD were managed in 157 hospitals (34%), although only 3 hospitals (2%) managed >10 cases per year.

Conclusions: In most hospitals in Japan, a limited number of ACHD patients have been followed up and specialized multi-disciplinary facilities for ACHD need to be established.

Key Words: Care programs; Congenital heart disease; Japan

Advances in diagnosis, medical treatment, and surgical repair have dramatically changed the survival patterns in congenital heart disease (CHD). Surgical mortality was 60–70% in the decade following the first intracardiac operation for CHD in 1952¹. Currently, surgical mortality in large CHD centers is 10% or less,² and more than 85% of babies born with cardiovascular anomalies can now expect to reach adulthood^{3,4}.

In Japan, the mortality rate from CHD declined from 3.7 per 100,000 persons in 1968 to 1.2 in 1997. Among the population aged 1–19 years, mortality declined from 3.1 per 100,000 population in 1968 to 1.1 in 1997. In contrast, among adults aged >20 years, that rate increased from 0.4 per 100,000 population in 1968 to 0.5 in 1997⁵. The adult population with CHD is estimated at more than 400,000, with a rate increase of 4–5% per year⁶.

Patients with at least moderate CHD are likely to undergo 1 or more surgical interventions³. With rare exception, notably patent ductus arteriosus, surgery is not curative, so most if not all operated patients have postoperative residua and sequelae that require life-long follow-up^{7–10}.

Cyanotic CHD in adults is a multisystem disorder^{11,12}. Risk stratification of pregnancy should be undertaken in virtually all women with CHD^{13,14}. Adult CHD (ACHD) patients

are particularly susceptible to common psychiatric illnesses, so appropriate screening and referral resources for management and follow-up must be available^{15,16}.

In 2000, the “Guidelines for Management of Congenital Heart Disease in Adults” were published by the Japanese Circulation Society (JCS), and revised in 2006^{6,17}. However, facilities for the care of ACHD have not yet been organized in Japan^{18,19}. This nationwide survey was therefore designed to investigate the status and function of care facilities for ACHD in Japan.

Methods

This study was conducted by the research group of the Japanese Society of Adult Congenital Heart Disease. Questionnaires regarding current status and resources of outpatient and inpatient services and management of pregnancy in ACHD patients were sent in May and June, 2007, to 1,033 training institutions, including 928 hospitals that were authorized by the JCS as “Training Hospitals for Board-Certified Members,” 175 hospitals to which councils of the Japanese Society of Pediatric Cardiology and Cardiac Surgery belonged, and 442 hospitals that were authorized by the Japanese Board of Cardiovascular Surgery as “Training Hospitals for Board-Certified Members of the Japanese Board of Cardiovascular Surgery”. If an institution had 2 or 3 board-certified members, for example, cardiology and cardiovascular surgery, their answers were merged.

The survey included: (1) outpatient services—number of patients, whether or not there is a special clinic for ACHD; (2) in-hospital services—number of hospitalized patients; (3) cardiac surgery for these patients—number of operations; (4) management of pregnancy and/or delivery—number of patients; and (5) whether the cardiologists recognized the JCS guidelines. The data were collected and analyzed at

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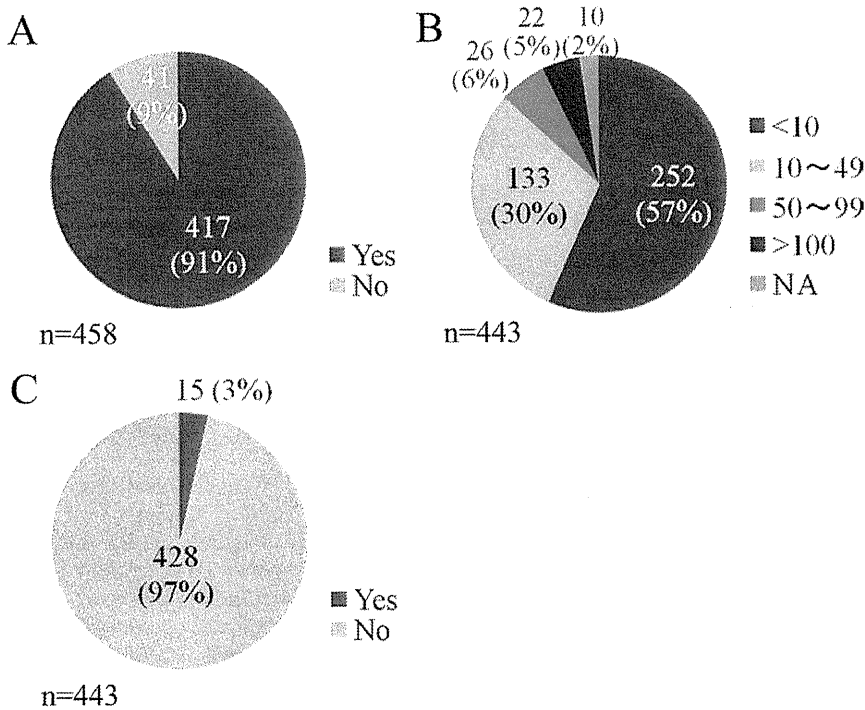


Figure 1. Outpatient services. (A) Number of hospitals which followed up adult congenital heart disease patients. (B) Number of patients who were followed up in each institution. (C) Number of hospitals which had specialized clinic. NA, not available.

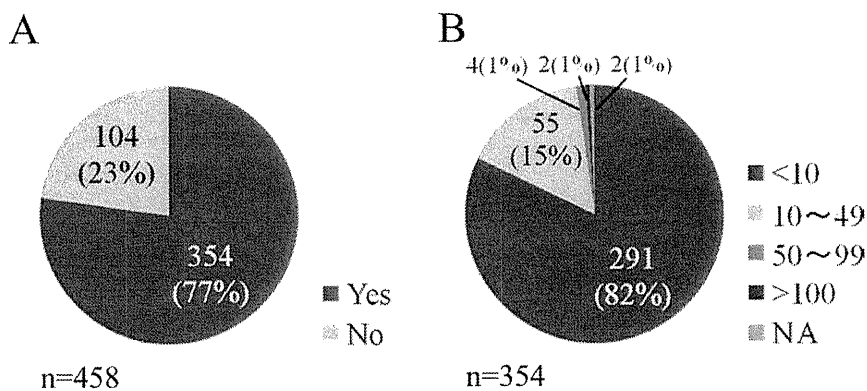


Figure 2. In-hospital services. (A) Number of hospitals admitting adult congenital heart disease patients. (B) Number of patients admitted to each hospital per year. NA, not available.

the Department of Adult Congenital Heart Disease and Pediatric Cardiology, Chiba Cardiovascular Center.

Ethical Considerations

This study did not deal with data from the medical records of individual patients, but conformed to the 2004 revised version of the Ethical Guidelines of Epidemiological Study by the Ministry of Education, Culture, Sports, Science and Technology and the Ministry of Health, Labor and Welfare of Japan.

Results

Satisfactory replies were obtained from 458 hospitals (44% of all solicited hospitals).

Outpatient Services

Data are summarized in **Figure 1**: 91% of the hospitals had outpatient clinics for ACHD, but 57% of these clinics followed less than 10 patients. Moreover, only 15 hospitals (3%) had specialized outpatient clinics.

In-Hospital Services

Data are summarized in **Figure 2**: 77% of the hospitals admitted ACHD patients over the course of 1 year, but 82% had less than 10 annual hospitalizations. Only 6 hospitals (2%) had more than 50 hospitalizations per year.

Cardiac Surgery

Data are summarized in **Figure 3**: 51% of the hospitals performed cardiac surgery for ACHD, although 81% had less than 10 cases per year. Only 6 hospitals (3%) had more than 20 cases per year.

Pregnancy and Delivery

Data are summarized in **Figure 4**: 34% of the hospitals had experience in the management of pregnancy and/or delivery in ACHD patients; however, 91% managed less than 10 cases per year.

Penetration Rate of Practice Guidelines

In 289 hospitals recognized the JCS guidelines, making a penetration rate of 63%.

