Usefulness of the Right Parasternal Approach to Evaluate the Morphology of Atrial Septal Defect for Transcatheter Closure Using Two-Dimensional and Three-Dimensional Transthoracic Echocardiography

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Background: The aim of this study was to demonstrate the feasibility and usefulness of addition of the right parasternal approach to the conventional left parasternal and apical approaches using two-dimensional (2D) and three-dimensional (3D) transthoracic echocardiography (TTE) for morphologic evaluation in cases of transcatheter closure of atrial septal defects (ASDs).

Methods: In 112 consecutive patients with ASDs, the morphology of the defects was evaluated for transcatheter closure in the right parasternal view in addition to the conventional left views using 2D and 3D TTE. Measurements of the maximal ASD diameter and detection of deficient rim obtained on 2D TTE were compared with those obtained by 2D transesophageal echocardiography. The shapes and locations of ASDs visualized by 3D TTE were compared with those visualized by 3D transesophageal echocardiography.

Results: In 88 patients (80.0%), optimal images from the right parasternal approach for morphologic evaluation of ASDs were obtained. Although there was a significant difference in maximal ASD diameter obtained only in the conventional left approach compared with transesophageal echocardiographic measurements ($P < .05$), when the right parasternal approach was applied, a significant difference was not found ($P = .18$), and the diagnostic concordance of the rim deficiency was improved from 85.2% to 90.9%. Three-dimensional TTE from the right parasternal approach improved visualization of the shape and location of ASDs from 65.5% to 74.5%.

Conclusions: Additional use of the right parasternal approach enables detailed morphologic evaluation for transcatheter closure of ASDs. In patients with suboptimal images on 3D TTE in the left conventional approach, additional 3D TTE in the right parasternal approach can improve the feasibility of obtaining optimal 3D images to evaluate the shapes and locations of ASDs. (J Am Soc Echocardiogr 2012;25:376-82.)

Keywords: Right parasternal approach, Transthoracic echocardiography, Transesophageal echocardiography, Atrial septal defect

Transcatheter closure of atrial septal defects (ASDs) has recently become established as a safe and effective treatment, and the procedure has become an alternative to a surgical approach.[1-5] Appropriate patient selection for transcatheter closure is the most important factor for success in this procedure,[4] and morphologic evaluation, including evaluation of maximal ASD diameter and surrounding rims by echocardiography, is essential. Although two-dimensional (2D) transthoracic echocardiography (TTE) in the left parasternal, apical, and subcostal views is routinely used for this purpose, previous studies have demonstrated that these views enable only limited morphologic evaluation of ASDs.[6-9] Real-time three-dimensional (3D) echocardiography, in which a comprehensible en face view of ASDs is obtained, has been available in a clinical setting.[10-12] Three-dimensional TTE is expected to improve understanding of the morphology of ASDs, but data are limited, and it is difficult to obtain good-quality images on 3D TTE using the left parasternal and apical approaches.[13-15] In this regard, 2D transesophageal echocardiography (TEE) and 3D TEE have been widely accepted and established as diagnostic modalities in evaluation of the morphology of ASDs for transcatheter closure because of their high-quality imaging[16-21]; however, TEE has a semi-invasive nature.

The right parasternal approach, in which the transducer is placed to the right of the sternum in the right lateral decubitus position, was

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Abbreviations

ASD = Atrial septal defect

TEE = Transesophageal echocardiography

3D = Three-dimensional

TTE = Transthoracic echocardiography

2D = Two-dimensional

reported to enable better visualization of ASDs and evaluation of the direction of shunt flow in patients with ASDs because it obtains a longitudinal vena cava superior-inferior plane of the interatrial septum.[22-26] In addition, 3D TTE in this approach might improve the feasibility of obtaining optimal en face images of ASDs. However, there have been limited data on the usefulness of the right parasternal approach using 2D and 3D TTE for morphologic evaluation in cases of transcatheter closure. Therefore, we sought to assess the usefulness of the right parasternal approach in addition to the conventional left parasternal and apical approaches in evaluating ASD morphology for the suitability of transcatheter closure using 2D and 3D TTE.

METHODS

Study Population

A total of 112 consecutive patients (40 men and 72 women) were prospectively evaluated for transcatheter closure of ASDs using the Amplatzer Septal Occluder (AGA Medical Corporation, Plymouth, MN) with 2D and 3D TTE. Two patients with ASDs other than the secundum type were excluded from this study (one had a superior sinus venosus ASD and the other had an unroofed coronary sinus ASD). Therefore, 110 patients were included in the study. All patients except for one were referred from other hospitals to our institution for transcatheter ASD closure. Age at the examination ranged from 6 to 84 years (mean, 46.1 ± 20.5 years). Two-dimensional TEE and 3D TEE were performed <3 days after TTE by a blinded observer. The study was approved by the local ethics committee.

