JSACHD
JAPANESE SOCIETY FOR ADULT CONGENITAL HEART DISEASE

第21回
日本成人先天性心疾患学会
総会・学術集会

The 21st Annual Scientific Meeting of Japanese Society for Adult Congenital Heart Disease

抄録集

メインテーマ

『健やかな成長から成熟へ　チームで支える』

会 期

2019年1月11日(金)～13日(日)

会 場

岡山コンベンションセンター

岡山市北区駅元町14番1号

会 長

伊藤　浩

岡山大学大学院医歯薬学総合研究科 循環器内科学 教授

主催事務局

岡山大学大学院医歯薬学総合研究科 循環器内科学
事務局長　杜　徳尚

〒700-8558　岡山県岡山市北区鹿田町2-5-1
TEL：086-235-7351

第21回日本成人先天性心疾患学会総会・学術集会開催に向けて

第21回日本成人先天性心疾患学会総会・学術集会 会長
岡山大学大学院医歯薬学総合研究科 循環器内科学 教授
伊藤　浩

この度、第21回日本成人先天性心疾患学会総会・学術集会を2019年1月11日（金）から13日（日）まで、岡山コンベンションセンターで開催させていただくこととなりました。

今回のテーマは「健やかな成長から成熟へ：チームで支える」です。複雑心奇形の術後患者が続々と成人期に移行しており、受け入れる体制の構築が迫られています。解剖学的特異性、多臓器障害、妊娠出産への対応、精神的あるいは社会的サポートの必要性など多くの問題があるために、循環器内科医やメディカルスタッフもとまどっているのではないでしょうか。本学術集会はそのような方を対象に、成人性心疾患（ACHD）患者を学ぶ場を提供することを大きな目的としています。本学術集会では一会場を教育セッションに当てました。一通り聴講すればACHDの全てがわかります。エビデンスが乏しいのがACHDのもう一つの問題です。臨床上の問題や疑問に関してはシンポジウム、ディベートやケースセッションを通じて何らかの答えを出していきたいと思います。人材育成のために専門医制度の開始が迫っています。是非、本学術集会を活用してください。

ACHDは新しい領域であり、日々新しいエビデンスが明らかになっています。海外からはOechslin先生をお招きして最先端の情報をご講演いただくとともに、シンポジウムではエキスパートから最先端のエビデンスお伝えしていきたいと思います。

本学術集会はAsian Pacific Society of ACHDと併催します。アジアにおいてもACHD患者は増加していますが、人材が育っていません。アジアの同胞をできるだけ助ける必要があります。そのこともあって、先生方には一般演題やシンポジウムの抄録を英語でお書きいただきました。学術集会の中にもEnglish Oral SessionやEnglish Poster Sessionを組むことで、アジアの医師との交流を深めていただければ幸いです。

お楽しみの企画もあります。2日目夜のウルトラクイズで症例を勉強しましょう。その後の18時半からは全員懇親会（無料）があります。岡山のB級グルメをご堪能ください。

晴れの国　岡山といわれているように冬でも温暖な気候であり、食材や観光資源にも恵まれています。学会の合間に岡山の観光を是非楽しまれては如何でしょうか。

多くの皆様の参加により有意義な学会になるよう鋭意努力させていただきます。皆様に学会会場でお目にかかれることを楽しみにしております。

Welcome to the 21st Annual Scientific Meeting of Japanese Society for Adult Congenital Heart Disease

President of 21th Annual Meeting of JSACHD
Hiroshi Ito M.D., Ph.D.
Professor
Department of Cardiovascular Medicine,
Okayama University Graduate School of Medicine

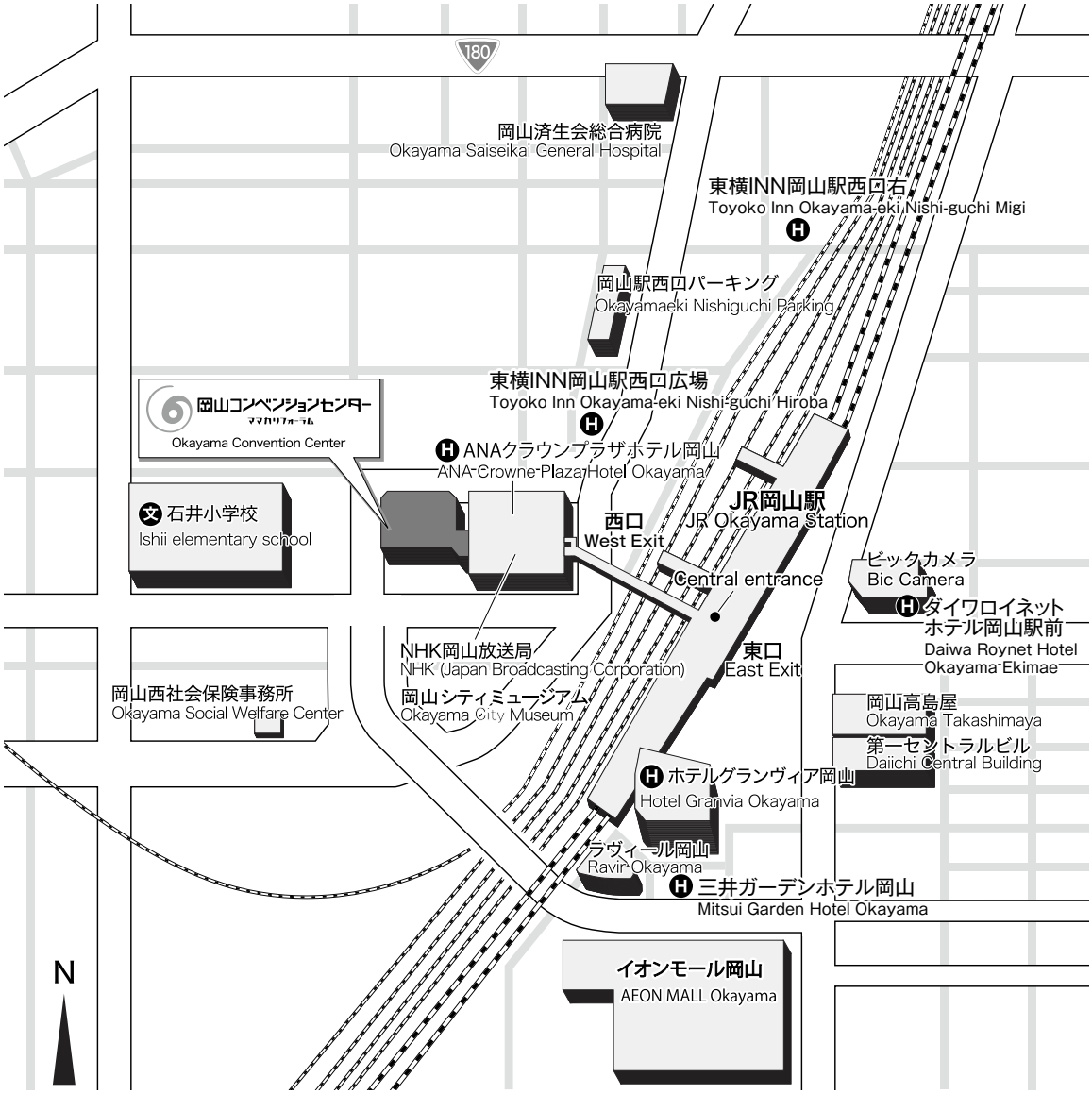
We are planning to hold the 21st scientific session of Japanese Society for Adult Congenital Heart Disease (JSACHD) from January 11th to 13th, 2019, and the 1st Asia Pacific Society for Adult Congenital Heart Disease (APSACHD) Symposium on January 12th, 2019 in Okayama Convention Center, Okayama, Japan.

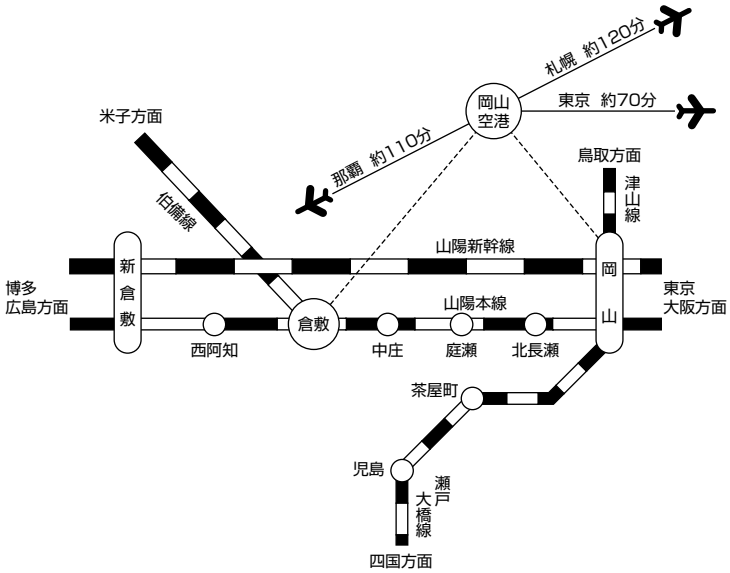
The theme of this conference is "From healthy growth to maturity: Support by multidisciplinary team". Postoperative patients with complicated congenital heart disease (CHD) are transitioning to adulthood one after another, and it is imperative to establish a system to accept them. Since there are many problems such as anatomical complexity, multiple organ failure, response to pregnancy and delivery, necessity of mental or social support, and so on, cardiologists and medical staffs may hesitate to accept them. It is the same for Asian countries. This scientific meeting aims to provide an opportunity to learn adult CHD (ACHD) from various aspects. The lack of evidence is another problem of ACHD. Regarding clinical problems and questions, some answers will be provided by experts through symposiums, debates and case sessions. We invite Dr. Oechslin to give me the latest information on ACHD. In addition to APSACHD, English sessions and English posters are held through the conference of JSACHD. You can make discussions and academic exchanges with Japanese experts.

A reception party is set after APSACHD, and you can enjoy Okayama's food and drinks and communication with Japanese doctors. Okayama is not cold even in winter, and it is blessed with delicious meals and tourism resources. How about enjoying sightseeing in Okayama by all means during or after the conference.

We are looking forward to seeing you at the conference.

会場へのアクセス／Access to the Venue





●新幹線

JR東京駅　⇔ JR岡山駅　約3時間30分
JR名古屋駅 ⇔ JR岡山駅　約1時間45分
JR新大阪駅 ⇔ JR岡山駅　約45分
JR広島駅　⇔ JR岡山駅　約40分
JR博多駅　⇔ JR岡山駅　約1時間45分

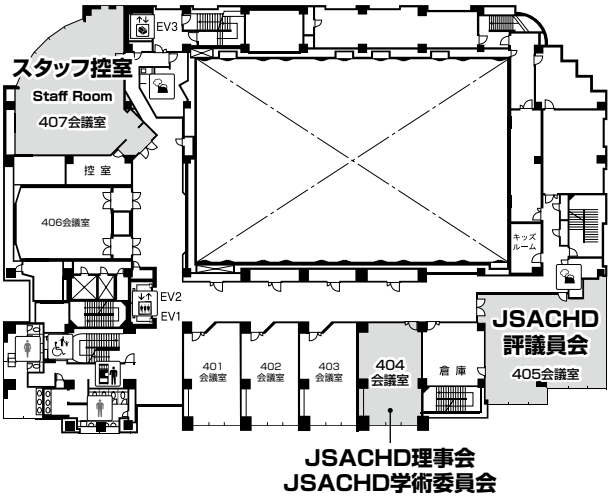
■ JRでお越しの方
JR岡山駅中央改札口から徒歩約3分

■ お車でお越しの方
岡山空港から約30分 岡山I.Cから約20分

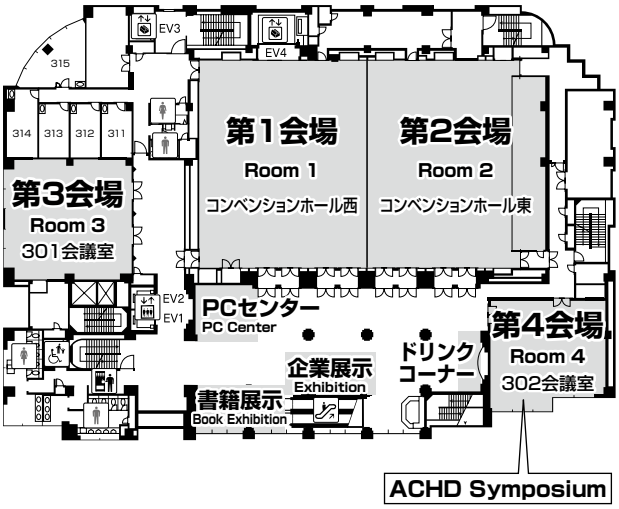
■ 飛行機でお越しの方
岡山空港より岡山駅行きのバスを
ご利用ください。
約40分（ノンストップバス約30分）
※バスはJR岡山駅西口に着きます。

会場内のご案内／Floor Guide

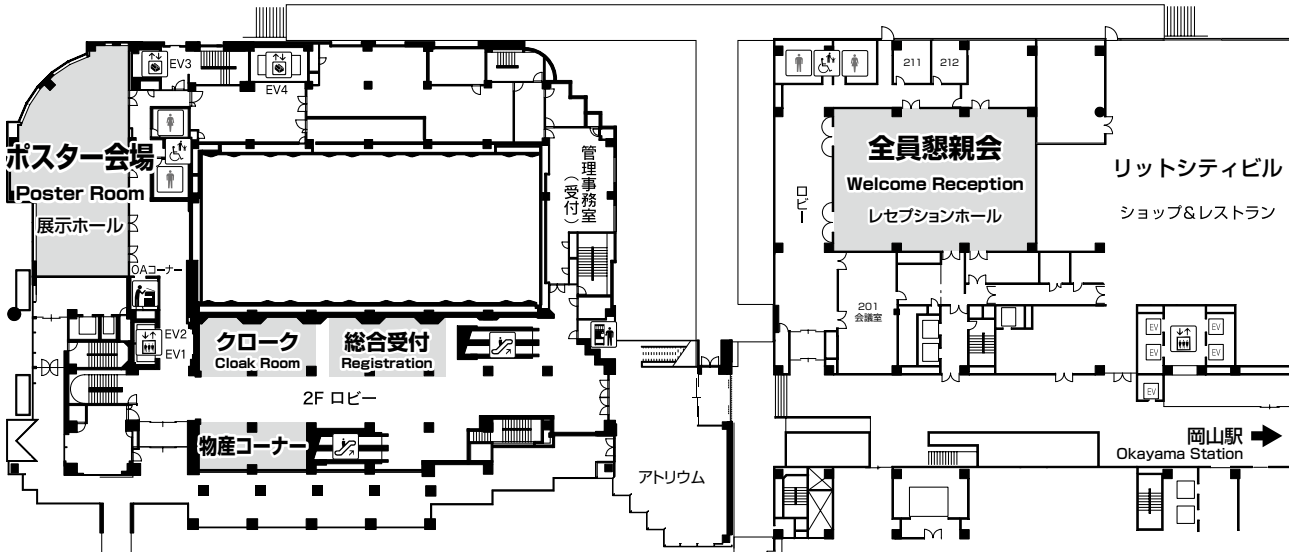
4th Floor



3rd Floor



2nd Floor



参加者の皆様へ

1. 会場

岡山コンベンションセンター
〒700-0024 岡山県岡山市北区駅元町14-1　TEL：086-214-1000

2. 参加受付

場　所：総合受付（岡山コンベンションセンター 2F ロビー）
日　時：1月11日（金）10:30～18:00
　　1月12日（土） 7:30～18:00
　　1月13日（日） 7:30～14:30

3. 参加費、抄録集販売など（現金受付のみ）

医師・歯科医師 …… 12,000円
多領域専門職（看護師・助産師・検査技師・心理士、他）…… 5,000円
一般企業 …… 12,000円
一般の方々（患者さん、ご家族）、学部学生* …… 無料
　*受付時に学生証をご呈示ください。
抄録集 …… 1,000円
※会場内では必ず参加証（兼領収書）に所属・氏名を記入のうえ、携帯してください。
※参加証（兼領収書）の再発行はできませんので大切に保管してください。
※学会員には抄録集を事前にお送りいたしますので、忘れずご持参ください。