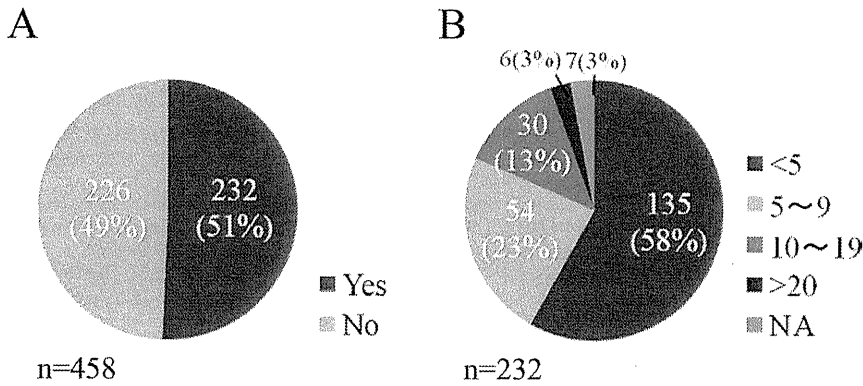


Figure 3. Cardiac surgery. (A) Number of hospitals in which surgery for ACHD was performed. (B) Number of cases of cardiac surgery for ACHD in each institution. ACHD, adult congenital heart disease; NA, not available.

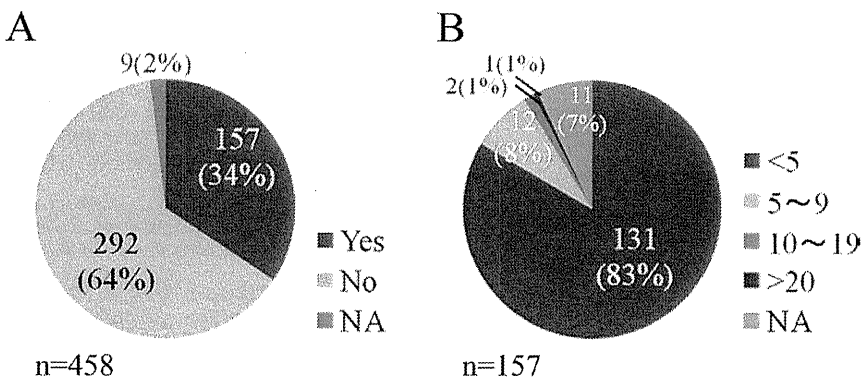


Figure 4. Management of pregnancy and/or delivery. (A) Number of hospitals managing pregnancy and/or delivery in adult congenital heart disease (CHD) patients. (B) Number of managed pregnant women with CHD per year. NA, not available.

Discussion

This is the first nationwide survey of ACHD care facilities in Japan. Most hospitals have been following a limited number of patients. In one-third of surveyed hospitals, pregnancy in women with CHD is managed, and in half of these hospitals, surgery for these patients is regularly undertaken. However, the number of hospitals in Japan with a large volume of the patients remains small. Special care systems and training programs should be established in Japan without delay.

Need for Specialized ACHD Care Facilities

The majority of patients with CHD now reach adulthood, so multidisciplinary facilities for the comprehensive care of this new patient population have emerged, including congenital cardiac imaging techniques, diagnostic and interventional catheterization, congenital cardiac surgery and anesthesia, heart failure management, transplantation, electrophysiology, reproductive and high-risk pregnancy services, genetics, pulmonary hypertension, hepatology, nephrology, hematology, and psychiatry.^{3,13,20-22}

More than 90% of the hospitals in this study followed ACHD patients in their outpatient clinics, but more than half followed less than 10 patients. More than 70% of surveyed hospitals had admitted patients in a year, but 82% had less than 10 hospitalizations per year. Many patients were probably managed by physicians who were not familiar with CHD.²³ Because of the complexities inherent in the comprehensive care of ACHD, especially patients with complex malformations, specialized tertiary care facilities have been developed in North America, Europe, and in Japan,^{24,25} but currently, only 15 hospitals in Japan have specialized outpatient clinics.

Cardiac Surgery and Pregnancy

Mortality is higher in centers with relatively low volume of surgical cases, highlighting the risk of performing the occasional operation on ACHD patients.²⁶ Surgery for most simple malformations can be undertaken in centers with relatively low volumes, but surgical outcomes are better when surgery is centralized to high-volume facilities. In this survey, less than 5 operations per year were undertaken in over 60% of hospitals that performed cardiac surgery in ACHD patients.

Most female patients can tolerate pregnancy under specialized care, but heart failure, arrhythmias, and thromboembolic complications are not rare during pregnancy, delivery and postpartum in these women.²⁷⁻²⁹ Patients at high risk should be managed in a high-risk perinatal facility staffed by a multidisciplinary team including an obstetrician, cardiologist, anesthesiologist, and pediatrician.³ In the current survey, only 3 hospitals (1%) had managed more than 10 pregnancies per year.

Recommended Guidelines for Management of ACHD

In the Canadian Cardiovascular Society Consensus Conference on Adult Congenital Heart Disease, full service centers that provide care to adults with congenital or heritable cardiovascular disease serve populations of 3-10 million. Regional specialized centers provide care to patients within the constraints of available resources, serving populations of up to 2 million.³⁰⁻³³ In the United States, the 32nd Bethesda Conference on Care of the Adult with Congenital Heart Disease organized by American College of Cardiology stated that there should be approximately 1 regional center per population of 5-10 million.³⁴ In the European Guidelines for Management of Grown Up Congenital Heart Disease, each specialized center served approximately 5-10 million

people, as in the United States.¹⁴

The JSC guidelines have not specified the desirable number of specialized centers.^{6,17} More than 20 centers are believed to be required based on the guidelines from other countries. In the current survey, 15 hospitals had a specialized outpatient clinic for ACHD. The data from this survey suggest that 20 specialized centers might exist in Japan, but collaborative care, even in these centers, has not yet been fully developed. Recommendations are needed regarding the number of pediatric or medical cardiologists dedicated to the care of this patient population, and the number of nurse specialists, residents or consultants for non-cardiac problems and educational programs.

The establishment of new specialized ACHD centers is necessary in addition to improvement of local facilities. Because most general cardiologists have virtually no experience in or understanding of the management of these patients, adult cardiology training programs should provide experience in CHD. Specialized centers should offer educational opportunities to general cardiologists so that they can contribute optimally to patient management.³⁵

As for recognition of the JSC guidelines, 63% hospitals answered positively, so circulation of the guidelines should be encouraged in order to improve and standardize management.

Conclusions

Most hospitals in Japan have been following limited numbers of ACHD patients, but specialized care for this expanding patient population has not been effectively organized. The establishment of multidisciplinary facilities for ACHD is mandatory to assure optimal prognosis for these patients.

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総括報告書

循環器疾患等生活習慣病対策総合研究事業
(研究課題名)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
(H21—循環器等(生習)—一般—016)
分担研究報告

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

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研究要旨

【目的】本邦の循環器内科における成人先天性心疾患診療の実態と今後の診療の意向を明らかにし、循環器内科を含めてチーム医療を提供できる集約化施設の候補施設を特定すること

【方法】今後、成人先天性心疾患診療において中心的な役割果たすことが期待される、循環器内科を対象に全国調査を行った。全国138の主要施設の循環器内科に調査票を郵送し、各施設の循環器内科における、現在の成人先天性心疾患の診療の実態、今後の診療に対する意向、成人先天性心疾患集約化施設に必要とされる施設基準の充足状況を尋ねた。

【結果】109施設(回収率79%)から回答を得た。回答の得られた施設のうち、現在、循環器内科において重症度の高い患者を含め全ての成人先天性心疾患患者を診療していると回答した施設は34施設、今後、全ての患者を診療する意向があると回答した施設は39施設であった。成人先天性心疾患診療集約化に必要とされる施設基準を全て満たし、今後、集約化施設として機能しうる施設は全国に14あったが、全国8つの地方のうち、2つの地方には集約化施設の候補となる施設が存在しなかった。全国平均では人口910万人につき1つの集約化施設が配備可能であることが示された。

【結論】本研究は、循環器内科における成人先天性心疾患診療の実態を明らかにし、集約化施設の候補となる施設を特定した本邦初の調査である。人口あたりの集約化施設の候補施設数は、欧米のガイドラインの基準を満たすものであった。しかし、2つの地方には候補施設が存在せず、成人先天性心疾患診療には地域格差がある可能性が示唆された。今後は、集約化候補施設における成人先天性心疾患診療チームの中心的役割を担う医師を育成するための教育体制整備が望まれる。また、本研究で指摘された成人先天性心疾患診療における地域格差については、診療機能の集約化と同時に、地域の医療機関と都市部の医療機関の連携体制整備が必要である。

研究目的

医療の発達により、成人期を迎える先天性心疾患患者(以下、成人先天性心疾患患者)が増えてきた。国内の成人先天性心疾患患者数は40万人と推定され、今後年

間9千人の増加が見込まれる。患者数の増加に伴い、欧米では成人先天性心疾患の診療体制に関するガイドラインが策定されており、これらのガイドラインが定める診療

体制の中心的役割を担うのが、**regional ACHD center** や **specialist center** と呼ばれる成人先天性心疾患を専門的に診療する地域中核施設(以下、集約化施設)である。集約化施設は、多職種医療を提供し、患者の生涯にわたるすべてのニーズに応えうる施設と定義されている。

