

抄録／ Abstracts

S1-1

沖縄におけるFontan術後成人患者の現状と課題

Current issues for adult Fontan patients in Okinawa

竹蓋 清高, 島袋 篤哉, 佐藤 誠一, 西畑 昌大, 塚原 正之, 内田 英利, 中矢代 真美
沖縄県立南部医療センター・こども医療センター 小児循環器内科

Kiyotaka Takefuta, Atsuya Shimabukuro, Seiichi Sato, Masahiro Nishibata, Masayuki Tsukahara, Hidetoshi Uchida, Mami Nakayashiro
Department of pediatric cardiology, Okinawa Nambu Medical Center・Children's Medical Center

Background: Okinawa has struggled to manage complex congenital heart disease (CCHD) due to historical and geographic factors. Since our hospital opened in 2006, CCHD patients in Okinawa have been concentrated to one institution. The diverse background of the adult Fontan patients continues to be a large issue.

Methods: From 2006 to 2018, 41 adult Fontan patients (>15yo) were identified from our hospital clinical database. Data was collected retrospectively.

Results: Patient demographics: male 28 (68%), median age 21yo, systemic right ventricle 22 (54%), 20% had SpO₂ < 93%. Follow-up heart catheterization was performed in 36 patients. Patients with surgery in mainland Japan was 43%. 15% had history of dropout. Complications: 33% of patients had arrhythmia, 2.5% had plastic bronchitis, 13% had Fontan associated liver disease, 15% had protein-losing enteropathy. Mortality: two patients died at 30 years and 33 years of age.

Conclusion: Adult Fontan patients in Okinawa have diverse past background which can be a factor for high dropout rate and morbidity. Collaboration with the previous hospitals is crucial for a better understanding and management of our cohort.

S1-2

女性とFontan

Women with Fontan Circulation

島田 衣里子, 篠原 徳子, 稲井 慶, 杉山 央
東京女子医科大学 循環器小児科

Eriko Shimada, Tokuko Shinohara, Kei Inai, Hisashi Sugiyama
Department of Pediatric Cardiology, Tokyo Women's Medical University

The gender/sex differences in prognosis have been well recognized in cardiovascular (CV) disease such as ischemic heart disease, heart failure, and stroke. A few studies have also been reported the impact of gender on several CV events among adults with congenital heart disease. In adult patients with Fontan circulation, not only the CV problems but also the various systemic complications due to the unique hemodynamics are well known. However, few studies have focused on whether the gender influences outcomes or CV events in Fontan patients. We should also consider about the specific problems in females such as menstruation, hormone therapy, gynecological surgery, pregnancy, and menopause. This talk will summarize the problems and the management with women after Fontan operation.

S1-3

術前から予想する failing Fontan

Prediction of failing Fontan physiology before completion of the procedure

豊野 学朋

秋田大学 小児科学講座

Manatomo Toyono

Department of Pediatrics, Akita University

The Fontan procedure is a reparative surgery when a 2-ventricle physiology is impossible. This can be usually performed as a 2 or more-stage procedure. A bidirectional Glenn anastomosis is performed as a previous stage and is followed by completion of the Fontan. Although important benefits have been fulfilled, a number of adverse results of the Fontan physiology have been recognized. After the bidirectional Glenn, many criteria can identify individuals who do not well after the Fontan (Table). Individuals who meet 1 or more of these criteria have a risk of failing Fontan physiology, if completed without adequate interventions. However, even the well-functioning Fontan physiology eventually fails as chronic non-pulsatile pulmonary flow, elevated systemic venous pressures, and limited cardiac output accelerate multi-organ failure. An earlier version of the Fontan procedure to be seen in adults includes direct right atrial appendage-to-pulmonary artery connection. This version is associated with right atrial enlargement, increased systemic venous pressure, atrial arrhythmia, formation of atrial thrombi and pulmonary embolism.

Table. Preoperative variables predicting failing Fontan physiology

Vascular properties	Ventricular function	Valve function	Arrhythmia	Anatomy
<ul style="list-style-type: none"> ● Elevated mean pulmonary artery pressure ● Elevated pulmonary vascular resistance ● Distortion or narrowing of the pulmonary arteries ● Elevated systemic vascular resistance 	<ul style="list-style-type: none"> ● Decreased systemic ventricular ejection fraction ● Dyssynchronous ventricular wall motion ● Elevated systemic ventricular end-diastolic pressure 	<ul style="list-style-type: none"> ● Significant systemic valve regurgitation 	<ul style="list-style-type: none"> ● Tachyarrhythmia ● Bradyarrhythmia 	<ul style="list-style-type: none"> ● Hypoplastic left heart syndrome ● Heterotaxy ● Single morphologically right ventricle ● Obstructive bulboventricular foramen

S1-4

成人期フォンタンに対する外科治療

Surgical approach for failed Fontan in adult

帯刀 英樹¹⁾, 坂本 一郎²⁾, 藤田 智¹⁾, 永田 弾³⁾, 筒井 裕之²⁾, 塩瀬 明¹⁾

1)九州大学病院 心臓血管外科, 2)九州大学病院 循環器内科, 3)九州大学病院 小児科

Hideki Tatewaki¹⁾, Ichiro Sakamoto²⁾, Satoshi Fujita¹⁾, Hazumu Nagata³⁾, Hiroyuki Tsutsui²⁾, Akira Shiose¹⁾

1) Department of Cardiovascular Surgery, Kyushu University Hospital, 2) Department of Cardiology, Kyushu University Hospital,

3) Department of Pediatrics, Kyushu University

Objective: Various complication including atrioventricular regurgitation, arrhythmias developed in adult after Fontan completion. We present our surgical results for failing Fontan in adult.

Methods: We reviewed 14 patients with single ventricle physiology who underwent surgical interventions since 2015. The types of initial Fontan completion were atriopulmonary connection in 3, lateral tunnel in 6, extracardiac conduit in 3 and intraatrial conduit in 2.

Results: The mean age at surgery was 30.2 years old (range, 20 to 46 years). For Fontan conversion in 11 patients, total cavopulmonary connection was performed with extracardiac conduit. Nine patients required surgical interventions for systemic ventricle system including atrioventricular valve repair in 7, aortic valve replacement in one and ascending aorta replacement in one. The NYHA functional class significantly improved in all patients. During study period, we employed our strategy including modification of surgical techniques and perioperative management to improve surgical outcomes.

Conclusions: This study demonstrated favorable outcomes after surgical interventions for adult patients with single ventricle physiology.

S1-5**Failing Fontan 予防のための内科治療
Medical management for Fontan circulation**

元木 博彦

信州大学医学部附属病院 成人先天性心疾患センター

Hirohiko Motoki

Adult Congenital Heart Disease Center, Shinshu University Hospital

Fontan operation is a final definitive palliative procedure for patients with complex congenital heart disease (CHD) with functional single ventricular (FSV) physiology. The goal of this procedure includes elimination of hypoxia and volume overload to the functional systemic ventricle. Total cavo-pulmonary connection (TCPC) is now a contemporarily main procedure for CHD patients with FSV physiology; however, persistent high central venous pressure (CVP) and low cardiac output deteriorate multi-organ function in Fontan patients with chronic heart failure (HF), "Failing Fontan". Deep understanding of adult Fontan pathophysiology as multiorgan diseases due to chronic venous hypertension as well as chronic heart failure is mandatory as to how to manage for expecting better long-term outcome in these patients.

S2-1**岡山大学病院ACHDセンターの試み
ACHD Management System of Okayama University Hospital**赤木 禎治, 杜 徳尚, 大月 審一, 増山 寿, 大西 秀樹, 木野村 賢, 大森 一弘, 笠原 真悟, 伊藤 浩
岡山大学病院 成人先天性心疾患センター

Teiji Akagi, Norihisa Toh, Shinichi Otsuki, Hisashi Masuyama, Hideki Onishi, Ken Kinomura, Kazuhiro Ohmori, Shingo Kasahara, Hiroshi Ito

Adult Congenital Heart Disease Center, Okayama University Hospital

Up to now, pediatric cardiologists have been contributed as mail roll for ACDH patient's management in Japan. However, in fact, such occasions are limited even in major hospital, because of daily patient's care as in each cardiac specialists. Additionally, it is still very difficult to establish the independent ACHD unit under the current Japanese condition. To resolve such limitations, we try to join the educational seminar by Japanese Society for ACHD, especially case discussion. Cardiac anatomy of complex heart disease, post-operative hemodynamic features were rare experiences for adult cardiologists. Also, management of arrhythmia, pregnancy and social health care system were commonly required for the management of ACHD patients. Especially in patients after Fontan procedure, we need more scientific evidence. To increase the adult congenital heart disease specialists, such education system should be functioned continuously.

Environments of ACHD patient's care has been changed rapidly. Roll of adult cardiologists is very important to establish the ACHD management system. To fulfill the national management system, human resource is essential especially the involvement of adult cardiologists.

S2-2

神戸大学ACHDセンターの実際 ～今日は全てみせちゃいます～ Real practice of adult congenital heart disease at Kobe University

松本 賢亮¹⁾, 城戸 佐知子²⁾, 鈴木 麻希子¹⁾, 須藤 麻貴子¹⁾, 平田 健一¹⁾

1) 神戸大学附属病院 循環器内科, 2) 兵庫県立こども病院 循環器内科

Kensuke Matsumoto¹⁾, Sachiko Kido²⁾, Makiko Suzuki¹⁾, Makiko Suto¹⁾, Ken-ichi Hirata¹⁾

1) Division of Cardiovascular Medicine, Department of Internal Medicine, Kobe University Graduate School of Medicine, Kobe, Japan.,

2) Division of Cardiology, Kobe Children's Hospital, Kobe, Hyogo, Japan.

One day, a pediatrician who is working at a neighboring children's hospital claimed us that "Recently, the number of the patients with grown-up congenital heart disease is extremely increasing. So, please support the medical care for these patients!". To comply with the proposal, "Kobe University Adult Congenital Heart Disease (ACHD) Center" was launched in 2013. Since then, a total number of the patients who are managed at our ACHD center have increased to more than 400 during 5 years. Today, in proportion to the increased number of the outpatients, our ACHD center has grown up to a "team" that consists of 3 internists specialized for cardiovascular medicine and 1 pediatrician for congenital heart disease. Through 5 years practice as ACHD center, we indeed had a lot of valuable experiences, and could build the tight relationship with other department, including obstetrics, cardiac surgery, radiology, and even with palliative care unit. Recently, we can extend our experience to the medical research, and to the education for the residents and even for the physicians who have other sub-specialty.

On the other hand, we sometimes experience difficulties in managing ACHD clinic, including understanding the complex anatomy, hemodynamics, and specialized therapy for ACHD patients, and also feel the difficulty in the establishment of the doctor-patients and/or doctor-parents relationships. And, another important problem is the increasing severity of the patients with ACHD.

Today, we would like to disclose the real practice of Kobe University ACHD center including both going well and wrong.

S2-3

小児循環器科のない施設におけるACHD診療

ACHD center without the department of pediatric cardiology

椎名 由美, 木島 康文, 福田 旭伸, 杉淵 景子, 小宮山 伸之, 丹羽 公一郎

聖路加国際病院 心血管センター

Yumi Shiina, Yasufumi Kijima, Terunobu Fukuda, Keiko Sugibuchi, Nobuyuki Komiyama, Koichiro Niwa
Cardiovascular Center, St.Luke's International Hospital

- Our center was started in 2011 by two pediatric cardiologists. Now 3 adult cardiologists (ACHD specialists) and 1 pediatric cardiologist (ACHD specialist) see patients both CHD and non-CHD (adult patients in general cardiology) patients.
- Adult patients are transferred from other children hospital, mainly from the eastern part of Tokyo and Chiba.
- There is no pediatric cardiovascular department in our own hospital. There is no pediatric surgeon as well.
- Our hospital is a private hospital, not a public one; therefore, patient-friendly service and ER system. Palliative care support system are well-established. On the other hand, it is important for cardiovascular center to make benefit.

S2-4

長野モデルの苦労話

Hardships around Nagano Model

元木 博彦

信州大学医学部附属病院 成人先天性心疾患センター

Hirohiko Motoki

Adult Congenital Heart Disease Center, Shinshu University Hospital

Recently, adult patients with congenital heart disease (ACHD) are facing urgent issues regarding healthcare systems in Japan. We have operated the ACHD center in Shinshu University since June 2013 in collaboration with Nagano children's hospital. Details of our efforts with respect to the running of this center is expected to provide useful information for every adult cardiologist and cardiovascular institute, which handles the care of patients with ACHD in daily practice. We sought to clarify the issues that were noted during the process of establishment of the ACHD care system.

S2-5

九州におけるACHD診療

ACHD Practice in Kyushu

坂本 一郎¹⁾, 石北 綾子¹⁾, 永田 弾²⁾, 帯刀 英樹³⁾, 梅本 真太郎¹⁾, 塩瀬 明³⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 小児科, 3) 九州大学病院 心臓血管外科

Ichiro Sakamoto¹⁾, Ayako Ishikita¹⁾, Hazumu Nagata²⁾, Hideki Tatewaki³⁾, Shintaro Umemoto¹⁾, Akira Shiose³⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Medicine, Kyushu University Hospital, 2) Department of Pediatrics, Kyushu University Hospital,

3) Department of Cardiovascular Surgery, Kyushu University Hospital

We started Adult Congenital Heart Disease (ACHD) outpatient clinic in September 2009. It has been conducting clinical care mainly focusing on the transition from Fukuoka Children's Hospital. Transition from a children's hospital was one of the major objectives, but recently introductions of undiagnosed cases and dropped out cases from nearby general hospitals are increasing. It has become able to play a certain role for adult cardiologists working at non-ACHD specialized hospital.

At the beginning of the ACHD outpatient clinic, hospitalized cases were few. It was necessary to do other practice than ACHD practice. Currently, both outpatient and hospitalized patients are increasing, manpower is insufficient for ACHD practice. Although there are few pediatricians in rural areas, adult cardiologist is not enough to focus on sub specialty such as ACHD. We must make a attractive system to learn ACHD practice effectively and invite young cardiologists.

S3-1

シャント閉鎖後PAHの治療戦略と注意点

Treatment strategy of PAH after shunt closure

相馬 桂^{1,2)}, 稲葉 俊郎¹⁾, 八尾 厚史^{1,3)}1) 東京大学医学部附属病院 循環器内科, 2) 東京大学医学部附属病院 22世紀医療センター コンピューター画像診断学,
3) 東京大学 保健健康推進本部Katsura Soma^{1,2)}, Toshiro Inaba¹⁾, Atsushi Yao^{1,3)}

1) Department of Cardiovascular Medicine, The University of Tokyo Hospital,

2) Department of Computational Diagnostic Radiology and Preventive Medicine, The University of Tokyo Hospital,

3) Division for Health Service Promotion, The University of Tokyo.

Treatment strategy for adult congenital heart disease with shunt-associated pulmonary arterial hypertension (ACHD-sPAH) is still under consideration. Previously, residual sPAH after shunt closure had been reported to be associated with a poor prognosis, suggesting that the shunt closure for ACHD-sPAH was considered as contraindication. However, recently PAH drugs have improved the hemodynamics and the prognosis of sPAH after shunt closure as well as sPAH without shunt closure or Eisenmenger syndrome. Pathophysiology and the effectiveness of PAH drugs are quite similar between idiopathic PAH (IPAH) and sPAH. In Japan, the pressure-oriented medical therapy with aggressive combined use of PAH drugs successfully improved the prognosis of IPAH. Therefore, we have been applying the similar strategy for ACHD-sPAH, and found the similar or even better results. It is concerned that only shunt closure is performed without medical treatment with PAH drugs, because we sometimes experienced worsening of sPAH after shunt closure. On the other hand, under appropriate medication, residual or worsened sPAH after shunt closure continuously and steadily improves over the long term. In this talk, we'd like to share the experience of treating PAH after shunt closure, and discuss on the treatment strategy after shunt closure.

S3-2

IPAH with small ASDのshunt閉鎖は禁忌でいいのか？

Should we leave shunt untreated in IPAH patients with small ASD?

松原 広己

国立病院機構岡山医療センター 循環器内科

Hiromi Matsubara

Department of Cardiology, National Hospital Organization Okayama Medical Center

The prognosis of Idiopathic Pulmonary Arterial Hypertension (IPAH) has been miserable. Even with recent development of many pulmonary hypertension specific drugs, 5 years survival rate of the patients in European countries is only about 60%. Thus, balloon atrial septostomy is still mentioned in European guidelines for pulmonary hypertension as a palliative or bridging procedure for severe right heart failure. In other word, small ASD in PAH patients would be regarded as un-closable under this circumstances. However, situation is totally different in Japan. Early initiated upfront combination therapy with rapid up-titration of parenteral prostanoid was resulted in remarkably improved outcomes. In our center, 15 years survival rate of the patients has been already reached about 80%. None of the patients died or lung transplanted who started treatment during past 10 years. Since most of the patients did not have any intracardiac shunt, it would be clear that there is no need to leave shunt untreated in IPAH patients with small ASD. In this presentation, the author at first summarizes state of the art of IPAH treatment and then shows long term outcome of ASD closure in IPAH patients.

S3-3

シャント性心疾患関連肺高血圧症に対する Treat and Repair の適応と方法

Indication and Strategy of Treat and Repair for patients with pulmonary arterial hypertension associated with congenital cardiac defects

赤木 達

岡山大学大学院医歯薬学総合研究科 循環器内科学

Satoshi Akagi

Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Science

Congenital cardiac defects such as atrial septal defects and ventricular septal defects are one of the causes of pulmonary arterial hypertension (PAH). Recently PAH-specific drugs have developed, which leads to concept of treat and repair strategy in treatment for patients with PAH associated with congenital cardiac defects. This strategy includes 'treatment' with PAH-specific drugs initially and then 'repair' by closure of the cardiac defect. Treat and repair strategy could prevent to proceed Eisenmenger syndrome and improve pulmonary hypertension after shunt closure. On the other hands, use of PAH-specific drugs under cardiac defects might cause to increase share stress in pulmonary arteries, which might worsen the pulmonary hypertension. We experienced 13 cases of treat and repair strategy in patients with PAH associated with cardiac defects. In this session we discuss the indication and strategy for treat and repair based on our experiences.

S3-4

たかがASD、されどASD、成人では

Don't despise ASD in adulthood as the simplest cardiac anomaly.

根本 慎太郎

大阪医科大学医学部 外科学講座胸部外科学小児心臓血管外科

Shintaro Nemoto

Department of Thoracic and Cardiovascular Surgery, Osaka Medical College

Atrial septal defect (ASD) is the simplest cardiac anomaly usually closed in childhood when the defect is considered to be clinically and hemodynamically significant. However, the defects are occasionally undetected in early life and suddenly become a subject of treatment discussion when patients grow old. Although closure of the defect is always considered as a mainstay to relief clinical symptoms together with hemodynamic improvement regardless of age, a certain proportion of patients do not receive the benefits of the closure. This unsolved reality of the current treatment of ASD is possibly attributed to the development or persistence of pulmonary arterial hypertension (PAH) after the closure in adulthood which is associated with a poor exercise capacity, a variety of morbidity, and unfortunate prognosis.

In this presentation of the symposium, the following subjects will be reviewed and discussed to provide a clue to fight against PAH in adult ASD; effect of age at closure and defect size on pulmonary artery pressure, prevalence of PAH before and after the closure, especially worsening of PAH after the closure, predictors of development or persistence of PH after the closure, and currently reported modifications and pitfalls of surgical or device closure.

S3-5

Eisenmenger 症候群の定義とその不可逆概念はこのままでいいのか？

Conventional "Eisenmenger syndrome" contains borderline hemodynamics in the current era

椎名 由美, 木島 康文, 福田 旭伸, 小宮山 伸之, 丹羽 公一郎

聖路加国際病院 心血管センター 循環器内科

Yumi Shiina, Yasufumi Kijima, Terunobu Fukuda, Nobuyuki Komiyama, Koichiro Niwa
Cardiovascular Center, St.Luke's International Hospital

Adults with congenital heart disease who was diagnosed with conventional "Eisenmenger syndrome" in their childhood show a wide variety of hemodynamics as follows:

A. Eisenmenger Syndrome

Large congenital systemic-to-pulmonary shunt resulting in pulmonary vasculopathy, increased PVR and shunt reversal. Cyanosis and erythrocytosis present, with mult-system involvement.

B. PAH associated with a predominant systemic-to-pulmonary shunt

Moderate to large shunts with mild or moderately increased PVR. May be correctable or non-correctable.

C. PAH associated with a small defect

Significantly elevated PVR in the presence of a small defect (ASD <2 cm diameter, VSD <1 cm diameter). Behaves similarly to idiopathic PAH

D. PAH following a repaired defect.

PAH persists after closure or develops/recurs following closure.

E. Segmental PH

Encompasses any condition with abnormal underlying cardiac or vascular anatomy, usually including varied sources of pulmonary blood supply, which results in distal pulmonary vascular disease that affects various lung segments to differing degrees.

In the current era of "treat and repair" strategy, the appropriate patients' selection is very important. PVR is between 2.3 and 4.6 WU (PVRi 4–8 WU/m²) is not straightforward. Clinical decision-making is individualized and may be debated.

S3-6

Eisenmenger 症候群に対する疾患標的療法の現況

Current status of disease targeting therapy for adult patients with Eisenmenger syndrome

坂崎 尚徳¹⁾, 丹羽 公一郎²⁾, 武田 充人³⁾, 小野 博⁴⁾, 高月 晋一⁵⁾, 堀米 仁志⁶⁾, 犬塚 亮⁷⁾, 福島 裕之⁸⁾, 森 善樹⁹⁾, 立野 滋¹⁰⁾, 市田 蓂子¹¹⁾, 糸井 利幸¹²⁾, 奥村 謙一¹²⁾, 小垣 滋豊¹³⁾, 脇 研自¹⁴⁾, 赤木 禎治¹⁵⁾, 須田 憲治¹⁶⁾

1) 兵庫県立尼崎総合医療センター 小児循環器内科, 2) 聖路加国際病院心血管センター 循環器内科,

3) 北海道大学 小児科, 4) 国立成育医療研究センター 小児循環器科,

5) 東邦大学医療センター大森病院 小児医療センター 小児科, 6) 筑波大学 小児科, 7) 東京大学 小児科,

8) 慶応義塾大学 小児科, 9) 聖隷浜松病院 小児循環器科, 10) 千葉県立循環器病センター 成人先天性心疾患診療部,

11) 富山大学 小児科, 12) 京都府立医科大学 小児循環器科, 13) 大阪大学 小児科, 14) 倉敷中央病院 小児科,

15) 岡山大学 循環器疾患診療部, 16) 久留米大学 小児科

Hisanori Sakazaki¹⁾, Koichiro Niwa²⁾, Atsuhito Takeda³⁾, Hiroshi Ono⁴⁾, Shinichi Takatsuki⁵⁾, Hitoshi Horigome⁶⁾,

Ryo Inuzuka⁷⁾, Hiroyuki Fukushima⁸⁾, Yoshiki Mori⁹⁾, Shigeru Tateno¹⁰⁾, Fukiko Ichida¹¹⁾, Toshiyuki Itoi¹²⁾,

Kenichi Okumura¹²⁾, Shigetoyo Kogaki¹³⁾, Kenji Waki¹⁴⁾, Teiji Akagi¹⁵⁾, Kenji Suda¹⁶⁾

1) Department of Pediatric Cardiology, Hyogo prefectural Amagasaki Medical Center,

2) Cardiovascular Center, St Luke's International Hospital, 3) Department of Pediatrics, Hokkaido University Hospital,

4) Department of Pediatric Cardiology, National Center for Child Health,

5) Department of Pediatrics, Toho University Omori Medical Center, 6) Department of Pediatrics, Tsukuba University Hospital,

7) Department of Pediatrics, Tokyo University Hospital, 8) Department of Pediatrics, Keio University Hospital,

9) Department of Pediatric Cardiology, Seirei Hamamatsu General Hospital,

10) Department of Adult congenital heart disease, Chiba Cerebral and Cardiovascular Center,

11) Department of Pediatric, Toyama University Hospital,

12) Department of Pediatric Cardiology and nephrology, University Hospital Kyoto prefectural University of Medicine,

13) Department of Pediatrics, Osaka University Hospital, 14) Department of Pediatrics, Kurashiki Central Hospital,

15) Department of Cardiovascular Medicine, Okayama University Hospital,

16) Department of Pediatrics, Kurume University Hospital

Background: Recently some articles on efficacy of disease targeting therapy (DTT) for adult patients with Eisenmenger syndrome were have been published. However, proper timing of initiation and escalation of DTT is unknown.

Method & Results: By the prospective observational multicenter study, clinical data of 66 patients (pts) treated with at least one DTT were examined. At the enrollment, 31pts were treated with single DTT (bosentan 38%, tadarafil 24%), 19pts with dual, and 10pts with triple DTT. 11pts started or escalated DTT proactively, and 10 of them were clinically stable at last clinical visits, despite of one with discontinuation of bosentan because of severe leg edema. Remaining one made the sift to terminal care because of uterus cancer. However, 15 pts started or escalated DTT after clinical deterioration, and 8 of them resulted in clinical improvement, 5 had no significant change and 2 had clinical worsening.

Conclusions: Proactive therapy may be acceptable to maintain clinical condition and initiation or escalation of DTT after deterioration was effective in half of patients.

S4-1

ACHD患者の妊娠前・妊娠管理

Pre-pregnancy, pregnancy management for ACHD patients

田中 博明

三重大学医学部 産科婦人科学

Hiroaki Tanaka

Department of Obstetrics and Gynecology, Mie University Faculty Medicine

Women with adult congenital heart disease (ACHD) present in a variety of states, depending on their underlying diseases and courses. Many of them, while appearing outwardly healthy, have various problems. During pregnancy, increased cardiac output accompanying elevated circulating blood volume can change hemodynamics in dynamic ways, such as by reducing vascular resistance. Therefore, the risks of pregnancy should be assessed in women with ACHD before they become pregnant. It is extremely important that care be provided by a team that includes an obstetrician, pediatric cardiovascular specialist, cardiovascular specialist, cardiovascular surgeon, anesthesiologist, and other specialists. Pregnancy should then be managed based on an understanding of the aforementioned changes in hemodynamics caused by pregnancy and by anticipating any cardiovascular events that could occur during pregnancy. Depending on the severity, many pregnant women with ACHD have preterm deliveries.

We surveyed pregnant women with complicating cardiovascular diseases at 424 perinatal care facilities in Japan over 1 year from April 2013 to March 2014, then investigated cardiovascular event risk factors in the 302 cases that were reported. New York Heart Association (NYHA) classification class I was associated with significantly lower incidence of cardiovascular events ([aOR] 0.02, 95%CI 0.01-0.07, $p < 0.001$). Pre-pregnancy mechanical valve replacement and pre-pregnancy drug administration were significant risk factors for cardiovascular events (valve replacement: [aOR] 87.17, CI 13.94-1690.51, $p < 0.001$; medication: [aOR] 4.48 CI, 1.14-25.79, $p = 0.04$). The number of pregnant women with cardiovascular complications is expected to increase due to an increase in women with ACHD of reproductive age, more geriatric pregnancies, advances in reproductive care, and other factors. This study indicates that at-risk women require not only perinatal management, but should receive care at specialized facilities able to provide sufficient cardiovascular management.

S4-2

ACHD 合併妊娠と児の合併症

Fetal and neonatal complications in women with congenital heart disease

堀内 縁

国立循環器病研究センター 周産期・婦人科

Chinami Horiuchi

Departments of Perinatology and Gynecology, National Cerebral and Cardiovascular Center

Cardiac adaptations are required during pregnancy in order to ensure adequate blood perfusion of the uteroplacental circulation. An already functionally deteriorated cardiovascular system may not adapt for such pregnancy-related hemodynamic changes. Indeed, pregnancy complicated with congenital heart disease (CHD) is associated with increased risk of adverse offspring outcomes, such as preterm birth, low birth weight and neonatal mortality. According to the recent reports, the predictors of offspring complications were NYHA functional class ≥ 3 , left heart obstruction, smoking, low oxygen saturation, cardiac medication, cyanotic heart disease at birth, mechanical valve prosthesis, cardiac complications during pregnancy, maternal decline in cardiac output during pregnancy and abnormal uteroplacental Doppler flow (UDF). UDF parameters in pregnant women with CHD were worse than those in healthy pregnant women. However, there was no strong event-specific predictor of fetal and neonatal complications. 2 Recurrence risk to offspring in women with CHD is also significantly higher than those without CHD.

Therefore, the preconception counseling in women with CHD is required, taking into account not only maternal cardiovascular complications but also offspring complications. It's important to the close monitoring of the fetal growth and the screening for fetal CHD.

S4-3

ACHD患者の分娩管理

The management of delivery in patients with ACHD

衛藤 英理子, 増山 寿

岡山大学大学院医歯薬学総合研究科 産科・婦人科学教室

Eriko Eto, Hisashi Masuyama

Department of Obstetrics & Gynecology, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Sciences

Along with medical advance, the adult congenital heart disease (ACHD) population is also growing. Similarly, the number of women with ACHD who reach child-bearing age has also increased. In pregnant women with cardiovascular complications, the most dynamic circulatory changes occur during labor, and careful management is necessary because of the risk to life. Vaginal delivery is generally recommended, but in some exceptional cases, caesarean section is chosen. As an obstetric treatment, vacuum extraction may be performed in order to shorten the second stage of labor. Administering epidural anesthesia during labor is useful for reducing the load in order to reduce cardiac output. We will present a case of pregnancy after Fontan surgery, in which epidural anesthesia was used during labor. We will also show the data in our hospital and summarize the literature findings and make current recommendations on managing delivery in patients with ACHD.

S4-4

ACHDの妊娠が自然予後に及ぼす影響

The effect of pregnancy with ACHD on the natural course of the disease

桂木 真司¹⁾, 丹羽 公一郎²⁾, 池田 智明³⁾

1) 榊原記念病院 産婦人科, 2) 聖路加国際病院 循環器内科, 3) 三重大学 産婦人科

Shinji Katsuragi¹⁾, Koichiro Niwa²⁾, Tomoaki Ikeda³⁾

1) Department of Obstetrics and Gynecology, Sakakibara Heart Institute, 2) Department of Cardiology, St.Luke's International Hospital,

3) Department of Obstetrics and Gynecology, Mie University

Pregnancy is a state of hyperstimulation of para-sympathetic node and related with hypertension, and tachycardia. And these hyperdynamic and hypervolemic changes by pregnancy may cause cardiac failure, cyanosis, and arrhythmia. We present three cases of ACHD complicated pregnancies. (1) In 26 y.o. congenital ASD women, IPAHA occurred suddenly at 26 weeks of gestation and tadalafil 40mg and Epoprostenol 40 mg was introduced during pregnancy. She delivered at 37 weeks by elective cesarean section. She is in NYHA class III after three years. (2) In 34 y.o. women with previous twice mitral valve repair, infectious endocarditis caused acute mitral valve dysfunction and cardiac failure, and the patient had mitral valve repair with fetus in the uterus at 10 weeks of gestation. She delivered via vagina at 39 weeks. (3) In 38 y.o. women with congenital severe AS, severe preeclampsia occurred at 36 weeks, and the patient dropped to NYHA class IV with cardiac failure with peripartum cardiomyopathy and the patient died in 6 months.

The prognosis of pregnancy with adult congenital heart diseases is well written in previous paper (ZAHARA Score 2010, CARPREG II 2018, modified WHO class, 2017). However complication with idiopathic pulmonary hypertension, infectious endocarditis, and preeclampsia dramatically changes the natural course of the disease.

S5-1

欧州ワーキンググループ 先天性心疾患に対する不整脈治療指針 2018

Opening Remarks.: A position paper of arrhythmias in congenital heart disease from EHRA, AEPC and ESC Working Group 2018.

立野 滋

千葉県循環器病センター 成人先天性心疾患診療部

Shigeru Tateno

Department of Adult Congenital Heart Disease, Chiba Cerebral and Cardiovascular Center

Arrhythmias are cause of morbidity, impaired quality of life, and mortality in adults with congenital heart disease (CHD). Optimal treatment strategies for arrhythmias are necessary to updated frequently due to growing experience and introduction of new techniques. Arrhythmias in congenital heart disease: a position paper of the EHRA, AEPC, ESC Working Group on Grown-up Congenital heart disease was published in 2018 that updated PACES/HRS Expert Consensus Statement on the Recognition and Management of Arrhythmias in Adult Congenital Heart Disease published in 2014.

Catheter ablation became to play more important role due to improving efficacy introduced by new 3D mapping system and irrigation catheter. On the other hand, long term amiodarone therapy is not advised in young CHD patients due to side frequent effects and may require discontinuation. One of other major revision is lead extraction that must be needed in young patients with CHD. Indication for lead extraction is expanded for noninfectious lead according to 2017 HRS expert consensus statement.

Further developments in technology for diagnosis and treatment for arrhythmia may improve morbidity and mortality of patients with CHD.

S5-2

成人先天性心疾患に対するマグネティックナビゲーションシステムの有用性

Usefulness of a Magnetic Navigation System for Patients with Adult Congenital Heart Disease

岡嶋 克則¹⁾, 中西 智之¹⁾, 市堀 博俊¹⁾, 米田 幸代¹⁾, 藤井 寛之¹⁾, 市川 靖士¹⁾, 辻本 誠長¹⁾, 園田 祐介¹⁾, 三和 圭介¹⁾, 下浦 広之¹⁾, 寺尾 侑也¹⁾, 金子 明弘¹⁾, 中岡 創¹⁾, 嘉悦 泰博¹⁾, 中村 浩彰¹⁾, 白木 里織¹⁾, 白井 丈晶¹⁾, 角谷 誠¹⁾, 圓尾 文子²⁾, 大西 祥男¹⁾

1) 加古川中央市民病院 循環器内科, 2) 加古川中央市民病院 心臓血管外科

Katsunori Okajima¹⁾, Tomoyuki Nakanishi¹⁾, Hirotochi Ichibori¹⁾, Sachiyo Yoneda¹⁾, Hiroyuki Fujii¹⁾, Yasushi Ichikawa¹⁾, Masanaga Tsujimoto¹⁾, Yusuke Sonoda¹⁾, Keisuke Miwa¹⁾, Hiroyuki Shimoura¹⁾, Yuya Terao¹⁾, Akihiro Kaneko¹⁾, Hajime Nakaoka¹⁾, Yasuhiro Kaetsu¹⁾, Hiroaki Nakamura¹⁾, Rio Shiraki¹⁾, Takeaki Shirai¹⁾, Makoto Kadotani¹⁾, Ayako Maruo²⁾, Yoshio Onishi¹⁾

1) Department of Cardiology, Kakogawa Central City Hospital.

2) Department of Cardiovascular Surgery, Kakogawa Central City Hospital

Background: Catheter ablation (CA) using a remote magnetic navigation system (MNS) may provide maximum benefit in patients with complex anatomy.

Objective: The aim of this study was to evaluate the clinical outcomes of CA using an MNS for congenital heart disease (ACHD) patients.

Methods: Consecutive six patients included TOF (n=2), TGA (n=1), AVSD (n=1), TAPVR (n=1), and inferior vena cava (IVC) interruption (n=1), [43 (18-66) years, two males] underwent CA using an MNS and three-dimensional electroanatomic mapping system were studied. Procedure outcomes were evaluated during a follow-up period of 15 (12-26) months.

Results: Paroxysmal atrial fibrillation in one patient with IVC interruption, and atrial tachycardias (AT) in the remaining five patients were targeted. Ablation catheter was introduced to targeted chambers (four right atrium, one left atrium, one pulmonary venous atrium) via azygous vein in one patient, retrograde aortic root in two patients and IVC in three patients. Acute CA success was obtained in all patients. Total procedure time was 205 (120-240) minutes and fluoroscopy time was 18 (4-33) minutes. Two of three patients with arrhythmia recurrence underwent 2nd CA procedure, and 1 patient was free from arrhythmic event. No complication occurred in this series.

Conclusion: CA using an MNS is considered to be safely performed with good clinical outcomes in ACHD patients.

S5-3**成人先天性心疾患患者におけるICD治療****ICD therapy in patients with ACHD**

西井 伸洋

岡山大学大学院医歯薬学総合研究科 先端循環器治療学講座

Nobuhiro Nishii

Department of Cardiovascular Therapeutics, Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences

In adult congenital heart disease (ACHD) patients, cardiovascular prognosis is a heart failure and a sudden death. Although ICD has been effective device to prevent sudden death, the indication of ICD in ACHD patients has not been shown. However, in 2014, Expert Consensus Statement on Management of Arrhythmias in ACHD was published, including secondary prevention and primary prevention. However, anatomy of ACHD patients is diverse, then, the implantation methods are also diverse. In this session, the various implantation methods would be focused on. Firstly, in the ACHD patients with four chamber anatomy, the implantation method of ICD is not different to the patients without ACHD. However, in patients with cardiac shunt, the risk of systemic emboli would raise, then, transvenous ICD implantation is thought to be a contraindication. Secondly, in patients without transvenous access, some methods are considered. One candidate is a subcutaneous ICD, especially in patients without necessity of pacing and anti-tachycardia pacing. To correctly use a S-ICD, screening ECG should be passed. ACHD patients are likely to have a bundle branch block and an inverted T wave, which sometimes fail for screening ECG. Another candidate for ICD implantation is open chest surgery. The ICD leads are fixed in various regions, such as through the transverse sinus, out of the pericardium, and in the thoracic cavity.

S5-4**成人先天性心疾患におけるリードマネジメント****Lead management for adult congenital heart disease**

庄田 守男

東京女子医科大学 循環器内科

Morio Shoda

Tokyo Women's Medical University Department of Cardiology

Adult patients with congenital heart disease are prone to bradyarrhythmias and life threatening tachyarrhythmias. Therefore, implantable cardiac pacing devices such as pacemaker, implantable cardioverter defibrillator and cardiac resynchronization therapy play an extremely important role. "Lead management" has been recognized as a key issue for better clinical outcomes because trouble-shooting of the implanted leads is much more difficult than that of the implanted device. Lead management should arise not at the timing of lead problems but at the implantation, at the device exchange and at the lead addition. Deep consideration should be paid to decide the pacing leads and the implantation site at the operation. Percutaneous lead extraction is an important option during the device exchange and the lead addition. New concepts of "lead management" will be discussed at this symposium.

S6-1

成人先天性心疾患に対するカテーテル治療のための画像診断 Cardiovascular imaging for the interventional cardiology in adult congenital heart disease

大山 伸雄^{1,2)}, Israel Valverde²⁾, Kuberan Pushparajah²⁾, 藤井 隆成¹⁾, 富田 英¹⁾

1) 昭和大学病院 小児循環器・成人先天性心疾患センター,

2) Department of Congenital Heart Disease, Evelina London Children's Hospital and St Thomas' Hospital, GSST NHS Foundation Trust, London, UK

Nobuo Oyama^{1,2)}, Israel Valverde²⁾, Kuberan Pushparajah²⁾, Takanari Fujii¹⁾, Hideshi Tomita¹⁾

1) Pediatric Heart Disease and Adult Congenital Heart Disease Center, Showa University Hospital, Tokyo, Japan,

2) Department of Congenital Heart Disease, Evelina London Children's Hospital and St Thomas' Hospital, GSST NHS Foundation Trust, London, UK

Cardiovascular imaging plays an important role in establishing the diagnosis, interventional management, follow-up after palliative or corrective surgery. There are various kinds of imaging modalities such as echocardiography, cardiac computed tomography (CCT), cardiac magnetic resonance (CMR) and fluoroscopic angiography. Three-dimensional (3D) images are useful, particularly giving information of intra- and extra-cardiac anatomy, coronary arteries, and vascular structures. 3D echocardiography (3DE) has priority for imaging of congenital heart disease with good spatial and temporal resolution. 3DE is particularly helpful for irregularly or asymmetrically shaped defects, as well as for complex lesions. Therefore, 3DE is recommended to assist interventional closure of selected ASDs and VSDs. CCT and CMR are non-invasive tools with high spatial resolution and powerful 3D reconstruction. 3D data sets are used for 3D printing nowadays, and 3D-models are useful in planning the catheterization intervention especially in cases with complex anatomy. 3D rotational angiography (3DRA) is a new and promising imaging technique in the cath lab. 3DRA fusion with live fluoroscopy are used as roadmap for coiling, stenting or percutaneous valve implantation. The aim of this talk is to give the latest topics of such cardiovascular imagings.

S6-2

日本における経皮的肺動脈弁置換術適応と考えられるデバイス毎の患者数の推定 Japanese single institutional survey of predicted frequency of candidates for transcatheter pulmonary valve implantation devices

藤本 一途¹⁾, 北野 正尚¹⁾, 坂口 平馬¹⁾, 大内 秀雄¹⁾, 津田 悦子¹⁾, 白石 公¹⁾, 黒崎 健一¹⁾, 中田 朋宏²⁾, 島田 勝利²⁾, 帆足 孝也²⁾, 市川 肇²⁾

1) 国立循環器病研究センター 小児循環器科, 2) 国立循環器病研究センター 小児心臓外科

Kazuto Fujimoto¹⁾, Masataka Kitano¹⁾, Hideo Ohuchi¹⁾, Etsuko Tshuda¹⁾, Isao Shiraiishi¹⁾, Kenichi Kurosaki¹⁾, Tomohiro Nakata²⁾, Masatoshi Shimada²⁾, Takaya Hoashi²⁾, Hajime Ichikawa²⁾

1) Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center, 2) Department of Pediatric Cardiovascular Surgery, National Cerebral and Cardiovascular Center

Background: To fit right ventricular outflow tract (RVOT) which has the anatomic heterogenicity, transcatheter pulmonary valve implantation (TPVI) devices are under development.

Objective: The aim of this study is to estimate the frequency of candidates for different type of TPVI devices associated with Tetralogy of Fallot (TOF) patients with RVOT dysfunction following native RVOT repair.

Method: Two hundred one TOF patients following RVOT repair who underwent angiography, weighed above 30Kg and excluded stenting for left pulmonary artery were investigated. Patients were divided to conduit repair group (Group I, 121patients) and native RVOT repair group (Group II, 80patients). In group I, we estimated the frequency of candidates for balloon-expandable device, Sapien3[®] (S). In group II, we investigated the frequency of candidates for self-expandable devices, Harmony[®] (H) and Venous P valve[®] (V). According to RVOT morphology obtained by angiography and surgical record (RVOT diameter, conduit diameter, bifurcation diameter, RVOT length and length of the stenosis), we studied the number of candidates for TPVI devices. In this study, we measured from maximum conduit diameter obtained by angiogram instead from sizing balloon dilation. In deployment of S without pre-stenting, we assumed length of the stenosis should be shorter than the device height. The morphological indication for TPVI (S, H, V) is conduit diameter ranged from 16 to 28mm, equal to or greater 22mm in systole and ranged from 14mm to 32mm, respectively. The regurgitation indication for H and V is regurgitation equal to or greater than moderate by echocardiography and RV end-diastolic volume index is greater than 150ml/m². The stenotic indication for S is stenosis with mean RVOT gradient equal to or greater than 35mmHg, and/or the ratio of right to left ventricular systolic pressure equal to or greater 0.7.

Results: Median diameters of RVOT, length of stenotic portion in group I and systolic pulmonary annulus valve diameter in group II in anteroposterior and lateral views were 16.6 × 18.3mm, 26.1 × 25.5mm, 24.4 × 21.5mm, respectively. In group I, The indication of TPVI (S) without pre-stenting was stenosis in 2 (2%), regurgitation in 16 (13%), and both in 18 (15%), respectively. The number of potential candidates for TPVI (S) with pre-stenting, which included length of stenotic portion less than 16mm was 81 (67%). In group II, three (3%), fifty (63%) patients were eligible for TPVI (H, V), respectively.

Conclusion: In Sapien3 without pre-stenting, the number of TOF patients eligible for TPVI following conduit repair was small. However, the number of patients eligible for Sapien3 with pre-stenting following conduit repair and Venus P valve following native RVOT repair might be large.

S6-3

ファロー四徴症の右室流出路不全に対する経カテーテル治療

Transcatheter pulmonary valve replacement in patients with tetralogy of Fallot

佐地 真育¹⁾, 高山 守正¹⁾, 高見澤 格¹⁾, 桃原 哲也¹⁾, 小林 匠²⁾, 吉敷 香菜子²⁾, 上田 知実²⁾, 矢崎 諭²⁾, 嘉川 忠博²⁾, 高梨 秀一郎³⁾, 和田 直樹⁴⁾, 安藤 誠⁴⁾, 高橋 幸宏⁴⁾, 磯部 光章¹⁾

1) 榊原記念病院 循環器内科, 2) 榊原記念病院 小児科, 3) 榊原記念病院 心臓血管外科, 4) 榊原記念病院 小児心臓外科

Mike Saji¹⁾, Morimasa Takayama¹⁾, Itaru Takamisawa¹⁾, Tetsuya Tobaru¹⁾, Takumi Kobayashi²⁾, Kanako Kishiki²⁾, Tomomi Ueda²⁾, Satoshi Yazaki²⁾, Tadahiro Yoshikawa²⁾, Shuichiro Takanashi³⁾, Naoki Wada⁴⁾, Makoto Ando⁴⁾, Yukihiro Takahashi⁴⁾, Mitsuaki Isobe¹⁾

1) Department of Cardiology, Sakakibara Heart Institute, 2) Department of Pediatrics, Sakakibara Heart Institute,

3) Department of Cardiovascular surgery, Sakakibara Heart Institute,

4) Department of Pediatric cardiovascular surgery, Sakakibara Heart Institute

Due to massive effort to improve outcomes in patients with congenital heart disease (CHD) by the multidisciplinary heart team approach consisting of pediatrician and pediatric cardiovascular surgeons, overall outcomes in patients with CHD has dramatically improved in last decades. CHD has been diagnosed in 12 thousand patients per year in Japan, and, surprisingly, 95% of them has become to be able to turn 20 years now. In the United States, 40 thousands CHD has been diagnosed every year, and 15% of them has been recognized to have Tetralogy of Fallot (TOF) with pulmonary stenosis. They often encounter cardiovascular events such as heart failure, or ventricular tachycardia 10 to 20 years after the surgery due to failed transannular patch, recognized as a gold standard surgical method to treat pulmonary stenosis in right ventricular outflow tract in patients with TOF. Therefore, routine follow-up using transthoracic echocardiography and cardiac magnetic resonance imaging are required as well as routine check-up for heart failure symptom. However, even with these imaging tests, it is still difficult to completely predict outcomes after their 2nd surgery, because right heart ventricular viability/remodeling is complicated, and careful attention is required in this population.

Transcatheter pulmonary valve replacement (TPVR) is the less invasive option without using cardiopulmonary bypass, and is anticipated to be an alternative way to surgical treatment in patients with previous sternotomies. TPVR using the Harmony valve is one of the investigational devices in an international multicenter prospective trials. TPVR will definitely change the treatment strategy in patients with TOF including timing of surgical interventions in a near future.

S6-4

卵円孔開存に対するカテーテル治療

Transcatheter Closure of Patent Foramen Ovale

高谷 陽一, 赤木 禎治, 中川 晃志, 中山 理絵, 三木 崇史, 伊藤 浩
岡山大学 循環器内科

Yoichi Takaya, Teiji Akagi, Koji Nakagawa, Rie Nakayama, Takashi Miki, Hiroshi Ito

Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine, Dentistry and Pharmaceutical Science

Patent foramen ovale (PFO) is linked with various diseases, including cryptogenic stroke and migraine. Especially, the relationship between PFO and cryptogenic stroke has become the focus of increasing interest, because recent randomized trials have demonstrated that transcatheter PFO closure is beneficial to reduce the recurrence of stroke compared with medical therapy. In Japan, transcatheter PFO closure will be introduced for secondary prevention of cryptogenic stroke. For the preventive therapy, it must be performed without any complications. In addition, accurately PFO diagnosis is essential. Transesophageal echocardiography remains the standard reference, but it is uncomfortable. Transthoracic echocardiography can be valuable for PFO screening due to noninvasive and easy availability. Furthermore, as transcatheter closure would be expanded, it is important to identify the high-risk PFO which is more likely to associate with cryptogenic stroke. We present about transcatheter PFO closure, including PFO diagnosis on echocardiography.

S7-1

ファロー四徴症を含む右室流出路再建

帆足 孝也

国立循環器病研究センター病院 小児心臓外科

Takaya Hoashi

Department of pediatric cardiovascular surgery, National Cerebral and Cardiovascular Center

S7-2

完全大血管転位術後の長期成績と問題点

Long-term Outcome and Complications after Surgical Repair of Complete Transposition of the Great Arteries

小谷 恭弘, 黒子 洋介, 立石 篤史, 笠原 真悟

岡山大学大学院医歯薬学総合研究科 心臓血管外科学講座

Yasuhiro Kotani, Yosuke Kuroko, Atsushi Tateishi, Shingo Kasahara

Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences

History of surgical treatment for complete transposition of the great arteries (TGA) always attracts the congenital heart surgeons as its drama, yet we are facing unprecedented issues after experience over the last half century. Atrial switch operation, such as Senning and Mustard operation was introduced in 1950s and had been performed as the first choice of surgery for TGA during that period. Arterial switch operation introduced by Jatene in 1975 was a game changer in the surgical treatment of TGA and remains the choice of surgery until today. Rastelli operation is performed for patients with TGA and pulmonary stenosis.

In arterial switch operation, atrial and ventricular function is usually preserved, but surgically-related problem, such as coronary ischemia, aortic valve regurgitation, and pulmonary stenosis can be a problem. Following atrial switch operation, systemic right ventricular dysfunction, systemic tricuspid regurgitation, atrial arrhythmia, and systemic/pulmonary venous obstruction are raised as issues. Right ventricle-to-pulmonary artery conduit stenosis, left ventricular outflow tract obstruction, and a residual ventricular septal defect are seen after Rastelli operation.

Surgery to address above issues can be an extremely challenge due to anatomic and physiologic uniqueness of post-surgical condition. Long-term survival and complications as well as surgical management for late issues are discussed.

S7-3

総肺静脈還流異常に対する外科治療の長期成績

Long-term results of surgical correction for total anomalous pulmonary venous connection

松久 弘典¹⁾, 大嶋 義博¹⁾, 日隈 智慧¹⁾, 岩城 隆馬¹⁾, 村上 優¹⁾, 城戸 佐知子²⁾, 田中 敏克²⁾

1) 兵庫県立こども病院 心臓血管外科, 2) 兵庫県立こども病院 循環器科

Hironori Matsuhisa¹⁾, Yoshihiro Oshima¹⁾, Tomonori Higuma¹⁾, Ryuma Iwaki¹⁾, Yu Murakami¹⁾, Sachiko Kido²⁾, Toshikatsu Tanaka²⁾

1) Department of cardiovascular surgery, Kobe Children's Hospital, 2) Department of cardiology, Kobe Children's Hospital

Introduction: Surgical repair of total anomalous pulmonary venous connection (TAPVC) is associated with significant mortality and morbidity. Especially, postoperative pulmonary venous stenosis (PVS) persists as a major determinant of long-term outcome.