Two-Dimensional TTE

Two-dimensional TTE was performed using a commercially available ultrasound system with a 3.5-MHz transducer (Vivid 7; GE Healthcare, Wauwatosa, WI). Right ventricular midcavity diameter was measured in the apical four-chamber view according to the guideline of American Society of Echocardiography.[27] In all patients, the morphology of ASDs was evaluated using TTE in the left lateral decubitus position from the left parasternal and apical approaches (conventional left approach). Then a transducer was positioned on the right parasternal border with the patient in the right lateral decubitus position (right parasternal approach). Maximal ASD diameter and the minimal diameter of surrounding rims were measured at end-systole by carefully sweeping the transducer from right to left and top to bottom of the interatrial septum in both approaches. Regarding the maximal ASD diameter, first, the ASD diameter was measured using the conventional left approach (ASD_L diameter), and then the ASD diameter was measured by the right parasternal approach (ASD_R diameter). The maximal ASD diameter was considered the maximal value from measurements by both approaches. The surrounding rims were classified according to location as superoanterior, inferoanterior, superoposterior, or inferoposterior. The superoanterior rim was measured as the distance between the aorta and the defect. The inferoanterior rim was measured as the distance from the atrioventricular valves. The inferoposterior rim was measured as the distance from the left atrial wall. The superoposterior rim was measured as the distance from the defect to the superior vena cava and to determine the inferoposterior rim as the distance from the defect to the inferior vena cava (Figure 1). Any rim length < 5 mm was considered deficient. First, the presence or absence of a deficient rim was evaluated using the conventional left approach, and then the right parasternal approach was used.

Three-Dimensional TTE

Three-dimensional TTE was performed after 2D TTE using a commercially available ultrasound system with a 3V transducer (Vivid 7). In all patients, the left parasternal approach was first chosen and optimized, and then loops from five consecutive cycles were acquired and digitally stored. In cases with suboptimal 3D images by the left parasternal approach, we attempted to obtain optimal 3D images using the right parasternal approach. In all patients, at least three acquisitions were performed, and the data set with the best image quality was chosen for analysis. The shapes and locations of ASDs were visually evaluated on the best 3D images.

Two-Dimensional and 3D TEE

Two-dimensional and 3D TEE were performed using a commercially available ultrasound system (iE33; Philips Medical Systems, Andover, MA). Maximal ASD diameter (ASD_{TEE} diameter) and minimal diameter of the surrounding rims were assessed at end-systole using both 2D TEE and 3D TEE, as previously reported.[18] To evaluate surrounding rims using 2D TEE, the superoanterior rim was measured as the distance between the aortic annulus and the defect in the horizontal plane at 0° to 30°. The inferoanterior rim was measured as the distance between the defect and atrioventricular valves in the four-chamber view at 135°. The longitudinal plane around 90° was used to determine the superoposterior rim as the distance from the defect to the superior vena cava and to determine the inferoposterior rim as the distance from the defect to the inferior vena cava (Figure 1). The rim length was considered deficient if the length was <5 mm.

Real-time 3D transesophageal echocardiographic data were obtained after a complete 2D transesophageal echocardiographic study. Real-time 3D zoom mode, which displays a smaller, magnified pyramidal data set, was used to evaluate the shapes and locations of ASDs as well as the rough relation to surrounding structures.

Two-dimensional and 3D transesophageal echocardiographic data were considered reference standards. In patients with optimal images obtained on both approaches, ASD_{TEE} diameter and detection of deficient rims obtained on 2D TEE were compared with those obtained on 2D TTE. The shapes and locations of ASDs using 3D TTE were compared with those obtained using 2D and 3D TEE.

Measurement Variability

ASD diameter and the minimal diameter of the surrounding rims obtained using TTE were measured by two independent observers and by one observer two times 1 month apart in 10 randomly selected patients to determine interobserver variability and intraobserver variability. Variability was assessed as the absolute difference between two measurements expressed as a percentage of their mean values.

Statistical Analysis

Categorical data are expressed as numbers and percentages and continuous data as mean ± SD. The significance of baseline differences