欧米の集約化施設の多くは、循環器内科を背景とする医師を中心に運営されている。他方、日本を含めたアジア諸国では、成人先天性心疾患患者の多くは小児循環器内科医によるフォローアップを受けており、欧米とアジアでは診療体制に差異が見られる。しかし、我々が以前行った研究結果からは、成人先天性心疾患診療に関わる本邦の医師の多くが、我が国でも欧米同様、循環器内科医が成人先天性心疾患診療の中心的役割を担うべきと認識していることが指摘されている。成人先天性心疾患患者が成人期に直面することの多い内科疾患への対応や、小児科医不足という本邦の現状を考えると、循環器内科医の成人先天性心疾患分野への参加は、今後不可欠となると考えられる。

しかし、現在どの程度の循環器内科医が成人先天性心疾患診療に従事しており、また、将来的にどの程度診療していく意向を有しているのかは明らかにされていない。そこで我々は今回、本邦の循環器内科における成人先天性心疾患診療の実態と今後の診療の意向を明らかにし、循環器内科を含めてチーム医療を提供できる集約化施設の候補施設を特定することを目的に、全国の循環器内科を対象とした質問紙調査を行った。

研究方法

1. 対象施設

下記のいずれかの適格基準を満たす全国 138 施設の循環器内科診療科長宛に、質問紙を郵送した。

- 1)大学病院である施設
- 2)成人先天性心疾患年間外来患者数 50 人以上の施設
- 3)成人先天性心疾患専門外来を有する施設

設

回答は診療科長本人、または同科に所属する医師のうち、成人先天性心疾患診療に興味を持つ医師に依頼した。調査票は郵送にて回収した。

2. 調査項目

循環器内科における成人先天性心疾患診療の実態として、「現在、循環器内科において、どの重症度の成人先天性心疾患患者まで診療しているか」、今後の診療の意向として、「今後、循環器内科において、どの重症度の成人先天性心疾患患者まで診療していく意向があるか」をそれぞれ尋ねた。選択肢はそれぞれ「重症度の高い患者を含め全ての患者まで」「中等度の患者まで」「軽度の患者まで」「診療していない(する意向はない)」の 4 件法とした。また循環器内科における成人先天性心疾患診療に関連する項目として、循環器内科が主体となった成人先天性心疾患専門外来設置に対する意向、成人先天性心疾患を専門とする医師養成に対する意向を尋ねた。

さらに、集約化施設の候補となる施設を特定するために、下記の施設基準の充足状況を尋ねた。循環器内科が今後、「重症度の高い患者を含め全ての患者まで」診療していく意向を有していること、小児心臓血管外科医が 2 名以上いること、小児循環器内科医が 1 名以上いること、現時点で成人先天性心疾患外来(小児科・循環器内科主体を問わず)がある、または将来設置の意向があること、カテーテル・不整脈・エコーを専門とする循環器内科医がいること、成人心疾患患者に対しカテーテル検査を年間 500 件以上・アブレーションを年間 20 件以上・ペースメーカー植え込み術を年間 20 件以上・植え込み型除細動器埋め込み術を年間 10 件以上実施していること、カルトエンサイトシステム・MRI・3DCT 等の設備を有すること、産科・精神科といった診療科が併設されていること。これらの施設基準の設定にあたっては、欧米のガイドライン、および先行研究の基準を踏襲した。

3. 分析

各項目について記述統計量を算出した。集約化施設の基準を満たす候補施設を特定後、全国、そして8つの地方毎に、候補施設あたりの人口を算出した。統計解析にはSPSS ver.16.0を用いた。

4. 倫理的配慮

本調査は東京大学大学院医学系研究科・医学部倫理委員会の承認を得て実施した。調査票への回答にあたっては、個人情報第三者に明かさなないこと、今後の診療に影響を及ぼさないことを文書にて対象者に説明し、調査票の返送をもって対象者の同意とした。

研究結果

1. 対象施設背景

調査票を郵送した138施設中、109施設から回答を得た(回収率 79.0%)。回答のあった施設のうち、83施設(76.1%)が大学病院、20施設(18.3%)が総合病院、6施設(5.5%)が循環器専門施設であった。

2. 循環器内科における成人先天性心疾患診療

循環器内科における成人先天性心疾患診療に関する結果を示す。109施設のうち、34施設(31.2%)が、現在、循環器内科において重症度の高い患者を含め全ての成人先天性心疾患患者を診療していると回答した。今後の診療については、37施設(33.9%)が、今後、循環器内科において重症度の高い患者を含め全ての成人先天性心疾患患者を診療していく意向があると回答した。

循環器内科が主体となった成人先天性心疾患専門外来設置に対する意向については、11施設(10.1%)が既に専門外来を設置していると回答し、10施設(9.2%)が、今後、設置の意向があると回答した。成人先天性心疾患を専門とする医師養成に対する意向については、23施設(21.1%)が養成の意向があると回答した。

3. 集約化候補施設の特定

集約化施設の施設基準の充足状況を示す。14施設(12.8%)が全ての施設基準を満たした。全項目のうち、対象施設の半数以

下しか施設基準を満たすことができなかった項目は、循環器内科が今後、「重症度の高い患者を含め全ての患者まで」診療していく意向を有していること(充足率 33.9%)、小児心臓血管外科医が2名以上いること(充足率 37.6%)、現時点で成人先天性心疾患外来(小児科・循環器内科主体を問わず)がある、または将来設置の意向があること(充足率 35.7%)の3つであり、この3つを全て満たす施設は19施設のみであった。

全ての施設基準を満たした14施設の地理的分布については、全国8つの地方のうち、6つの地方には、集約化施設の候補となる施設が1施設以上存在し、各地方における1候補施設あたりの人口は、690万人から2360万人であった。一方で、北海道と四国には、候補となる施設が存在しなかった。日本全国における1候補施設あたりの人口は910万人であった。

考察

本調査は、循環器内科における成人先天性心疾患診療に着目した本邦初の調査である。本調査において我々は、全国の主要施設の循環器内科を幅広くリクルートするよう努めた。本調査の回収率は80%弱であり、この数字は欧米の同様の研究と比較しても同等かそれ以上である。このことから、本研究の結果は、本邦における成人先天性心疾患診療の実態を一定程度、反映していると考えられる。

1. 循環器内科における成人先天性心疾患診療

先行する欧州の調査では、集約化施設の90%に、循環器内科を背景とする医師が配置されていることが報告されている。他方、本調査では、成人先天性心疾患診療に積極的な姿勢を有する循環器内科は全体の三分の一に留まっていた。本邦においては成人先天性心疾患診療に積極的な姿勢を有する循環器内科は限られているというのが現状であろう。今後は、成人先天性心疾患に興味を持つ循環器内科医の絶対数を増やすために、循環器内科医を対象としたセミナー・研修の充実が求められる。

循環器内科が成人先天性心疾患診療に積極的な姿勢を有している施設においては、今後、循環器内科と、小児循環器内科、小児心臓血管外科等が連携して、成人先天性心疾患診療におけるチーム医療のモデルケースを確立することが求められる。また、23施設の循環器内科が、成人先天性心疾患専門医を育成していく意向を示しており、今後は、成人先天性心疾患診療におけるチーム医療の中心的役割を担う医師を育成するための教育体制整備も望まれる。近年、欧米では成人先天性心疾患専門医の教育プログラムが整備されつつあり、本邦においても、医療制度や文化的背景を考慮した教育プログラムの策定が急務である。

2. 集約化施設の確立とネットワーク構築

我々が欧米のガイドライン、および先行研究の基準をもとに作成した施設基準のうち、充足率が低かった項目は、循環器内科が今後、「重症度の高い患者を含め全ての患者まで」診療していく意向を有していること、小児心臓血管外科医が2名以上いること、現時点で成人先天性心疾患外来(小児科・循環器内科主体を問わず)がある、または将来設置の意向があることの3つであった。このうち、1つ目の循環器の診療の意向と3つ目の専門外来設置の有無に関しては、循環器内科の意向が大きく反映される項目である。しかし2つ目の小児心臓血管外科医の配置状況に関しては、循環器内科の問題と言うよりは、小児心臓血管外科の問題と言えるだろう。成人先天性心疾患患者は再手術を必要とする者が多く、さらに、小児心臓血管外科医と成人の心臓血管外科医が成人先天性心疾患に対する手術を行った場合、成人の心臓血管外科医が行った方が、病院内死亡率が高いという報告がある。これらのデータからも、小児心臓血管外科医の存在は、成人先天性心疾患集約化施設に不可欠と考えられる。今後、集約化施設を選定していく際、小児心臓血管外科医の存在は、最も重要な施設基準の1つ

