Asia-Pacific pediatric cardiac society: My vision for the next decade
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ABSTRACT
Recently, further expansion of patients with congenital heart disease (CHD) and adults with CHD (ACHD) and evolution of specialized care facilities for pediatric cardiology and ACHD can be anticipated in the Asia-Pacific countries. These patients have a lot of problems to be solved now and in next decade, and they include various medical issues including issues specific to oriental populations, psychosocial issues, establishment of CHD care facilities and so on. We now have two established societies namely, Asia-Pacific Pediatric Cardiology Society (APPCS) and Asia Pacific Society for Adult Congenital Heart Disease (APSACHD). Through these Societies, we should solve these problems by promoting international collaborative studies and support projects to facilitate the rapid progress that is desperately needed in the field in the Asia Pacific Region.

Keywords: Adult congenital heart disease, Asia-Pacific pediatric cardiology society, Asia-Pacific society for adult congenital heart disease, congenital heart disease, future prospect

BRIEF HISTORY OF THE ASIA-PACIFIC PEDIATRIC CARDIOLOGY SOCIETY
It took several years with a lot of discussions of country delegates for establish Asia-Pacific Pediatric Cardiology Society (APPCS). Several meetings have been held among founding members of the Society and finally APPCS was officially established in 2005. Formal congress was started in Thailand (Bangkok; 2006), followed by Korea (Jeju Island; 2008), Japan (Chiba; 2010) and Taiwan (Taipei; 2012). India is hosting this meeting in New Delhi (March 2014).

Recently, adult congenital heart disease (ACHD) field in Asia-Pacific region is also evolving as observed in the North America and Europe, and pediatric cardiologists in our region have been joining the team for taking care of adolescents, adults, and even elder patients with CHD. In this context, I will touch on the future prospect not just in pediatric cardiology field, but also in ACHD field in this review.

BRIEF HISTORY OF THE ASIA-PACIFIC SOCIETY FOR ADULT CHD
Asia-Pacific Society for Adult CHD (APSACHD) was established at the 2nd Congress of APPCS in Jeju Island in Korea in 2008. After that it has been held combined with APPCS at the same place and time. From the beginning, APSACHD has been held under the endorsement of ISACHD (International Society for Adult CHD) and the ESC GUCH (Grown-up CHD) Working Group. The 2nd Congress of APSACHD was held in Chiba (2010), Japan, followed by Taiwan, 2012. In India, there will be special session on ACHD during 5th Congress of APPCS. The APSACHD currently includes 15 Asia-Pacific countries such as Australia, China, India, Indonesia, Japan, Korea, Malaysia, New Zealand, Pakistan, Philippines, Singapore, Taiwan, Thailand, Turkey, and Vietnam [Figure 1].

CHD IN DEVELOPING COUNTRIES IN ASIA
In selected developing countries such as Afghanistan, Myanmar, Bhutan, Nepal, Laos, and Cambodia CHD care is available but limited, virtually nonexistent, or rudimentary. Healthcare is very basic and suboptimal, but is gradually improving together with rapid economic growth. In India, well over 10,000 patients undergo congenital heart surgery each year. Fully 180,000 children with CHD are born annually in India. It is estimated that India needs 200 centers doing...
1,000 cases/year, but in reality there are only about 30 centers now.\textsuperscript{[2]} India is vast country with limited resources, uneven population distribution, and few specialized centers.\textsuperscript{[3]} Also, there are no organized ACHD centers, and it is difficult to travel from remote areas for follow-up appointments. There are special challenges in Pakistan, where severe CHD lesions seem unusually common, where neurodevelopmental problems are frequent, and where the challenges of tuberculosis and malnutrition are prevalent. There is only one center for caring ACHD in Pakistan. Possibly telemedicine and/or medical support are necessary in these Asia-Pacific developing countries in near future.