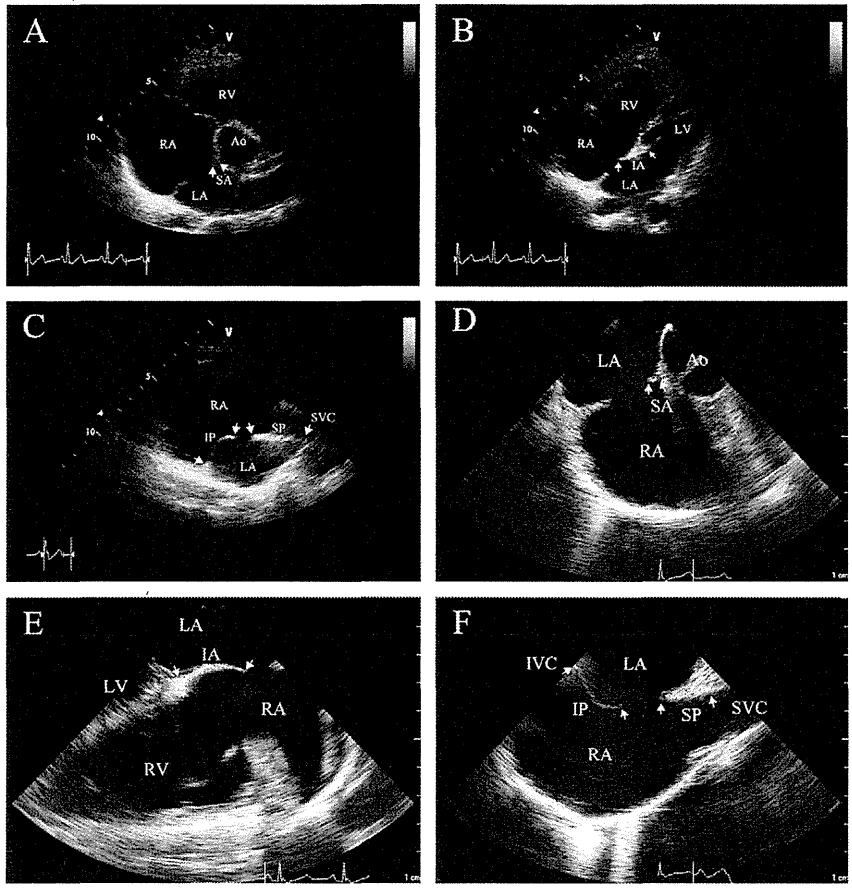

Figure 1 Measurement of maximal ASD diameter and the surrounding rims on 2D TTE and 2D TEE. The surrounding rim is measured from mark (*white arrow*) to mark (*white arrow*). **(A)** Left parasternal short-axis view, **(B)** left parasternal four-chamber view, **(C)** right parasternal longitudinal view, **(D)** short-axis transesophageal echocardiographic view (0°–30°), **(E)** four-chamber transesophageal echocardiographic view (135°), **(F)** biatrial transesophageal echocardiographic view (90°). The surrounding rims are measured at end-systole (*white arrow*). *Ao*, Aorta; *IA*, inferoanterior rim; *IP*, inferoposterior rim; *IVC*, inferior vena cava; *LA*, left atrium; *LV*, left ventricle; *RA*, right atrium; *RV*, right ventricle; *SA*, superoanterior rim; *SP*, superoposterior rim; *SVC*, superior vena cava.

was determined using paired and unpaired *t* tests as appropriate. Categorical variables are expressed as counts and percentages and were compared using χ^2 or Fisher's exact tests as appropriate. Comparisons between measurements were done using Pearson's linear regressions analysis. The agreement of the two methods was evaluated using the Bland-Altman test. *P* values $< .05$ were considered statistically significant. Statistical analyses were done using SPSS version 18.0 (SPSS, Inc., Chicago, IL).

RESULTS

Baseline Characteristics of Study Population

Table 1 shows the baseline characteristics and 2D transthoracic echocardiographic parameters of the study population. All patients showed hemodynamically significant atrial shunts or the presence of right atrial and ventricular volume overload.

Feasibility of 2D TTE in the Right Parasternal Approach

Two-dimensional TTE with the conventional left approach enabled the detection of shunt flow on color-flow Doppler imaging and visualization of the optimal images for the measurement of defects in all patients. Detection of shunt flow in the right parasternal approach on color-flow Doppler images was successful in 102 patients (92.7%). Optimal images with the right parasternal approach for measurements of defects and surrounding rims were visualized in 88 patients (80.0%). When all patients were divided into two groups according to age, <40 years ($n = 42$; mean age, 24.1 ± 10.4 years) and ≥40 years ($n = 68$; mean age, 59.6 ± 11.5 years), the percentage of patients in whom optimal images to measure ASD diameter and surrounding

Table 1 Baseline characteristics and transthoracic echocardiographic parameters of the study population ($n = 110$)

Variable	Value
Men/women	39/71
Age (y)	46.1 ± 20.5 (6–84)
Height (m)	1.58 ± 0.12 (1.13–1.83)
Weight (kg)	54 ± 12.3 (17–92)
Body surface area (m^2)	1.53 ± 0.22 (0.75–2.14)
Right ventricular midcavity diameter (mm)	41.8 ± 5.4 (28–55)
Pulmonary flow/systemic flow ratio	2.4 ± 0.7 (1.2–4.1)

Data are expressed as numbers or as mean ± SD (range).

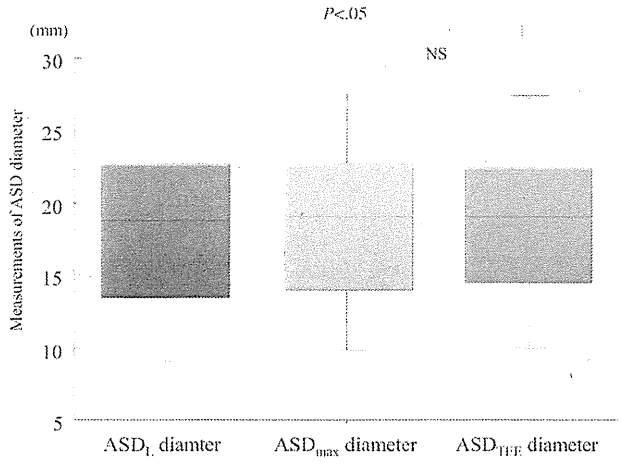

Figure 2 Box plots showing the comparison ASD_L diameter obtained with the conventional left approach (*red box*), maximal ASD diameter obtained with both the left conventional and right parasternal approaches (*orange box*), and ASD_{TEE} diameter (*blue box*).

rim were obtained in the right parasternal approach was significantly higher in those aged <40 years than in those aged ≥40 years (90.5% vs 73.5%, $P = .033$).

Data for the 88 patients in whom optimal 2D transthoracic echocardiographic images for measurements of ASD diameter and surrounding rims were obtained by both approaches were analyzed in our study.

Morphologic Evaluation with 2D TTE and TEE

Maximal ASD diameters between ASD_L diameter, maximal ASD diameter, and ASD_{TEE} diameter were compared in 88 patients with optimal images from the right parasternal approach. There was a small but significant difference between ASD_L diameter and ASD_{TEE} diameter (18.5 ± 6.9 vs 19.0 ± 6.9 mm, $P < .05$). However, when the diameter obtained with the right parasternal approach was taken into account in addition to the diameter obtained with the conventional left approach, a significant difference was not found between measurements of maximal ASD diameter and ASD_{TEE} diameter (18.8 ± 6.7 mm, $P = .18$; Figure 2). Bland-Altman analysis showed the smallest mean absolute differences and narrower limits of agreement when the measurement from the right parasternal approach was added to that from the conventional left approach (Figure 3).