として認識されるべきだろう。

調査票に回答した109施設のうち、全ての施設基準を満たし、集約化施設の候補となりうる施設は全国に14施設あった。日本全国で考えると、人口910万人につき1つの集約化施設がある計算になる。この910万人という数字は、人口300万～1000万につき1つの集約施設が必要とする欧米のガイドラインの基準を、かろうじて満たすものである。候補施設の地理的分布に目を向けると、施設は概ね地方の人口にそって分布しており、人口が最も少ない北海道と四国においては、候補施設が存在しないという結果であった。これまで成人先天性心疾患診療体制整備は集約化という観点から語られることが多かったが、人口の少ない地方においては、集約化を担いうる施設が現状では存在せず、集約化は現実的とは言えない可能性がある。今後、人口の少ない地方においては、ウェブベースの遠隔医療体制を整備し、都市部の医療機関との連携により診療の質を担保するといった対応が求められよう。

結論

本研究は、循環器内科における成人先天性心疾患診療の実態を明らかにし、集約化施設の候補となる施設を特定した本邦初の調査である。人口あたりの集約化施設の候補施設数は、欧米のガイドラインの基準を満たすものであった。しかし、2つの地方には候補施設が存在せず、成人先天性心疾患診療には地域格差がある可能性が示唆された。今後は、集約化候補施設における成人先天性心疾患診療チームの中心的役割を担う医師を育成するための教育体制整備が望まれる。また、本研究で指摘された成人先天性心疾患診療における地域格差については、診療機能の集約化と同時に、地域の医療機関と都市部の医療機関の連携体制整備が必要である。

健康危険情報

該当せず。

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知的財産権の出願・登録状況

なし



Status and Future Needs of Regional Adult Congenital Heart Disease Centers in Japan

— A Nationwide Survey —

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Background: Although the prevalence of adult congenital heart disease (ACHD) in Japan continues to rise, the number and geographic distribution of facilities potentially serving as regional ACHD centers remains unknown. We examined trends in ACHD care in Japan to identify needs and to determine potential regional responses to this growing patient population.

Methods and Results: A descriptive, cross-sectional, nationwide survey was conducted to assess the status and needs of cardiology specialists related to providing ACHD care. Questionnaires were mailed to 138 cardiology departments located in 8 geographical regions throughout Japan; respondents were asked to document the status and future direction of ACHD care for each facility. Of the 109 facilities that responded, approximately one-third currently treat or plan to treat all ACHD patients. Fourteen facilities (12.8%) fulfilled all criteria for becoming regional ACHD centers. Although each regional center was projected to serve a population of 9.1 million, in 2 regions, no centers possessed the necessary care structure.

Conclusions: Our findings revealed a shortage of adult cardiologists dedicated to ACHD care. Moreover, basic as well as formal fellowship ACHD training was deemed necessary. In Japan, the number of potential regional ACHD centers has just reached international standards. However, based on the geographic gaps documented here, a strategy other than regional centralization might be required to deliver adequate ACHD care to rural areas. (*Circ J* 2011; **75**: 2220–2227)

Key Words: Adult congenital heart disease; Congenital heart disease; Physician training; Regional ACHD centers; Rural medicine

Advances in medical treatment, cardiac surgery, intensive care, and non-invasive diagnosis over the last 50 years have led to enormous growth throughout the world in the number of adults with congenital heart disease (CHD).^{1–4} In Japan, it is estimated that there are at least 400,000 adult CHD (ACHD) patients; this number is expected to increase by approximately 9,000 patients annually.⁵ In response to the increasing numbers of ACHD patients globally, several North American and European task forces, as well as numerous expert panels, have developed “best practice” recommendations for the management of ACHD patients.^{6–15}

At the core of these recommendations, the “regional ACHD

center,” also called a “specialist center” in Europe, is expected to play a key role in coordination of the ACHD care delivery system.^{10,13,15,16} Regional ACHD centers are large, multidisciplinary centers of excellence that are capable of meeting all potential needs of ACHD patients over their lifetimes. In Europe and North America, a regional ACHD center is usually directed by an ACHD cardiologist with an adult cardiology background.^{17,18} Alternatively, in Japan, most ACHD patients are followed by pediatric cardiologists,¹⁹ even though our previous qualitative study revealed that many Japanese physicians dedicated to ACHD care expected adult cardiologists to play the central role in treating ACHD.²⁰ Thus, it appears that recruiting adult cardiologists to ACHD care is

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Table 1. Recommendations for Optimal ACHD Care^{6,8*}

1. An ACHD referral center must employ at least 1, preferably 2, cardiologist(s) specifically trained and educated in the care of adults with CHD.
2. Specialized ACHD centers should provide care in connection with pediatric cardiology and/or congenital cardiac surgery.
3. Specialist centers[†] must treat sufficient numbers of patients and perform a sufficient number of procedures to be effective as well as develop and maintain high levels of performance.
4. General adult cardiac facilities and non-specialist centers should have an established referral relationship with a specialist center.
5. A minimum of 2 cardiac surgeons trained in and practicing adult and pediatric cardiac surgery are required.
6. The optimal activity for a pediatric and congenital heart surgeon is 125 operations per year. Specifically, for ACHD, a minimum of 50 operations per year is recommended.
7. A fully equipped electrophysiology laboratory staffed by properly trained electrophysiologists with experience in detecting arrhythmias inherent to CHD and with experience in pacemaker technology, ablation technology, and defibrillator implantation must be available.
8. An ACHD referral center must employ at least 1 nurse specialist that is trained and educated in the care of ACHD patients.

*Complying with recommendations 5–8 is critical in achieving an optimal ACHD care program.

[†]For specialist centers, recommendations 1–3 should be fulfilled.

ACHD, Adult Congenital Heart Disease; CHD, congenital heart disease.

a pressing issue in providing multidisciplinary care to ACHD patients in Japan. Moreover, the extent to which adult cardiologists in Japan are currently dedicated to ACHD care and the extent to which they are willing to participate in such care remain unknown.

Released in 2000, the “Guidelines for Management of Congenital Heart Disease in Adults” suggest the necessity of specialized regional ACHD care facilities.²¹ However, although a previous nationwide survey of ACHD care facilities clarified the number of outpatient visits, admissions, cardiac surgeries, and obstetric deliveries that were related to ACHD patients in each facility,²² the exact number and geographic distribution of facilities with the potential to provide multidisciplinary care as regional ACHD centers, have not yet been investigated. Our previous study identified the various types of facilities that could potentially serve as regional ACHD centers in Japan: general hospitals, such as university hospitals; facilities with pediatric cardiac surgeons; and facilities treating large numbers of patients.²⁰

The present nationwide survey was therefore designed to investigate the current status, needs, and future direction of ACHD care in adult cardiology departments in Japan with the aim of identifying the number and geographic distribution of facilities with the potential to become regional ACHD centers.

Methods

Participating Facilities

In the present study, a descriptive, cross-sectional, nationwide survey was conducted by mailing survey questionnaires to the Directors of adult cardiology departments in 138 facilities located in 8 geographical regions throughout Japan. Directors were asked to complete the questionnaires themselves or to delegate the response to a physician who was dedicated to or interested in ACHD care in their department. The facilities selected to receive the mailing included those that met any of the following institutional criteria: (1) university hospitals; (2) facilities with ≥ 50 ACHD outpatients; and (3) facilities with ACHD-specialized outpatient clinics. Information on eligible facilities was provided by our previous nationwide survey, which gathered data regarding the status and function of ACHD care facilities in Japan.²² Criterion (3) was adopted

because facilities with ACHD-specialized outpatient clinics were assumed to be treating a sufficient number of ACHD patients. Because the number of pediatric cardiac surgeons employed at each facility was not available prior to the survey, we did not use this as inclusion criteria, but we did include an item about it on the questionnaire.

ACHD Care in Adult Cardiology Departments

The European Guidelines for Management of Grown-Up Congenital Heart Disease and the 32nd Bethesda Conference on Care of Adult Congenital Heart Disease state that patients requiring ACHD care can be categorized into 3 types: (1) patients with severe CHD, as represented by patients with single ventricular physiology²³ who require care provided exclusively by a specialist center; (2) patients with moderate CHD, as represented by patients with tetralogy of Fallot²⁴ who can receive care from appropriate general adult cardiac facilities in strong collaboration with a specialist center; and (3) patients with mild CHD, as represented by patients with isolated congenital aortic valve disease who can be managed by non-specialist centers that have access to specialized care if needed.^{6,10}

To document the current status and future direction of ACHD care in adult cardiology departments in Japan, we asked participating facilities to what extent these 3 types of patients were currently being treated and to what extent they would be treated in the future. We also asked participating facilities whether they had an ACHD-specialized outpatient clinic directed by an adult cardiologist and whether they intended to participate in or send a trainee to a formal ACHD fellowship training program, if available. Furthermore, we conducted a “needs assessment” pertaining to the needs and barriers perceived by cardiology specialists involved in the care of ACHD patients. We asked participants about educational as well as organizational enhancements that would promote the optimal delivery of ACHD care. Responses to each item related to the education and organization of the health-care system and were rated via a 5-point Likert-type scale ranging from “very much necessary” to “not necessary at all.”