Methods: One hundred eleven patients with TAPVC from 1996 to 2018 were included (BV: 75, SV: 36). Our current surgical strategy (preoperative CT and selective application of primary sutureless repair) was established around 2010. Thus, study period was divided into 2 categories: era 1 (1996-2010), and era 2 (2011-2018).

Results: For patients with BV, 5-year survival was 69% in era 1, and 97% in era 2 ($P = 0.003$). For patients with SV, 5-year survival was 21% in era 1, and 76% in era 2 ($P = 0.0005$). Postoperative PVS was present in 25 patients (era 1: 12, era 2: 13). Since 2011, we started scheduled multiple reintervention for postoperative PVS. Three-year survival after PVS was 33% in era 1, and 100% in era 2 ($P = 0.0008$).

Conclusions: CT based surgical strategy for TAPVC provided significant survival benefit. Multiple reintervention also appears to confer a significant survival benefit to patients with postoperative PVS.

S7-4

肺動脈閉鎖兼正常心室中隔のFontan成績

Long-term outcomes of Fontan survivors for pulmonary atresia with intact ventricular septum

小田 晋一郎¹⁾, 中野 俊秀¹⁾, 石川 友一²⁾, 倉岡 彩子²⁾, 杉谷 雄一郎²⁾, 角 秀秋¹⁾

1) 福岡市立こども病院 心臓血管外科, 2) 福岡市立こども病院 循環器科

Shinichiro Oda¹⁾, Toshihide Nakano¹⁾, Yuichi Ishikawa²⁾, Ayako Kuraoka²⁾, Yuichiro Sugitani²⁾, Hideaki Kado¹⁾

Department of Cardiovascular Surgery¹⁾ and Cardiology²⁾, Fukuoka Children's Hospital, Fukuoka, Japan

Objectives: Previous studies have reported a risk of death after Fontan for patients with pulmonary atresia with intact ventricular septum (PA-IVS) associated with right ventricle-dependent coronary circulation (RVDCC). We reviewed our experience of these patients after Fontan.

Methods: Between 1981 and 2017, 127 patients with PA-IVS underwent single-ventricle palliation (n=89), biventricular repair (n=29), one and a half repair (n=7), and unrepaired (n=2). Sixty-eight patients had extracardiac conduit Fontan procedure. The catheterization measurements and clinical data were reviewed.

Results: No death occurred after Fontan completion. Thirteen patients (20%) had RVDCC. Those with RVDCC had a higher Fontan pressure (10.4 ± 2.0 vs 9.0 ± 1.8 mmHg; $P=0.015$). There was no difference in LVEDP, LVEDV, LVEF, cardiac output, and SaO₂ between RVDCC and non-RVDCC. Fifteen (17%) death occurred before Fontan procedure. The 20-years age survival was 75% (RVDCC) vs 89% (non-RVDCC) ($P=0.077$). All patients were on cardiovascular medications (ACE inhibitors: 91%, Beta-blockers: 29%, Warfarin: 98%, Aspirin: 91%).

Conclusions: Long-term survival was excellent after Fontan completion. Aggressive medical treatment with cardioprotective agents and anticoagulants may be effective for preventing myocardial ischemia and death. Active surveillance for inter-stage mortality may be necessary.

S8-1

循環器疾患における緩和ケア Palliative care for heart disease

水野 篤, 福田 旭伸, 丹羽 公一郎
聖路加国際病院 循環器内科

Atsushi Mizuno, Terunobu Fukuda, Koichiro Niwa
Department of Cardiology, St.Luke's International Hospital

Palliative care is essentially needed in all patients with life-threatening disease. Unfortunately, palliative care is not adequately supplied to many patients with heart disease comparing those with cancer. Furthermore, for many non-palliative care specialist, palliative care is too vague to deliver in clinical practice. Therefore, we have several studies about the quality of care about palliative care in heart disease. We developed several quality indicators for palliative care and performed bereaved family survey, which might imply some important clinical implications for many clinical practitioners.

Considering the rapid increasing number of adult congenital heart disease patients, we should focus not only the treatment for long-term survival but also the palliative care for many worse prognosis patients.

S8-2

ACHD 患者に対する緩和ケアマインドを持った日常診療 Daily Medical Practice with Palliative Care Mind for Adults with Congenital Heart Disease

中澤 誠
総合南東北病院 小児・生涯心臓疾患研究所

Makoto Nakazawa
Southern Tohoku General Hospital, Pediatric and Lifelong Congenital Cardiology Institute

Palliative care is, in general, a practice for patients at terminal or near-terminal stages, but should be differently considered in adult congenital heart disease (ACHD) patients because they are always not free from the fear of possibility of own premature death after when they realize, at some time during growth, that their heart problems could be life threatening. The fear becomes a real issue at every time when any cardiac problems such as arrhythmia or heart failure comes out or when their friends with similar conditions become worse or died. These emotional movements are in sharp contrast to acquired diseases such as cancer. In general, when people realize own death, they feel several issues of dreadful fear and emotional movement, which including fear for pain, fear for loneliness, fear of leaving the family, uneasiness going into never experienced area, regret for ending the life before accomplishment, fear of own disappearance from the world. Many of them are common also in ACHD patients, who however have especially strong emotions of anxiety of leaving the family and of regret of premature death, both of which they anticipated to face.

In daily practice of ACHD patients, they often ask us about their own future, as one of my patients directly asked me how long further she may survive. It is not a question simply about the life span but also about how they may spend the rest of life. Obviously we do not have an right answer to such a question, but what we are able to do is to imagine their fear and anxiety as listed above, and to express our mind of sympathy "Kokoro wo yoseru" (to place heart along with patient). To do so, we need to understand the death in an aspect of thanatology.

Thanatology is defined as to learn broad aspects of the death and as to consider how to live, i.e. the way of life as a human, and the practice of thanatology is the support and help to people/patient who is living with death and dying and to people or family who are left behind the death. This is exactly the practice of palliative care.

In my talk in this session, I will touch first on the general aspect of thanatology and secondly on issues related to ACHD patients, that is only from my own experience but not from the literatures since almost no study was published. As described above, ACHD patients have always the fear and anxiety in their mind, nonetheless they will not express directly their mind to us and, instead, they imply their fear with indirect words or questions. We do not necessarily have to reply to each of them, but we have to understand the patients' deep thought, and we have to support what they want as much as we are able to do even though the support may possibly shorten their life span.

S8-3

緩和医療と心肺移植

Palliative Medicine in heart/lung transplant candidates or recipients

福島 教偉

国立循環器病研究センター 移植医療部

Norihide Fukushima

Department of Transplant Medicine, National Cerebral and cardiovascular Center

Heart and lung transplantation (HLT_x) is an established therapeutic option for many patients with combined advanced therapy-resistant cardiac and pulmonary diseases and a limited long-term prognosis. Patients with end-stage heart and lung disease awaiting transplantation may experience a complex constellation of physical, psychosocial, and emotional symptoms, including dyspnea, cough, anxiety, depression, and insomnia. Given the disease burden faced by these patients, involvement of a palliative care team would seem appropriate even in many other developed countries. But in Japan, only 76 brain dead organ donation were performed in 2017 and only three HLT_x were done for one patient with complex congenital heart anomaly with severe pulmonary hypertension (PH) and two patients with restrictive cardiomyopathy with severe PH in Osaka University since January 2009. Due to extremely severe organ shortage. Nearly half of HLT_x candidates died during waiting for HLT_x. Most of possible candidates for HLT_x died without listing for HLT_x at Japan Organ Transplant Network. Therefore, it may be more important for HLT_x candidates or recipients to establish palliative care system.

In this talk, current status and future aspects of HLT_x in Japan will be introduced and palliative care for these patients will be discussed.

S8-4

緩和ケアにおける看護師の役割

The Role of Nurse for Palliative Care in Adult Congenital Heart Disease

河野 由枝¹⁾, 高田 弥寿子¹⁾, 庵地 雄太²⁾, 濱谷 康弘²⁾

1) 国立循環器病研究センター 看護部, 2) 国立循環器病研究センター 心臓血管内科 心不全科

Yukie Kawano¹⁾, Yasuko Takada¹⁾, Yuta Anchi²⁾, Yasuhiro Hamatani²⁾

1) Department of Nursing, National Cerebral and Cardiovascular Center.

2) Division of Heart Failure, Department of Cardiovascular Medicine, National Cerebral and Cardiovascular Center

The nurse have 4 roles in palliative care as follows; 1. Symptom management, 2. Decision support that respects values and wishes of the patient and family members, 3. Family care, 4. Coordinate among the medical team. In the field of adult congenital heart disease, transitional support gradually enhances the patients' autonomy. Whereas, decision making and support should be performed based on the patients' comprehension of their disease and the patients' dependence on their parents. At the end-of-life and terminal stage, we sometimes have difficulty in managing the patients' symptom because of complicated hemodynamics of adult congenital heart diseases. Thus, multidisciplinary-term approach is crucial for palliative care in patients with adult congenital heart disease. In this session, we would like to talk about the role of nurse for palliative care in adult congenital heart disease.

ES-1

高尿酸血症への対応は必要か

Does it need to take care of hyperuricemia?

桑原 政成¹⁾、丹羽 公一郎²⁾、久留 一郎³⁾

1) 虎の門病院 集中治療科・循環器センター内科, 2) 聖路加国際病院 心血管センター,
3) 鳥取大学大学院 機能再生医科学専攻再生医療学部門

Masanari Kuwabara¹⁾, Koichiro Niwa²⁾, Ichiro Hisatome³⁾

1) Intensive Care Unit and Department of Cardiology, Toranomon Hospital, 2) Cardiovascular center, St. Luke's International Hospital,
3) Division of Regenerative Medicine and Therapeutics, Tottori University Graduate School of Medical Sciences

Hyperuricemia is often accompanied by obesity, metabolic syndrome, hypertension, diabetes mellitus, dyslipidemia, chronic renal disease, heart failure, cardiovascular disease and adult congenital heart disease (ACHD). Some basic and clinical studies showed the causality between uric acid and hypertension or chronic kidney disease, but we cannot conclude whether uric acid is an independent risk factor for heart failure or cardiovascular disease.

The serum uric acid level is known to vary significantly depending on meals, lifestyle, gender, and previous use of medicine including diuretics. Based on these facts, it is believed that the uric acid level partly reflects the lifestyle origins of the disease, and it merely serves as a marker of cardiovascular disease. Furthermore, since female hormones lower the serum uric acid levels, they tend to increase after menopause, and the evaluation of uric acid becomes more difficult. The number of confounding factors involved in evaluating serum uric acid levels complicates the sole analysis of uric acid. Indeed, there are a few intervention studies that focused only on uric acid.

Recently, some intervention studies were reported. EXACT-HF trial showed that allopurinol, a xanthine oxidase inhibitor, was not able to improve clinical status, exercise capacity, quality of life, or left ventricular ejection fraction at 24 weeks in high-risk heart failure patients with reduced ejection fraction. FEATHER study showed that feboxostat, another xanthine oxidase inhibitor, was not able to improve kidney function compared to placebo, even though feboxostat prevented gout attack. In contrast, FREED study showed that feboxostat prevented the development of kidney disease compared to control, but there was no difference on cardiovascular disease, heart failure, atrial fibrillation, and cardiovascular mortality between the two groups. The evidence between uric acid and heart failure or cardiovascular disease is still controversial. In this session, I will introduce recent evidence about uric acid.

ES-2

ACHD患者に対する心臓リハビリテーション

Why and How? Cardiac Rehabilitation for ACHD Patients

肥後 太基¹⁾、坂本 一郎¹⁾、石北 綾子¹⁾、永富 将太²⁾、樋口 妙²⁾、筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 リハビリテーション部

Taiki Higo¹⁾, Ichiro Sakamoto¹⁾, Ayako Ishikita¹⁾, Syouta Nagatomi²⁾, Tae Higuchi²⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Medicine, Kyushu University Hospital, 2) Department of Rehabilitation, Kyushu University Hospital

It is well-known that the exercise capacity in ACHD patients is lower than healthy controls. The reason of this impairment of exercise capacity seems to be multifactorial; relatively sedative life style, impairment of chronotropic response, desaturation of arterial blood gas, and so on. Although there has not been established evidence of the beneficial effects of regular exercise training on survival rate in ACHD patients, the significance of keeping or improving exercise capacity are obvious. Cardiac rehabilitation has been developed not only in the aim of improving prognosis, but in that of improving exercise capacity and quality of life in patients with various heart disease. Furthermore, supervised cardiac rehabilitation may have more favorable effect on psychological conditions of patients and their care-givers. What is matter is not "to do or not to do exercise", but "how to do exercise". Exercise prescription based on the results of cardio-pulmonary exercise test seems to be useful to decide the intensity of exercise training for ACHD patients.

ES-3

高齢者ファローのいつまで手術するか、外科医の立場から
Surgical repair in elderly patient with Tetralogy of Fallot

笠原 真悟

岡山大学大学院医歯薬学総合研究科 心臓血管外科

Shingo Kasahara

Department of Cardiovascular surgery, Okayama University

In recent years there has been a considerable increase in adult patients with congenital heart disease as a result of the success made in cardiac surgery during the 30 to 40 years. Among these patients, tetralogy of Fallot (TOF) is a frequent diagnosis because technical progress and improved postoperative management have led to improved results after complete repair in infancy. However, there are still a certain number of order patients with uncorrected TOF. These patients survive until adulthood because of favorable morphologic condition such as well-balanced systemic/pulmonary perfusion. Chronic hypoxemia stimulates the development of large systemic-to-pulmonary arterial collaterals. These collaterals or preexisting shunts, or both, lead to increased pulmonary artery blood flow, and together with myocardial hypertrophy and myocardial hypoxia, they lead to reduced biventricular function. Impaired ventricular function, severe hypoxemia, and tricuspid regurgitation are associated with a higher operative mortality, whereas the influence of age at the time of operation on operative mortality is controversially discussed.

We reviewed our experience to assess risk factors for operative morbidity and to determine the benefit of total correction of TOF in adolescent and adult patients according the functional status at follow up.

ES-4

妊娠出産 ハイリスク症例への対応
The clinical management of high-risk pregnancy

神谷 千津子

国立循環器病研究センター 周産期・婦人科

Chizuko A. Kamiya

Department of Perinatology and Gynecology, National Cerebral and Cardiovascular Center

Recent advanced clinical practice concerning congenital heart disease (CHD) has enabled most women with CHD to reach childbearing age, preserving normal daily life. Therefore, the number of pregnancies with CHD is increasing. The various advanced operative methods, including Fontan operation, are widely spread and the range and severity of CHD in pregnant women are expanding. The clinical management of pregnancies with CHD requires comprehensive knowledge and experience.

Among women with Fontan circulation, pregnancy and delivery are possible in a case with NYHA class I or II and favorable cardiac function retaining sinus rhythm, but the miscarriage rate is high up to 40-50%. There is no consensus on use of anticoagulant and antiplatelet therapies during pregnancy, and further accumulation of cases is required to evaluate the use of this therapy. Maternal cyanosis effect on fetal outcomes. Ability Index, hemoglobin, and arterial oxygen saturation before the pregnancy were factors related to live birth rates. Women with mechanical valve should face to high-risk of valve thrombosis in pregnancy or fetal poor outcomes.

A multidisciplinary team, which is composed of trained obstetricians, adult and pediatric cardiologists, anesthesiologists, midwives and other specialists involved, is necessary for the management of the women with high pregnancy risks.

JS2-1

先天性心疾患における房室弁逆流の定量評価

Assessment of atrioventricular valve regurgitation in CHD

新居 正基

静岡県立こども病院 循環器科

Masaki Nii

Department of Cardiology, Mt Fuji Shizuoka Children's Hospital

The severity of atrioventricular valve (AVV) regurgitation is assessed in two aspects. One is quantitative assessment of regurgitation volume, and the other is the assessment of functional reserve of the ventricle. The latter is evaluated based on an end-diastolic/systolic ventricular volume or myocardial performance analyzed by tissue Doppler or strain. Echocardiography has been one of the main modalities for the assessment of functional reserve of the ventricle, which is important to determine the timing of intervention on AVV, or to predict the prognosis. On the other hand, echocardiography has limitations to assess AVV regurgitation quantitatively, therefore it requires multiple semiquantitative indices to assess AVV regurgitation. In this presentation, conventional and new quantitative assessments of AVV regurgitation are discussed.

JS2-2

狭窄病変の定量における問題点と意義。

Evaluation of stenotic lesions in the patient with ACHD

山澤 弘州¹⁾, 武田 充人¹⁾, 泉 岳¹⁾, 佐々木 理¹⁾, 谷口 宏太¹⁾, 岩野 弘幸²⁾, 石森 直樹²⁾, 加藤 伸康³⁾

1) 北海道大学大学院医学研究院 小児科, 2) 北海道大学大学院医学研究院 循環器内科, 3) 北海道大学大学院医学研究院 循環器外科

Hirokuni Yamazawa¹⁾, Atsuhito Takeda¹⁾, Gaku Izumi¹⁾, Osamu Sasaki¹⁾, Kouta Taniguchi¹⁾, Hiroyuki Iwano²⁾, Naoki Ishimori²⁾, Nobuyasu Katou³⁾

1) Department of Pediatrics, Faculty of Medicine and Graduate School of Medicine, Hokkaido University,

2) Department of Cardiology, Faculty of Medicine and Graduate School of Medicine, Hokkaido University,

3) Department of Cardiovascular surgery, Faculty of Medicine and Graduate School of Medicine, Hokkaido University

Accurate evaluation of a burden on upstream regions of the stenotic site, such as ventricle, atrium, and lung, are important for quantification of stenotic lesion. According to the Japanese guideline, using echocardiography and cardiac catheterization are the main method for those quantification. It is identified that there are theoretical pitfalls in those method. The value of the pressure gradient derived from cardiac catheterization is often used to evaluate the valve stenosis. However, it is known that this value does not accurately reflect hemodynamic abnormality. Furthermore, echocardiography does not accurately represent the load of the ventricle due to difference in shape of stenosis, such as supravalvular stenosis, and double-chambered right ventricle. In addition to those theoretical pitfalls, technical limitation can also affect quantification, especially in the cases of adults because it is difficult to properly adjust the incident angle of the echocardiographic beam to the measurement site due to anatomical difference. It is important to integrate those quantification, symptoms, physical examination and the result of other tests, such as electrocardiograms, to properly evaluate stenotic lesion.

JS2-3

右室同期不全の定量化

Quantification of right ventricular dyssynchrony

石津 智子¹⁾, 瀬尾 由広²⁾, 山田 優²⁾, 中澤 直美¹⁾, 川松 直人²⁾, 町野 智子²⁾, 堀米 仁志³⁾

1) 筑波大学 臨床検査医学, 2) 筑波大学 循環器内科, 3) 筑波大学 小児科

Tomoko Ishizu¹⁾, Yoshihiro Seo²⁾, Yu Yamada²⁾, Naomi Nakazawa¹⁾, Naoto Kawamatsu²⁾,

Tomoko Machino-Ohtsuka²⁾, Hitoshi Horigome³⁾

1) Department of Clinical Laboratory Medicine, University of Tsukuba, 2) Department of Cardiology, University of Tsukuba,

3) Department of Pediatrics, University of Tsukuba

Cardiac resynchronization therapy (CRT) is the effective non-pharmacological treatment in certain patients with left side heart failure accompanied left bundle branch block. Recently, the application of CRT to the right-side heart disease of congenital heart defect has been focused. Because of the heterogeneity of its anatomy and the history of surgical intervention, the detailed assessment for the dyssynchrony in each patient is warranted. Speckle tracking two-dimensional echocardiography derived strain-time curve in each segment can evaluate the desynchronized/discoordinated wall motion quantitatively. Furthermore, three-dimensional speckle tracking echocardiography is the new non-invasive application for right ventricle. Especially, isochrone activation mapping to show the way of mechanical activation propagation on the right ventricle may help to understand the dyssynchrony pattern visually and may give us the information for optima pacing site. In the present session, we will present the case with tetralogy Fallot with complete right bundle branch block, congenitally corrected transposition of great arteries, TGA treated with atrial switch operation, and right uni-ventricle with wide QRS duration. We will also review the recent literatures and discuss the future application of echocardiographic non-invasive dyssynchrony imaging.

JS2-4

周術期不整脈治療における病態評価 ～ multimodality の活用～

Pathological examination using multimodality in perioperative arrhythmic treatment

梶山 葉¹⁾, 糸井 利幸¹⁾, 瀧上 雅雄²⁾, 杉山 裕章²⁾, 中西 直彦²⁾, 中村 猛²⁾, 白石 裕一²⁾, 白山 武司²⁾, 的場 聖明²⁾, 板谷 慶一³⁾, 前田 吉宣³⁾, 山岸 正明³⁾, 夜久 均⁴⁾

1) 京都府立医科大学 小児科, 2) 京都府立医科大学 循環器内科, 3) 京都府立医科大学 小児心臓血管外科,

4) 京都府立医科大学 心臓血管外科

Yo Kajiyama¹⁾, Toshiyuki Itoi¹⁾, Masao Takigami²⁾, Hiroaki Sugiyama²⁾, Naohiko Nakanishi²⁾, Takeshi Nakamura²⁾,

Hirokazu Shiraishi²⁾, Takeshi Shirayama²⁾, Satoaki Matoba²⁾, Keiichi Itatani³⁾, Yoshinobu Maeda³⁾,

Masaaki Yamagishi³⁾, Hitoshi Yaku⁴⁾

1) Department of Pediatrics, Kyoto Prefectural University of Medicine, 2) Department of Cardiovascular Medicine,

3) Department of Pediatric Cardiovascular Surgery, 4) Department of Cardiovascular Surgery

We present strategies of perioperative arrhythmic treatments using multimodal pathological examination for adult patients with congenital heart disease. Grown-up patients with congenital heart disease may have symptomatic and/or asymptomatic arrhythmic events related to myocardial damages due to preceding surgical treatments and prolonged cardiac disturbance. Development of a surgical strategy hence requires multimodal estimates of arrhythmic substrate prior to surgical re-interventions. Recent technological advances in electrophysiology (EP), echocardiography, computed tomography (CT) and cardiovascular magnetic resonance (CMR) as well as their combined use allow for detailed cardio-pathological estimations. The combination of CMR with EP voltage mapping provides excellent myocardial imaging of viability and can be of great help in surgical decision making. For patients with a pacemaker treatment, however, CT and echocardiography are preferable to CMR imaging due to disturbances of electrical leads and generator. We should be careful in combining cardio-pathological tests based on specificities of individual patients.

JS2-5

体心室右室での定量化への挑戦とその有用性

Challenges in the echocardiographic assessment of systemic right ventricle

杜 徳尚, 赤木 禎治, 伊藤 浩
岡山大学 循環器内科

Norihisa Toh, Teiji Akagi, Hiroshi Ito
Department of Cardiovascular Medicine, Okayama University

Systemic right ventricle (RV) is a unique physiology in congenital heart disease. Mainly, 2 different types of congenital heart disease provide the unusual physiology: congenitally corrected transposition of the great arteries (ccTGA) and transposition of the great arteries (TGA) after arterial switch operation. Since RV function is a crucial determinant of long-term outcomes in patients with systemic RV, accurate quantification of systemic RV is mandatory. Although the current gold standard for noninvasive measurement of systemic RV function is cardiac magnetic resonance imaging (CMR) which provides comprehensive assessment of RV size and function, CMR has several limitations. Therefore, echocardiography plays a central role in the evaluation of systemic RV; however, echocardiographic quantification of systemic RV is still a tough challenge. In this session, we will summarize the clinical utility of echocardiography to assess systemic RV.

CD1

身体的・社会的フレイルを伴う部分肺静脈還流異常症合併高齢女性の一例

A case of elderly woman with partial anomalous pulmonary venous connection accompanied by physical and social frailty.

川松 直人¹⁾, 石津 智子²⁾, 中澤 直美²⁾, 山本 昌良²⁾, 町野 智子²⁾, 瀬尾 由広²⁾, 堀米 仁志³⁾, 平松 祐司⁴⁾, 家田 真樹²⁾

1) 水戸済生会総合病院 循環器内科, 2) 筑波大学 循環器内科, 3) 筑波大学 小児科, 4) 筑波大学 心臓血管外科

Naoto Kawamatsu¹⁾, Tomoko Ishizu²⁾, Naomi Nakazawa²⁾, Masayoshi Yamamoto²⁾, Tomoko Machino²⁾,

Yoshihiro Seo²⁾, Hitoshi Horigome³⁾, Yuji Hiramatsu⁴⁾, Masaki Ieda²⁾

1) Department of Cardiology, Mito Saiseikai General Hospital, 2) Department of Cardiology, Faculty of Medicine, University of Tsukuba,

3) Department of Child Health, Graduate School of Comprehensive Human Sciences, University of Tsukuba,

4) Department of Cardiovascular Surgery, University of Tsukuba

A 70-year-old woman who is 146 cm tall and weighs 33.5 kg (BSA 1.18m², BMI 15.7) presented with worsening shortness of breath. She lives with her 85-years-old husband and has no other relatives.

She has been prescribed diuretics for many years with edema, sense of easy fatigue, and dyspnea from her late 40s. Her first heart failure hospitalization was at the age of 64, and then she repeated exacerbations of heart failure.

At the age of 67, she was diagnosed as pulmonary hypertension because the estimated systolic right ventricular pressure was 61 mmHg in the transthoracic echocardiography, and beraprost and sildenafil were added.

In the follow-up echocardiography, atrial septal defect was pointed out, but pulmonary vasodilators and diuretics were continuously prescribed without further evaluation. Her heart failure symptoms gradually worsened and came to see us in the emergency outpatient frequently.

Her right heart catheterization to reevaluate her condition showed mild pulmonary hypertension with a mean pulmonary artery pressure of 27 mmHg and a pulmonary capillary wedge pressure of 8 mmHg, and Qp/Qs of 4:1.

As a result of transesophageal echocardiogram and chest CT, findings that two atrial septal defects and right upper and lower pulmonary veins were refluxed to the right atrium were confirmed.

I would like to receive your opinion for this case with physical and social frailty.

この症例をどうする？
皆で考えよう。

CD2-1

未修復不完全型房室中隔欠損に拘束型心筋症を合併した挙児希望女性の一例

A Case of a Woman with Unrepaired Partial Atrioventricular Septal Defect Complicated with Restrictive Cardiomyopathy Who Wishes for a Baby, Watchful Waiting or Surgery?

小永井 奈緒^{1,3)}, 福井 重文²⁾, 浅野 遼太郎^{2,3)}, 上田 仁²⁾, 辻 明宏²⁾, 大郷 剛²⁾

1) 国立循環器病研究センター 小児循環器科, 2) 国立循環器病研究センター 心臓血管内科,

3) 熊本大学医学教育部 循環器先進医療学分野

Nao Konagai^{1,3)}, Shigefumi Fukui²⁾, Ryotaro Asano^{2,3)}, Jin Ueda²⁾, Akihiro Tsuji²⁾, Takeshi Ogo²⁾

1) Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center,

2) Department of Cardiovascular Medicine, National Cerebral and Cardiovascular Center,

3) Department of Advanced Cardiovascular Medicine, Kumamoto University Graduate School of Medical Sciences

A 34-year-old woman, who was diagnosed as atrial septal defect (ASD) when she was one year old, was presented to our hospital for further examination of her symptoms of New York Heart Association functional class II and evaluation of feasibility of pregnancy. Echocardiography revealed primum ASD and mitral valve cleft, while mitral regurgitation was limited to slight degree. Plasma BNP level was 68.3 pg/dl. Catheterization indicated that Qp/Qs was 1.81 and right ventricle (RV) was mildly enlarged (end-diastolic volume index of 110 ml/m²), whereas left ventricular (LV) end-diastolic volume index was small (59 ml/m²). Pressure curve of both ventricles showed dip and plateau pattern with high RV end-diastolic pressure of 16 mmHg and that of LV of 15 mmHg, which resulted in mildly elevated mean pulmonary arterial pressure of 21 mmHg. Endomyocardial biopsy indicated progressive interstitial fibrosis, suggesting the possibility of restrictive cardiomyopathy. Refractory left heart failure could be considered after surgical repair of primum ASD because of increased LV volume load, especially in the setting of pregnancy. On the other hand, if unrepaired, it is difficult to predict interatrial shunting ratio considering the change in plasma volume and vascular resistance during pregnancy. In this session, I would like to discuss possible treatment options and the management of pregnancy in this case.

皆で考えよう。この症例をどうする？

CD2-2

門脈圧亢進症を合併した心内修復術後ファロー四徴症の進行性血行動態増悪に対する治療戦略を如何に考えるか？

What is a Way Out of the Progressive Hemodynamic Crisis in an Adult Case of Repaired Tetralogy of Fallot with Portal Hypertension?

藤田 鉄平¹⁾, 小坂橋 俊美¹⁾, 前川 恵美¹⁾, 郡山 恵子¹⁾, 宮本 隆司²⁾, 宮地 鑑²⁾, 阿古 潤哉¹⁾

1) 北里大学病院 循環器内科, 2) 北里大学 心臓血管外科

Teppey Fujita¹⁾, Toshimi Koitabashi¹⁾, Emi Maekawa¹⁾, Keiko Ryo Kooriyama¹⁾, Takashi Miyamoto²⁾, Kagami Miyazi²⁾, Jyunya Ako²⁾

1) Department of Cardiovascular Medicine, Kitasato University Hospital,

2) Department of Cardiovascular Surgery, Kitasato University Hospital

We present a case of a 21-year-old woman who underwent intracardiac repair for Tetralogy of Fallot (TOF) at the age of 11 months, tricuspid valve replacement (TVR) at 3 and 4 years old, and the third-time TVR (Carpentier-Edwards 23 mm) and pulmonary valve replacement (Carpentier-Edwards 19 mm) at 14 years old. She was consulted to the department of internal medicine as transition to the adult care when she was 19 years old. At that time she already had New York Heart Association functional Class II m heart failure symptom. A thorough examination demonstrated right ventricular (RV) dilatation (end-diastolic-volume 267 ml and end-systolic-volume 209 ml), RV dysfunction (ejection fraction 22%), pulmonary valve stenosis (maximum pressure gradient 40 mmHg, and the index of the RV to left ventricular systolic pressure 0.7), tricuspid stenosis (mean pressure gradient 9 mmHg), and significantly high right atrial pressure (27 mmHg). Moreover, serious systemic complications were revealed. The long-term chronic right heart failure caused liver dysfunction with ascites, splenomegaly, portosystemic shunt, and portal hypertension. In this high-risk case for surgical interventions, the treatment strategy should be carefully and thoroughly discussed.

皆で考えよう。
この症例をどうする？

D1

Fontanでの抗凝固療法：必要 vs. 不要
Anticoagulations for Fontan patients: Do you USE or NOT?

安田 謙二
 島根大学医学部 小児科
 Kenji Yasuda
 Department of Pediatrics, Shimane University Faculty of Medicine

血栓塞栓症は、心不全、不整脈、蛋白漏出性胃腸症、Fontan 関連肝疾患などと並び、Fontan 術後の重要な合併症のひとつです¹⁾。Fontan 術後患者では凝固因子、凝固線溶系、血小板機能の異常があり、凝固促進因子と抗凝固因子とのアンバランスが、血栓塞栓症の危険因子となる可能性について報告されています²⁾³⁾。一方咯血や脳出血といった易出血性に関連した合併症も散見され、長期の抗凝固療法が関連していると報告されています⁴⁾。こうした状況にも関わらず、現在のところ Fontan 術後患者に対する抗凝固療法について、コンセンサスの得られた標準的治療はなく、抗凝固薬投与の有無、投与期間（開始、中止、再開）、投与量（目標プロトロンビン時間）等に関しては、施設毎の判断に委ねられています。Fontan 術後患者での抗凝固療法は必要なのでしょうか、あるいは不要なのでしょうか？本セッションでは“A Never-Ending Debate?”⁵⁾とも言われるこのテーマを取り上げ、ハイボリュームセンターでご活躍のお二人の先生にご発表頂き、“A Resolved Issue”を目指し、深く議論したいと思えます。

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D2

TOF術後PRに対する成人期PVR：人工弁の種類・サイズを選択
PVR for repaired TOF in Adult: How to choose prosthesis and its size

河田 政明
 自治医科大学とちぎ子ども医療センター・成人先天性心疾患センター 小児・先天性心臓血管外科
 Masaaki Kawada
 Pediatric and Congenital Cardiovascular Surgery, Jichi Medical Center Tochigi, Jichi Adult Congenital Heart Center, Jichi Medical University

Fallot 四徴症 (TOF) 心内修復術後の続発症の中でも最も注目される肺動脈弁逆流 (PR) に対する肺動脈弁位人工弁置換術 (PVR) はすでに多くの症例で行われており、今後も増加が予想される。心臓 MRI (CMR) による右室容積が PVR の適応決定にしばしば用いられるが、PVR の際選択される人工弁については術前の臨床症状の乏しさに加え、左心系での人工弁置換術と異なる形態・血流動態などの特徴や、経験の少なさなどからいわゆるエビデンスが不足している状態であり、生体弁 vs. 機械弁、ウシ心膜弁 vs. ブタ大動脈弁、適切なサイズの選択など不明の点も多い。さらに近年本邦で優れた臨床成績を示している sinus を有する PTFE 弁付き PTFE 心外導管 (Yamagishi, Miyazaki) や遠からず導入が期待されるカテーテルによる人工弁挿入など話題は多い。こうした多くの検討項目を頭の中に置いた議論は必ずや明日からの診療に役立つと思われる。国内外での多数の経験や情報、独自の評価法などユニークな視点を持った演者にこれらの問題に対する最近の知見や展望を提示いただき、術前・周術期・術後それぞれの立場での注意点を認識して、本邦での成人期 PVR の質的向上を目指したい。

D3

複雑 ACHDでの妊娠：ここまで可能 vs. ここから不可能
Perinatal care of ACHD patients: Possible vs. Impossible

市川 肇
 国立循環器病研究センター 小児心臓外科
 Hajime Ichikawa
 National Cerebral and Cardiovascular Center Pediatric Cardiovascular Surgery

先天性心疾患の成績の向上により、90%以上が成人し、ファロー四徴症以上の複雑な先天性心疾患患者が生殖可能年齢に到達する。子供を持つことの意味は個人にゆだねられるが女性の妊娠出産への願望はその病態に関係なく多くの場合存在する。シンプルな心房中隔欠損症や心室中隔欠損症で遺残病変の無い場合は健常人と同等の安全性での妊娠出産が可能であるが、それ以上の疾患では詳細な血行動態の評価を行うことが母体および胎児の生命の安全に必要である。また母体の遺残病変や非生理学的循環のため妊娠出産に厳密な全身管理を必要とするような場合、妊娠出産が母体の余命に及ぼす影響は未だに明らかではない。本ディベートセッションではガイドライン上の複雑 ACHD 患者における妊娠出産について取り上げるとともに単心室循環、チアノーゼ性心疾患患者での妊娠出産にも言及しその安全限界について本分野でのエキスパートに議論していただきます。フロアからの質問・意見も歓迎いたします。

OE1-1

Fontan術後の血栓塞栓症予防にワーファリンとアスピリン併用療法は有効である
Combination Therapy with Warfarin and Aspirin is Effective for Thromboembolic Prophylaxis in Patients after the Fontan Operation

梅本 真太郎¹⁾, 坂本 一郎¹⁾, 大谷 規彰¹⁾, 石北 綾子¹⁾, 兒玉 祥彦^{2,3)}, 永田 弾³⁾, 井手 友美¹⁾, 石川 司朗²⁾, 大賀 正一²⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 福岡市立こども病院 循環器科, 3) 九州大学病院 小児科

Shintaro Umemoto¹⁾, Ichiro Sakamoto¹⁾, Kisyo Ohtani¹⁾, Ayako Ishikita¹⁾, Yoshihiki Kodama^{2,3)}, Hazumu Nagata³⁾, Tomomi Ide¹⁾, Shiro Ishikawa²⁾, Shoichi Ohga²⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Medicine, Kyushu University Hospital, 2) Department of Cardiology, Fukuoka Children's Hospital,

3) Department of Pediatrics, Kyushu University Hospital

Introduction: Thromboembolism is one of major complications in Fontan circulation. However, the optimal management strategy remains unestablished, and the efficacy and safety of combination therapy with warfarin and aspirin has not been evaluated.

Method and Results: We retrospectively analyzed 129 patients (median age 24, range: 15–40, years) who had been treated with warfarin and aspirin after the Fontan operation in our hospital. Thromboembolic events were defined as cerebral infarction, pulmonary embolism, deep vein thrombosis and other systemic embolism. Target PT-INR was within 1.5 to 2.0. The median duration of follow-up after the Fontan operation was 19.2 (5.1–27.0) years and thromboembolic events occurred in 4 patients. The thromboembolic event free rate was 99.2%, 98.4% and 97.2% at 10, 15 and 20 years, respectively. On the other hand, major bleeding events occurred in 25 patients (male: female=7:18). The major bleeding event free rate was 96.9%, 91.9% and 76.8% at 10, 15 and 20 years, respectively.

Conclusion: Combination therapy with warfarin and aspirin may be effective for preventing thromboembolic events in patients after the Fontan operation.

OE1-2

成人先天性心疾患における Early Vascular Aging の成因
Early vascular aging in adult patients with congenital heart disease

村上 智明¹⁾, 堀端 洋子²⁾, 立野 滋²⁾, 川副 泰隆²⁾, 丹羽 公一郎³⁾

1) 千葉県こども病院, 2) 千葉県循環器病センター, 3) 聖路加国際病院

Tomoaki Murakami¹⁾, Yoko Horibata²⁾, Shigeru Tateno²⁾, Yasutaka Kawasoe²⁾, Koichiro Niwa³⁾

1) Department of Cardiology, Chiba Children's Hospital, 2) Chiba Cardiovascular Center, 3) St. Luke's International Hospital

The concept of early vascular aging (EVA) seems to be a promising tool for clinical guidance in individuals at increased cardiovascular risk. We analyzed the early vascular aging in adult patients with congenital heart disease.

64 patients with congenital heart disease aged over 20 years were enrolled for the analysis. The EVA was defined as over 5 percentile of age-gender matched control of brachial-ankle pulse wave velocity (baPWV).

The baPWV was 1281 ± 330 cm/s and 10 patients met the definition of EVA. The EVA was significantly correlated with age [years] (OR 1.088; CI 1.031-1.148; $p=0.002$), systolic blood pressure [mmHg] (1.130; 1.049-1.217; 0.001), fasting blood sugar [mg/dl] (1.066; 1.081-1.117; 0.007), hemoglobin A1c [%] (8.088; 1.288-50.782; 0.026), uric acid [mg/dl] (1.831; 1.036-3.235; 0.037), LDL cholesterol [mg/dl] (1.024; 1.002-1.047; 0.035), and triglyceride [mg/dl] (1.013; 1.004-1.022; 0.006).

In patients with EVA, many factors which have been reported as risk factors for cardiovascular disease in general population were elevated. The patients age had a significant impact on EVA, although the definition of EVA contains the patients age.

OE1-3

肺循環とFontan術後遠隔期の心血管イベントの関連

Pulmonary circulation related to late cardiovascular events after Fontan operation

宗内 淳¹⁾, 渡辺 まみ江¹⁾, 杉谷 雄一郎¹⁾, 川口 直樹¹⁾, 松岡 良平¹⁾, 藤井 俊輔¹⁾, 安東 勇介²⁾, 落合 由恵²⁾
 1) JCHO九州病院 小児科, 2) JCHO九州病院 心臓血管外科

Jun Muneuchi¹⁾, Mamie Watanabe¹⁾, Yuichiro Sugitani¹⁾, Naoki Kawaguchi¹⁾, Ryohei Matsuoka¹⁾, Shunsuke Fujii¹⁾, Yusuke Ando²⁾, Yoshie Ochiai²⁾

1) Department of Pediatrics, Japan Community Healthcare Organization Kyushu Hospital, 2) Department of Cardiovascular Surgery, JCHO Kyushu Hospital

Aim: To clarify factors related to cardiovascular events (CVE) late after Fontan operation.

Methods: We studied cardiac catheterization and cardiopulmonary test in 76 Fontan subjects, and compared hemodynamic parameters between subjects with and without late CVE.

Results: Age at Fontan operation and examinations was 3 (2-4) years and 9 (8-13) years, respectively. During the follow-up of 15 (11-16) years, there were 8 CVEs including 3 deaths, 2 heat failure and 2 protein losing enteropathy. There was no significant difference in peak VO₂ (with vs without CVE: 30.3 [26.1-36.1] vs 28.1 [23.4-31.0] ml/min/kg), peak HR (147 [135-164] vs 136 [116-152] bpm), cardiac index (4.0 [3.6-4.6] vs 3.8 [3.0-4.4] L/min/m²), pulmonary arterial index (225 [189-302] vs 216 [186-255] mm²/m²) and pulmonary arterial resistance (1.07 [0.73-1.31] vs 1.04 [0.78-1.66] WU · m²). However, central venous pressure was increased (10 [9-12] vs 16 [13-16] L/min/m², P < 0.001) and pulmonary arterial compliance was decreased (51.0 [36.0-75.0] vs 38.9 [25.7-45.9] mm²/mmHg/m², P < 0.001).

Conclusions: Lower pulmonary arterial compliance and elevated central venous pressure are related to late CVE after Fontan operation.

OE1-4

Ebstein病の中長期予後についての検討

Mid-term clinical outcomes in adult patients with surgically operated Ebstein anomaly

杜 徳尚¹⁾, 赤木 禎治¹⁾, 小谷 恭弘²⁾, 横濱 ふみ¹⁾, 黒子 洋介²⁾, 馬場 健児³⁾, 大月 審一³⁾, 笠原 真悟²⁾, 伊藤 浩¹⁾

1) 岡山大学 循環器内科, 2) 岡山大学 心臓血管外科, 3) 岡山大学 小児循環器科

Norihisa Toh¹⁾, Teiji Akagi¹⁾, Yasuhiro Kotani²⁾, Fumi Yokohama¹⁾, Yosuke Kuroko²⁾, Kenji Baba³⁾, Shin-ichi Otsuki³⁾, Shingo Kasahara²⁾, Hiroshi Ito¹⁾

1) Department of Cardiovascular Medicine, Okayama University, 2) Department of Cardiovascular Surgery, Okayama University, 3) Department of Pediatric Cardiology, Okayama University

Background: We sought to review our experience of adults with operated Ebstein anomaly (EA).

Methods: 35 adult patients with operated EA were identified. Unfavorable cardiovascular events included death, heart failure admission, ventricular arrhythmia, and re-intervention.

Results: Median age at the first clinical visit was 17 years. On the initial visit, 24 patients had tricuspid valve (TV) repair, 2 patients had TV replacement, 4 patients had one-and-a-half ventricular repair, and 5 patients had Fontan palliation. Median follow-up was 7.1 years. One patient died after the re-operation. Six patients experienced heart failure admission and 3 of 6 required reoperations. One patient experienced sustained ventricular tachycardia followed by reoperation. One patient underwent re-TV replacement. Event-free survival was 94%, 90%, 77%, and 64% at 1, 5, 10, and 15 years. In multivariable Cox proportional hazards analysis, serum BNP at the initial visit > 85 pg/ml was independently predictive for unfavorable cardiovascular events.

Conclusions: Adult patients with surgically operated EA continue to have high rates of morbidity and mortality, with need for re-operations.

OE1-5

Fontan循環とその他の成人先天性心疾患での肝病変の特徴の比較

Unique features of hepatic disease in adults with Fontan circulation: A comparison with congenital heart disease patients after two-ventricular repair

杜 徳尚¹⁾, 大西 秀樹²⁾, 赤木 禎治¹⁾, 竹内 康人²⁾, 中村 進一郎²⁾, 横濱 ふみ¹⁾, 高谷 陽一¹⁾, 小谷 恭弘³⁾, 黒子 洋介³⁾, 笠原 真悟³⁾, 岡田 弘之²⁾, 伊藤 浩¹⁾

1) 岡山大学 循環器内科, 2) 岡山大学 消化器・肝臓内科学, 3) 岡山大学 心臓血管外科

Norihia Toh¹⁾, Hideki Onishi²⁾, Teiji Akagi¹⁾, Yasuto Takeuchi²⁾, Shinichiro Nakamura²⁾, Fumi Yokohama¹⁾, Yoichi Takaya¹⁾, Yasuhiro Kotani³⁾, Yosuke Kuroko³⁾, Shingo Kasahara³⁾, Hiroyuki Okada²⁾, Hiroshi Ito¹⁾

1) Department of Cardiovascular Medicine, Okayama University,

2) Department of Gastroenterology and Hepatology, Okayama University,

3) Department of Cardiovascular Surgery, Okayama University

Background: We sought to explore the features and predictors of liver disease in Fontan physiology in comparison with other adult congenital heart disease (ACHD) patients.

Methods: A total of 89 ACHD patients (51 with Fontan and 38 with two-ventricular-repair [2VR]) were enrolled in the study. Echo, abdominal ultrasound, FibroScan, cardiac catheterization, and labo-parameters were evaluated. Cardiac events included death, heart failure, and arrhythmic events.

Results: BNP, hepatic fibrosis findings, and central venous pressure (CVP) were significantly elevated in Fontan. FibroScan value correlated with Fontan/definitive operation duration, systemic ventricular ejection fraction, and CVP (all $p < 0.05$). Multiple logistic regression analysis showed that Fontan physiology was the predictor of elevated FibroScan (> 12.5 kPa) (OR, 16.1; 95%CI, 3.3 to 79.0). However, in Fontan and 2VR, FibroScan value showed no significant difference between patients with and without cardiac events ($p=0.992$ and 0.741 , respectively).

Conclusion: Hepatic structural abnormality is common in Fontan compared with other ACHDs and Fontan physiology is an independent predictor of liver fibrosis.

OE1-6

フォンタン術後妊娠の胎盤病理

The placental pathology of women with functional single ventricle after Fontan palliation

小西 妙¹⁾, 大郷 恵子²⁾, 植田 初江²⁾, 神谷 千津子¹⁾, 澤田 雅美¹⁾, 塩野入 規¹⁾, 中西 篤史¹⁾, 堀内 縁¹⁾, 釣谷 充弘¹⁾, 岩永 直子¹⁾, 吉松 淳¹⁾

1) 国立循環器病研究センター 周産期婦人科, 2) 国立循環器病研究センター 病理部

Tae Konishi¹⁾, Keiko Ohta Ogo²⁾, Hatsue Ishibashi Ueda²⁾, Chizuko A Kamiya¹⁾, Masami Sawada¹⁾,

Tadasu Shionoiri¹⁾, Atsushi Nakanish¹⁾, Chinami Horiuchi¹⁾, Mitsuhiro Tsuritani¹⁾, Naoko Iwanaga¹⁾, Jun Yoshimatsu¹⁾

1) Department of Perinatology and Gynecology, National Cerebral and Cardiovascular Center,

2) Department of Pathology, National Cerebral and Cardiovascular Center

Objectives: Pregnancies with Fontan circulation have high risks of miscarriage, premature delivery and fetal growth restriction. The purpose of the current study is to investigate its pathology by the examination of placentae in women with Fontan circulation.

Methods: Five pregnancies with Fontan circulation delivered at our institution between 2009 and 2018 were retrospectively reviewed.

Results: Maternal median SpO₂ was 94 [range: 91-96] %, central venous pressure was 10 [9-11] mmHg, cardiac index (CI) was 2.75 [2.25-3.67] L/min/m² at pre-pregnancy. The median gestational age at delivery was 34 [30-37] weeks. There were 3 cases of preterm delivery and fetal growth restriction (FGR). All cases of Fetal/Placental weight ratio were smaller compared to each gestational age. Histopathologically, all placentae showed increased syncytial knots, formation of vasculo-syncytial membrane, villous vascular congestion, villous stromal fibrosis, and villous branching.

Conclusion: In all cases, weight of placentae tended to be lighter compared with their fetal weight. All placentae with Fontan circulation showed histological findings of placental hypoxia and maternal under-perfusion.

OE1-7

先天性心疾患合併妊娠における産後BNPと授乳状況の関連についての検討

Relations between breastfeeding and postpartum changes in brain natriuretic peptide among mothers with congenital heart disease

松坂 優, 神谷 千津子, 横内-小西 妙, 澤田 雅美, 塩野入 規, 中西 篤史, 堀内 縁, 釣谷 充弘, 岩永 直子, 吉松 淳

国立循環器病研究センター 周産期・婦人科部

Yu Matsuzaka, Chizuko A Kamiya, Tae Yokouchi Konishi, Masami Sawada, Tadasu Shionoiri, Atsushi Nakanishi, Chinami Horiuchi, Mitsuhiro Tsuritani, Naoko Iwanaga, Jun Yoshimatsu

Department of Perinatology and Gynecology, National Cerebral and Cardiovascular Center

Introduction: Brain natriuretic peptide (BNP) is a biomarker that reflects ventricular blood volume. The aim of this study was to evaluate how breastfeeding affects postpartum changes in BNP among mothers with congenital heart disease (CHD).

Methods: A retrospective review of patients with CHD who received perinatal care at our institute between 2017 and 2018 was conducted. Clinical data about patients' backgrounds, obstetrical outcomes, breastfeeding status, and postpartum BNP levels were collected.

Results: 32 women were primarily breastfeeding (breastfed group) and 22 primarily formula feeding (formula group). The average levels of BNP within one week postpartum and at one month postpartum were not statistically different between the two groups (47.89 and 38.24 mg/dL respectively in the breastfed group and 71.14 and 38.46 mg/dL in the formula group; $p=0.086, 0.987$). Average BNP levels decreased by 9.65 mg/dL in the breastfed group compared to 32.68 mg/dL in the formula group, showing a marginal difference ($p=0.058$).

Conclusion: Our study suggests that postpartum ventricular volume reduction is possibly greater in women that don't breastfeed, but further studies are needed.