**NUMBER OF PATIENTS WITH CHD IN THE ASIA-PACIFIC REGION**

On several previous reports, the number of neonates born with CHD is similar in different races and different countries.\textsuperscript{[4]} Survival rate of these patients is improving in these days. In Japan, estimated number of children with CHD is now 400,000; this number is smaller than adults with CHD. The formal registration system on CHD is not working in Asia-Pacific countries; however, we should work together to establish registration system in CHD in future. The annual number of cardiovascular surgery in CHD by the Japanese registry has been constant around 9,000-10,000 in past decade.\textsuperscript{[5]}

**THE NUMBER OF ACHD IN ASIA-PACIFIC AREA**

The number of CHD and ACHD patients in Japan based on the death certificates is as follows: A total of 622,800 patients, including 305,000 children (49%) and 318,000 adults (51%) were estimated to be alive in 1997. From 1997 to 2007, there has been an estimated increase of 9,000 adults every year, and in 2013, 454,000 adults are estimated to be alive [Figure 2].\textsuperscript{[6-9]} The prevalence of ACHD population in Korea, Taiwan, and Thailand in 2000 is 22-26, 20, and 32% of total CHDs, respectively.\textsuperscript{[1]} In Singapore, the number of CHD in adults (~15,000) is higher comparing with CHD in children (~5,000) in 2008. The number of ACHD patients with moderate or greater severity was 36.6% in Korea and 32% in Japan.\textsuperscript{[1]} In the other regions in Asia-Pacific, owing to the development of pediatric cardiology and cardiovascular surgery, the number of children and adults with CHD is constantly increasing.

**PROBLEMS ON CHD FACILITIES AND HUMAN RESOURCES IN THE ASIA-PACIFIC AREA**

In the Asia-Pacific area, other than selected countries where pediatric heart care has not developed, there is a society for pediatric cardiology in each country. Also, pediatric cardiology facilities have been developed in most countries those belonged to APPCS. The number
of countries belonged to APPCS is now increased to 19. However, the number of cardiovascular surgeons in most of the countries is still quite inadequate.

ACHD facilities have generally not yet been developed in this area. Also, tertiary care facilities for ACHD like those developed in the North America and Europe is rare.\[1\] However, 13 countries in this area already opened at least one outpatient clinic for ACHD. The number of countries that had ACHD clinic was only seven in 2007, so the number is growing rapidly.\[10,11\] Directors in most of Asia-Pacific facilities are pediatric cardiologists. Recently, adult cardiologists have joined ACHD services in some areas including Japan and Korea. In the Japanese Society for Adult CHD (JSACHD), 15% of registrants for the annual congress are adult cardiologists. Many adults with the history of Kawasaki disease (KD) and coronary artery lesion need to be followed by adult cardiologists. Adult cardiologists do not often know a lot about CHD and KD, so education of adult cardiologists is now very important. Catheter intervention has been growing, but even so, cardiovascular surgeons in CHD field, especially expert surgeons on CHD are lacking in most of the Asia countries not just in the developing countries.\[1\] Future developments in the field of surgery are mandatory, education of young surgeons and international support is also mandatory at this point of time.

CHD, SPECIFIC TO ASIA‑PACIFIC REGION‑ORIENTAL VSD, KD, AND OTHER CARDIAC DISORDERS

It is well-known that the incidence of neonates with CHDs is almost similar internationally, but the incidence in each type of CHD is different between Asia and the North America/Europe. The incidence of coarctation of the aorta and Marfan syndrome is higher in North America/Europe, but ventricular septal defect (VSD) especially subpulmonary (outlet, conus, or subarterial) VSD is much more prevalent in Asia (29-38% of total VSD) [Figure 3].\[12‑16\] In subpulmonary VSD, right coronary cusp prolapse (RCCP) will develop and result in aortic regurgitation (AR). Also, natural closure of VSD rarely happens in subpulmonary VSD. Therefore, in many cases with over moderate sized VSD, early surgical closure of the defect is inevitably. However, the natural history of this CHD especially small defect is not yet unclear. It is not known whether RCCP and AR will progressive or not. Indication of surgery or intervention has not yet established in these patients with small sized VSD with VSD size of 5 mm or smaller and/or RCCP. In future, we should work together to identify natural history of this disease and fix the guidelines. KD is also much more prevalent in Japan and Asian countries, it happens over in 10,000 children every year in these days, with >200,000 children already affected, one-third of whom need follow-up. However, tendency of having cardiovascular disease (CVD) in KD patients in future has not yet been identified.\[17\]

FUTURE PROSPECT OF CHD IN ASIA‑PACIFIC AREA

There were many scientifically interesting topics evolving in the Asia-Pacific region. Important topics include hybrid approach, cardiopulmonary transplantation, long-term problems after Fontan procedure, catheter intervention and catheter ablation, pulmonary hypertension, school-based screening on CVD including metabolic syndrome, imaging, updated cardiovascular surgery including current strategy for hypoplastic left heart syndrome, arrhythmia, and sudden death in postoperative tetralogy of Fallot (TOF), KD, infectious cardiac diseases, genetics, perinatal/fetal cardiology, aortopathy, psychosocial issues, and regenerative medicine and long-term issues and ACHD.