4. 年会費・新入会受付

筆頭演者は本学会会員に限ります。未入会の方は必ず日本成人先天性心疾患学会事務局へ入会手続きを行ってください。なお、会期中は下記受付にて入会手続きを承ります。
＜日本成人先天性心疾患学会事務局＞
　〒162-0801 東京都新宿区山吹町358-5　株式会社国際文献社内
　TEL：03-6824-9380　E-mail：jsachd-post@bunken.co.jp
　電話受付：平日9:00～17:00まで（12:00～13:00を除く）
＜会期中の受付場所＞
　岡山コンベンションセンター 2F 総合受付

5. クローク

場　所：総合受付（岡山コンベンションセンター 2F ロビー）
日　時：1月11日（金）10:30～18:20
　　1月12日（土） 7:30～18:15*
　　1月13日（日） 7:30～15:40
*全員懇親会時は、懇親会会場 2F レセプションホール前のクロークをご利用ください。

6. 単位取得

・日本小児循環器学会専門医：8単位
・日本循環器学会専門医：3単位
　※申請期間は、循環器専門医認定4年目の認定更新手続き時です。
・日本小児科学会／日本専門医機構専門医（新制度）iii小児科領域講習：1単位
　※指定セッションの受講が必要です。
　　指定セッション：教育講演13「実際の診療体制と問題点」
　　日時：1月13日（日）11:00～12:00
　　会場：第2会場（岡山コンベンションセンター 3F コンベンションホール東）

7. 全員懇親会

会　場：岡山コンベンションセンター 2F レセプションホール
日　時：1月12日（土）18:30〜20:30
参加費：無料

8. ランチョンセミナーに関して

整理券の配布はございません。セミナー入場時にお弁当をお受け取りください。

9. 第1回ACHDウルトラクイズにご参加のお願い

会　場：第2会場（岡山コンベンションセンター 3F コンベンションホール東）
日　時：1月12日（土）17:00〜18:00
参加費：無料
この度、学会の会期中に施設対抗のウルトラクイズと題したクイズ大会を行います。全員懇親会の前に学会としての一体感を高めようというレクレーション的な企画となります。第1会場では学術委員会シンポジウム、第4会場ではThe 1st Asia Pacific Society for ACHD Symposiumが行われている時間ですが、少しでも多くの人に集まっていただきたいと思い企画しましたので、何卒ご容赦ください。
以下、概要でございます。

1) 主催者側が指定させて頂いた施設の他、公募でどの施設でもご自由に参加できます。また、異なる施設の方々でチームを組んでいただいても構いません。学会ホームページにて公募をさせていただいております。
2) 出題者は極秘です。
3) 1チームの構成は最大3人までです。内訳としては、どの科や職種の方が入って頂いても構いません。もちろんお一人で参加して頂いても結構です。
4) 問題数は10問を予定しており、各々4択でA〜Dの回答用紙を挙げて頂きます。
5) 問題の点数は、難易度に合わせて1点から3点までに分かれます。
6) 回答はして頂きますが、各チームの総得点はその場では発表いたしません。全員懇親会の際に大会長より発表させて頂きます。
7) 1位から5位まで豪華賞品を用意する予定です。
8) あくまでレクレーション企画ですので、ゆるい雰囲気でご参加いただければと思っています。もちろん、応援もwelcomeです。
9) 年齢制限：45歳

10. Wi-Fiコーナー

岡山コンベンションセンター内では、Wi-Fiデータ通信がご利用いただけます。
【SSID】mamakari
※パスワードの入力は不要です。
※一部、場所や回線状況によっては繋がりにくい場合がございます。

11. 会期中の問い合わせ先

総合受付（岡山コンベンションセンター 2F ロビー）
TEL：086-214-1000

12. その他

1) 会場内では、携帯電話をマナーモードに設定してください。
2) 会場内は全館禁煙です。
3) 会長の許可の無い掲示・展示・印刷物の配布・録音・写真撮影・ビデオ撮影は固くお断りいたします。

座長・演者の皆様へ

1. 進行情報

セッション	発表	質疑	総合討論
シンポジウム1	15分	3分	−
シンポジウム2	12分	−	30分
シンポジウム3〜6・8	12分	3分	−
シンポジウム7	10分	5分	−
日本循環器学会ジョイントセッション	JS1-1〜JS1-4：12分 JS1-5：20分	JS1-1〜JS1-4：3分 JS1-5：5分	−
学術委員会シンポジウム	15分	5分	−
日本心エコー図学会共同企画	12分	3分	15分
教育講演1〜13・15	20分 (質疑含む)		−
教育講演14	EL14-1：10分／EL14-2：15分 (質疑含む)		−
一般演題 (口演)	7分	3分	−
一般演題 (ポスター)	4分	2分	−
ACHD Symposium	7分	3分	

・その他の口頭セッションは担当セッションの座長の先生に一任しております。
・発表終了1分前に黄色ランプ、終了・超過時には赤色ランプを点灯してお知らせします。
・円滑な進行のため、時間厳守でお願いします。
・演台上には、モニター、キーボード、マウス、レーザーポインターを用意いたします。
演台に上がると最初のスライドが表示されますので、その後の操作は各自でおこなってください。

2. 座長の皆さまへ

Ⅰ. 口演セッション

1) 担当セッション開始予定時刻の15分前までに、会場内前方の「次座長席」にご着席ください。
2) 次座長席に着かれましたら進行席スタッフにその旨お伝えください。(進行席は次座長席近くに設置いたします。)

Ⅱ. ポスターセッション

1) 担当セッション開始予定時刻の15分前までに、ポスター会場前の「座長受付」にお立ち寄りいただき、座長用リボンと指示棒をお受取りください。
2) アナウンスはいたしませんので、担当セッションのパネルの前で待機していただき、所定の時刻より開始してください。活発な討論が行われますよう、お取り計らいをお願いいたします。
3) セッションの時間厳守にご協力をお願いいたします。
4) セッション終了後、指示棒は「座長受付」にご返却ください。

3. 発表者の皆さまへ

Ⅰ. 利益相反の開示

1) 口演発表は、発表スライドの最初(または演題・発表者などを紹介するスライドの次)に、ポスター発表は最下段に、演題発表に関連する過去1年間におけるCOIを開示してください。筆頭演者のみならず共同演者も対象となります。
2) 開示基準やCOI開示スライド等の詳細につきましては、本会ホームページをご確認ください。
http://www.med-gakkai.org/jsachd2019/coi/

Ⅱ. 口演セッション　試写・発表方法

1) 口演発表はすべてPC発表(PowerPoint)のみといたします。
英語でのご発表の場合、発表スライドは英語で作成をお願いいたします。
また、日本語でのご発表の場合でも、海外からの参加者との情報共有のため、発表スライドは出来るだけ英語で作成をお願いいたします。

2) 発表データは、Windows PowerPoint 2007～2016のバージョンで作成してください。
3) PowerPointの「発表者ツール」は使用できません。発表用原稿が必要な方は各自ご準備ください。
4) 発表時間の10分前に次演者席にご着席ください。
5) 個人情報が特定される発表は禁止します。

＜PC発表データの受付＞

学会当日に発表データの受付を行います。セッション開始30分前までに各会場前のPCセンターにて、発表データの試写ならびに受付をお済ませください。

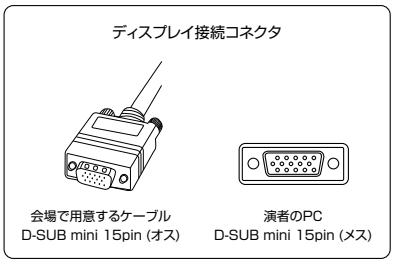
場　所：岡山コンベンションセンター　3F ロビー
日　時：1月11日（金）10:30～18:00
　　1月12日（土）　7:30～18:00
　　1月13日（日）　7:30～14:20

＜データ発表の場合＞

1) 作成に使用されたPC以外でも必ず動作確認を行っていただき、USBフラッシュメモリーでご持参ください。
2) フォントは文字化け、レイアウト崩れを防ぐため下記フォントを推奨いたします。
MSゴシック，MSPゴシック，MS明朝，MSP明朝
Arial，Century，Century Gothic，Times New Roman
3) 発表データは学会終了後、事務局で責任を持って消去いたします。

＜PC本体持込みによる発表の場合＞

1) Macintoshで作成したものと動画・音声データを含む場合は、ご自身のPC本体をお持込みください。
2) 会場で用意するPCケーブルコネクタの形状は、D-SUB mini 15pin（図参照）です。この出力端子を持つPCをご用意いただくか、この形状に変換するコネクタを必要とする場合には必ずご持参ください。デジタル出力（HDMI）の出力端子しか無いPCはHDMI→D-SUBの変換アダプターも必要です。電源ケーブルもお忘れなくお持ちください。



（図）

3) 再起動をすることがありますので、パスワード入力は“不要”に設定してください。
4) スクリーンセーバーならびに省電力設定は事前に解除しておいてください。
5) 動画データ使用の場合は、Windows Media Playerで再生可能であるものに限定いたします。

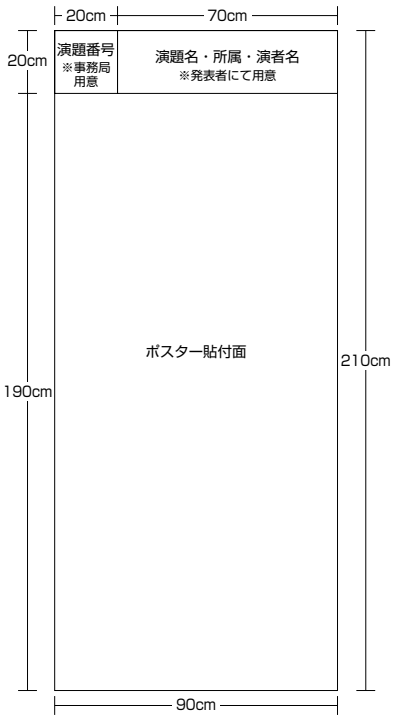
Ⅲ．ポスターセッション

1) ポスターの貼付スペースは、横90cm×縦190cmの範囲内とします。タイトル・氏名・所属は、パネル上方の演題番号の横に横70cm×縦20cmでおさまるようにご自身でご準備ください。演題番号と画鋲は、事務局で準備いたします。
2) 英語でのご発表の場合、掲示ポスターは英語で作成をお願いいたします。また、日本語でのご発表の場合でも、海外からの参加者との情報共有のため、掲示ポスターは出来るだけ英語で作成をお願いいたします。
3) ポスター貼付・撤去スケジュール

発表日	貼付	撤去
1月11日（金）	10:30～12:40	17:54～18:15
1月12日（土）	7:30～9:30	16:54～18:00
1月13日（日）	7:30～9:30	12:19～13:40

※ポスター撤去時間は厳守してください。時間になり次第、処分いたします。

4) 演者受付は不要です。セッション開始15分前までにご自身のパネルの前で待機してください。



理事会・総会・各種会合

1. JSACHD理事会

日　時：1月11日（金）10:00～12:00
会　場：4F 404会議室

2. JSACHD評議員会

日　時：1月11日（金）12:00～12:30
会　場：4F 405会議室

3. JSACHD学術委員会

日　時：1月12日（土）7:30～8:10
会　場：4F 404会議室

4. JSACHD総会

日　時：1月13日（日）13:45～14:15
会　場：第1会場（3F コンベンションホール西）

5. 看護ワーキング情報交換会

日　時：1月13日（日）8:30～10:00
会　場：第3会場（3F 301会議室）

6. 全員懇親会

日　時：1月12日（土）18:30～20:30
会　場：2F レセプションホール
参加費：無料

General Information for Participants

1. Conference Venue

Okayama Convention Center
(address: 14-1, Ekimotomachi, Kita-ku, Okayama city, Okayama 700-0024, Japan)

2. On-site Registration

Venue: "Lobby", the 2nd floor of Okayama Convention Center
Registration Hours: Day 1, Friday, January 11th 10:30-18:00
Day 2, Saturday, January 12th 7:30-18:00
Day 3, Sunday, January 13th 7:20-14:30

3. Registration Fee

Please note that credit cards are not available. We accept cash, Japanese Yen, only.
Medical Doctor, Dentist …… 12,000 JPY
Nurse, Technician and Psychologist etc. …… 5,000 JPY
Company Personnel …… 12,000 JPY
Patient and their Family and Undergraduate student* …… Free of charge
*Student needs to provide their valid student ID cards.
All registered delegate, and accompanying persons are requested to wear their registration card during the period of the conference and its social events.

4. Cloakroom

A cloakroom service for coats and reasonably-sized luggage is available the following hours. It is located in the 2nd floor. Items of value should not be left in the cloakroom. Please make sure you collect all your belongings at the end of each day.
Day 1, Friday, January 11th 10:30-18:20
Day 2, Saturday, January 12th 7:30-18:15*
Day 3, Sunday, January 13th 7:30-15:40
* Please note that a cloak room service, located in the front of "Reception Hall", 2nd floor of the congress venue, is available for participants of Welcome Reception to be held at 18:30-20:30 on January 12.

5. Welcome Reception

Venue: Reception Hall, 2nd floor of Okayama Convention Center
Schedule: 18:30-20:30, Saturday, January 12
Participation Fee: Free of charge

6. Luncheon Seminar

We do not provide the number of tickets for lunch boxes.

7. Wi-Fi

Free Wi-Fi "mamakari" is available without pass word.

8. Information Desk

Venue: "Lobby", 2nd floor of Okayama Convention Center
Phone: 086-214-1000

9. Others

1) Please turn your mobile phones on silent mode during the congress.
2) Smoking in the congress venue is prohibited.
3) Photography, recording, distribution of printed matters, exhibition and positing without the congress president's approval are prohibited.

Guideline for Chairs and Speakers

1. Time Allocation

Session	Presentation (minutes)	Discussion (minutes)	General Discussion (minutes)
Symposium 1	15	3	nil
Symposium 2	12	nil	30
Symposium 3-6 & 8	12	3	nil
Symposium 7	10	5	nil
Joint Session with Japanese Circulation Society	JS1-1～1-4：12 JS1-5：20	JS1-1～1-4：3 JS1-5：5	nil
Symposium by Scientific Committee	15	5	nil
Cooperative Project with Japanese Society of Echocardiology	12	3	15
Educational Lecture 1-13 & 15	20 including discussion		nil
Educational Lecture14	EL14-1：10 minutes including discussion EL14-2：15 minutes including discussion		nil
Oral Session	7	3	nil
Poster Session	4	2	nil
APSACHD Symposium	7	3	nil

・The time allocation of oral sessions except for the above will be left by the chairs.
・When the yellow lamp lights, it means that one minute remains. When the red lamp lights, it means the end of your presentation.
・Please be punctual and carefully follow the allotted time limits.·
・The speakers' podium is equipped PC, monitor, keyboard, mouse and laser pointer

2. To Chairs

Ⅰ. Oral Session

1) Chairs should take a seat in the front row of the room specially reserved for the next session's chair at least 15 minutes prior to the session that he/she is scheduled to chair.
2) Please inform a congress staff of chairs' name when arriving the room.

Ⅱ. Poster Session

1) Chairs should register and take a ribbon and pointer at the poster registration desk, located in front of poster session venue, at least 15 minutes prior to the session. After your session, please return the pointer to the registration desk.
2) As there will be no announcement or cue, please start the session at the appointed time. We request your cooperation to ensure that your session proceeds according to the prescribed time limit/schedule.

3. To Presenters

Ⅰ. Disclosure of COI

1) COI status will be disclosed at the beginning of the slide or poster at the time of the presentation.
2) Please refer to the sample slides of COI disclosure and confirm the details described in the Guidelines for conflict of interest (COI) at the congress website. http://www.med-gakkai.org/jsachd2019/coi/.