TEE demonstrated that 17 patients (19.3%) had centrally positioned ASD, 55 (62.5%) had superoanterior rim deficiencies, three (3.4%) had inferoposterior deficiencies, five (5.7%) had both superoanterior and inferoposterior deficiencies, and eight (9.1%) had multiple ASDs. Although the detection of a deficient rim showed concordance in 75 patients (85.2%) between TTE with the conventional left approach and 2D TEE, diagnostic concordance was improved to 90.9% by adding the right parasternal approach. In nine patients with inferoposterior rim deficiencies, diagnostic accuracy of the rim deficiency was improved from 66.7% to 100% when the right parasternal approach was added to the conventional left approach.

Evaluation of ASDs with 3D TTE

Although 3D TTE from the left parasternal approach could visualize the 3D optimal image for understanding the shape and location of

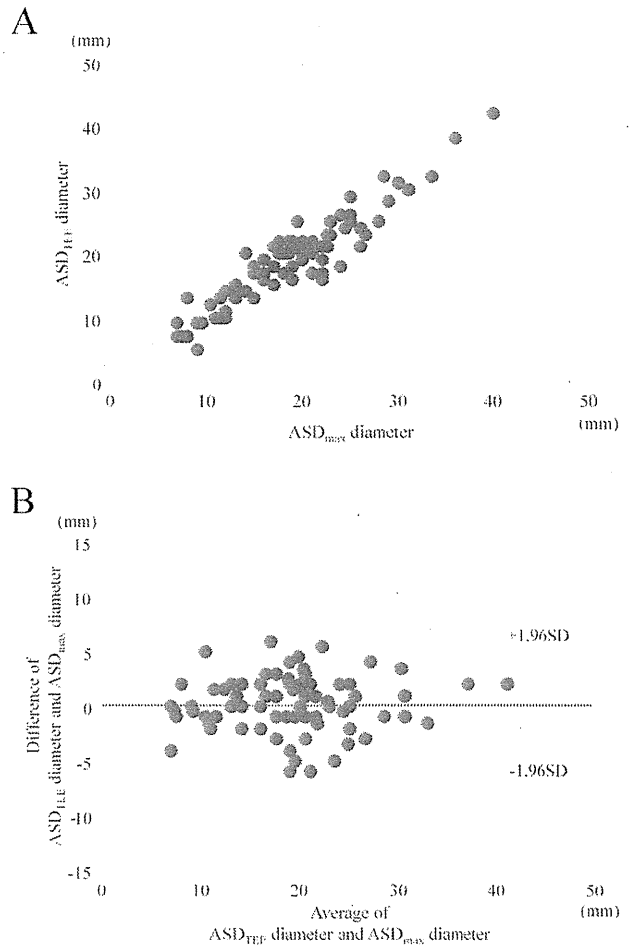

Figure 3 **(A)** Correlations of maximal ASD diameter measured by 2D TTE versus 2D TEE. **(B)** Bland-Altman plot of ASD diameter difference between the measurements on 2D TTE and those on 2D TEE as a function of the average measurements. The *thick continuous line* and *dotted line* indicate the mean ± 1.96 SD of the difference, respectively.

ASDs in only 72 patients (65.5%), the use of the right parasternal approach improved the visualization of optimal 3D TTE images to 74.5% (Figures 4 and 5).

Measurement Variability

Interobserver and intraobserver variability were 1.1% and 0.6%, respectively, for ASD diameter measured by 2D TTE; 6.1% and 6.8%, respectively, for superoposterior rim measurement by 2D TTE; and 9.7% and 8.0%, respectively, for inferoposterior rim measurement by 2D TTE.

DISCUSSION

Our study demonstrated that 2D TTE with the addition of the right parasternal approach to the conventional left approach is feasible and enables evaluation of the morphology of ASDs for transcatheter closure with satisfactory accuracy compared with evaluation by 2D TEE. In particular, the right parasternal approach contributes greatly

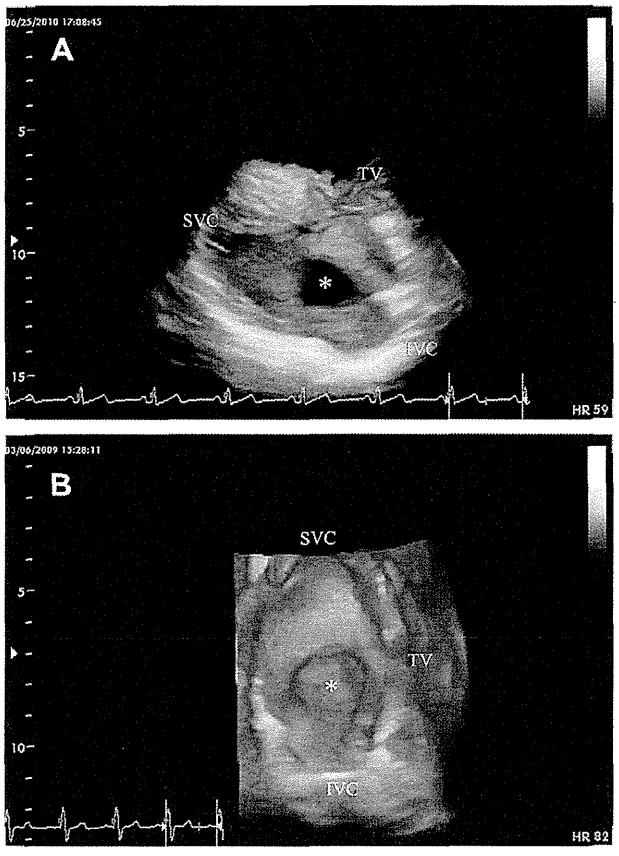

Figure 4 Three-dimensional transthoracic echocardiography of various shapes of the secundum-type ASD (*asterisk*) viewed from the right atrium. **(A)** Superoanterior rim deficient from the left parasternal approach, **(B)** inferoposterior rim deficient from the right parasternal approach. *IVC*, Inferior vena cava; *SVC*, superior vena cava; *TV*, tricuspid valve.