CURRENT STATUS AND FUTURE PROSPECT IN PEDIATRIC CARDIOLOGY

Education for young pediatric cardiologists, fellows, and residents

The number of pediatric patients has been continuously growing in this area. However, in reality, the absolute number of pediatric cardiologists and cardiovascular surgeons is small considering regional requirements. Training and education, therefore, should be focused on CHD fellows in this area, who will represent the next generation of pediatric
cardiologists and cardiovascular surgeons that will assume responsibility for this patient population.\textsuperscript{16,19} Especially for developing country, education tools on CHD for young fellows, physicians, and patients are necessary. Recently, telemedicine has been developed especially in the field of perinatal medicine. We should apply this convenient method in our field in Asia-Pacific not just for patients care, but for case discussions and consultation for specialists. Because, the number of specialists in this region is limited, these web-based techniques are currently important and convenient.

**Fetal diagnosis**

Fetal cardiac diagnosis has been widely available in these days. In general, obstetricians, perinatologists, or pediatric cardiologists perform fetal echo following guidelines on fetal diagnosis. Comprehensive diagnosis in most of significant cardiac diseases in fetuses is easily obtained nowadays. By transportation of mother and fetus to the institutions where delivery and cardiovascular surgery are available, treatment for CHD in proper timing can be done. Fetus with significant arrhythmia can also be managed through maternal intravenous antiarrhythmic medication. Early termination of pregnancy or abortion can be possible if fetus has severe CHD or in fetus with multiple anomalies. The indications and timings for termination of pregnancy for fetuses with CHD are not clear and depend on specific legal and ethical considerations.\textsuperscript{20}

**Cardiovascular surgery**

Recently, the results of surgery for complete transposition of the great arteries, Fontan procedure, revised total cavopulmonary connection (TCPC), hypoplastic left heart syndrome (HLHS), and other complex disorders have become satisfactory in several countries in Asia-Pacific. Especially in HLHS, the initial surgery has been improving in survival and hybrid approach (bilateral pulmonary artery banding with patent ductus arteriosus stenting or prolonged prostaglandin E1 infusion) has been developed.\textsuperscript{21,22} And now long-term follow-up issues of these patients after repair are the focus of research. However, in most of regions in Asia, surgical procedure is not satisfactory yet. Also, the number of surgeon is limited in most of areas in Asia, so recruitment of surgeons and their education has become important. It is extremely important to encourage more and younger surgical fellows to pursue pediatric cardiovascular surgery as a career. We should work together under APPCS to promote this movement in this area for solving this problem.

**Catheter intervention**

Even if the long-term prognosis is not clarified, in some disorders such as atrial septal defect, pulmonary stenosis, and other simple disorders; catheter intervention is the treatment of choice and replacing the role of surgery. Recent focus of intervention is for severe CHD such as severe aortic stenosis with borderline left ventricle, balloon aortic valvuloplasty as one of hybrid approaches.\textsuperscript{23} Also catheter ablation for children with Wolff-Parkinson-White (WPW), paroxysmal supraventricular tachycardia, and other tachyarrhythmias is useful and widely available. However, specialized services and personnel for these techniques are still inadequate in number in Asia, so future development and education is necessary.

**Arrhythmias and cardiac school screening**

Cardiac school screening has been developed in East Asian countries especially in Japan and Taiwan; therefore, incidence of arrhythmia\textsuperscript{24} in children has been well-known in these countries. Also it is useful to detect QT prolongation in school children for prevention of sudden death. Mode and type of QT prolongation has been studied, and gene analysis can be performed in these countries. Also cardiac school screening is useful for detecting those cardiomyopathies that cannot be diagnosed without this. In spite of limited awareness of the cost vs benefits of cardiac screening, it will perhaps be beneficial for preventing sudden death of school children. Hopefully cardiac screening in children could develop in Asian countries.