Ⅱ. Oral Session

1) All presentation materials should be prepared in Microsoft PowerPoint 2007 to 2016 version for Windows PC.
2) It's not available to use "Presentation Tool" in the Power Point. Please prepare the speech draft by yourself.
3) When the presenter ahead of you takes the stage, please be seated in the Next Speaker seat at least 10 minute before your presentation starts.
4) It is prohibited to specify personal information in your presentation.

<PC Preview Desk>

Speakers should preview their presentation and complete a run-through of the connection and operation at the PC Preview Desk located on the 3rd floor, at least 30 minutes before their sessions start.

PC Preview Desk Opening Hours: Day 1, Friday, January 11th 10:30-18:00
Day 2, Saturday, January 12th 7:30-18:00
Day 3, Sunday, January 13th 7:30-14:20

<Notes for presentation data>

1) Save your presentation data on USB memory. Please be sure that your file is compatible with the Windows operating system.
2) Please use only the standard fonts provided with Microsoft PowerPoint for Windows as follows: Arial, Century, Century Gothic and Times New Roman
3) After the conference, all presentation data installed on the conference supplied PCs will be deleted.

< Notes for speakers with their own PC >

1) You are advised to bring your own laptop PC if your presentation materials including video/movies cannot be prepared in the above mentioned format or your are using a Mcintosh.
2) Your computer must be equipped with a Dsub 15 pin video output. Remember to bring your own power adapter and cables.
3) Your password should be inactivated.
4) All energy-saving functions such as screen-savers and sleep/power-saving modes should be disabled on the PC to be used for the presentation.
5) In case of use of movies, the version of Windows Media Player and the movie file must be included in the SAME FOLDER as the presentation file.

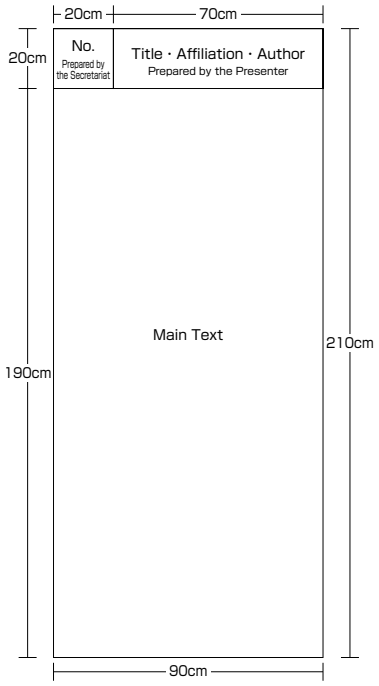
Ⅲ. Poster Session

1) The secretariat will prepare a panel with your poster number. Tacks for putting up posters will be prepared at each panel. Poster Mounting area is H190cm × W90cm. Please prepare a slip with title, name(s) of author(s), and affiliation(s) with the size of H20cm × W70cm.
2) Schedule of Mounting and Removal

Date	Poster Mounting Hours	Removal Hours
Friday, January 11th	10:30-12:40	17:54-18:15
Saturday, January 12th	7:30-9:30	16:54-18:00
Sunday, January 13th	7:30-9:30	12:19-13:40

Note: All posters should be taken down from the boards by the above time. Conference secretariat will not assume responsibility for possible loss of posters left on the board.

3) It is not necessary to register at the registration desk. Please stand by 15 minutes before the session starts.



日程表

1月11日 (金)

第1会場	第2会場	第3会場	第4会場	ポスター会場
3F コンベンションホール西	3F コンベンションホール東	3F 301会議室	3F 302会議室	2F 展示ホール

第1会場（3F コンベンションホール西）

12:30～12:40 **開会の辞**

12:40～13:40
ランチョンセミナー1
座長：筒井裕之
演者：矢野雅文
共催：ノバルティス ファーマ㈱ メディカル本部

13:50～15:20
シンポジウム1
「Failing Fontan」
座長：先崎秀明／坂本一郎
演者：竹蓋清高／島田衣里子
豊野学朋／帯刀英樹
元木博彦

15:30～16:30
コーヒーブレイクセミナー1
座長：庄田守男
演者：西井伸洋
共催：日本メドトロニック㈱

16:40～18:10
シンポジウム2
「ご当地ACHDセンター診療の実際；上手くいってることはもちろん、そうでないことも全て見せます」
座長：伊藤　浩／平田健一
演者：赤木禎治／松本賢亮
椎名由美／元木博彦
坂本一郎

第2会場（3F コンベンションホール東）

12:40～13:40
ランチョンセミナー2
座長：中村一文
演者：赤木　達
共催：グラクソ・スミスクライン㈱

13:50～14:50
教育講演1
「先天性心疾患、まずは解剖と病態を識る その1」
座長：白石　公
演者：仁田　学／松本賢亮
中川直美

14:55～15:55
教育講演2
「先天性心疾患、まずは解剖と病態を識る その2」
座長：三浦　大
演者：久持邦和／近藤麻衣子
櫻井寛久

16:00～17:00
教育講演3
「ACHDの診療－Back To Basics」
座長：森　善樹
演者：水野　篤／相馬　桂
岡　岳文

17:10～18:10
教育講演4
「CHDの手術と成人期での注意点 その1」
座長：市川　肇
演者：笠原真悟／上村秀樹
黒子洋介

第3会場（3F 301会議室）

11:30～12:30
一般演題（口演）1
「カテーテルインターベンション」
座長：大月審一／佐地真育
ｺﾒﾝﾃｰﾀｰ：中川晃志

13:50～14:50
一般演題（口演）2
「術後遠隔期の諸問題とチーム医療」
座長：新井禎彦／堀端洋子
ｺﾒﾝﾃｰﾀｰ：岡本吉生

15:20～16:00 **ディベート1**
「Fontanでの抗凝固療法：必要 vs. 不要」
座長：安田謙二
演者：栄徳隆裕／根岸　潤

16:00～17:00
一般演題（口演）3
「Fontan」
座長：満下紀恵／中野俊秀
ｺﾒﾝﾃｰﾀｰ：戸田孝子

第4会場（3F 302会議室）

10:00～12:00
JSACHD理事会
(4F 404会議室)

12:00～12:30
JSACHD評議員会
(4F 405会議室)

13:50～14:50
Case Session 1
座長：上村秀樹
演者：Alexander Van De Bruaene
June Huh
ｺﾒﾝﾃｰﾀｰ：Erwin Oechslin

15:00～16:30
Oral Presentation 1 (English)
「Long-term outcomes in ACHD」
座長：篠原徳子／杜　徳尚
基調講演：Erwin Oechslin
ｺﾒﾝﾃｰﾀｰ：神谷千津子
Erwin Oechslin

16:30～17:00
皆で考えよう。この症例をどうする？1
座長：白井丈晶　演者：川松直人
ｺﾒﾝﾃｰﾀｰ：安田謙二／久持邦和
圓尾文子／仁田　学

ポスター会場（2F 展示ホール）

10:30～12:40
ポスター貼付

12:40～17:00
ポスター閲覧

17:00～17:54
Poster Presentation (English)
PE1～PE3

17:54～18:15
ポスター撤去

ハンズオンセミナー
「ACHDにおける心腔内エコーの応用」
実施企業：ジョンソン・エンド・ジョンソン㈱
日本光電工業㈱
会期中、3F 企業展示ブースにて展示機器を体験していただけます。

日程表

1月12日 (土)

第1会場 (3F コンベンションホール西)

8:30～10:00
シンポジウム3
「ACHDの肺高血圧治療の最前線」
座長：中村一文／八尾厚史
演者：相馬　桂／松原広己
赤木　達／根本慎太郎
椎名由美／坂崎尚徳

10:05～11:05
スポンサードセミナー
座長：三好　亨
演者：東　幸仁
共催：田辺三菱製薬㈱

11:10～12:40
日本循環器学会ジョイントセッション
「先天性心疾患の成人への移行医療に関する提言を具体化する」
座長：三谷義英／赤阪隆史
演者：三谷義英／安河内聰
八尾厚史／青木美千代
谷口顕信

12:50～13:50
ランチョンセミナー3
座長：伊藤　浩
演者：赤木禎治
共催：第一三共㈱

13:55～14:25　**会長講演**
座長：丹羽公一郎
演者：伊藤　浩

14:35～15:15
海外招聘講演1
座長：森田紀代造
演者：Erwin Oechslin

15:25～16:25
コーヒーブレイクセミナー2
座長：伊藤　浩
演者：上村史朗
共催：日本ベーリンガーインゲルハイム㈱

16:30～18:00
学術委員会シンポジウム
「実臨床のcontroversial」
座長：稲井　慶／大内秀雄
演者：桑原政成／肥後太基
笠原真悟／神谷千津子

第2会場 (3F コンベンションホール東)

8:30～9:30
教育講演5
「CHDの手術と成人期での注意点 その2」
座長：松尾浩三
演者：小谷恭弘／本淨修己
猪飼秋夫

9:35～10:35
教育講演6
「Fontan循環を理解する」
座長：安河内聰
演者：本淨修己／大内秀雄
大西秀樹

10:40～11:40
教育講演7
「ACHDのイメージング」
座長：城戸佐知子
演者：杜　徳尚／椎名由美
板谷慶一

11:40～12:40
教育講演8
「心臓カテーテル検査」
座長：鎌田政博
演者：加藤温子／馬場健児
佐地真育

12:50～13:50
ランチョンセミナー4
座長：赤阪隆史
演者：小板橋紀通
共催：ブリストル・マイヤーズスクイブ㈱
ファイザー㈱

13:55～14:55
教育講演9
「冠動脈疾患と結合織異常」
座長：市田蕗子
演者：山村健一郎／小林　徹
犬塚　亮

14:55～15:55
教育講演10
「妊娠」
座長：桂木真司
演者：神谷千津子／照井克生
兵藤博信

15:55～16:55
教育講演11
「不整脈」
座長：簇　義仁
演者：宮﨑　文／芳本　潤
西井伸洋

17:00～18:00
第1回 ACHDウルトラクイズ

第3会場 (3F 301会議室)

8:30～9:30
シンポジウム4
「ACHD患者の妊娠・出産そして授乳の問題を考える」
座長：池田智明／増山　寿
演者：田中博明／堀内　縁
衛藤英理子／桂木真司

9:35～10:35
一般演題(口演)4
「妊娠・出産・診療支援」
座長：富松宏文／小口秀紀
ｺﾒﾝﾃｰﾀｰ：堀内　縁

10:40～11:40
一般演題(口演)5
「予後」
座長：櫻井　一／黒子洋介
ｺﾒﾝﾃｰﾀｰ：永田　弾

11:40～12:40
シンポジウム5
「ACHDの不整脈治療」
座長：庄田守男／立野　滋
演者：立野　滋／岡嶋克則
西井伸洋／庄田守男

12:50～13:50
ランチョンセミナー5
座長：小垣滋豊
演者：土井　拓
共催：日本新薬㈱
アクテリオン ファーマシューティカルズ ジャパン㈱

13:55～14:35　**ディベート2**
「TOF術後PRに対する成人期PVR：人工弁の種類・サイズの選択」
座長：河田政明
演者：小谷恭弘／板谷慶一

14:45～15:35
一般演題(口演)6
「心不全」
座長：田邊一明／福本義弘
ｺﾒﾝﾃｰﾀｰ：稲葉俊郎

15:55～16:55
皆で考えよう。この症例をどうする?2
座長：小板橋俊美／福田旭伸
演者：小永井奈緒／藤田鉄平
ｺﾒﾝﾃｰﾀｰ：立野　滋／河田政明
森田紀代造／
神谷千津子

第4会場 (3F 302会議室)

7:30～8:10
JSACHD学術委員会
(4F 404会議室)

8:55～18:00
The 1st Asia Pacific Society for ACHD Symposium (English)

ポスター会場 (2F 展示ホール)

7:30～9:30
ポスター貼付

9:30～16:00
ポスター閲覧

16:00～16:54
ポスター発表
PJ1～PJ4

16:54～18:00
ポスター撤去

ハンズオンセミナー
「ACHDにおける心腔内エコーの応用」
実施企業：ジョンソン・エンド・ジョンソン㈱
日本光電工業㈱
会期中、3F 企業展示ブースにて展示機器を体験していただけます。

18:30～20:30　**全員懇親会**（2F レセプションホール）

日程表

1月13日㊐

第1会場（3F コンベンションホール西）

8:30～9:30
シンポジウム6
「カテーテルインターベンションの最新治療」
座長：坂本一郎／赤木禎治
演者：大山伸雄／藤本一途
佐地真育／高谷陽一

9:35～11:05
日本心エコー図学会共同企画
「ACHDの病態の定量評価に挑む」
座長：石津智子／杜　徳尚
演者：新居正基／山澤弘州
石津智子／梶山　葉
杜　徳尚

11:10～11:40　**海外招聘講演2**
座長：筒井裕之
演者：Mei-Hwan Wu

11:45～12:30
成人先天性心疾患専門医制度解説セッション
座長：丹羽公一郎
演者：市田蕗子／森田紀代造
八尾厚史

12:40～13:40
ランチョンセミナー6
座長：丹羽公一郎
演者：八尾厚史
共催：アクテリオン ファーマシューティカルズ ジャパン㈱
日本新薬㈱

13:45～14:15
JSACHD総会

14:20～15:20
シンポジウム8
「ACHDの緩和医療」
座長：丹羽公一郎／福嶌教偉
演者：水野　篤／中澤　誠
福嶌教偉／河野由枝

15:20～15:30　**閉会の辞**

16:00～17:00
市民公開講座
「大人になった先天性心疾患患者さんへ 困ったこと・不安なことがありますか？」
座長：伊藤　浩／笠原真悟
演者：赤木禎治／小谷恭弘
榎本淳子／患者様

第2会場（3F コンベンションホール東）

8:30～9:50
一般演題（口演）7
「外科」
座長：宮地　鑑／大嶋義博
ｺﾒﾝﾃｰﾀｰ：帯刀英樹

9:55～10:55
教育講演 12
「肺高血圧と心不全－How to tackle critical situations」
座長：坂崎尚徳
演者：赤木　達／建部俊介
坂本一郎

11:00～12:00
教育講演 13*
「実際の診療体制と問題点」
座長：賀藤　均
演者：落合亮太／元木博彦
檜垣高史

12:05～12:30　**教育講演 14**
「知っておきたいコメディカルの知識」
座長：落合亮太
演者：祇園由美／榎本淳子

12:40～13:40
ランチョンセミナー7
座長：檜垣高史
演者：高谷陽一／七里　守
共催：日本ライフライン㈱

14:20～15:20
教育講演 15
「細部まで見落とさない－Common oversight」
座長：脇　研自
演者：山岸敬幸／高谷陽一
大森一弘

第3会場（3F 301会議室）

8:30～10:00
看護ワーキング情報交換会

10:05～10:45　**ディベート3**
「複雑ACHDでの妊娠：ここまで可能 vs. ここから不可能」
座長：市川　肇
演者：吉松　淳／兵藤博信

10:50～12:20
シンポジウム7
「小児心臓血管外科手術の長期成績」
座長：笠原真悟／山岸正明
演者：帆足孝也／小谷恭弘
松久弘典／小田晋一郎

14:20～15:20
一般演題（口演）8
「患者ケア」
座長：水野芳子／木島康文
ｺﾒﾝﾃｰﾀｰ：森　善樹

第4会場（3F 302会議室）

8:30～10:10
Oral Presentation 2 (English)
「Surgical and interventional strategy for ACHD」
座長：小谷恭弘／米田正始
基調講演：Worakan Promphan
ｺﾒﾝﾃｰﾀｰ：平田康隆
Worakan Promphan

10:10～10:50
Case Session 2
座長：赤木禎治
演者：Lucy Eun／牧　尉太
ｺﾒﾝﾃｰﾀｰ：Seung Woo Park

ポスター会場（2F 展示ホール）

7:30～9:30
ポスター貼付

9:30～11:25
ポスター閲覧

11:25～12:19
ポスター発表
PJ5～PJ8

12:19～13:40
ポスター撤去

ハンズオンセミナー
「ACHDにおける心腔内エコーの応用」
実施企業：ジョンソン・エンド・ジョンソン㈱
日本光電工業㈱
会期中、3F 企業展示ブースにて展示機器を体験していただけます。