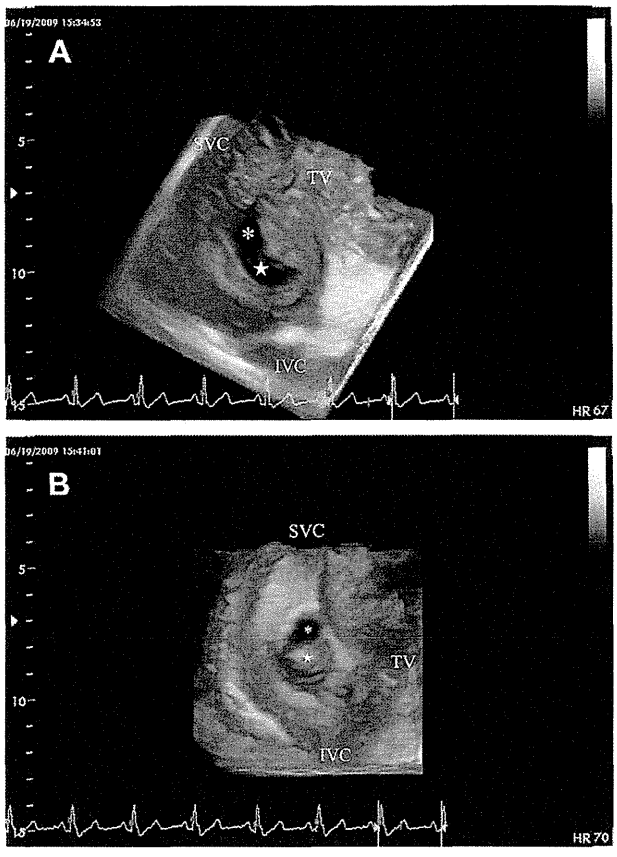

Figure 5 Three-dimensional TTE of the secundum-type ASD (*asterisk*) with an atrial septal aneurysmal (ASA) viewed from the right atrium. **(A)** The ASA (*white star*) was dropped out from the left parasternal approach **(B)** but was clearly visualized from the right parasternal approach. *IVC*, Inferior vena cava; *SVC*, superior vena cava; *TV*, tricuspid valve.

to the identification of rim deficiency, especially in patients with inferoposterior rim deficiencies. In terms of the acquisition of optimal 3D transthoracic echocardiographic images, the addition of the right parasternal approach to the conventional left approach can improve the feasibility of ASD morphologic evaluation.

Feasibility of the Right Parasternal Approach

Previous studies have shown that the right parasternal approach is a reliable technique for detection of ASDs.[22-26] Iliceto *et al.*[25] reported that ASDs were identified using the right parasternal approach in 13 of 17 patients (76.5%) and that the right parasternal approach improved the feasibility of 2D TTE for the detection of ASDs. In our study, we could detect ASDs on color-flow Doppler imaging by 2D TTE using the right parasternal approach with high sensitivity (92.7%). Advances in the technology of echocardiography may have greatly contributed to its high feasibility compared with previous studies. In our study, optimal images in the right parasternal approach were obtained more frequently in younger patients than in older patients, who sometimes have obesity or lung disease. A previous study also demonstrated that the right parasternal view was often easily obtainable in neonates and young children.[26] Therefore, especially in younger patients with intolerance of TEE, TTE including the right parasternal approach can contribute to evaluating ASD morphology for transcatheter closure.

Morphologic Evaluation of ASDs

Previous studies have demonstrated that appropriate patient selection is essential for successful transcatheter closure of ASDs using the Amplatzer Septal Occluder.[16,17] Two crucial parameters, maximal ASD diameter to choose an appropriately sized device and tissue rim dimensions all around the defect to optimize placement of the device, should be measured to select patients for transcatheter closure of ASDs in addition to detection of atrial shunts or echocardiographic findings of right ventricular volume overload.[12] Although TEE is considered the gold standard in evaluating ASD morphology for the suitability of transcatheter closure, TEE has a semi-invasive nature. TTE used to perform a detailed morphologic evaluation of ASDs before TEE can lead to avoiding oversight and shortening transesophageal examination time. Therefore, detailed morphologic evaluation, including evaluation of maximal ASD diameter and surrounding rims by TTE, is important. In our study, the use of the additional right parasternal approach in TTE improved measurements of maximal ASD diameter and detection of rim deficiencies and enabled morphologic evaluation comparable with that obtained by TEE. In the conventional left approach, because the

direction of the ultrasound beam is almost parallel to the interatrial septum, there is frequent dropout of interatrial septal echoes in the region of the mid portion (fossa ovalis). Previous studies have shown that echo dropout in the region of the mid portion frequently occurs and can lead to false diagnoses of large defects.[22,28,29] One of those previous studies showed that the maximal ASD diameter measured with 2D TTE was larger than that measured with 2D TEE and that there was a poor correlation between these measurements because of dropout in the region of the fossa ovalis.[29] The subcostal approach is another useful method for visualizing perpendicularly the interatrial septum.[28] Although 2D TTE from the subcostal approach enables the detection of shunt flow across ASDs easily in pediatric patients,[25,28] this approach can provide suboptimal images or incomplete clinical information with regard to morphologic evaluation for transcatheter ASDs closure, especially in adult patients, because of the limited echocardiographic window.[6-8] The right parasternal approach is a method that can provide better evaluation of the structure of the interatrial septal because the ultrasound beam passes in a plane perpendicular to the interatrial septal.[22-26] The use of the right parasternal approach provided better visualization of the superior vena cava and inferior vena cava entering into the right atrium. The right parasternal approach contributed greatly to the detection of deficient rims in the present study, particularly for the inferoposterior rim, which is sometimes difficult to visualize clearly by TEE. A previous study showed that inferior rim deficiency was a significant factor associated with unsuccessful transcatheter closure.[30] Therefore, the additional right parasternal view can contribute to appropriate patient selection and the prediction of procedural results for transcatheter ASD closure.