**Pulmonary arterial hypertension**

PAH is a common complication in PAH-CHD, and contributes to morbidity and mortality. There are no clear current guidelines for the treatment of PAH-CHD. There are a few trials to date investigating PAH treatment specifically on patients with CHD. However, the available data seems to demonstrate that with the advent of PAH-targeted therapies; the quality of life, exercise capacity, and outcomes in these patients is improving. In addition, PAH targeted therapies may be useful in selected patients for a combined medical-surgical approach to treatment. In patients with CHD and borderline PAH those are inoperable for a long time, repair-and-treat strategy (defect closure after targeted therapy) are applied in some patients now. Pretreating borderline PAH patients with advanced pulmonary vasodilators has the potential to demonstrate the reactivity of the pulmonary vascular bed and possibly close the cardiac defects. However, it can induce left to right shunting resulted in cardiac failure due to volume overload and this cannot be generally recommended until evidence of its safety, efficacy, and durability becomes available. So we should work together to collect data on this in future.\textsuperscript{23}

**Guidelines**

Genes in Asia-Pacific people are possibly different from Caucasians and other races. Recommended international normalized ratio (INR) levels for warfarin is different for different regions. Medical guidelines could possibly be partially different among countries due to genetic, medical system, or social differences. In Japanese Society of Pediatric Cardiology and Cardiovascular Surgery, there
are several guidelines, but these are a bit different from those in the North America and Europe. We should have multicenter international studies and collect data together with a view to and then we could establish Asia-Pacific guidelines.

**CURRENT STATUS AND FUTURE PROSPECT IN ADULTS WITH CHD**

Because of major advances in surgical and medical management, ever-increasing numbers of patients with CHD reach adulthood. With few exceptions, however, reparative surgery is not “curative”, and is specific residua and sequelae that require lifelong surveillance are common. In patients with CHD after repair, even if perfectly timed and correctly executed, disease-specific and/or operative procedure-specific anatomical and functional abnormalities can progress. These abnormalities are classified as residua, sequelae, and complication. Residua are observed before surgery and it continues after that such as right ventricular outflow tract obstruction in repaired TOF. Sequelae are not observed before surgery, but can happen after that, such as pulmonary regurgitation (PR) in repaired TOF. Complication is not observed before surgery, but it can happen unexpectedly due to surgery (nerve palsy). In each CHD, there are specific residua and sequelae especially in moderate to severe CHD after repair. Because of these complications and a host of key issues that include pregnancy and delivery, and noncardiac surgery, hepatitis, psychosocial problems such as depression, cognitive abnormalities, insurance, and extracardiac complications inherent in the comprehensive care of these patients, proper follow-up, and management are mandatory in future [Tables 1 and 2].

The most common cause of death is sudden death possibly arrhythmic followed by cardiac failure and operation including reoperation.[26]

In these contexts, I will discuss several topics that are important in clinical practice and research in ACHD field.

**Cardiac failure**

Chronic heart failure syndrome is widespread in ACHD. RV failure is common in this patient population [Table 3]. Possible pathogenetic factors for heart failure in ACHD are shown in Table 4. Chronotropic incompetence and exercise intolerance are present even in asymptomatic patients. Volume/pressure overload, with neurohormonal activation, impaired autonomic nervous function and operative scar in ACHD may all play a contributory role for CHF followed by arrhythmias and SCD. Atrial/ventricular chamber dilatation, fibrosis and dysfunction may be risk factors for cardiac failure, sustained arrhythmia and sudden death. However, clinical implication of these abnormalities remains unknown. In considering therapy for cardiac failure or sometimes for coexisting arrhythmias, care must be taken for the underlying hemodynamic substrate in each cardiac disease, particularly repairable one that might favor a surgical or catheter based approach to treatment. Typical ACHD substrates for CHF, arrhythmias and SCD are as follows; severe aortic stenosis and/or AR (superimposed coarctation of the aorta), Ebstein’s disease, congenital corrected transposition of the great arteries, TOF after repair, extra conduit repair (Rastelli procedure), complete transposition of the great arteries after Mustard or Senning operation, single-ventricle physiology, Fontan surgery, and unrepaired or post palliated cyanotic CHD.

Management of CHF in ACHD includes medication, cardiac resynchronization therapy (CRT), transplantation and reoperation, and others. Reoperation combined with arrhythmia surgery such as reoperation of repaired TOF, post extra conduit repair, etc., is popular in ACHD field, and it is different from acquired heart disease. In patients with repaired TOF with sustained ventricular tachycardia (VT) and history of syncope, reoperation with cryoablation of VT is recommended instead of implantable cardioverter defibrillator (ICD) that stabilizes hemodynamics and reduces the incidence of recurrent VT.

