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. (Circ J 2009; 73: 1147–1150)

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 1952! Currently, surgical mortality in large CHD centers is 10% or less; 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 1997! The adult population with CHD is estimated at more than 400,000, with a rate increase of 4–5% per year.6

Patients with at least moderate CHD are likely to undergo 1 or more surgical interventions.3 With rare exception, notably patent ductus arteriosus, surgery is not curative, so most if not all operated patients have postoperative residual 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 2006.17 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) cardiology 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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**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%.

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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.
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.

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. 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 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. 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. 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. 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. 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. 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.\textsuperscript{14} The JSC guidelines have not specified the desirable number of specialized centers.\textsuperscript{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.\textsuperscript{15} 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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References