Definition of Facilities With the Potential to Become Regional ACHD Centers

To identify facilities with the potential to become regional

Table 2. Background of Responding Facilities

	n or means	% or ±SD
Regions		
Hokkaido	4	3.7
Tohoku	4	3.7
Kanto	48	44
Chubu	13	11.9
Kinki	16	14.7
Chugoku	9	8.3
Shikoku	3	2.8
Kyusyu & Okinawa	12	11
Types of facilities		
University hospitals	83	76.1
General hospitals	20	18.3
Cardiovascular centers	6	5.5
Age of responding physicians*	48.4	6.4
Board-certified member of the Japanese Circulation Society	100	91.7

N=109. *n=107.

ACHD centers, we assessed the structure of each participating facility. Based on the definitions for optimal ACHD care structure established by previous studies (see Table 1 for details),^{18,25} we adopted the following criteria to delineate potential regional ACHD centers: having an adult cardiology department that plans to treat all ACHD patients, including

patients with severe CHD; employing at least 1 pediatric cardiologist; employing at least 2 pediatric cardiac surgeons; currently possessing an ACHD-specialized outpatient clinic or planning to establish one in the near future; having a sufficiently staffed and equipped electrophysiology laboratory; and having other supportive services, such as obstetrics and psychiatry, available for patients.

Because the purpose of this study was to assess the potential of each facility to provide multidisciplinary care as a regional ACHD center, we did not include the presence of an ACHD cardiologist in our definition (although it was included in the definition established by previous studies).^{18,25} Instead, we considered the presence of either an adult cardiologist planning to treat all ACHD patients or a pediatric cardiologist as a candidate future ACHD cardiologist. Another difference between the criteria adopted in this study and those in our previous study is that we substituted the criterion of either the presence of an ACHD-specialized outpatient clinic or a plan to have one in the near future for the number of annual ACHD outpatient visits. We made this change because we felt it would be difficult for adult cardiologists in Japan to know the total number of ACHD outpatients followed in their facilities because, in most facilities, the majority of ACHD patients are followed by pediatric cardiologists. In addition, we did not include the presence of nurse specialists in our definition because such specialists are very scarce in Japan at present, and almost none of the facilities were expected to employ them.

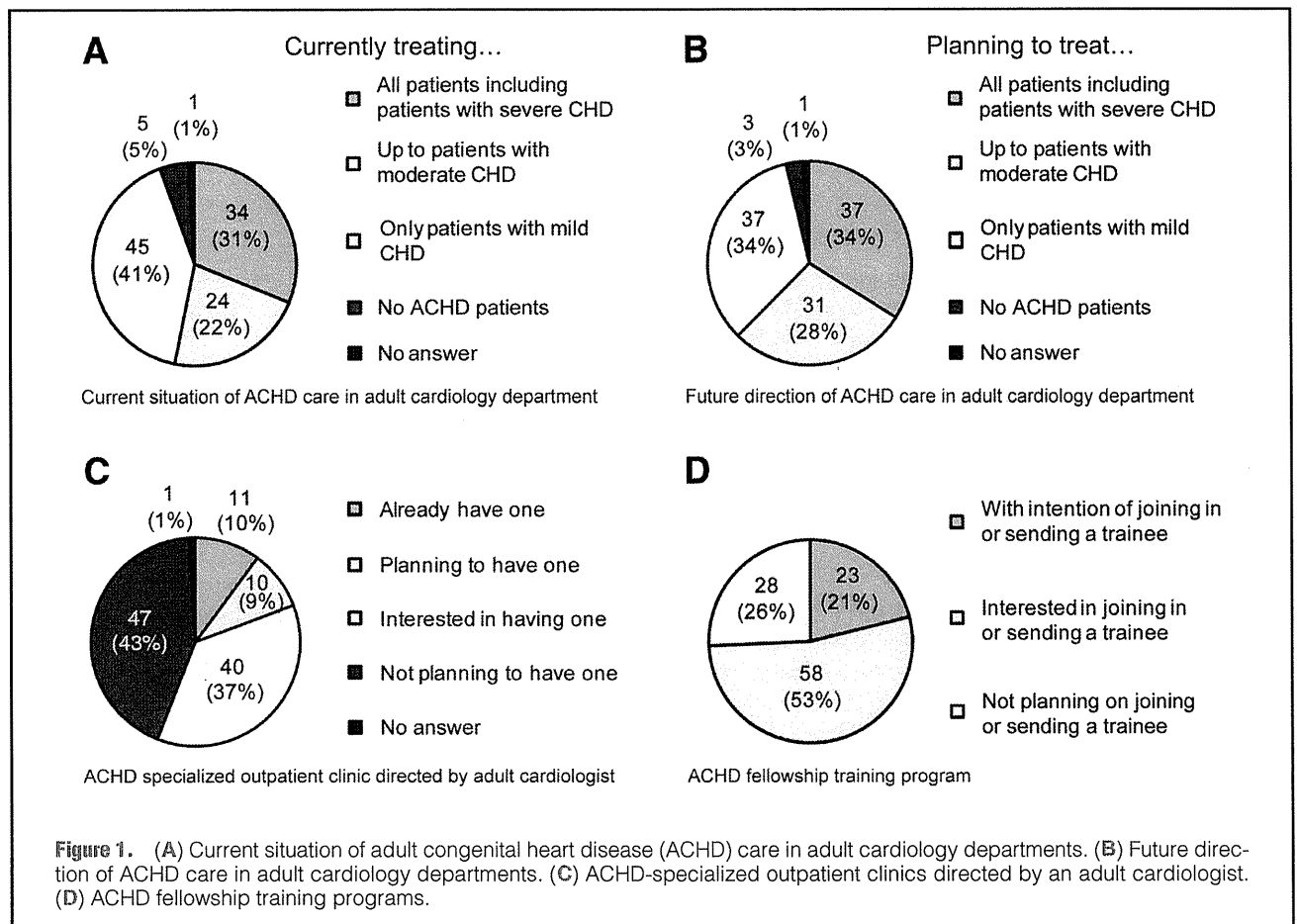


Figure 1. (A) Current situation of adult congenital heart disease (ACHD) care in adult cardiology departments. (B) Future direction of ACHD care in adult cardiology departments. (C) ACHD-specialized outpatient clinics directed by an adult cardiologist. (D) ACHD fellowship training programs.