＊日本小児科学会／日本専門医機構専門医（新制度）指定セッション

Time Table

January 11 (Friday)

Room 1 3F Convention Hall West	Room 2 3F Convention Hall East	Room 3 3F Conference room 301	Room 4 3F Conference room 302	Poster Room 2F Exhibition Hall
12:30-12:40 **Opening remark**		11:30-12:30 **Oral Presentation 1** "Catheter intervention" Chairs: Shinich Ohtsuki Saji Mike		10:30-12:40 Poster Mounting
12:40-13:40 **Luncheon Seminar 1** Chair: Hiroyuki Tsutsui Speaker: Masafumi Yano Cosponsored by Novartis Pharma K.K. Medical Division	12:40-13:40 **Luncheon Seminar 2** Chair: Kazufumi Nakamura Speaker: Satoshi Akagi Cosponsored by Glax SmithKline K.K.			12:40-17:00
13:50-15:20 **Symposium 1** "Failing Fontan" Chairs: Hideaki Senzaki Ichiro Sakamoto	13:50-14:50 **Educational Lecture 1** "Anatomy and pathology of CHD 1" Chair: Isao Shiraishi	13:50-14:50 **Oral Presentation 2** "Long-term issues after surgery and multi-disciplinary care" Chairs: Sadahiko Arai Yoko Horibata	13:50-14:50 **Case session 1 in English** Chair: Hideki Uemura	Poster Viewing
	14:55-15:55 **Educational Lecture 2** "Anatomy and pathology of CHD 2" Chair: Masaru Miura	15:20-16:00 **Debate 1** Chair: Kenji Yasuda	15:00-16:30 **Oral Presentation 1 in English** "Long-term outcomes in ACHD" Chairs: Tokuko Shinohara Norihisa Toh	
15:30-16:30 **Coffee break Seminar 1** Chair: Morio Shoda Speaker: Nobuhiro Nishii Cosponsored by Medtronic Japan Co., Ltd.	16:00-17:00 **Educational Lecture 3** "Management of Adults with Congenital Heart Disease: Back to Basic" Chair: Yoshiki Mori	16:00-17:00 **Oral Presentation 3** "Fontan" Chairs: Norie Mitsushita Toshihide Nakano	16:30-17:00 **Case Discussion 1** Chair: Takeaki Shirai	
16:40-18:10 **Symposium 2** "How to manage ACHD center: Experience of the advanced center" Chairs: Hiroshi Ito Ken-ichi Hirata	17:10-18:10 **Educational Lecture 4** "CHD Surgery 1; What we should know about CHD Surgery for ACHD Care" Chair: Hajime Ichikawa			17:00-17:54 **Poster Presentation in English** PE1-PE3 17:54-18:15 Poster Removal

Time Table

January 12 (Saturday)

	Room 1 3F Convention Hall West	Room 2 3F Convention Hall East	Room 3 3F Conference room 301	Room 4 3F Conference room 302	Poster Room 2F Exhibition Hall
9:00	8:30-10:00 **Symposium 3** "The Forefront of Therapeutic Strategy for ACHD-PAH" Chairs: Kazufumi Nakamura Atsushi Yao	8:30-9:30 **Educational Lecture 5** "CHD Surgery 2; What we should know about CHD Surgery for ACHD Care" Chair: Kozo Matsuo	8:30-9:30 **Symposium 4** "Management during pregnancy, delivery and breastfeeding in women with adult congenital heart disease" Chairs: Tomoaki Ikeda Hisashi Masuyama	**The 1st Asia Pacific Society for ACHD Symposium (English)** 8:55-9:00 **Welcome Remarks** 9:00-10:20 **Oral Abstracts** Chairs: Teiji Akagi Gu Hong	7:30-9:30 Poster Mounting
10:00		9:35-10:35 **Educational Lecture 6** "Current management of adults with Fontan circulation" Chair: Satoshi Yasukochi	9:35-10:35 **Oral Presentation 4** "Pregnancy, delivery and medical care system" Chairs: Hirofumi Tomimatsu Hidenori Oguchi		9:30-16:00
11:00	10:05-11:05 **Sponsored Seminar** Chair: Toru Miyoshi Speaker: Yukihito Higashi Cosponsored by Mitsubishi Tanabe Pharma Corporation	10:40-11:40 **Educational Lecture 7** "ACHD imaging: choice of modalities" Chair: Sachiko Kido	10:40-11:40 **Oral Presentation 5** "Prognosis" Chairs: Hajime Sakurai Yosuke Kuroko	10:20-11:10 **ACHD Management & Education System** Chairs: Seung Woo Park Atsushi Yao	
12:00	11:10-12:40 **Joint Session with Japanese Circulation Society** "Toward realizing the statements on the transition issues of congenital heart disease" Chairs: Yoshihide Mitani Takashi Akasaka	11:40-12:40 **Educational Lecture 8** "The role of cardiac catheterization in ACHD" Chair: Masahiro Kamada	11:40-12:40 **Symposium 5** "Arrhythmia Management for ACHD" Chairs: Morio Shoda Shigeru Tateno	11:10-11:50 **Optimal Arrhythmia Management in ACHD** Chairs: Aya Miyazaki Mei Hwan Wu 12:00-12:40 **Surgical review for ACHD specialists** Chairs: Shingo Kasahara Gi Beom Kim	Poster Viewing
13:00	12:50-13:50 **Luncheon Seminar 3** Chair: Hiroshi Ito Speaker: Teiji Akagi Cosponsored by Daiichi Sankyo Company, Limited.	12:50-13:50 **Luncheon Seminar 4** Chair: Takashi Akasaka Speaker: Norimichi Koitabashi Cosponsored by Bristol-Myers Squibb K.K. / Pfizer Japan Inc.	12:50-13:50 **Luncheon Seminar 5** Chair: Shigetoyo Kogaki Speaker: Hiraku Doi Cosponsored by Nippon Shinyaku Co., Ltd. / Actelion Pharmaceuticals Japan Ltd.		
14:00	13:55-14:25 **Presidential Lecture** Chair: Koichiro Niwa 14:35-15:15 **Int'l Invited Lecture 1 in English** Chair: Kiyozo Morita Speaker: Erwin Oechslin	13:55-14:55 **Educational Lecture 9** "Connective tissue disease and coronary artery disease" Chair: Fukiko Ichida	13:55-14:35 **Debate 2** Chair: Masaaki Kawada 14:45-15:35 **Oral Presentation 6** "Heart failue" Chairs: Kazuaki Tanabe Yoshihiro Fukumoto	14:00-14:30 **Pulmonary Hypertension in Asia** Chairs: Kei Inai Worakan Promphan	
15:00		14:55-15:55 **Educational Lecture 10** "Pregnancy in ACHD" Chair: Shinji Katsuragi		15:20-16:20 **Management of RVOT in Repaired TOF** Chairs: Jae Yong Choi Hideaki Senzaki	
16:00	15:25-16:25 **Coffee Break Seminar 2** Chair: Hiroshi Ito Speaker: Shiro Uemura Cosponsored by Nippon Boehringer Ingelheim	15:55-16:55 **Educational Lecture 11** "Management of arrhythmia in ACHD" Chair: Yoshihito Hata	15:55-16:55 **Case Discussion 2** Chairs: Toshimi Koitabashi Terunobu Fukuda	16:20-17:20 **Special Issues in ACHD** Chairs: Lucy Eun, et al.	16:00-16:54 **Poster Presentation** PJ1-PJ4
17:00	16:30-18:00 **Symposium with ACHD Academic Committee** "Controversial topics in the ACHD clinical practice" Chairs: Kei Inai Hideo Ohuchi	17:00-18:00 **The 1st ACHD Ultra Quiz**		17:20-18:00 **Future Prescribe of ACHD management in Asia** Chairs: Pultra Sukman Koichiro Niwa	16:54-18:00 Poster Removal
18:00					

18:30-20:30 **Welcome Reception** (Reception Hall, 2nd floor)

Time Table

January 13 (Sunday)

Room 1 — 3F Convention Hall West

8:30-9:30
Symposium 6
"Up to date of catheter intervention for ACHD patients"
Chairs: Ichiro Sakamoto
Teiji Akagi

9:35-11:05
Joint Seminar with Japanese Society of Echocardiography
"Current and future challenges in quantification of ACHD Echo"
Chairs: Tomoko Ishizu
Norihisa Toh

11:10-11:40
Int'l Invited Lecture 2 in English
Chair: Hiroyuki Tsutsui
Speaker: Mei-Hwan Wu

11:45-12:30
Guidance report of Subspecialty training systems of Japanese Society for ACHD
Chair: Koichiro Niwa

12:40-13:40
Luncheon Seminar 6
Chair: Koichiro Niwa
Speaker: Atsushi Yao
Cosponsored by Actelion Pharmaceuticals Japan Ltd. / Nippon Shinyaku Co., Ltd.

13:45-14:15
JSACHD General Assembly

14:20-15:20
Symposium 8
"End of life care"
Chairs: Koichiro Niwa
Norihide Fukushima

15:20-15:30 **Closing Remark**

16:00-17:00
Open Conference with Citizens
Chairs: Hiroshi Ito
Shingo Kasahara

Room 2 — 3F Convention Hall East

8:30-9:50
Oral Presentation 7
"Surgery"
Chairs: Kagami Miyaji
Yoshihiro Oshima

9:55-10:55
Educational Lecture 12
"Pulmonary hypertension and heart failure in ACHD"
Chair: Hisanori Sakazaki

11:00-12:00
Educational Lecture13
"Ideal clinical care system in ACHD"
Chair: Hitoshi Kato

12:05-12:30
Educational Lecture 14
"Up-to-date information"
Chair: Ryota Ochiai

12:40-13:40
Luncheon Seminar 7
Chair: Takashi Higaki
Speakers: Youichi Takaya
Mamoru Nanasato
Cosponsored by Japan Lifeline Co., Ltd.

14:20-15:20
Educational Lecture 15
"Common oversight in ACHD care"
Chair: Kenji Waki

Room 3 — 3F Conference room 301

8:30-10:00
Nursing Care Session

10:05-10:45
Debate 3
Chair: Hajime Ichikawa

10:50-12:20
Symposium 7
"Long term outcome following repair of congenital heart disease"
Chairs: Shingo Kasahara
Masaaki Yamagishi

14:20-15:20
Oral Presentation 8
"Patient care"
Chairs: Yoshiko Mizuno
Yasufumi Kijima

Room 4 — 3F Conference room 302

8:30-10:10
Oral Presentation 2 in English
"Surgical and interventional strategy for ACHD"
Chair: Yasuhiro Kotani
Masashi Komeda

10:10-10:50
Case session 2 in English
Chair: Teiji Akagi

Poster Room — 2F Exhibition Hall

7:30-9:30
Poster Mouting

9:30-11:25
Poster Viewing

11:25-12:19
Poster Presentation
PJ5-PJ8

12:19-13:40
Poster Removal

プログラム

1月11日 ㊎　　第1会場 [3F コンベンションホール西]

12：30～12：40　開会の辞

会長：**伊藤 浩**（岡山大学大学院医歯薬学総合研究科 循環器内科学）

12：40～13：40　ランチョンセミナー1

座長：**筒井 裕之**（九州大学 大学院医学研究院 循環器内科学）
共催：ノバルティス ファーマ株式会社 メディカル本部

LS1　**細胞内カルシウム制御による心不全・不整脈治療戦略**
矢野 雅文
山口大学大学院医学系研究科 器官病態内科学

13：50～15：20　シンポジウム1

Failing Fontan

座長：**先崎 秀明**（北里大学医学部 小児循環器集中治療学）
坂本 一郎（九州大学大学院医学研究院 循環器内科学）

S1-1　**沖縄におけるFontan術後成人患者の現状と課題**
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

S1-2　**女性とFontan**
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

S1-3　**術前から予想するfailing Fontan**
豊野 学朋
秋田大学 小児科学講座

S1-4　**成人期フォンタンに対する外科治療**
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

S1-5　**Failing Fontan予防のための内科治療**
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

15：30～16：30　コーヒーブレイクセミナー1

座長：庄田 守男（東京女子医科大学 循環器内科／信州大学医学部 循環器内科学教室）
共催：日本メドトロニック株式会社

CBS1　先天性心疾患患者におけるデバイス治療の有用性について
西井 伸洋
岡山大学大学院医歯薬学総合研究科 循環器内科学

16：40～18：10　シンポジウム2

ご当地ACHDセンター診療の実際；上手くいってることはもちろん、そうでないことも全て見せます

座長：伊藤　浩（岡山大学大学院医歯薬学総合研究科 循環器内科学）
平田 健一（神戸大学大学院医学研究科 循環器内科学分野）

S2-1　岡山大学病院ACHDセンターの試み
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

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～
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

S2-3　小児循環器科のない施設におけるACHD診療
椎名 由美，木島 康文，福田 旭伸，杉渕 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

S2-4　長野モデルの苦労話
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

S2-5　九州におけるACHD診療
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

1月11日㊎　第2会場［3F コンベンションホール東］

12：40～13：40　ランチョンセミナー2

座長：中村 一文（岡山大学大学院医歯薬学総合研究科 循環器内科学）
共催：グラクソ・スミスクライン株式会社

LS2　肺高血圧症治療薬の登場でEisenmenger症候群の治療は変わったか？
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

13：50～14：50　教育講演1

先天性心疾患、まずは解剖と病態を識る　その1

座長：**白石 公**（国立循環器病研究センター 教育推進部・小児循環器部）

EL1-1　Simple lesion ASD/VSD診断のpitfall

仁田 学

横浜市立大学医学部 循環器・腎臓・高血圧内科学教室

EL1-2　心エコー図を用いた成人先天性心疾患の解剖と病態の理解

松本 賢亮

神戸大学大学院医学研究科 内科学講座・循環器内科学分野

EL1-3　右室の異常 (TOF, PAVSD and PAIVS)

中川 直美

広島市立広島市民病院 循環器小児科

14：55～15：55　教育講演2

先天性心疾患、まずは解剖と病態を識る　その2

座長：**三浦 大**（東京都立小児総合医療センター）

EL2-1　大血管の異常 (TGA and ccTGA)

久持 邦和

広島市立広島市民病院 心臓血管外科

EL2-2　両大血管右室起始とは？

近藤 麻衣子

岡山大学病院 小児科／小児循環器科

EL2-3　左室流出路の異常 (CoA and HLHS)

櫻井 寛久

中京病院 心臓血管外科

16：00～17：00　教育講演3

ACHDの診療－Back To Basics

座長：**森 善樹**（北里大学メディカルセンター 小児科）

EL3-1　先天性心疾患の身体所見から学ぶ

水野 篤

聖路加国際病院 循環器内科

EL3-2　成人期チアノーゼ性心疾患の評価と管理

相馬 桂

東京大学医学部附属病院 循環器内科

EL3-3　心肺運動負荷試験の基本と検査結果の読み方

岡 岳文

一般財団法人津山慈風会津山中央病院 循環器内科

17：10～18：10　　**教育講演4**

CHDの手術と成人期での注意点　その1

座長：**市川 肇**（国立循環器病研究センター病院 小児心臓外科）

EL4-1　**房室中隔欠損症の手術と成人期での問題点**
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

EL4-2　**成人先天性心疾患領域の大血管転位**
上村 秀樹
奈良県立医科大学附属病院 先天性心疾患センター

EL4-3　CoA and Truncus
黒子 洋介
岡山大学大学院医歯薬学総合研究科 心臓血管外科

1月11日㊎　　第3会場 [3F 301会議室]

11：30～12：30　　**一般演題（口演）1**

カテーテルインターベンション

座長：**大月 審一**（岡山大学病院 小児循環器科）
佐地 真育（榊原記念病院 循環器内科）
コメンテーター：**中川 晃志**（岡山大学大学院医歯薬学総合研究科 循環器内科学）

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**
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

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**
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）岡山大学病院行 超音波センター

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**
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

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**
小木曽 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

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

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)]，吉眞 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

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

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)]，髙田 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

