



アジアセッション



13th Annual Meeting of Japanese Society for Adult Congenital Heart Disease

AS-1 Midterm Follow-up result of transcatheter ASD closure in adults severe pulmonary hypertension: The Role of Advanced Medical Therapy for Pulmonary Arterial Hypertension

Nam Kyun Kim¹⁾, Su-Jin Park¹⁾, Lucy Y. Eun²⁾, Jae Young Choi¹⁾

¹⁾ Division of Pediatric Cardiology, Severance Cardiovascular Hospital, Yonsei University Health System, Seoul Korea

²⁾ Myongji Hospital Cardiovascular Center, Kwandong University Health System

Background: Atrial septal defect (ASD) is one of the most common congenital diseases, but symptoms usually do not surface earlier in life, therefore many patients are diagnosed in adulthood. Furthermore, patients with large ASD may have concomitant various degree of pulmonary arterial hypertension (PAH), which may cause complications. The purpose of this study was to investigate the progress of patients with ASD and severe PAH after transcatheter closure of the defect.

Method: During the 5 year period between June, 2005 and May, 2010, total of 435 ASD patients underwent transcatheter closure in our institute. Patients with mean systolic pulmonary artery pressure (PAP) over 60 mmHg at the time of defect closure were classified as severe PAH, and we reviewed these patients retrospectively.

Results: Sixty-six patients had concomitant PAH (15.3%, systolic PAP >40 mmHg) and 14 patients (3.2%) had severe PAH. Among them, 9 patients needed selective pulmonary vasodilator therapy after defect closure. The mean age in these 9 patients was 30.7 ± 10.8 years, mean systolic PAP decreased from 83.3 ± 14.4 to 71.7 ± 10.4 mmHg, after the closure. Mean Qp/Qs ratio was 1.8 ± 0.6 and Rp/Rs ratio was 0.4 ± 0.3 . Of the 9 patients, 4 patients were successfully weaned off the medication, but 5 patients are still under advanced medical therapy.

Conclusion: Transcatheter closure of ASD is safe and efficacious in selected group of patients with severe PAH. New pulmonary vasodilators may expand indication of closure and improve clinical outcome.

AS-2 Marriage among Taiwanese Adults with Congenital Heart Disease

Yu-Chuan Hua¹⁾, Chun-An Chen²⁾, Chun-Wei Lu^{2) 3)}, Li-Li Mo⁴⁾, Jou-Kou Wang^{1) 2)}, Hung-Chi Lue^{1) 2)}, Mei-Hwan Wu^{1) 2)}

¹⁾ Cardiac Children's Foundation, Taiwan; ²⁾ Department of Pediatrics, and ³⁾ Adult Congenital Heart Center, National Taiwan University Hospital, Taipei, Taiwan; ⁴⁾ Department of Social Work, SooChow University, Taipei, Taiwan.

In a previous study, we have found that marriage rate is lower among Adult Congenital Heart Disease (ACHD) compared to the general population. Marital status was an important factor underlying differences in QoL between male and female patients. Therefore, this study conducted a combined quantitative and qualitative survey to further investigate marriage-related issues in this unique patient population. Between January to March 2010, 210 valid questionnaires were collected from ACHD patients (patient age: 33.3 ± 11.1 years; proportion of female patients: 67.6 %). Most research subjects had a mild form of CHD (47.6 %), and good functional status (58.6 %). Most patients had undergone either surgical or transcatheter repair of their cardiac defects (81.4 %). Compared to the general Taiwanese population, marriage rate for ACHD was low overall (45 % vs. 66 %). Most research subjects were unmarried (n = 116, 55.2 %), and the percentage of married female patients significantly exceeded the percentage of married male patients (56 % vs. 19 %, $P < 0.001$). Common medical-related reasons for remaining unmarried included: (1) Pregnancy could worsen cardiac function (31.1% of female patients); (2) Children born to ACHD might also have CHD (24.1%; women vs men: 27.9% vs 20.0%, $P = \text{NS}$); (3) Giving a potentially negative impression to others due to CHD (18.1%; women vs men: 21.3% vs 14.5%, $P = \text{NS}$), and (4) Limited physical capacity to handle family duties (15.5%; women vs men: 24.6% vs 5.5%, $P = 0.005$). Married men seemed to be more satisfied with their marriages than women were. Regarding satisfaction with marriage-related issues, married ACHD appeared less satisfied with their income and educational or occupational achievements (percentage of satisfaction: 38.3% and 33.0%, respectively). In conclusion, psychosocial concerns about CHD were common among both unmarried and married ACHD. Significant gender discrepancies were observed in marriage rate and marriage satisfaction among ACHD. Additionally, severity of CHD may be related to patient concerns and decisions about marriage.