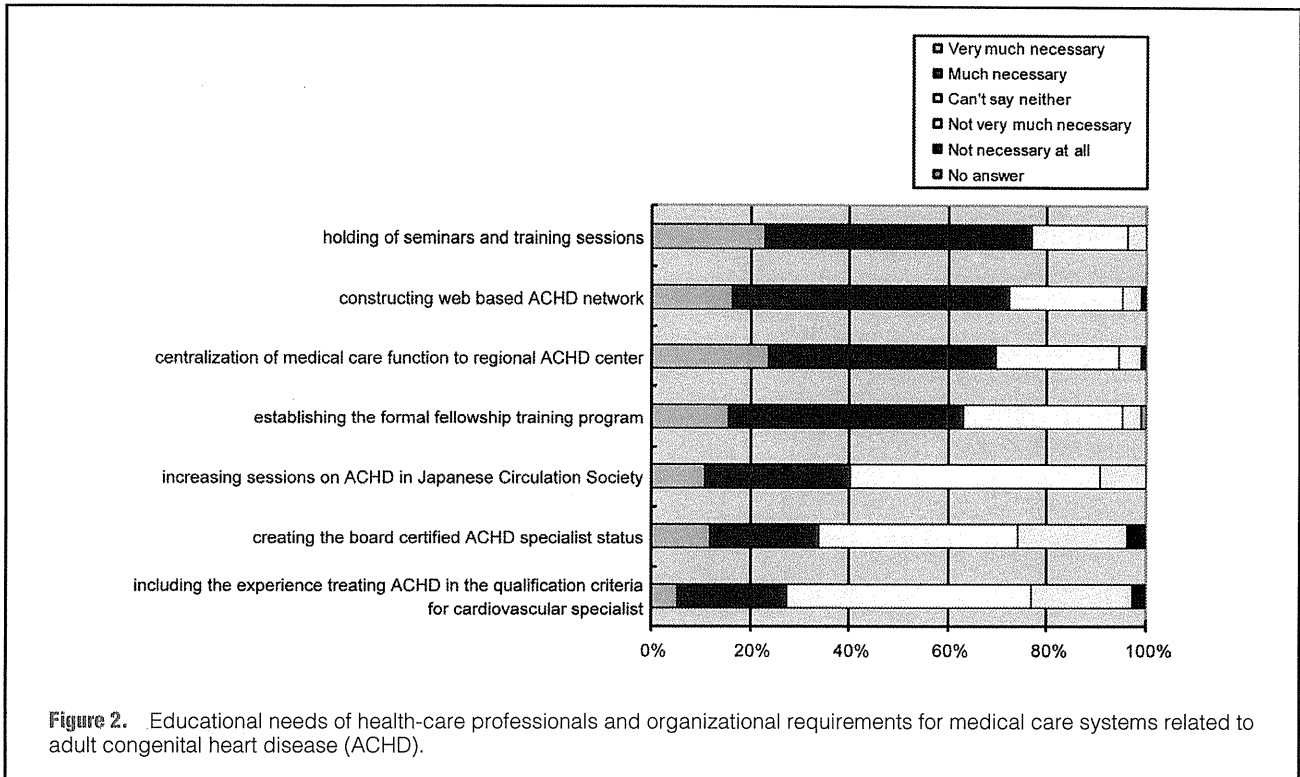
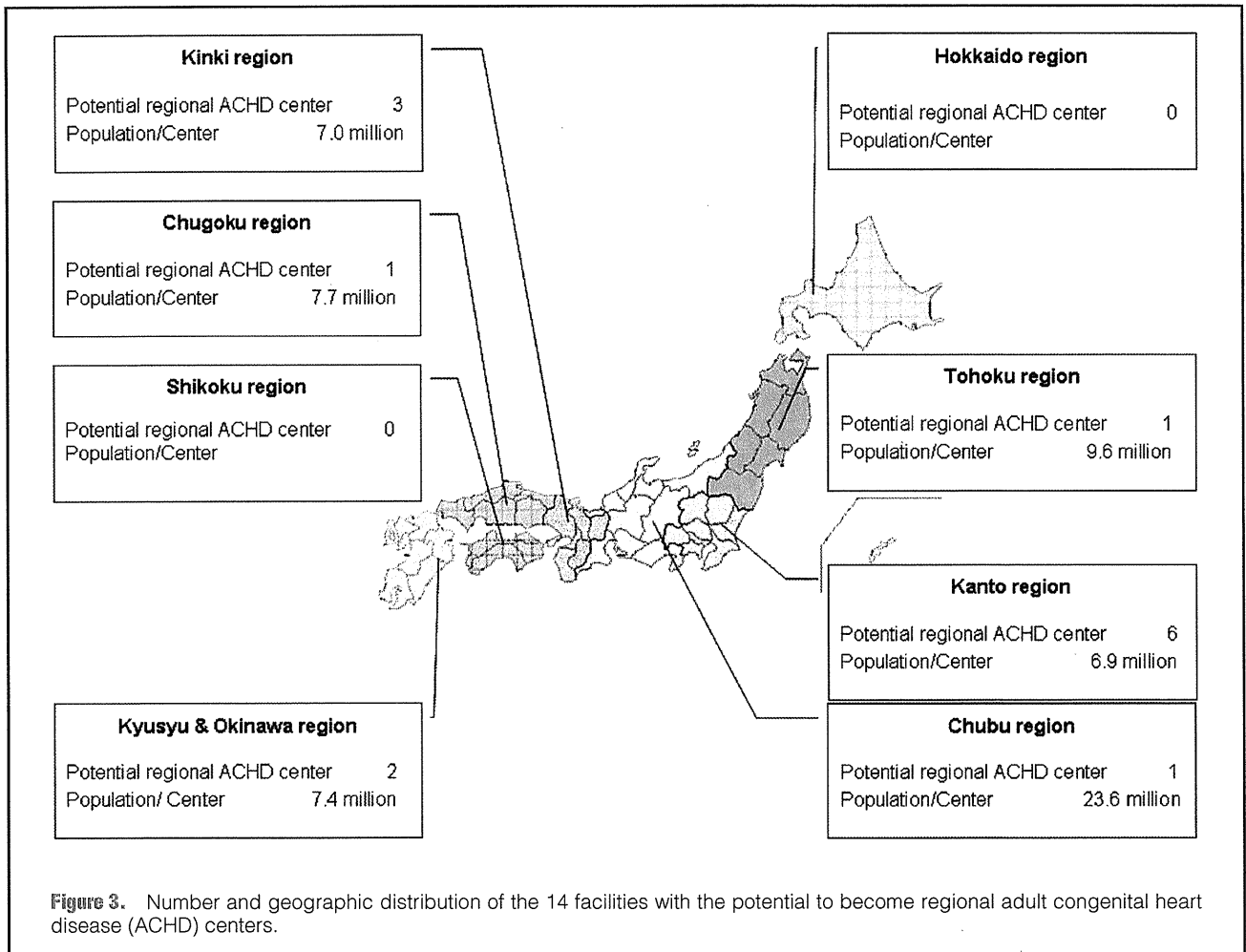


Figure 2. Educational needs of health-care professionals and organizational requirements for medical care systems related to adult congenital heart disease (ACHD).

	No. of facilities meeting this criterion	
	N	(%)
Health-care professionals		
Adult cardiology department planning to treat all ACHD patients, including severe patients	37	33.9
≥1 pediatric cardiologist	74	67.9
≥2 pediatric cardiac surgeons	41	37.6
ACHD-specialized outpatient clinic		
ACHD-specialized outpatient clinic	29	26.6
Planning to have ACHD-specialized outpatient clinic directed by an adult cardiologist	10	9.2
Sufficiently staffed and equipped electrophysiology service		
≥1 adult cardiologist specializing in cardiac catheterization	108	99.1
≥1 adult cardiologist specializing in arrhythmias	100	91.7
≥1 adult cardiologist specializing in echocardiography	88	80.7
≥500 cardiac catheterizations per year*	83	76.1
≥20 ablations per year*	87	79.8
≥20 pacemaker insertions per year*	100	91.7
≥10 implantable cardioverter defibrillator implantations per year*	76	69.7
CARTO, EnSite system available	81	74.3
Magnetic Resonance Imaging available	87	79.8
3-dimensional computer tomography available	99	90.8
Other supportive service available		
Intensive care unit	106	97.2
Cerebral surgery	103	94.5
Obstetrics	100	91.7
Psychiatry	95	87.2
No. of facilities with optimal ACHD care structure		
No. of facilities fulfilling all the criteria	14	12.8

*Total number of procedures conducted in an adult cardiology department (not specific to ACHD).
Abbreviations see in Table 1.
N=109.



Statistical Analysis

Data were analyzed with SPSS v16.0 (SPSS Inc, Chicago, IL, USA). Nominal-level data were expressed as percentages, and means and standard deviations were calculated for continuous variables. After identifying the facilities having the potential to become regional ACHD centers, we calculated the population that each center was expected to serve in each of the 8 regions in Japan.

Ethical Considerations

This study was carried out with approval from the Ethics Committee of the Tokyo University Graduate School of Medicine/Faculty of Medicine. We explained to participants via a letter of intent that we would never force them to participate in the survey, that they would not suffer any disadvantage even if they did not provide consent, and that we would never disclose personally identifiable information to third parties.

Results

ACHD Care in Adult Cardiology Departments

Of the 138 facilities invited to participate in this study, satisfactory replies were obtained from 109 facilities in all 8 regions in Japan (a response rate of 79.0%). Backgrounds of responding facilities are shown in Table 2. Of the responding facilities, 83 (76.1%) were university hospitals, 20

(18.3%) were general hospitals, and 6 (5.5%) were cardiovascular centers. The mean age of the responding physician at each facility was 48.4 ± 6.4 years.

Thirty-four facilities (31.2%) reported that they were currently treating all ACHD patients, including patients with severe CHD. In the future, 37 facilities (33.9%) reported that they planned to treat all ACHD patients in their adult cardiology departments. Eleven departments (10.1%) included an ACHD-specialized outpatient clinic directed by an adult cardiologist, and 10 (9.2%) planned to have one in the near future. Twenty-three facilities (21.1%) intended to participate in or send a trainee to a formal ACHD fellowship training program, if available. These data are summarized in Figure 1.