13：50～14：50　一般演題（口演）2

術後遠隔期の諸問題とチーム医療

座長：**新井 禎彦**（旭川荘療育・医療センター）
堀端 洋子（済生会熊本病院 循環器内科（心臓血管センター））
コメンテーター：**岡本 吉生**（香川県立中央病院 小児科）

OJ2-1　肺気腫を伴った右肺静脈-門脈短絡の一例

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

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

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

OJ2-3　マスタード術後遠隔期の体心室右室機能

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

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

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

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

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

OJ2-6　MRI strainによる無症候性術後ファロー四徴症の肺動脈弁置換至適タイミングの検討

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

15：20～16：00　ディベート1

Fontanでの抗凝固療法：必要 vs. 不要

座長：**安田 謙二**（島根大学医学部 小児科）

D1-1　**『Fontanでの抗凝固療法は必要』**

栄徳 隆裕

岡山大学大学院医歯薬学総合研究科 小児医科学

D1-2　**『Fontanでの抗凝固療法は不要』**

根岸 潤

国立循環器病研究センター病院 小児循環器科

16：00～17：00　一般演題（口演）3

Fontan

座長：**満下 紀恵**（静岡県立こども病院 循環器センター）
中野 俊秀（福岡市立こども病院 心臓血管外科）
コメンテーター：**戸田 孝子**（山梨大学医学部 小児科）

OJ3-1　**Fontan術後遠隔期の心腔内エコーViewFlexによる導管内観察**

松岡 良平，宗内 淳，相良 優佳，藤井 俊輔，川口 直樹，杉谷 雄一郎，渡邉 まみ江

地域医療機能推進機構JCHO九州病院 小児科

OJ3-2　**Fontan循環における門脈血流の減少**

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)]，渡邉 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

OJ3-3　**成人Fontan患者における下垂体機能評価**

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)]，鵜池 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

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**

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)]，黒嵜 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

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**

村岡 衛[1)]，永田 弾[1)]，坂本 一郎[2)]，江口 祥美[1)]，福岡 将治[1)]，鵜池 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

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**

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

13：50～14：50 Case Session 1

座長：**上村 秀樹**（奈良県立医科大学附属病院 先天性心疾患センター）
コメンテーター：Erwin Oechslin（Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre）

CS1-1 **『Challenges related to PLE』**
Alexander Van De Bruaene
Department of Cardiovascular Diseases, University Hospitals Leuven, Leuven, Belgium

CS1-2 **『Added value of exercise CMR, 3D segmentation and modeling in decision making』**
Alexander Van De Bruaene
Department of Cardiovascular Diseases, University Hospitals Leuven, Leuven, Belgium

CS1-3 **『LVAD implantation for heart failure in an adult with repaired CC-TGA』**
June Huh
Samsung Medical Center, Sungkyunkwan University School of Medicine

15：00～16：30 Oral Presentation 1 (English)

Long-term outcomes in ACHD

座長：**篠原 徳子**（東京女子医科大学病院 心臓病センター 循環器小児・成人先天性心疾患科）
杜　徳尚（岡山大学 循環器内科）
コメンテーター：**神谷 千津子**（国立循環器病研究センター 周産期・婦人科部）
Erwin Oechslin（Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre）

基調講演 **Unique long-term complications in ACHD: What should we know?**
Erwin Oechslin
Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre

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**
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

OE1-2 **成人先天性心疾患におけるEarly Vascular Agingの成因**
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

OE1-3 **肺循環とFontan術後遠隔期の心血管イベントの関連**
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

OE1-4　Ebstein病の中長期予後についての検討

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

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

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

OE1-6　フォンタン術後妊娠の胎盤病理

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

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

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

16：30～17：00　**皆で考えよう。この症例をどうする？1**

座長：**白井 丈晶**（加古川中央市民病院 循環器内科）
コメンテーター：**安田 謙二**（島根大学医学部 小児科）
久持 邦和（広島市立広島市民病院 心臓血管外科）
圓尾 文子（加古川中央市民病院 心臓血管外科）
仁田　学（横浜市立大学医学部 循環器・腎臓・高血圧内科学教室）

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』

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

1月11日㊎　[4F 404会議室]

10：00～12：00　**JSACHD理事会**

1月11日㊎　[4F 405会議室]

12：00～12：30　**JSACHD評議員会**

8：30～10：00　シンポジウム3

ACHDの肺高血圧治療の最前線

座長：中村 一文（岡山大学大学院医歯薬学総合研究科 循環器内科学）
八尾 厚史（東京大学 保健・健康推進本部）

S3-1　シャント閉鎖後PAHの治療戦略と注意点

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

S3-2　IPAH with small ASDのshunt閉鎖は禁忌でいいのか？

松原 広己
国立病院機構岡山医療センター 循環器内科

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

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

S3-4　たかがASD、されどASD、成人では

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

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？

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

S3-6　Eisenmenger症候群に対する疾患標的療法の現況

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 蕗子[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

10：05～11：05　スポンサードセミナー

座長：三好 亨（岡山大学病院 循環器内科）
共催：田辺三菱製薬株式会社

SS　糖尿病と血管内皮機能：SGLT2阻害薬の意義

東 幸仁
広島大学原爆放射線医科学研究所 ゲノム障害医学研究センター／広島大学病院 未来医療センター

11：10～12：40　日本循環器学会ジョイントセッション

先天性心疾患の成人への移行医療に関する提言を具体化する

座長：**三谷 義英**（三重大学大学院医学系研究科 小児科学）
赤阪 隆史（和歌山県立医科大学 循環器内科）

JS1-1　関連８学会合同提言の背景と日本循環器学会の取り組み
三谷 義英
三重大学大学院医学系研究科 小児科学

JS1-2　生涯医療の中の移行医療連携システム構築－長野モデルからの提案
安河内 聰
長野県立こども病院 循環器センター

JS1-3　移行の受け手側の役割と日本の現状、そして学校保健
八尾 厚史[1)]，落合 亮太[2)]，村田 俊樹[3)]，榎本 淳子[4)]，赤木 禎治[5)]，白石 公[6)]，丹羽 公一郎[7)]
1) 東京大学 保健・健康推進本部，2) 横浜市立大学学術院医学群医学研究科 看護学専攻 がん・先端成人看護学，
3) 東京都心臓病の子どもを守る会／東京心友会，4) 東洋大学 文学部 教育学科，5) 岡山大学 循環器内科，
6) 国立循環器病研究センター 小児循環器科，7) 聖路加国際病院 心臓血管センター

JS1-4　先天性心疾患患者の移行期の現状と課題 ～患者・家族の立場から～
青木 美千代
一般社団法人 全国心臓病の子どもを守る会 理事

JS1-5　厚生労働省の取り組み
谷口 顕信
厚生労働省健康局難病対策課

12：50～13：50　ランチョンセミナー３

座長：**伊藤 浩**（岡山大学大学院医歯薬学総合研究科 循環器内科学）
共催：**第一三共株式会社**

LS3　成人先天性心疾患における抗凝固療法の重要性と意義
赤木 禎治
岡山大学病院 循環器内科

13：55～14：25　会長講演

座長：**丹羽 公一郎**（聖路加国際病院 心血管センター）

PL　『成人先天性心疾患!?：循環器内科医の覚悟』
伊藤 浩
岡山大学大学院医歯薬学総合研究科 循環器内科学

14：35～15：15　海外招聘講演1

座長：**森田 紀代造**（東京慈恵会医科大学 心臓外科学講座）

II1　**『Evaluation and management of cyanotic ACHD: Pitfalls and tricks!』**

Erwin Oechslin

Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre

15：25～16：25　コーヒーブレイクセミナー2

座長：**伊藤 浩**（岡山大学大学院医歯薬学総合研究科 循環器内科学）

共催：**日本ベーリンガーインゲルハイム株式会社**

CBS2　**循環器抗血栓療法のトレンド －心房細動合併虚血性心疾患の治療戦略－**

上村 史朗

川崎医科大学 循環器内科学

16：30～18：00　学術委員会シンポジウム

実臨床のcontroversial

座長：**稲井　慶**（東京女子医科大学 心臓病センター 循環器小児・成人先天性心疾患科）

大内 秀雄（国立循環器病研究センター 小児循環器科 成人先天性心疾患科）

ES-1　**高尿酸血症への対応は必要か**

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

ES-2　**ACHD患者に対する心臓リハビリテーション**

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

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**

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

ES-4　**妊娠出産　ハイリスク症例への対応**

神谷 千津子

国立循環器病研究センター 周産期・婦人科

8：30～9：30　教育講演5

CHDの手術と成人期での注意点 その2

座長：**松尾 浩三**（千葉県循環器病センター 心臓血管外科）

EL5-1　**成人期におけるTOFに対する外科治療：適応と戦略**
小谷 恭弘
岡山大学大学院医歯薬学総合研究科 心臓血管外科

EL5-2　Evolution of surgical repair and current outcome in Ebstein anomaly
本淨 修己
The Hospital for Sick Children

EL5-3　**懐かしのCHD手術－今では稀だが、知っておくべき術式**
猪飼 秋夫
静岡県立こども病院 心臓血管外科

9：35～10：35　教育講演6

Fontan循環を理解する

座長：**安河内 聰**（長野県立こども病院 循環器センター）

EL6-1　Fontan operation: Indication, techniques, and outcome
本淨 修己
The Hospital for Sick Children

EL6-2　**成人期に到達したFontan患者の心臓評価**
大内 秀雄
国立循環器病研究センター 小児循環器科 成人先天性心疾患科

EL6-3　**Fontan関連肝疾患（FALD）とは？**
大西 秀樹
岡山大学病院 消化器内科

10：40～11：40　教育講演7

ACHDのイメージング

座長：**城戸 佐知子**（兵庫県立こども病院 循環器内科）

EL7-1　**成人期に心エコーで初めて診断されるCHD－見逃していませんか？**
杜 徳尚
岡山大学 循環器内科

EL7-2　**心臓MRIの基本とACHDへの応用**
椎名 由美
聖路加国際病院 心血管センター

EL7-3　ACHDのイメージングに基づく手術戦略

板谷 慶一

京都府立医科大学大学院医学研究科 心臓血管外科学

11：40～12：40　教育講演8

心臓カテーテル検査

座長：**鎌田 政博**（広島市立広島市民病院 循環器小児科）

EL8-1　先天性心疾患でのカテーテル検査と血行動態の読み方

加藤 温子

中京病院 小児循環器科

EL8-2　成人期の複雑先天性心疾患に対するカテーテル治療

馬場 健児

岡山大学病院 IVRセンター 小児循環器部

EL8-3　SHDに対するカテーテルインターベンション－日本で出来ること

佐地 真育

榊原記念病院 循環器内科

12：50～13：50　ランチョンセミナー4

座長：**赤阪 隆史**（和歌山県立医科大学附属病院 循環器内科）

共催：ブリストル・マイヤーズ スクイブ株式会社／ファイザー株式会社

LS4　血栓症とDOAC

小板橋 紀通

群馬大学医学部附属病院 循環器内科

13：55～14：55　教育講演9

冠動脈疾患と結合織異常

座長：**市田 蕗子**（富山大学医学部 小児科教室）

EL9-1　冠動脈起始異常とその治療適応

山村 健一郎

九州大学医学部 小児科／トロント総合病院

EL9-2　川崎病の診断、治療と長期予後

小林 徹

国立成育医療研究センター 臨床研究センター 企画運営部

EL9-3　Marfan症候群と類縁疾患の診断と管理

犬塚 亮

東京大学 小児科

14：55～15：55　教育講演10

妊娠

座長：**桂木 真司**（榊原記念病院 産婦人科）

EL10-1　ACHDでの妊娠前評価から妊娠・産後管理まで

神谷 千津子

国立循環器病研究センター 周産期・婦人科部

EL10-2　ACHDでの分娩様式と麻酔選択

照井 克生

埼玉医科大学総合医療センター 産科麻酔科

EL10-3　ACHD女性と避妊

兵藤 博信

東京都立墨東病院 産婦人科

15：55～16：55　教育講演11

不整脈

座長：**簇 義仁**（昭和大学病院 小児循環器・成人先天性心疾患センター）

EL11-1　ACHDでの上室性不整脈の特徴と対策

宮﨑 文

天理よろづ相談所病院 小児循環器科・先天性心疾患センター

EL11-2　ACHDにおける心室性不整脈－その状態と治療

芳本 潤

静岡県立こども病院 循環器センター

EL11-3　成人先天性心疾患患者におけるデバイス治療

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

17：00～18：00　第1回ACHDウルトラクイズ

8：30～9：30　シンポジウム4

ACHD患者の妊娠・出産そして授乳の問題を考える

座長：池田 智明（三重大学大学院医学系研究科 産科婦人科学）
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）

S4-1　ACHD患者の妊娠前・妊娠管理
田中 博明
三重大学医学部 産科婦人科学

S4-2　ACHD合併妊娠と児の合併症
堀内 縁
国立循環器病研究センター 周産期・婦人科

S4-3　ACHD患者の分娩管理
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

S4-4　ACHDの妊娠が自然予後に及ぼす影響
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

9：35～10：35　一般演題（口演）4

妊娠・出産・診療支援

座長：富松 宏文（東京女子医科大学病院 心臓病センター 循環器小児科）
小口 秀紀（トヨタ記念病院 周産期母子医療センター 産科）
コメンテーター：堀内　縁（国立循環器病研究センター 周産期・婦人科部）

OJ4-1　肥大型心筋症と妊娠・出産
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

OJ4-2　成人先天性心疾患女性の妊娠・分娩期の産科合併症
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

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
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

OJ4-4　成人先天性心疾患チームの立ち上げ6か月の現状

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

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

廣瀬 圭一[1)]，猪飼 秋夫[1)]，長門 久雄[1)]，村田 眞哉[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

OJ4-6　当院におけるACHD患者移行における現状と諸問題

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)]，大薗 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 心臓血管外科学，4) 大阪急性期・総合医療センター 小児科

10：40～11：40　一般演題（口演）5

予後

座長：**櫻井　一**（中京病院 心臓血管外科）
黒子 洋介（岡山大学大学院医歯薬学総合研究科 心臓血管外科）
コメンテーター：**永田　弾**（九州大学 小児科）

OJ5-1　ファ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

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

OJ5-2　Transcatheter PVR時代を前にしたSurgical PVR

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，簇 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

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

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，黒嵜 健一
国立循環器病研究センター 小児循環器科

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

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

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

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

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

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

11：40～12：40　シンポジウム5

ACHDの不整脈治療

座長：庄田 守男（東京女子医科大学 循環器内科）
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）

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**

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

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**

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

S5-3　**成人先天性心疾患患者におけるICD治療**

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

S5-4　**成人先天性心疾患におけるリードマネジメント**

庄田 守男
東京女子医科大学 循環器内科

12：50～13：50　ランチョンセミナー5

座長：小垣 滋豊（大阪急性期・総合医療センター 小児科新生児科）
共催：日本新薬株式会社／アクテリオン ファーマシューティカルズ ジャパン株式会社

LS5　**小児循環器医が考える成人先天性心疾患診療のポイント**
～移行期医療と肺循環に注目した循環動態の理解を中心に～

土井 拓
天理よろづ相談所病院 小児科／先天性心疾患センター

13：55～14：35　ディベート2

TOF術後PRに対する成人期PVR：人工弁の種類・サイズの選択

座長：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）

D2-1　**『肺動脈弁置換の最近の流れについて』**

小谷 恭弘
岡山大学大学院医歯薬学総合研究科 心臓血管外科

D2-2　**『血流評価を用いた人工弁の種類やサイズなどについて』**

板谷 慶一
京都府立医科大学大学院医学研究科 心臓血管外科学

14：45～15：35　一般演題（口演）6

心不全

座長：田邊 一明（島根大学医学部 内科学講座（内科学第四））
福本 義弘（久留米大学医学部 内科学講座 心臓・血管内科部門）
コメンテーター：稲葉 俊郎（東京大学医学部附属病院 循環器内科）