OE2-1

4D flow MRIを用いた右室機能と血行動態から見た成人先天性心疾患の肺動脈弁手術適応
Surgical Indication for Pulmonary Valve Disease in Adult Congenital Heart Disease Based on Right Ventricular Hemodynamics Assessed with 4D flow MRI

板谷 慶一¹⁾, 山岸 正明²⁾, 前田 吉宣²⁾, 藤田 周平²⁾, 本宮 久之²⁾, 高柳 佑士²⁾, 夫 悠²⁾, 森地 裕子¹⁾, 宮崎 翔平³⁾, 梶山 葉⁴⁾, 瀧上 雅雄⁵⁾, 中西 直彦⁵⁾, 的場 聖明⁵⁾, 夜久 均¹⁾

1) 京都府立医科大学 心臓血管外科 心臓血管血流解析学, 2) 京都府立医科大学 小児心臓血管外科, 3) Cardio Flow Design Inc., 4) 京都府立医科大学 小児科, 5) 京都府立医科大学 循環器内科

Keiichi Itatani¹⁾, Masaaki Yamagishi²⁾, Yoshinobu Maeda²⁾, Shuhei Fujita²⁾, Hisayuki Hongu²⁾, Yuji Takayanagi²⁾, Haruka Fu²⁾, Hiroko Morichi¹⁾, Shohei Miyazaki³⁾, Yo Kajiyama⁴⁾, Masao Takigami⁵⁾, Naohiko Nakanishi⁵⁾, Satoaki Matoba⁵⁾, Hitoshi Yaku¹⁾

1) Department of Cardiovascular Surgery, Kyoto Prefectural University of Medicine,

2) Pediatric Cardiovascular Surgery, Kyoto Prefectural University of Medicine, 3) Cardio Flow Design Inc.,

4) Department of Pediatrics, Kyoto Prefectural University of Medicine,

5) Department of Cardiology, Kyoto Prefectural University of Medicine

Background: Surgical Indication for pulmonary valve in adult congenital heart disease is still unclear. 4D flow MRI was used to assess right ventricle (RV) hemodynamics.

Method: 21 patients (28.3 ± 13.7 years old) with tetralogy of Fallot or its related disease, transposition of great arteries, two chamber RV, and post Ross state were enrolled, and classified into pulmonary stenosis (PS, N=6), regurgitation (PR, N=5), stenosis with regurgitation (PSR, N=9), and those after RV outflow reconstruction or valve replacement (N=7). 4D flow MRI was performed to evaluate RV end diastolic and systolic volume (RVEDV/RVESV), cardiac output (CO), regurgitation fraction (RF), and flow energy loss (EL). Analysis of variance was used for statistical analysis.

Results: No statistical difference was observed in age and body size. PR group had significantly lower CO and higher RV volume than other groups. EL was significantly reduced after the surgery (to 1.9 ± 0.4 mW), and was higher in PSR and PR than in PS (7.6, 5.7, and 4.9 mW, respectively).

Conclusions: Regurgitation caused RV deterioration, so early intervention is preferred in those with RF over 15% and EL over 3.0 mW.

OE2-2

成人先天性心疾患における心臓MRIを用いたNative T1とECV値の有用性

Native T1 and Extracellular Volume at Cardiac Magnetic Resonance in Adults with Congenital Heart Disease

椎名 由美¹⁾, 谷口 宏太³⁾, 長尾 充展⁴⁾, 高橋 辰徳⁵⁾, 河窪 正照⁶⁾, 稲井 慶²⁾

1) 聖路加国際病院 心血管センター, 2) 東京女子医大 循環器小児科, 3) 北海道大学 小児科, 4) 東京女子医大 放射線科, 5) 山形大学 小児科, 6) 九州大学大学院医学研究院 保健学部門

Yumi Shiina¹⁾, Kota Taniguchi³⁾, Michinobu Nagao⁴⁾, Tatsunori Takahashi⁵⁾, Masateru kawakubo⁶⁾, Kei Inai²⁾

1) Cardiovascular Center, St. Lukes International Hospital,

2) Department of Pediatric Cardiology, Tokyo Women's Medical University, 3) Department of Pediatrics, Hokkaido University,

4) Department of Diagnostic Imaging and Nuclear Medicine, 5) Department of Pediatric Cardiology, Yamagata University,

6) Department of Health Sciences, Faculty of Medical Sciences, Kyushu University

Myocardial fibrosis is considered to be a substrate for fatal ventricular arrhythmias in congenital heart diseases. A novel technique on cardiac magnetic resonance (CMR), T1 mapping, can characterize diffuse interstitial myocardial changes.

1. MOLLI (Modified Look-Locker Inversion-recovery) vs saturation recovery methods: RV native T1 value and ECV using MOLLI method showed good correlation with RV EF. As previous reports, our study also showed that saturation recovery method yields higher accuracy, lower precision, and similar reproducibility compared with MOLLI for T1 measurement. Both sequences have similar reproducibility for ECV quantification.

2. Native T1 value and ECV of systemic RV are higher than healthy controls: These values were similar to those in dilated cardiomyopathy.

3. Native T1 value, extracellular volume and GLS in repaired TOF: Native T1 value and ECV using MOLLI in adults with TOF significantly correlated with biventricular global longitudinal strain (GLS), suggesting that native T1 value and ECV reflect potential biventricular dysfunction.

OE2-3

成人先天性心疾患における3Dプリンティングモデルの利用

Applications of three-dimensional printed modeling of adult congenital heart disease

三好 亨¹⁾, 杜 徳尚¹⁾, 赤木 禎治¹⁾, 三木 崇史¹⁾, 小山 靖史²⁾, 伊藤 浩¹⁾

1) 岡山大学病院 循環器内科, 2) 桜橋渡邊病院 放射線科

Toru Miyoshi¹⁾, Norihisa Toh¹⁾, Teiji Akagi¹⁾, Takashi Miki¹⁾, Yasushi Koyama²⁾, Hiroshi Ito¹⁾

1) Department of Cardiovascular Medicine, Okayama University, 2) Department of Radiology, Sakurabashi Watanabe Hospital

Background: Current diagnostic assessment tools remain suboptimal in demonstrating complex morphology of adult congenital heart disease (ACHD). This limitation has posed several challenges in preoperative planning, communication in medical practice, and medical education. This study aims to investigate the impact of 3D printed model of ACHD in the above three areas.

Methods: Using cardiac CT data, patient-specific 3D models including atrioventricular septal defect, truncus arteriosus, and tetralogy of Fallot after surgery etc. were printed. Survey was conducted by Likert-type questionnaires to cardiologists, cardiac surgeons, nurses, and medical students.

Results: The complex cardiac anatomy can be accurately replicated in 3D printed model. With these 3D models, feedback shown in the questionnaires found the model to be helpful in facilitating preoperative planning, enhancing patient-doctor communication, and learning the pathology quicker with better understanding.

Conclusions: 3D printed ACHD models could serve as an excellent tool in the field of clinical practice and medical education.

OE2-4

大動脈縮窄症に対する外科手術とカテーテル治療の比較

Comparison between surgery and catheter intervention for management of coarctation of the aorta in adults.

加藤 温子¹⁾, 佐藤 純¹⁾, 吉井 公浩¹⁾, 森本 美仁¹⁾, 吉田 修一朗¹⁾, 西川 浩¹⁾, 大橋 直樹²⁾, 櫻井 寛久²⁾, 野中 利通²⁾, 櫻井 一²⁾

1) JCHO中京病院 中京こどもハートセンター 小児循環器科, 2) JCHO中京病院 中京こどもハートセンター 心臓血管外科

Atsuko Kato¹⁾, Jun Sato¹⁾, Kimihiro Yoshii¹⁾, Yoshihito Morimoto¹⁾, Shuichiro Yoshida¹⁾, Hiroshi Nishikawa¹⁾, Naoki Ohashi²⁾, Takahisa Sakurai²⁾, Toshimichi Nonaka²⁾, Hajime Sakurai²⁾

1) Department of Pediatric Cardiology, JCHO Chukyo Hospital, 2) Department of Cardiovascular Surgery, JCHO Chukyo Hospital

Background: Stent implantation in coarctation of the aorta (CoA) has been considered as the first choice of treatment in the world, but not in Japan. We sought to compare safety and cost-effectiveness between surgery and catheter intervention in adult CoA.

Methods: All the patients were included with CoA older than 15 years old who underwent surgery or catheter intervention in our hospital from 2006 to 2018 and compared according to the treatment.

Results: There were 4 surgical cases and 1 stent case. Surgical intervention included interpose with an ePTFE graft (n=1), end-to-end anastomosis (n=2), and patch enlargement (n=1). A PALMAZ large stent was implanted in a patient. The procedure time and length of hospital stay were longer in surgical patients than a stent patient (499 ± 183 minutes vs 86 minutes, 19 ± 5 days vs 6 days). There were two major complications in surgical patients. The medical cost was more in surgical patients (3.0 ± 0.5 million yen vs 1.1 million yen)

Conclusion: Stent implantation in CoA can provide less invasive intervention with shorter hospital stay and less medical cost, and should be regarded as the prime option for adult patients.

OE2-5

二尖大動脈弁の再建手術とMICSの可能性

Repair of Reconstruction of Bicuspid Aortic Valve - including consideration of minimally invasive approaches

米田 正始, 藤原 祥司, 氏家 敏巳

医誠会病院 心臓血管外科

Masashi Komeda, Shoji Fujiwara, Toshimi Ujiie

Department of Cardiovascular Surgery, Iseikai Hospital

Purpose: Understanding of the geometry of bicuspid aortic valve (BAV) in patients with aortic regurgitation (AR) has been improved. Based on it, we try to improve the surgery for BAV with AR.

Method: Out of 62 patients whom we reconstructed the aortic valve in the past 8 years, 26 had BAV. Among them, 18 had repair (Repair Group, 16 men, 34 ± 14y.o.), 6 pericardial reconstruction (Pericard Group, 5 men, 43 ± 6y.o.), 2 valve sparing root reconstruction (Root Group, 2 females, 54 ± 12y.o.). In the repair, cusp plication and STJ/VAJ adjustment were done to make effective height (eH) ≥ 10mm.

Results: There was no hospital death. Five patients of Repair Group had MICS approaches. In 2 patients, pressurized regurgitation test helped accurate evaluation. AR changed from 3.4 ± 0.6 degree (preop.) to 1.1 ± 0.8* (postop.) in Repair Group, from 4.0 ± 0.0 to 0.2 ± 0.4* in Pericard Group, from 3.0 ± 1.4 to 0.5 ± 0.7 in Root Group. (*p ≤ .05 vs. preop. by t-test). There were no late death with the follow-up of 37 ± 22 months.

Conclusions: Results of the repair improved after introducing eH adjustment and MICS approach has become feasible. Pressurized regurgitation test seems useful.

OE2-6

成人先天性心疾患手術における MICS の役割

Minimally Invasive Cardiac Surgery for Simple Congenital Heart Surgery in the Adults

小谷 恭弘, 川田 幸子, 堀尾 直裕, 小林 泰幸, 田井 龍太, 迫田 直也, 辻 龍典, 後藤 拓弥, 黒子 洋介,
新井 禎彦, 笠原 真悟
岡山大学 心臓血管外科

Yasuhiro Kotani, Sachiko Kawada, Naohiro Horio, Yasuyuki Kobayashi, Ryuta Tai, Naoya Sakoda, Tatsunori Tsuji,
Takuya Goto, Yosuke Kuroko, Sadahiko Arai, Shingo Kasahara
Department of Cardiovascular Surgery, Okayama University

Objective: We sought to see the surgical outcome of MICS in simple congenital heart surgery in the adults. Patients: Retrospective study was performed in 95 patients > 16 years old who underwent congenital heart surgery between 1999 and 2017. Fifty three out of 95 (56%) patients underwent MICS. Cardiopulmonary bypass (CPB) was routinely established by means of a central cannulation. Results: Diagnosis included atrial septal defect (ASD) in 53 patients and ventricular septal defect (VSD) in 15 patients. Twenty (21%) patients had a right thoracotomy approach to close ASD. Three females having VSD had a submammary skin incision. There were no death and no CPB-related complications. When comparing between a right thoracotomy and a median sternotomy in ASD closure, CPB time was significantly longer in right thoracotomy approach (44 [34-54] min vs. 36 [30-46] min, p=0.019), but aortic cross clamping time was comparable (18 [15-25] min vs. 15 [11-20] min, p=0.071). Conclusions: MICS was performed in more than half of simple congenital heart surgery in the adults. Right thoracotomy approach can be performed without having longer myocardial ischemic time compared to median sternotomy.

OE2-7

左室緻密化障害 (LVNC) への外科治療

Surgical treatment of left ventricular non-compaction (LVNC)

米田 正始, 藤原 祥司, 氏家 敏巳
医誠会病院 心臓血管外科

Masashi Komeda, Shoji Fujiwara, Toshimi Ujiiie
Department of Cardiovascular Surgery, Iseikai Hospital

Background: LVNC is a disease with poor prognosis, but, those who reach adult age may have curable part of the disease.

Methods: We reviewed 7 patients who had surgery for LVNC in the past 10 years (3 males, median age 66): 3 had LV repair for dilated cardiomyopathy (DCM Group), 2 removal of ventricular stenosis (Stenosis Group), and 2 valve surgery (Valve Group).

Results: There was no hospital mortality. In DCM Group, there was no late death with patients' survival for 10yrs, 1yr and 5 months. LVDd changed from 60 ± 2mm (preop.) to 53 ± 9 (postop.), EF from 25 ± 3% to 32 ± 6, and RV pressure from 39 ± 20mmHg to 37 ± 7, respectively. In Stenosis Group, 1 patient survived for 5 years and follow-up was lost thereafter while the other patient who had preop. RV pressure of 80mmHg had transplant 4 years postoperatively. In Valve Group, patients' symptom improved, but one patient 84 y.o. male with preop. Dd63mm and EF23% died of heart failure 15 months postoperatively.

Conclusions: For patients with LVNC, surgery seems to be effective for DCM Group and Valve Group. Simplified LV repair may be helpful. Preoperative pulmonary hypertension may carry high risk.

OE2-8

成人期多脾症候群に対する手術の経験：肺血管病理所見との対比

Surgical experience of Adults with Polysplenia syndrome: Clinical relevance with lung biopsy findings

河田 政明¹⁾, 吉積 功¹⁾, 鷗垣 伸也¹⁾, 片岡 功一²⁾, 関 満²⁾, 岡 健介²⁾, 松原 大輔²⁾, 今井 靖³⁾,
甲谷 友幸³⁾, 久保田 香菜³⁾

1) 自治医科大学とちぎ子ども医療センター・成人先天性心疾患センター 小児・先天性心臓血管外科, 2) 小児科, 3) 循環器内科

Masaaki Kawada¹⁾, Ko Yoshizumi¹⁾, Shin-ya Ugaki¹⁾, Ko-ichi Kataoka²⁾, Mitsuru Seki²⁾, Kenske Oka²⁾,

Daisuke Matsubara²⁾, Yasushi Imai³⁾, Tomoyuki Kabutoya³⁾, Kana Kubota³⁾

1) Pediatric and Congenital Cardiovascular Surgery, Jichi Adult Congenital Heart Center, 2) Pediatric Cardiology,

3) Cardiovascular Medicine

Not a few polysplenia syndrome patient survive into adulthood without reparative surgery. We reviewed experience of adult cases in this 10-year interval. Through 2007 to 2017, 4 adults (ages; 26 to 36 years, 2 males and 2 females) referred for surgery. One had a biventricular morphology and other three cases basically had single ventricle morphology and hemodynamics. All suffered from various rhythm disturbances. All survived surgery and followed up for up to 9 years through biventricular repair in 1 and single stage TCPC completion in another. Remaining 2 stayed at palliated stages. Pacing required in all (AAI 2, DDD 1, VVI 1). Palliated patients showed SpO₂ around 82 to 88%. Medial thickening and scattered thrombo-occlusion and recanalization with irregular intimal fibrosis were common in lung specimens. Patients, unrepaired until adulthood, commonly have maintained pulmonary blood flow and as its consequence pulmonary arteries have reactive thickening of media and luminal damages from long-standing cyanosis and polycythemia. Surgical treatment of unrepaired adults yields a reasonable outcome. Pulmonary vascular findings offer informations for selection of surgical approach.

OJ1-1

経胸壁心エコー図によるPFO診断における腹部圧迫法とバブルカットオフ値の有用性
Importance of Abdominal Compression Valsalva Maneuver and Microbubble Cutoff of Transthoracic Echocardiography for Detecting Patent Foramen Ovale

高谷 陽一¹⁾, 渡辺 修久²⁾, 池田 まどか²⁾, 赤木 禎治¹⁾, 中川 晃志¹⁾, 中山 理絵¹⁾, 杜 徳尚¹⁾, 伊藤 浩¹⁾
1) 岡山大学 循環器内科, 2) 岡山大学病院 検査部

Yoichi Takaya¹⁾, Nobuhisa Watanabe²⁾, Madoka Ikeda²⁾, Teiji Akagi¹⁾, Koji Nakagawa¹⁾, Rie Nakayama¹⁾, Norihisa Toh¹⁾, Hiroshi Ito¹⁾
1) Department of Cardiovascular Medicine, Okayama University, 2) Division of Medical Support, Okayama University Hospital

Background: Although transthoracic echocardiography (TTE) is useful to diagnose patent foramen ovale (PFO), optimal methodologies are not established. We aimed to evaluate the efficacy of abdominal compression Valsalva maneuver (VM) and microbubble (MB) cutoff.

Methods: We analyzed 101 patients with suspected PFO. TTE was performed at spontaneous VM and abdominal compression VM. PFO was defined as positive if ≥ 1 or ≥ 5 MBs were seen in left chambers.

Results: Of 101 patients, 62 were confirmed PFO by transesophageal echocardiography and/or catheterization. The sensitivity of TTE in detecting PFO was 92% at spontaneous VM and 100% at abdominal compression VM, when ≥ 1 MB was used as the cutoff. The sensitivity was 85% at spontaneous VM and 100% at abdominal compression VM, when ≥ 5 MBs was used. At abdominal compression VM, the specificity was increased in the cutoff of ≥ 5 MBs compared with ≥ 1 MB (85% vs. 49%). The abdominal compression VM with the cutoff of ≥ 5 MBs provided the higher accuracy of 94%.

Conclusions: TTE with the criteria of ≥ 5 MBs cutoff at abdominal compression VM provides the excellent accuracy for PFO diagnosis.

OJ1-2

心房中隔欠損症患者における右室容積と exercise capacity の関連性について
Relationship between right ventricular volume and exercise capacity in patients with atrial septal defect

中山 理絵¹⁾, 高谷 陽一¹⁾, 赤木 禎治¹⁾, 渡辺 修久²⁾, 池田 まどか²⁾, 中川 晃志¹⁾, 杜 徳尚¹⁾, 伊藤 浩¹⁾
1) 岡山大学 循環器内科, 2) 岡山大学病院 超音波センター

Rie Nakayama¹⁾, Yoichi Takaya¹⁾, Teiji Akagi¹⁾, Nobuhisa Watanabe²⁾, Madoka Ikeda²⁾, Koji Nakagawa¹⁾, Norihisa Toh¹⁾, Hiroshi Ito¹⁾
1) Department of Cardiovascular Medicine, Okayama University, 2) Division of Medical Support, Okayama University Hospital

Background and Purpose: Atrial septal defect (ASD) with right ventricular (RV) dilatation is considered to be the indication for defect closure, however few studies have been reported the relation between RV dilatation and reduction in cardiopulmonary function. Therefore, we examined the relationship between RV volume and exercise capacity in adult ASD patients.

Methods: We analyzed 56 patients with ASD. Magnetic resonance imaging and Symptom-limited cardiopulmonary exercise test were performed, and we evaluated the relationship between RV volume and maximal oxygen uptake (VO_2 max). Additionally, we estimated the cutoff value of the RV volume that could cause the reduction in cardiopulmonary function using the ROC curve.

Results: There was a correlation between RV end diastolic volume (RVEDV) index and VO_2 max. The optimal cutoff value of RVEDV index at less than 80% of predicted VO_2 max was 112 ml/m^2 .

Conclusions: In adult ASD patients, there was a relationship between the dilatation of RVEDV and the reduction in cardiopulmonary function. Moreover, ASD patients with RVEDV index more than 112 ml/m^2 may be having heart failure symptoms, therefore should be performed ASD closure.

OJ1-3

より質の高い診療を目指して、チームで支える成人先天性心疾患カテーテル治療

Collaborative strategy of catheter interventions for higher quality of life in patients with adult congenital heart disease

片岡 功一^{1,2,3)}, 河田 政明^{1,3,4)}, 松原 大輔²⁾, 岡 健介²⁾, 古井 貞浩²⁾, 安済 達也²⁾, 関 満²⁾, 佐藤 智幸²⁾, 今井 靖^{3,5)}, 甲谷 友幸^{3,5)}, 久保田 香菜^{3,5)}

1) 自治医科大学とちぎ子ども医療センター 小児手術・集中治療部, 2) 自治医科大学とちぎ子ども医療センター 小児科,
3) 自治医科大学成人先天性心疾患センター, 4) 自治医科大学とちぎ子ども医療センター 小児・先天性心臓血管外科,
5) 自治医科大学 循環器内科

Koichi Kataoka^{1,2,3)}, Masaaki Kawada^{1,3,4)}, Daisuke Matsubara²⁾, Kensuke Oka²⁾, Sadahiro Furui²⁾, Tatsuya Anzai²⁾, Mitsuru Seki²⁾, Tomoyuki Sato²⁾, Yasushi Imai^{3,5)}, Tomoyuki Kabutoya^{3,5)}, Kana Kubota^{3,5)}

1) Pediatric Operating Suite and Intensive Care Unit, Jichi Children's Medical Center Tochigi,
2) Department of Pediatrics, Jichi Children's Medical Center Tochigi,
3) Adult Congenital Heart Center, Jichi Medical University,
4) Pediatric and Congenital Cardiovascular Surgery, Jichi Children's Medical Center,
5) Cardiovascular Medicine, Jichi Medical University

Objective: Based on the characteristics of our children's medical center located at the university hospital, we discuss good collaborative strategies of catheter interventions (CI) for ACHD.

Methods: We retrospectively examined 62 ACHD patients who underwent CI since December 2010.

Results: The patient's age ranged from 16 to 78 yrs. 44 ASD closure including 4 atypical cases: 2 right to left shunt (1 Ebstein's anomaly, 1 Critical PS after PTPV) and 2 paradoxical cerebral embolism, 12 PDA closure including 2 using Vascular Plug, and 6 others: 3 PTA for PS, 1 MAPCAs occlusion, 1 PAVM occlusion, 1 VV collaterals occlusion after TCPC. Many patients had comorbidities: hypertension, arrhythmia, renal failure and so on. All CI, discussed at regular conference including pediatric cardiologists, adult cardiologists and cardiovascular surgeons, was successful without any serious complications.

Discussion: Pediatric cardiologists specialize in anatomy and pathophysiology of CHD, adult cardiologists are familiar with comorbidities in adults, and surgeons can overview from surgical standpoint. Multidisciplinary collaboration within the facility is important for higher quality medical care.

OJ1-4

心房細動合併心房中隔欠損症に対し、閉鎖前にアブレーションを行うことの有効性

The efficacy of catheter ablation before transcatheter closure of atrial septal defects in patients with atrial fibrillation ≥40 years of age

小木曾 正隆¹⁾, 江島 浩一郎¹⁾, 杉山 央²⁾, 萩原 誠久¹⁾

1) 東京女子医科大学 循環器内科, 2) 東京女子医科大学 小児循環器科

Masataka Ogiso¹⁾, Koichiro Ejima¹⁾, Hisashi Sugiyama²⁾, Nobuhisa Hagiwara¹⁾

1) Department of Cardiology, Tokyo Women's Medical University,
2) Department of pediatric cardiology, Tokyo Women's Medical University

Background: Atrial fibrillation (AF) and atrial septal defect (ASD) often coexist in elderly patients, with AF adversely affecting the prognosis of patients with ASD. Catheter ablation is an effective therapeutic option for patients with AF. However, little is known about the efficacy of catheter ablation before transcatheter closure of ASD in elderly patients with AF.

Methods and results: Among 205 consecutive patients who underwent transcatheter closure of ASD at our hospital between April 2007 and March 2018, we retrospectively identified 31 patients with paroxysmal AF (pAF) ≥ 40 years of age (mean age, 62.1 ± 9.0 years; male, 48.4%), including 17 patients who underwent catheter ablation for AF (the ablation group) and 14 patients who did not before transcatheter closure of ASD (the non-ablation group). In the follow-up period (2.3 ± 2.1 years), the ablation group had significantly fewer recurrence than the non-ablation group (2 cases vs. 8 cases, log-rank p = 0.039).

Conclusion: The catheter ablation prior to transcatheter closure of ASD might be effective for the elderly patients with pAF.

OJ1-5

当院における成人ASD患者に対するカテーテル閉鎖術の現状

The outcomes among adults post transcatheter atrial septal defect closure

田中 秀門¹⁾, 桑原 直樹¹⁾, 面家 健太郎^{1,4)}, 寺澤 厚志¹⁾, 山本 哲也¹⁾, 後藤 浩子¹⁾, 桑原 尚志¹⁾, 片桐 絢子²⁾, 腰山 宏²⁾, 岩田 祐輔²⁾, 竹内 敬昌²⁾, 吉真 孝^{3,4)}, 小野 浩司³⁾, 野田 俊之³⁾

1) 岐阜県総合医療センター 小児循環器科内科, 2) 岐阜県総合医療センター 小児心臓外科, 3) 岐阜県総合医療センター 循環器内科, 4) 岐阜県総合医療センター 先天性心疾患診療科

Hideto Tanaka¹⁾, Naoki Kuwabara¹⁾, Kentaro Omoya^{1,4)}, Atsushi Terazawa¹⁾, Tetsuya Yamamoto¹⁾, Hiroko Goto¹⁾, Takashi Kuwahara¹⁾, Junko Katagiri²⁾, Hiroshi Koshiyama²⁾, Yusuke Iwata²⁾, Takamasa Takeuchi²⁾, Takashi Yoshitane^{3,4)}, Koji Ono³⁾, Toshiyuki Noda³⁾

1) Department of Pediatric Cardiology, Gifu Prefectural General Medical Center,

2) Department of Pediatric Cardiac Surgery, Gifu Prefectural General Medical Center,

3) Department of Cardiology, Gifu Prefectural General Medical Center,

4) Department of ACHD, Gifu Prefectural General Medical Center

Background: Recently, Transcatheter Closure (TC) has become the main therapy for many secundum atrial septal defects (ASD). In our hospital, all adult patients are treated with TC by pediatric cardiologists.

Purpose: We report the outcome of TC for adults (> 16years old) and the way how to work together with adult cardiologists in our institution.

Methods: We retrospectively analyzed 28 patients treated with TC from July 2008 to July 2018.

Results: 19 were introduced from adult cardiologist in our institution. 8 were directly introduced from adult cardiologist in other institutions. Some past medical history were recorded (HT: 5, DM: 2, arrhythmia [PVC, non-sustained VT, PAF, Af]: 4, and more). 11 were diagnosed by medical examination, 15: by cardiac symptom, 1: by stroke, 1: by deep vein thrombosis. 18 were examined by cardiac catheter, 3 were treated arrhythmia with catheter ablation before TC. Device implantation was successful in all patients. 1 case of retroperitoneal hematoma was reported as major complication.

Conclusions: It is difficult to treat ASD with TC in adult patients with past illness and severe complications. We necessary cooperate with cardiologists.

OJ1-6

当院における成人動脈管開存症に対する経カテーテル的閉鎖術の検討

Transcatheter closure for patent ductus arteriosus in adults at our hospital

赤澤 祐介¹⁾, 鈴木 萌子¹⁾, 中尾 恭久¹⁾, 東 晴彦¹⁾, 佐々木 康浩¹⁾, 藤井 昭¹⁾, 上谷 晃由¹⁾, 青野 潤¹⁾, 永井 啓行¹⁾, 西村 和久¹⁾, 井上 勝次¹⁾, 池田 俊太郎¹⁾, 宮田 豊寿³⁾, 森谷 友造³⁾, 千阪 俊行³⁾, 高田 秀実^{2,3)}, 檜垣 高史^{2,3)}, 石井 榮一^{2,3)}, 山口 修¹⁾

1) 愛媛大学大学院医学系研究科 循環器・呼吸器・腎高血圧内科学, 2) 愛媛大学大学院医学系研究科 地域小児・周産期学講座, 3) 愛媛大学大学院医学系研究科 小児科学講座

Yusuke Akazawa¹⁾, Moeko Suzuki¹⁾, Yasuhisa Nakao¹⁾, Haruhiko Higashi¹⁾, Yasuhiro Sasaki¹⁾, Akira Fujii¹⁾, Teruyoshi Uetani¹⁾, Jun Aono¹⁾, Takayuki Nagai¹⁾, Kazuhisa Nishimura¹⁾, Katsuji Inoue¹⁾, Shuntaro Ikeda¹⁾, Toyohisa Miyata³⁾, Tomozou Moritani³⁾, Toshiyuki Chisaka³⁾, Hidemi Takata^{2,3)}, Takashi Higaki^{2,3)}, Eiichi Ishii^{2,3)}, Osamu Yamaguchi¹⁾

1) Ehime University Graduate School of Med, Dept of Cardiology, Pulmonology, Hypertension & Nephrology,

2) Ehime University Graduate School of Med, Dept of Regional Ped. and Perinatology,

3) Ehime University Graduate School of Medicine, Department of Pediatrics

Background: There have been only a few reports of evaluation of the treatment of transcatheter closure for the patent ductus arteriosus (PDA) in adults.

Method: Out of 123 PDA patients who have undergone transcatheter closure at our hospital from Jan. 1997 to Sep. 2018, 15 patients (12.2%) at the age of 15 years or older were retrospectively studied.

Result: Patients (1 male, 14 females) with a median age of 54 (15-84) years were analyzed. PDA was found during a medical checkup or study of another disease (8 cases, 53%), and the symptom of heart failure and/or atrial fibrillation (4 cases, 27%). Coil (5 cases, 33%)/AMPLATZER[®] Duct Occluder (ADO) (10 cases, 67%) was placed. The procedural success was 100%. The LVEDV decreased 1 month after the closure (p=0.007). Complications were: arteriovenous fistula at a puncture site (1 case), hemolysis due to residual leak (2 cases). ADO placements were performed in both hemolysis cases that had long ampulla lengths (24.4mm, 14.8mm) with calcification.

Conclusion: PDA in adults involves of the risk of residual leak and hemolysis because the malapposition of ADO device occurs due to PDA size and presence of calcification.

OJ2-1

肺気腫を伴った右肺静脈-門脈短絡の一例

Congenital Veno-Portal Shunt Associated with Pulmonary Emphysema

福田 旭伸¹⁾, 木島 康文¹⁾, 金村 宙昌²⁾, 小宮山 伸之¹⁾, 丹羽 公一郎¹⁾

1) 聖路加国際病院 循環器内科, 2) 聖路加国際病院 呼吸器内科

Terunobu Fukuda¹⁾, Yasufumi Kijima¹⁾, Hiroaki Kanemura²⁾, Nobuyuki Komiyama¹⁾, Yoichiro Niwa¹⁾

1) Department of Cardiology, St'lukes International Hospital,

2) Division of Pulmonary Medicine, Thoracic Center, St'lukes International Hospital

A-27-year-old asymptomatic man was referred us for the evaluation of pulmonary emphysema which was incidentally found at medical check-up. Contrast computed tomography showed localized pulmonary emphysema in the right inferior lobe, and coincidentally found aberrant vessels connecting the right inferior pulmonary vein (RIPV) and the portal vein (PoV) forming the esophageal varices. Four pulmonary veins normally combined with the left atrium, which implicate denial for the anomalous pulmonary venous connection. Gadolinium enhanced magnetic resonance imaging and angiography revealed the blood flow running from RIPV to PoV, the direction of which was opposite to the secondary formed varices. Direct measurement of PoV pressure by catheterization showed 8 mmHg as normal. There was no pulmonary hypertension and normal left atrial pressure. Based on these findings, simultaneous presence of the pulmonary emphysema and the aberrant vessels was considered to be a congenital malformation. We would like to discuss whether some interventions for the aberrant vessel are needed or not in this extremely rare case.

OJ2-2

右室流出路再建術の術式が遠隔期の右室機能に与える影響；心臓 MRI による定量評価

Impact on Right Ventricular Parameters of Procedure of Right Ventricular Outflow Tract Reconstruction: Assessment by Cardiac Magnetic Resonance

脇 研自¹⁾, 佐藤 一寿¹⁾, 荻野 佳代¹⁾, 林 知宏¹⁾, 小坂田 皓平²⁾, 大家 理伸²⁾, 福 康志²⁾, 門田 一繁²⁾, 新垣 義夫¹⁾

1) 公益財団法人 大原記念倉敷中央医療機構倉敷中央病院 小児科,

2) 公益財団法人 大原記念倉敷中央医療機構倉敷中央病院 循環器内科

Kenji Waki¹⁾, Kazuhisa Sato¹⁾, Kayo Ogino¹⁾, Tomohiro Hayashi¹⁾, Kohei Osakada²⁾, Masanobu Ohya²⁾, Yasushi Fuku²⁾, Kazushige Kadota²⁾, Yoshio Arakaki¹⁾

1) Department of Pediatrics, Kurashiki Central Hospital, 2) Department of Cardiology, Kurashiki Central Hospital

Purpose: To evaluate the difference in right ventricular (RV) parameters among the patients who underwent three different procedures as RV outflow tract reconstruction (RVOTR) by using cardiac magnetic resonance (CMR) and echocardiography.

Patients and Methods: In 53 patients (mean age 26.2years ± 8.8, 15-59years) after RVOTR, RVEDVI, RVESVI, Pulmonary regurgitant fraction (PRF%) by CMR, and TRPG (mmHg) by echocardiography. Patients were divided into three groups, groupA (commissurotomy: n = 12), groupB (transannular repair: n=27), and groupC (Rastelli: n = 14, Carpentier-Edward 7, Xenomedica 3, Yamagishi 3, Homograft 1).

Results: RVEDVI (131.6ml/m² ± 30.8, p=0.052) and PRF (40.1% ± 13.1, p < 0.001) in groupB were higher than in groupA (117.7ml/m² ± 38.3, 20.9% ± 12.0,) and groupC (96.0ml/m² ± 26.1, 14.5% ± 11.1). TRPG was significantly higher in groupC (51.9mmHg ± 26.7) compared to groupA (29.1mmHg ± 10.6, p < 0.01), however, there was no significant difference compared with that in groupB (44.5mmHg ± 18.6, p=0.50).

Conclusion: Unlike transannular repair, Rastelli repair may be protective in terms of RV dilatation even if implanted valve function is almost lost.

OJ2-3

マスタートド術後遠隔期の体心室右室機能

Systemic right ventricular function in adult patients with transposition of the great arteries after Mustard operation

坂崎 尚徳¹⁾, 石原 温子¹⁾, 豊田 直樹¹⁾, 稲熊 洸太郎¹⁾, 藤原 慶一²⁾, 前田 登史²⁾, 加藤 おと姫²⁾, 植野 剛²⁾, 渡辺 謙太郎²⁾, 大野 暢久²⁾

1) 兵庫県立尼崎総合医療センター 小児循環器内科, 2) 兵庫県立尼崎総合医療センター 心臓血管外科

Hisanori Sakazaki¹⁾, Haruko Ishihara¹⁾, Naoki Toyota¹⁾, Koutaro Inaguma¹⁾, Keiichi Fujiwara²⁾, Toshi Maeda²⁾, Otohime Katou²⁾, Go Ueno²⁾, Kentarou Watanabe²⁾, Nobuhisa Ohno²⁾

1) Department of Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical Center,

2) Department of Cardiovascular Surgery, Amagasaki General Medical Center

background: Systemic right ventricular (SRV) dysfunction in adult patients (pts) with transposition of the great arteries (TGA) after Mustard operation is a major of concern. We aim to evaluate the long term SRV function in our cohort.

methods: We examined SRV function in 12 pts after Mustard operation, aged 43+/-2.3years, by echocardiography, catheter examination, and/or MRI.

Results: Two pts died because of heart failure. Two pts underwent stent implantation of baffle stenosis. One pts underwent pulmonary valve repair for infectious endocarditis. Four pts had severe RV dysfunction, two of those died, and one had paroxysmal atrial fibrillation. Especially one patient has progressed severe diastolic dysfunction with severe pulmonary hypertension. However, in eight patients, RV function was preserved.

Conclusion: SRV function was preserved in 75% of pts after Mustard operation, but progressing SRV diastolic dysfunction was also one of causes of clinical worsening, besides SRV systolic dysfunction.

OJ2-4

Fallot四徴症修復術後の成人の大動脈基部拡大と弾性低下に関する前向きコホート研究

The prospective cohort research for aortic root dilatation and non-elasticity after surgical repair in adults with Tetralogy of Fallot

永峯 宏樹¹⁾, 三浦 大¹⁾, 石津 智子²⁾, 小野 博³⁾, 立野 滋⁴⁾, 前田 潤⁵⁾, 山岸 敬幸⁵⁾, 丹羽 公一郎⁶⁾

1) 東京都立小児総合医療センター 循環器科, 2) 筑波大学 臨床検査医学, 3) 国立成育医療研究センター 循環器科,

4) 千葉県循環器病センター 小児科, 5) 慶應義塾大学 小児科, 6) 聖路加国際病院心血管センター 循環器内科

Hiroki Nagamine¹⁾, Masaru Miura¹⁾, Tomoko Ishizu²⁾, Hiroshi Ono³⁾, Shigeru Taten⁴⁾, Jun Maeda⁵⁾, Takayuki Yamagishi⁵⁾, Kouichirou Niwa⁶⁾

1) Tokyo Metropolitan Children's Medical Center, 2) Laboratory Sport Medicine, Faculty of Medicine, University of Tsukuba,

3) National Center for Child Health and Development, 4) Chiba Cerebral and Cardiovascular Center,

5) Department of Pediatrics, School of Medicine, Keio University, 6) Cardiovascular Center, St Luke's International Hospital

Background: We planned a multicenter cooperative prospective cohort study (TRANSIT) for adults over the age of 20 who underwent repair surgery with diagnosis of TOF for Japanese.

Method: The image of UCG at the initial examination was evaluated by central analysis and the Valsalva (Val) was evaluated. Furthermore, Aortic dilatation (AD) was defined as Val 40 mm or more, and the risk factor between the group with AD (AD group) and the group without AD (the NL group) was examined.

Results: The subjects of the initial survey were 110 cases, ages 20 to 54 years (34y ± 9m), 69 males (63%), Chromosomal aberration 20 cases, right aortic arch 10 cases, PA/VSD 11 cases. Val is 35.0 ± 5.8 mm (112 ± 15% of N). There were 21 cases (19%) in the AD group. Male, aortic pulmonary artery shunt history, PA/VSD, right aortic arch and chromosome abnormality considered as AD risk factors were examined, but there was no significant difference between AD group and NL group.

Discussion: Although AD was seen in 18% of Japanese adults after TOF surgery, it was similar to that reported overseas, but no significant risk factors could be pointed out from the results of the initial survey.

OJ2-5 左室性単心室症に対するseptation術後の遠隔期にone and one half repair施行した2例 Two cases of double inlet left ventricle after septation converted to one and one half (1.5 repair) ventricular repair

武井 陽¹⁾, 上田 知実¹⁾, 小林 匠¹⁾, 吉敷 香菜子¹⁾, 稲毛 章郎¹⁾, 浜道 裕二¹⁾, 矢崎 諭¹⁾, 嘉川 忠博¹⁾, 豊原 啓子²⁾, 竹内 大二²⁾, 高橋 幸宏¹⁾

1) 榊原記念病院, 2) 東京女子医科大学病院

Akira Takei¹⁾, Tomomi Ueda¹⁾, Takumi Kobayashi¹⁾, kanako kishiki¹⁾, Akio Inage¹⁾, Yuji Hamamichi¹⁾, Satoshi Yazaki¹⁾, Tadahiro Yoshikawa¹⁾, Keiko Toyohara²⁾, Daiji Takeuchi²⁾, Yukihiko Takahashi¹⁾

1) Sakakibara Heart Institute, 2) Tokyo Women's Medical University Hospital

Introduction: Few cases reported about re-operation after Septation to single ventricle, as operation itself is rare. case:

Case1: 30 years old, male. He was diagnosed {S, L, L}, DILV, TGA, CoA. When he was 2 months, he was done CoA repair, PA banding. At 7 years, he was done Septation operation and PMI. At 24 years, the severe symptoms of atrial arrhythmia (AT, AFL) was appeared, frequently he needed to be treated by Cardioversion and EA. He was examined, the result were RV volume 57.8ml/m² (60% of Normal), RVEF 55%, TRIII, CVP 16mmHg, LVEF 40%.

Case2: 39 years old, male. He was diagnosed {S, L, L}, DILV, TGA, RAoA. When he was 8 years, he was done Septation operation, PMI. At 32 years, the general fatigue, and AT was appeared, he was done EA. He was examined, the result were RV volume 52% of Normal, RVEF 59%, TRIII, CVP 12mmHg, LVEF 55%.

Both cases was small right ventricle size, they should be done operation of right atrial enlargement caused by right heart overload. They were operated 1.5 repair, right atrium plication TVR and PMI.

Conclusion: We Experienced two cases that single ventricle performed Septation operation. They were done 1.5 repair in postoperative remote period for right heart overload.

OJ2-6 MRI strainによる無症候性術後フォロー四徴症の肺動脈弁置換至適タイミングの検討 Consideration of optimal timing for pulmonary valve replacement in asymptomatic repaired tetralogy of Fallot using feature tracking MR strain

稲毛 章郎¹⁾, 吉敷 香菜子¹⁾, 水野 直和²⁾, 小林 匠¹⁾, 浜道 裕二¹⁾, 上田 知実¹⁾, 矢崎 諭¹⁾, 嘉川 忠博¹⁾

1) 榊原記念病院 小児循環器科, 2) 榊原記念病院 放射線科

Akio Inage¹⁾, Kanako Kishiki¹⁾, Naokazu Mizuno²⁾, Takumi Kobayashi¹⁾, Yuuji Hamamichi¹⁾, Tomomi Ueda¹⁾, Satoshi Yazaki¹⁾, Tadahiro Yoshikawa¹⁾

1) Division of Pediatric Cardiology, Sakakibara Heart Institute, 2) Department of Radiology, Sakakibara Heart Institute

Objective: To consider optimal timing for pulmonary valve replacement (PVR) in asymptomatic repaired tetralogy of Fallot (TOF) using feature tracking magnetic resonance.

Methods: 50 repaired TOF patients were divided into two groups, RV end-diastolic volume (EDV) exceeding 160 ml/m² (group A, n=25) and the group not exceeding 160 ml/m² (group B, n=25). Novel CMR software was used to perform peak global longitudinal strain (GLS) and peak global circumferential strain (GCS) at the RV mid-cavity.

Results: Mean age of group A was 27.1±7.9 years and 31.4±14.2 years in group B. GLS was reduced in both groups compared to normal value (p<0.01). GCS was preserved in group A, however it decreased in group B (p<0.05). GLS and GCS decreased with increasing RV volumes in both groups excluding the correlation between GLS and RVEDV and decreasing RV ejection fraction in both groups.

Conclusions: Preservation of the circumferential strain is important in maintaining RV function in group B. The optimal timing for PVR is considered before RVEDV exceeds 160 ml/m² in asymptomatic repaired TOF patients. However, RV end-systolic volume should also be simultaneously considered in deciding.

OJ3-1

Fontan術後遠隔期の心腔内エコーViewFlexによる導管内観察

Intracardiac echocardiography is feasible to observe the conduit late after Fontan operation.

松岡 良平, 宗内 淳, 相良 優佳, 藤井 俊輔, 川口 直樹, 杉谷 雄一郎, 渡邊 まみ江
地域医療機能推進機構 JCHO九州病院 小児科

Ryohei Matsuoka, Jun Muneuchi, Yuka Sagara, Shunsuke Fuji, Naoki Kawaguchi, Yuichirou Sugitani,
Mamie Watanabe
Department of Pediatrics, Japan Community Healthcare Organization Kyushu Hospital

Background: Intracardiac echocardiography (ICE) has been used in the management of CHD.

Case1: A 34-year-old male with single ventricle visited due to acute abdomen. He underwent extracardiac TCPC at 13 years of age, and had the history of brain abscess, renal infarction. CT revealed renal infarction and a suspicion of thrombosis in the extracardiac conduit, but ICE revealed normal in the conduit.

Case2: A 22-year-old male with single ventricle presented cyanosis with systemic saturation of 86%. He underwent lateral tunnel TCPC at 1 years of age. ICE revealed a leakage from the margin of the conduit. However the device closure of the leakage seemed to be difficult. He was followed with anticoagulation of warfarin.

Case3: A 9-year-old female with common canal was diagnosed with infective endocarditis followed by dental therapy. CT showed a defect within the conduit, suspicious of infective thrombus. We performed biopsy which guided by ICE. Histopathological finding showed white thrombus without any pathogen. We performed curettage of thrombus.

Conclusions: ICE is a feasible imaging modality for real-time dynamic observation of the conduit after Fontan operation.

OJ3-2

Fontan循環における門脈血流の減少

A decrease in portal venous flow in the Fontan circulation

杉谷 雄一郎¹⁾, 宗内 淳¹⁾, 藤井 俊輔¹⁾, 松岡 良平¹⁾, 川口 直樹¹⁾, 渡邊 まみ江¹⁾, 安東 勇介²⁾, 落合 由恵²⁾
1) 地域医療機能推進機構九州病院 小児科, 2) 地域医療機能推進機構九州病院 心臓血管外科

Yuichirou Sugitani¹⁾, Jun Muneuchi¹⁾, Shunsuke Fujii¹⁾, Ryohei Matsuoka¹⁾, Naoki Kawaguchi¹⁾, Mamie Watanabe¹⁾,
Yusuke Ando²⁾, Yosie Ochiai²⁾

1) Department of Pediatrics, Japan Community Healthcare Organization Kyushu Hospital,

2) Department of CVS, Japan Community Healthcare Organization Kyushu Hospital

Objective: The precise pathophysiology of Fontan associated liver disease (FALD) remains unknown. We aim to study the relationship between FALD and portal circulation evaluated by magnetic resonance imaging (MRI).

Methods: We performed MRI in 24 subjects (13 subjects after Fontan operation and 11 subjects with biventricular circulation). We measured cardiac index and portal venous flow (PVF) corrected by BSA, and compared these parameters between subjects after Fontan operation and with biventricular circulation.

Results: Age at MRI was 10.3 (4.6-34.5) years. Cardiac index and PVF index was 3.1 (1.2-4.9) and 0.32 (0.14-0.82) L/min/m², respectively. PVF index was significantly reduced in Fontan subjects compared with that in biventricular subjects [0.23 (0.14-0.51) vs 0.35 (0.22-0.82), p=0.04], whereas there was no difference in cardiac index between the groups [2.8 (1.2-4.9) vs 3.3 (2.4-4.3)].

Conclusions: A decrease in PVF is a unique finding in Fontan subjects.

OJ3-3 成人Fontan患者における下垂体機能評価

Pituitary research on Fontan patients

長友 雄作¹⁾, 永田 弾¹⁾, 坂本 一郎²⁾, 江口 祥美¹⁾, 村岡 衛¹⁾, 福岡 将治¹⁾, 鶴池 清¹⁾, 平田 悠一郎¹⁾, 大賀 正一¹⁾

1) 九州大学病院 小児科, 2) 九州大学病院 循環器内科

Yusaku Nagatomo¹⁾, Hazumu Nagata¹⁾, Ichiro Sakamoto²⁾, Yoshimi Eguchi¹⁾, Mamoru Muraoka¹⁾, Shoji Fukuoka¹⁾, Kiyoshi Uike¹⁾, Yuichiro Hirata¹⁾, Shouichi Ohga¹⁾

1) Department of Pediatrics, Kyushu University Hospital, 2) Department of Cardiology, Kyushu University Hospital

Objective: The purpose of this study is to research the pituitary function in Fontan patients.

Methods: We measured each pituitary-associated hormone (thyroid, adrenal, gonad and growth) at outpatient clinic in adult Fontan patients.

Results: 15 males and 10 females were included in the patients. Clinical data in their last examination demonstrated that SpO₂ 94.4 ± 4.0%, CVP 10.4 ± 2.9 mmHg and CI 3.0 ± 1.6 L/min/m². DHEA-S was as low as 131 ± 54 (standard 159-538) in males and 72 ± 41 (standard 92-399) in females. IGF-1 was 134 ± 32, including 15 patients who demonstrated lower than their age standard. fT₄ 1.5 ± 0.2 ng/dl, TSH 2.6 ± 1.5, ACTH 37 ± 20, Cortisol 7.1 ± 2.8, GH 1.3 ± 2.9 (male), 0.3 ± 0.5 (female), LH 3.4 ± 1.8 (male), 8.1 ± 6.3 (female), FSH 5.3 ± 2.5 (male), 10.0 ± 3.3 (female), PRL 10.4 ± 5.3 (male), 12.5 ± 7.7 (female), all of which were within normal range.

Conclusion: Although to assess the pituitary function accurately is too difficult due to its diurnal variation or interaction with lower organs, high CVP may be associated with pituitary function in Fontan patients. Further examination such as each hormone loading test is required.

OJ3-4 フォンタン術後患者の体静脈側副血行路に対する塞栓術の治療効果

Effects of Embolization for Systemic Venous Collaterals in Fontan Patients

三池 虹¹⁾, 大内 秀雄^{1,2)}, 中島 公子¹⁾, 鈴木 大¹⁾, 根岸 潤¹⁾, 岩朝 徹¹⁾, 坂口 平馬¹⁾, 白石 公¹⁾, 黒寄 健一¹⁾

1) 国立循環器病研究センター 小児循環器科, 2) 国立循環器病研究センター 成人先天性心疾患科

Hikari Miike¹⁾, Hideo Ohuchi^{1,2)}, Kimiko Nakajima¹⁾, Dai Suzuki¹⁾, Jun Negishi¹⁾, Tohru Iwasa¹⁾, Heima Sakaguchi¹⁾, Isao Shiraishi¹⁾, Kenichi Kurosaki¹⁾

1) Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center,

2) Department of Adult Congenital Heart Disease, National Cerebral and Cardiovascular Center

Background: Development of systemic to pulmonary venous collaterals (VVC) may progress with arterial oxygen desaturation in patients after Fontan procedure. Catheter embolization (CE) is a standard treatment for the collaterals, but there have being few studies assessing the clinical effects.

Purpose: To assess clinical benefit of CE to prevent arterial desaturation by cardiopulmonary exercise testing (CPX) before and after CE.

Methods and Result: We performed CE for VVC and PAVF in 16 Fontan patients and compared the results of peripheral oxygen saturation (SPO₂) and exercise tolerance based on CPX before and after CE. Rest SPO₂ increased 1.9 ± 0.6% (p < 0.01) and lowest exercise-induced SPO₂ increased 3.6 ± 1.0% (p=0.002) after CE. Although the exercise-induced decline of SPO₂ decreased 1.8% after the CE, the decline didn't reach statistical significance. SPO₂ variations didn't depend on the site of VVC and there was no significant change in peak VO₂.