Figure 2 shows the perceived educational needs of health-care professionals and the organizational requirements deemed necessary for achieving an optimal ACHD care system. Educational opportunities, such as "holding of seminars and training sessions" and "establishing the formal ACHD fellowship training program", were rated as "very much necessary" or "much necessary" by more than half of the respondents. In contrast, items related to the qualifications of ACHD health-care professionals, such as "creating a board-certified ACHD specialist status" and "including experience in treating ACHD in the qualification criteria for cardiovascular specialists", were supported by less than half of the participants. Items related to organization of a care delivery system for ACHD patients, such as "constructing a web-based ACHD network"

and “centralization of medical care function to regional ACHD centers”, were highly supported.

Facilities With the Potential to Become Regional ACHD Centers

Table 3 shows to what extent the criteria for becoming a regional ACHD center were fulfilled by the participating facilities. Fourteen facilities (12.8%) fulfilled all applicable criteria. Criteria fulfilled by less than half of the participating facilities included the existence of an adult cardiology department that planned to treat all ACHD patients (33.9%), the presence of at least 2 pediatric cardiac surgeons (37.6%), and the presence of either an ACHD-specialized outpatient clinic or the existence of a plan to have one in the near future (35.7%). Applying only these 3 criteria, the number of facilities that were qualified to serve as regional ACHD centers was limited to 19.

The geographic distribution of the 14 facilities fulfilling all criteria throughout the 8 regions and the populations that each facility was expected to serve in each region are shown in Figure 3. In 6 of the 8 regions, each facility was expected to serve a population of 6.9–23.6 million, while in the other 2 regions, there were no facilities with the optimal care structure necessary. Thus, in Japan as a whole, each existing center was projected to serve a population of 9.1 million.

Discussion

This is the first nationwide survey focusing on ACHD care in adult cardiology departments in Japan. We identified the number and geographic distribution of facilities with the potential to become regional ACHD centers. We surveyed a wide range of facilities throughout the nation as candidates for potential regional ACHD centers, and the response rate was higher than in 2 previous studies.^{18,25} Hence, we believe that the results of the current study are representative of ACHD care in adult cardiology departments in Japan and that they cover most of the facilities with the potential to become regional ACHD centers.

Providing Multidisciplinary ACHD Care

A previous study in Europe reported that in 90% of ACHD specialist centers and in 78% of non-specialist centers, ACHD care was provided by specialized cardiologists with an adult cardiology background.¹⁸ In contrast, the current study showed that only about one-third of adult cardiology departments in the surveyed facilities were currently treating or planning to treat all ACHD patients, including patients with severe CHD. Moreover, only 11 facilities had ACHD-specialized outpatient clinics directed by an adult cardiologist. These results confirm the findings of previous studies suggesting that multidisciplinary ACHD care is not provided in every facility, even those at university hospitals.^{19,22} Because a multidisciplinary approach that includes both adult and pediatric cardiologists is indispensable for optimal ACHD care, facilities with adult cardiology departments disposed to treat all ACHD patients should take a leadership role in establishing models for providing multidisciplinary ACHD care in cooperation with pediatric cardiologists and other health-care professionals.

Organization of Different Levels of Education in ACHD Care

The need for educational opportunities was revealed to be high among adult cardiologists. However, the number of facilities with the actual intention of joining or sending a trainee

to formal ACHD fellowship training programs was low. This result reflects the current Japanese situation in that many adult cardiologists are interested in gaining knowledge in areas of ACHD, but few actually plan to treat ACHD as their subspecialty. In the current situation, offering a casual and easily joined opportunity to learn about ACHD care to a wide range of adult cardiologists might be necessary to increase the number of adult cardiologists interested in treating ACHD. In recent years, the organization of training in the care of ACHD patients has been enthusiastically discussed in Western countries, and 3 different levels of training have been recommended.²⁶ Seminars and training related to the care of ACHD patients—opportunities that were requested by the majority of respondents in this study—might correspond to “level 1 training”, which was defined as training to provide adult cardiologists with sufficient knowledge to recognize moderate and severe CHD as well as to obtain appropriate consultations or referrals to regional ACHD centers.²⁶ In the mean time, because the idea of establishing formal ACHD fellowship training programs was also supported by the majority of study participants, a concrete curriculum for formal ACHD fellowship training, perhaps corresponding to “levels 2 and 3 training”, should be developed to nurture sufficiently trained ACHD cardiologists.

Establishing Regional ACHD Centers and Networks

Fourteen centers fulfilled all applicable criteria for becoming regional ACHD centers. Because we focused on ACHD care in adult cardiology departments in this study, detailed criteria related to this area, such as the annual number of catheter interventions and implantable cardioverter defibrillator implantations, were included in the criteria. However, when we looked at the data, we found that these criteria were fulfilled by the majority of participating facilities. Other than criteria related to adult cardiology, the criterion that appeared to be the most difficult for participating facilities to fulfill was employment of at least 2 pediatric cardiac surgeons. Because in-hospital death rates for ACHD patients operated on by pediatric cardiac surgeons were reported to be lower than death rates for ACHD patients operated on by non-pediatric cardiac surgeons,²⁷ the presence of a sufficient number of pediatric cardiac surgeons should be considered one of the most important factors in the screening of potential regional ACHD centers.

The fact that 14 facilities fulfilled all applicable criteria implies that, in Japan as a whole, each center would be expected to serve a population of 9.1 million. This number is compatible with the standard suggested at The Canadian Cardiovascular Society Consensus Conference on Adult Congenital Heart Disease, which called for one regional ACHD center per 3–10 million people; it is also in agreement with the standard suggested at the 32nd Bethesda Conference on Care of Adult Congenital Heart Disease and in the European Guidelines for Management of Grown-Up Congenital Heart Disease, which called for one regional ACHD center per 3–10 million people.^{7,10,13} Because this is the first nationwide survey focusing on ACHD care in adult cardiology departments in Japan and no other previous data exist, we cannot judge whether the number of facilities with the potential to become regional ACHD centers in Japan is increasing. However, in 2 of the 8 regions in Japan, there were no facilities with the potential to become regional ACHD centers. This result suggests that there are regional differences in the potential to deliver specialized ACHD care and that there is sufficient opportunity to improve the ACHD care delivery

system, especially in rural areas. The 2 regions without potential regional ACHD centers are the areas with the first and second smallest populations in Japan. In our previous study, we pointed out that centralization of medical functions in rural areas is not feasible due to the small number of patients in such regions.²⁰ To solve this problem, it will be necessary to develop an intraregional health-care delivery model, such as one that uses a web-based telemedicine system, rather than promoting centralization uniformly throughout all the regions. Further research specifically focused on regional medicine is therefore needed.

Study Limitations

This study had several limitations. First, because we targeted ACHD care in adult cardiology departments in this study, we did not investigate to what extent other departments in each facility, such as pediatric cardiology and pediatric cardiac surgery departments, were offering ACHD care and how they collaborate with each other. As the majority of ACHD patients are apparently now followed by pediatric cardiologists in most facilities, the results of this study might only reflect one aspect of the current ACHD care situation in each facility. Because a regional ACHD center is expected to be multidisciplinary, cooperation between health-care professionals is essential. Future research is needed to evaluate the multidisciplinary functioning of ACHD-specialized teams in each facility, not merely those in the adult cardiology department.

Second, as mentioned above, the criteria applied for identifying the facilities with the potential to become regional ACHD centers in this study were not exactly the same as those adopted in the previous studies. Indeed, because we have little evidence about which elements in the structure and organization of ACHD care facilities contribute to better patient outcomes, there is no "gold standard" with regard to the criteria for regional ACHD centers. Future studies should link structural and organizational elements to mortality and morbidity data in order to clarify optimal ACHD care structure.

Finally, although we discussed the sufficient number of potential regional ACHD centers in the context of population and geographic distribution, there are many other factors that must be considered, such as the time required for patients to access the nearest regional ACHD center and the referral relationships between regional ACHD centers and other facilities. These factors should be evaluated, especially in rural areas, in order to determine whether a regional ACHD center should be established in every region or whether establishing strong referral relationships between regional ACHD centers in urban areas and non-specialist centers in rural areas is more efficacious.

Conclusions

In this nationwide survey, we investigated the current status and future direction of ACHD care in adult cardiology departments in Japan and identified the number and geographic distribution of facilities with the potential to become regional ACHD centers.

The results of the current study clarified the limited number of adult cardiology departments in Japan that currently treat or plan to treat ACHD. Offering a casual and easily utilized opportunity to learn about ACHD care to a large number of adult cardiologists and establishing a formal ACHD fellowship training program for those who are interested in

treating ACHD were thought to be necessary. Even though the number of facilities with the potential to become regional ACHD centers in Japan has just reached international standards, there are significant differences in the number of potential ACHD centers available in each region. Medical policy other than consolidation of medical functions in regional ACHD centers might be required for the coordination of ACHD care systems in less populated rural areas.