OJ6-1　TCPC術後の非薬物的不整脈治療

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

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

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

OJ6-3　成人先天性心疾患におけるサルコペニア

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

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

藤田 鉄平[1)]，小板橋 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

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

安河内 聰[1)]，武井 黄太[1)]，瀧聞 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

15：55～16：55　皆で考えよう。この症例をどうする？2

座長：小板橋 俊美（北里大学医学部 循環器内科学）
福田　旭伸（聖路加国際病院 心血管センター）
コメンテーター：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）
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）
森田 紀代造（東京慈恵会医科大学 心臓外科学講座）
神谷 千津子（国立循環器病研究センター 周産期・婦人科部）

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』

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

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？』

藤田 鉄平[1)]，小板橋 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

1月12日 (土)　第4会場 [3F 302会議室]

8：55～18：00	The 1st Asia Pacific Society for ACHD Symposium (English)

1月12日 (土)　[4F 404会議室]

7：30～8：10	JSACHD学術委員会

8：30～9：30　シンポジウム6

カテーテルインターベンションの最新治療

座長：坂本 一郎（九州大学大学院医学研究院 循環器内科学）
赤木 禎治（岡山大学病院 循環器内科）

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

大山 伸雄[1,2]，Israel Valverde[2]，Kuberan Pushparajah[2]，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, GSTT NHS Foundation Trust, London, UK

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

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]，黒嵜 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

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

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

S6-4　卵円孔開存に対するカテーテル治療

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

9：35～11：05　日本心エコー図学会共同企画

ACHDの病態の定量評価に挑む

座長：石津 智子（筑波大学 臨床検査医学）
杜　徳尚（岡山大学 循環器内科）

JS2-1　先天性心疾患における房室弁逆流の定量評価

新居 正基
静岡県立こども病院 循環器科

JS2-2　狭窄病変の定量における問題点と意義。

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

JS2-3　右室同期不全の定量化

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

JS2-4　周術期不整脈治療における病態評価 ～multimodalityの活用～

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

JS2-5　体心室右室での定量化への挑戦とその有用性

杜 徳尚，赤木 禎治，伊藤 浩
岡山大学 循環器内科

11：10～11：40　海外招聘講演2

座長：**筒井 裕之**（九州大学大学院医学研究院 循環器内科学）

II2　『Current Arrhythmia Management for ACHD Patients』

Mei-Hwan Wu
National Taiwan University Children's Hospital

11：45～12：30　成人先天性心疾患専門医制度解説セッション

座長：**丹羽 公一郎**（聖路加国際病院 心血管センター）

GS-1　『専門医制度（内科系）』

市田 蕗子
富山大学医学部 小児科教室

GS-2　『専門医制度（外科系）』

森田 紀代造
東京慈恵会医科大学 心臓外科学講座

GS-3　『専門医修練施設』

八尾 厚史
東京大学 保健・健康推進本部

12：40～13：40　ランチョンセミナー6

座長：**丹羽 公一郎**（聖路加国際病院 心血管センター）
共催：アクテリオン ファーマシューティカルズ ジャパン株式会社／日本新薬株式会社

LS6　循環器内科医による最新のACHD-PAH治療戦略

八尾 厚史
東京大学 保健・健康推進本部

13：45～14：15　JSACHD総会

14：20～15：20　シンポジウム8

ACHDの緩和医療

座長：**丹羽 公一郎**（聖路加国際病院 心血管センター）
福嶌　教偉（国立循環器病研究センター病院 移植医療部）

S8-1　**循環器疾患における緩和ケア**
水野 篤，福田 旭伸，丹羽 公一郎
聖路加国際病院 循環器内科

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**
中澤 誠
総合南東北病院 小児・生涯心臓疾患研究所

S8-3　**緩和医療と心肺移植**
福嶌 教偉
国立循環器病研究センター 移植医療部

S8-4　**緩和ケアにおける看護師の役割**
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

15：20～15：30　閉会の辞

会長：**伊藤 浩**（岡山大学大学院医歯薬学総合研究科 循環器内科学）

16：00～17：00　市民公開講座

大人になった先天性心疾患患者さんへ 困ったこと・不安なことがありますか？

座長：**伊藤　浩**（岡山大学大学院医歯薬学総合研究科 循環器内科学）
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）

OC-1　**『成人先天性心疾患の内科診療』**
赤木 禎治
岡山大学病院 循環器内科

OC-2　**『先天性心疾患手術で目指すもの：外科医からみた現状と今後』**
小谷 恭弘
岡山大学病院 心臓血管外科

OC-3　**『大人になった先天性心疾患患者への心理学的アプローチ』**
榎本 淳子
東洋大学文学部 教育学科

OC-4　**『患者様から』**

8：30～9：50　一般演題（口演）7

外科

座長：**宮地　鑑**（北里大学病院 心臓血管外科）
大嶋 義博（兵庫県立こども病院 心臓血管外科）
コメンテーター：**帯刀 英樹**（九州大学病院 心臓血管外科）

OJ7-1　ファロー四徴症再手術時の右室流出路再建法

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

OJ7-2　成人期Ebstein病に対する外科的治療経験

宮本 隆司[1)]，藤田 鉄平[2)]，小板橋 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)]，桒田 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

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

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)]，中嶌 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

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

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 修一朗[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

OJ7-5　成人先天性心疾患患者に対するBentall手術

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

OJ7-6　Lateral Tunnel TCPCに対するExtracardiac TCPC conversionの検討

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

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

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

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

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

9：55～10：55　教育講演12

肺高血圧と心不全－How to tackle critical situations

座長：**坂崎 尚徳**（兵庫県立尼崎総合医療センター 小児循環器内科）

EL12-1　ACHDに伴う肺高血圧の特徴とその治療

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

EL12-2　Eisenmenger症候群の診断と治療

建部 俊介

東北大学病院 循環器内科

EL12-3　心不全治療における薬物療法の功罪

坂本 一郎

九州大学病院 循環器内科

11：00～12：00　教育講演13

実際の診療体制と問題点

座長：**賀藤 均**（国立成育医療研究センター）

EL13-1　望まれる診療体制と本邦の現状 －横浜市立大学を例に－

落合 亮太

横浜市立大学大学院医学研究科 看護学専攻

EL13-2　診療中断をなくすために～移行医療のコツ

元木 博彦

信州大学医学部 循環器内科学教室

EL13-3　成人先天性心疾患診療において必要な社会保障制度

檜垣 高史

愛媛大学大学院医学系研究科 地域小児・周産期学講座

12：05～12：30　教育講演14

知っておきたいコメディカルの知識

座長：**落合 亮太**（横浜市立大学大学院医学研究科 看護学専攻）

EL14-1　緊急時に備えたカテ室看護師の役割

祇園 由美

岡山大学病院 放射線部

EL14-2　ACHD診療に重要な心理の問題

榎本 淳子

東洋大学文学部 教育学科

12：40～13：40　**ランチョンセミナー7**

成人期心房中隔欠損に対するカテーテル治療：治療適応と長期予後

座長：**檜垣 高史**（愛媛大学大学院医学系研究科 地域小児・周産期学講座）
共催：日本ライフライン株式会社

LS7-1　**成人期ASDの臨床像と長期予後**
高谷 陽一
岡山大学病院 循環器内科

LS7-2　**Occlutech Figulla Flex IIによるASD閉鎖のメリットは？**
七里 守
名古屋第二赤十字病院 第二循環器内科

14：20～15：20　**教育講演15**

細部まで見落とさない－Common oversight

座長：**脇 研自**（倉敷中央病院 小児科）

EL15-1　**染色体異常を伴ったACHDの診療で注意するべき点**
山岸 敬幸
慶應義塾大学医学部 小児科

EL15-2　**成人ASDの特徴と問題点**
高谷 陽一
岡山大学大学院医歯薬学総合研究科 循環器内科学

EL15-3　**成人先天性心疾患患者の口腔を知る ～感染性心内膜炎リスクを下げるためにできること～**
大森 一弘
岡山大学病院 歯周科

1月13日㊐　第3会場［3F 301会議室］

8：30～10：00　**看護ワーキング 情報交換会**

10：05～10：45　**ディベート3**

複雑ACHDでの妊娠：ここまで可能 vs. ここから不可能

座長：**市川 肇**（国立循環器病研究センター 小児心臓外科）

D3-1　**『複雑ACHDでの妊娠：ここまで可能』**
吉松 淳
国立循環器病研究センター 周産期・婦人科部

D3-2　『複雑ACHDでの妊娠：ここから不可能』

兵藤 博信
東京都立墨東病院 産婦人科

10：50～12：20　シンポジウム7

小児心臓血管外科手術の長期成績

座長：**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）
山岸 正明（京都府立医科大学大学院医学研究科 心臓血管・小児心臓血管外科学）

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

S7-2　完全大血管転位術後の長期成績と問題点

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

S7-3　総肺静脈還流異常に対する外科治療の長期成績

松久 弘典[1)]，大嶋 義博[1)]，日隈 智憲[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

S7-4　肺動脈閉鎖兼正常心室中隔のFontan成績

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

14：20～15：20　一般演題（口演）8

患者ケア

座長：**水野 芳子**（東京情報大学 看護学科）
木島 康文（聖路加国際病院 循環器内科）
コメンテーター：**森　善樹**（北里大学メディカルセンター 小児科）

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

猪又 竜
先天性心疾患患者

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

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

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）

福間 睦子，桂木 真司
榊原記念病院

OJ8-4　Fontan術後成人患者における筋力測定の検討

近野 宏知[1)]，久松 智子[1)]，加藤 秀典[1)]，高橋 雅文[1)]，朴 要俊[2)]，呉 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

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

梶濱 あや，島田 空知，中右 弘一
旭川医科大学 小児科

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

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 Methatham[2]，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学部門，
5) 自治医科大学成人先天性心疾患センター

1月13日㊐　第4会場［3F 302会議室］

8：30～10：10　Oral Presentation 2 (English)

Surgical and interventional strategy for ACHD

座長：**小谷 恭弘**（岡山大学 心臓血管外科）
米田 正始（医誠会病院 心臓血管外科）

コメンテーター：**平田 康隆**（東京大学医学部附属病院 心臓外科）
Worakan Promphan（Queen Sirikit National Institute of Child Health, Bangkok, Thailand）

基調講演　Transcatheter management RVOT in adult ToF

Worakan Promphan
Queen Sirikit National Institute of Child Health, Bangkok, Thailand

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

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

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

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

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

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

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

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

OE2-5　二尖大動脈弁の再建手術とMICSの可能性

米田 正始，藤原 祥司，氏家 敏巳
医誠会病院 心臓血管外科

OE2-6　成人先天性心疾患手術におけるMICSの役割

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

OE2-7　左室緻密化障害（LVNC）への外科治療

米田 正始，藤原 祥司，氏家 敏巳
医誠会病院 心臓血管外科

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

河田 政明[1)]，吉積 功[1)]，鵜垣 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

10：10～10：50　Case Session 2

座長：**赤木 禎治**（岡山大学病院 循環器内科）
コメンテーター：Seung Woo Park（Department of Cardiology Samsung Medical Center）

CS2-1　『Altered biventricular hemodynamic forces in a man with repaired Tetralogy of Fallot』

Lucy Eun
Division of Pediatric Cardiology, Department of Pediatrics, Yonsei University College of Medicine ACHD clinic, Severance Cardiovascular Hospital, Yonsei University Health System, Seoul, Korea

CS2-2　『Fontan型手術後のハイリスク妊娠症例 ～妊娠継続を切望した流産症例を振り返る～』

牧 尉太
岡山大学大学院医歯薬学総合研究科 産科・婦人科学教室

ポスターセッションプログラム

1月11日 ㊎　　ポスター会場［2F 展示ホール］

17：00～17：48　Poster Presentation 1 (English)

座長：**加藤 温子**（中京病院 小児循環器科）

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

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

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

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

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

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

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

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

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

宮﨑 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

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

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

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

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

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

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

17：00～17：54　Poster Presentation 2 (English)

座長：**帆足 孝也**（国立循環器病研究センター病院 小児心臓外科）

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

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

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

康 秀貞
CHA醫科學大學校 小兒靑少年科學敎室

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

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

PE2-4　甲状腺機能亢進がフォンタン循環へ及ぼす影響

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

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

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

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

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

PE2-7　著明な収縮期雑音により、心疾患を疑われた一例

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

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

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)]，栁川 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

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

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

17：00～17：54　Poster Presentation 3 (English)

座長：**石戸 美妃子**（東京女子医科大学病院 心臓病センター 循環器小児科）

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

郡山 恵子[1)]，小板橋 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

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

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

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－

小永井 奈緒，大内 秀雄，根岸 潤，坂口 平馬，岩朝 徹，白石 公，黒嵜 健一

国立循環器病研究センター 小児循環器科

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

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

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

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

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

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

PE3-7　18年間通院中断していたフォンタン術後の一例

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

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

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

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－

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) 東京逓信病院 小児科

16：00～16：48 一般演題（ポスター）1

座長：**牧 尉太**（岡山大学大学院医歯薬学総合研究科 産科・婦人科学教室）

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

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

PJ1-2 拡張相早期の肥大型心筋症合併妊娠の一例

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

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

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 尉太[2)]，増山 寿[2)]
1）広島市民病院，2）岡山大学病院 産婦人科

PJ1-4 心内膜床欠損症修復術後妊娠の一例

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

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

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

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

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

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

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 蕗子[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

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

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 眞知子[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

16：00～16：48　一般演題（ポスター）2

座長：**鈴木 嗣敏**（大阪市立総合医療センター 小児不整脈科）

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

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

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

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，樗木 晶子，筒井 裕之
九州大学病院 循環器内科

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

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

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

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)]，黒嵜 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

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

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

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

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)]，髙田 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

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

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

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

田尻 雄二朗[1)]，宮﨑 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

16：00～16：42　一般演題（ポスター）3

座長：**久持 邦和**（広島市民病院 心臓血管外科）

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

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

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

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

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

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

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

池内 博紀[1)]，椛沢 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

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

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

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

池内 博紀[1)]，椛沢 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

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

椛沢 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

16：00～16：54　一般演題（ポスター）4

座長：**星合 美奈子**（山梨県立中央病院 小児循環器病センター）

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

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

PJ4-2　成人期に手術介入した三心房心の2例

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

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

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)]，渡邉 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

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～

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

PJ4-5　Fontan術後遠隔期に鋳型気管支炎を発症した一例

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

PJ4-6　PAVSD術後遠隔期にVT/VFを合併した１例

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

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

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)]，槙田 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

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

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

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

齋藤 義弘[1)]，相馬 桂[1)]，齊藤 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

11：25～12：19 一般演題 (ポスター) 5

座長：永田 弾（九州大学病院 小児科）

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

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

PJ5-2 心房中隔欠損症のデバイス閉鎖における医療連携

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 穣，新田 順一
さいたま赤十字病院 循環器内科

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

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

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

中島 公子，大内 秀雄，北野 正尚，根岸 潤，黒嵜 健一
国立循環器病研究センター

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

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

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

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

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

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

PJ5-8 Scimitar症候群亜型に対する低侵襲外科治療の経験

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

PJ5-9 大動脈拡張を伴った大動脈二尖弁症例の妊娠例

小板橋 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

11：25～12：13　一般演題（ポスター）6

座長：**石森 直樹**（北海道大学病院 循環器内科）

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

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

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

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 穣，新田 順一
さいたま赤十字病院 循環器内科

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

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

PJ6-4　エピネフリン誘発性QT延長症候群合併妊娠の1例

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

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

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

PJ6-6　CRTが著効した修正大血管転位の1例

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

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

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

PJ6-8　Ross後のASR・LVOTOに対するApico-aortic bypass

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

11：25～12：19　一般演題（ポスター）7

座長：**豊野 学朋**（秋田大学 小児科学講座）

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

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

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

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

PJ7-3　Takeuchi法術後長期遠隔期を迎えたBWG症候群の１例

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

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

前川 恵美，小板橋 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

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

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

PJ7-6　卵円孔開存にdevice closureを施行したPlatpnea-Orthodexia syndromeの１例

金子 幸栄[1)]，中嶌 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

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

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

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

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

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

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

11：25～12：07　一般演題（ポスター）8

座長：**中川 直美**（広島市立広島市民病院 循環器小児科）

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

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

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

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

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

前村 健治[1)]，小板橋 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 心臓血管外科学

PJ8-4　Fontan術後の月経異常にどう対処するか－1例報告

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

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

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

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

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

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

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

Program (English Session)