Conclusions: CE for VVC has short-term clinical benefit of both preventing exercise-induced hypoxia and raising resting SPO₂ in Fontan patients. However, the CE-related benefit to exercise capacity needs further study.

OJ3-5 フォンタン術後遠隔期の腎機能低下と蛋白尿に関する研究

Renal dysfunction and proteinuria in patients with Fontan operation

村岡 衛¹⁾, 永田 弾¹⁾, 坂本 一郎²⁾, 江口 祥美¹⁾, 福岡 将治¹⁾, 鶴池 清¹⁾, 長友 雄作¹⁾, 平田 悠一郎¹⁾, 石北 綾子²⁾, 筒井 裕之²⁾, 大賀 正一¹⁾

1) 九州大学病院 小児科, 2) 九州大学病院 循環器内科

Mamoru Muraoka¹⁾, Hazumu Nagata¹⁾, Ichiro Sakamoto²⁾, Yoshimi Eguchi¹⁾, Shoji Fukuoka¹⁾, Kiyoshi Uike¹⁾, Yusaku Nagatomo¹⁾, Yuichiro Hirata¹⁾, Ayako Ishikita²⁾, Hiroyuki Tsutsui²⁾, Shouichi Ohga¹⁾

1) Department of Pediatrics, Kyushu University Hospital, 2) Department of Cardiovascular medicine, Kyushu University Hospital

Background: Fontan patients with high venous pressure and low cardiac output are at high risk of kidney injury.

Methods: Patients were recruited from Kyushu University Hospital between 2009 and 2018. 185 Fontan patients (97 males) were identified. Demographic, cardiac and kidney data were collected.

Results: The median age of patients was 21 (18-46) years. Fontan operations were performed at the median age of 57 (8-292) months, and the median postoperative period was 16 (4-36) years. Fontan types included atriopulmonary connection (n=20, 10.8%), lateral tunnel (n=54, 29.2%) and extracardiac conduit (n=111, 60%). 33 patients (18.2%) had renal dysfunction (eGFR < 90 mL/min/1.73m²) and 43 patients (26.1%) had proteinuria. Patients with renal dysfunction were older at Fontan procedure (72 vs 57 months, p=0.039), showed lower postoperative cardiac index (2.4 vs 2.8 L/min/m², p=0.0082) and longer postoperative period (18 vs 16 years, p=0.0072). Patients with proteinuria were also older at procedure (87 vs 56 months, p < 0.001), with higher venous pressure (11.5 vs 9 mmHg, p=0.0012).

Conclusions: Timing of Fontan operation may influence on the long-term risk of kidney injury.

OJ3-6 フォンタン術後遠隔期血行動態予後における肺動脈サイズの影響：肺動脈最小径の重要性

Influence of Pulmonary Artery Size on the Late Hemodynamic Outcome After Fontan Operation: Importance of Minimal Diameter of Pulmonary Artery

田中 敏克¹⁾, 城戸 佐知子¹⁾, 林 賢¹⁾, 久保 慎吾¹⁾, 上村 和也¹⁾, 三木 康暢¹⁾, 松岡 道生¹⁾, 亀井 直哉¹⁾, 小川 禎治¹⁾, 富永 健太¹⁾, 大嶋 義博²⁾

1) 兵庫県立こども病院 循環器内科, 2) 兵庫県立こども病院 心臓血管外科

Toshikatsu Tanaka¹⁾, Sachiko Kido¹⁾, Ken Hayashi¹⁾, Shingo Kubo¹⁾, Kazuya Uemura¹⁾, Yasunobu Miki¹⁾, Michio Matsuoka¹⁾, Naoya Kamei¹⁾, Yasuharu Ogawa¹⁾, Kenta Tominaga¹⁾, Yoshihiro Oshima²⁾

1) Department of Cardiology, Kobe Children's Hospital, 2) Department of Cardiovascular Surgery, Kobe Children's Hospital

Background: There are few studies about the influence of PA size on the late outcome after Fontan operation. The aim of this study is to evaluate if the PA size affects the late hemodynamic outcomes in the Fontan population.

Methods: This study was conducted retrospectively on 49 patients over 18 years of age undergoing follow-up catheterization. Their median age was 22 years (range, 18 to 39 years). Data on PS index (PSI) (measuring the diameters of right and left PA at the narrowest portion of proximal to the origin of upper lobe branch, and sum of them, then dividing by BSA), PA index (PAI), cardiac index (CI), CVP were analyzed.

Results: The narrowest diameter of right and left PA was 13.4 mm (8.1 to 23.2mm), and 11.6mm (7.3 to 20.0mm). PSI was 17.3 ± 2.7mm/m², PAI was 228 ± 67mm²/m². PSI was correlated positively with CI (p < 0.05, R=0.30), however, there were no correlations with between PSI and CVP, between PAI and both CI and CVP. Conclusion: These results show the possibility that PSI is more useful clinical indicator of late outcome after Fontan operation than PAI, and minimal diameter of PA is an important factor for Fontan hemodynamics.

OJ4-1 肥大型心筋症と妊娠・出産

Maternal and Fetal Outcomes in Pregnancy Complicated with Hypertrophic Cardiomyopathy

桂木 真司¹⁾, 河村 卓弥¹⁾, 吉田 純¹⁾, 中尾 真大¹⁾, 池田 智明²⁾, 小野 良子¹⁾, 鈴木 僚¹⁾, 川端 伊久乃¹⁾, 高見澤 格¹⁾, 高梨 秀一郎¹⁾, 高山 守正¹⁾

1) 榊原記念病院 産婦人科, 2) 三重大学

Shinji Katsuragi¹⁾, Takuya Kawamura¹⁾, Atsushi Yoshida¹⁾, Masahiro Nakao¹⁾, Tomoaki Ikeda²⁾, Ryoko Ono¹⁾, Ryo Suzuki¹⁾, Ikuno Kawabata¹⁾, Itaru Takamizawa¹⁾, Syuichiro Takanasi¹⁾, Morimasa Takayama¹⁾

1) Department of Obstetrics and Gynecology, Sakakibara Heart Institute, 2) Mie University

Background: There is an increased risk of arrhythmia or cardiac failure in Hypertrophic Cardiomyopathy (HCM) patients with left ventricular outlet (LVOT) obstruction during pregnancy, and postpartum.

Methods and Results: The subjects were 15 HCM patients wishing for a baby who were managed at one institution. In 10 HOCM (hypertrophic obstructive cardiomyopathy) cases, beta blockade worked well in 2, otherwise we performed 6 myectomies and 1 PTSMA (percutaneous transluminal septal myocardial ablation). After SRT (septal reduction therapy) and beta blockade treatment, women with NYHA class II became class I in 7 cases out of 9, median [interquartile range] pressure gradient of LVOT changed from 85 [64-102] to 15 [10-20] mmHg, $p < 0.001$. 11 women became pregnant and delivered at 38 [37-38] gestational weeks. In 2 cases, atrial flutter, and non-sustained ventricular tachycardia occurred during pregnancy.

Conclusion: More care is needed for women with class II or more with obstruction. Medical and surgical treatment for SRT resolves the heart failure, but meticulous care is still necessary for safe pregnancy and delivery.

OJ4-2 成人先天性心疾患女性の妊娠・分娩期の産科合併症

Obstetric complication in the women with adult congenital heart disease

兵藤 博信, 竹田津 史野, 新田 慧, 藤野 佐保, 中里 紀彦, 齋藤 悦子, 布施 由紀子, 岩佐 加波, 若佐谷 敦, 彦坂 慈子, 船倉 翠, 井上 知子, 今田 信哉, 三浦 紫保, 砂川 空広, 笠松 高弘, 久具 宏司

東京都立墨東病院 産婦人科

Hironobu Hyodo, Fumino Taketazu, Satoshi Nitta, Saho Fujino, Norihiko Nakazato, Etsuko Saito, Yukiko Fuse, Kanami Iwasa, Atsushi Wakasaya, Chikako Hikosaka, Midori Funakura, Tomoko Inoue, Shinya Imada, Shiho Miura, Sorahiro Sunagawa, Takahiro Kasamatsu, Koji Kugu

Department of Obstetrics and Gynecology, Tokyo Metropolitan Bokutoh Hospital

Most of the pregnancy cases in the women with ACHD may be uneventful pregnancy if the primary cardiac condition is good. However, it may sometimes possibly affect obstetric complication.

29-year-old, primipara woman with repaired ASD delivered vaginally with forceps because of prolonged labor in 38th week. She subsequently suffered postpartum hemorrhage over 6 L and received massive infusion and transfusion. Although she fell into DIC and pulmonary edema, transfusion and diuretic recovered her within five days and the cardiac condition did not deteriorated.

27-year-old, primipara woman with VSD, who had had infective endocarditis two years before and had received subsequent surgical repair, developed preeclampsia with mild TR and AR. She delivered by c-section in 37th week because of failed induction. She also developed pulmonary edema and the condition recovered within three days by diuretic and antihypertensive.

Pulmonary edema may be developed in some obstetric condition by cardiogenic or non-cardiogenic reason. Ultrasonocardiography may be helpful for differential diagnosis. The treatment may be started with ordinary procedure if prior condition is good.

OJ4-3 ACHD 合併妊娠の分娩時における、分娩第2期短縮を目的とした吸引分娩の有用性の検討 The evaluation of operative delivery for the purpose of shortening 2nd stage of labor in pregnancy with adult congenital heart disease

中西 篤史, 神谷 千津子, 田路 明彦, 月永 理恵, 松坂 優, 水野 祐紀子, 澤田 雅美, 塩野入 規, 小西 妙, 堀内 縁, 釣谷 充弘, 岩永 直子, 吉松 淳
国立循環器病研究センター 周産期婦人科

Atsushi Nakanishi, Chizuko Kamiya, Akihiko Touji, Rie Tsukinaga, Yuu Matsuzaka, Yukiko Mizuno, Masami Sawada, Tadashi Shionoiri, Tae Konishi, Chinami Horiuchi, Mitsuhiko Tsuritani, Naoko Iwanaga, Jun Yoshimatsu
Department of Perinatology and Gynecology, National Cerebral and Cardiovascular Center

Background: Operative delivery is thought to mitigate cardiac load through shortening of the second stage of labor; however, the effect on ACHD pregnancies is not clarified. The aim of this study is to evaluate whether operative delivery shortens the 2nd stage of labor and mitigates cardiac load in ACHD pregnancies.

Methods: The subjects were pregnant women with ACHD who gave their first vaginal birth to a singleton. We evaluated the duration of the 2nd stage of labor, BNP, blood loss and neonatal injuries.

Results: 6 cases of vaginal delivery and 14 cases of operative delivery were identified. The duration of the 2nd stages of labor (min.) were not significantly different (46 (25-54) vs 62 (22-71), $p=0.87$). The post-partum BNP (pg/ml) of the operative deliveries (89.9 (49.4-128.8)) was higher than the vaginal deliveries (59.3 (44.1-61.2)), but the difference was not significant ($p=0.25$). The blood loss of operative deliveries was similar to vaginal deliveries. No neonatal injuries were observed.

Conclusions: In the study, we could not demonstrate that operative delivery shortens the 2nd stage of labor and mitigates cardiac load when compared with vaginal delivery.

OJ4-4 成人先天性心疾患チームの立ち上げ6か月の現状 Developing multidisciplinary team for patients with adult congenital heart disease

圓尾 文子¹⁾, 白井 丈晶²⁾, 佐藤 有美³⁾, 山本 真由子¹⁾, 角谷 誠²⁾, 坂本 敏仁¹⁾, 脇山 英丘¹⁾, 大保 英文¹⁾, 山口 眞弘¹⁾

1) 加古川中央市民病院 心臓血管外科, 2) 加古川中央市民病院 循環器内科, 3) 加古川中央市民病院 小児科

Ayako Maruo¹⁾, Takeaki Shirai²⁾, Yumi Sato³⁾, Mayuko Yamamoto¹⁾, Makoto Kadotani²⁾, Toshihito Sakamoto¹⁾, Hidetaka Wakiyama¹⁾, Hidefumi Obo¹⁾, Masahiro Yamaguchi¹⁾

1) Department of Cardiovascular Surgery, Kakogawa Central City Hospital,

2) Department of Cardiology, Kakogawa Central City Hospital, 3) Department of Pediatrics, Kakogawa Central City Hospital

Our hospital opened in August 2016 uniting two municipal hospitals. Department of the pediatric cardiology launched in 2015 prior to the practice and surgery for congenital heart disease started at the bran-new hospital. Since then, the number of the patients with adult congenital heart disease (ACHD) has been increasing. In April 2018 we developed the multidisciplinary team to offer comprehensive care. Core member consist of cardiologists, surgeon, nurses, physical therapist, echocardiography technician, and medical accounting clerk. At the beginning, the kickoff meeting was held to clarify the purpose of the team and 137 staffs attended to it. Regular team meeting is opened twice a month. Lecture of topics in ACHD and case presentation of patients requiring discussion and involvement of the team are given alternately. Number of outpatients with ACHD is 270. Nine patients underwent surgery at our hospital including 2 patients who had resisted re-do surgery for long time after interference of the team. All patients were offered cardiac rehabilitation. Process of team development and subjects in the future of our ACHD team will be discussed.

OJ4-5

静岡県立病院機構内二病院でのACHD共同手術体制の現状

The results of the joint operations in Shizuoka prefectural hospitals

廣瀬 圭一¹⁾, 猪飼 秋夫¹⁾, 長門 久雄¹⁾, 村田 真哉¹⁾, 今井 健太¹⁾, 菅野 勝義¹⁾, 石道 基典¹⁾, 太田 恵介¹⁾, 植木 力²⁾, 山中 憲²⁾, 佐藤 博文²⁾, 平野 雅大²⁾, 恒吉 裕史²⁾, 坂本 喜三郎¹⁾

1) 静岡県立こども病院 心臓血管外科, 2) 静岡県立総合病院

Keiichi Hirose¹⁾, Akio Ikai¹⁾, Hisao Nagato¹⁾, Masaya Murata¹⁾, Kenta Imai¹⁾, Kazuyoshi Kannno¹⁾, Motonori Ishido¹⁾, Keisuke Ota¹⁾, Chikara Ueki²⁾, Ken Yamanaka²⁾, Hirofumi Sato²⁾, Masahiro Hirano²⁾, Hiroshi Tuneyoshi²⁾, Kisaburo Sakamoto¹⁾

1) Department of Cardiovascular Surgery, Mt. Fuji Shizuoka Children's Hospital,

2) Department of Cardiovascular Surgery, Shizuoka General Hospital

background: As for the medical management including surgery for the patients with ACHD, it may be difficult for most hospitals other than that has both adult/congenital cardiologists/cardiovascular surgeons. Between Shizuoka general hospital and Mt.Fuji Shizuoka Children's hospital, medical stuff and information have shared for years. And joint cardiovascular surgeries have started since 2015 autumn at Shizuoka general hospital.

purpose: We examined the contents and the results of the joint operations.

patients and methods: 12 joint operations were performed and median age at operation was 52.5 years old (Male3, Female9). The original diagnosis were TOF/PAVSD6, VSD ± PS3, AVSD/DORV/Ebstein 1 (each). The procedures were PVR/RVOTR7, MVP/TAP4, Bentall2, VSD closure2, etc. (included multiple choice).

results: There was no early mortality. One late mortality was occurred 11 months after the surgery due to AML. General conditions in other patients have been feasible and most of them were followed in Shizuoka general hospital.

conclusions: The results of the joint operations were feasible in the present study. Our joint project may become more important in the future.

OJ4-6

当院におけるACHD患者移行における現状と諸問題

Transition problems of ACHD patients: A single center experience for the past 5 years

石田 秀和¹⁾, 成田 淳¹⁾, 石井 良¹⁾, 水流 宏文¹⁾, 石垣 俊¹⁾, 橋本 和久¹⁾, 木村 幸嗣¹⁾, 吉原 千華¹⁾, 木戸 高志³⁾, 平 将生³⁾, 上野 高義³⁾, 澤 芳樹³⁾, 塚本 泰正²⁾, 坂田 泰史²⁾, 小垣 滋豊^{1,4)}, 大藪 恵一¹⁾

1) 大阪大学大学院医学系研究科 小児科学, 2) 大阪大学大学院医学系研究科 循環器内科学,

3) 大阪大学大学院医学系研究科 心臓血管外科, 4) 大阪急性期・総合医療センター 小児科

Hidekazu Ishida¹⁾, Jun Narita¹⁾, Ryo Ishii¹⁾, Hirofumi Tsuru¹⁾, Suguru Ishigaki¹⁾, Kazuhisa Hashimoto¹⁾, Yukitsugu Kimura¹⁾, Chika Yoshihara¹⁾, Takashi Kido³⁾, Masaki Taira³⁾, Takayoshi Ueno³⁾, Yoshiki Sawa³⁾, Yasumasa Tsukamoto²⁾, Yasushi Sakata²⁾, Shigetoyo Kogaki^{1,4)}, Keiichi Ozono¹⁾

1) Department of Pediatrics, Osaka University Graduate School of Medicine,

2) Department of Cardiology, Osaka University Graduate School of Medicine,

3) Department of Cardiovascular Surgery, Osaka University Graduate School of Med,

4) Department of Pediatrics, Osaka General Medical Center

Since many adult congenital heart disease (ACHD) patients have various systemic complications, they sometimes hard to move from pediatric outpatient clinic to ACHD clinic. In this study, we analyzed the real situation of ACHD transition for the past 5 years in our hospital. From 2012 to 2017, 330 ACHD patients (above 20 years old) visited the follow-up clinic of our pediatric cardiology unit. 240 cases were still followed-up. Among them, 142 cases were successfully transferred to ACHD clinic (transition group), whereas 63 patients were still followed-up by the pediatric cardiologist (non-transition group). Other 35 cases were followed-up only by post-surgery clinic. In the transition group, 18% of the patients had systemic complications, such as chromosomal abnormality, hypoxic encephalopathy, mental retardation, or other organ dysfunction. However in the non-transition group, 71% of patients had such complications. The multivariate analysis demonstrated that chromosomal abnormality, mental retardation, and pulmonary hypertension were the significant risk factors for transition. In contrast, single ventricle physiology did not affect the transition in our hospital.

OJ5-1

フォロー四徴症心内修復術後患者に対する下肢陽圧負荷装置を用いた前負荷予備能の検討

Non-invasive assessment of preload reserve using the leg-positive pressure maneuver in patients with repaired tetralogy of Fallot

須藤 麻貴子¹⁾, 松本 賢亮¹⁾, 鈴木 麻希子¹⁾, 城戸 佐知子²⁾, 平田 健一¹⁾

1) 神戸大学医学部 循環器内科, 2) 兵庫県立こども病院 小児科

Makiko Suto¹⁾, Kensuke Matsumoto¹⁾, Makiko Suzuki¹⁾, Sachiko Kido²⁾, Ken-ichi Hirata¹⁾

1) Departments of Cardiovascular Medicine, Kobe University, 2) Pediatrics, Kobe Children's Hospital

Introduction: In the patients with repair of TOF, early detection of RV dysfunction and lack of hemodynamic reserve is important.

Purpose: The purpose was to investigate the preload reserve and reveal its determinant.

Methods: Forty patients with repaired TOF and 30 normal controls were recruited. Echocardiographic parameters were obtained both at rest and during leg-positive pressure (LPP) stress.

Results: LV ejection fraction was slightly decreased and RV functional parameters were significantly impaired in the patients and no difference was observed in forward SV between two groups at baseline. The LPP stress significantly increased forward SV from 73 ± 14 to 83 ± 15 mL/m² in normal controls, but increase in forward SV was quite impaired (from 77 ± 20 to 80 ± 20 mL/m²) in the patients. Multiple logistic regression analysis revealed that more than moderate pulmonary regurgitation was an independent determinant factor for the lack of preload reserve.

Conclusions: Even in the patients with repaired TOF, preload reserve was significantly impaired. The presence of significant pulmonary regurgitation was an important determinant of blunted preload reserve in these patients.

OJ5-2

Transcatheter PVR時代を前にしたSurgical PVR

Surgical Pulmonary Valve Replacement in Consideration of Future Transcatheter Valve-in-Valve Implantation

樽井 俊, 宮原 義典, 長岡 孝太, 山口 英貴, 清水 武, 伊吹 圭二郎, 柿本 久子, 籾 義仁, 藤井 隆成,

石野 幸三, 佐野 俊二, 富田 英

昭和大学病院 小児循環器・成人先天性心疾患センター

Suguru Tarui, Yoshinori Miyahara, Kouta Nagaoka, Hideki Yamaguchi, Takeshi Shimizu, Keijiro Ibuki,

Hisako Kakimoto, Yoshihito Hata, Takanari Fujii, Kozo Ishino, Shunji Sano, Hideshi Tomita

Pediatric Heart Disease and Adult Congenital Heart Disease Center, Showa University Hospital

Recently, transcatheter valve-in-valve (VIV) implantation within dysfunctional bioprosthetic valves (BPV) has emerged as an alternative to redo surgery. We performed surgical pulmonary valve replacement (SPVR) in three cases considering future VIV implantation.

During SPVR, we used as large size of BPV as possible, to ensure inner diameter for VIV implantation. The BPV was positioned proximally to the native annulus and was slightly tilted posteriorly. Gore-Tex was used as trans-annular patch. An 27mm-BPV for 67-year-old male with BSA of 1.74m², and 23mm-BPVs for 21- and 41-year-old females with BSA of 1.39m² and 1.43m², respectively, were used. Although rather small BPVs were used in two cases due to limited thoracic spaces, cardiologists affirmed at least 18mm-Melody or 20mm-SAPIEN valves could be implanted into 23mm-BPVs, those of which were acceptable for their BSA. Also, even larger valves might be implanted by the most recent technique of intentional fracture of BPV frame.

When SPVR is performed, it is crucial that surgeons discuss with experienced cardiologists to determine appropriate BPV size and position. We report our experience along with some literature review.

OJ5-3 血清プレセプシン値は成人先天性心疾患において病態と予後を反映する**High Plasma Presepsin Level Predicts Morbidity and Mortality in Adults with Congenital Heart Disease.**

豊島 由佳, 大内 秀雄, 羽山 陽介, 三池 虹, 鈴木 大, 根岸 潤, 岩朝 徹, 坂口 平馬, 白石 公, 黒寄 健一
国立循環器病研究センター 小児循環器科

Yuka Toyoshima, Hideo Ohuchi, Yohsuke Hayama, Hikari Miike, Dai Suzuki, Jun Negishi, Tohru Iwasa,
Heima Sakaguchi, Isao Shiraiishi, Ken-ichi Kurosaki
Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center

Background: Presepsin is a fragment of CD14 released in the process of phagocytosis of bacteria and reflects inflammatory state due to gut bacterial translocation.

Purpose: To measure plasma levels of Presepsin (PSEP, pg/mL) and clarify the possible association of right-sided heart failure (HF) pathophysiology and their prognosis in adults with congenital heart disease (ACHD).

Methods: We prospectively measured PSEP in 134 consecutive ACHD patients. We compared the PSEP with their clinical profiles, hemodynamics, and prognosis.

Results: The median PSEP was 294. On multivariate analysis, high central venous pressure, high cardiac index and Fontan circulation were independently associated with the high PSEP ($p < 0.05$). High PSEP was also associated with the high incidence of unexpected hospitalization (USH), including death (HR 1.05 per 100, 95%CI : 1.03-1.08, $p=0.0006$). The receiver-operator characteristic analysis revealed the cut-off value of 513 and the patients with high PSEP (≥ 513) have 3.3 times higher rate of USH than those with low PSEP.

Conclusions: High PSEP reflects right-sided HF pathophysiology and predicts morbidity and mortality in ACHD patients.

OJ5-4 成人先天性心疾患患者の予後予測におけるMELD-XI Scoreの有用性**Prognostic Utility of Model for End-Stage Liver Disease Excluding INR (MELD-XI) Score in Patients with Adult Congenital Heart Disease**

紺野 亮¹⁾, 建部 俊介¹⁾, 杉村 宏一郎¹⁾, 佐藤 公雄¹⁾, 青木 竜男¹⁾, 三浦 正暢¹⁾, 鈴木 秀明¹⁾, 山本 沙織¹⁾,
佐藤 遥¹⁾, 照井 洋輔¹⁾, 安達 理²⁾, 木村 正人³⁾, 齋木 佳克²⁾, 下川 宏明¹⁾

1) 東北大学 循環器内科, 2) 東北大学 心臓血管外科, 3) 東北大学 小児科

Ryo Konno¹⁾, Shunsuke Tatebe¹⁾, Koichiro Sugimura¹⁾, Kimio Satoh¹⁾, Tatsuo Aoki¹⁾, Masanobu Miura¹⁾,
Hideaki Suzuki¹⁾, Saori Yamamoto¹⁾, Haruka Sato¹⁾, Yosuke Terui¹⁾, Osamu Adachi²⁾, Masato Kimura³⁾,
Yoshikatsu Saiki²⁾, Hiroaki Shimokaw¹⁾

1) Department of Cardiovascular Medicine, Tohoku University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Tohoku University Graduate School of Medic,

3) Department of Pediatrics, Tohoku University Graduate School of Medicine

Background: The Model for End-Stage Liver Disease eXcluding INR (MELD-XI) score has been reported to predict outcomes in heart failure (HF) patients. Thus, we examined the prognostic value of MELD-XI score in ACHD patients.

Methods and Results: We retrospectively examined 639 ACHD patients (median age, 31 years) who visited Tohoku University hospital from 1995 to 2015. MELD-XI score was calculated as follows; $11.76 \times \ln(\text{creatinine}) + 5.11 \times \ln(\text{total bilirubin}) + 9.44$. The composite endpoint (CE) was defined as cardiovascular death, HF hospitalization, and lethal ventricular arrhythmias. In Kaplan-Meier analysis, the high MELD-XI group (> 10.4) had significantly worse event-free survival compared with the low MELD-XI group (≤ 10.4) (log-rank, $P < 0.001$). Univariable Cox regression analysis showed that MELD-XI score was significantly associated with the CE (HR 1.48, $P < 0.001$). In multivariable analysis, MELD-XI score remained a significant predictor of the CE (HR 1.47, $P < 0.001$).

Conclusions: These results indicate that MELD-XI score is useful to predict late cardiovascular outcomes in ACHD patients. ACHD patients with high MELD-XI score need to be closely followed.

OJ5-5

成人期に到達した修正大血管転位の長期予後についての検討

The natural and unnatural course of adult survivors of congenitally corrected transposition of the great arteries

横濱 ふみ, 杜 徳尚, 赤木 貞治, 伊藤 浩
岡山大学医学部 循環器内科

Fumi Yokohama, Norihisa Toh, Teiji Akagi, Hiroshi Ito
Department of Cardiology, Okayama University

Background: There are limited data on the long-term clinical outcomes and complications in adults with congenitally corrected transposition of the great arteries (ccTGA) in Japan. We sought to evaluate long-term trends in morbidity and mortality in adults with ccTGA.

Methods: Nineteen adults with ccTGA age over 18 years were identified from our database. Surgical history, clinical, and outcome data were analyzed. Unfavorable cardiovascular events included death, heart failure, arrhythmia, and surgical intervention.

Results: The median age at latest follow-up was 47 years. During the observation period, one patient died suddenly at the age of 67, 11 patients had heart failure (HF), 5 had atrioventricular block (AVB), 4 had atrial tachyarrhythmia, 9 had surgical intervention. At 50 years of follow-up, freedom from HF was 0.52, freedom from AVB was 0.74, freedom from atrial tachyarrhythmia was 0.84, freedom from surgical intervention was 0.54.

Conclusions: Although ccTGA patients who survive to adulthood have low mortality up to 50 years old, they are exposed to the risk of heart failure, AVB, and atrial tachyarrhythmia.

OJ5-6

成人先天性心疾患における感染性心内膜炎の傾向、リスクファクター、合併症について

Trend, risk factors and complications of infective endocarditis in adult congenital heart disease

朝貝 省史, 佐藤 正規, 原田 元, 島田 衣里子, 石戸 美妃子, 篠原 徳子, 稲井 慶, 富松 宏文, 杉山 央
東京女子医科大学 循環器小児科

Seiji Asagai, Masaki Satoh, Gen Harada, Eriko Shimada, Mikiko Ishido, Tokuko Shinohara, Kei Inai,
Hirofumi Tomimatsu, Hisashi Sugiyama
Department of Pediatric Cardiology, Tokyo Women's Medical University

Background: Although a high number of patients with congenital heart disease (CHD) reached adulthood, adult CHD remain a potential lifelong risk factor for infective endocarditis (IE).

Methods and Results: Between 2003 and 2018, 46 cases (40 patients) of IE were observed. Age ranged from 15 to 56 (median 34.5 years old). There were 33 cases with a history of surgery and Rastelli procedure was most common with 19. Streptococcal species were found in 32 (70%), Staphylococcal species were in 8 (17%). Complications were: intracranial complication 3 (7%), embolism of pulmonary arteries 16 (35%) and presence of heart failure 7 (15%). 9 (20%) cases required surgical intervention. Mortality were 6 (13%). The determinant factors significantly associated with the need for surgical intervention were detection of vegetation in aortic valve and RVOT, and presence of heart failure. The determinant factors significantly associated with mortality were Staphylococcal species, intracranial complication and presence of heart failure.

Conclusions: Even with medical progress, the complication and mortality rate remains high. Presence of heart failure contribute in need for surgical intervention and mortality.

OJ6-1

TCPC術後の非薬物的不整脈治療

Non-pharmacological hybrid therapy for arrhythmias after total cavo-pulmonary connection

豊原 啓子¹⁾, 竹内 大二¹⁾, 稲井 慶¹⁾, 篠原 徳子¹⁾, 杉山 央¹⁾, 庄田 守男²⁾

1) 東京女子医科大学 循環器小児科, 2) 東京女子医科大学 循環器内科

Keiko Toyohara¹⁾, Daiji Takeuchi¹⁾, Kei Inai¹⁾, Tokuko Shinohara¹⁾, Hisashi Sugiyama¹⁾, Morio Shoda²⁾

1) Department of Pediatric Cardiology, Tokyo Women's Medical University,

2) Department of Cardiology, Tokyo Women's Medical University

Backgrounds: Both bradycardia and tachycardia are commonly observed in patients after total cavo-pulmonary connection (TCPC) operation, because of complex cardiac conduction system and surgical procedure.

Epicardial leads placement with a sternotomy or thoracotomy has mostly been used to establish a permanent cardiac pacing therapy.

The tachycardia substrates usually exist in pulmonary venous atrium (PVA), therefore the access to PVA for catheter ablation (CA) is very difficult.

Results: Nine patients were enrolled.

Their age was nine to forty years old.

Four patients after TCPC conversion had sick sinus syndrome and four patients had AV block.

We performed catheter ablation in 7 patients. We could reach the PVA by transaortic and transseptal approach. We could eliminate all tachycardias, but 43% patients had recurrence.

All 9 patients underwent device implantation. All 9 patients underwent device implantation: DDD-pacemaker in 6, CRT-P in 2 and subcutaneous ICD in 1.

An endovascular implantation of a permanent atrial pacing lead was performed in one case.

Conclusion: It is important to perform non-pharmacological anti-arrhythmic hybrid therapy for TCPC patients.

OJ6-2

成人先天性心疾患患者において、native T1 と細胞外液量は左房容積と関連がある

Native T1 and extracellular volume are associated with larger left atrial volume in adult congenital heart disease patients

高橋 辰徳, 小田切 徹州, 安孫子 雅之, 鈴木 康太

山形大学医学部 小児科

Tatsunori Takahashi, Teshu Otagiri, Masayuki Abiko, Kouta Suzuki

Department of Pediatrics, Yamagata University Faculty of Medicine

Introduction: Native T1 and extracellular volume (ECV) derived from T1 mapping enabled to assess degree of myocardial fibrosis. However, data on T1 mapping in adult congenital heart disease (ACHD) patients are still insufficient.

Methods: We enrolled consecutive patients with ACHD from October 2017 to August 2018 and performed per protocol cardiac MRI. Native T1 and ECV of systemic ventricle were assessed.

Results: A total of 12 patients (Fontan (F): 4, cyanotic (C): 1 and biventricular heart with systemic left ventricle (B): 7) were enrolled. Two normal control (N) were also investigated. The median native T1 (msec) were 1079 (F), 1180 (C), 1018 (B) and 957 (N). The median ECV (%) were 30 (F), 37 (C), 29 (B) and 27 (N). Among 12 ACHD patients, native T1 and ECV positively correlated with BNP ($R^2 = 0.61$ and 0.55 , respectively), with indexed left atrial volume ($R^2 = 0.53$ and 0.47 , respectively), and were worse in patients with NYHA II/III (median: 1119 and 32, respectively) compared with NYHA I (median: 1018 and 27, respectively).

Conclusion: Myocardial fibrosis is associated with larger left atrial volume in ACHD patients. It may be due to diastolic dysfunction.

OJ6-3 成人先天性心疾患におけるサルコペニア

Sarcopenia in Adults with Congenital Heart Disease: Preliminary Study of Nutritional Status, Dietary Intake, and Resistance Training

椎名 由美, 松元 紀子, 岡村 大介, 木島 康文, 福田 旭伸, 川松 直人, 丹羽 公一郎
聖路加国際病院 循環器内科

Yumi Shiina, Noriko Matsumoto, Daisuke Okamura, Kijima Yasufumi, Terunobu Fukuda, Naoto Kawamatsu,
Koichiro Niwa
St.Luke's International Hospital

Aims: (1) to assess the nutritional status and dietary intake (2) compare the body composition and nutritional intake between sarcopenia and non-sarcopenia (3) evaluate the effects of resistance training and amino acid intake in adults with CHD.

Methods: Study 1: In 172 adults with CHD, the Food Frequency Questionnaire was used, and body composition analysis was conducted. Study 2: 30 of 172 adult patients with CHD were divided into two groups: amino acid intake plus resistance training (group A) and amino acid intake only (group B) for 2 months.

Results: Study 1: Skeletal muscle mass index was lower in adults with CHD compared to healthy Japanese. Calorie, protein, and fat intake in adults with CHD was higher than controls. Study 2: In group A, body fat percentage, oedema index, and NT-proBNP improved, and body weight, skeletal muscle mass index, and basic metabolism increased after the intervention. There was no improvement after intervention for group B.

Conclusions: Appropriate nutritional education and resistance training guidelines should be provided in CHD.

OJ6-4 三尖弁置換術後に肝不全の急性増悪を認めたエブスタイン病の一例

A case of Ebstein Disease with acute-on-chronic liver failure after tricuspid valve replacement

藤田 鉄平¹⁾, 小坂橋 俊美¹⁾, 石末 成哉¹⁾, 前川 恵美¹⁾, 郡山 恵子¹⁾, 福西 琢真²⁾, 齋木 宏文³⁾,
宮本 隆司²⁾, 先崎 秀明³⁾, 宮地 鑑²⁾, 阿古 潤哉¹⁾

1) 北里大学病院 循環器内科, 2) 北里大学 心臓血管外科, 3) 北里大学 小児科

Teppey Fujita¹⁾, Toshimi Koitabashi¹⁾, Naruya Ishizue¹⁾, Emi Maekawa¹⁾, Keiko Ryo Kooriyama¹⁾, Takuma Fukunishi²⁾,
Hirofumi Saiki³⁾, Takashi Miyamoto²⁾, Hideaki Senzaki³⁾, Kagami Miyazi²⁾, Jyunya Ako¹⁾

1) Department of Cardiovascular Medicine, Kitasato University Hospital,

2) Department of Cardiovascular Surgery, Kitasato University Hospital,

3) Medical Department of Pediatrics of Kitasato University Hospital

Background: Right heart failure causes liver dysfunction. Liver function is an important factor for perioperative management of cardiac surgery.

Case: A 73-year-old male with Ebstein disease. He had severe tricuspid regurgitation, alcoholic cirrhosis and chronic kidney disease. The Child-Pugh score and indocyanine green retention showed that liver function was preserved. However, there was an episode that hepatic encephalopathy developed when diuretic dose was increased. The surgical repair of the tricuspid valve was expected to improve right heart failure and liver dysfunction. Hence, he underwent tricuspid valve replacement. Right atrial pressure decreased to 8 mmHg and improved heart failure, but hypoalbuminemia and hepatic encephalopathy was progressed, and ascites developed. He was died due to liver failure on 80 days after the operation.

Conclusion: Prediction of postoperative liver function is difficult. The Child-Pugh score may be underestimated the risk of death in acute-on-chronic liver failure after surgery.

OJ6-5

小児—成人医療施設連携協定下での成人先天性心疾患のカテーテル治療

Catheter intervention for ACHD patients at children's Hospital under the collaborative medical contract with nearby adult medical center

安河内 聡¹⁾, 武井 黄太¹⁾, 瀧間 浄宏¹⁾, 岡村 達¹⁾, 沼田 隆佑¹⁾, 小山 智史¹⁾, 大日方 晴香¹⁾, 田中 登¹⁾, 米原 恒介¹⁾, 元木 博彦²⁾

1) 長野県立こども病院 循環器センター, 2) 信州大学成人先天性心疾患センター

Satoshi Yasukochi¹⁾, Kouta Takei¹⁾, Kiyohiro Takigiku¹⁾, Masaru Okamura¹⁾, Ryusuke Numata¹⁾, Satoshi Koyama¹⁾, Haruka Obinata¹⁾, Noboru Tanaka¹⁾, Kosuke Yonehara¹⁾, Hirohiko Motoki²⁾

1) Heart Center, Nagano Children's Hospital, 2) Adult Congenital Heart Disease Center, Shinshu University

The demand for catheter intervention (CI) for ACHD patients has been increasing even in local city, but not all ACHD care-given institute could provide the necessary CI because of no license to conduct the device closure. We report our practice under the collaborative medical contract (CMC) between the children's hospital (CH) who has the CI license and the adult facility who has not.

Of 30 ACHD pts (44y as a mean) underwent various CIs at CH after CMC in 2014, 19 pts (55y), referred from ACHD center, underwent percutaneous closure of atrial septal defect (ASD)(14), percutaneous closure of arterial duct: ADO (4), percutaneous closure of coronary arterial fistula: CAF (1). The rest 11 pts (25y) were follow-up pts at CH.

There was no complications. 3 had preprocedural ablation for atrial fibrillation at ACHD center before transferring to CH.

In conclusion, at local area with limited medical resources, the tight collaboration between pediatric and adult medical institution under CMC could be one of solution to provide the better medical practice besides transferring to the other institution outside of the area.

OJ7-1

ファロー四徴症再手術時の右室流出路再建法

Right ventricular outflow reconstruction in reoperation of Tetralogy of Fallot

打田 俊司¹⁾, 鎌田 真弓¹⁾, 浪口 謙治¹⁾, 康 利章¹⁾, 泉谷 裕則¹⁾, 宮田 豊寿²⁾, 渡部 竜助²⁾, 森谷 友造²⁾, 千阪 俊行²⁾, 太田 雅明²⁾, 高田 秀実²⁾, 赤澤 祐介³⁾, 檜垣 高史²⁾

1) 愛媛大学大学院医学系研究科 心臓血管・呼吸器外科, 2) 愛媛大学大学院医学系研究科 小児科,

3) 愛媛大学大学院医学系研究科 循環器内科

Shunji Uchida¹⁾, Mayumi Kamada¹⁾, Kenji Namiguchi¹⁾, Toshiaki Kan¹⁾, Hironori Izutani¹⁾, Toyohisa Miyata²⁾, Ryusuke Watanabe²⁾, Tomozou Moritani²⁾, Toshiyuki Chisaka²⁾, Masaaki Ohta²⁾, Hidemi Takada²⁾, Yusuke Akazawa³⁾, Takashi Higaki²⁾

1) Department of Cardiovascular and Thoracic Surgery, Ehime University, 2) Department of Pediatrics, Ehime University,

3) Department of Cardiology, Ehime University

Backgrounds: In many reoperation cases for TOF in ACHD, there is no operation record of previous surgery, the RVOTR procedure and applied materials are unknown and surgeon must have various surgical options for these situations. We review the transition of the RVOTR of 7 recent cases.

Cases: Between 2016 and 2018, seven adult cases (M:5, F:2) of reoperation for repaired TOF were investigated. Average age 45 y.o., BW 70.3 kg, BSA 1.78 m². PVR was performed in 6 pts and PV plasty was performed in 1. For PVR, CEP 25 mm: 3, CEP MAGNA EASE 25 mm: 2, 23 mm: 1 were applied. For RVOTR, PTFE graft, patch and Hemashield patch were used, but recently we selected Triplex graft for integrally reconstruction of RVOT to the PA. Operation was performed on beating in 4, under cardiac arrest in 3.

Conclusions: Reoperation for repaired TOF has various features depending on adhesion, tissue calcification, and leftward deviation of RVOT. Recently, in reoperation of TOF, we used CEP MAGNA EASE for PVR, and Triplex graft for integrally reconstruction from the RVOT to the PA. Furthermore, this material has certain advantages of good manipulability and hemostasis.

OJ7-2 成人期Ebstein病に対する外科的治療経験

Three case reports of patients experiencing Ebstein disease correction from NYHA I to NYHA IV

宮本 隆司¹⁾, 藤田 鉄平²⁾, 小坂橋 俊美²⁾, 福西 琢真¹⁾, 松井 謙太¹⁾, 中村 優飛¹⁾, 豊田 真寿¹⁾, 石堂 博敬²⁾, 栗田 聖子³⁾, 高梨 学³⁾, 齋木 宏文³⁾, 菅本 健司³⁾, 先崎 秀明³⁾, 阿古 潤哉²⁾, 宮地 鑑¹⁾

1) 北里大学医学部 心臓血管外科, 2) 北里大学医学部 循環器内科, 3) 北里大学医学部 小児科

Takashi Miyamoto¹⁾, Teppei Fujita²⁾, Toshimi Koitabashi²⁾, Takuma Fukunishi¹⁾, Kenta Matsui¹⁾, Yuuhi Nakamura¹⁾, Makoto Toyoda¹⁾, Hiroataka Ishidou²⁾, Seiko Kuwata³⁾, Manabu Takanashi³⁾, Hirofumi Saiki³⁾, Kenji Sugamoto³⁾, Hideaki Senzaki³⁾, Jyunya Ako²⁾, Kagami Miyaji¹⁾

1) Department of Cardiovascular Surgery, Kitasato University School of Medicine.,

2) Department of Cardiology, Kitasato University School of Medicine.,

3) Department of Pediatrics, Kitasato University School of Medicine.

In patients with Ebstein anomaly, severe tricuspid regurgitation significantly increases preload on the right ventricle. This anomaly has always been difficult to classify, because of a specific pathophysiology involving tricuspid valve regurgitation and a primary abnormality in the development of the right ventricle, frequently associated with a right-to-left shunt at atrial level. In early disease, the body attempts to maintain cardiac output of the right ventricle by dilating it and increasing cardiac contractility. In advanced disease, tricuspid regurgitation severely expands both the right atrium and right ventricle, ultimately resulting in right ventricle failure. These conditions are considered to be an operative indication for Ebstein disease in adulthood. Therefore, we reviewed 3 cases of surgical patients with Ebstein anomaly and right ventricular and biventricular dysfunction from NYHA I to NYHA IV. Surgical prognosis can be improved through aggressive preoperative treatment, vasoactive and antiarrhythmia medications, and comprehensive measures designed to reduce myocardial injury, prevent myocardial edema, and reduce pre- and afterload on the right ventricle.

OJ7-3 ファロー四徴症に対する肺動脈弁置換術後中期遠隔期の心室機能

Midterm Ventricular Function after Pulmonary Valve Replacement for Repaired Tetralogy of Fallot

小出 昌秋¹⁾, 國井 佳文¹⁾, 立石 実¹⁾, 奥木 聡志¹⁾, 櫻井 陽介¹⁾, 曹 宇晨¹⁾, 中嶋 八隅²⁾, 金子 幸栄²⁾, 井上 奈緒¹⁾, 齋藤 秀輝³⁾, 杉浦 亮³⁾, 森 善樹⁴⁾

1) 聖隷浜松病院 心臓血管外科, 2) 聖隷浜松病院 小児循環器科, 3) 聖隷浜松病院 循環器科, 4) 北里大学メディカルセンター 小児科

Masaaki Koide¹⁾, Yoshifumi Kunii¹⁾, Minoru Tateishi¹⁾, Satoshi Okugi¹⁾, Yosuke Sakurai¹⁾, Uchen Chao¹⁾,

Yasumi Nakashima²⁾, Sachie Kaneko²⁾, nao Inoue¹⁾, Hideki Saito³⁾, Ryo Sugiura³⁾, Yoshiki Mori⁴⁾

1) Department of Cardiovascular Surgery, Seirei Hamamatsu General Hospital,

2) Department of Pediatric Cardiology, Seirei Hamamatsu General Hospital,

3) Department of Cardiology, Seirei Hamamatsu General Hospital, 4) Department of Pediatrics, Kitasato University Medical Center

Background: Although early results of PVR after TOF repair have been described, information about midterm to late postoperative ventricular function is lacking. This study was designed to characterize right ventricular (RV) remodeling midterm after PVR.

Method: Retrospective analysis of MRI data from 2007 to 2017 in 17 patients (29 studies) who underwent PVR was done. Age at PVR was 33.2 ± 12.0 y and interval between TOF repair and PVR was 27.8 ± 8.5 y. Average observation period after PVR was 50.8 ± 38.8 months.

Results: No early death or late death. One reoperation was done for prosthetic valve endocarditis. Preoperative RVEDVI was 196.4 ± 51.7 ml/m² and postoperative RVEDVI (1yr / 3yrs) were 116.4 ± 28.6 / 119.4 ± 36.7 ml/m² which showed marked reduction at 1yr and no change thereafter. Preoperative RVEF was $42.7 \pm 4.5\%$ and postoperative RVEF (1yr / 3yrs) were 41.8 ± 8.0 / $44.2 \pm 12.6\%$ revealing no improvement or worsening. LVEDVI and LVEF showed no change 1yr and 3yrs after PVR.

Discussion: Although RVEF did not improve at midterm after PVR, RV volume reduction was preserved for more than three years. These data may support appropriate timing of PVR for repaired TOF.

OJ7-4

修正大血管転位症に対する成人期三尖弁手術5例の経験

Outcomes of tricuspid valve surgery in five adult patients with congenitally corrected transposition of the great arteries

櫻井 一¹⁾, 野中 利通¹⁾, 櫻井 寛久¹⁾, 杉浦 純也¹⁾, 大沢 拓哉¹⁾, 和田 侑星¹⁾, 大橋 直樹²⁾, 西川 浩²⁾, 吉田 修一郎²⁾, 加藤 温子²⁾, 森本 美仁²⁾, 吉井 公浩²⁾, 佐藤 純²⁾

1) JCHO中京病院 心臓血管外科, 2) JCHO中京病院 小児循環器科

Hajime Sakurai¹⁾, Toshimichi Nonaka¹⁾, Takahisa Sakurai¹⁾, Junya Sugiura¹⁾, Takuya Osawa¹⁾, Yuson Wada¹⁾, Naoki Ohashi²⁾, Hiroshi Nishikawa²⁾, Shuichiro Yoshida²⁾, Atsuko Kato²⁾, Yoshihito Morimoto²⁾, Kimihiro Yoshii²⁾, Jun Sato²⁾

1) Department of Cardiovascular Surgery, JCHO Chukyo Hospital, 2) Department of Pediatric Cardiology, JCHO Chukyo Hospital

Background: In adult patients with congenitally corrected transposition of the great arteries (ccTGA) and significant tricuspid regurgitation, anatomical repair is almost impossible, and tricuspid valve (TV) surgery is the only surgical option barring heart transplantation.

Methods and Results: We reviewed 5 adult patients (>18 years) with ccTGA, undergoing TV surgery between 1999 and 2018 at our institution. Two of the 5 patients had ventricular septal defects and pulmonary stenosis and had undergone conventional Rastelli procedures previously. Preoperative TV regurgitation was moderate to severe in all patients. One tricuspid plasty and 5 tricuspid valve replacements were performed for these 5 patients. Mean duration of follow-up was 8.9 ± 6.9 years. Age at surgery ranged between 19 to 61 years, with a mean age of 30 ± 18 years. There were no early deaths. However, one patient died 9 years after surgery because of arrhythmia. The postoperative right ventricular ejection fraction of all patients was less than 45%.

Conclusions: The early results of tricuspid surgery were almost satisfactory. However, postoperative right ventricular function was still unacceptable.

OJ7-5

成人先天性心疾患患者に対するBentall手術

Bentall's procedure for adults with congenital heart disease

前田 登史¹⁾, 藤原 慶一¹⁾, 加藤 おと姫¹⁾, 渡辺 謙太郎¹⁾, 植野 剛¹⁾, 吉澤 康祐¹⁾, 大野 暢久¹⁾, 稲熊 洸太郎²⁾, 豊田 直樹²⁾, 石原 温子²⁾, 坂崎 尚徳²⁾

1) 兵庫県立尼崎総合医療センター 心臓血管外科, 2) 兵庫県立尼崎総合医療センター 小児循環器内科

Toshi Maeda¹⁾, Keiichi Fujiwara¹⁾, Otohime Kato¹⁾, Kentaro Watanabe¹⁾, Go Ueno¹⁾, Kosuke Yoshizawa¹⁾, Nobuhisa Ohno¹⁾, Kotaro Inaguma²⁾, Naoki Toyota²⁾, Haruko Ishihara²⁾, Hisanori Sakazaki²⁾

1) Cardiovascular Surgery, Hyogo Prefectural Amagasaki General Medical Center,

2) Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical Center

Currently, the number of repaired/unrepaired ACHD is increasing. Aortic root dilatation may progress in a few patients.

Between 2008 and 2018, Bentall's procedures for aortic root dilatation were performed in 7 patients (age at operation ranged: 22.6 years ~ 44.6 years) with ACHD. Associated congenital heart disease included VSD: 3 (repaired: 2), TF (repaired): 1, PA+VSD (repaired): 1, MR (repaired): 1 and PDA (repaired): 1. 4 patients associated with chromosomal abnormalities: Marfan syndrome in 2, Loeys-Dietz syndrome in 1 and 22q11.2 deletion in 1. Handmade composite graft with mechanical valve was used in all patients. Coronary artery transfer was performed by Carrel patch technique in all. In 5 patients, concomitant procedures (VSD/ASD closure: 2, PVR: 1, RV-PA conduit exchange: 1, TVP: 1, CABG: 1) were performed. There was one operative death (PA+VSD: alveolar hemorrhage and RV failure). During follow-up periods, there was 1 reoperation (re-PVR for prosthetic endocarditis).

In Bentall's procedure, concomitant procedures were required in ACHD patients. Although they were complex in patients with conotruncal anomalies, these postoperative results are satisfactory.