As the pathophysiology of CHF, exercise tolerance intolerance, cardiac dysfunction, and neurohormonal abnormalities and symptoms are very similar between CHD and acquired heart disease as mentioned above [Table 5], so management strategy for CHF in acquired heart disease such as angiotensin converting enzyme (ACE) inhibitors, angiotensin receptor blocker (ARB), and beta-blocker can be applied to ACHD. These medications can work in patients with left ventricular failure; however, no evidence for efficacy in right ventricular failure.[27-31] So in future large studies should be performed to address these questions. CRT can work in CHD and heart failure, but experience is still limited and specific features of CHD such as right ventricular failure, complete right bundle branch block (CRBBB) (not left BBB) and coronary vein abnormalities will possibly make CRT difficult.[32] Further experience in future will be necessary. Regarding CRT, there are several reports on its usefulness for ACHD. Because of the current limitations in the scope of cardiac transplantation, the future use of CRT and development of artificial heart may have a promising future.

**Arrhythmias**

Arrhythmia and sudden cardiac death in ACHD especially repaired TOF is a big issue. In patients with severe PR associated with right ventricular dysfunction are prone to die suddenly following ventricular tachycardia. Measurement of QRS duration and right ventricular size by MRI and inducible VT by electrophysiological study, all of these are advocated and have been proved useful for prediction of repaired TOF and SCD.[33] Reoperation with cryoablation for VT is effective for prevention of these crucial events. ICD is also used for prevention of sudden death as primary and secondary indication.[34] It works, but inappropriate firing and high frequency of complications
are still observed in some cases. Supraventricular tachyarrhythmias such as atrial flutter (AFL) or atrial fibrillation (Af) are common in ACHD patients long after repair, especially in those who have residual and/or sequelae lesions. Antiarrhythmic medication, catheter ablation including catheter maze procedure can work for some, but there is no consensus regarding management of Af in ACHD. Role of new oral anticoagulants for Af in Fontan and other disorders is not clear. Guidelines not only on these anticoagulants and antiplatelet agents, but also the role of catheter and surgical cryoablation in ACHD should be established in future.

**Pulmonary hypertension**

With the introduction of new pulmonary hypertension specific therapies and better pathophysiologic understanding of the disease, ACHD and associated PAH can benefit from improved quality of life and survival.[35,36]

We should work together to establish guidelines for PH management and operability of CHD and PH in ACHD. But these medications are still expensive, and many of them are not available in Asia-Pacific countries.[35] But further experience of these medication and future development of this kind of medication is anticipated. The pathophysiology of systemic complications in cyanotic ACHD, those are major morbidity and mortality, should be further investigated and proper management should be established for improvement of QOL in these patients.

**Pregnancy and delivery**

Half of the ACHD patients are female. As a result, pregnancy and delivery are important events in their lives. During pregnancy and delivery, hemodynamic and other physiologic changes can happen [Table 6]. Cardiac disease is a leading cause of maternal death in some countries and most affected women with cardiac disease have CHD. The number of such patients at risk is expected to grow. Women with PAH, severe left ventricular outflow tract stenosis (LVOTS), cyanotic CHD, aortic root dilatation, cardiac dysfunction, and mechanical valve carry a high risk for pregnancy and delivery [Table 7]. Most frequent complications are CHF or arrhythmia followed by thrombosis. Risk stratification regarding pregnancy in CHD patients relates to functional status of the patient and lesion specificity. Timely pre-pregnancy counseling should be offered to all women with CHD to prevent avoidable pregnancy-related risks and that allows patients to plan

**Table 3: Cardiac failure; especially right ventricular failure in adult congenital heart disease: Background, morphology, and physiology**

| Right ventricle: Subpulmonary RV and systemic RV |
|-----------------------------------------------|
| Systemic RV with/without AV valve regurgitation |
| Complete transposition of the great arteries post-atrial switch |
| Congenital corrected transposition of the great arteries |
| Single RV |
| Subpulmonary RV |
| L-R shunt (atrial septal defect) |
| Tricuspid regurgitation (Ebstein’s disease and tetralogy of Fallot post repair) |
| Pulmonary stenosis, pulmonary hypertension, and Eisenmenger syndrome |
| Pulmonary regurgitation (tetralogy of Fallot post repair) |