January 11 (Friday) Room 4 [3F Conference room 302]

13:50-14:50 Case Session 1 (English)

Chair: **Hideki Uemura** (Congenital Heart Disease Center, Nara Medical University)
Commentator: **Erwin Oechslin** (Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre)

CS1-1 Challenges related to PLE
Alexander Van De Bruaene
Department of Cardiovascular Diseases, University Hospitals Leuven, Leuven, Belgium

CS1-2 Added value of exercise CMR, 3D segmentation and modeling in decision making
Alexander Van De Bruaene
Department of Cardiovascular Diseases, University Hospitals Leuven, Leuven, Belgium

CS1-3 LVAD implantation for heart failure in an adult with repaired CC-TGA
June Huh
Samsung Medical Center, Sungkyunkwan University School of Medicine

15:00-16:30 Oral Presentation 1 (English)

Long-term outcomes in ACHD

Chairs: **Tokuko Shinohara** (Department of Pediatric Cardiology and Adult Congenital Cardiology, Tokyo Women's Medical University)
Norihisa Toh (Department of Cardiovascular Medicine, Okayama University)
Commentators: **Chizuko Kamiya** (National Cerebral and Cardiovascular Center Department of Perinatology and Gynecology)
Erwin Oechslin (Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre)

Keynote Lecture Unique long-term complications in ACHD: What should we know?
Erwin Oechslin
Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre

OE1-1 Combination Therapy with Warfarin and Aspirin is Effective for Thromboembolic Prophylaxis in Patients after the Fontan Operation
Shintaro Umemoto[1)], Ichiro Sakamoto[1)], Kisyo Ohtani[1)], Ayako Ishikita[1)], Yoshihiki Kodama[2,3)], Hazumu Nagata[3)], Tomomi Ide[1)], Shiro Ishikawa[2)], Shoichi Ohga[2)], Hiroyuki Tsutsui[1)]
1) Department of Cardiovascular Medicine, Kyushu University Hospital,
2) Department of Cardiology, Fukuoka Children's Hospital,
3) Department of Pediatrics, Kyushu University Hospital

OE1-2 Early vascular aging in adult patients with congenital heart disease

Tomoaki Murakami[1)], Yoko Horibata[2)], Shigeru Tateno[2)], Yasutaka Kawasoe[2)], Koichiro Niwa[3)]

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

OE1-3 Pulmonary circulation related to late cardiovascular events after Fontan operation

Jun Muneuchi[1)], Mamie Watanabe[1)], Yuichiro Sugitani[1)], Naoki Kawaguchi[1)], Ryohei Matsuoka[1)], Shunsuke Fujii[1)], Yusuke Ando[2)], Yoshie Ochiai[2)]

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

OE1-4 Mid-term clinical outcomes in adult patients with surgically operated Ebstein anomaly

Norihisa Toh[1)], Teiji Akagi[1)], Yasuhiro Kotani[2)], Fumi Yokohama[1)], Yosuke Kuroko[2)], Kenji Baba[3)], Shin-ichi Otsuki[3)], Shingo Kasahara[2)], Hiroshi Ito[1)]

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

OE1-5 Unique features of hepatic disease in adults with Fontan circulation: A comparison with congenital heart disease patients after two-ventricular repair

Norihia Toh[1)], Hideki Onishi[2)], Teiji Akagi[1)], Yasuto Takeuchi[2)], Shinichiro Nakamura[2)], Fumi Yokohama[1)], Yoichi Takaya[1)], Yasuhiro Kotani[3)], Yosuke Kuroko[3)], Shingo Kasahara[3)], Hiroyuki Okada[2)], Hiroshi Ito[1)]

1) Department of Cardiovascular Medicine, Okayama University,
2) Department of Gastroenterology and Hepatology, Okayama University,
3) Department of Cardiovascular Surgery, Okayama University

OE1-6 The placental pathology of women with functional single ventricle after Fontan palliation

Tae Konishi[1)], Keiko Ohta Ogo[2)], Hatsue Ishibashi Ueda[2)], Chizuko A Kamiya[1)], Masami Sawada[1)], Tadasu Shionoiri[1)], Atsushi Nakanish[1)], Chinami Horiuchi[1)], Mitsuhiro Tsuritani[1)], Naoko Iwanaga[1)], Jun Yoshimatsu[1)]

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

OE1-7 Relations between breastfeeding and postpartum changes in brain natriuretic peptide among mothers with congenital heart disease

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

January 12 (Saturday) Room 1 [3F Convention Hall West]

12:50-13:50 Luncheon Seminar 3 (Japanese and English)

Chair: **Hiroshi Ito** (Department of Cardiovascular Medicine, Okayama University Graduate School of Medicine)
Cosponsor: **Daiichi Sankyo**

LS3 **Clinical implication of anticoagulation management in adult patients with congenital heart disease**

Teiji Akagi

Department of Cardiovascular Medicine, Okayama University Hospital

14:35-15:15 International Invited Lecture 1 (English)

Chair: **Kiyozo Morita** (Jikei University School of Medicine, Cardiovascular Surgery)

II1 **Evaluation and management of cyanotic ACHD: Pitfalls and tricks!**

Erwin Oechslin

Director, Adult Congenital Heart Disease Program The Bitove Family Professor of Adult Congenital Heart Disease Peter Munk Cardiac Centre

January 12 (Saturday) Room 4 [3F Conference room 302]

8:55-18:00 The 1st Asia Pacific Society for ACHD Symposium (English)

January 13 (Sunday) Room 1 [3F Convention Hall West]

11:10-11:40 International Invited Lecture 2 (English)

Chair: **Hiroyuki Tsutsui** (Department of Cardiovascular Medicine, Faculty of Medical Sciences, Kyushu University)

II2 **Current Arrhythmia Management for ACHD Patients**

Mei-Hwan Wu

National Taiwan University Children's Hospital

8:30-10:10 Oral Presentation 2 (English)

Surgical and interventional strategy for ACHD

Chairs: **Yasuhiro Kotani** (Department of Cardiovascular Surgery, Okayama University)
Masashi Komeda (Department of Cardiovascular Surgery, Iseikai Hospital)
Commentators: **Yasutaka Hirata** (Department of Cardiac Surgery, The University of Tokyo Hospital)
Worakan Promphan (Queen Sirikit National Institute of Child Health, Bangkok, Thailand)

Keynote Lecture Transcatheter management RVOT in adult ToF

Worakan Promphan
Queen Sirikit National Institute of Child Health, Bangkok, Thailand

OE2-1 Surgical Indication for Pulmonary Valve Disease in Adult Congenital Heart Disease Based on Right Ventricular Hemodynamics Assessed with 4D flow MRI

Keiichi Itatani[1)], Masaaki Yamagishi[2)], Yoshinobu Maeda[2)], Shuhei Fujita[2)], Hisayuki Hongu[2)], Yuji Takayanagi[2)], Haruka Fu[2)], Hiroko Morichi[1)], Shohei Miyazaki[3)], Yo Kajiyama[4)], Masao Takigami[5)], Naohiko Nakanishi[5)], Satoaki Matoba[5)], Hitoshi Yaku[1)]
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

OE2-2 Native T1 and Extracellular Volume at Cardiac Magnetic Resonance in Adults with Congenital Heart Disease

Yumi Shiina[1)], Kota Taniguchi[3)], Michinobu Nagao[4)], Tatsunori Takahashi[5)], Masateru kawakubo[6)], Kei Inai[2)]
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

OE2-3 Applications of three-dimensional printed modeling of adult congenital heart disease

Toru Miyoshi[1)], Norihisa Toh[1)], Teiji Akagi[1)], Takashi Miki[1)], Yasushi Koyama[2)], Hiroshi Ito[1)]
1) Department of Cardiovascular Medicine, Okayama University,
2) Department of Radiology, Sakurabashi Watanabe Hospital

OE2-4 Comparison between surgery and catheter intervention for management of coarctation of the aorta in adults.

Atsuko Kato[1)], Jun Sato[1)], Kimihiro Yoshii[1)], Yoshihito Morimoto[1)], Shuichiro Yoshida[1)], Hiroshi Nishikawa[1)], Naoki Ohashi[2)], Takahisa Sakurai[2)], Toshimichi Nonaka[2)], Hajime Sakurai[2)]
1) Department of Pediatric Cardiology, JCHO Chukyo Hospital,
2) Department of Cardiovascular Surgery, JCHO Chukyo Hospital

OE2-5 **Repair of Reconstruction of Bicuspid Aortic Valve - including consideration of minimally invasive approaches**

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

OE2-6 **Minimally Invasive Cardiac Surgery for Simple Congenital Heart Surgery in the Adults**

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

OE2-7 **Surgical treatment of left ventricular non-compaction (LVNC)**

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

OE2-8 **Surgical experience of Adults with Polysplenia syndrome: Clinical relevance with lung biopsy findings**

Masaaki Kawada[1)], Ko Yoshizumi[1)], Shin-ya Ugaki[1)], Ko-ichi Kataoka[2)], Mitsuru Seki[2)], Kenske Oka[2)], Daisuke Matsubara[2)], Yasushi Imai[3)], Tomoyuki Kabutoya[3)], Kana Kubota[3)]
1) Pediatric and Congenital Cardiovascular Surgery, Jichi Adult Congenital Heart Center,
2) Pediatric Cardiology, 3) Cardiovascular Medicine

10:10-10:50 Case Session 2 (English)

Chair: **Teiji Akagi** (Department of Cardiovascular Medicine, Okayama University Hospital)
Commentator: **Seung Woo Park** (Department of Cardiology Samsung Medical Center)

CS2-1 **Altered biventricular hemodynamic forces in a man with repaired Tetralogy of Fallot**

Lucy Eun
Division of Pediatric Cardiology, Department of Pediatrics, Yonsei University College of Medicine ACHD clinic, Severance Cardiovascular Hospital, Yonsei University Health System, Seoul, Korea

CS2-2 **A high-risk case of Pregnancy After Fontan surgery. "Reviewing to a miscarriage case at 14 weeks in spite of wishing to continue pregnancy."**

Jota Maki
Department of Obstetrics and Gynecology Okayama University Graduate School of Medicine

Poster Session Program (English Session)

January 11 (Friday) Poster Room [2F Exhibition Hall]

17:00-17:48 Poster Presentation 1 (English)

Chair: **Atsuko Kato** (Department of Pediatric Cardiology, Chukyo Children's Heart Center, JCHO Chukyo Hospital)

PE1-1 **The anti-coagulant management of the pregnant complicating the mechanical valve and anti-phospholipid antibody syndrome**

Naomi Nakazawa[1)], Tomoko Ishizu[1)], Yasutoshi Boku[1)], Daisuke Hidaka[2)], Hidekazu Nishikii[3)], Mana Obata[4)], Chiho Tokunaga[5)], Yuichi Hasegawa[3)], Yayoi Miyazono[2)], Yoshihiro Seo[1)], Masaki Ieda[1)]

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

PE1-2 **Ventricular Tachycardia rotating Widespread Low Voltage Zone in Anterior Wall of Right Ventricle in An Adult Patient with Repaired Tetralogy of Fallot**

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

PE1-3 **Interval change of QTc duration in patients of long QT syndrome**

Tomoyuki Kabutoya[1,2)], Yasushi Imai[1,3)], Kana Kubota[1)], Kazuomi Kario[1)]

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

PE1-4 **A case of ablation, pacemaker implantation and medication for supraventricular tachycardia in patient with Fontan operation**

Nobuhiro Nishii[1)], Takahiro Eitoku[2)], YUsuke Shigemitsu[2)], Yoshimasa Morimoto[1)], Saori Asada[1)], Masakazu Miyamoto[1)], Norihisa Tou[1)], Koji Nakagawa[1)], Atsuyuki Watanabe[1)], Hiroshi Morita[1)], Shingo Kasahara[3)], Hiroshi Ito[1)]

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

PE1-5 **The cardiac resynchronization therapy for a patient after biventricular repair with a interventricular dyssynchrony and subpulmonary conduction delay.**

Aya Miyazaki[1)], Hayato Matsutani[2)], Makoto Miyake[1)], Kazuo Yamanaka[3)], Tadashi Ikeda[4)], Kazuyo Kuwano[2)], Hiraku Doi[1)]

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

PE1-6 Two Cases of Cardiac Resynchronization Therapy Responder in Corrected Transposition of Great Arteries Using Epicardial and Transvenous Leads

Shintaro Umemoto[1)], Ichiro Sakamoto[1)], Ayako Ishikita[1)], Hideki Tatewaki[2)], Hazumu Nagata[3)], Shoichi Ohga[3)], Akira Shiose[2)], Hiroyuki Tsutsui[1)]

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

PE1-7 Worsened remnant pulmonary arterial hypertension after 30 years of shunt closure operation for VSD & PDA

Kana Kubota[1)], Tomoyuki Kabutoya[1,2)], Shuichi Ueno[1,3)], Ken Kono[1)], 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

PE1-8 Overexpression of EP4 in pulmonary artery aneurysm in patient with pulmonary arterial hypertension associated with patent ductus arteriosus

Satoshi Akagi[1)], Utako Yokoyama[2)], Kentaro Ejiri[1)], Kazufumi Nakamura[1)], Hiroshi Ito[1)]

1) Department of Cardiovascular Medicine, Okayama University Hospital,
2) Department of Cardiovascular Research Institute, Yokohama City University

17:00-17:54 Poster Presentation 2 (English)

Chair: **Takaya Hoashi** (National Cerebral and Cardiovascular Center)

PE2-1 An emergency operation for erosion rupture of left atrial wall and aortic dissection after ASD closure by Figulla Flex 2

Kosuke Yoshizawa[1)], Keiichi Fujiwara[1)], Toushi Maeda[1)], Otohime Kato[1)], Kentaro Watanabe[1)], Go Ueno[1)], Nobuhisa Ohno[1)], Masao Imai[2)]

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

PE2-2 The effect of bicuspid aortic valve on the blood pressures of young women with Turner syndrome

Soojung Kang

Department of Pediatrics, CHA University School of Medicine, Seongnam, South Korea

PE2-3 A Treatment Strategy for Mitral Stenosis Associated with Patient-Prosthesis-Mismatch After Mitral Valve Replacement; a Case Report

Minako Kagimoto[1)], Manabu Nitta[1)], Tabito Kino[1)], Yusuke Matsumoto[1)], Sae Teranaka[1)], Kiwamu Iwata[1)], Masayoshi Kiyokuni[1)], Naohiro Komura[1)], Daisuke Kamimura[1)], Atsuichiro Shigenaga[1)], Junya Hosoda[1)], Katsumi Matsumoto[1)], Teruyasu Sugano[1)], Tomoaki Ishigami[1)], Toshiyuki Ishikawa[1)], Daisuke Machida[2)], Munetaka Masuda[2)], Koichi Tamura[1)]

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

PE2-4 Impact of hyperthyroidism on Fontan circulation

Hazumu Nagata[1)], Ichiro Sakamoto [2)], Shintaro Umemoto[2)], Ayako Ishikita[2)], Yoshimi Eguchi[1)], Mamoru Muraoka[1)], Shoji Fukuoka[1)], Yusaku Nagatomo[1)], Yuichiro Hirata[1)], Hiroyuki Tsutsui [2)], Shouichi Ohga[1)]

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

PE2-5 Coronary Artery Bypass Grafting Concomitant with Pulmonary Valve Replacement for a Patient with Repaired Tetralogy of Fallot

Sae Teranaka[1)], Manabu Nitta[1)], Hikari Noda[1)], Tabito Kino[1)], Yusuke Matsumoto[1)], Minako Kagimoto[1)], Rie Nakashima[1)], Kiwamu Iwata[1)], Masayoshi Kiyokuni[1)], Naohiro Komura[1)], Daisuke Kamimura[1)], Atsuichiro Shigenaga[1)], Junya Hosoda[1)], Katsumi Matsumoto[1)], Teruyasu Sugano[1)], Tomoaki Ishigami[1)], Toshiyuki Ishikawa[1)], Daisuke Machida[2)], Munetaka Masuda[2)], Koichi Tamura[1)]

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

PE2-6 Current Oral Condition of Patients with Adult Congenital Heart Disease in ACHD Center/Okayama University Hospital

Kazuhiro Omori[1,3)], Norihisa Toh[2,3)], Shinsuke Kochi[1)], Tadashi Yamamoto[1)], Teiji Akagi[2,3)], Hiroshi Ito[2,3)], Shogo Takashiba[1)]

1) Department of Periodontics and Endodontics, Okayama University Hospital,
2) Department of Cardiovascular Medicine, Okayama University Hospital,
3) ACHD Center, Okayama University Hospital

PE2-7 A suspected case of cardiac anomaly because of a marked systolic ejection murmur

Yusuke Matsumoto[1)], Manabu Nitta[1)], Tabito Kino[1)], Sae Teranaka[1)], Minako Kagimoto[1)], Kiwamu Iwata[1)], Masayoshi Kiyokuni[1)], Naohiro Komura[1)], Junya Hosoda[1)], Atsuichiro Shigenaga[1)], Daisuke Kamimura[1)], Katsumi Matsumoto[1)], Teruyasu Sugano[1)], Tomoaki Ishigami[1)], Toshiyuki Ishikawa[1)], Daisuke Machida[2)], Munetaka Masuda[2)], Koichi Tamura[1)]

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

PE2-8 A case of severe pulmonary congestion exacerbated by contrast computed tomography long after intracardiac repair of tetralogy of Fallot.