OJ7-6

Lateral Tunnel TCPCに対するExtracardiac TCPC conversionの検討

Results of Extracardiac Total Cavopulmonary Connection Conversion for the adult patients performed Lateral Tunnel Total Cavopulmonary Connection

黒子 洋介¹⁾, 新井 禎彦¹⁾, 小谷 恭弘¹⁾, 杜 徳尚²⁾, 笠原 真悟¹⁾, 伊藤 浩²⁾

1) 岡山大学病院 心臓血管外科, 2) 岡山大学病院 循環器内科

Yosuke Kuroko¹⁾, Sadahiko Arai¹⁾, Yasuhiro Kotani¹⁾, Norihisa Toh²⁾, Shingo Kasahara¹⁾, Hiroshi Ito²⁾

1) Department of Cardiovascular Surgery, Okayama University Hospital,
2) Department of Cardiovascular Medicine, Okayama University Hospital

Background: There has been only limited number of reports regarding TCPC conversion (TCPCC) for lateral tunnel Fontan (LT) compared with Atriopulmonary connection Fontan (APC). We present our surgical outcome of TCPCC for LT.

Patients: Between January 1991 and June 2018, 23 patients over 18 years old underwent TCPC conversion in our hospital. Previous operations were 15 APC and 8 LT. In LT-TCPC conversion patients, Mean interval between LT to TCPCC was 217 months and mean symptomatic period to TCPCC was 45 months. Cause of TCPCC were 5 hypoxemia by baffle leak, 2 arrhythmia and 1 heart failure. All patient underwent extracardiac conduit TCPCC.

Result: There was 1 early mortality due to hypoxic encephalopathy. 2 patient underwent pacemaker implantation after TCPCC. All 7 patients who survived TCPCC showed significant improvement of causative symptom for TCPCC.

Conclusion: We achieved excellent improvement of symptom in TCPCC for LT. Contrary to other reports, we have not observed arrhythmia and lateral tunnel enlargement as major cause of TCPCC so far. Hypoxemia caused by baffle leak was leading cause of TCPCC in LT patient and well controlled by TCPCC.

OJ7-7

ファロー四徴症術後患者に対する肺動脈弁置換術の早期成績と問題点

Early outcome and problems after pulmonary valve replacement in patients with repaired tetralogy of Fallot

白石 修一, 杉本 愛, 高橋 昌, 土田 正則

新潟大学大学院医歯学総合研究科 呼吸循環外科学分野

Shuichi Shiraishi, Ai Sugimoto, Masashi Takahashi, Masanori Tsuchida

Division of Thoracic and Cardiovascular Surgery, Niigata University Medical School

OBJECTIVE: To evaluate early outcome of pulmonary valve replacement (PVR) in patients with repaired TOF.

METHODS: We retrospectively reviewed the records of 23 patients who underwent surgical PVR using bioprosthetic valve after TOF repair (median age 32, 11-68 years), followed up for 3 ± 2 years. Mean interval between TOF repair and PVR was 27 years.

RESULTS: There was no in-hospital and late death, no re-operation related to pulmonary valve. Serum BNP significantly decreased (95 ± 105 vs. 40 ± 36 pmol/L), CTR decreased (61.5 ± 8.2 vs. $55.7 \pm 6.5\%$), and QRS width shortened (184 ± 26 vs. 168 ± 19 ms). On the latest UCG, no patient had significant pulmonary insufficiency, peak velocity at the pulmonary valve was 1.9 ± 0.4 m/s. CMRI showed significant decrease of RVEDVI from 210 ± 59 to 119 ± 40 ml/m². Re-admission due to acute left heart failure occurred in one, though the symptom was disappeared after medication. Recurrent VT occurred after PVR in two patients who had previously undergone ICD implantation.

CONCLUSION: Early outcome of PVR after TOF repair was acceptable. RVEDVI, QRS width and BNP were improved. Careful follow-up for recurrent VT and left heart failure is necessary.

OJ7-8 ファロー四徴症に対するPVRにおける同時手術手技の検討

Concomitant procedures in pulmonary valve replacement for tetralogy of Fallot

加藤 おと姫¹⁾, 藤原 慶一¹⁾, 前田 登史¹⁾, 渡辺 謙太郎¹⁾, 植野 剛¹⁾, 吉澤 康祐¹⁾, 大野 暢久¹⁾, 稲熊 洸太郎²⁾, 豊田 直樹²⁾, 石原 温子²⁾, 坂崎 尚徳²⁾

1) 兵庫県立尼崎総合医療センター 心臓血管外科, 2) 兵庫県立尼崎総合医療センター 小児循環器内科

Otohime Kato¹⁾, Keiichi Fujiwara¹⁾, Toshi Maeda¹⁾, Kentaro Watanabe¹⁾, Go Ueno¹⁾, Kosuke Yoshizawa¹⁾, Nobuhisa Ohno¹⁾, Kotaro Inaguma²⁾, Naoki Toyoda²⁾, Haruko Ishihara²⁾, Hisanori Sakazaki²⁾

1) Department of Cardiovascular Surgery, Hyogo Prefectural Amagasaki General Medical Center,

2) Department of Pediatric Cardiology, Hyogo Prefectural Amagasaki General Medical

Background: Patients with repaired tetralogy of Fallot (rTF) with/without PA atresia often require pulmonary valve replacement (PVR) in adulthood. Some patients require other surgical interventions for sequela and age-related problems. I review concomitant procedures at PVR for rTF.

Object: I reviewed adult patients with rTF who underwent PVR from since 2004.

Results: PVR was performed on 22 patients (M:F=9:14), totaling 23 cases. The patients' ages were 18-68 (average 36.6) years. There were 27 concomitant procedures, including TAP/TVP (9), ablation (7), ASD closure (3), MVP (3), Bentall (2), MVR (1), Asc Ao replacement (1), and CABG (1) (duplicate inclusion). The average operation time, CPB time, and cardiac arrest time were 413/636, 126/228, 0/90 min, respectively (without concomitant procedures / with concomitant procedures). There were 2 early deaths (lung bleeding and MOF) and 1 late death (heart failure).

Conclusion: At ACHD operation, some concomitant procedures for adhesions, age-related and aortic lesions were required.

OJ8-1 CHDの認知度の低さに起因するライフステージの諸問題

Life stage problems caused by low awareness of CHD.

猪又 竜

先天性心疾患患者

Ryu Inomata

Patient of CHD (TGA3)

先天性心疾患患者はほとんどが成人を迎えられる時代になった。しかしながら、先天性心疾患は社会ではほとんど知られておらず、患者は就学、就職、結婚などのライフステージで様々な問題に向き合うことになる。

学校現場では、教員は先天性心疾患を含めた障害児対応を学んできていない場合が多く、実際に障害児を受け持つと過剰な対応をしてしまうことが多い。また、企業も採用拒否をすることもあり、見た目では障害があることがわからない先天性心疾患患者に対してどうサポートすれば良いのかわからない。

認知度が高ければこのような様々な問題は少なくなるはずなので、私は企業、学校、行政等へ先天性心疾患の啓発活動を行っている。啓発活動の中で見えてきた一般社会の反応を紹介する。

企業役員「雇用を考える中で、当社は障害を持つ方は無理だと考えておりましたが、もう一度可能な仕事はある事を気づかされました。かかわり方を考え直したいと思います。」

中学生「自分にも得意、不得意があるので、障害というのも意外と単純なものなのかなと思います。自分は今まで先入観で障害者を見てしまっていたので、困っていたら助けてあげるなど同じ仲間として接していきたいです。」

ダイバーシティや共生社会という言葉が多く使われるようになった今こそ先天性心疾患の啓発が必要である。

OJ8-2 先天性心疾患の子どものひとり立ちに向けた父親の思い**Fathers Story Preparing Children with Congenital Heart Disease for Transitions to Independent Living**北村 千章¹⁾, 野澤 祥子¹⁾, 西條 竜也²⁾

1) 新潟県立看護大学, 2) 飯山赤十字病院

Kitamura Chiaki¹⁾, Shoko Nozawa¹⁾, Tatsuya Nishijo²⁾

1) Niigata College of Nursing, 2) Red Cross Iiyama Hospital

先天性心疾患の子どもの親に関する先行研究の対象は母親の場合が多く、父親の視点は明らかになっていない。先天性心疾患のある子どもと家族は、子どもの成長過程における母子・家族関係の問題が指摘されている。本研究では、父親がどのように子どもの自立に向けて取り組んでいるかを明らかにするため、全国の中でも先進的に成人移行期支援に取り組んでいる施設を有している、A県心臓病の子どもを守る会の支部に所属している父親を対象とした。

分析の結果、父親の思いとは 1. 可能性がある限り子どもが生きられる方法を選びたい 2. 子どものわずかな可能性をいつも信じたい 3. 子どもにはいつも他者に助けてほしいと言えるようになってほしい 4. 子どもには自分で病気のことを説明できるようになってほしい 5. 子どもが自分で医師に相談できるようになってほしい 6. 子どもにはできないことよりもできることを見つけてほしい 7. 社会の中で理不尽なことがあることも体験もした方がよい 8. 一度は親から離れて生活してほしい、の8つのカテゴリーに分類された。

成人先天性心疾患患者は、進学や就業、結婚といった社会的自立の程度が、一般に比べ劣る。子どもたちの90%以上が成人する現在において、成人先天性心疾患患者は、社会の中で自立して生きていく力を身につける必要がある。そのためには、子どものひとり立ちに向けて、幼少期から自立を意識した父親のかかわりが重要である。

OJ8-3 先天性心疾患患児を持つ母の1ヶ月健診時エジンバラスケール(A病院 113例)**A report concerning the marks answered by mothers after childbirth in one month on the Edinburgh Postnatal Depression Scale (EPDS)**

福間 睦子, 桂木 真司

榊原記念病院

Mutsuko Fukuma, Shinji Katsuragi

Sakakibara Heart Insutitute

A病院は循環器専門病院として2014年より産科開設し胎児心疾患を持つ妊産婦の受け入れを行っている。周産期に児と母が離れ離れにならず児は早期より高度専門医療を受けられ、母は児の刻々と変化する状況をそばで見守っている。今回1か月健診時のエジンバラスケールの集計を行い、胎児心疾患症例の一部をまとめ以下の結果を得た。2014年9月～2017年10月に1か月健診を受けた373人の平均年齢は33.5歳、初産婦が212人(56.8%)、経産婦161人(43.2%)、経膈分娩277人(74.3%)、帝王切開96人(25.7%)、エジンバラスケール平均得点は5.8点であり質問10(希死念慮)が0点以外の方が22人(5.8%)いた。そのうち先天性心疾患を持つ母は113人(30.3%)、エジンバラスケール平均得点は7.4点であり、9点以上の方が45人(39.8%)質問10(希死念慮)0点以外の方が10人(8.8%)おり、それ以外の方256人(69.7%)の、平均5.2点、9点以上48人(18.6%)質問10(希死念慮)12人(4.7%)であった。疾患のある児は虐待を受ける割合が高く、母親のストレスは大きいという現状が明らかになっており、妊娠期から家族だけでなく医療スタッフのサポートを受け、継続して親子ケアを行うことで家族が健全に暮らしていけるように支援する必要性があると思われた。

OJ8-4 Fontan術後成人患者における筋力測定の見直し

Consideration of muscle strength measurement in fontan postoperative adult patients

近野 宏知¹⁾, 久松 智子¹⁾, 加藤 秀典¹⁾, 高橋 雅文¹⁾, 朴 要俊²⁾, 吳 龍梅²⁾, 石津 智子²⁾, 松原 宗明³⁾, 小池 朗²⁾

1) 筑波大学附属病院 リハビリテーション部, 2) 筑波大学附属病院 循環器内科, 3) 筑波大学附属病院 心臓血管外科

Hiroto Konno¹⁾, Tomoko Hisamatu¹⁾, Hidenori Kato¹⁾, Masafumi Takahashi¹⁾, Yasutoshi Boku²⁾, Longmei Wu²⁾, Tomoko Ishizu²⁾, Muneaki Matsubara³⁾, Akira Koike²⁾

1) Rehabilitation Department, University of Tsukuba Hospital, 2) Department of Cardiology, University of Tsukuba Hospital,

3) Department of Cardiovascular Surgery, University of Tsukuba Hospital

【背景】Fontan手術は術式の改良により長期予後が向上しているが、術後遠隔期の問題に骨格筋の機能異常がある。長期間にわたる活動量の低下が骨格筋量の減少、筋力の低下につながっていると考えられるが、術後遠隔期の筋力に関する報告は少ない。

【目的】外来心臓リハビリテーションを導入したFontan術後遠隔期患者の筋力と運動能を測定する。

【対象と方法】2000年～2012年にFontan手術(全例TCPC)を施行された患者4例(男性2例)、平均年齢21.0 ± 4.1歳を対象とし、2017年11月～2018年2月に心肺運動負荷試験を施行し、同時期に握力と膝伸展筋力を測定した。

【結果】握力: 23.2 ± 4.3kg、膝伸展筋力体重比: 0.49 ± 0.08kgf/kg、peak VO₂: 16.6 ± 2.0ml/min/kg(基準値の56.0 ± 11.2%)であり、健常成人と比べ筋力および運動耐容能の低下を認めた。

【考察】肺循環への駆出心室の欠如がFontan循環の特徴であることから、肺循環維持はFontan術後遠隔期の重要な要素であり骨格筋ポンプが果たす役割は大きい。一方で、握力と膝伸展筋力は最高酸素摂取量の重要な規定因子である。全身の筋力を推定する上で重要な評価指標である。Fontan術後患者における筋力の低下は骨格筋ポンプ作用を低下させ、更には肺循環へも悪影響を及ぼしていると考えられる。

【結語】Fontan術後患者における筋力評価は、遠隔期管理の重要な要素であり、その測定は遠隔期管理をより改善する上で有用であると考えられる。

OJ8-5 当院小児科に通院する成人先天性心疾患患者の服薬状況と薬効理解

Medication Adherence among Patients with Adult Congenital Heart Disease.

梶濱 あや, 島田 空知, 中右 弘一

旭川医科大学 小児科

Aya Kajihama, Sorachi Shimada, Kouichi Nakau

Department of Pediatrics, Asahikawa Medical University

Background: The patient with adult congenital heart disease (ACHD) has increased due to advances in medical procedures. To adopt a healthy lifestyle, they need to have good medication adherence.

Objective: To evaluate the illness cognition and medication adherence of ACHD patients.

Methods: Original questionnaire was distributed to the ACHD patients who visited our outpatient from July to December 2017. Patients with mental retardation were excluded.

Results: We enrolled 76 patients, 36 (48%) took medication but half of them couldn't answer their own disease. Furthermore, only 12 (33%) took their medicine steadily. Polypharmacy wasn't associated with poor adherence. There was difference in disease acceptance scores between good and poor adherence group ($p < 0.01$). Almost all of patients who took diuretic or anticoagulant had understanding of medication effect. But who took ACEi (18), 55% answered the effect as antihypertensive, 22% as unknown, and only 22% gave correct answer as cardioprotection.

Conclusion: The results showed many ACHD patients had a passive behavior to treatment and poor medicine adherence. Education focusing on meaning of medication would support to ameliorate the adherence.

OJ8-6

成人先天性心疾患患者における薬物療法コンサルテーション

Clinical pharmaceutical consultation for the patients with adult congenital heart diseases: lessons from our institutional cases

今井 靖^{1,2,5)}, 甲谷 友幸^{1,5)}, 久保田 香菜^{1,5)}, 今井 利美²⁾, 牛島 健太郎²⁾, 早川 朋子²⁾, 根岸 経太^{1,2)}, 永野 達也²⁾, Thanachai Methatham²⁾, 相澤 健一^{1,2)}, 片岡 功一^{3,5)}, 関 満^{3,5)}, 渡部 智紀¹⁾, 小森 孝洋¹⁾, 横山 靖浩¹⁾, 横田 彩子¹⁾, 藤村 昭夫²⁾, 苅尾 七臣^{1,5)}, 河田 政明^{4,5)}

1) 自治医科大学 内科学講座循環器内科学部門, 2) 自治医科大学 薬理学講座臨床薬理学部門, 3) 自治医科大学 小児科学講座, 4) 自治医科大学 外科学講座小児先天性心臓血管外科部門, 5) 自治医科大学成人先天性心疾患センター

Yasushi Imai^{1,2,5)}, Tomonori Kabutoya^{1,5)}, Kana Kubota^{1,5)}, Toshimi Imai²⁾, Kentaro Ushijima²⁾, Tomoko Hayakawa²⁾, Keita Negishi^{1,2)}, Tatsuya Nagano²⁾, Methatham Thanachai²⁾, Kenichi Aizawa^{1,2)}, Kouichi Kataoka^{3,5)}, Mitsuru Seki^{3,5)}, Tomonori Watanabe¹⁾, Takahiro Tomori¹⁾, Yasuhiro Yokoyama¹⁾, Ayako Yokota¹⁾, Akio Fujimura²⁾, Kazuomi Kario^{1,5)}, Masaaki Kawada^{4,5)}

1) Cardiovascular Medicine, Jichi Medical University, 2) Clinical Pharmacology, Jichi Medical University,

3) Pediatrics, Jichi Medical University, 4) Pediatric and Congenital Cardiovascular Surgery, Jichi Medical University,

5) Adult Congenital Heart Disease Center, Jichi Medical University

The number of the patients with adult congenital heart diseases has been increased remarkably, but some have to continue medical therapies after growing up. For female patients with cardiovascular disorders, if they are allowed to expect pregnancy, we should carefully select drugs to minimize the risk of harm to a mother and fetus, especially in the most critical term, the first trimester of pregnancy. In our clinical pharmacology division, we have provided consultation about drug use during pregnancy or lactation on the basis of the pharmacological knowledge and experiences guided with available databases. We have had 250-300 consultation cases per year in the whole of our hospital and approximately half of them are related to pregnancy or lactation, including antithrombotic and cardiovascular drugs for heart failure or complicated hypertension, although we should pay attention to the preexisting diseases themselves which might affect fetal or neonatal status. Here we would like to introduce our representative consultation cases and provide up-coming information about the aforementioned matters.

PE1-1

抗リン脂質抗体症候群を合併した機械弁僧帽弁置換術後妊婦の抗凝固療法戦略

The anti-coagulant management of the pregnant complicating the mechanical valve and anti-phospholipid antibody syndrome

中澤 直美¹⁾, 石津 智子¹⁾, 朴 要俊¹⁾, 日高 大介²⁾, 錦井 秀和³⁾, 小畠 真奈⁴⁾, 徳永 千穂⁵⁾, 長谷川 雄一³⁾, 宮園 弥生²⁾, 瀬尾 由広¹⁾, 家田 真樹¹⁾

1) 筑波大学 循環器内科, 2) 筑波大学 小児科, 3) 筑波大学 血液内科, 4) 筑波大学 産婦人科, 5) 筑波大学 心臓血管外科

Naomi Nakazawa¹⁾, Tomoko Ishizu¹⁾, Yasutoshi Boku¹⁾, Daisuke Hidaka²⁾, Hidekazu Nishikii³⁾, Mana Obata⁴⁾, Chiho Tokunaga⁵⁾, Yuichi Hasegawa³⁾, Yayoi Miyazono²⁾, Yoshihiro Seo¹⁾, Masaki Ieda¹⁾

1) Department of Cardiology, University of Tsukuba, 2) Department of Pediatrics, University of Tsukuba,

3) Department of Hematology, University of Tsukuba, 4) Department of Obstetrics and Gynecology, University of Tsukuba,

5) Department of Cardiovascular surgery, University of Tsukuba

Forty-year-old female with unexpected pregnancy was referred to our hospital. At 19, she underwent mechanical mitral valve replacement for infectious endocarditis. The blood sample test showed abnormal extended activated partial thromboplastin time (APTT), subsequently led to the diagnosis as anti-phospholipid antibody syndrome (APS). Since APS inhibits the APTT-monitoring and she had already passed the period of organogenesis, we controlled her coagulability with warfarin in the rest of pregnant period and replaced to heparin on the day before the cesarean section (CS). Though valve thrombosis was suspected by transesophageal echocardiography just before CS without hemodynamical change, CS was preceded. Thereafter, we resumed warfarin immediately after the hemostasis, confirming the mechanical valve by echocardiography in perioperative period. She and her baby discharged on the 21st postoperative day without any major complication.

We herein report the high-risk pregnant complicating the mechanical valve and APS. As the antithrombotic management of a pregnant with mechanical valve have many problems, an adequate explain in advance and a tailor-made protocol are mandatory.

PE1-2

右室前壁の広範な低電位領域を旋回する心室頻拍を呈したファロー四徴症修復術後の一例

Ventricular Tachycardia rotating Widespread Low Voltage Zone in Anterior Wall of Right Ventricle in An Adult Patient with Repaired Tetralogy of Fallot

中野 智彰, 塚本 泰正, 南口 仁, 小津 賢太郎, 溝手 勇, 水野 裕八, 彦惣 俊吾, 坂田 泰史

大阪大学大学院医学系研究科 循環器内科学

Tomoaki Nakano, Yasumasa Tsukamoto, Hitoshi Minamiguchi, Kentaro Ozu, Isamu Mizote, Yuya Mizuno,

Shungo Hikoso, Yasushi Sakata

Department of Cardiovascular Medicine, Osaka University Graduate School of Medicine

A 50-year-old woman with Repaired Tetralogy of Fallot (ToF) presented palpitation and diagnosed with ventricular tachycardia(VT). She underwent right ventricular outflow tract (RVOT) repair and patch closure of ventricular septal defect at 3 years old. She was hospitalized for heart failure caused by pulmonary regurgitation and underwent RVOT reintervention at 49 years old.

In three-dimensional electroanatomical mapping, the clinical VT was rotating widespread low voltage zone (LVZ) in the anterior wall of the right ventricle. Concealed entrainment was established by pacing from many sites in the LVZ and the VT was terminated by a radiofrequency application near the exit of the circuit.

However VTs with repaired ToF are typically caused by surgical incision, this VT may occur from the LVZ because of RV remodeling caused by long-term RV volume over load.

We experienced an unusual case of VT with repaired ToF.

PE1-3

QT延長症候群の心電図におけるQTc間隔の経時的変化

Interval change of QTc duration in patients of long QT syndrome

甲谷 友幸^{1,2)}, 今井 靖^{1,3)}, 久保田 香菜¹⁾, 苅尾 七臣¹⁾

1) 自治医科大学 内科学講座 循環器内科学, 2) 自治医科大学 成人先天性心疾患センター, 3) 自治医科大学 臨床薬理学

Tomoyuki Kabutoya^{1,2)}, Yasushi Imai^{1,3)}, Kana Kubota¹⁾, Kazuomi Kario¹⁾

1) Division of Cardiovascular Medicine, Department of Medicine, Jichi Medical University School of Medicine,

2) Adult Congenital Heart Disease Center, Jichi Medical University,

3) Clinical Pharmacology, Jichi Medical University School of Medicine

Background: QT duration changes by aging. However, a detail of interval change of QTc duration in individuals of long QT syndrome remains unclear.

Methods: We studied 19 outpatients of long QT syndrome (female 16 patients, male 3 patients, average age 27 ± 6 years). We investigated electrocardiography at latest visit and 5 years before, and compared QTc duration. We divided patients into young group (< 30 years, $N=9$) and elder group (≥ 30 years, $N=10$).

Results: In overall, QTc durations did not changed from 5 years ago and latest visit (467.6 ± 47.2 vs. 467.5 ± 37.0 ms, $P=0.99$). Although QTc durations did not changed from 5 years ago and latest visit (471.4 ± 60.7 vs. 457.8 ± 41.7 ms, $P=0.29$) in young group, QTc durations increased from 5 years ago to latest visit (464.1 ± 34.0 vs. 476.3 ± 31.8 ms, $P=0.042$) in elder group.

Conclusions: We should carefully follow electrocardiography especially in elder patients of long QT syndrome.

PE1-4

Fontan患者の上室性不整脈に対し、アブレーション、デバイス、薬物療法を行った一例

A case of ablation, pacemaker implantation and medication for supraventricular tachycardia in patient with Fontan operation

西井 伸洋¹⁾, 栄徳 隆裕²⁾, 重光 祐輔²⁾, 森本 芳正¹⁾, 浅田 早央莉¹⁾, 宮本 真和¹⁾, 杜 徳尚¹⁾, 中川 晃志¹⁾, 渡辺 敦之¹⁾, 森田 宏¹⁾, 笠原 真悟³⁾, 伊藤 浩¹⁾

1) 岡山大学大学院医歯薬学総合研究科 循環器内科, 2) 岡山大学大学院医歯薬学総合研究科 小児循環器科,

3) 岡山大学大学院医歯薬学総合研究科 心臓血管外科

Nobuhiro Nishii¹⁾, Takahiro Eitoku²⁾, YUsuke Shigemitsu²⁾, Yoshimasa Morimoto¹⁾, Saori Asada¹⁾,Masakazu Miyamoto¹⁾, Norihisa Tou¹⁾, Koji Nakagawa¹⁾, Atsuyuki Watanabe¹⁾, Hiroshi Morita¹⁾, Shingo Kasahara³⁾,Hiroshi Ito¹⁾

1) Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine, Dentistry,

2) Department of pediatric cardiology, Okayama University,

3) Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine

A case was 30 years old female diagnosed as polysplenia, double outlet right ventricle, mitral atresia, hypoplastic left heart syndrome. She was carried out TCPC conversion through lateral tunnel Fontan. Her rhythm was junctional rhythm with repetitive PAC short run, which was consistent with palpitation. She was also suffered from desaturation due to pulmonary arteriovenous fistula. And re-operation was planned to change the bloodstream of pulmonary artery. Before operation, hemodynamics was examined in a couple of pacing configuration. Hemodynamics was better in A pace V sense than that in junctional rhythm. Then, pacemaker implantation was also planned during operation. Because it was uncertain whether pacemaker implantation and medication could eliminate PAC, catheter ablation was performed. The approach to the systemic atrium was from internal jugular vein through pulmonary artery by Brockenbrough method. Several kinds of PAC were recognized, and main PAC was ablated, but elimination of all PAC was not achieved. After re-operation, her rhythm was A pace V sense and beta blocker was prescribed. After that, repetitive PAC short run was disappeared.

PE1-5 心室間同期不全・右室伝導遅延を呈した二心室修復後左室体心室の心臓再同期療法
The cardiac resynchronization therapy for a patient after biventricular repair with a interventricular dyssynchrony and subpulmonary conduction delay.

宮崎 文¹⁾, 松谷 勇人²⁾, 三宅 誠¹⁾, 山中 一朗³⁾, 池田 義⁴⁾, 桑野 和代²⁾, 土井 拓¹⁾

1) 天理よろづ相談所病院 先天性心疾患センター, 2) 天理よろづ相談所病院 臨床検査部, 3) 天理よろづ相談所病院 心臓血管外科, 4) 京都大学 心臓血管外科

Aya Miyazaki¹⁾, Hayato Matsutani²⁾, Makoto Miyake¹⁾, Kazuo Yamanaka³⁾, Tadashi Ikeda⁴⁾, Kazuyo Kuwano²⁾, Hiraku Doi¹⁾

1) Congenital Heart Disease Center, Tenri Hospital, 2) Department of Clinical Laboratory, Tenri Hospital, 3) Department of Cardiacvascular Surgery, Tenri Hospital, 4) Department of Cardiovascular Surgery, Graduate School of Medicine, Kyoto University

Background: In general adults, the role of intraventricular synchrony of left ventricle (LV) is more important than interventricular synchrony. The LV conduction delay is the major factor for response to cardiac resynchronization therapy (CRT).

Case: The case was a 31-year-old male with right isomerism after biventricular (BiV) repair with a systemic LV. He showed LV and RV dysfunction (LV end-diastolic volume index (EDVI) 154 ml/m²; LV ejection fraction (EF) 25%; RV EDVI 110 ml/m²; RVEF 35%). The QRS duration was 196 ms due to the LV anterior wall pacing. The peak longitudinal strain delay (pLSD) between mid-septal and LV free wall was 76 ms, but that between LV and RV free wall was 183 ms. In the intracardiac electrogram, the delay of RV free wall from QRS onset was 120 ms. We performed CRT implanting another pacing lead on the RV free wall. The pLSD between LV and RV free wall shortened to 32 ms. Cardiothoracic ratio decreased from 58.9 to 53.3%.

Discussion: CRT was effective for the patient with a BiV with a systemic LV showing the interventricular dyssynchrony and subpulmonary conduction delay. We must evaluate the detailed mechanism of heart failure in each patient.

PE1-6 心外膜リードと経静脈リードを用いCRTが有効であった修正大血管転位症の2症例
Two Cases of Cardiac Resynchronization Therapy Responder in Corrected Transposition of Great Arteries Using Epicardial and Transvenous Leads

梅本 真太郎¹⁾, 坂本 一郎¹⁾, 石北 綾子¹⁾, 帯刀 英樹²⁾, 永田 弾³⁾, 大賀 正一³⁾, 塩瀬 明²⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 心臓血管外科, 3) 九州大学病院 小児科

Shintaro Umemoto¹⁾, Ichiro Sakamoto¹⁾, Ayako Ishikita¹⁾, Hideki Tatewaki²⁾, Hazumu Nagata³⁾, Shoichi Ohga³⁾, Akira Shiose²⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Medicine, Kyushu University Hospital,

2) Department of Cardiovascular Surgery, Kyushu University Hospital, 3) Department of Pediatrics, Kyushu University Hospital

Introduction: Patients with corrected transposition of great arteries (cTGA) and systemic right ventricle (RV) suffers from heart failure. Some patients were implanted cardiac resynchronization therapy (CRT), but it is difficult to implant because of anatomical issues. We experienced 2 cases of CRT responder with cTGA using epicardial and transvenous leads.

Case 1: A 26-year-old female. She underwent pacemaker implantation due to complete AV block (CAVB) using epicardial V pacing lead. In 25 years old, pacemaker was upgraded from VVI to DDD using transvenous lead. In 26 years old, she underwent TVR and AVR, concomitant with CRT upgraded using transvenous leads and epicardial lead. Her QRS duration decreased from 204msec to 122msec.

Case 2: A 39-year-old female. She was implanted DDD pacemaker using transvenous leads due to CAVB, but suffered from heart failure in 39 years old. Her pacemaker was upgraded to CRT-P using transvenous leads and previously implanted epicardial lead. Her QRS duration decreased from 260msec to 146msec.

Conclusion: CRT using epicardial and transvenous leads was considered to be useful for patients with cTGA and systemic RV.

PE1-7

心室中隔欠損・動脈管開存のシャント閉鎖術後30年経過して増悪した遺残肺高血圧症**Worsened remnant pulmonary arterial hypertension after 30 years of shunt closure operation for VSD & PDA**久保田 香菜¹⁾, 甲谷 友幸^{1,2)}, 上野 修市^{1,3)}, 河野 健¹⁾, 今井 靖^{1,2,4)}, 河田 政明^{2,5)}, 苅尾 七臣^{1,2)}

1) 自治医科大学 内科学講座循環器内科部門, 2) 自治医科大学附属病院 成人先天性心疾患センター, 3) うえのクリニック,

4) 自治医科大学 薬理学講座臨床薬理学部門, 5) 自治医科大学とちぎ子ども医療センター 小児・先天性心臓血管外科

Kana Kubota¹⁾, Tomoyuki Kabutoya^{1,2)}, Shuichi Ueno^{1,3)}, Ken Kono¹⁾, Yasushi Imai^{1,2,4)}, Masaaki Kawada^{2,5)}, Kazuomi Kario^{1,2)}

1) Division of Cardiovascular Medicine, Jichi Medical University School of Medicine,

2) Center for Adult Congenital Heart Diseases, Jichi Medical University Hospital, 3) Ueno Clinic,

4) Division of Clinical Pharmacology, Jichi Medical University School of Medicine,

5) Department of Cardiovascular surgery, Jichi Children's Medical Center Tochigi

A 53-year-old woman had a two-year history of exertional dyspnea (WHO-FC II). At the age of nine, she underwent catheterization, which confirmed that her diagnosis was VSD and she had no surgical indication. When she was 22 years old, she strongly hoped pregnancy and underwent VSD closure and PDA ligation surgery. She became conscious of exertional dyspnea in her fifties and was hospitalized with suspected PAH. The right heart catheterization showed elevation of the mPAP (=55 mmHg) and PVR (=983 dyne·s·cm⁻⁵). Whereas, macitentan and riociguat were administered sequentially, right heart failure became apparent. After treatment with dobutamine and tolvaptan, her mPAP and PVR improved to 47 mmHg and 622 dyne·s·cm⁻⁵. Next, we are considering adding prostacyclin or related drugs.

There are few reports about the treatment for the aforementioned disease conditions. "Treat and repair" for Eisenmenger syndrome has also been actively discussed, and we believe her clinical course will suggest some therapeutic options for remnant PAH caused by pressure and volume overload.

PE1-8

PDA関連肺動脈性肺高血圧症に合併した肺動脈瘤ではEP4受容体が高発現している**Overexpression of EP4 in pulmonary artery aneurysm in patient with pulmonary arterial hypertension associated with patent ductus arteriosus**赤木 達¹⁾, 横山 詩子²⁾, 江尻 健太郎¹⁾, 中村 一文¹⁾, 伊藤 浩¹⁾

1) 岡山大学大学院医歯薬学総合研究科 循環器内科学, 2) 横浜市立大学 循環制御医学講座

Satoshi Akagi¹⁾, Utako Yokoyama²⁾, Kentaro Ejiri¹⁾, Kazufumi Nakamura¹⁾, Hiroshi Ito¹⁾

1) Department of Cardiovascular Medicine, Okayama University Hospital,

2) Department of Cardiovascular Research Institute, Yokohama City University

A 29-year-old man was diagnosed with pulmonary arterial hypertension associated with patent ductus arteriosus (PDA). Contrast-enhanced computed tomography demonstrated large pulmonary artery (PA) aneurysm with a diameter of 72 mm. His mean PA pressure was 44 mm Hg. PDA was closed with coil. Seven years after the diagnosis, he was transported to our hospital with sudden chest pain. Contrast-enhanced computed tomography demonstrated the expansion of a PA aneurysm with a diameter of 105 mm and a dissection in the main PA. The patient underwent emergency surgery for reconstruction of the right ventricular outflow tract and to perform bilateral PA plication. We investigated elastic fiber formation and prostaglandin E receptor type 4 (EP4), which is increased in abdominal aortic aneurysm, expression in the resected PA. EP4 expression was enhanced in the PA aneurysm, dissection, and PDA. EP4 was not expressed in the normal PA. PDA is known to be the most frequent congenital heart defects associated with a PA aneurysm.

EP4 overexpression in PA and PDA might contribute to the formation of a PA aneurysm and dissection in patients with pulmonary arterial hypertension.

PE2-1

Figulla Flex2によるASD閉鎖後の左房壁浸食、大動脈解離に対し緊急手術を行った一例
An emergency operation for erosion rupture of left atrial wall and aortic dissection after ASD closure by Figulla Flex 2

吉澤 康祐¹⁾, 藤原 慶一¹⁾, 前田 登史¹⁾, 加藤 おと姫¹⁾, 渡辺 謙太郎¹⁾, 植野 剛¹⁾, 大野 暢久¹⁾, 今井 逸雄²⁾

1) 兵庫県立尼崎総合医療センター 心臓血管外科, 2) 兵庫県立尼崎総合医療センター 循環器内科

Kosuke Yoshizawa¹⁾, Keiichi Fujiwara¹⁾, Toshi Maeda¹⁾, Otohime Kato¹⁾, Kentaro Watanabe¹⁾, Go Ueno¹⁾, Nobuhisa Ohno¹⁾, Masao Imai²⁾

1) Cardiovascular Surgery, Hyogo Prefectural Amagasaki General Medical Center,

2) Cardiology, Hyogo Prefectural Amagasaki General Medical Center

A 46-years-old male came to our hospital to check his ASD. He took steroid for atopic dermatitis. The majored diameter of ASD was 13.8 mm and the aortic and superior rims were absent. FF2 (19.5 mm) was placed with a flare shape on the aortic side. He was discharged 4 days after the placement. Two months later he was transferred emergently to our hospital for sudden chest pain during exercising. Enhanced CT revealed pericardial effusion and an ulcer like projection of the aortic wall. Emergency operation was performed. After CPB establishment and cross-clump, we removed the FF2 via right atrial incision. Two small perforations were found at the cranial side of the left atrium and were repaired with direct stiches. The ASD was closed with ePTFE patch. After de-clumped, the aortic wall was inspected carefully, and we found a massive breeding from the backside wall. We cross-clumped again and transected the aorta transversely. There was a laceration on the intima and it extended to the orifice of the LMT. We repaired the orifice and replaced the ascending aorta with an artificial graft. He was discharged 27 days after the operation, and doing well now 2 months after the operation.

PE2-2

ターナー症候群の若年女性の血圧に及ぼす二尖大動脈弁の影響

The effect of bicuspid aortic valve on the blood pressures of young women with Turner syndrome

康 秀貞

CHA 醫科學大學校 小兒青少年科學教室

Soojung Kang

Department of Pediatrics, CHA University School of Medicine, Seongnam, South Korea

The effect of bicuspid aortic valve on the blood pressure in young women with Turner syndrome is still unclear. We investigated the effect of bicuspid aortic valve on the blood pressures in young women with Turner syndrome, and compared the results with those of young women with Turner syndrome without bicuspid aortic valve. We analyzed the ambulatory blood pressure data and echocardiographic indices of 5 women with Turner syndrome who had bicuspid aortic valve, and 8 women with Turner syndrome who did not have bicuspid aortic valve. The mean age, weight, and height were not significantly different between the two groups. In addition, left ventricular ejection fraction and left ventricular mass indexed to body surface area were similar between the two groups. Mean systolic and diastolic blood pressures were similar between the two groups. Both systolic and diastolic blood pressure load were elevated in the two groups, but were similar between the two groups. The effect of bicuspid aortic valve on the blood pressures of young women with Turner syndrome were minimal. Longer durations of follow up would be needed to further elucidate this.

PE2-3

機械弁置換後の狭小僧帽弁に対する治療方針決定に難渋している症例

A Treatment Strategy for Mitral Stenosis Associated with Patient-Prosthesis-Mismatch After Mitral Valve Replacement; a Case Report

鍵本 美奈子¹⁾, 仁田 学¹⁾, 木野 旅人¹⁾, 松本 祐介¹⁾, 寺中 紗絵¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 上村 大輔¹⁾, 重永 豊一郎¹⁾, 細田 順也¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Minako Kagimoto¹⁾, Manabu Nitta¹⁾, Tabito Kino¹⁾, Yusuke Matsumoto¹⁾, Sae Teranaka¹⁾, Kiyamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Daisuke Kamimura¹⁾, Atsuichiro Shigenaga¹⁾, Junya Hosoda¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine

Background: There are limited surgical options for a mitral stenosis (MS) with small annulus.

Case: A 32-year-old male was diagnosed as MS associated with patient-prosthesis-mismatch (PPM). Soon after birth he was diagnosed as incomplete atrioventricular septal defect and underwent 4 times of mitral surgeries for mitral regurgitation. At the age of 7, mechanical mitral valve replacement (MVR) with using St-Jude Medical bileaflet valve (21mm) as a 4th surgery. A transthoracic echocardiogram revealed 2.6 m/sec of peak velocity. A cardiac catheterization showed 19 mmHg of pulmonary artery wedge pressure and 11.3 mmHg of mean transvalvular pressure gradient. The MV area was measured as 0.95 cm² by Gorlin's equation, suggesting severe MS. In addition, mean pulmonary artery pressure elevated to 33 mmHg. The leaflets were mobile with no pannus and thrombus, therefore we diagnosed as PPM accompanied with somatic growth. Aortic and mitral valve replacement with Manouguian procedure is planned to gain a larger-size of mitral annulus.

Conclusion: A patient after MVR during childhood should be taken into consideration double valve replacement in adulthood even if the aortic valve is intact.

PE2-4

甲状腺機能亢進がフォンタン循環へ及ぼす影響

Impact of hyperthyroidism on Fontan circulation

永田 弾¹⁾, 坂本 一郎²⁾, 梅本 真太郎²⁾, 石北 綾子²⁾, 江口 祥美¹⁾, 村岡 衛¹⁾, 福岡 将治¹⁾, 長友 雄作¹⁾, 平田 悠一郎¹⁾, 筒井 裕之²⁾, 大賀 正一¹⁾

1) 九州大学病院 小児科, 2) 九州大学病院 循環器内科

Hazumu Nagata¹⁾, Ichiro Sakamoto²⁾, Shintaro Umemoto²⁾, Ayako Ishikita²⁾, Yoshimi Eguchi¹⁾, Mamoru Muraoka¹⁾, Shoji Fukuoka¹⁾, Yusaku Nagatomo¹⁾, Yuichiro Hirata¹⁾, Hiroyuki Tsutsui²⁾, Shouichi Ohga¹⁾

1) Department of Pediatrics, Kyushu University Hospital, 2) Department of Cardiovascular Medicine, Kyushu University Hospital

Background: Fontan circulation could be impaired in some situations. It is unclear whether thyroid function impacts on Fontan circulation.

Case: A 34 year-old woman was referred to our institution for follow-up care at 31 years of age. She was diagnosed as tricuspid atresia at birth. At 3 years of age, Fontan procedure was performed. After surgery, NYHA was class I. The cardiac catheterization at the age of 32 years showed optimal hemodynamic status.

Laboratory data revealed high TSH with normal free T4. Oral thyroid hormone treatment was initiated to maintain the levels of TSH within normal range. With dose up, she presented with general fatigue, tachycardia, excessive sweating and edema. Chest X-ray showed cardiomegaly. The level of BNP was 134 pg/ml. Administration of diuretics and the discontinuation of levothyroxine improved the symptoms.

Conclusion: Although thyroid hormone modulate the homeostasis mediated by thyroid receptor, hyperthyroidism would be associated with water excretion and vasoconstriction leading to pulmonary hypertension. Thyroid hormone therapy might be considered in selected patients.

PE2-5

ファロー四徴症心内修復術後遠隔期に肺動脈弁置換術と冠血行再建を行った1例

Coronary Artery Bypass Grafting Concomitant with Pulmonary Valve Replacement for a Patient with Repaired Tetralogy of Fallot

寺中 紗絵¹⁾, 仁田 学¹⁾, 野田 光里¹⁾, 木野 旅人¹⁾, 松本 祐介¹⁾, 鍵本 美奈子¹⁾, 中島 理恵¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 上村 大輔¹⁾, 重永 豊一郎¹⁾, 細田 順也¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Sae Teranaka¹⁾, Manabu Nitta¹⁾, Hikari Noda¹⁾, Tabito Kino¹⁾, Yusuke Matsumoto¹⁾, Minako Kagimoto¹⁾, Rie Nakashima¹⁾, Kiwamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Daisuke Kamimura¹⁾, Atsuichiro Shigenaga¹⁾, Junya Hosoda¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine, 2) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine

Background: Patients with adult congenital heart disease (ACHD) are now increasing in number and aging.

Case: A 52-year-old male with fatigability was referred to our department as transition. He underwent intracardiac repair of Tetralogy of Fallot at the age of 4. He has received medication for hypertension, hyperlipidemia and hyperuricemia for more than 20 years and also pointed out to be diabetic. Cardiac MRI revealed 117 ml/m² of right ventricular end-diastolic volume index and 40% of pulmonary regurgitation (PR) fraction, suggested moderate PR. In addition, coronary angiogram showed atherosclerosis both in left anterior descending branch and left circumflex branch, the fractional flow reserve of the former was 0.75, suggested significant stenosis. Therefore, we performed one-stage surgery of coronary artery bypass grafting concomitant with pulmonary valve replacement. Our case highlighted the necessity of considering comorbidities that were likely to occur among elder ACHD patients.

Conclusion: Since patients with ACHD are now aging, we should care about not only the condition related to CHD but also cardiological impairment related to aging like general population.

PE2-6

当院における成人先天性心疾患患者の口腔状態の現況

Current Oral Condition of Patients with Adult Congenital Heart Disease in ACHD Center/Okayama University Hospital

大森 一弘^{1,3)}, 杜 徳尚^{2,3)}, 高知 信介¹⁾, 山本 直史¹⁾, 赤木 禎治^{2,3)}, 伊藤 浩^{2,3)}, 高柴 正悟¹⁾

1) 岡山大学病院 歯周科, 2) 岡山大学病院 循環器内科, 3) 岡山大学病院 成人先天性心疾患センター

Kazuhiro Omori^{1,3)}, Norihisa Toh^{2,3)}, Shinsuke Kochi¹⁾, Tadashi Yamamoto¹⁾, Teiji Akagi^{2,3)}, Hiroshi Ito^{2,3)}, Shogo Takashiba¹⁾

1) Department of Periodontics and Endodontics, Okayama University Hospital,

2) Department of Cardiovascular Medicine, Okayama University Hospital, 3) ACHD Center, Okayama University Hospital

Background: Infective endocarditis (IE) is one of the serious complications for adult congenital heart disease (ACHD). Oral bacterial infections are involved in the onset of IE, however, there is no report on the oral condition of ACHD.

Method: During the survey period (from May 2017 to August 2018), we examined oral condition of patients visiting ACHD Center/Okayama University Hospital.

Results: The number of patients examined was 14 (age: 32.7 ± 12.8 years old, female ratio: 71.4%, BMI: 22.2 ± 4.9kg/m², CRP value: 0.25 ± 0.44mg/dL; the value showed mean ± SD). Meanwhile, the oral condition was as follows (current teeth: 28.4 ± 2.6, teeth that required treatment: 2.1 ± 3.0, morbidity of severe periodontitis: 28.6%). In addition, the periodic dental examination rate at family dentistry was 35.7%.

Discussion: A large amount of infected foci existed in the oral cavity of ACHD patients without family dentistry. In addition, as age of patients increased, the oral condition tended to deteriorate. Therefore, in order to decrease the risk of developing IE, it is strongly recommend to provide appropriate dental information from early childhood, and to carry out the periodic dental checkup.

PE2-7

著明な収縮期雑音により、心疾患を疑われた一例

A suspected case of cardiac anomaly because of a marked systolic ejection murmur

松本 祐介¹⁾, 仁田 学¹⁾, 木野 旅人¹⁾, 寺中 紗絵¹⁾, 鍵本 美奈子¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 細田 順也¹⁾, 重永 豊一郎¹⁾, 上村 大輔¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Yusuke Matsumoto¹⁾, Manabu Nitta¹⁾, Tabito Kino¹⁾, Sae Teranaka¹⁾, Minako Kagimoto¹⁾, Kiwamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Junya Hosoda¹⁾, Atsichiro Shigenaga¹⁾, Daisuke Kamimura¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine

Background: A heart murmur can be an opportunity to detect a cardiac anomaly.

Case: A 20-year-old slender male presented to our hospital for investigating of heart murmur that had been pointed out at a medical checkup at his university. Although he was completely asymptomatic, he was suspected to have cardiac anomaly and transferred to our hospital. The Levine Grade 4/6 systolic ejection murmur with thrill was heard at the upper left sternal border. Thus, the murmur varied with respiration, markedly exacerbated in expiration and remitted in inspiration. Neither echocardiogram nor computed tomography showed any abnormalities. We also performed catheter examination on him, which proved pressure gradient (PG) change depends on respiration status; the PG between pulmonary artery and right ventricle (RV) was 7mmHg in expiration but no PG can be seen in inspiration. We concluded the main cause of his respiration-dependent-varied murmur was due to his skinny constitution. During expiration, RV outflow tract (RVOT) would be physically compressed by his tight thorax and released during inspiration.

Conclusion: A very mild RVOT stenosis can be a cause of marked systolic ejection murmur.

PE2-8

ファロー四徴症の心内修復術遠隔期に造影CTで高度肺うっ血を呈した一例

A case of severe pulmonary congestion exacerbated by contrast computed tomography long after intracardiac repair of tetralogy of Fallot.

柏村 健^{1,2)}, 武田 ルイ¹⁾, 西田 耕太¹⁾, 木村 新平¹⁾, 林 由香¹⁾, 保屋野 真¹⁾, 柳川 貴央¹⁾, 高野 俊樹¹⁾, 尾崎 和幸¹⁾, 白石 修一³⁾, 南野 徹¹⁾

1) 新潟大学大学院医歯学総合研究科 循環器内科学, 2) 新潟大学大学院医歯学総合研究科 先進心肺血管治療学講座,

3) 新潟大学大学院医歯学総合研究科 呼吸循環外科学分野

Takeshi Kashimura^{1,2)}, Rui Takeda¹⁾, Kota Nishida¹⁾, Shinpei Kimura¹⁾, Yuka Hayashi¹⁾, Makoto Hoyano¹⁾, Takao Yanagawa¹⁾, Toshiki Takano¹⁾, Kazuyuki Ozaki¹⁾, Shuichi Shiraishi³⁾, Tohru Minamino¹⁾

1) Department of Cardiovascular Biology and Medicine, Niigata University,

2) Department of Advanced Cardiopulmonary Vascular Therapeutics, Niigata University,

3) Division of Thoracic and Cardiovascular Surgery, Niigata University

Case: A 37-year-old female with repaired tetralogy of Fallot (TOF) was admitted to the hospital due to progressive dyspnea and lung congestion. Echocardiography revealed a flat interventricular septum and a peak pressure gradient of tricuspid valve regurgitation (TR-PG) was 97 mmHg. Diuretics improved her symptom and the cause of elevated TR-PG was explored by contrast computed tomography. Intravenous injection of contrast medium exacerbated lung congestion and PaCO₂ elevated to 149mmHg. Noninvasive positive pressure ventilation and diuretics improved the extreme hypercapnia. Ten days later, cardiac catheterization revealed pulmonary hypertension (56/20 [34] mmHg), mild pulmonary valve stenosis (peak-peak pressure gradient 16 mmHg), mildly high pulmonary artery wedge pressure (13mmHg), and mildly high pulmonary vascular resistance (3.2 Wood Unit). Fluid-challenge (normal saline 1mL/kg/min) caused dyspnea, and LVEDP rose from 14mmHg to 19mmHg in two minutes with only 75mL of normal saline.

Conclusion: Adult patients with repaired TOF may have multiple causes of high TR-PG. Left ventricular diastolic dysfunction should be considered in those with pulmonary congestion.