Considerable experience and operator skills are necessary for evaluating ASD morphology precisely using 2D TTE, equivalent to 2D TEE. There are some issues of technique and some pitfalls. The ultrasound beam should be passed as perpendicularly to the interatrial septum as possible. In addition, gain adjustment using time-gain compensation, focus position, and the use of zoom mode (high frame rate) should be set for morphologic evaluation. In addition, control of respiration and body position should be required to avoid potential artifacts such as dropout and side lobe.

Three-Dimensional Echocardiography Using the Right Parasternal Approach

ASDs are known to have complex geometry that may be elliptical, ovoid, or multiple defects or fenestrations.[20,21] Three-dimensional echocardiography provides more spatial anatomic information without the need for mental 2D reconstruction. There have been several studies on the usefulness for assessing ASDs of 3D transesophageal echocardiographic reconstruction[10-12] and real-time 3D TEE.[18-21] In addition, some previous studies have demonstrated possible usefulness of 3D TTE for evaluating ASD morphology.[18,21,22] Acar *et al.*[31] reported a high correlation between 3D transthoracic and 3D transesophageal echocardiographic measurements of maximal ASD diameter in pediatric patients. Van den Bosch *et al.*[13] demonstrated that real-time 3D TTE enabled reliable assessment of the dimensions of ASDs and the exact location and extent of the surrounding rim, and they reported an excellent correlation of real-time 3D transthoracic echocardiographic findings compared with surgical and 2D transesophageal echocardiographic measurements of ASDs in pediatric and relatively young adult patients. Chen *et al.*[14] reported that 3D TTE could provide accurate diagnosis of sinus venosus ASDs. In terms of the usefulness of 3D TTE in the right parasternal approach for evaluating ASD morphology, although there was one case report, studies with a sufficient number patients and including patients with a wide age range have been limited. In the present study, we demonstrated that the use of the right parasternal approach in addition to the conventional left approach improved the feasibility of visualization of satisfactory 3D images, even in adult patients.

Limitations

This study had some limitations. First, the number of patients in this study was relatively small to conclude whether the extent of variation in ASD type was taken into account. Second, almost all patients in the present study were referred from other hospitals to our institution for transcatheter closure of ASDs. Therefore, patient selection bias could have existed before enrollment. Third, visualizing ASDs and surrounding rims using the right parasternal approach requires a learning curve. In this study, interobserver variability and intraobserver variability were quantitatively evaluated by two skilled operators. Finally, evaluation in the subcostal view may improve the feasibility and accuracy of TTE.

CONCLUSIONS

The use of the right parasternal approach enables detailed morphologic evaluation, especially for longitudinal ASD diameter, superoanterior rim diameter, and inferoposterior rim diameter of ASDs in a longitudinal vena cava superior-inferior plane of the interatrial septum. In patients with suboptimal images with the conventional left approach, additional 3D TTE in the right parasternal approach can improve the feasibility of obtaining optimal 3D images to evaluate the shapes and locations of ASDs.

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IMAGES IN CARDIOLOGY

Combined Subaortic and Mid-ventricular Obstruction With Significant Aortic Stenosis Diagnosed by Triphasic Doppler Flow Pattern

Multiple Levels of Left Ventricular Outflow Tract Obstruction

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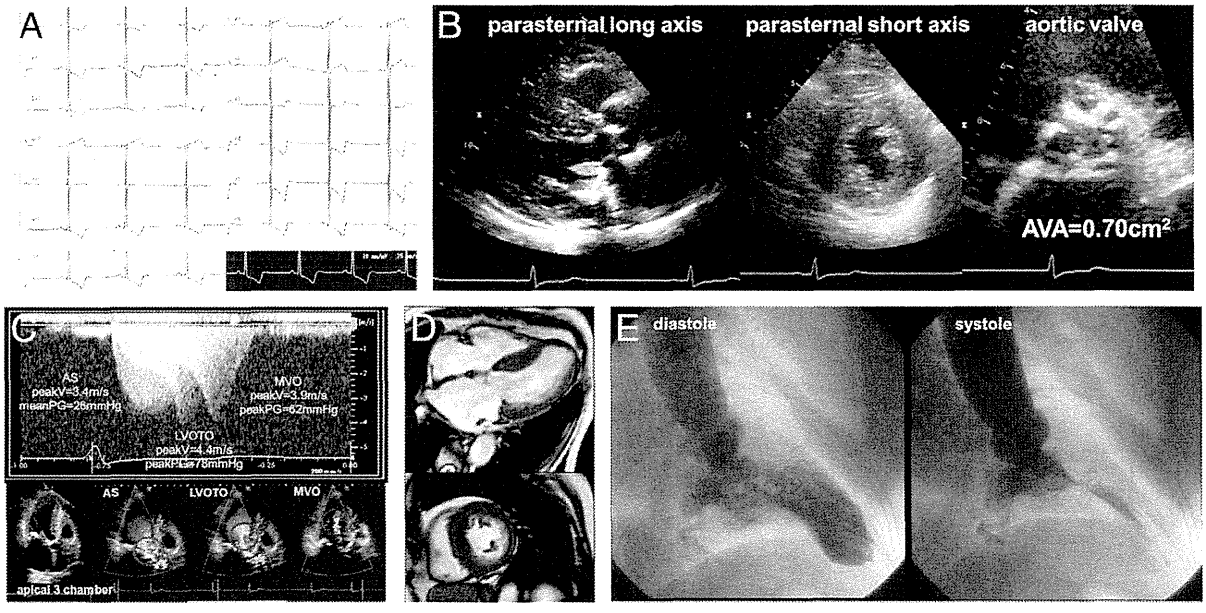