AS-3 Issues of surgical cases with adult congenital heart disease in stand-alone children' s hospital

Satoshi Yasukochi¹⁾, Kiyohiro Takigiku¹⁾, Hikoro Matsui¹⁾, Nao Inoue¹⁾, Shigerou Watanabe¹⁾, Hiromitsu Mori¹⁾, Yasuko Morimoto¹⁾, Yorikazu Harada²⁾, Takahiko Sakamoto²⁾

Department of ¹⁾ Pediatric Cardiology and ²⁾ Cardiovascular surgery
Nagano Children' s Hospital

Background: the number of the surgical cases with congenital heart disease (CHD) in stand-alone children' s hospital (SA-CH) was remarkably increased with good survival rate and outcome, but we faced the difficult situation and management of such postsurgical patients, especially in the case of re-do, if the children' s hospital does not have any affiliated hospital with adult CHD department.

Aim: to investigated the clinical and social issues of the surgical cases older than 18 years old who underwent cardiac surgery either at our hospital or at referral hospital.

Subjects: Of 3328 CHD surgery since 1993, 26 patients >18yo were operated at our hospital and 5 were referred to the other hospital outside the prefecture. Results: the twenty one cases of 26 underwent open heart surgery (initial procedure (11) , re-do (13) , others five cases had pacemaker implantation (4) and removal of graft (1) . The five of 21 underwent TCPC conversion from modified Fontan operation. The surgical cases >18yo increased from 15 years after opening our hospital. There was no surgical death but one with cardiac resynchronization therapy was lost by congestive heart failure.

Issues: Although the surgical outcome was good, the extra-cardiac complications after surgery (cholecystitis (1) , liver cirrhosis (1) , depression (1)) became problematic because of inexperience of medical management and nursing for the adult in the pediatric ward. Those who were referred to the other hospital required the further professional surgery, especially in the field of aortic arch reconstruction. Beside these issue, the major problem is a lack of network of ACHD care beyond the border of the prefecture, especially for those who required the re-do surgery, because there is few cardiac surgeon who can manage ACHD patients in ACHD care center usually far from the patients' hometown.

L-3 History and Current Care for Adult Congenital Heart Disease in Korea

Jae Young Choi, M.D. and Heung Jae Lee, M.D.*

Division of Pediatric Cardiology, Severance Cardiovascular Hospital, Yonsei University Health System, Seoul, Korea *GUCH Clinic, Cardiac and Vascular Center, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Introduction

In the western countries, the survival of patients with malformed heart started to rapidly increase since the introduction of cardiopulmonary bypass in the 1950's [1], and it was 10 to 15 years later when the concept of 'GUCH clinic' first surfaced [2]. In Korea, palliative surgery for cyanotic congenital heart disease started in 1957, and successful intracardiac repair using cardiopulmonary bypass have been performed since early 1960's in several institutes. However, the initiation of special facility for adults with congenital heart disease appeared more than 30 years later, and in the following decade additional 4 adult congenital heart disease clinics (ACHDC) had been established. Nowadays, the cumulative adult population with CHD is estimated to be about 150,000 to 200,000 [3, 4], but recent survey shows that current care for ACHD in Korea still appears to be inadequate. The lack of attention and investment to this group of patients exist, as experienced in leading ACHD facilities [5], and this phenomenon seems to have been even worse in Korea. We reviewed the history and current affairs in the care for ACHD patients, and speculated how primary physicians responsible for these patients perceive care of ACHD in Korea.