**Left ventricle**

| Pressure overload |
| Coarctation of the aorta and aortic stenosis |
| Volume overload |
| Aortic regurgitation (bicuspid aortic valve, tetralogy of Fallot, or cyanotic congenital heart disease with pulmonary atresia or stenosis) |
| Left to right shunting (ventricular septal defect and patent ductus arteriosus) |
| Mitral regurgitation |

**Table 4: Possible pathogenetic factors for heart failure in adult congenital heart disease**

| Cyanosis |
| Pressure/volume overload |
| Residua and sequelae after repair |
| Poor intraoperative myocardial preservation |
| Artificial material (large ventricular patch) |
| Ventricular incisions/scar |
| Arhythmias (bradyarrhythmias and tachyarrhythmias) |
| Abnormal ventricular-ventricular interaction |
| Myocardial ischemia (ventricular hypertrophy and abnormal coronary supply) |
their lives. Adequate care during pregnancy, delivery, and the postpartum period requires a multidisciplinary team approach with cardiologists, obstetricians, and anesthesiologists. Successful pregnancy is feasible for most women with CHD at relatively low risk when appropriate counseling and optimal care are provided. We need more data on pregnancy and delivery in ACHD. Already registration system on pregnancy and delivery in cardiac disease including ACHD has been started in Europe under European Society of Cardiology and in Japan under JSACHD.

We should also collect data among Asian countries. Collaboration of multidisciplinary team will be necessary for the care of pregnancy and delivery, and it should be established in Asian-Pacific countries. In Asian countries, still rheumatic heart disease is prevalent, so proper management plan for female with this disorder is also important.

**Psychosocial issues**

Currently, most researches on the psychosocial aspects of ACHD patients are conducted in Europe and the North America using questionnaires and interviews. However, the results of these studies are inconsistent. Some questionnaire studies showed no differences in emotional and social functioning between ACHD patients and the reference group,

while the psychiatric diagnostic interview with adult patients has indicated that one- to two-thirds of these patients have some psychosocial issues. Apparently, these reported differences in psychosocial adjustment depend on the country in which patients reside and might reflect an aspect of the particular sociocultural environment, such as the healthcare system and mental health treatment. In Japanese Study, ACHD patients often have psychosocial difficulties, and the psychosocial factors those influence patient’s mental health are social problem-solving, independence, and self-esteem. However, patients have poor abilities in all of those areas and hence, they run the risk of losing their mental health. But these trends may be different from country to country in Asia-Pacific, so we should collect data together and analyze them in future.

**Future possibility of acquired CVD**

Recently several papers have been published regarding early onset CVD in ACHD patients including coarctation of the aorta and other coronary artery abnormalities and post coronary artery manipulation surgery such as Jatene and Ross procedure. In general, cyanotic CHD is preventive for CVD [Table 8]; however, even in these patients, CVD and atherosclerosis can happen in presence of risk factors for ischemic cardiac disease such as hypertension, dyslipidemia, diabetes mellitus, and metabolic syndrome. In patients with CHD, the incidence of obese and metabolic syndrome is higher than general population in Japan [Table 9].

Imaging modalities including CT and other laboratory technique have been developed for early detection of CVD, so we should conduct a research on this for future prevention of CVD in ACHD and KD patients.

**Establishment of caring system of ACHD**

Regarding caregivers for ACHD, proper transition from pediatric cardiologists and cardiovascular surgeons to ACHD specialists and/or cardiologists who are well-trained in the field of ACHD is necessary. Provision of comprehensive care by multidisciplinary teams including adult and pediatric cardiologists and cardiovascular surgeons was the fundamental feature in care facilities for ACHD. Also anesthesiologists have a major role on taking care of ACHD patients during cardiac and noncardiac surgery. Training and education should be focused on the ACHD fellows who represent the next generation of cardiologists and cardiovascular surgeon that will assume responsibility for this patient population.
developing countries, the number of ACHD will grow in near future, so system of transition and caring for ACHD is necessary to start at this point of time.\cite{53} Working closely together in Asia-Pacific countries is important towards this goal.