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A 74-year-old woman was referred to our hospital for evaluation of increasing dyspnea and fatigue. Her electrocardiogram showed marked left ventricular (LV) hypertrophy and T-wave inversion over leads V_3 to V_6 **(A)**. Transthoracic echocardiography demonstrated significant aortic stenosis (AS) and gross asymmetrical LV hypertrophy **(B**, Online Videos 1, 2, and 3), which caused combined subaortic and mid-ventricular obstruction. Doppler echocardiography demonstrated triphasic severe pressure gradients through the LV outflow tract (LVOT), mid-peaking symmetric velocity, and 2 asymmetric late-peaking "dagger-shaped" velocities. The subaortic gradient reached a peak in mid-systole, and the mid-ventricular gradient reached a peak in late systole and persisted to early diastole (**C**, Online Videos 4 and 5). Cardiac magnetic resonance imaging showed marked asymmetrical LV hypertrophy **(D)**. A left ventriculogram also revealed dynamic mid-cavity and LVOT obliteration **(E)** (Online Video 6). Coronary angiography showed normal vessels.

The patient was diagnosed with multiple levels of LVOT obstruction with significant AS. Septal myectomy and aortic valve replacement were successfully performed. This case highlighted the significance of meticulous examination by Doppler echocardiography for evaluation of dynamic LVOT obstruction.

Pediatric perfusion in Japan: 2010 practice survey

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Abstract

We report here Japan's first pediatric perfusion survey. It covers practices from January 2007 through December 2009. Of the 70 congenital heart centers contacted, 53 (76%) completed the survey. They reported performing 3,379 pediatric cardiopulmonary bypass (CPB) procedures in 2009, 3,408 in 2008, and 3,358 in 2007. Twenty-eight percent of all centers used CPB circuits with a priming volume between 151-200 ml. All centers used pre-bypass ultrafiltration and only 6% used retrograde autologous priming. A biomaterial-coated circuit was used by 78% of the centers, a roller pump as the arterial pump by 91%, vacuum-assisted venous drainage by 39%, dilutional ultrafiltration by 48%, and modified ultrafiltration at the end of the procedure by 30%. A regional oxygen saturation monitor was used by 69% of the centers and high flow (150-200 ml/kg/min) management with alpha-stat blood gas control was standard during moderate to normothermic CPBs. Crystalloid cardioplegia solution was used as myocardial protection by 56% of the centers, electronic recording of monitoring data by 51%. The centers performed 98 pediatric extracorporeal membrane oxygenation procedures in 2007, 109 in 2008, and 119 in 2009; 58% of the centers used a centrifugal pump. This survey provides a description of the current practice in Japan. Future surveys will identify trends and rate of change in practice.

Keywords

perfusion; cardiopulmonary bypass; survey; pediatric; congenital heart surgery

Introduction

The history of perfusion is marked by serious challenges and major accomplishments. John Gibbon's development of the cardiopulmonary bypass (CPB) machine in 1953 marked the beginning of cardiopulmonary support during cardiovascular surgery[1]. The technique of CPB was widely implanted by Kirklin, Lillehei, and others[2-4]. It has become an indispensable tool for cardiovascular surgery, whose progress has been astonishing. In congenital heart surgery, the improved results and lower mortality rate that have followed from evidence-based perfusion (EBP) practices are of special note.

In 2005, with the goal of providing an infrastructure for collaboration between healthcare professionals interested in the analysis of outcomes of treatments provided to patients with congenital cardiac disease, and the ultimate aim of improvement in the quality of care provided to these patients[5], the International Consortium for Evidence-Based Perfusion was established as a collaboration of perfusion societies, clinicians, and industry to improve the delivery of care and outcomes for patients worldwide[6]. It should lead to the highest quality of comprehensive care to all congenital heart disease patients[5].

An estimated 9,000 congenital heart surgery procedures are conducted annually in Japan[7]. To date, little is known about actual perfusion management and the organization framework for perfusion in Japan. A clinical database covering multiple congenital heart centers that collects data on perfusion performance would enable us to establish a database that would lead to improved EBP techniques. However, we have, as yet, no basis for a pediatric perfusion database in Japan. The aim of the survey

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was to determine current practice across Japanese centers. The unit of analysis for this study was individual centers.

Materials and Methods

We sent a 45-question perfusion survey in June 2010 by government mail and electronic mail to chief perfusionists in 70 congenital heart centers in Japan. We asked the recipients to fill in blanks and check all applicable boxes based on the predominant practices in their center. We clarified inconsistencies in the survey results by contacting respondents by e-mail or by phone.