Takeshi Kashimura[1,2)], Rui Takeda[1)], Kota Nishida[1)], Shinpei Kimura[1)], Yuka Hayashi[1)], Makoto Hoyano[1)], Takao Yanagawa[1)], Toshiki Takano[1)], Kazuyuki Ozaki[1)], Shuichi Shiraishi[3)], Tohru Minamino[1)]

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

PE2-9 Pathological findings of a growing coronary aneurysm in Noonan syndrome

Yoshito Ogihara[1)], Naoki Fujimoto[1)], Junko Ishiura[1)], Hiroyuki Ohashi[2)], Naoki Yamamoto[3)], Hisato Ito[3)], Naoto Kurita[1)], Kaoru Dohi[1)], Hirofumi Sawada[2)], Yoshihide Mintani[2)], Aoki Yoko[5)], Kyoko Imanaka Yoshida[4)], Masaaki Ito[1)]

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

17:00-17:54 Poster Presentation 3 (English)

Chair: **Mikiko Ishido** (Department of Pediatric Cardiology Tokyo Women's Medical University)

PE3-1 A Case of Secundum Atrial Septal Defect with an Unusual Right Atrial Membrane

Keiko Ryo Koriyama[1)], Toshimi Koitabashi[1)], Emi Maekawa[1)], Teppei Fujita[1)], Takashi Miyamoto[2)], Tadashi Kitamura[2)], Kagami Miyaji[2)], Junya Ako[1)]
1) Department of Cardiology, Kitasato University,
2) Department of Cardiovascular Surgery, Kitasato University

PE3-2 Management of bilateral partial anomalous pulmonary venous connection

Shinji Ogawa[1)], Masanobu Maeda[1)], Kan Kaneko[1)], Yasuhide Okawa[2)], Keita Kikuchi[1)]
1) Department of Cardiovascular Surgery, Ichinomiya Nishi Hospital, 2) Toyohashi Heart Center

PE3-3 A Case of Disastrous Lung Injury in a One-lung Fontan Patient: Successful Treatment in Acute Phase and Negative Impact on Hemodynamics

Nao Konagai, Hideo Ohuchi, Jun Negishi, Heima Sakaguchi, Toru Iwasa, Isao Shiraishi, Kenichi Kurosaki
Department of Pediatric Cardiology, National Cerebral and Cardiovascular Center

PE3-4 Successful Radiofrequency Catheter Ablation for Ventricular Tachycardia in a Patient with Double-chambered Right Ventricle.

Tabito Kino[1)], Minako Kagimoto[1)], Manabu Nitta[1)], Yusuke Matsumoto[1)], Sae Teranaka[1)], Yuka Taguchi[1)], Kiwamu Iwata[1)], Masayoshi Kiyokuni[1)], Naohiro Komura[1)], Junya Hosoda[1)], Atsuichiro Shigenaga[1)], Daisuke Kamimura[1)], Katsumi Matsumoto[1)], Teruyasu Sugano[1)], Tomoaki Ishigami[1)], Toshiyuki Ishikawa[1)], Daisuke Machida[2)], Munetaka Masuda[2)], Koichi Tamura[1)]
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

PE3-5 An adult case of portal hypertension after the occlusion of congenital portosystemic shunt

Jun Maeda, Kazuki Yamamoto, Kousei Araki, Kazuki Kodo, Hiroyuki Yamagishi
Department of Pediatrics, Keio University School of Medicine

PE3-6 Renal abscess in Eisenmenger syndrome with a patent ductus arteriosus

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

PE3-7 Post Fontan Operation Patient Lost to Follow-Up for 18 Years

Masaki Kodaira[1)], Makoto Tanaka[1)], Ryota Tabei[1)], Yohei Numasawa[1)], Akihito Saito[2)], Katsura Soma[2)], Atsushi Yao[2)], Issei Komuro[1)]
1) Department of Cardiology,
2) Department of Cardiovascular Medicine, The University of Tokyo Hospital

PE3-8 A successful Fontan type operation for a patient with tricuspid atresia combined with congenital left lung agenesis; 15 years follow-up

Kunikazu Hisamochi[1)], Masahiro Kamada[2)], Takuya Kawabata[1)], Naomi Nakagawa[2)], Yukiko Ishiguchi[2)]
1) Department of Cardiovascular Surgery, Hiroshima Citizens Hospital,
2) Department of Pediatric Cardiology, Hiroshima Citizens Hospital

PE3-9 The usefulness of MRA for check up aortitis syndrome patient -a case report with brachial artery obstruction-

Yasunori Horiguchi[1)], Atsuko Suzuki[2)]

1) Department of Pediatrics, International University of Health and Welfare Atami Hospital,
2) Department of Pediatrics, Tokyo Teishin Hospital

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 mulita-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
● Elevated mean pulmonary artery pressure ● Elevated pulmonary vascular resistance ● Distortion or narrowing of the pulmonary arteries ● Elevated systemic vascular resistance	● Decreased systemic ventricular ejection fraction ● Dyssynchronous ventricular wall motion ● Elevated systemic ventricular end-diastolic pressure	● Significant systemic valve regurgitation	● Tachyarrhythmia ● Bradyarrhythmia	● Hypoplastic left heart syndrome ● Heterotaxy ● Single morphologically right ventricle ● Obstructive bulvoventricular 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)]
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Hideki Tatewaki[1)]，Ichiro Sakamoto[2)]，Satoshi Fujita[1)]，Hazumu Nagata[3)]，Hiroyuki Tsutsui[2)]，Akira Shiose[1)]
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 contemporally 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

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Kensuke Matsumoto[1)]，Sachiko Kido[2)]，Makiko Suzuki[1)]，Makiko Suto[1)]，Ken-ichi Hirata[1)]
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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，杉渕 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

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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

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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

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3) 東京大学 保健健康推進本部

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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 sever 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 sate 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
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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

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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/m^2) 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

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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

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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

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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 $\geqq 3$, 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

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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

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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 arrythmia. We present three cases of ACHD complicated pregnancies. (1) In 26 y.o. congenital ASD women, IPAH occurred suddenly at 26 weeks of gestation and tadalafil 40㎎ 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.

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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

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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

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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

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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

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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

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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/m^2. 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

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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

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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

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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

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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

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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 form 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: era1 (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

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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 SaO_2 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

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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

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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 threating. 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

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Heart and lung transplantation (HLTx) 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 braid dead organ donation were performed in 2017 and only three HLTx 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 HLTx candidates died during waiting for HLTx. Most of possible candidates for HLTx died without listing for HLTx at Japan Organ Transplant Network. Therefore, it may be more important for HTx candidates or recipients to establish palliative care system.
In this talk, current status and future aspects of HLTx 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

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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?

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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 febuxostat prevented gout attach. In contrast, FREED study showed that febuxostat 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

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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

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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

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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

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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

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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

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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/disco-ordinated 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

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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

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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.

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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.

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?

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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/m^2), whereas left ventricular (LV) end-diastolic volume index was small (59 ml/m^2). 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.

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?

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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.

D1 Fontanでの抗凝固療法：必要 vs. 不要

Anticoagulations for Fontan patients: Do you USE or NOT?

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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)]。一方喀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います。

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D2 TOF術後PRに対する成人期PVR：人工弁の種類・サイズの選択

PVR for repaired TOF in Adult: How to choose prosthesis and its size

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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

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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思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

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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

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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

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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/m^2), pulmonary arterial index (225 [189-302] vs 216 [186-255] mm^2/m^2) and pulmonary arterial resistance (1.07 [0.73-1.31] vs 1.04 [0.78-1.66] WU・m^2). However, central venous pressure was increased (10 [9-12] vs 16 [13-16] L/min/m^2, $P < 0.001$) and pulmonary arterial compliance was decreased (51.0 [36.0-75.0] vs 38.9 [25.7-45.9] mm^2/mmHg/m^2, $P < 0.001$).
Conclusions: Lower pulmonary arterial compliance and elvated 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

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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

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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

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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/m^2 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

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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

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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

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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

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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.

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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 treament.
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

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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

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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)

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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

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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

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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

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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_2max). 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_2max. The optimal cutoff value of RVEDV index at less than 80% of predicted VO_2max 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

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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

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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 transcatherter 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. 8cases, log-rank p = 0.039).
Conclusion: The catheter ablation prior to transcatherter 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

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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

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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

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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

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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), goupB (transannular repair: n=27), and groupC (Rastelli: n = 14, Carpenteir-Edward 7, Xenomedica 3, Yamagishi 3, Homograft 1).
Results: RVEDVI (131.6ml/m^2 ± 30.8, p=0.052) and PRF (40.1% ± 13.1, p < 0.001) in groupB were higher than in goupA (117.7ml/m^2 ± 38.3, 20.9% ± 12.0,) and groupC (96.0ml/m^2 ± 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

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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

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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

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Introduction: Few cases reported about re-operaion 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 2months，he was done CoA repair，PA banding.At 7years，he was done Septation operation and PMI.At 24years，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/m^2 (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 8years，he was done Septation operation，PMI.At 32years，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 plicationTVR 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ァ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

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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/m^2 (group A, n=25) and the group not exceeding 160 ml/m^2 (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/m^2 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.

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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

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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 resonace imaging (MRI).
Methods: We performed MRI in 24 subjects (13 subjects after Fontan operation and 11 subjects with biventricular circuation). 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/m^2, 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 differencein 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

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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/m^2. 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

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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

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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/1.73m^2) 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/m^2, 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

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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/m^2, PAI was 228 ± 67mm^2/m^2. 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

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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

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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. Ultrasonocadiography 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

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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

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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 resuls of the joint operations in Shizuoka prefectural hospitals

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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

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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 ファ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

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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/m^2 in normal controls, but increase in forward SV was quite impaired (from 77 ± 20 to 80 ± 20 mL/m^2) 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

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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 1.74m^2, and 23mm-BPVs for 21- and 41-year-old females with BSA of 1.39m^2 and 1.43m^2, 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.

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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 ($\geqq 513$) 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

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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 × ln (creatinine) + 5.11 × ln (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 ($\leqq 10.4$) (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

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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

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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

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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

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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

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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 heathy 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

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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 presser 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

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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 deice 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 (ASO)(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

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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 m^2. 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

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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

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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/m^2 and postoperative RVEDVI (1yr / 3yrs) were 116.4 ± 28.6 / 119.4 ± 36.7ml/m^2 which showed marked reduction at 1yr and no change thereafter. Preoperative RVEF was 42.7 ± 4.5% and postoperative RVEF (1yr / 3yrs) were 41.8 ± 8.0 / 44.2 ± 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

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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

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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

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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

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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 ± 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 ± 0.4m/s. CMRI showed significant decrease of RVEDVI from 210 ± 59 to 119 ± 40 ml/m^2. 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

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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、不得意があるように、障害というのも意外と単純なものなのかと思います。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

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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と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)

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榊原記念病院

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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%）、経膣分娩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検討

Consideration of muscle strength measurement in fontan postoperative adult patients

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【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.

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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 steadly.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 unknowing, 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

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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

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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

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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

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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 ± 37.0ms, P=0.99). Although QTc durations did not changed from 5 years ago and latest visit (471.4 ± 60.7 vs. 457.8 ± 41.7ms, P=0.29) in young group, QTc durations increased from 5 years ago to latest visit (464.1 ± 34.0 vs. 476.3 ± 31.8ms, 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

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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.

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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/m^2; LV ejection fraction (EF) 25%; RVEDVI 110 ml/m^2; 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

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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

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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 repeir" 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

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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

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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

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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

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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 cm^2 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

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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 1. 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

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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/m^2 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

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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/m^2, 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

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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.

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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

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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

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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 posterily 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

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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

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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 hematothorax 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.

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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

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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

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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

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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. Furthemore, 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

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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 1 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－

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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 hs 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 patinet 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.

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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

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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.

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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

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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

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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

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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

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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

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【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）合併妊娠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

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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

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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

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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

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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.

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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-S^{TM} 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-S^{TM} 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

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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

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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; CHA_2DS_2-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.

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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 deference 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.

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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.

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A 60-year-old man was admitted to Saku Central Hospital because of right ventricular disfunction. He was diagnosed of Tetralogy of Fallot and performed original Blalock-Taussig sunt operation in 6 years old and total correction in 20 years old. Preoperative magnet Image Resonance showed RVEDVI was 185ml/m^2 and LVEDVI was 75.4ml/m^2. 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.

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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/m^2, LVEDVI 110.2 ml/m^2, LVEF 45 %, RVEDVI 79.4 ml/m^2, 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.

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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 (SpO_2 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

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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

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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/m^2. 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

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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

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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

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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.

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Background: Sleep apnea syndrome (SAS)is assocated with cardiovascular adverse events. We present 3 cases of adult patients after Fontan opeartion in whom aggressive introductions of noninvasive positve 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/m^2 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

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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術後遠隔期に鋳型気管支炎を発症した一例

A case of plastic bronchitis late after Fontan procedure

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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/m^2. 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

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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/m^2, 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

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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 atriopulmonay connection (APC) Fontan surgery who resulted in severe heart failure due to atrial tachycardia (AT)

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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

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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

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【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【良い友達の選択】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の感覚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

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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.

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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

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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

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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

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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.

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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 remifentanil 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

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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

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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

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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 sigmod septum was shown. Therefore we diagnosed as not HOCM but discrete type subvalvular aortic stenosis with sigmoid septum. Aftet the surgical resection of subaortic septal muscle in addition to mechanical aortic valve replacement, LVOTO improved, just 10mHg 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 remarkedly in post-operation of CABG and ASD closure.

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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/m^2, Qp/Qs 2.07, PVR 2.56WU・m^2. 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 remarkedly (CI 1.53L/min/m^2、PVR 7.2WU・m^2、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

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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

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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

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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 m^2. 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

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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

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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

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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

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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・m^2, 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・m^2). 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

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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/m^2. 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

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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

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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

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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/m^2, 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Platpnea-Orthodexia syndromeの1例

Platypnea-orthodeoxia syndrome in an elder women treated by device closure for patent foramen ovale.

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Platypnea-orthodeoxia 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 devise 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

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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

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A 74 years old woman came to our hospital 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/m^2. 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

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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

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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 bioprosthesis had progressed and echocardiogram 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 VO_2, which was equivalent to 60% of predicted normal values for her age. After 3 months of exercise training (ET), peak VO_2 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

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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

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Background: The causes of pulmonary arterial pulmonary 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－1例報告

How to deal with menstrual disorder in a patient with Fontan circulation?

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【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・プロゲスチン療法（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.

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【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

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【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

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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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