**Aortopathy: New clinical entity**

Bicuspid aortic valve and/or coarctation of the aorta are consistently associated with abnormalities of the medial layers of the aortic wall. Medial abnormalities in ascending aorta were prevalent in a variety form of CHD such as single ventricle, persistent truncus arteriosus, transposition of the great arteries, hypoplastic left heart syndrome, and TOF, encompassing a wide age range, and may predispose to dilatation, aneurysm, rupture, and AR; necessitating aortic valve and root surgery.\cite{54,55} This dilatation can develop in CHD patients even without a stenotic region. These CHD patients exhibit ongoing dilatation of the aortic root and reduced aortic elasticity and increased aortic stiffness\cite{56} that may relate to intrinsic properties of the aortic root. The concept of aortic dilatation amounts to a paradigm shift from so-called post-stenotic dilatation, to primary intrinsic aortopathy. The aortic dilatation and increased stiffness can induce aortic dilatation and AR, provoke left ventricular hypertrophy, reduce coronary artery flow, and eventually result in left ventricular failure in future. We can recognize this association of aortic pathophysiological abnormality, aortic dilatation and aorto-left ventricular interaction as a new clinical entity: “Aortopathy”. We should work together to follow these patients focusing on progression of aortic root dilatation as natural or postoperative history. Also we should find the medication for preventing this dilatation and stiffness using beta blocker/ACE inhibitor or angiotensin II receptor antagonist (ARB).

### COLLABORATION BETWEEN ASIA-PACIFIC AND THE NORTH AMERICA, EURO, AND OTHER INTERNATIONAL REGIONS

We, APPCS and APSACHD, should have a regular biannual meeting. In addition, we should be working together on clinical practice and researches inside Asia-Pacific and beyond collaborate with Association of European Pediatric Cardiology (AEPC) and CVD in the Young (CVDY) in AHA. I believe international collaboration is important to promote friendship, mutual understanding, and new development of CHD field; and we should have a regular joint session among these groups. By having these sessions, all speakers and attendants will feel close each other and are easily able to share the data and various issues. So, I strongly feel that it is a proper timing (joint sessions are establishing internationally) for us to make a great step forward for close international collaboration in research and clinical support from now and then. Especially we are happy that young people such as fellows, young physicians, nurses, co-medicals and technicians, and patients can join and enjoy this trend.

| Table 8: Paucity of coronary atherosclerosis and influencing factors for low incidence of atherosclerosis in cyanotic ACHD |
|--------------------------------------------------|
| Adults with CHD have low levels of total cholesterol, LDL, and HDL cholesterol |
| Low levels of lipid profile persist after surgical elimination of cyanosis influencing factors including anti-atherogenic properties of hypoxemia |
| Hypocholesterolemia |
| Increased nitric oxide |
| Increased serum bilirubin levels |
| Low platelets account |
| **ACHD:** Adult congenital health disease; **LDL:** Low density lipoprotein; **HDL:** High density lipoprotein |

| Table 9: Clinical Manifestations of cardiovascular disease in ACHD |
|--------------------------------------------------|
| Adult congenital heart disease patients have a similar prevalence of risk factors for CVD as general population |
| Cyanotic ACHD has lower incidence of CVD |
| Acanotic ACHD has similar prevalence of CVD as general population |
| Increased risk of coronary atherosclerosis is observed in congenital coronary artery anomalies, complete transposition of the great arteries post arterial switch, and coarctation of the aorta |
| Aortopathy may be an additional risk factor for CVD |
| Prevention for CVD is even more important in young adults with CHD |
| **CVD:** Cardiovascular disease; **ACHD:** Adult congenital health disease |

Now it is the moment to nail down this movement. We should work together to keep continuing this with thoughtfulness and mutual respect, and walk hand in hand for the promising future. Also we should educate and develop medical staffs in pediatric cardiology and ACHD field in Asia-Pacific region beyond country barriers.

### SUMMARY

- Formal education and training systems for CHD practitioners are still lacking in developing countries in Asia. Collaborative work and unconditional support on these problems among us, Asia-Pacific is mandatory
- Further expansion of this population and evolution of specialized care facilities for pediatric cardiology and ACHD can be anticipated in the Asia-Pacific countries
- Training and education focused on the trainees who represent the next generation of pediatric and adult cardiologists that will assume responsibility for this patient population is necessary
- We now have biannual Congress of the APPCS and APSACHD and these societies are actively moving forward in cooperation with the Societies in Euro and North America
- Collaboration, research work, and medical support for developing countries have begun and should develop
- We should walk together to make further progress of our mutual relationship toward activities such as international collaborative studies and support project
- The next APPCS and APSACHD Congress will be held
in New Delhi in 2014 followed by Shanghai in 2016. We will work together to keep our regional and international collaborations progressing towards a promising future.

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