PE2-9

冠動脈瘤の増大が確認されたヌーナン症候群の1例～冠動脈瘤の病理報告

Pathological findings of a growing coronary aneurysm in Noonan syndrome

荻原 義人¹⁾, 藤本 直紀¹⁾, 石浦 純子¹⁾, 大橋 啓之²⁾, 山本 直樹³⁾, 伊藤 久人³⁾, 栗田 泰郎¹⁾, 土肥 薫¹⁾, 澤田 博文²⁾, 三谷 義英²⁾, 青木 洋子⁵⁾, 今中 恭子⁴⁾, 伊藤 正明¹⁾

1) 三重大学大学院 循環器・腎臓内科学, 2) 三重大学大学院 小児科学, 3) 三重大学大学院 胸部心臓血管外科学, 4) 三重大学大学院 修復再生病理学, 5) 東北大学大学院 遺伝医療学分野

Yoshito Ogihara¹⁾, Naoki Fujimoto¹⁾, Junko Ishiura¹⁾, Hiroyuki Ohashi²⁾, Naoki Yamamoto³⁾, Hisato Ito³⁾, Naoto Kurita¹⁾, Kaoru Dohi¹⁾, Hirofumi Sawada²⁾, Yoshihide Mintani²⁾, Yoko Aoki⁵⁾, Kyoko Imanaka Yoshida⁴⁾, Masaaki Ito¹⁾

1) Department of Cardiology and Nephrology, Mie University, 2) Department of Pediatrics, Mie University,

3) Department of Thoracic Cardiovascular Surgery, Mie University, 4) Department of Pathology and Matrix Biology, Mie University,

5) Department of Medical Genetics, Tohoku University School of Medicine

A 44-year-old man with Noonan syndrome was admitted to our hospital for the evaluation and treatment of a growing coronary aneurysm.

Three years before this admission, he was diagnosed as Noonan syndrome by genetic screening which was complicated with hypertrophied cardiomyopathy and coronary aneurysms. A CT scan showed an anomalous left anterior descending artery (LAD) with retrograde flow arising from the distal portion of the left circumflex due to a defect of the proximal portion of the LAD. The left main trunk (LMT) saccular aneurysm (37 × 27 mm) was the biggest and slightly calcified, and contained a thrombus. Therefore, warfarin anticoagulation was initiated.

On this admission, the CT scan showed the size of the LMT aneurysm increased to 52 × 35 mm. The patient underwent surgical ligation of the aneurysm and coronary artery bypass grafting because of the risk of rupture of the aneurysm. The post-operative course was uneventful.

To the best of our knowledge, this is the first case of Noonan syndrome in whom the expansion of the aneurysm was confirmed by imaging tests during follow-up. In addition, we report the pathological findings of coronary aneurysm in Noonan syndrome.

PE3-1

右心房内に膜様構造物を認めた二次孔型心房中隔欠損症の一例

A Case of Secundum Atrial Septal Defect with an Unusual Right Atrial Membrane

郡山 恵子¹⁾, 小坂橋 俊美¹⁾, 前川 恵美¹⁾, 藤田 鉄平¹⁾, 宮本 隆司²⁾, 北村 律²⁾, 宮地 鑑²⁾, 阿古 潤哉¹⁾

1) 北里大学医学部 循環器内科, 2) 北里大学医学部 心臓血管外科

Keiko Ryo Koriyama¹⁾, Toshimi Koitabashi¹⁾, Emi Maekawa¹⁾, Teppei Fujita¹⁾, Takashi Miyamoto²⁾,

Tadashi Kitamura²⁾, Kagami Miyaji²⁾, Junya Ako¹⁾

1) Department of Cardiology, Kitasato University, 2) Department of Cardiovascular Surgery, Kitasato University

Background: Transcatheter atrial septal defect (ASD) closure is gaining popularity in the treatment for ASDs because it is less invasive. However surgical treatment is selected in definite populations.

Case: A 44-year-old woman who was diagnosed ASD previously came to our hospital because of the feeling of palpitation. Transthoracic echocardiography showed that the right heart was dilated because of the volume overload by the shunt flow through the ASD. Transesophageal echocardiography revealed the ASD type was suitable for the transcatheter ASD closure, but there was an unusual membranous structure in the right atrium (RA). It attached posteriorly at the RA free wall almost 5mm far from the inter-atrial septum (IAS), and infero-anteriorly at IAS. The membranous structure and posterior IAS are almost parallel and the gap between them was supposed to be a disturbance for device deployment. Finally we decided to select the surgical treatment.

Conclusion: We experienced a case of the typical secundum ASD for which we selected the surgical therapy due to an unusual membranous structure in RA.

PE3-2

両側肺静脈還流異常症に対してゴアテックス人工血管を用いて再建した1例
Management of bilateral partial anomalous pulmonary venous connection

小川 真司¹⁾, 前田 正信¹⁾, 金子 完¹⁾, 大川 育秀²⁾, 菊地 慶太¹⁾

1) 一宮西病院 心臓血管外科, 2) 豊橋ハートセンター

Shinji Ogawa¹⁾, Masanobu Maeda¹⁾, Kan Kaneko¹⁾, Yasuhide Okawa²⁾, Keita Kikuchi¹⁾

1) Department of Cardiovascular Surgery, Ichinomiya Nishi Hospital, 2) Toyohashi Heart Center

Bilateral partial anomalous venous connections are rare. Here, we present a patient who underwent the modified Warden procedure to re-route the superior vena cava with the partial anomalous veins to the left atrium and reconstructed the innominate vein and superior vena cava with a polytetrafluoroethylene conduit to the right atrial appendage. The procedure was successfully treated without using foreign materials in the pulmonary venous route. Furthermore, it prevented the obstruction of the blood flow.

PE3-3

片肺フォンタン循環患者の重症胸部外傷の1例 –急性期治療と慢性期血行動態への影響–
A Case of Disastrous Lung Injury in a One-lung Fontan Patient: Successful Treatment in Acute Phase and Negative Impact on Hemodynamics

小永井 奈緒, 大内 秀雄, 根岸 潤, 坂口 平馬, 岩朝 徹, 白石 公, 黒崎 健一

国立循環器病研究センター 小児循環器科

Nao Konagai, Hideo Ohuchi, Jun Negishi, Heima Sakaguchi, Toru Iwasa, Isao Shiraishi, Kenichi Kurosaki

Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center

Backgrounds: Respiration plays an important role in the determinant of cardiac output in Fontan patients.

Case: A 39-year-old male patient with a history of pulmonary atresia treated with Fontan operation had developed complete thrombotic occlusion of left pulmonary artery at his age of 24. The catheterization had revealed failing Fontan status with central venous pressure of 15 mmHg. He was taken to our hospital because of respiratory failure due to multiple rib fractures, lung contusion and hemothorax on the right "available" side caused by a traffic accident. Hypoventilation led to critical decrease in ventricular preload via pulmonary circulation which resulted in cardiogenic shock. He was intubated and treated with inotropic agents. Although he was successfully weaned from mechanical ventilation in four weeks, the reduced intrathoracic space and weakness of inspiratory muscle, with additional decrease in %VC from 64% to 35%, may pose a great burden on respiratory pump function as well as the pulmonary gas exchange.

Comments: We'd like to share our rare, but thought-provoking experience to manage critically sicker patients with failed Fontan circulation.

PE3-4

右室二腔症に合併した心室頻拍に対してカテーテルアブレーションで根治に成功した一例
Successful Radiofrequency Catheter Ablation for Ventricular Tachycardia in a Patient with Double-chambered Right Ventricle.

木野 旅人¹⁾, 鍵本 美奈子¹⁾, 仁田 学¹⁾, 松本 祐介¹⁾, 寺中 紗絵¹⁾, 田口 有香¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 細田 順也¹⁾, 重永 豊一郎¹⁾, 上村 大輔¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Tabito Kino¹⁾, Minako Kagimoto¹⁾, Manabu Nitta¹⁾, Yusuke Matsumoto¹⁾, Sae Teranaka¹⁾, Yuka Taguchi¹⁾, Kiwamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Junya Hosoda¹⁾, Atsuichiro Shigenaga¹⁾, Daisuke Kamimura¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine

Background: Double-chambered right ventricle (DCRV) has an abnormal muscle band (AMB) in RV and associates with chest discomfort and right-side heart failure, however, fatal arrhythmias rarely develop.

Case: A 28-year-old female suffered from sustained ventricular tachycardia (VT) and was referred to our hospital. At the age of 3 months, she was diagnosed as ventricular septal defect (VSD) and DCRV. Thereafter VSD closed spontaneously, and surgical intervention was not performed because of insignificant pressure gradient in the RV (35 mmHg). After the admission to our hospital, the cardiac catheterization revealed just 30 mmHg of the pressure gradient across the RV outflow tract, suggesting no progression of DCRV. A radiofrequency catheter ablation (RFCA) was successfully performed. The target VT was documented not at an AMB site but just above the pulmonary valve, which is often accordant with an idiopathic VT. Therefore we conclude that the VT was idiopathic, unrelated to a DCRV.

Conclusion: An idiopathic VT can occur regardless of the progression of DCRV. RFCA plays an important role to manage it.

PE3-5

先天性門脈体循環シャントを閉塞後に門脈圧亢進をきたした成人例
An adult case of portal hypertension after the occlusion of congenital portosystemic shunt

前田 潤, 山本 一希, 荒木 耕生, 古道 一樹, 山岸 敬幸

慶應義塾大学医学部 小児科

Jun Maeda, Kazuki Yamamoto, Kousei Araki, Kazuki Kodo, Hiroyuki Yamagishi

Department of Pediatrics, Keio University School of Medicine

Congenital portosystemic shunt (CPSS) is often associated with pulmonary arterial hypertension (PAH). To date, no hepatic complication has been reported after therapeutic occlusion of the CPSS. A 25-year-old female was diagnosed as the intrahepatic portal atresia by abdominal imaging studies at the age of five years. She developed PAH at the age of 12 years and was sequentially administered pulmonary vasodilators, but PAH persisted. At the age of 22 years, a percutaneous balloon occlusion of the CPSS demonstrated the diminutive intrahepatic portal vein within tolerable range of portal pressure. After the staged surgical banding of the CPSS, the forward flow of the intrahepatic portal vein without obstruction was recognized. However, she developed melena and hematemesis due to esophageal varices and treated with urgent endoscopic ligation. Thereafter her ascites was remarkably worsened, resulting in anuria that required continuous hemodiafiltration. To our knowledge, this is the first case showing refractory portal hypertension after the successful closing of CPSS. The aggressive PAH therapy may be necessary for such case to consider liver transplantation.

PE3-6

動脈管開存症・アイゼンメンジャー症候群に腎膿瘍を合併した一例

Renal abscess in Eisenmenger syndrome with a patent ductus arteriosus

木島 康文, 福田 旭伸, 椎名 由美, 小宮山 伸之, 丹羽 公一郎

聖路加国際病院 循環器内科

Yasufumi Kijima, Terunobu Fukuda, Yumi Shiina, Nobuyuki Komiyama, Koichiro Niwa

Department of Cardiovascular Medicine, St. Luke's International Hospital

Eisenmenger syndrome (ES) with a patent ductus arteriosus (PDA) refers to a shunt associated with high pulmonary vascular resistance (PVR), reversal or bi-directional shunting flow at the great vessel, and cyanosis. Infection is a major cause of death in patients with ES. A 32-years-old female with ES induced by untreated huge PDA was referred to our hospital. Degree of oxygen saturation was 92% at rest. A catheterization study showed a mean pulmonary artery pressure of 102 mmHg and a PVR of 37 Wood Units. Six minute-walk distance was 210 m presenting the lowest saturation level of 40% when measured in the right upper extremity. She presented with a high fever and bacteriemia caused by klebsiella pneumoniae 4 weeks after the initial presentation. An enhanced CT demonstrated abscess in her right kidney which required emergent percutaneous drainage. During admission her oxygen saturation levels got unstable dropping to a level of 75% even with oxygen inhalation when she developed a high fever. Six weeks intravenous antibiotics was provided prior to her discharge. This case highlights the severity of infection and difficulty of management when an ES patient is infected.

PE3-7

18年間通院中断していたフォンタン術後の一例

Post Fontan Operation Patient Lost to Follow-Up for 18 Years

小平 真幸¹⁾, 田中 誠¹⁾, 田部井 亮太¹⁾, 沼澤 洋平¹⁾, 斎藤 暁人²⁾, 相馬 桂²⁾, 八尾 厚史²⁾, 小室 一成¹⁾

1) 足利赤十字病院 循環器内科, 2) 東京大医学部附属病院 循環器内科

Masaki Kodaira¹⁾, Makoto Tanaka¹⁾, Ryota Tabei¹⁾, Yohei Numasawa¹⁾, Akihito Saito²⁾, Katsura Soma²⁾,

Atsushi Yao²⁾, Issei Komuro¹⁾

1) Department of Cardiology, Japanese Red Cross Ashikaga Hospital, 2) Department of Cardiovascular Medicine, The University of Tokyo Hospital

A 28-year-old female patient was referred to our hospital for hepatomegaly from a local clinic. A thorough medical history revealed that her chief complaint was palpitation and shortness of breath, which led to consultation to our cardiology department. Physical examination and chest radiogram suggested signs of heart failure. Electrocardiogram demonstrated atrial flutter with tachycardia. Although the patient was able to inform us that she had undergone cardiac surgery in her childhood, she could not explain in detail. Furthermore, she had not been seen by any cardiologist for 18 years. Computed tomography (CT) and echocardiography revealed that she had received Atriopulmonary Fontan operation for pulmonary stenosis. The CT result also ruled out thrombus and we decided to perform electric cardioversion. Her cardiac rhythm was successfully converted to sinus rhythm after a single electric defibrillation. Amiodarone was prescribed to maintain her sinus rhythm and she was discharged two weeks later. Currently, we are preparing for a Total Cavopulmonary Connection conversion.

This case conveys the risk of lost to follow up among patients who have undergone Fontan operation.

PE3-8 先天性左肺無形性症を伴う三尖弁閉鎖症に対するフォンタン手術例：術後15年目の報告
A successful Fontan type operation for a patient with tricuspid atresia combined with congenital left lung agenesis; 15 years follow-up

久持 邦和¹⁾, 鎌田 政博²⁾, 川畑 拓也¹⁾, 中川 直美²⁾, 石口 由希子²⁾

1) 広島市民病院 心臓血管外科, 2) 広島市民病院 循環器小児科

Kunikazu Hisamochi¹⁾, Masahiro Kamada²⁾, Takuya Kawabata¹⁾, Naomi Nakagawa²⁾, Yukiko Ishiguchi²⁾

1) Department of Cardiovascular Surgery, Hiroshima Citizens Hospital,

2) Department of Pediatric Cardiology, Hiroshima Citizens Hospital

Congenital unilateral lung agenesis is a very rare anomaly. Its incidence is reported as about 1 in 100,000 births. Previously, we have successfully performed a Fontan type procedure for a patient of Tricuspid atresia (Ib) with congenital left lung agenesis, and have reported the case twice, first after surgery and second at 10-year follow-up period. At this time, 15 years have passed since the surgery, we would like to report the data as mid to long term condition of this rare case. The patient underwent bidirectional Glenn procedure as first palliation at 4-year-old, followed by Fontan type operation at 6-year-old. Since then, she has been in stable physical condition except one Fontan related hospital admission at 15-year-old. At that time, she underwent coil embolization to veno-venous collateral vessels, due to desaturation. Her oxygen saturation level was decreased from 93% to 86%, and was recovered to 94% after coil embolization. She is now 22-year-old part-time worker, in NYHA class I and taking only aspirin. Even with unilateral lung agenesis, the conditions are met; Fontan type operation may be feasible and achieve good mid to long term result.

PE3-9 大動脈炎症候群におけるMRAの有用性 – 上腕動脈閉塞例での経験 –
The usefulness of MRA for check up aortitis syndrome patient – a case report with brachial artery obstruction –

堀口 泰典¹⁾, 鈴木 淳子²⁾

1) 国際医療福祉大学熱海病院 小児科, 2) 東京通信病院 小児科

Yasunori Horiguchi¹⁾, Atsuko Suzuki²⁾

1) Department of Pediatrics, International University of Health and Welfare Atami Hospital,

2) Department of Pediatrics, Tokyo Teishin Hospital

Purpose: We report usefulness of MRA for aortitis syndrome patient.

Patient: Our patient is a 37 years old female who has been suffering from aortitis. At the onset she was 13 years old. Her symptoms were low grade fever, general fatigue and body weight loss.

Acute phase reactants were high (WBC 7800 CRP 6.6 ESR 94 mm/h). Dilatation of ascending aorta and stenosis of main branch of aortic arch, were found by angiography. Steroid and aspirin were started.

Clinical course has been good. Medication has been tapered steadily. Once a year MRA has been performed.

Left radial artery pulsation has been somewhat weak than right. But no symptom has been noticed. But left brachial artery obstruction was found by MRA.

Discussion: In aortitis, arterial stenotic lesion is found at main branch points of aorta. But in this patient, stenotic lesion was far from the aorta. So MRA must be done at extremities too.

Conclusion: 1) A patient of aortitis with left brachial artery obstruction is reported

2) MRA is useful tool to detect it.

3) In aortitis patients MRA must be done not only at near aortic portion but also at extremities.

PJ1-1

塞栓術を経て妊娠出産に至ったびまん性肺動静脈瘻の一例

Pregnancy and delivery in a patient after percutaneous closure of unilateral diffuse pulmonary arteriovenous malformations: a case report.

荻野 佳代¹⁾, 上田 和利¹⁾, 佐藤 一寿¹⁾, 林 知宏¹⁾, 脇 研自¹⁾, 新垣 義夫¹⁾, 大家 理伸²⁾, 福 康志²⁾, 門田 一繁²⁾, 清川 晶³⁾, 長谷川 雅明³⁾

1) 倉敷中央病院 小児科, 2) 倉敷中央病院 循環器内科, 3) 倉敷中央病院 産婦人科

Kayo Ogino¹⁾, Kazutoshi Ueda¹⁾, Kazutoshi Sato¹⁾, Tomohiro Hayashi¹⁾, Kenji Waki¹⁾, Yoshio Arakaki¹⁾, Masanobu Ohya²⁾, Yasushi Fuku²⁾, Kazushige Kadota²⁾, Hikaru Kiyokawa³⁾, Masaaki Hasegawa³⁾

1) Department of Pediatrics, Kurashiki Central Hospital, 2) Department of Cardiovascular Medicine, Kurashiki Central Hospital,

3) Department of Obstetrics and Gynecology, Kurashiki Central Hospital

We report a 24-year woman of diffuse multiple pulmonary arteriovenous malformation (PAVM) with cyanosis. The patient could complete pregnancy and delivery followed by embolization. She was diagnosed at the age of 1 by cyanosis. It was judged that there was no treatment indication because of diffuse type. She increasingly felt exertional fatigue and hoped to have a baby. We re-evaluated and found that PAVM was confined to the left lower lobe. Coil embolization was performed for 11 lesions. Her oxygen saturation improved from 83% to 94%. After 8 months, she got pregnant naturally. In consideration of the left to right shunt remaining, heparin injection was started for thrombosis prevention, and oxygen therapy was continued. In cardiopulmonary exercise test, the peak VO₂ was 14.8 ml/kg/min, the lowest saturation was 91%. On the 40th week of gestation, she had a healthy neonate with a body weight of 3,195 g by normal vaginal delivery. Her saturation was monitored during delivery. It is said that the maternal cardiorespiratory function and the severity of cyanosis are related to the prognosis of the fetus. The embolization for right to left shunt permitted to have a baby.

PJ1-2

拡張相早期の肥大型心筋症合併妊娠の一例

Successful Pregnancy in a Woman with an Early Stage of the Dilated Phase of Hypertrophic Cardiomyopathy

中尾 真大¹⁾, 奥村 亜純¹⁾, 河村 卓弥³⁾, 小野 良子¹⁾, 鈴木 僚¹⁾, 川端 伊久乃¹⁾, 吉田 純¹⁾, 桂木 真司¹⁾, 石黒 まや²⁾, 佐地 真育²⁾, 高見澤 格²⁾, 高山 守正²⁾

1) 榊原記念病院 産婦人科, 2) 榊原記念病院 循環器内科, 3) 三重大学医学部 産科婦人科学教室

Masahiro Nakao¹⁾, Asumi Okumura¹⁾, Takuya Kawamura³⁾, Ryoko Ono¹⁾, Ryo Suzuki¹⁾, Ikuno Kawabata¹⁾, Atsushi Yoshida¹⁾, Shinji Katsuragi¹⁾, Maya Ishiguro²⁾, Mike Saji²⁾, Itaru Takamisawa²⁾, Morimasa Takayama²⁾

1) Department of Obstetrics and Gynecology, Sakakibara Heart Institute, 2) Department of Cardiology, Sakakibara Heart Institute,

3) Department of Obstetrics and Gynecology, Mie University Faculty of Medicine

Introduction: Women with the dilated phase of hypertrophic cardiomyopathy (D-HCM) are vulnerable to volume overload and thought to be difficult to tolerate the pregnancy.

Case: A 34-year-old nulliparous woman with an early stage of D-HCM and NYHA functional class 2 had an oral beta-blocker. The echocardiograms showed the chamber dilatation and mild impairment of LV systolic function (LAD=47mm, LVDD/Ds=51/36mm, LVEF=51%, TRPG=18mmHg). The CMR performed two years before the pregnancy showed extensive late Gd enhancement in the LV myocardium. She had an exacerbation of LA dilatation in the second trimester of pregnancy and started the oral diuretics. She developed paroxysmal AF at 37 weeks of gestation and delivered a healthy male infant by cesarean section. After the delivery, she did not develop AF or heart failure, continuing the beta-blocker and the diuretics and discharged on the day 12.

Discussions: The volume changes during the pregnancy and the peripartum period have a strong potential for arrhythmias or heart failures in the patients with D-HCM. However, an early stage of D-HCM might be able to tolerate the pregnancy with effective use of beta-blockers and diuretics.

PJ1-3

妊娠20週に急性大動脈解離、大動脈弁輪拡張症を発症し生児を得た Marfan 症候群の一例
A case of acute aortic dissection and annulo-aortic ectasia in a pregnant woman with Marfan syndrome at the 20th week of gestation.

三島 桜子¹⁾, 森川 恵司¹⁾, 石田 理¹⁾, 入江 恭平¹⁾, 清水 かれん¹⁾, 築澤 良亮¹⁾, 久保 倫子¹⁾, 植田 麻衣子¹⁾, 片山 陽介¹⁾, 原賀 順子¹⁾, 関野 和¹⁾, 依光 正枝¹⁾, 上野 尚子¹⁾, 中西 美恵¹⁾, 児玉 順一¹⁾, 牧 耐太²⁾, 増山 寿²⁾

1) 広島市民病院, 2) 岡山大学病院 産婦人科

Sakurako Mishima¹⁾, Keiji Morikawa¹⁾, Makoto Ishida¹⁾, Kyohei Irie¹⁾, Karen Shimizu¹⁾, Yoshiaki Tsukizawa¹⁾, Rinko Kubo¹⁾, Maiko Ueda¹⁾, Yousuke Katayama¹⁾, Junko Haraga¹⁾, Madoka Sekino¹⁾, Masae Yorimitsu¹⁾, Naoko Ueno¹⁾, Yoshie Nakanishi¹⁾, Junichi Kodama¹⁾, Jota Maki²⁾, Hisashi Masuyama²⁾

1) Department of Obstetrics and Gynecology, Hiroshima City Hiroshima Citizens Hospital,

2) Department of Obstetrics and Gynecology, Okayama University Hospital

A 35-year-old woman was referred to a local medical center due to acute onset of chest back pain at 20 weeks of gestation. She was diagnosed with acute aortic dissection (Stanford B, De Bacay type 3b) by CT angiography and the administration of Ca antagonist was started to control her hypertension. The echocardiography revealed annulo-aortic ectasia and severe aortic regurgitation. She was getting to suffer from respiratory failure and was transferred to our hospital by helicopter four days after diagnosis. We considered acute aorta dissection and annulo-aortic ectasia complicated with Marfan syndrome which developed under the increase of afterload by pregnancy, following heart failure and lung edema. Bentall operation with a bioprosthetic heart valve was performed two days after transfer. After the operation, her prenatal course and fetal growth had been favorable. We performed a selective cesarean section at 30 weeks of gestation to prevent maternal morbidity. A 1463 gm male baby was delivered with a 5 min Apgar score of 7. She was discharged six days after operation and her baby on the 64th day. She is being followed up aorta dissection and is going to have more operation.

PJ1-4

心内膜床欠損症修復術後妊娠の一例
A case of pregnant woman with repaired atrioventricular septal defect

古橋 芙美, 真川 祥一, 二井 理文, 鳥谷部 邦明, 田中 博明, 池田 智明
三重大学 産婦人科

Fumi Furuhashi, Shoichi Magawa, Masafumi Nii, Kuniaki Toriyabe, Hiroaki Tanaka, Tomoaki Ikeda
Department of Obstetrics, Mie University

Purpose: One of complications in women with repaired atrioventricular septal defect (AVSD) is mitral valve stenosis (MS). Pregnant women with severe MS requires strict management during perinatal.

Case: 35 years old, primipara. After the repair of AVSD (4 years old: ASD patch closure + MV cleft suture + VSD direct closure + PDA ligation, 30 years old: MVP + DDD, PMI), She was regularly followed up in medical center. At 26 gestational weeks, hospitalization management was began for intensive maternal management. The evaluation of maternal cardiovascular status was performed with brain natriuretic peptide every 1 week and sonography and electrocardiogram every 2 weeks. At 33/2 gestational weeks, it became difficult to control the uterine contraction, and she had an emergency cesarean section in general anesthesia. After cesarean section, she was taken to the intensive care unit for strict management. No cardiovascular events occurred in postpartum. She was discharged on the 14th day.

Conclusion: A woman with severe MS due to the repair of AVSD had live birth without developing serious cardiovascular events by strictly management.

PJ1-5

当院で経験した先天性心疾患合併妊娠症例についての検討

Evaluation of cases of pregnancy with adult congenital heart disease

入江 恭平¹⁾, 三島 桜子¹⁾, 上野 尚子¹⁾, 清水 かれん¹⁾, 築澤 良亮¹⁾, 久保 倫子¹⁾, 森川 恵司¹⁾, 植田 麻衣子¹⁾, 片山 陽介¹⁾, 原賀 順子¹⁾, 関野 和¹⁾, 依光 正枝¹⁾, 中西 美恵¹⁾, 石田 理¹⁾, 児玉 順一¹⁾, 牧 尉太²⁾, 増山 寿²⁾

1) 広島市立広島市民病院 産科・婦人科, 2) 岡山大学病院 産科婦人科

Kyohei Irie¹⁾, Sakurako Mishima¹⁾, Naoko Ueno¹⁾, Karen Shimizu¹⁾, Yoshiaki Tsukizawa¹⁾, Rinko Kubo¹⁾, Keiji Morikawa¹⁾, Maiko Ueda¹⁾, Yosuke Katayama¹⁾, Junko Haraga¹⁾, Madoka Sekino¹⁾, Masae Yorimitsu¹⁾, Yoshie Nakanishi¹⁾, Makoto Ishida¹⁾, Junichi Kodama¹⁾, Jota Maki²⁾, Hisashi Masuyama²⁾

1) Department of Obstetrics and Gynecology, Hiroshima City Hiroshima Citizens Hospital,

2) Department of Obstetrics and Gynecology, Okayama University Hospital

OBJECTIVE: Pregnancies in patients with adult congenital heart disease (ACHD) have been increased. We aimed to discuss the outcomes of pregnancy with ACHD in our hospital.

METHODS: We examined a total of 29 patients (33 deliveries) with congenital heart disease who delivered in our hospital from 2014 to 2018, through medical records.

RESULTS: The commonest disease was VSD seen in 7 cases followed by ASD in 5 cases. We examined TGA 4 cases, TOF 2 cases, PA/IVS, and others (Ebstein's malformation, CoA/VSD, Marfan syndrome, Coronary artery fistula, bicuspid AoV). Cesarean section (CS) was performed in 15 cases including 3 emergency CS due to non-reassuring fetal status. We used preventive antibiotics without 2 cases (naturally closed VSD). We haven't experience infective endocarditis with these cases.

One case in Ebstein's malformation needed a general anesthesia and ICU management after CS for severe TR. One bicuspid AoV case was detected because of her cardiomegaly on chest X-ray routinely performed on admission.

CONCLUSION: Pregnancies in ACHD patients are at high risk for complications, and we should experience and discuss more cases to control pregnancies in ACHD safely.

PJ1-6

妊娠初期に感染性心内膜炎, 急性心不全を発症し生体弁置換を施行した一例

A case of Severe Infection Endocarditis and Cardiac Failure that needed Mitral Valve Replacement during the first trimester Pregnancy

桂木 真司, 中尾 真大, 奥村 亜純, 吉田 純, 小野 良子, 鈴木 僚, 川端 伊久乃, 藤巻 晴香, 石黒 まや, 古市 結富子, 清水 篤, 加瀬川 均

榊原記念病院 産婦人科

Shinji katuragi, Masahiro Nakao, Asumi Okumura, Atsushi Yoshida, Ryoko Ono, Ryou Suzuki, Ikuno Kawabata, Haruka Fujimaki, Maya Ishiguro, Yuko Furuichi, Atsushi Shimizu, Hitoshi Kasegawa

Department of Obstetrics and Gynecology, Sakakibara Heart Institute of Okayama

Case: The patient had received mitral valve (MV) repair at 16 years old (y.o.), and MV replacement (Normo valve) at 29 y.o. At 34 y.o. she experienced high fever, and was referred to our hospital. On admission she was in NYHA class IV, echocardiography revealed severe MV regurgitation, and vegetation was observed. We performed MV replacement with prosthetic valve. Streptococcus sanguinis was admitted from mother's blood and the removed valve, and antibiotics were given for 6 weeks. Drip infusion therapy by heparin was continued for 3 months after operation. Transient decrease of ejection fraction (EF)(40-45%), increased mean MV pressure gradient (8-10mmHg) were observed during pregnancy. We performed planned vaginal delivery under epidural anesthesia with antibiotics to prevent infectious endocarditis (IE) at 39 weeks. After delivery 4 months, she is in NYHA class I, and the cardiac function is good with EF 55%. The neurological and physical development of the newborn were good at 4 months old.

Conclusion: In severe IE with cardiac failure in pregnancy, we could save mother and fetus's lives by performing cooperated and intensive medical, surgical, and perinatal cares.

PJ1-7

妊娠出産を経験した遺残狭窄を有する大動脈縮窄症の一例

Successful pregnancy and delivery in a patient with significant pressure gradient after surgical repair of the aortic coarctation

平井 忠和¹⁾, 福田 信之¹⁾, 田中 修平¹⁾, 城宝 秀司¹⁾, 米田 哲³⁾, 福田 香織³⁾, 市田 露子²⁾, 絹川 弘一郎¹⁾

1) 富山大学 第二内科, 2) 富山大学 小児科, 3) 富山大学 産婦人科

Tadakazu Hirai¹⁾, Nobuyuki Fukuda¹⁾, Shuhei Tanaka¹⁾, Shuji Joho¹⁾, Satoshi Yoneda³⁾, Kaori Fukuda³⁾, Fukiko Ichida²⁾, Koichiro Kinugawa¹⁾

1) Second Department of Internal Medicine, University of Toyama, 2) Department of Pediatrics, University of Toyama,

3) Department of Obstetrics and Gynecology, University of Toyama

Women with a repaired coarctation of the aorta (CoA) have a relatively low risk of maternal cardiovascular complications during pregnancy, but residual stenosis of the aorta could be existed in some patients.

A 30 years old female patient had undergone surgical repair of complex CoA involving atrial septal defect, ventricular septal defect, and patent ductus arteriosus during infancy, but the resting pressure gradient across the CoA remained 30mmHg in postoperative follow-up catheter examination. Although residual coarctation was considered as moderate risk of pregnancy, she insisted on pregnancy and childbearing because of no clinical symptom. During pregnancy, there was no hypertension or adverse cardiovascular events, while mild increase of BNP (~44.1pg/ml) and sporadic premature ventricular contractions were observed during second-trimester. She spent her last month of pregnancy in our hospital under close observation, and a successful delivery was achieved without drug treatment. While significant CoA is regarded as a contraindication to pregnancy, successful pregnancy and delivery could be expected in mild to moderate residual stenosis of the CoA.

PJ1-8

先天性心疾患合併妊婦での左室拡張能指標の経時的変化と周産期心血管イベントについて

Temporal changes in the left ventricular diastolic function and peripartum cardiac events in pregnant women with congenital heart disease

福光 梓¹⁾, 宗内 淳²⁾, 金子 育美¹⁾, 小川 明希¹⁾, 奥田 知世¹⁾, 村田 真知子¹⁾, 秋光 起久子¹⁾,

渡辺 まみ江²⁾, 川上 剛史³⁾, 伊藤 浩司^{1,4)}

1) 地域医療機能推進機構九州病院 中央検査室, 2) 地域医療機能推進機構九州病院 小児循環器科,

3) 地域医療機能推進機構九州病院 産婦人科, 4) 地域医療機能推進機構九州病院 循環器内科

Azusa Fukumitsu¹⁾, Jun Muneuchi²⁾, Ikumi Kaneko¹⁾, Aki Ogawa¹⁾, Tomoyo Okuda¹⁾, Machiko Murata¹⁾, Kikuko Akimitsu¹⁾, Mamie Watanabe²⁾, Takeshi Kawakami³⁾, Kouji Itou^{1,4)}

1) Department of Clinical Laboratory, Japan Community Health care Organization Kyushu Hospital,

2) Department of Pediatric Cardiology, JCHO Kyushu Hospital, 3) Department of Gynecology, JCHO Kyushu Hospital,

4) Department of Cardiology, JCHO Kyushu Hospital

【背景と目的】 妊娠後期の左室拡張能低下は産褥心筋症をはじめとする妊娠関連心機能障害の指標となる。先天性心疾患(CHD)合併妊婦における左室拡張能指標の変化と予後について検討した。

【対象と方法】 2004年以降に当院で周産期管理中に心エコー図検査を施行したCHD合併妊婦94例(心血管イベントあり22例、なし72例)を対象として、妊娠初期、後期、産褥期の各期における左室拡張能指標(左房容積、E/A、E/e')を後方視的に追跡し、周産期管理中の母体心血管イベントとの関連について検討した。心血管イベントは不整脈、加療を要した心不全とした。

【結果】 イベントあり群において妊娠初期、後期、産褥期における各変化は、左房容積(ml): 44 vs 67 vs 48 (p<0.001)、E/A: 1.96 vs 1.36 vs 1.37 (p<0.001)、E/e': 9.66 vs 10.48 vs 11.67 (p=0.35)であるのに対し、イベントなし群では左房容積(ml): 42 vs 45 vs 45 (p=0.163)、E/A: 2.13 vs 1.67 vs 1.60 (p<0.05)、E/e': 10.86 vs 9.86 vs 11.30 (p=0.08)であった。両群ともに妊娠初期から後期にかけてE/Aは低下し、E/e'は変化しなかったが、イベントあり群では妊娠後期の左房容積が有意に大きかった(p<0.001)。

【考察】 妊娠経過中の左室拡張能指標は心血管イベントの関与で異なる推移を呈し、特に左房負荷増大に伴う変化は心血管イベント発症の予測指標になりうる可能性が示唆された。

PJ2-1

経静脈的に心房リードの植え込みを行った心外導管を用いた TCPC 術後の一例

Tranvenous Atrial Lead implantation in Patient with Toral CavoPulmonary Connection Using Extracardiac Conduit

坂本 一郎¹⁾, 向井 靖¹⁾, 梅本 真太郎¹⁾, 石北 綾子¹⁾, 出口 裕子¹⁾, 永田 弾²⁾, 帯刀 英樹³⁾, 大賀 正一³⁾, 塩瀬 明³⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 小児科, 3) 九州大学病院 心臓血管外科

Ichiro Sakamoto¹⁾, Yasushi Mukai¹⁾, Shintaro Umemoto¹⁾, Ayako Ishikita¹⁾, Hiroko Deguchi¹⁾, Hazuki Nagata²⁾, Hideki Tatewaki³⁾, Shonichi Ohga³⁾, Akira Shiose³⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Surgery, Kyushu Univesity, 2) Department of Pediatrics, Kyushu University Hospital,

3) Department of Cardiovascular Surgery, Kyushu Univesity Hospital

We reported a 15-year-old male with pulmonary atresia without intact ventricular septum defect and hypoplastic right ventricle. He had received a Total CavoPulmonary Connection (TCPC) using extracardiac conduit (EC) at the age of 4 years. At the age of 2 years, an VVI pacemaker (PM) using epicardial ventricular (V) lead had been implanted because of transient AV block. At 13 years old, epicardial V lead re-implantation via left 6th intercostal space thoracotomy was performed because of disconnection of V lead. At this time, atrial (A) lead implantation was attempted but could not be implanted for technical issue. He became symptomatic because the ratio of V pacing was increased for progressive AV block. Because he was not a candidate for open surgery except for A lead implantation, we attempted to A lead implantation via transvenous approach. A lead was implanted at main pulmonary artery via right subclavian vein and connected to generator at left side chest. In this procedure, he got AV sequential pacing and became asymptomatic. Transvenous A lead implantation might be one of alternatives for patients with TCPC using EC who need A pacing and not candidate for surgery.

PJ2-2

下大静脈欠損, 右胸心, 房室中隔欠損症術後の心房頻拍にアブレーションを行った1例

Successful catheter ablation of atrial tachycardia in a patient with surgically corrected cAVSD with IVC defect

庄島 耀子, 坂本 一郎, 池田 翔大, 河合 俊輔, 石北 綾子, 長岡 和宏, 坂本 和生, 林谷 俊児, 向井 靖, 樽木 晶子, 筒井 裕之

九州大学病院 循環器内科

Yoko Shojima, Ichiro Sakamoto, Shota Ikeda, Shunsuke Kawai, Ayako Ishikita, Kazuhiro Nagaoka,

Kazuo Sakamoto, Shunji Hayashidani, Yasushi Mukai, Akiko Chishaki, Hiroyuki Tsutsui

Department of Cardiovascular Medicine, Kyushu University Hospital

A 27-year-old male, who was diagnosed complete atrioventricular septal defect with inferior vena cava (IVC) defect and dextrocardia, was performed biventricular repair at 1 years old. He was performed fourth open heart surgery until 17 years old. He had performed epicardial VVI permanent pacemaker implantation at 10 years old for complete atrioventricular block. The index radiofrequency catheter ablation was performed at 17 years old for atrial tachycardia (AT) via transfemoral approach but AT was recurred. After AAI pacemaker was added via transvenous, AT was decreased. However AT was reappeared at 27 years old. We retried catheter ablation via internal jugular approach. The Ripple map in the right atrium (RA) during the AT showed a figure-8 pattern. Clinical AT was diagnosed as incisional macro-reentrant AT. Fragmented potential was identified at RA and radiofrequency (RF) catheter ablation was performed. AT was terminated by first ablation and could not be inducible. AT did not recur thereafter. Catheter ablation was useful even in patients with repaired complex congenital heart disease.

PJ2-3

QT延長症候群に対して着用型除細動器を使用した1例

A case of long QT syndrome with wearable cardioverter defibrillator

齋藤 俊祐, 甲谷 友幸, 久保田 香菜, 今井 靖, 苅尾 七臣

自治医科大学附属病院 循環器内科

Shunsuke Saito, Tomoyuki Kabutoya, Kana Kubota, Yasushi Imai, Kazuomi Kario

Division of Cardiovascular Medicine, Jichi Medical University School of Medicine

Case presentation: A 20s woman was referred to our hospital due to ventricular tachycardia. She had fainted during taking with her friends and had been carried to other hospital 4 days before transferring to our hospital. She was checked by brain CT, and the finding of CT was normal. Nextday, she had fainted supine, and fell from the bed. She felt dyspnea and carried to other hospital. Her electrocardiogram showed QT prolongation and polymorphic ventricular tachycardia. She had fainted and recovered by automated external defibrillator shocks. She underwent temporary cardiac pacing and medication of magnesium, and was referred to our hospital. She had no events of ventricular tachycardia after introducing beta blocker. We presented the choices of ICD and WCD, she hoped WCD therapy. She acquired the WCD care during her hospitalization and discharged. During three months observation, no ventricular tachycardia was detected (Average wearing time 23 hours). She had no ventricular events for 3 years with oral medication of beta blocker and potassium. The result of the gene examination later had sent us and diagnosed as LQT 1 with KCNQ1 674Q missense.

PJ2-4

顕著な肺動脈拡大を伴う Eisenmenger 症候群に肺化膿症を合併した1例

A case of lung abscess with Eisenmenger syndrome with extremely dilated pulmonary artery

鈴木 大¹⁾, 岩朝 徹¹⁾, 大内 秀雄¹⁾, 坂口 平馬¹⁾, 白石 公¹⁾, 津田 悦子¹⁾, 黒崎 健一¹⁾, 山田 修^{1,2)}

1) 国立循環器病研究センター 小児循環器科, 2) 国立循環器病研究センター 臨床病理科

Dai Suzuki¹⁾, Toru Iwasa¹⁾, Hideo Ohuchi¹⁾, Heima Sakaguchi¹⁾, Isao Shiraishi¹⁾, Etsuko Tsuda¹⁾, Kenichi Kurosaki¹⁾, Osamu Yamada^{1,2)}

1) Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center,

2) Department of Pathology, National Cerebral and Cardiovascular Center

35-years-old man with Eisenmenger syndrome developed severe lung abscess. His pulmonary artery was extremely dilated and there was in situ thrombi in both pulmonary arteries. He required several unscheduled hospitalization from recurrent hemoptysis.

He presented high fever, hypoxia and dyspnea, his C reactive protein was elevated as high as 35mg/dl. Chest X-ray and computed tomography revealed lung abscess in his left upper lobe. Standard antibiotic therapy was not successful. Because the lung abscess became enlarged and close to left pulmonary artery, the risk of rupture of pulmonary artery was considered. Intensive care and combination therapy of antibiotics was effective. Cultivation could not detect any suspicious bacteria. The follow up CT revealed his left upper bronchi and left superior pulmonary vein were occluded by the compression from dilated pulmonary artery. We thought this problem made the respiratory infection complicated.

In the patients with Eisenmenger syndrome, severe respiratory infection was not major complication, but we should take care of the serious respiratory infection in the patient with extremely dilated pulmonary vessels.

PJ2-5

Mustard術後の静脈狭窄と房室ブロックにステントとペースメーカー留置術を行った一例

A case of stent and pacemaker implantation for superior and inferior stenosis late after Mustard operation.

麻生 健太郎¹⁾, 中野 茉莉恵¹⁾, 桜井 研三¹⁾, 水野 将徳¹⁾, 高野 誠²⁾, 三村 秀文³⁾

1) 聖マリアンナ医科大学 小児科, 2) 聖マリアンナ医科大学 循環器内科, 3) 聖マリアンナ医科大学 放射線科

Kentaro Aso¹⁾, Marie Nakano¹⁾, Kenzo Sakurai¹⁾, Masanori Mizuno¹⁾, Makoto Takano²⁾, Hidefumi Mimura³⁾

1) Department of Pediatrics, St. Marianna University School of Medicine,

2) Department of Cardiology, St. Marianna University School of Medicine,

3) Department of Radiology, St. Marianna University School of Medicine

We experienced patient who underwent pacemaker and stent implantation for advanced atrioventricular block and vena cava stenosis that occurred late after Mustard operation.

Case: A 41-year-old female patient with transposition of the great artery who underwent a Mustard operation at age of 2 years. She recognize dyspnea on exertion since 39 years of age. Advanced AV block was observed on electrocardiogram so We planned pacemaker implantation (PMI). Prior to PMI Cardiac CT was conducted and confirmed inferior and superior vena cava stenosis. Stenosis is highly likely to proceed further we decided to do PMI after placing the stent in the stenosed region. Niti-STM gastroduodenal stent was used and PMI was performed 4 days after stent implantation. The pacemaker setting was DDD mode with basic heart rate to 80 bpm.

Discussion: Along with an increase in adult congenital heart disease patients, opportunities for stent placement for venous stenosis are expected to increase. Currently there are no stents that have official indications in vein in Japan. Although it is off label use, Niti-STM gastroduodenal stent is suitable for treatment against venous stenosis.

PJ2-6

上大静脈症候群に対する自己拡張型ステント留置術

Self-expandable stent placement for treatment of superior vena cava syndrome

赤澤 祐介¹⁾, 鈴木 萌子¹⁾, 中尾 恭久¹⁾, 東 晴彦¹⁾, 佐々木 康浩¹⁾, 藤井 昭¹⁾, 上谷 晃由¹⁾, 青野 潤¹⁾, 永井 啓行¹⁾, 西村 和久¹⁾, 井上 勝次¹⁾, 池田 俊太郎¹⁾, 宮田 豊寿³⁾, 森谷 友造³⁾, 千阪 俊行³⁾, 高田 秀実^{2,3)}, 打田 俊司⁴⁾, 檜垣 高史^{2,3)}, 石井 榮一^{2,3)}, 山口 修¹⁾

1) 愛媛大学大学院医学系研究科 循環器・呼吸器・腎高血圧内科学, 2) 愛媛大学大学院医学系研究科 地域小児・周産期学講座,

3) 愛媛大学大学院医学系研究科 小児科学講座, 4) 愛媛大学大学院医学系研究科 心臓血管・呼吸器外科学

Yusuke Akazawa¹⁾, Moeko Suzuki¹⁾, Yasuhisa Nakao¹⁾, Haruhiko Higashi¹⁾, Yasuhiro Sasaki¹⁾, Akira Fujii¹⁾, Teruyoshi Uetani¹⁾, Jun Aono¹⁾, Takayuki Nagai¹⁾, Kazuhisa Nishimura¹⁾, Katsuji Inoue¹⁾, Shuntaro Ikeda¹⁾, Toyohisa Miyata³⁾, Tomozou Moritani³⁾, Toshiyuki Chisaka³⁾, Hidemi Takata^{2,3)}, Shunji Uchita⁴⁾, Takashi Higaki^{2,3)}, Eiichi Ishii^{2,3)}, Osamu Yamaguchi¹⁾

1) Ehime University Graduate School of Medicine, Department of Cardiology, Pulmonology, Hypertension & Nephrology,

2) Ehime University Graduate School of Medicine, Department of Regional Ped. and Perinatology,

3) Ehime University Graduate School of Medicine, Department of Pediatrics,

4) Ehime University Graduate School of Medicine, Department of Cardiovascular and Thoracic Surgery

A 66-year-old man was diagnosed with partial anomalous pulmonary venous return (PAPVR) during the treatment of right glottis cancer at age 63. Cardiac catheterization revealed Qp/Qs=2.6, mean PAP=23 mmHg. He underwent modified Williams method. 2 years after the operation, he was diagnosed with superior vena cava (SVC) syndrome based on facial edema after awakening. We suspected the stenosis has occurred due to the compression of SVC (14mm PTFE graft interposed) by pulmonary vein (12mm ringed PTFE graft) accompanying SVC, chest wall and aorta. While his symptoms improved after the balloon angioplasty procedure, restenosis was observed within a few days. We then performed self-expandable stent (SMART Control[®] 14mm×60mm) placement for SVC. We met the challenge of keeping the both flow of PV return and SVC return during the treatment by confirming the patency of pulmonary vein with CT during balloon expansion. We also successfully prevented the stent migration by expanding it from the side of SVC and placing it flared end. SVC syndrome is one of the problems after surgical repair for PAPVR. We report self-expandable stent placement for the treatment of SVC syndrome.

PJ2-7

中間報告：成人先天性心疾患患者における直接作用型経口抗凝固薬の有効性に関する検討
Interim Report of a Multicenter Prospective Cohort Study: Effect of Direct Oral Anticoagulant in Adult Congenital Heart Disease Patients

増田 慶太¹⁾, 石津 智子²⁾, 青沼 和隆²⁾, 家田 真樹²⁾

1) 横浜労災病院 不整脈科, 2) 筑波大学医学医療系 循環器内科

Keita Masuda¹⁾, Tomoko Ishizu²⁾, Kazutaka Aonuma²⁾, Masaki Ieda²⁾

1) Department of Heart Rhythm Management, Yokohama Rosai Hospital,

2) Department of Cardiology, Faculty of Medicine, University of Tsukuba

Background and Objective: Data about the use of direct oral anticoagulant (DOAC) in adult congenital heart disease (ACHD) patients are lacking. The purpose of this multicenter prospective cohort study was to evaluate the effectiveness and safety of DOAC as compared with warfarin in ACHD patients.

Results: As of August 2018, 37 patients (age 40 ± 17 years; 23 men; CHA₂DS₂-VASc score 1.5 ± 1.0) from 9 institutes were registered. The details of the underlying heart diseases were as follows: UVH in 9 patients, TOF in 8, ASD in 5, VSD in 4, AVSD in 3, and other in 8. Of the patients, 28 (76%) had moderate or severe disease complexity. For anticoagulation, 22 patients took warfarin (warfarin group) and the other 15 took DOAC (DOAC group). The indication for anticoagulation was post-valve replacement in most patients in the warfarin group, whereas it was arrhythmia in all the patients in the DOAC group. No major bleeding events were observed in both groups during the follow-up periods.

Conclusion: DOAC was commonly used in the clinical situation of ACHD patients and favorable results were obtained during short-term follow-up. Further case registration and follow-up are ongoing.

PJ2-8

左右肺動脈の高低差に起因する Platypnea-Orthodeoxia Syndrome のフォンタン循環の1例
A case with Platypnea-Orthodeoxia Syndrome caused by height difference between right and left pulmonary arteries in the Fontan circulation.

田尻 雄二郎¹⁾, 宮崎 文^{1,2)}, 三宅 誠²⁾, 齊藤 瞬¹⁾, 樋垣 諒¹⁾, 三木 直木¹⁾, 御前 隆³⁾, 土井 拓^{1,2)}

1) 天理よろづ相談所病院 小児科, 2) 天理よろづ相談所病院 先天性心疾患センター, 3) 天理よろづ相談所病院 放射線部

Yujirou Tajiri¹⁾, Aya Miyazaki^{1,2)}, Makoto Miyake²⁾, Shun Saito¹⁾, Ryo Higaki¹⁾, Naoki Miki¹⁾, Takashi Misaki³⁾, Hiraku Doi^{1,2)}

1) Department of Pediatrics, Tenri Hospital, 2) Congenital Heart Disease Center, Tenri Hospital,

3) Department of Radiology, Tenri Hospital

Background: Platypnea-Orthodeoxia Syndrome (POS) is a syndrome exhibiting tachypnea and hypoxia in standing, caused by right to left (RL) shunt or ventilation-perfusion mismatch (VQM). POS was reported as a complication in Fontan circulation (FC) with RL shunt.

Case: The case was a 22-year-old woman with situs inversus after the Fontan operation. Her oxygen saturation was 93% during supine and 88% during standing. No significant RL shunt was observed. We performed pulmonary ventilation/perfusion scintigraphy during supine and sitting. No ventilation abnormalities were observed. However, the perfusion during sitting was reduced at the whole right lung and the apex of both lungs. The R/L ratio was 1.44 during supine and 0.52 during sitting. The proximal right pulmonary artery (rPA) was located more cranial than the left pulmonary artery (lPA). Then, we diagnosed her desaturation as POS caused by postural VQM which came from the height difference between rPA and lPA.