The History of ACHDC in Korea

The rapid achievement of successful cardiac surgeries in early 1960's including repair of ASD, VSD and TOF did not reach general popularity due to the high medical cost and low income per capita in Korea. As a consequence of establishment of national health insurance system in 1977 and the Korea Heart Foundation in 1984, the number of cardiac surgery rapidly increased, and as a result, the cumulated CHD population reaching up to an adult or adolescent age dramatically increased since 1990's. Professor Heung Jae Lee in the Samsung Medical Center, Sungkyunkwan University pioneered in this field to establish Korea's first ACHDC in 1995.

Current Surroundings of ACHDC in Korea and the Result of Brief Questionnaire

There are five regularly operating specialized ACHDC in Korea. All are located in the capital area including 4 centers in Seoul city. In average, around 1000 (660-1636) patients a year visit each of the ACHDC in these centers. The most frequent diagnosis was ASD (32.4%), followed by VSD (22.7%), TOF / DORV (21.7%), PDA (5.2%), univentricular heart (4.5%), d-TGA (3.3%), AVSD (3.3%), and pulmonary stenosis (2.6%), in order of frequency. Remaining patients (4.3%) had Ebstein anomaly, congenitally corrected TGA, coarctation of the aorta, Marfan syndrome, etc. However, the proportion of each disease showed wide range of variation according to the settings of different institute. Each clinic essentially consists of pediatric cardiologist, adult cardiologist, cardiovascular surgeon who majors in CHD, and specialized nurse practitioner in terms of human resources. Nevertheless, only 1 center had dedicated physician and nurse practitioner exclusively affiliated to ACHDC, and in most other centers, the personnel are adjunct to another department at the same time. In most centers, the care for ACHD patients was conducted also in pediatric OPD, frequently. In the remaining centers excluding the 5 mentioned above, the number of patients visiting OPD with ACHD was below 200 per year. Other than the 5 preexisting ACHDC nationwide, 3 other centers positively responded to the plan in building a new ACHDC in the near future. The obstacles for building an optimal ACHDC were said to be as follows; 1) Lack of public support from the government and institution (medical cost, etc.), 2) Fixed perception of adult cardiologists, 3) Fixed perception of pediatric cardiologists, 4) Lack of changes in the public and/or patient's perception, 5) Lack of regional centers with easy access for ACHD patients. In centers currently operating ACHD replied that 1) clause was the biggest obstacle and 1), 2), and 3) were most common multiply checked clauses.

Conclusion

ACHDC in Korea still remains to be under an inadequate environment, and support of human resources, material resources, and public resources are still under challenge. For an effective management of ACHDC, learning from worldwide pioneering institutes in this field is foremost, and striving to reach a social consensus is also called for.

References

1. Dearani JA, Connolly HM, Martinez R, Fontanet H, Webb GD. Caring for adults with congenital cardiac disease: successes and challenges for 2007 and beyond. *Cardiol Young* 2007;17 Suppl 2:87-96.
2. Webb G. The long road to better ACHD care. *Congenit Heart Dis* 2010;5:198-205.
3. Lee HJ. The past, present and the future of adult congenital heart disease. Current status and problems. *Korean Circulation J* 2010;40 Suppl 1:192-6.
4. Lee HJ. An overview of congenital heart disease in adults. *Cardiovasc Update* 2005;7:8-13.
5. Niwa K, Perloff JK, Webb GD, Murphy D, Liberthson R, Warnes CA, Gatzoulis MA. Survey of specialized tertiary care facilities for adults with congenital heart disease. *Int J Cardiol* 2004;96:211-6.