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循環器疾患等生活習慣病対策総合研究事業研究事業
(研究課題名) 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
(H23—循環器等(生習)—一般—016)
分担研究報告

成人先天性心疾患患者診療に対する循環器内科ネットワークの確立

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研究要旨

近年の手術技術/内科的管理の進歩により、先天性心疾患(congenital heart disease : CHD)患者の予後は著明に改善した。本邦ではすでに成人に達した CHD(ACHD)患者は約 40 万人に達するとされ、今後も増加することが予想され、ACHD 患者を小児科/胸部外科から(成人)循環器内科医師へ引き継いでいくことが早急に望まれる。そのためには、ACHD 集約施設の確立を急ぐ必要がある。今回、我々のアンケート調査結果をもとに、全国主要施設の循環器内科からなる ACHD 診療体制循環器内科ネットワークの確立を目指した。

研究目的

近年、外科治療と診断技術、内科管理の進歩により、心室中隔欠損症やファロー四徴症に代表される先天性心疾患患者の多くが、成人期を迎えるようになってきた^{1,2}。成人になった先天性心疾患患者、つまり成人先天性心疾患(adult congenital heart disease :ACHD)患者の数は本邦で現在少なくとも約 40 万人と推定され、今後年間約 9000 人の増加が見込まれている²。ACHD 患者数の増加に伴い、小児期から成人期以降までの継続的な医療に対する需要は高まっており、治療・経過観察を含む疾患に関する総合的な理解とそれに基づいた医療システムの構築が必要とされている。

欧米では 1970 年代から ACHD 専門施設が設立されており、多くが循環器科医の主導のもと、小児科医と共同で運営されている^{3,4}。他方、本邦では ACHD 専門の診療科は少なく、多くの患者は小児科医による経過観察を受けている。

本研究の第一段階において、東京大学医学部付属病院において 2008 年より開設した循環器内科医師による ACHD 専門外来の特徴を調べたところ、成人 CHD 外来への

紹介は小児科および心臓外科からが主体で、複雑心奇形が大半であった。また、成人期に手術を行なうことが予想されるような外科的に完全修復されていない患者も多く存在し、手術(根治)不可能なアイゼンメンジャー症候群/肺高血圧症の患者やフォンタン循環といった特異な循環動態を有する患者も少数ながら存在していた。このような成人循環器医師にとって不慣れなケースが多いため、今後成人循環器内科医師の専門的トレーニングが必要であるのはもちろんであるが、外来開設当初は、こういった症例の経験豊富な小児科や心臓外科との提携は必須と考えられた。最終的には、女性科(産婦人科)などすべての科との連携も必要となることも考慮すると、ACHD 外来開設は、総合的医療ができる大学病院/国立病院といった大規模な総合病院でかつ CHD 患者をすでに豊富に診ている施設で行なわれることが必須であると考えられた。

そこで、我々は本研究班において、今後、成人 CHD 専門外来を担いうる施設の特定と、専門外来設置の意向を有する循環器科医の特定、さらには特定された循環器科医

に対して専門外来設置に向けた働きかけを行うことを検討した。まず、アンケート調査を全国主要 138 施設に郵送し、約 80% の回答を得ることに成功し、その結果を *Circulation Journal* に掲載するに至った⁵。その結果からは、全国に集約施設基準を満たしうる施設は 14 施設しかなく、現状では日本の ACHD 診療に十分な集約施設は確保できないと思われた。技術的状況や小児科医師・先天性心疾患専門外科医師などは概ね確保できてはいたが、とくに不十分な理由としては循環器内科として ACHD 専門外来の設置もしくは設置する意向が少ないこと(約 36%)が第一に挙げられた。

以上の調査結果をもとに、ACHD 診療体制の構築で、まず重要視されねばならない要因を挙げ、その対策を講じることが必要と考えられた。1.すでに 40 万人以上 ACHD 患者が存在し今後増加の一途をたどることが予想される。

2.これまで通りの小児科医師主体の医療では許容力として破綻する可能性が高い。

3.循環器内科医師による診療開始が望まれるが、以下の問題が存在し/予想される。

a.紹介患者の多くが専門的治療の必要性があり、循環器内科医師のみでは対応が不十分と思われる。

b.循環器内科に ACHD 診療に加わる意思が積極的にはない。

この 3 つの点を考慮すると、やはり循環器内科医師による ACHD 診療への介入がまず必須であると考えられるが、循環器内科医師のみでは ACHD 診療は難しいということになる。しかしながら、東京大学医学部附属病院では、実際に ACHD 診療を開始して、十分に成果が上がっている。その大きな特長としては、問題点 3a に対して、小児循環器内科医師との外来併診により対応している点である。特に、問題症例に関してはこの方法により、両科の協力のもと診療ができ、同時に循環器内科医師の実地トレーニングも行えるようになった。医療享受者としての患者側のデメリットはなく、

成人疾患の診療をスムーズに受けることができるメリットそしてなにより併診により成人医療への移行に伴う心理的ストレス軽減につながると考えられた。問題点 3b の理由として考えられる要因は、循環器内科医師の日常診療の多忙、ACHD 診療に対する知識的および実臨床上の不安といった点が 3b を生んでいる大きな要因と考えるが、東京大学での実績経験による他施設への具体的な働きかけを行うことで循環器内科医師による ACHD 診療体制の確立は進むものと思われた。

そこで、すでに診療体制が稼働している東京大学医学部附属病院での方法を参考にして、全国の主要 7 施設に循環器内科医師による ACHD 診療体制を確立することを目的とした働きかけを行い、実際の診療を稼働することを試みた。

研究方法

先の報告により比較的十分に集約施設の基準を満たす 7 施設に対し依頼状を送付し、ACHD 診療開始に向けての参加を呼び掛けるとともに、今後の ACHD 診療体制形成のための循環器内科議決機関としての活動することの了解を求めた。

倫理面への配慮

本研究は、新たな診療体制確立に向けた医師による議決機関の形成の呼びかけであり、倫理上考慮する必要を認めない。

研究結果

東京大学に事務局を置いての 7 施設への呼びかけによりすべての施設からの合意の返事を受けることに至った。2011 年 12 月 10 日に開催した第一回成人先天性心疾患対策委員会において、全施設参加のもと各施設の循環器内科での ACHD 専門外来の開設を小児科医師との協力のもと進めていくことで合意に至った。また、地域性および人口などの要素を加味して、さらに細かく診療体制を形成する方向で意見が一致し、参加施設の増加を考慮することの合意を得た。

考察

今回の循環器内科医師による全国規模でのACHD診療体制の推進は、非常に画期的な試みと言える。我々の研究⁵から、2009年の時点では、全国主要施設のうち循環器内科医師によるACHD専門外来は2箇所にしかな認められていない（公表外データ）。今回、その数を大きく進める結果となっただけではなく、この委員会の設置は、循環器内科医師のこの分野への注目度を高める効果が期待できる。先の我々の報告では、10施設からしか専門外来の設置の意向がなかったことを考えると、この委員会の設置は次に依頼をする予定施設に対して大きな影響力を持つと思われる。

また、循環器内科専門外来運営において、小児科医師との連携を行うことを基本合意としたことは、我々の事前調査でも指摘されている循環器内科医師のACHDに関する知識や経験不足を補うに十分と考えられ、患者の不利益はなくせるものと考えられる。それに加えて、循環器内科医師の成人医療に関する知識や技術をスムーズにACHD患者に提供できるメリットも加わることになり、小児科医師のみの診療と比較しても医療の質の向上を望むことができる。

しかしながら、40万人以上とも言われるACHD患者（うち約13万人が中等症以上）²の診療体制としてはまだまだ不十分であり、今後参加施設のさらなる追加加入が必要である。最終的には、本委員会に属する施設を十分な数まで増加させ、各施設が各地域での中心となり、地域ネットワークの形成へとつなげていく必要がある。

そして更に、ACHD専門とする循環器内科医師を養成する教育プログラムならびに専門医制度の確立を経て、循環器内科医師単独での診療体制へと進化することで、より多くのマンパワーが発揮できるACHD医療体制への進化を目指す必要がある。

結論

今回の研究では、40万人というACHD患者への循環器内科による対応が急務とされる状況下で、患者に対する不利益を生じない実際の診療推進のきっかけを作成できたものと考えられる。今後、この組織を発展させ、全国規模による推進を行うことがACHD診療問題の解決への近道であると予想される。今後引き続きこの研究の推進が望まれる。

健康危険情報

該当せず。

研究発表

1. 論文発表：なし
2. 学会発表：なし

知的財産権の出願・登録状況 (予定を含む。)

1. 特許取得：なし
2. 実用新案登録：なし
3. その他：なし

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