Discussion: The effect of gravity on hemodynamics in FC is greater than that of two ventricles, due to the lack of a sub-pulmonary ventricle pump. The height difference between rPA and lPA may cause POS in FC.

PJ3-1

**修正大血管転位症修復術後の三尖弁置換術において左側左房アプローチが有用だった一例
The left-sided left atriotomy in systemic tricuspid valve replacement for the repaired corrected transposition of the great arteries.**

阿瀬 孝治, 新川 武史, 松村 剛毅, 中山 祐樹, 宝亀 亮悟, 小林 慶, 新浪 博士
東京女子医科大学病院 心臓血管外科

Koji Aze, Takeshi Shinkawa, Goki Matsumura, Yuki Nakayama, Ryogo Hoki, Kei Kobayashi, Hiroshi Niinami
Department of Cardiovascular Surgery, Tokyo Women's Medical University

We report a 22-year-old male with past medical history of conventional repair for corrected transposition of the great arteries with left ventricle-to-pulmonary artery conduit and epicardial pacemaker placement at 4 years of age and systemic tricuspid valve repair at 20 years of age. He underwent transvenous cardiac resynchronization therapy system placement for residual tricuspid regurgitation and atrial tachycardia, which was complicated with infectious endocarditis a year later. Removal of infected materials and tricuspid valve replacement were indicated after successful preoperative antibiotics therapy. Even it was thought to be difficult to achieve an adequate view of tricuspid valve due to mesocardia and atrioventricular groove on the horizontal plane, an excellent view was obtained through left-sided left atriotomy. The patient successfully underwent tricuspid valve replacement, conduit replacement and removal of transvenous system. As conclusion, preoperative antibiotics therapy and left-sided left atriotomy approach were useful in this case.

PJ3-2

**ファロー四徴症根治40年後にPVR、TAPを施行した60歳男性の一治験例
A 60 years-old case of pulmonary and tricuspid regurgitation 40 years after total correction for Tetralogy of Fallot.**

豊田 泰幸¹⁾, 矢崎 善一²⁾, 竹村 隆広¹⁾, 柳澤 聖²⁾

1) 佐久医療センター 心臓血管外科, 2) 佐久医療センター 循環器内科

Yasyuki Toyoda¹⁾, Yosikazu Yazaki²⁾, Takahiro Takemura¹⁾, Takashi Yanagisawa²⁾

1) Department of Cardiovascular Surgery, Saku Central Hospital, Advanced Care Center,

2) Department of Cardiology, Saku Central Hospital Advanced Care Center

A 60-year-old man was admitted to Saku Central Hospital because of right ventricular dysfunction. He was diagnosed of Tetralogy of Fallot and performed original Blalock-Taussig shunt operation in 6 years old and total correction in 20 years old. Preoperative magnet Image Resonance showed RVEDVI was 185ml/m² and LVEDVI was 75.4ml/m². Ultrasound cardiography showed severe pulmonary and tricuspid regurgitation. Pulmonary valve replacement and tricuspid valve annuloplasty was performed. Postoperative was uneventful.

PJ3-3 PA/VSDに対する姑息術40年後の肺動脈瘤と大動脈基部拡大に対し二期的根治に至った1例
A successful surgical case of Rastelli procedure combined with valve-sparing aortic root replacement for PA/VSD in adult.

安東 悟央¹⁾, 加藤 伸康¹⁾, 新井 洋輔¹⁾, 橘 剛¹⁾, 稗田 哲也¹⁾, 下地 章夫¹⁾, 石垣 隆弘¹⁾, 杉本 聡¹⁾, 関 達也¹⁾, 新宮 康栄¹⁾, 大岡 智学¹⁾, 加藤 裕貴²⁾, 久保田 卓¹⁾, 松居 喜郎¹⁾

1) 北海道大学病院 循環器・呼吸器外科, 2) 北海道大学病院 先進急性期医療センター・救急科

Norihiro Ando¹⁾, Nobuyasu Kato¹⁾, Yosuke Arai¹⁾, Tsuyoshi Tachibana¹⁾, Tetsuya Hieda¹⁾, Akio Shimoji¹⁾, Takahiro Ishigaki¹⁾, Satoshi Sugimoto¹⁾, Tatsuya Seki¹⁾, Yasushige Shingu¹⁾, Tomonori Ooka¹⁾, Horoki Kato²⁾, Suguru Kubota¹⁾, Yoshiro Matsui¹⁾

1) Department of Cardiovascular and Thoracic Surgery, Hokkaido University,

2) Hokkaido University Hospital Emergency and Critical Care Center

There are few reports about definitive repair for pulmonary atresia and ventricular septal defect (PA/VSD) over 40 years old. A male in his 40s underwent Waterston operation for PA/VSD at the age of one. Medical follow-up was continued without definitive repair. One year ago, CT scan showed a huge right pulmonary artery aneurysm with over 90mm, and he had urgent operation, which was aneurysmectomy with graft replacement. And then, NYHA classification was 2, and catheter examination showed Qp/Qs 2.7, Rp 4.5 unit/m², LVEDVI 110.2 ml/m², LVEF 45 %, RVEDVI 79.4 ml/m², and RVEF 46 %. And more, aortic root was dilated 57 mm in diameter. So, we decided definitive repair for PA/VSD and aortic root aneurysm. Extracorporeal circulation was set up by F-F bypass. Under cardiac arrest with moderate hypothermia, we underwent VSD closure using ePTFE patch, RVOT reconstruction using composite graft with bioprosthetic valve, and aortic root replacement using Valsalva graft. Post-operative UCG showed mild AR, no residual shunt and no RVOTS without PR. He had good clinical course and discharged on POD 34. Now, he has no symptoms in six months (NYHA classification 1).

PJ3-4 両大血管右室起始症術後の左上大静脈遺残を rerouting することで酸素化改善を得た1例
A case of remarkable oxygenation improvement by rerouting persistent left superior vena cava with previous double outlet right ventricle operation.

池内 博紀¹⁾, 桜沢 政司¹⁾, 岡嶋 良和²⁾, 川副 泰隆²⁾, 立野 滋²⁾, 森島 宏子²⁾, 武智 史恵²⁾, 松尾 浩三¹⁾

1) 千葉県循環器病センター 心臓血管外科, 2) 千葉県循環器病センター 小児科

Hiroki Ikeuchi¹⁾, Masashi Kabasawa¹⁾, Yoshitomo Okajima²⁾, Yasutaka Kawasoe²⁾, Shigeru Tateno²⁾, Hiroko Morishima²⁾, Fumie Takechi²⁾, Kozo Matsuo¹⁾

1) Department of Cardiovascular Surgery, Chiba Cerebral and Cardiovascular Center,

2) Department of Pediatrics, Chiba Cerebral and Cardiovascular Center

A 42-year-old man with previous operation for double outlet right ventricle was admitted to our hospital. However persistent left superior vena cava (PLSVC) and unroofed coronary sinus had been recognized after previous operation, any treatment had not performed and cyanosis remained (SpO₂ 85%). Cardiac catheterization showed small right ventricle (%RVEDV 48%), right ventricular outflow tract (RVOT) stenosis. We assessed PLSVC rerouting and RVOT repair could be performed. PLSVC was anastomosed directly to the right atrial appendage because there were juxtaposition of atrial appendages. After weaning from cardiopulmonary bypass, right-to-left shunt through the foramen ovale and deterioration of oxygenation were revealed. Therefore, foramen ovale closure was added and improvement of oxygenation was achieved. Postoperative course was good and cyanosis improved (SpO₂ 95%). We should not overlook PLSVC because it can occur cyanotic organ damage. It is difficult to predict right ventricular capacity load after rerouting PLSVC. Because of that, gradual control of interatrial communication under repetitive assessment of right ventricular function and oxygenation are required.

PJ3-5

心外導管型フォンタンへの移行に加えて自己弁温存大動脈基部置換を併施した一例

A successful surgical case of extra-cardiac Fontan conversion combined with valve-sparing aortic root replacement for PA/IVS and aortic root aneurysm

加藤 伸康¹⁾, 橘 剛²⁾, 新井 洋輔¹⁾, 新宮 康栄¹⁾, 加藤 裕貴³⁾, 大岡 智学¹⁾, 久保田 卓¹⁾, 泉 学⁴⁾, 山澤 弘州⁴⁾, 武田 充人⁴⁾, 松居 喜郎¹⁾

1) 北海道大学大学院医学研究院 循環器呼吸器外科, 2) 神奈川県立こども医療センター 心臓血管外科,
3) 北海道大学病院 先進急性期医療センター, 4) 北海道大学大学院医学研究院 小児科

Nobuyasu Kato¹⁾, Tsuyoshi Tachibana²⁾, Yousuke Arai¹⁾, Yasushige Shingu¹⁾, Hiroki Kato³⁾, Tomonori Ooka¹⁾,
Suguru Kubota¹⁾, Gaku Izumi⁴⁾, Hirokuni Yamazawa⁴⁾, Atsuto Takeda⁴⁾, Yoshiro Matsui¹⁾

1) Department of Cardiovascular and Thoracic Surgery, Hokkaido University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Kanagawa Children's Medical Center,

3) Department of Emergency and Critical Care Center, Hokkaido University Hospital,

4) Department of Pediatrics, Hokkaido University Graduate School of Medicine

There is no report of extra-cardiac Fontan conversion (EC-FC) combined with valve-sparing aortic root replacement (David operation). He is a 24-year-old man with pulmonary atresia and intact ventricular septum (PA/IVS) after Fontan operation. He had achieved lateral tunnel Fontan operation at six years old after twice systemic pulmonary shunt procedures (one month and two years old) and bi-directional Glenn (five years old). Recently, he had a palpitation and diagnosed atrial tachycardia by electrophysiology study. And more, CT scan showed progressive aortic root aneurysm with 54mm in diameter, which had been 50mm 4 years ago. We decided EC-FC combined with David operation. Re-median sternotomy was done and cardio-pulmonary bypass was established with ascending aorta and bicaval cannulation. We used 28mm Valsalva graft for David operation, and 22mm Gore-Tex graft for EC-FC. A huge right atrium was resected and plicated, and non-functional right ventricle was also plicated. Post-operative status was stable. He was worn from respirator on POD1, and left ICU on POD2. Now he is undergoing rehabilitation for discharge.

PJ3-6

未治療単心室、巨大肺動脈瘤に対する肺動脈基部置換術、絞扼術

Pulmonary root replacement with valved conduit and conduit banding for untreated single right ventricle and huge pulmonary artery aneurysm

池内 博紀¹⁾, 花沢 政司¹⁾, 岡嶋 良知²⁾, 川副 泰隆²⁾, 立野 滋²⁾, 森島 宏子²⁾, 武智 史恵²⁾, 松尾 浩三¹⁾

1) 千葉県循環器病センター 心臓血管外科, 2) 千葉県循環器病センター 小児科

Hiroki Ikeuchi¹⁾, Masashi Kabasawa¹⁾, Yoshitomo Okajima²⁾, Yasutaka Kawasoe²⁾, Shigeru Tateno²⁾,

Hiroko Morishima²⁾, Fumie Takechi²⁾, Kozo Matsuo¹⁾

1) Department of Cardiovascular Surgery, Chiba Cerebral and Cardiovascular Center,

2) Department of Pediatrics, Chiba Cerebral and Cardiovascular Center

A 16-years-old Chinese man with untreated single ventricle visited our hospital. However he had been regarded as inoperable, his family had sought for any possibility of surgical intervention for him. After several examination at our hospital, he diagnosed single right ventricle(S, X, L), huge pulmonary artery aneurysm (76mm), patent ductus arteriosus (PDA), severe pulmonary regurgitation. Although pulmonary hypertension was revealed (118/57mmHg), pulmonary vascular resistance was calculated unexpectedly low as much as 7.3 wood unit/m². Therefore, we scheduled palliative surgery and aneurysm resection. Through a median sternotomy, PDA ligation and pulmonary artery root replacement with valved expanded polytetrafluoroethylene conduit were performed. After replacement of pulmonary artery, we gradually banded distal of conduit. Improvement of oxygenation was achieved when it banded at 55cm in diameter. Postoperative course was good and pulmonary hypertension and cyanosis improved. Three years have passed without reoperation and administration of heart failure.

PJ3-7

先天性心疾患の術後遠隔期にバルサルバ洞動脈瘤破裂を疑われた一例

A case of suspected Valsalva sinus aneurysm rupture in the postoperative period for congenital heart disease

梶沢 政司^{1,2)}, 松尾 浩三^{1,2)}, 池内 博紀¹⁾, 伊藤 貴弘¹⁾, 岡嶋 良知^{2,3)}, 川副 泰隆^{2,3)}, 森島 宏子^{2,3)}, 武智 史恵^{2,3)}, 立野 滋^{2,3)}

1) 千葉県循環器病センター 心臓血管外科, 2) 成人先天性心疾患診療部, 3) 小児科

Masashi Kabasawa^{1,2)}, Kozo Matsuo^{1,2)}, Hiroki Ikeuchi¹⁾, Takahiro Ito¹⁾, Yoshitomo Okajima^{2,3)}, Yasutaka Kawasoe^{2,3)}, Hiroko Morishima^{2,3)}, Fumie Takechi^{2,3)}, Shigeru Tateno^{2,3)}

1) Department of Cardiovascular Surgery, Chiba Cerebral and Cardiovascular Center,

2) Department of Adult Congenital Heart Disease, 3) Department of Pediatrics

The Patient was 57-year-old man. He has been performed some aortic valve surgery for uncertain congenital heart disease at 9 years old. He did not receive any follow-up after surgery. After 47 years after surgery, he consulted a near hospital because of respiratory distress and leg edema. Since abnormality of the aortic valve was suspected, he was referred to our hospital. He was diagnosed as heart failure because of Valsalva sinus aneurysm rupture by echocardiography and CT, therefore we performed surgery as early as possible. Cardiopulmonary bypass was established by femoral incision and median re-sternotomy was performed. After dissection of adhesion, the aorta was clamped and cardioplegia was injected. When the ascending aorta was transected and the lumen was observed, Dacron patch was sewn on the sino-tubular junction on the head side of non-coronary cusp, and the right atrium wall adhered to this back, penetrated at two places. The leaflets and commissures had marks of repair, both of which were thickened. modified Bentall procedure was performed (26mm Triplex + 23mm Magna Ease). Postoperative course was good.

PJ4-1

先天性一尖弁に伴う高度大動脈閉鎖不全症に対して弁形成術を施行した2例

Two cases who underwent aortic valve repair for severe aortic regurgitation associated with congenital unicuspid aortic valve

川村 廉, 儀武 路雄, 松村 洋高, 山城 理仁, 中尾 充貴, 宇野 吉雅, 篠原 玄, 木南 寛造, 長堀 隆一, 坂東 興, 森田 紀代造, 國原 孝

東京慈恵会医科大学附属病院 心臓外科

Ren Kawamura, Michio Yoshitake, Youkou Matsumura, Masahito Yamashiro, Mitsutaka Nakao, Yoshimasa Uno,

Gen Shinohara, Hiroo Kinami, Ryuichi Nagahori, Ko Bando, Kiyozo Morita, Takashi Kuniyara

Department of Cardiac Surgery, Jikei University School of Medicine

Aortic valve replacement has been the standard surgical treatment for aortic regurgitation, however, life-long anticoagulation after mechanical valve replacement for young patients appears major concern. Recently aortic valve repair has emerged as an attractive alternative to eliminate anticoagulation therapy.

We experienced 15- and 23-year-old male patients with severe aortic regurgitation due to the congenital unicuspid aortic valve who underwent aortic valve repair. Echocardiography showed LVDd/Ds were 59.2/39.2 mm in case 1 and 69.8/50.7 mm in case 2. Bicuspidization was performed by using glutaraldehyde-treated autologous pericardial patches. External suture annuloplasty and replacement of the ascending aorta was also performed. Postoperative echocardiography showed trivial and mild regurgitation without relevant stenosis. LVDd/Ds were 44.5/32.1 mm in case 1 and 59.3/51.5 mm in case 2. Their postoperative courses were uneventful and they were discharged home at the 16th and 15th postoperative day.

Long-term durability of bicuspidization of the unicuspid aortic valve using an autologous pericardium is unclear. Careful follow-up survey is thus mostly important.

PJ4-2 成人期に手術介入した三心房心の2例

Surgical repair of cor triatriatum sinister in adulthood: case report

岩瀬 友幸¹⁾, 小泉 淳一¹⁾, 上田 寛修²⁾, 高橋 信³⁾, 小山 耕太郎³⁾, 金 一¹⁾

1) 岩手医科大学 心臓血管外科, 2) 岩手医科大学 循環器内科, 3) 岩手医科大学 循環器小児科

Tomoyuki Iwase¹⁾, Jyunichi Koizumi¹⁾, Hironobu Ueda²⁾, Shin Takahashi³⁾, Kotaro Oyama³⁾, Hajime Kin¹⁾

1) Department of Cardiovascular Surgery, Iwate medical University, 2) Department of Cardiology, Iwate Medical University, 3) Division of Pediatric Cardiology, Iwate Medical University

Case 1. 33 year-old female. Arrhythmia was pointed out when she was 16 years old in medical checkup. Cor triatriatum was point out by echocardiography (but no other congenital heart defect). PCWP was elevated up to 16mmHg and further elevation of PCWP was measured after infusion of beta stimulator. We recommended the surgical treatment, but she didn't accept. There was no symptom during the perinatal period at age 32 years. After the delivery, we told the risk of arrhythmia, thrombotic disease, and heart failure, and then she accepted the surgical treatment. We performed membrane resection.

Case 2. 19 year-old male. He had chest pain once a month from junior high school, and the symptom wasn't improved recently. There was no abnormality in ECG, but had an obstructive membrane in left atrium in echocardiography. PCWP was elevated up to 22mmHg without any other abnormality in Catheter exam. He accepted the surgical repair, performed excision of cor triatriatum membrane.

Summary. Cor triatriatum is rare congenital heart disorder. In these two cases, there was no complication after surgical treatment. It is necessary to performed proper diagnosis and appropriate operations.

PJ4-3 成人Fontan術後患者に対する積極的な在宅非侵襲性陽圧換気療法の効果

The aggressive treatment to sleep apnea with in-home Noninvasive Positive Pressure Ventilation improves the hemodynamics of Fontan patients.

杉谷 雄一郎¹⁾, 宗内 淳¹⁾, 藤井 俊輔¹⁾, 松岡 良平¹⁾, 川口 直樹¹⁾, 渡邊 まみ江¹⁾, 安東 勇介²⁾, 落合 由恵²⁾

1) 地域医療機能推進機構九州病院 小児科, 2) 地域医療機能推進機構九州病院 心臓血管外科

Yuichiro Sugitani¹⁾, Jun Muneuchi¹⁾, Shunsuke Fujii¹⁾, Ryohei Matsuoka¹⁾, Naoki Kawaguchi¹⁾, Mamie Watanabe¹⁾, Yusuke Ando²⁾, Yosie Ochiai²⁾

1) Department of Pediatrics, Japan Community Healthcare Organization Kyushu Hospital, 2) Department of CVS, Japan Community Healthcare Organization Kyushu Hospital

Background: Sleep apnea syndrome (SAS) is associated with cardiovascular adverse events. We present 3 cases of adult patients after Fontan operation in whom aggressive introductions of noninvasive positive pressure ventilation (NIPPV) improve the hemodynamic status.

Case1: A 16-year-old man who underwent Fontan operation had dyspnea on exertion. He gained 13kg in weight for 3 years. He was diagnosed as SAS in polysomnography. He had a 15kg weight loss and cardiac index by MRI increased from 2.3 to 3.1L/min/m² by treatment with in-home NIPPV.

Case2: A 42-year-old man with single ventricle gained a 13kg weight after Fontan completion. He also had sleep apnea, headache and fatigue in the day time. Although the criteria of SAS was not fulfilled, the treatment to sleep disorder with in-home NIPPV started and subsequently symptoms improved.

Case3: A 34-year-old man with Fontan circulation gained a 11kg weight for three years. He developed cerebellar infarction. At hospitalization, he was diagnosed as SAS. He had PSVT during sleep. However, PSVT was not detected after NIPPV started. Conclusion: The treatment to SAS with in-home NIPPV improved the hemodynamics and QOL for Fontan patients.

PJ4-4

成人期Fontan手術後合併の肺動静脈瘻に対する治療 ～肝静脈還流の修正～

Redirection of hepatic venous return for treatment of pulmonary arteriovenous malformations after adult Fontan completion

稲熊 洸太郎¹⁾, 坂崎 尚徳¹⁾, 豊田 直樹¹⁾, 石原 温子¹⁾, 前田 登史²⁾, 加藤 おと姫²⁾, 渡辺 謙太郎²⁾, 植野 剛²⁾, 吉澤 康祐²⁾, 大野 暢久²⁾, 藤原 慶一²⁾, 板谷 慶一³⁾

1) 兵庫県立尼崎総合医療センター 小児循環器内科, 2) 兵庫県立尼崎総合医療センター 心臓血管外科,
3) 京都府立医科大学 心臓血管外科・心臓血管血流解析学講座

Kotaro Inaguma¹⁾, Hisanori Sakazaki¹⁾, Naoki Toyoda¹⁾, Haruko Ishihara¹⁾, Toshi Maeda²⁾, Otohime Katou²⁾, Kentro Watanabe²⁾, Go Ueno²⁾, Kosuke Yoshizawa²⁾, Nobuhisa Oono²⁾, Keiichi Fujiwara²⁾, Keiichi Itatani³⁾

1) Hyogo Prefectural Amagasaki General Medical Center, Pediatric Cardiology,
2) Hyogo Prefectural Amagasaki General Medical Center, Cardiovascular Surgery,
3) Kyoto Prefectural University of Medicine, Department of Cardiovascular Surgery

Pulmonary arteriovenous malformations (PAVM) can develop after Fontan completion when there is unequal distribution of hepatic venous return between the two lungs. This results in progressive cyanosis and the need for reintervention. We retrospectively reviewed the clinical data on two patients.

Case 1 is a 46-year old female with polysplenia, SRV, IVC interruption with azygos continuation, right SVC and left hepatic vein. At 36 years of age, Fontan completion was performed. 10 years later, cyanosis worsened due to right sided PAVM. Surgical translocation of SVC was performed to correct the distribution of hepatic venous flow.

Case 2 is a 49-year old female with MA, TGA, VSD, PS. At 40 years of age, Fontan completion was performed with fenestrated extra-cardiac conduit. 8 years later, cyanosis worsened due to left sided PAVM. Percutaneous occlusion of fenestration was performed to redirect hepatic venous flow.

Redirection of hepatic venous return was successful in two patients with improvement in oxygen saturation. There were no recorded short-term complications. Furthermore studies are needed to determine the efficacy.

PJ4-5

Fontan術後遠隔期に鑄型気管支炎を発症した一例

A case of plastic bronchitis late after Fontan procedure

石北 綾子¹⁾, 坂本 一郎¹⁾, 梅本 真太郎¹⁾, 永田 弾²⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 小児科

Ayako Ishikita¹⁾, Ichiro Sakamoto¹⁾, Shintaro Umemoto¹⁾, Hazumu Nagata²⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiology, Kyushu University, 2) Department of Pediatrics, Kyushu University

Plastic bronchitis is characterized by the formation of exudative air way cast. It is rare but life-threatening complications after Fontan procedure.

A 37-year-old male with DORV and pulmonary stenosis underwent total cardiopulmonary connection with lateral tunnel (LT) at age 15. Leakage from lateral tunnel had observed just after the operation, but he was asymptomatic. His CVP was 10 mmHg at age 33. He suffered from hemoptysis at age 35 that required coil embolization and withdrawal of anticoagulation. At age 36, tracheobronchial tree cast was discharged. A pathological study indicated the cast was non-inflammatory mutin-rich sputa and he was diagnosed as plastic bronchitis. His CVP became 20mmHg and cardiac output decreased to 2.1 L/min/m². Except known-leakage from LT to SA, any indications for surgical therapy could not be observed. Diuretics and an anticoagulant therapy were restarted.

The medical literatures for plastic bronchitis are limited. Here we report a case of plastic bronchitis late after Fontan procedure.

PJ4-6 PAVSD術後遠隔期にVT/VFを合併した1例**Cardiac arrest and ventricular fibrillation in a woman with repaired pulmonary atresia with ventricular septal defect: a case report**横濱 ふみ¹⁾, 杜 徳尚¹⁾, 赤木 貞治¹⁾, 衛藤 弘城²⁾, 黒子 洋介²⁾, 小谷 恭弘²⁾, 笠原 真悟²⁾, 伊藤 浩¹⁾

1) 岡山大学 循環器内科, 2) 岡山大学 心臓血管外科

Fumi Yokohama¹⁾, Norihisa Toh¹⁾, Teiji Akagi¹⁾, Koki Eto²⁾, Yosuke Kuroko²⁾, Yasuhiro Kotani²⁾, Shingo Kasahara²⁾, Hiroshi Ito¹⁾

1) Department of Cardiology, Okayama University, 2) Department of Cardiovascular Surgery, Okayama University

A 34-year-old woman with right ventricular outflow (RVOT) reconstruction and unifocalization for pulmonary atresia with ventricular septal defect (PA/VSD) and major aortopulmonary collateral arteries was referred to our institution for further treatment. She had been treated for heart failure and pulmonary hypertension and was admitted to the local hospital after out-of-hospital cardiac arrest. Bystander cardiopulmonary resuscitation was performed followed by automated external defibrillator shocks with return of spontaneous circulation. After the initial treatment, she was transferred to our hospital. Cardiac magnetic resonance imaging demonstrated severe pulmonary regurgitation with a regurgitant fraction of 70%, dilated right ventricular (RV) with an end-diastolic volume index of 189 ml/m², and decreased RV ejection fraction of 14%. Cardiac catheterization showed elevated systolic pulmonary artery pressure of 50 mmHg. She underwent pulmonary valve replacement and RVOT reconstruction followed by implantable cardioverter defibrillator implantation and adjustment of pulmonary hypertension drugs. In this report, we will review outcomes after the surgical repair of PA/VSD.

PJ4-7 フォンタン術後遠隔期に急性心筋梗塞を来した1症例**A Rare Case of Acute Myocardial Infarction (AMI) in a Patient with Fontan Circulation**島袋 篤哉¹⁾, 佐藤 誠一¹⁾, 西畑 昌大¹⁾, 塚原 正之¹⁾, 内田 英利¹⁾, 竹蓋 清高¹⁾, 中矢代 真美¹⁾, 宮城 文音²⁾, 勝連 朝史²⁾, 平良 良集²⁾, 槇田 徹²⁾, 大城 克彦²⁾, 宮良 高史²⁾, 田場 洋二²⁾, 當真 隆²⁾

1) 沖縄県立南部医療センター・こども医療センター 小児循環器内科,

2) 沖縄県立南部医療センター・こども医療センター 循環器内科

Atsuya Shimabukuro¹⁾, Seiichi Sato¹⁾, Masahiro Nishibata¹⁾, Masayuki Tsukahara¹⁾, Hidetoshi Uchida¹⁾, Kiyotaka Takefuta¹⁾, Mami Nakayashiro¹⁾, Ayane Miyagi²⁾, Tomofumi Katsuren²⁾, Ryosyu Taira²⁾, Toru Makita²⁾, Katsuhiko Ooshiro²⁾, Takashi Miyara²⁾, Youji Taba²⁾, Takashi Touma²⁾

1) Department of Pediatric Cardiology, Okinawa Prefectural Nanbu Children Medical Center,

2) Department of Cardiology, Okinawa Prefectural Nanbu Medical Center

Introduction: It is the general belief that the serum-lipid levels of cyanotic congenital heart disease patients including patients with Fontan circulation are low and therefore there is low risk of developing ischemic heart disease.

Case: We report a 42-year-old man with criss-cross heart, pulmonary atresia and straddling mitral valve who underwent APC-Fontan in childhood and TCPC conversion in Adulthood. He presented with two days of intermittent chest pain and the initial 12-lead ECG revealed ST segment depression in V1-2. He was emergently transported to the cardiac catheterization laboratory where coronary angiography revealed severe stenosis of left circumflex branch due to atherosclerotic plaque and was recovered by stent implantation. He had a past history of protein-losing enteropathy for which he was taking oral glucocorticoid steroids for over 5 years. Therefore, the cause for coronary atherogenesis could be considered related to side effect of steroids.

Conclusion: We should be careful of chest pain even in Fontan patients on long-term glucocorticoids, which could cause acute myocardial infarction associated with coronary artery atherogenesis and vascular remodeling.

PJ4-8

心房頻拍による心不全をきたし救急受診したAPCフォンタン術後ドロップアウト症例

A case of lost to follow up after atriopulmonary connection (APC) Fontan surgery who resulted in severe heart failure due to atrial tachycardia (AT)

戸田 孝子¹⁾, 喜瀬 広亮¹⁾, 河野 洋介¹⁾, 吉沢 雅史¹⁾, 小泉 敬一¹⁾, 星合 美奈子²⁾, 犬飼 岳史¹⁾

1) 山梨大学医学部 小児科, 2) 山梨県立中央病院 小児科

Takako Toda¹⁾, Hiroaki Kise¹⁾, Yousuke Kouno¹⁾, Masashi Yoshizawa¹⁾, Keiichi Koizumi¹⁾, Minako Hoshiai²⁾, Takeshi Inukai¹⁾

1) Department of Pediatrics, Yamanashi University, 2) Department of Pediatrics, Yamanashi Prefectural Central Hospital

Fontan operation is associated with long-term various complications.

Case: 30 years old female with tricuspid atresia. She had APC Fontan operation at 3 years old. She stopped going to the hospital after 20 years old. At 30 years old, she was taken to emergency visit of our hospital by ambulance with palpitation and dyspnea. Electrocardiography showed AT with heart rate 160 bpm. Central venous pressure (CVP) was elevated to 28 mmHg. Although she was administered antiarrhythmic drugs, AT did not stop, so she was achieved by synchronized direct-current shocks. An echocardiography revealed right atrial gross dilation with large thrombus. She was administered heparin, warfarin and aspirin then thrombus disappeared. CVP was still elevated, macitentan and tadalafil were administered. After that, on cardiac catheterization, mean pulmonary artery pressure was decreased to 10mmHg, and pulmonary resistance was 2.1WU. Finally, she underwent a conversion operation to total cavopulmonary connection (TCPC).

Conclusion: Continuous follow-up is important for patients after Fontan operation. We should educate patients from childhood to prevent dropout during adulthood.

PJ4-9

Fontan術後肝障害における動的中心静脈圧評価の重要性

The venous pressure during exercise in Fontan associated liver disease

齋藤 義弘¹⁾, 相馬 桂¹⁾, 齋藤 暁人¹⁾, 稲葉 敏郎¹⁾, 網谷 英介¹⁾, 犬塚 亮²⁾, 八尾 厚史¹⁾, 小室 一成¹⁾

1) 東京大学医学部附属病院 循環器内科, 2) 東京大学医学部附属病院 小児科

Yoshihiro Saito¹⁾, Katura Souma¹⁾, Akito Saito¹⁾, Toshiro Inaba¹⁾, Eisuke Amiya¹⁾, Ryou Inutuka²⁾, Atushi Yao¹⁾, Issei Komuro¹⁾

1) Department of Cardiovascular Medicine, Tokyo University, 2) Department of Pediatrics, Tokyo University

Fontan associated liver disease (FALD) is the serious complication in patients living long under Fontan circulation. High central venous pressure (CVP) has been thought to be a risk for FALD progression, while only long exposure to Fontan circulation has been proved to be the risk. Even in the patients whose CVP at rest is not so high, FALD could progress. Whereas, we evaluated CVP during exercise in 2 cases of FALD.

Case 1 is a 22-year-old male with single right ventricle. TCPC-Fontan surgery was performed in infancy. Since his fibro-scan score significantly increased from 20 to 29, we performed hemodynamic analysis showing that IVC pressure and PAP increased from 13 to 23 and from 7 to 13 mmHg, respectively, with a pressure gradient between IVCP and PAP during exercise.

Case 2 is a 20-year-old female with Pulmonary Atresia. TCPC-Fontan surgery was performed in infancy. Abdominal echocardiogram showed FS of 23.5 and some space occupied lesions. Hemodynamics analysis showed that IVCP and PAP equally elevated from 12 to 21mmHg during exercise.

High CVP during exercise might be a risk factor for FALD progression, although we need more numbers of data and adjusted control patients.

PJ5-1

成人先天性心疾患患者が認識する学校生活における支援

Recognition of Support in School Life with Adult Congenital Heart Disease Patients

仁尾 かおり¹⁾, 藤澤 盛樹²⁾, 原口 昌宏³⁾

1) 三重大学大学院医学系研究科 看護学専攻, 2) 千里金蘭大学, 3) 東京医療保健大学

Kaori Nio¹⁾, Seiki Fujisawa²⁾, Masahiro Haraguchi³⁾

1) Course of Nursing Science, Mie University Graduate School of Medicine, 2) Senri Kinran University,

3) Tokyo Healthcare University

【目的】 学校生活における重要他者の支援に対する ACHD 患者の認識や期待、支援の現状を明らかにする。

【方法】 ACHD 患者7名を対象に、(1)個別インタビュー、(2)友人と合同のフォーカス・グループインタビューを行い、分析にはKJ法を用いた。

【結果】 研究参加者は男性2名・女性5名、27～39歳(平均30.1歳)、病名はSV、TOF各2名、DORV、ECD、VSD・PH各1名であった。

学校生活では【病気を理解し周囲へ意思表示するのは<自分>】が基盤である。【<自分>の言葉による病気の開示】により【学校生活に参加できるための先生の采配】がなされる。【自らの言葉による病気の開示】は当事者と友人双方が信頼し合い【良い友達を選択】という好循環となり、【友達からの按配良い気配り】につながる。また【友達からの救いの一言】により体調悪化を免れることもある。一方、【内部障害の理解されにくさ】により、【病気説明の難しさ】を感じ、支援に対するニードと友人や先生に食い違いが生じることで、【ニードとサポートのバランスが難しい】と認識していた。

【考察】 学校生活での支援は、「<自分>がカギになるが難しい」と認識していると考えられた。当事者自身が病気を理解し、病気のことを開示し、支援に対する意思表示をすることで、友達や先生との良い循環が生まれるが、理解されにくい内部障害であり、支援に対するニードと実際のサポートにはアンバランスが生じやすいと考える。

PJ5-2

心房中隔欠損症のデバイス閉鎖における医療連携

Inter-hospital management of ASD closure procedure with occlusion device between general and pediatric hospitals

狩野 実希, 矢野 弘崇, 新田 義一, 加藤 駿一, 高野 寿一, 池ノ内 孝, 村田 和也, 松田 隼治, 高宮 智正,

加藤 信孝, 稲村 幸洋, 根木 謙, 佐藤 明, 大和 恒博, 松村 穰, 新田 順一

さいたま赤十字病院 循環器内科

Miki Kanoh, Hirotaka Yano, Giichi Nitta, Shunichi Kato, Toshikazu Kono, Takashi Ikenouchi, Kazuya Murata,

Junji Matsuda, Tomomasa Takamiya, Nobutaka Kato, Yukihiro Inamura, Ken Negi, Akira Sato, Tsunehiro Yamato,

Yutaka Matsumura, Junichi Nitta

Department of Cardiology, Saitama Red Cross Hospital

Atrial septal defect (ASD) is one of congenital heart diseases that is often diagnosed in patients' adulthood and percutaneous device occlusion with AMPLATZER™ Septal Occluder (ASO) is now a common practice for closure. This procedure is highly specialized and is only allowed in the carefully selected hospitals where the standards for facilities and operators defined by two Japanese Intervention Societies that are JPEC and CVIT, are met and our hospital is indeed disqualified. Fortunately, a pediatric hospital adjacent to our hospital is granted a qualification for this procedure but the most of its patients tends to be young in general and experiences for adult patients are scarce. To provide the treatments for our adult patients, two hospitals now operate an inter-hospital management program, in which catheter procedures are performed in the pediatric hospital and then patients are transferred to our hospital for postoperative care. This unique program allows patients in our neighborhood not only to seek a tertiary care center far away from their home but also have convenient medical access nearby.

PJ5-3

左卵巣切除に踏み切ったファロー四徴症の術後例：多診療科連携の問題点

Who should make decision?: A female case with chronic ovarian hemorrhage after repair of ToF.

手島 秀剛¹⁾, 松口 一道²⁾, 本村 秀樹³⁾, 永田 弾⁴⁾

1) 市立大村市民病院 小児科, 2) 市立大村市民病院 婦人科, 3) 長崎医療センター 小児科, 4) 九州大学 小児科

Hidetaka Teshima¹⁾, Kazumichi Matsuguchi²⁾, Hideki Motomura³⁾, Hazumu Nagata⁴⁾

1) Division of Pediatrics, Omura Municipal Hospital, 2) Division of Gynecology, Omura Municipal Hospital, 3) Division of Pediatrics, Nagasaki Medical Center, 4) Department of Pediatrics, Kyushu University

The case is 24 years old, female with 22q11.2 deletion syndrome. She had repaired her cardiac anomaly (Tetralogy of Fallot with pulmonary atresia and MAPCA) at the age of 10 and had been followed up by pediatricians over a long term because of residual VSD and aortic/pulmonary insufficiency. Since adolescents often complained abdominal pain and lumbago, she was consulted with general internal medicine, urology and gynecology department, then diagnosed with chronic left ovarian bleeding. She was to be followed up, but her quality of life had declined because of sustained symptoms. So finally primary care physician, a pediatrician, recommended surgical intervention, and then left ovariectomy was done. Her symptoms subsided and QOL was remarkably improved. The role of the primary care doctor is important in medical treatment involving many medical departments.

PJ5-4

Amplatzer Vascular Plug での肺動静脈瘻塞栓後に胸膜炎を併発したフォンタン術後の2例
Embolization of Pulmonary Arteriovenous Fistula (PAVF) with Amplatzer Vascular Plug (AVP)
Caused Pleurisy in Patients after Fontan procedure

中島 公子, 大内 秀雄, 北野 正尚, 根岸 潤, 黒崎 健一

国立循環器病研究センター

Kimiko Nakajima, Hideo Ohuchi, Masataka Kitano, Jun Negishi, Kenichi Kurosaki
Department of Pediatrics, National Cardiovascular Center

Background: PAVF in patients after Fontan procedure require intervention for prevention to complication. We report two cases of Fontan patients with pleurisy due to embolization of PAVF with AVP.

Case1: The patient underwent Fontan operation at four years of age. 23 years following the operation, the patient developed progressive cyanosis and heart failure. Catheterization confirmed the presence of PAVF as the cause of cyanosis and heart failure. Embolization of PAVF with AVP was performed successfully. A week later, she developed pleural effusion and painful pleurisy and needed a chest cavity drainage.

Case2: The patient with heterotaxy underwent Fontan operation at nine years of age. 27 years following the operation, the patient developed progressive cyanosis due to PAVF. Embolization of PAVF with AVP was performed successfully. Three days after, he developed pleural effusion and painful pleurisy and needed addition of diuretic. He used oral opioid pain medication.

Discussion: In embolization of PAVF, pleuritic chest pain, the most common procedural complication (5-13 percent), is usually self-limiting, but occasionally a analgesic is necessary for more protracted pain.

PJ5-5

A型大動脈弓離断, 完全大血管転位II型に対する大血管スイッチ術後遠隔期のBentall手術 Bentall procedure after the arterial switch operation for interrupted aortic arch type A and transposition of the great artery type II

落合 由恵, 安東 勇介, 浮池 宣史, 馬場 啓徳, 久原 学, 中田 悠介, 徳永 滋彦
JCHO九州病院 心臓血管外科

Yoshie Ochiai, Yusuke Ando, Yoshifumi Fuke, Hironori Baba, Manabu Hisahara, Yusuke Nakata, Shigehiko Tokunaga
Department of Cardiovascular Surgery, JCHO Kyushu Hospital

We report a case of Bentall procedure following the arterial switch operation for interrupted aortic arch type A (IAA-A) and transposition of the great artery type II (TGA II). On postnatal day 4, the patient underwent Blalock-Park arch plasty and pulmonary artery (PA) banding. Subsequently, at 8 months of age, he underwent arterial switch operation with Lecompte maneuver and closure of the ventricular septal defect. He developed aortic regurgitation (AR) gradually. Because of the progression of moderate AR at 21 years of age, he underwent aortic valve replacement (AVR) using SJM 27 mm and enlargement of the main PA. He remained in a good clinical condition. However, he developed a large aortic root aneurysm at 73 mm in size 10 years after the AVR. Therefore, we performed the Bentall procedure with Carbomedics 25 mm with Carbo-Seal Valsalva graft 28 mm at 31 years of age on the third sternotomy incision. We exposed the aortic root by transecting the right PA and re-excised the left and right coronary bottoms. Because the large aortic root aneurysm was replaced with the graft, the right PA was reconstructed by direct re-anastomosis.

PJ5-6

突然死回避のために外科的治療が必要な大動脈壁内走行を伴う左冠動脈起始異常の2症例 Two cases of anomalous aortic origin of a coronary artery with an intramural course which need surgical repair to avoid sudden death

辻 龍典¹⁾, 小谷 恭弘¹⁾, 迫田 直也¹⁾, 田井 龍太¹⁾, 小林 泰幸¹⁾, 川田 幸子¹⁾, 堀尾 直裕¹⁾, 後藤 拓弥¹⁾, 黒子 洋介¹⁾, 新井 禎彦¹⁾, 笠原 真悟¹⁾, 杜 徳尚²⁾, 赤木 禎治²⁾, 伊藤 浩²⁾

1) 岡山大学医学部附属病院 心臓血管外科, 2) 岡山大学医学部附属病院 循環器内科

Tatsunori Tsuji¹⁾, Yasuhiro Kotani¹⁾, Naoya Sakota¹⁾, Ryuuta Tai¹⁾, Yasuyuki Kobayashi¹⁾, Sachiko Kawada¹⁾, Naohiro Horio¹⁾, Takuya Gotou¹⁾, Yousuke Kuroko¹⁾, Sadahiko Arai¹⁾, Sinngo Kasahara¹⁾, Norihisa Tou²⁾, Sadaharu Akagi²⁾, Hiroshi Itou²⁾

1) Department of Cardiovascular Surgery, Okayama University, 2) Department of Cardiovascular Medicine, Okayama University

Early detection of congenital coronary anomalies by mass electrocardiographic screening of school-aged children is very hard. However, congenital coronary anomalies are recognized as the most important causes of exercised-induced sudden cardiac death of the young after cardiomyopathies. Anomalous aortic origin of left main coronary artery (AAOLCA) is said to be about 5-7% of congenital coronary anomalies. It is necessary to recognize that AAOLCA especially with an intramural course is a high risk of myocardial ischemia or sudden cardiac death. Therefore, surgical repair should be chosen to avoid myocardial ischemia or sudden cardiac death because the risk is unpredictable even if patients have no subjective symptom or no ischemic finding by exercise electrocardiographic. We describe two cases of AAOLCA with an intramural course having avoided sudden death by surgical repair. Both cases needed much time to be operated not being correctly diagnosed even though the patients had had a detailed cardiac examination for fainting. Correct diagnosis and appropriate surgical repair are important for AAOLCA with an intramural course to avoid myocardial ischemia or sudden cardiac death.

PJ5-7

開腹を余儀なくされたファロー四徴症根治術後患者に対する腹腔鏡下肝切除術の麻酔経験
Anesthetic management of laparoscopic hepatectomy which needed to be converted to open surgery in a patient with repair of tetralogy of Fallot.

住江 誠¹⁾, 帯刀 英樹²⁾, 塩瀬 明²⁾, 辛島 裕士³⁾

1) 九州大学病院 手術部, 2) 九州大学病院 心臓血管外科, 3) 九州大学大学院医学研究院 麻酔・蘇生学

Makoto Sumie¹⁾, Hideki Tatewaki²⁾, Akira Shiose²⁾, Yuji Karashima³⁾

1) Operating Rooms, Kyushu University Hospital, 2) Department of Cardiovascular Surgery, Kyushu University Hospital, 3) Department of Anesthesiology and Critical Care Medicine, Graduate School of Medicine

Case Presentation: A 54-year-old female patient was scheduled for laparoscopic hepatectomy for hepatocellular carcinoma. The medical history included corrective repair of tetralogy of Fallot at 7 years of age and pulmonary valve replacement at 40 years of age. The preoperative examination showed severe tricuspid regurgitation and moderate pulmonary valve regurgitation. General anesthesia was induced successfully and maintained with desflurane and remifentanyl under standard monitoring, direct radial artery pressure and central venous pressure (CVP). As soon as the intraperitoneal examination started under pneumoperitoneum, conversion from laparoscopic to open surgery was decided because of the congestive liver with CVP as high as 15-20mmHg. Although CVP was decreased to 13-17mmHg with open surgery, massive bleeding as much as 3,000ml occurred during liver resection which was successfully treated without any critical event.

Conclusions: In this case, elevated CVP interfered with the strategy of surgery and caused massive bleeding. More proactive intervention, such as administration of nitric oxide, nitroglycerin, and PDE III inhibitor could have avoided these drawbacks.

PJ5-8

Scimitar 症候群亜型に対する低侵襲外科治療の経験
Minimally Invasive Cardiac Surgery for a Patient with Scimitar Syndrome

仁田 学¹⁾, 木野 旅人¹⁾, 松本 祐介¹⁾, 寺中 紗絵¹⁾, 鍵本 美奈子¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 細田 順也¹⁾, 重永 豊一郎¹⁾, 上村 大輔¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 石川 善啓²⁾, 町田 大輔^{1,3)}, 益田 宗孝³⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・呼吸器外科, 3) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Manabu Nitta¹⁾, Tabito Kino¹⁾, Yusuke Matsumoto¹⁾, Sae Teranaka¹⁾, Minako Kagimoto¹⁾, Kiwamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Junya Hosoda¹⁾, Atsuichiro Shigenaga¹⁾, Daisuke Kamimura¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Yoshihiro Ishikawa²⁾, Daisuke Machida^{1,3)}, Munetaka Masuda³⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine, 2) Department of Thoracic Surgery, Yokohama City University Graduate School of Medicine, 3) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine

Background: Scimitar syndrome with "dual drainage", anomalous vein connection to inferior vena cava (IVC) and left atrium (LA) is called "scimitar variant".

Case: A 19-year-old female suffered from hemoptysis and was referred to our hospital. At the age of 16, she was diagnosed as scimitar syndrome in another hospital. The cardiac computed tomography revealed scimitar vein (SV) drained to IVC. In addition both upper and lower right PVs also drained to LA, suggesting "scimitar variant". Cardiac catheterization demonstrated normal pulmonary artery pressures (PAp) with a calculated Qp/Qs of 1.7. In general, surgical re-routing of the PVs to the LA is recommended. However in our case with dual drainage, just occlusion of the SV was thought to be effective. The balloon occlusion test of the SV demonstrated insignificant hemodynamic changes; right PA wedge pressure from 11 to 14 mmHg and mean right PAp from 16 to 15mmHg. Therefore video-assisted thoracoscopic ligation of scimitar vein, minimally invasive surgery was successfully performed.

Conclusion: Minimally invasive cardiac surgery can be an another treatment option in the patient with "scimitar variant".

PJ5-9

大動脈拡張を伴った大動脈二尖弁症例の妊娠例

A Case of Pregnant Patient with Bicuspid Aortic Valve and Dilated Ascending Aorta

小坂橋 俊美¹⁾, 藤田 鉄平¹⁾, 郡山 恵子¹⁾, 前川 恵美¹⁾, 望月 純子²⁾, 海野 信也²⁾, 阿古 潤哉¹⁾

1) 北里大学医学部 循環器内科学, 2) 北里大学医学部 産科学

Toshimi Koitabashi¹⁾, Teppei Fujita¹⁾, Keiko Kooriyama¹⁾, Emi Maekawa¹⁾, Junko Mochiduki²⁾, Nobuya Unno²⁾, Junya Ako¹⁾

1) Department of Cardiovascular Medicine, Kitasato University School of Medicine,

2) Department of Obstetrics, Kitasato University School of Medicine

Background: Ascending aortic dilatation is important in bicuspid aortic valve (BAV) disease, with increased risk of aortic dissection. Likewise, pregnancy is known as a risk factor of the aortic dissection; pregnancy itself is linked to a 25-fold increased risk of aortic dissection among young women. However, the impact of the pregnancy in the BAV patients is unclear, and neither risk stratification nor the management for the BAV patients with pregnancy are established.

Case: A 37-year-old pregnant woman with BAV was consulted to our section for the management of the dilated ascending aorta which is measured as 47 mm by transthoracic echocardiography. Her systolic blood pressure was around 90 mmHg without medications and had been stabled during pregnancy. The aortic diameter was assessed frequently by echocardiography and remained unchanged. In 39 weeks of gestation, she delivered by the painless vaginal childbirth method without any problem. The postpartum was uneventful.

Conclusion: We experienced a case of BAV patient with dilatation of ascending aorta successfully delivered without surgical procedure.

PJ6-1

Discrete Type Subaortic StenosisとS字状心室中隔とによって引き起こされた左室流出路狭窄

Left Ventricular Outflow Tract Obstruction Induced by Sigmoid Septum in Addition to Discrete Type Subvalvular Aortic Stenosis

野田 光里¹⁾, 仁田 学¹⁾, 木野 旅人¹⁾, 松本 祐介¹⁾, 寺中 紗絵¹⁾, 鍵本 美奈子¹⁾, 岩田 究¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 細田 順也¹⁾, 重永 豊一郎¹⁾, 上村 大輔¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科

Hikari Noda¹⁾, Manabu Nitta¹⁾, Tabito Kino¹⁾, Yusuke Matsumoto¹⁾, Sae Teranaka¹⁾, Minako Kagimoto¹⁾, Kiwamu Iwata¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Junya Hosoda¹⁾, Atsuihiro Shigenaga¹⁾, Daisuke Kamimura¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University School of Medicine,

2) Department of Cardiovascular Surgery, Yokohama City University School of Medicine

Background: Sigmoid septum is a non-negligible cause of left ventricular outflow tract obstruction (LVOTO).

Case: A 69-year-old female presented with progressive dyspnea on exertion and was referred to our department. At the age of 63, she was diagnosed as hypertrophic obstructive cardiomyopathy (HOCM) with peak pressure gradient (PG) of 67mmHg. The recent transthoracic echocardiogram revealed peak PG of 105mmHg and moderate aortic regurgitation, which suggested worsening LVOTO. We also found a membranous ridge tissue just below the aortic valve, which narrowed LVOT. In addition, a diminished angle between ascending aorta and interventricular septum, so called sigmoid septum was shown. Therefore we diagnosed as not HOCM but discrete type subvalvular aortic stenosis with sigmoid septum. After the surgical resection of subaortic septal muscle in addition to mechanical aortic valve replacement, LVOTO improved, just 10mmHg of PG. The biopsy specimen of the resected muscle showed myocardial hypertrophy with interstitial fibrosis, which was discordant with HCM.

[Conclusion] Sigmoid septum is an important cause of LVOTO. It is necessary to observe carefully about the morphology of LVOT.

PJ6-2 CABG合併ASD閉鎖術後経過において肺高血圧症増悪により重篤な転機を辿った一例 A case of severe outcome for pulmonary hypertension worsening remarkably in post-operation of CABG and ASD closure.

加藤 駿一, 矢野 弘崇, 新田 義一, 高野 寿一, 池ノ内 孝, 村田 和也, 松田 準治, 狩野 実希, 加藤 信孝, 高宮 智正, 稲村 幸洋, 根木 謙, 佐藤 明, 大和 恒博, 松村 穰, 新田 順一
さいたま赤十字病院 循環器内科

Shunichi Kato, Hiroataka Yano, Giichi Nitta, Toshikazu Kouno, Takashi Ikenouchi, Kazuya Murata, Junji Matsuda, Miki Kanou, Nobutaka Kato, Tomomasa Takamiya, Yukihiko Inamura, Ken Negi, Akira Sato, Tsunehiro Yamato, Yutaka Matsumura, Junichi Nitta
Department of Cardiology, Japanese Red Cross Saitama Hospital

A 69 year-old female patient started to undergo dialysis for chronic Glomerular nephritis at 43 year-old. and she presented with hypotension during dialysis and dyspnea for pulmonary congestion. Her Right heart system was expanded, her atrial septum was deficit, and ejection fraction of left ventricular was 48% by modified Simpson method. Her coronary artery was stenosed in the all three vessels. Right heart catheter (RHC) reported that CI 1.97L/min/m², Qp/Qs 2.07, PVR 2.56WU · m². mPAP 27mmHg, PAWP 17mmHg, we thought that pulmonary hypertension (PH) existed because not shunt of ASD, but Passive post-capillary PH. Therefore, the CABG and ASD surgical closure was performed simultaneously. After this surgery, the one of graft was damaged, and V-A ECMO was performed. And then her hemodynamics had been stable gradually, V-A ECMO was weaning in post-operative day (POD)6. But until POD7, saturation of oxygen was decreased, and pulmonary congestion got worse. We performed RHC again, and the PVR was increased remarkably (CI 1.53L/min/m², PVR 7.2WU · m², mPAP 30mmHg, PAWP 19mmHg), and she passed away in POD20 because worsening of PH and progression of sepsis from leg gangrene.

PJ6-3 肺動脈絞扼術及びCRT-D植え込みが行われた修正大血管転位の一例 Effect of pulmonary artery banding and CRT-D for congenitally corrected transposition of the great arteries with severe tricuspid regurgitation

奥田 真一¹⁾, 濱田 頼臣¹⁾, 周布 陽子¹⁾, 和田 靖明¹⁾, 杜 徳尚²⁾, 伊藤 浩²⁾, 笠原 真悟³⁾, 矢野 雅文¹⁾

1) 山口大学大学院医学系研究科 器官病態内科学, 2) 岡山大学大学院医歯薬学総合研究科 循環器内科,
3) 岡山大学大学院医歯薬学総合研究科 心臓血管外科

Shinichi Okuda¹⁾, Yoriomi Hamada¹⁾, Yoko Sufu¹⁾, Yasuaki Wada¹⁾, Norihisa Toh²⁾, Hiroshi Ito²⁾, Shingo Kasahara³⁾, Masafumi Yano¹⁾

1) Department of Medicine and Clinical Science, Yamaguchi University Graduate School of Medicine,

2) Department of Cardiovascular Medicine, Okayama University, 3) Department of Cardiovascular Surgery, Okayama University

A 42-year-old man was referred to our hospital for dyspnea and pulmonary edema. He diagnosed of congenitally corrected transposition of the great arteries (ccTGA) at 5 years old and had a history of admission of heart failure (HF) 6 years ago. After pharmacological therapy for chronic HF including beta-blockade and diuretics, symptoms were improved to NYHA II. However, transthoracic echocardiography (TTE) revealed dilatation of right ventricle (RVDd:87mm) with reduced RVEF (35%) and severe tricuspid regurgitation (TR). After discussion of surgical treatment, pulmonary artery banding (PAB) and cardiac resynchronization therapy defibrillator (CRT-D) were performed. After surgery, the value of BNP was reduced and TTE parameters were improved such as decrease of TR and increase of RVEF (45%), probably caused by the phenomenon that PAB elevates the left ventricular pressure, reduces leftward septal shift, prevents further tricuspid annular dilation, and subsequent by improvement of HF. About 1.5 years after surgery, no history of administration of HF was shown. Treatment of TR and improvement of the systemic RV's systolic function by PAB and CRT-D could be useful for ccTGA.

PJ6-4

エピネフリン誘発性QT延長症候群合併妊娠の1例

A case of pregnant woman with epinephrine-induced long QT syndrome

鳥谷部 邦明, 古橋 芙美, 真川 祥一, 島田 京子, 真木 晋太郎, 金田 倫子, 二井 理文, 田中 佳世,
田中 博明, 池田 智明
三重大学 産婦人科

Kuniaki Toriyabe, Fumi Furuhashi, Shoichi Magawa, Kyoko Shimada, Shintaro Maki, Michiko Kaneda, Masafumi Nii,
Kayo Tanaka, Hiroaki Tanaka, Tomoaki Ikeda
Department of Obstetrics and Gynecology, Mie University

Introduction: The activation of the sympathetic nervous system increases during pregnancy. Therefore, pregnancy may be the risk of polymorphic ventricular tachycardia in the pregnant women with epinephrine-induced long QT syndrome (LQTS). We report a case of pregnant woman with epinephrine-induced LQTS.

Case: She caused cardiopulmonary arrest at the age of 12. She was diagnosed epinephrine-induced LQTS, and implantable cardioverter-defibrillator (ICD) was implanted. She was pregnant at the age of 22. No cardiovascular event was caused during pregnancy. Epidural anesthesia was started after labor onset. She delivered 3052 g baby by vacuum extraction without the cardiovascular event. At the age of 24, she was pregnant again. No cardiovascular event was caused during pregnancy. Epidural anesthesia was started after labor onset as on the previous occasion. She delivered 3192 g baby by vacuum. No cardiovascular event was caused during pregnancy and labor.

Conclusion: The pregnancy may be no risk in pregnant woman with epinephrine-induced long QT syndrome. Epidural anesthesia would be effective in labor more activated the sympathetic nervous system than antepartum.

PJ6-5

心臓カテーテル検査後に造影剤腎症をきたした肺動脈閉鎖症

Contrast induced nephropathy after cardiac catheterization in a patient with pulmonary atresia

鈴木 康太, 高橋 辰徳, 安孫子 雅之, 小田切 徹州, 三井 哲夫
山形大学医学部 小児科

Kota Suzuki, Tatsunori Takahashi, Masayuki Abiko, Teshu Otagiri, Tetsuo Mitsui
Department of Pediatrics, Yamagata University Faculty of Medicine

A 30 years old woman with pulmonary atresia was undergone modified Blalock - Taussig shunt at 3 months of age, and her oxygen saturation was 80%. She admitted because of acute exacerbation of chronic heart failure. Fluid restriction and intravenous diuretic therapy were not effective, so Milrinone were added on day 18. She received cardiac catheterization on day 22, and Iohexol was used a total of 3.4 ml/kg. Before the catheterization, serum creatinine was 0.76 mg/dl, and estimate glomerular filtration rate was 69.9 ml/min/1.73 m². On the next day, oliguria occurred, and the serum creatinine increased to 1.62 mg/dl. We diagnosed as contrast induced nephropathy (CIN). 48 hours after the catheterization, she became anuria and serum creatinine markedly increased to 3.86 mg/dl. Moreover, systemic edema and pleural effusion appeared, therefore continuous hemodiafiltration (CHDF) began on day 25. After that, serum creatinine and anuria improved. CHDF was terminated on day 33. In the case of chronic cyanosis, prophylactic hydration should be performed before and after cardiac catheterization, even if the mild renal dysfunction, because of high risk of CIN.

PJ6-6 CRTが著効した修正大血管転位の1例

A case of congenitally corrected transposition of the great arteries successfully treated with cardiac resynchronization therapy

市川 啓之, 西井 伸洋, 三好 章仁, 杜 徳尚, 三好 亨, 中村 一文, 赤木 禎治, 伊藤 浩
岡山大学 循環器内科

Keishi Ichikawa, Nobuhiro Nishii, Akihito Miyoshi, Norihisa Toh, Toru Miyoshi, Kazufumi Nakamura, Teiji Akagi, Hiroshi Ito
Department of Cardiovascular Medicine, Okayama University

Congenitally corrected transposition of the great arteries (ccTGA) is rare form of congenital heart disease. In these patients, bradyarrhythmia, tachyarrhythmia and systemic right ventricular dysfunction are common. Although cardiac resynchronization therapy (CRT) in patients with systemic left ventricle has been established, the effect of CRT in patients with systemic right ventricle remain unclear.

A 46-year-old man with a history of hypertension was referred to our hospital due to exacerbation in dyspnea on exertion. Electrocardiogram demonstrated complete atrioventricular block. A transthoracic echocardiogram showed ccTGA, and mildly reduced systolic RV function (ejection fraction 45%). Only left ventricular pacing could induce dyssynchrony of right ventricle and exacerbate heart failure. Therefore, CRT was implanted by transvenous approach. Follow-up assessment at 6 months revealed a significant improvement in functional class (NYHA I), an increase of ejection fraction of the systemic right ventricle and a significant improvement of exercise tolerance. CRT can be a valuable option for the management of ccTGA.

PJ6-7 成人期に診断された血管輪に伴う気管支・食道狭窄の一例

Adult case of tracheal and esophageal compression due to vascular ring

石北 綾子¹⁾, 坂本 一郎¹⁾, 梅本 真太郎¹⁾, 永田 弾²⁾, 帯刀 英樹³⁾, 塩瀬 明³⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 小児科, 3) 九州大学病院 心臓血管外科

Ayako Ishikita¹⁾, Ichiro Sakamoto¹⁾, Shintaro Umemoto¹⁾, Hazumu Nagata²⁾, Hideki Tatewaki³⁾, Akira Shiose³⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiology, Kyushu University, 2) Department of Pediatrics, Kyushu University,

3) Department of Cardiovascular Surgery, Kyushu University

Vascular ring is a congenital anomaly in which the encircled aortic arch and its branches compress the trachea or esophagus.

A 19-year-old female presented with dysphagia and dyspnea. She was pointed out a chest radiography abnormality at age 17 and then vascular ring was suspected in CT. At age 18, she complained of worsening dysphagia, and dyspnea on exercise. Contrast-enhanced CT revealed that right aortic arch with retroesophageal components, i.e. left brachiocephalic artery, left subclavian artery, and ligamentum arteriosum extended from a diverticulum on the upper descending thoracic aorta surrounded the trachea and esophagus. Bronchoscopy and upper gastrointestinal endoscopy identified tracheal and esophagus compression at the vascular ring site. Esophagography confirmed the compression of esophagus and delay of swallowing. Consequently, a surgical operation was indicated for this patient.

We experienced adult case of tracheal and esophageal compression due to vascular ring who suffered from progressive symptom.

PJ6-8

Ross後のASR・LVOTOに対するApico-aortic bypass

Apico-aortic conduit bypass in patients with ASR and LVOTO after Ross procedure with RVOTR using Freestyle aortic root bioprosthesis

坂本 一郎¹⁾, 石北 綾子¹⁾, 帯刀 英樹²⁾, 出口 裕子¹⁾, 山本 泰史¹⁾, 梅本 真太郎¹⁾, 永田 弾³⁾, 大賀 正一³⁾, 塩瀬 明²⁾, 筒井 裕之¹⁾

1) 九州大学病院 循環器内科, 2) 九州大学病院 心臓血管外科, 3) 九州大学病院 小児科

Ichiro Sakamoto¹⁾, Ayako Ishikita¹⁾, Hideki Tatewaki²⁾, Hiroko Deguchi¹⁾, Taishi Yamamoto¹⁾, Shintaro Umemoto¹⁾, Hazumu Nagata³⁾, Shouichi Ohga³⁾, Akira Shiose²⁾, Hiroyuki Tsutsui¹⁾

1) Department of Cardiovascular Medicine, Kyushu University Hospital,

2) Department of Cardiovascular Surgery, Kyushu University Hospital, 3) Department of Pediatrics, Kyushu University Hospital

A 35-year-old female with prior Ross operation was referred to our hospital. She was born with bicuspid aortic valve and subaortic stenosis. She was performed aortic commissurotomy and resection of fibrous membrane at the age of 12 years. But her LV pressure was not decreased, Ross operation with RVOTR using Freestyle aortic root bioprosthesis was performed at the age of 16 years. However, her symptom and LV pressure were not improved. She became orthopnea and complicated renal dysfunction due to high CVP and low cardiac output.

We performed apico-aortic conduit bypass using SJM regent 19mm and J-graft 22mm, because AVR with LVOT reconstruction was not enough for small annulus size and median sternotomy was high risk for previously used Freestyle valve. She needed long-term mechanical ventilation and continuous hemodiafiltration. But post-operative intensive care improved her general condition. Although severe aortic regurgitation was remained, she was discharged three months after the operation with improved symptom, renal function and systolic LV-aorta pressure gradient.

PJ7-1

肺血管拡張薬投与後、遺残VSDシャントを閉鎖しえたPA-VSDの成人症例

A case of treat and repair: Residual VSD Closure for an Adult Patient with Pulmonary Atresia with VSD after Treatment with Pulmonary Vasodilators

塚本 泰正¹⁾, 中野 智彰¹⁾, 平 将生²⁾, 成田 淳³⁾, 溝手 勇¹⁾, 大谷 朋仁¹⁾, 上野 高義²⁾, 彦惣 俊吾¹⁾, 坂田 泰史¹⁾

1) 大阪大学大学院医学系研究科 循環器内科学, 2) 大阪大学大学院医学系研究科 心臓血管外科学,

3) 大阪大学大学院医学系研究科 小児科学

Yasumasa Tsukamoto¹⁾, Tomoaki Nakano¹⁾, Masaki Taira²⁾, Jun Narita³⁾, Isamu Mizote¹⁾, Tomohito Ohtani¹⁾, Takayoshi Ueno²⁾, Shungo Hikoso¹⁾, Yasushi Sakata¹⁾

1) Department of Cardiovascular Medicine, Osaka University School of Medicine,

2) Department of Cardiovascular Surgery, Osaka University School of Medicine,

3) Department of Pediatrics, Osaka University School of Medicine

We present the case of a 40-year-old female patient with pulmonary atresia with ventricular septum defect (PA-VSD) and major aorto-pulmonary collateral arteries (MAPCAs). The patient underwent right and left BT shunt when she was 7 and 8 years old. Then she underwent right ventricle outflow tract (RVOT) repair and MAPCAs ligation at age 10. Because of her bilateral pulmonary arteries hypoplasia and high pulmonary vascular resistance, it was judged that the VSD closure carried a high risk at that time. Because RVOT stenosis progressed with age, she received catheter examination at age 35. The pressure gradient between RV and PA was 66mmHg, her PVRI was measured at 5.60 Wood Units · m², and Qp/Qs ratio was 1.1. Administration of bosentan was initiated with an expectation of decline in PVR. At age 39, hemodynamic data showed decrease in PVRI (1.95 Wood Units · m²). We then decided to perform re-RVOT repair and VSD closure. Cyanosis markedly subsided after the operation.

We suggest that this is an unusual therapeutic strategy for residual lesion of a patient with PA/VSD, after treatment with pulmonary vasodilators reduced PVRI and the accompanying procedural risk.

PJ7-2 心内修復術を施行したDown症候群, 完全型房室中隔欠損症, 肺動脈絞扼術後の成人手術例 An adult surgical case of Down syndrome, complete AVSD, postoperative state of PA banding

森下 寛之¹⁾, 江連 雅彦¹⁾, 長谷川 豊¹⁾, 山田 靖之¹⁾, 星野 丈二¹⁾, 岡田 修一¹⁾, 金澤 祐太¹⁾, 加我 徹¹⁾, 山下 英治²⁾, 村上 淳²⁾, 宮本 隆司³⁾

1) 群馬県立心臓血管センター 心臓血管外科, 2) 群馬県立心臓血管センター 循環器内科, 3) 北里大学医学部 心臓血管外科

Hiroyuki Morishita¹⁾, Masahiko Ezure¹⁾, Yutaka Hasegawa¹⁾, Yasuyuki Yamada¹⁾, Joji Hoshino¹⁾, Shuichi Okada¹⁾, Yuta Kanazawa¹⁾, Toru Kaga¹⁾, Eiji Yamashita²⁾, Jun Murakami²⁾, Takashi Miyamoto³⁾

1) Division of Cardiovascular Surgery, Gunma Prefectural Cardiovascular Center,

2) Department of Cardiology, Gunma Prefectural Cardiovascular Center,

3) Department of Cardiovascular Surgery, Kitasato University School of Medicine

Introduction: Intracardiac repair (ICR) for complete atrioventricular septal defect (cAVSD) is generally applied during infancy. Here is a report of the case we experienced in applying the ICR on an adult patient.

Case: A 32 year-old man diagnosed with Down syndrome and cAVSD, he received pulmonary artery banding (PAB) at 7 months old and continued outpatient clinic thereafter. At around age 30, his conditions worsened with breathing difficulty and he was referred to our hospital. His pulmonary artery mean pressure was 16mmHg, and the cardiac catheterization indicated Qp/Qs was 0.46 and pulmonary vascular resistance was 7.9 U/m². After medical therapy, we decided to switch to ICR. The surgery was performed in two-patch repair. While no left-sided atrioventricular valve regurgitation was noted, weaning from cardiopulmonary bypass was difficult due to reduced pulmonary blood flow. He was returned to ICU with ECMO. The decrease in pulmonary perfusion continued, and no improvement resulted after introduction of Treprost. On the 17th postoperative day, the patient was deceased.

Summary: Despite the PAB, pulmonary blood flow was not secured and the patient was lost.

PJ7-3 Takeuchi法術後長期遠隔期を迎えたBWG症候群の1例 A case of BWG syndrome with long-term after the Takeuchi procedure

山内 博貴¹⁾, 前田 正信¹⁾, 倉石 建治²⁾

1) 一宮西病院, 2) 大垣市民病院

Hirota Yamauchi¹⁾, Masanobu Maeda¹⁾, Kenji Kuraishi²⁾

1) Ichinomiya Nishi Hospital, 2) Ogaki Municipal Hospital

A 32-year-old man was diagnosed with Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) also known as Bland-White-Garland (BWG) syndrome at the age of six. It was pointed out that there was an abnormality in the electrocardiogram with an ischemic change while having a health checkup at school. The left coronary orifice was located on the left side of the pulmonary artery and it was difficult to implant the coronary artery directly. So the Takeuchi procedure was selected as a left coronary artery reconstruction surgery. After postoperative periodic follow-up, his regular visit was interrupted for school, changing his address and etc. However 26 years passed without cardiovascular events. This patient came to our hospital asking for health evaluation with employment. Electrocardiogram, echocardiogram, Coronary artery CT and catheter examination showed good coronary artery reconstruction without ischemic change. Only mild to moderate pulmonary artery stenosis was noted. We report the details of this case with discussion about long-term outcome of Takeuchi procedure for children. To tell the truth, this is my own case report.

PJ7-4

心房リードの追加により運動時周期性呼吸変動が改善した房室中隔欠損症術後の1例

An atrial lead implantation improve exercise oscillations ventilation in a case of postoperative atrioventricular septal defect

前川 恵美, 小坂橋 俊美, 矢崎 麻由, 前村 健治, 大木 卓巳, 藤田 鉄平, 池田 祐毅, 鍋田 健, 郡山 恵子, 成毛 崇, 阿古 潤哉
北里大学医学部 循環器内科学

Emi Maekawa, Toshimi Koitabashi, Mayu Yazaki, Kenji Maemura, Takumi Ooki, Teppei Fujita, Yuuki Ikeda, Takeshi Nabeta, Keiko Kohriyama, Takashi Naruke, Junya Ako
Department of Cardiovascular Medicine, Kitasato University School of Medicine

Background: In ACHD cases with complicated background, it's difficult to diagnose the causes of shortness of breath (SOB).

Case: The patient was a 25-year-old female who diagnosed as partial atrioventricular septal defect, and underwent intracardiac repair surgery and mechanical mitral valve replacement. After surgery a DDD pacemaker was implanted in a patient with complete atrioventricular block. The pacemaker mode was switched to VDD at 21 years of age due to elevating the threshold of the atrial lead. Gradually, SOB developed but didn't presented sign of prosthetic valve dysfunction.

Decreasing exercise tolerance and presenting with exercise oscillations ventilation (EOV) during cardiopulmonary exercise testing (CPX) suspected it caused decreasing heart rate response. There was no remarkable change in the result of CPX in patient switched to VVIR. So an atrial lead was implanted for switch to DDD. As a result, CPX demonstrated the patient was EOV disappeared.

Conclusion: In this case, an atrial lead implantation reveal EOV disappeared and to effect of the relation between atrium and ventricular. CPX is useful for detecting the causes of SOB and the point of intervention.

PJ7-5

先天性左冠動脈起始異常術後肺動脈弁逆流に対し血流解析を基に二弁修復しえた成人例

Right heart valves repair in an adult patient with severe pulmonary and tricuspid regurgitation after repair of congenital anomalous origin of LMT

瀧上 雅雄¹⁾, 板谷 慶一²⁾, 中西 直彦¹⁾, 森地 裕子²⁾, 中路 康介³⁾, 中村 猛¹⁾, 梶山 葉⁴⁾, 的場 聖明¹⁾, 夜久 均²⁾, 山岸 正明⁵⁾

1) 京都府立医科大学付属病院 循環器内科, 2) 京都府立医科大学付属病院 心臓血管外科, 3) 京都府立医科大学付属病院 放射線科, 4) 京都府立医科大学付属病院 小児科, 5) 京都府立医科大学付属病院 小児心臓血管外科

Masao Takigami¹⁾, Keiichi Itatani²⁾, Naohiko Nakanishi¹⁾, Yuko Morichi²⁾, Kosuke Nakaji³⁾, Takeshi Nakamura¹⁾, Yo Kajiyama⁴⁾, Satoaki Matoba¹⁾, Hitoshi Yaku²⁾, Masaaki Yamagishi⁵⁾

1) Department of Cardiovascular Medicine, Kyoto Prefectural University of Medicine.

2) Department of Cardiovascular Surgery, Kyoto Prefectural University of Medicine.

3) Department of Radiology, Kyoto Prefectural University of Medicine.

4) Department of Pediatrics, Kyoto Prefectural University of Medicine.

5) Department of Pediatric Cardiovascular Surgery, Kyoto Prefectural University of Medicine

The patient is a 29 year-old female, who underwent Takeuchi repair for Bland-White-Garland syndrome when 2 months old. Repeated residual coronary shunt closure was required, resulting in pulmonary stenosis, which was released a large patch placed when she was 14 years old. However, this patch caused severe pulmonary regurgitation (PR), right ventricular (RV) enlargement, and tricuspid regurgitation (TR). Coronary angiogram showed flow stagnation in Takeuchi route. CFD model found vortices moving in the route and estimated FFR was sufficiently high. 4D flow MRI revealed PR and TR fraction 43.4% and 68.5%, respectively, RV EDV was 189.35 ml/m², and energy loss was 6.57mW, around 6 times higher than that in normal controls. The left interventricular pressure difference from color M mode in echocardiograph was 5.5mmHg, with acceptable left ventricular diastolic sucking force. She underwent RV outflow reconstruction and tricuspid valve plasty, which successfully controlled regurgitation. She discharged at 15 day after the operation without complication. We discuss the assessment of adult congenital heart disease (ACHD) based on blood flow imaging and ACHD heart team.

PJ7-6

**卵円孔開存に device closure を施行した Platypnea-Orthodexia syndrome の 1 例
Platypnea-orthodexia syndrome in an elder women treated by device closure for patent foramen ovale.**

金子 幸栄¹⁾, 中嶋 八隅¹⁾, 井上 奈緒¹⁾, 森 善樹^{1,3)}, 後藤 雅之²⁾, 杉浦 亮²⁾

1) 聖隷浜松病院 小児循環器科, 2) 聖隷浜松病院 循環器科, 3) 北里大学メディカルセンター 小児科

Sachie Kaneko¹⁾, Yasumi Nakajima¹⁾, Nao Inoue¹⁾, Yoshiki Mori^{1,3)}, Masayuki Goto²⁾, Ryo Sugiura²⁾

1) Department of Pediatric Cardiology, Seirei Hamamatsu General Hospital,

2) Department of Cardiology, Seirei Hamamatsu General Hospital, 3) Department of Pediatrics, Kitasato University Medical Center

Platypnea-orthodexia syndrome (POS) is a rare but important form of dyspnea. It is characterized by dyspnea and hypoxia occurs in the upright position and improves with recumbency.

A 79-year-old woman was referred due to dyspnea. The oxygen saturation (SpO₂) was 99% but decreased around 80% in the standing. The transthoracic echocardiography (TTE) showed no intra-cardiac shunt but moderate aortic regurgitation with enlargement of sinus of valsalva. Computed tomography revealed no abnormalities of lung, air way and coronary arteries. In day 10, she presented with dysarthria and right arm paralysis. MRI confirmed multiple cerebral infarctions. We performed transesophageal echocardiography (TEE) and repeated TTE with a microbubble test changing the position. The POS associated with aortic elongation and PFO was confirmed. We performed device closure of PFO using a 25mm Amplatzer Cribriform. Her dyspnea improved with increase of SpO₂ (92-95%) in the standing.

The PFO must be actively investigated in the presence of position dependent hypoxia. The microbubble test with TTE and TEE is useful and the device closure is a safe and effective technique for elder person as previously reported.

PJ7-7

川崎病冠動脈瘤に血栓閉塞をきたした ST 上昇型心筋梗塞患者の一例

A case report: ST-elevation myocardial infarction caused by thrombotic occlusion of giant coronary artery aneurysm following Kawasaki disease

大家 理伸¹⁾, 小坂田 皓平¹⁾, 羽原 誠二¹⁾, 福 康志¹⁾, 荻野 佳代²⁾, 林 知宏²⁾, 脇 研自²⁾, 小宮 達彦³⁾, 門田 一繁¹⁾, 新垣 義夫²⁾

1) 倉敷中央病院 循環器内科, 2) 倉敷中央病院 小児科, 3) 倉敷中央病院 心臓血管外科

Masanobu Ohya¹⁾, Kohei Osakada¹⁾, Seiji Habara¹⁾, Yasushi Fuku¹⁾, Kayo Ogino²⁾, Tomohiro Hayashi²⁾, Kenji Waki²⁾, Tatsuhiko Komiya³⁾, Kazushige Kadota¹⁾, Yoshio Aragaki²⁾

1) Department of Cardiology, Kurashiki Central Hospital, 2) Department of Pediatrics, Kurashiki Central Hospital,

3) Department of Cardiovascular surgery, Kurashiki Central Hospital

A 10-years-old girl presented to the emergency department with acute chest pain. The patient had a known history of Kawasaki disease and giant coronary aneurysms detected by follow-up coronary angiography 2 years before. An initial electrocardiogram showed ST-segment elevation in leads V₂ to V₅ and emergent coronary angiography was performed. It showed coronary thrombotic occlusion at a giant aneurysm in the left anterior descending artery (LAD). We performed thrombotic aspiration repeatedly and injected tissue-plasminogen activator into the LAD. Although coronary reperfusion was available at that time, follow-up coronary angiography showed LAD reocclusion and the myocardial viability was none by cardiac scintigraphy. Three years later, she received coronary artery bypass grafting for right coronary artery and LAD. She was planned to receive cardiac resynchronization therapy, but had cardiac pulmonary arrest due to ventricular fibrillation and needed percutaneous cardiopulmonary assist. She has been waiting for heart transplantation for 2 years, with left ventricular assist device. We would like to discuss the optimal timing and contents of treatments for this patient.

PJ7-8

造影CTにより診断にいたった部分肺静脈還流異常合併心房中隔欠損症の一例

A case of valid computed tomography for the diagnosis of atrial septal defect complicated partial anomalous pulmonary venous drainage

宮澤 聡明, 岡田 拓也, 大石 岳, 木暮 武仁, 佐藤 陽, 宮内 尊徳, 篠崎 雅人
IMSグループ横浜旭中央総合病院 循環器科

Satoaki Miyazawa, Takuya Okada, Gaku Ooishi, Takehito Kogure, Akira Sato, Takenori Miyauchi, Masato Shinozaki
Department of Cardiology, IMS group Yokohama Asahi Chuo General Hospital

A 74 years old woman came to our hospital for dyspnea on effort. Her echocardiography showed mild pulmonary hypertension, and Qp/Qs ratio was 3.1. But we could not find out shunt disease in her echogram. We performed computed tomography, which showed sinus venosus type atrial septal defect and partial anomalous pulmonary venous drainage. Her diagnostic cardiac catheterization showed that Qp/Qs ratio was 2.01, left-to-right shunt ratio was 55.4%, left-to-right shunt ratio was 10.3%, pulmonary vascular resistance was 1.79 unit/m². We assessed that she required surgical operation.

Computed tomography was very valid for the pulmonary hypertension case suspected shunt disease.

PJ7-9

成人期に Fontan 手術を行った右室型単心室、両大血管右室起始症の2例

The two cases of extracardiac TCPC (E-TCPC) in adulthood for double-outlet right ventricle (DORV) and single right ventricle (SRV) in our hospital

廣瀬 和俊¹⁾, 相馬 桂¹⁾, 佐々 達郎¹⁾, 齊藤 暁人¹⁾, 稲葉 俊郎¹⁾, 石川 友一²⁾, 犬塚 亮³⁾, 平田 康隆⁴⁾, 八尾 厚史⁵⁾, 小室 一成¹⁾

1) 東京大学医学部附属病院 循環器内科, 2) 福岡市立こども病院 循環器科, 3) 東京大学医学部附属病院 小児科, 4) 東京大学医学部附属病院 心臓外科, 5) 東京大学保健・健康推進本部

Kazutoshi Hirose¹⁾, Katsura Souma¹⁾, Tatsuro Sassa¹⁾, Akihito Saito¹⁾, Toshiro Inaba¹⁾, Yuichi Ishikawa²⁾, Ryo Inuzuka³⁾, Yasutaka Hirata⁴⁾, Atsushi Yao⁵⁾, Issei Komuro¹⁾

1) Department of Cardiovascular Medicine, The University of Tokyo Hospital, 2) Fukuoka Children's Hospital, Dept of Cardiology, 3) Department of Pediatrics, The University of Tokyo Hospital, 4) Department of Cardiac Surgery, The University of Tokyo Hospital, 5) Division for Health Service Promotion, The University of Tokyo

In adulthood, the indication of E-TCPC for cyanotic congenital heart disease has not been established yet. We report two adult cases of E-TCPC. Case1 is a woman with single atrium, SRV, DORV, and pulmonary atresia. She underwent Blalock Taussig shunts in infancy, and was referred to our hospital due to dyspnea in adulthood. Although RV ejection fraction slightly decreased, bidirectional Glenn and E-TCPC were serially performed because of low pulmonary arterial pressure and pulmonary vascular resistance. She has been treated with sildenafil and carvedilol for 9 years. Case2 is a man with DORV, pulmonary stenosis, atrial septal defect, SRV, common atrioventricular valve (CAVV) and total anomalous pulmonary venous connection. In adulthood, his cyanosis-related symptoms got worse as CAVV regurgitation became severe. He underwent multi-staged surgeries, finally leading to fenestrated TCPC with CAVV replacement. Postoperative cardiac catheterization revealed RV systolic dysfunction and right-to-left shunt through fenestration. Therefore, he has been treated with carvedilol. Indication of E-TCPC in adults and appropriate medications for Fontan circulation will be discussed.

PJ8-1

心臓外科治療だけでは成人先天性心疾患患者の運動耐容能を十分改善しない

Only Surgical Treatment of Cardiac Impairment is Insufficient for Improvement of Exercise Tolerance in Patients with Adult Congenital Heart Disease

岩田 究¹⁾, 仁田 学¹⁾, 岡村 正嗣³⁾, 木野 旅人¹⁾, 松本 祐介¹⁾, 寺中 紗絵¹⁾, 鍵本 美奈子¹⁾, 清國 雅義¹⁾, 小村 直弘¹⁾, 細田 順也¹⁾, 重永 豊一郎¹⁾, 上村 大輔¹⁾, 松本 克己¹⁾, 菅野 晃靖¹⁾, 石上 友章¹⁾, 石川 利之¹⁾, 町田 大輔²⁾, 益田 宗孝²⁾, 中村 健³⁾, 田村 功一¹⁾

1) 横浜市立大学大学院医学研究科 病態制御内科学, 2) 横浜市立大学大学院医学研究科 外科治療学・心臓血管外科, 3) 横浜市立大学大学院医学研究科 リハビリテーション科

Kiwamu Iwata¹⁾, Manabu Nitta¹⁾, Masatsugu Okamura³⁾, Tabito Kino¹⁾, Yusuke Matsumoto¹⁾, Sae Teranaka¹⁾, Minako Kagimoto¹⁾, Masayoshi Kiyokuni¹⁾, Naohiro Komura¹⁾, Junya Hosoda¹⁾, Atsuihiro Shigenaga¹⁾, Daisuke Kamimura¹⁾, Katsumi Matsumoto¹⁾, Teruyasu Sugano¹⁾, Tomoaki Ishigami¹⁾, Toshiyuki Ishikawa¹⁾, Daisuke Machida²⁾, Munetaka Masuda²⁾, Ken Nakamura³⁾, Koichi Tamura¹⁾

1) Department of Medical Science and Cardiorenal Medicine, Yokohama City University Graduate School of Medicine,

2) Department of Cardiovascular Surgery, Yokohama City University Graduate School of Medicine,

3) Department of Rehabilitation Medicine, Yokohama City University Graduate School of Medicine

Background: In most patients with adult congenital heart disease, exercise capacity is limited because of sequelae or remote term complications and sometimes need further surgery.

Case: A 22-year-old female suffered from repeated syncope and was referred to our department. At the age of 1, she was diagnosed as double outlet right ventricle with dextrocardia, and intracardiac surgical correction was performed at the age of 5. Thereafter due to development of aortic insufficiency, she underwent bioprosthetic aortic valve replacement (b-AVR) at the age of 15. 7 years later, structural valve deterioration of bioprosthetic aortic valve replacement showed 4.7 m/sec of peak velocity and 50 mmHg of mean pressure gradient, suggesting severe aortic stenosis. Therefore 2nd b-AVR was performed. Cardiopulmonary exercise test at 6 months after the surgery showed 18.6 ml/kg/min of peak VO₂, which was equivalent to 60% of predicted normal values for her age. After 3 months of exercise training (ET), peak VO₂ elevated to 22.1 ml/kg/min (75% of normal).

Conclusion: Both surgical Treatment and subsequent internal treatment including ET are necessary to improve exercise capacity.

PJ8-2

残存心房間右左シャントに対してASDデバイス閉鎖を施行した純型肺動脈閉鎖の成人例

Percutaneous ASD Closure for Interatrial Right-to-left Shunt in Adult Patient with Repaired Pulmonary Atresia with Intact Ventricular Septum

関 満¹⁾, 片岡 功一^{1,2,3)}, 久保田 香菜^{3,4)}, 安済 達也¹⁾, 古井 貞浩¹⁾, 岡 健介¹⁾, 松原 大輔¹⁾, 佐藤 智幸¹⁾, 甲谷 友幸^{3,4)}, 今井 靖^{3,4)}, 河田 政明^{2,3,5)}

1) 自治医科大学とちぎ子ども医療センター 小児科, 2) 自治医科大学とちぎ子ども医療センター 小児手術・集中治療部,

3) 自治医科大学成人先天性心疾患センター, 4) 自治医科大学 循環器内科,

5) 自治医科大学とちぎ子ども医療センター 小児・先天性心臓血管外科

Mitsuru Seki¹⁾, Koichi Kataoka^{1,2,3)}, Kana Kubota^{3,4)}, Tatsuya Anzai¹⁾, Sadahiro Furui¹⁾, Kensuke Oka¹⁾, Daisuke Matsubara¹⁾, Tomoyuki Satoh¹⁾, Tomoyuki Kabutoya^{3,4)}, Yasushi Imai^{3,4)}, Masaaki Kawada^{2,3,5)}

1) Department of Pediatrics, Jichi Childrens Medical Center,

2) Pediatric Operating Suite and Intensive Care Unit, Jichi Childrens Medical Center,

3) Adult Congenital Heart Disease Center, Jichi Medical University, 4) Cardiovascular Medicine, Jichi Medical University,

5) Pediatric and Congenital Cardiovascular Surgery, Jichi Childrens Medical Center

Introduction: We report ASD device closure for improving desaturation due to interatrial right-to-left shunt in adult patient with biventricular repaired pulmonary atresia with intact ventricular septum.

Case: The patient is an 18-year old male. He underwent percutaneous balloon pulmonary valvuloplasty at 1 month after birth. Although this treatment was effective, SpO₂ remained around 90%. Since the desaturation due to interatrial right-to-left shunt was persistent, ASD device closure was planned. Qp/Qs was 0.7 and RVEDP was 7mmHg with no pulmonary valve stenosis. The test occlusion of interatrial communication with sizing balloon for 10 minutes showed that SaO₂ increased from 83 to 95% and RAP increased from 8 to 9 mmHg without systemic hypotension. ASD was successfully occluded using Figulla Flex II (21mm diameter). At 6 months-follow-up, the patient's oxygenation is remarkably improved and no right heart failure finding is recognized.

Conclusion: Prior test occlusion is essential for safe ASD closure in adult patient with cyanosis.

PJ8-3

慢性骨髄性白血病と肺動脈性肺高血圧を合併したファロー四徴症術後の一例

A Case of Repaired Tetralogy of Fallot with Chronic Myelogenous Leukemia and Pulmonary Arterial Hypertension

前村 健治¹⁾, 小坂橋 俊美¹⁾, 藤田 鉄平¹⁾, 郡山 恵子¹⁾, 前川 恵美¹⁾, 齋木 宏文²⁾, 宮本 隆司³⁾, 宮地 鑑³⁾, 先崎 秀明³⁾, 阿古 潤哉¹⁾

1) 北里大学医学部 循環器内科学, 2) 北里大学医学部 小児科学, 3) 北里大学医学部 心臓血管外科

Kenji Maemura¹⁾, Toshimi Koitabashi¹⁾, Teppei Fujita¹⁾, Keiko Kooriyama¹⁾, Emi Maekawa¹⁾, Hirohumi Saiki²⁾, Takashi Miyamoto³⁾, Kagami Miyaji³⁾, Hideaki Senzaki³⁾, Junya Ako¹⁾

1) Department of Cardiovascular Medicine, Kitasato University School of Medicine,

2) Department of Pediatrics, Kitasato University School of Medicine,

3) Department of Cardiovascular Surgery, Kitasato University School of Medicine

Background: The causes of pulmonary arterial hypertension are diverse.

Case: A 35-year-old woman with repaired tetralogy of Fallot and left pulmonary artery hypoplasia. At 29 years of age, she started taking dasatinib for Chronic Myelogenous Leukemia (CML). After that, the echocardiogram revealed increase of right ventricular systolic pressure. At 33 years of age, right heart catheterization showed high pulmonary vascular resistance (PVR) (5.1 woods). In addition to therapeutic agents for pulmonary hypertension (PHTx), dasatinib was changed to another medicine. PVR decreased to 3.0 woods.

Conclusion: Dasatinib is known to cause pulmonary hypertension. In this case, PVR was improved due to stop of dasatinib and administration of PHTx.

PJ8-4

Fontan術後の月経異常にどう対処するかー1例報告

How to deal with menstrual disorder in a patient with Fontan circulation?

大津 幸枝¹⁾, 高井 泰²⁾, 江良 澄子²⁾, 黄 海鵬²⁾, 先崎 秀明³⁾, 岩本 洋一⁴⁾, 石戸 博隆⁴⁾, 増谷 聡⁴⁾

1) 埼玉医科大学総合医療センター 看護部, 2) 埼玉医科大学総合医療センター 産婦人科, 3) 北里大学 小児科,

4) 埼玉医科大学総合医療センター 小児循環器部門

Yukie Otsu¹⁾, Yasushi Takai²⁾, Sumiko Era²⁾, Haipeng Huang²⁾, Hideaki Senzaki³⁾, Youiti Iwamoto⁴⁾, Hiroataka Isido⁴⁾, Satoshi Masutani⁴⁾

1) Nursing Department, Saitama Medical University Saitama Medical Center,

2) Obstetrics and Gynecology, Saitama Medical University Saitama Medical Center, 3) Pediatrics, Kitasato University,

4) Pediatric Cardiology, Saitama Medical University Saitama Medical Center

【はじめに】 治療法の向上により、成人期に到達する先天性心疾患 (ACHD) が増加している。女性 ACHD 患者で月経異常の頻度は低くなく、ときに介入を要するが、ACHD 領域での注目は必ずしも高くない。今回、Fontan 術後の月経異常に対し、婦人科と連携し、治療選択に熟慮中の症例を提示する。

【症例】 18歳 (高校3年生)、体重50kg。右側相同、右室型単心室を有し、3歳でFontan型手術を施行された。15歳で初経発来したが、月経不順を伴う過多月経のため産婦人科を紹介受診した。少量のエストラジオール (E2) の分泌はみられたが排卵はなく、骨密度の減少を指摘された。本来周期的エストロゲン・プロゲステロン療法 (いわゆるカウフマン療法) が考慮されるが、血栓リスクを考慮すると方針決定は容易でなく、骨密度減少に対する食事療法から治療を開始し、今後の治療法を検討中である。

【考察】 E2低値は骨粗鬆症につながり、無排卵症は不妊症や子宮体癌リスクを増加させる。月経異常を来さない正常なホルモン環境は、生体維持に重要である。本症例のような病態にカウフマン療法が有効と考えられるが、血栓リスクを考慮すると一律の方針決定はできない。今後、ACHD 患者の月経異常について、リスク・ベネフィットを勘案したガイドラインが望まれる。月経異常は非常にデリケートな話題のため、婦人科や多職種が連携し、サポートすることが重要と考えられた。

PJ8-5

65歳でチアノーゼ性心疾患に対し初回手術を受けた患者の心臓リハビリテーションの経験

Cardiac rehabilitation for patient who underwent initial surgery for cyanotic heart disease at 65 years old.

大西 伸悟¹⁾, 圓尾 文子²⁾, 山本 真由子²⁾, 白井 丈明³⁾, 嘉悦 泰博³⁾, 金子 明弘³⁾, 川崎 健作¹⁾,
大西 和子¹⁾, 角谷 誠³⁾, 大西 祥男³⁾

1) 加古川中央市民病院 リハビリテーション室, 2) 加古川中央市民病院 心臓血管外科, 3) 加古川中央市民病院 循環器内科

Shingo Ohnishi¹⁾, Ayako Maruo²⁾, Mayuko Yamamoto²⁾, Takeaki Shirai³⁾, Yasuhiro Kaetsu³⁾, Akihiro Kaneko³⁾,
Kensaku Kawasaki¹⁾, Kazuko Ohnishi¹⁾, Makoto Kadotani³⁾, Yoshio Ohnishi³⁾

1) The Department of Rehabilitation, Kakogawa Central City Hospital.

2) The Department of Cardiovascular Surgery, Kakogawa Central City Hospital.

3) The Department of Cardiovascular Medicine, Kakogawa Central City Hospital

【はじめに】65歳でVSD、PSにより初回手術を受けたチアノーゼ性心疾患患者の心臓リハビリテーション(以下心リハ)を実施する機会を得たので報告する。

【症例】65歳女性。12歳から労作時にチアノーゼが出現。中学校入学時の健康診断でASD、VSDを診断されたが手術を希望しなかった。25歳時と45歳時のカテーテル検査にてFallot四徴症と診断されるも手術は拒否していた。今回、労作時呼吸困難感が悪化し他院受診の結果、手術適応と判断され当院に紹介された。精査の結果VSD、severe PSを認め手術目的にて入院。手術は、肺動脈弁置換術、右室流出路形成、心室中隔欠損閉鎖が行われた。術直後は左室容量増大による左心不全にてDOB、pacing開始。術後翌日に右室肥大と右室拡張不全による右心不全も認めた。術後2日目に人工呼吸器離脱、術後3日目より理学療法開始。術後3日目に30m歩行を達成、術後10日目には200m歩行を達成したが、術後11日目から関節痛や下腿浮腫が悪化し、歩行距離延長は行わず最低限度の活動までとした。術後14日目の身体運動検査にて握力20.3kg、膝伸展筋力0.33kgf/kg、片脚立ち6.76秒、10m歩行11.6秒でFrailtyの状態を呈していた。術後15日目に退院となった。

【結語】本症例は通常の術後心リハに沿った経過で離床がすすめられた。しかし、心不全症状が出現しやすく入院中に200m以上の歩行練習は実施できなかった。Frailty改善に向けた長期的な心リハ介入の必要性が示唆された。

PJ8-6

正常心電図所見を示す心房中隔欠損症(ASD)の特徴についての検討

The features of ASD with normal ECG

井波 礼香¹⁾, 富松 宏文²⁾, 芦原 京美³⁾, 黒川 文夫¹⁾, 神田 かおり¹⁾, 小島 幸子¹⁾, 三浦 ひとみ¹⁾,
稲井 慶²⁾, 杉山 央²⁾

1) 東京女子医科大学 中央検査部, 2) 東京女子医科大学 循環器小児科, 3) 東京女子医科大学 循環器内科

Ayaka Inami¹⁾, Hirofumi Tomimatsu²⁾, Kyomi Ashihara³⁾, Fumio Kurokawa¹⁾, Kaori Kanda¹⁾, Sachiko Kojima¹⁾,
Hitomi Miura¹⁾, Kei Inai²⁾, Hisashi Sugiyama²⁾

1) Tokyo Women's Medical University, Central Clinical Laboratory, 2) Tokyo Women's Medical University, Pediatric Cardiology,

3) Tokyo Women's Medical University, Cardiology

【背景】心房中隔欠損症(ASD)では心電図異常が出現しないことがある。

【目的】正常心電図を呈するASD患者の特徴を明らかにすること。

【対象】2006年1月から2017年12月までの間に当院循環器小児科にてカテ治療をした19歳以上のASD患者連続130人中、Qp/Qs>2の44人。

【方法】対象を正常心電図群(N群)と異常心電図群(D群)にわけ、身長、体重、BSA、CTR、Qp/Qs、デバイスサイズ、右室圧(RVp)、右室径、胸郭縦長、胸郭横幅径などを2群間で比較した。心電図正常の判定はミネソタコードによる自動診断の結果を用い、胸郭径は身長で補正した。

【結果】N群21人、D群23人であった。N群vsD群の比較ではQp/Qs(2.6vs2.8)、デバイスサイズ(20vs20mm)に有意差は認めなかった。しかし、胸郭の横幅/身長は、N群vsD群=0.16vs0.17と有意差を認めた(P=0.018)。**【考察】**自動診断では21人(48%)が正常と判定された。2群間で欠損孔のサイズや右心負荷の程度に差はなかったが、胸郭の横幅/身長はN群で有意に小さく、心電図所見に差が出るのは体型による影響が考えられた。

【結語】治療適応があるASDであるにもかかわらず正常心電図を示す例が48%あり、ミネソタコードを用いたASDの診断には注意が必要であり、その要因として体型が関与している可能性が示唆された。

PJ8-7

心内修復術未施行の肺高血圧合併成人DORV症例に対する集学的治療の検討と経過
Multidisciplinary therapy for an adult case of DORV complicated by pulmonary arterial hypertension who hadn't undergone intracardiac repair in infancy

常盤 洋之¹⁾, 相馬 桂¹⁾, 廣瀬 和俊¹⁾, 齊藤 暁人¹⁾, 稲葉 俊郎¹⁾, 牧 尚孝¹⁾, 犬塚 亮²⁾, 石川 友一³⁾, 辻 重人⁴⁾, 柴田 深雪⁴⁾, 近藤 良一⁴⁾, 益澤 明広⁴⁾, 平田 康隆⁴⁾, 八尾 厚史⁵⁾, 小室 一成¹⁾

1) 東京大学医学部附属病院 循環器内科, 2) 東京大学医学部附属病院 小児科, 3) 福岡市立こども病院 循環器科, 4) 東京大学医学部附属病院 心臓外科, 5) 東京大学保健・健康推進本部

Hiroyuki Tokiwa¹⁾, Katsura Souma¹⁾, Kazutoshi Hirose¹⁾, Akihito Saito¹⁾, Toshiro Inaba¹⁾, Hisataka Maki¹⁾, Ryo Inuzuka²⁾, Yuichi Ishikawa³⁾, Shigeto Tsuji⁴⁾, Miyuki Shibata⁴⁾, Ryouichi Kondo⁴⁾, Akihiro Masuzawa⁴⁾, Yasutaka Hirata⁴⁾, Atsushi Yao⁵⁾, Issei Komuro¹⁾

1) Department of Cardiovascular Medicine, The University of Tokyo Hospital,

2) Department of Pediatrics, The University of Tokyo Hospital, 3) Fukuoka Children's Hospital, Dept of Cardiology,

4) Department of Cardiac Surgery, The University of Tokyo Hospital,

5) Division for Health Service Promotion, The University of Tokyo

We report a 38-year-old woman diagnosed with DORV, subpulmonary VSD and subpulmonary stenosis. A BT shunt was created at 10 days of age. Mustard operation was planned at 2 years of age, but her right atrium was small; thus, ASD creation was performed instead.

PAH developed with increasing age. Although ERA and PDE5i were administered, hypoxemia progressed, and she was referred to our hospital.

We suspected that intracardiac repair would improve hypoxemia, and PAH was relatively mild (mPAP 37 mmHg, PVR 4.9 WU); thus, surgical therapy was acceptable.

Considering the operative risk, we performed the Mustard operation and VSD partial closure. Hypoxemia was ameliorated after surgery, but refractory pulmonary edema and pleural effusion emerged. Examination revealed that severe TR occurred after surgery, along with atrial baffle stenosis.

To reduce the burden of re-operation, we planned catheter intervention for baffle stenosis before TVR. An iliac artery stent was placed, and stenosis was alleviated sufficiently. TVR was performed under the stable conditions.

A favorable outcome was achieved by careful combination therapy with surgery, catheter intervention, and drug therapy.

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東京都新宿区山吹町 358-5
株式会社国際文献社内
日本成人先天性心疾患学会事務局
TEL 03-6824-9380
FAX 03-5227-8631