The survey, which enquired about perfusion practices from January 2007 through December 2009, covered pediatric caseloads, CPB circuits (priming volume, solution, coating), CPB components (arterial pump, oxygenator, heart-lung machine), CPB monitoring (regional oxygen saturation monitor, in-line monitor, electronic recording), CPB management (perfusion flow, temperature, filtration, vacuum-assisted venous drainage, blood transfusion), blood gas management, hypothermia technique, myocardial protection (cardioplegia solution, temperature), pediatric extracorporeal membrane oxygenation (ECMO) and employment of perfusionists.

Results

Of the 70 centers contacted, 53 (76%) completed the survey.

Program demographics

Procedures. The respondents reported 3,379 pediatric CPB procedures in 2009, 3,408 in 2008, and 3,358 in 2007. The median number of CPB procedures performed per center per year was 50 (range, 4-445) and the mean caseload was 0-50 for 72% of the perfusionists, 51-100 for 21%, and 101-150 for 5%.

CPB circuit

Priming volume. Table 1 shows hospital priming volume of the CPB circuit.

Table 1. Priming volume of CPB circuit used by hospitals during pediatirc cardiac surgery in Japan, 2007-2009.

Priming Volume (mL)	% Hospital share (no. of centers)
100-150	6 (3)
151-200	28 (15)
201-250	21 (11)
251-300	28 (15)
301<	17 (9)

Priming solution. The most frequently used crystalloid priming solutions were Ringer's lactate solution (39% of centers), Ringer's acetate solution (22%), bicarbonated Ringer's solution (18%), bicarbonate replacement fluid (11%), normal saline (4%) and others (6%). For standard colloid priming solutions, usage was 25% albumin (56% of centers), 20% albumin (9%), 5% albumin (18.5%), fresh frozen plasma (2%), low molecular weight dextran (3.7%) and no usage (10.8%). The drugs most frequently added to the priming solution were heparin, mannitol, sodium bicarbonate, antibiotics, and steroids. All centers employed pre-bypass ultrafiltration for blood priming-28% of the centers used 500 mL and 25% used 1000 mL. Only 6% of the centers employed retrograde autologous priming, and 55% used priming solutions at 36-37°C.

Coated circuit. A coated circuit was used by 78% of the centers-49% used poly-2-methoxyethylacrylate coating and 29% used a heparin coating.

CPB components

Arterial pump. A roller pump was used as the arterial pump by 91% of the centers, a centrifugal pump by 9%.

Oxygenator. Table 2 shows hospital usage of oxygenators. The data include plural responses.

Table 2. Oxygenators used by hospitals during pediatric cardiac surgery in Japan, 2007-2009.

	Oxygenator	% Hospital share (no. of centers)
SORIN	D100	2 (1)
	D901	15 (8)
	D902	38 (20)
	D905	8 (4)
TERUMO	RX05	62 (33)
	RX15	21 (11)
	RX25	4 (2)
	FX05	26 (14)
	FX15	13 (7)
	FX25	4 (2)
MAQUET	Quadrox	2 (1)
JMS	Oxia-IC	38 (20)
	Oxia-LP	2 (1)
MERA	Excelung-kids	8 (4)
	HPO-05 RHFC	2 (1)
	HPO-06	2 (1)
NIPRO	Biocube 2000	8 (4)
	Biocube 4000	2 (1)
MEDOS	Hilite 2800	9 (5)

Table 3. Heart-lung machine used by hospitals during pediatric cardiac surgery in Japan, 2007-2009.

	Heart-lung machine	% Hospital share (no. of centers)
SORIN	S3	28.3 (15)
	S5	9.4 (5)
TERUMO	APS-1	5.7 (3)
MAQUET	HL-30	7.5(4)
Technowood	Compo3	17.0 (9)
MERA	HAS	15.1(8)
	AS-R	5.7 (3)
	HAS-2	1.9 (1)
Others (unknown)		9.4 (5)

Heart-lung machine. Table 3 shows hospital usage of a heart-lung machine.

CPB monitoring

Regional oxygen saturation. Regional oxygen saturation monitors such as INVOS (Somanetics, Troy, MI, USA) were used by 69% of the centers; 60% were set in the frontal lobe, 14% in both the frontal lobe and posterior regions.

In-line monitor. Thirty-four percent of the centers used in-line, continuous, arterial blood gas monitoring, 28% used in-line continuous venous blood gas monitoring, 23% used intermittent arterial blood gas monitoring, and 12% used intermittent venous blood gas monitoring and others (no response: 3%).

Electronic recording. Electronic recording was used by 51% of the centers.

CPB management

CPB flow. For routine normothermic neonatal CPB flow rate, 10% of the centers used 200 ml/kg/min, 12% used 180-200 ml/kg/min, 20% used 180 ml/kg/min, 10% used 150-180 ml/kg/min, 8% used 150 ml/kg/min, 4% used 120ml/kg/min, 8% used 80-120 ml/kg/min, 4% used 3.0 L/min/m^2, 4% used 2.8-3.0 L/min/m^2, 7% used 2.6-2.8 L/min/m^2, 2% used 2.5 L/min/m^2 and others (no response: 11%).

Temperature. For intraoperative temperature monitoring, 39% of centers used the nasopharynx, 86% the rectum, 33% the esophagus, 37% the bladder, 33% the skin, 15% a palmoplantar site, 80% the arterial blood of the circuit, and 78% the venous blood of the circuit. (These data include plural responses).

Filtration. During CPB, 48% of the centers used dilutional ultrafiltration and, at the end of CPB, 30% used modified ultrafiltration.

VAVD. Vacuum-assisted venous drainage was used by 39% of the centers.

Blood transfusion. The minimum acceptable hematocrit level during moderate hypothermia (28-30°C) was reported as 23.5±4.5% (range, 16%-35%), and the minimum acceptable level during deep hypothermia (18-22°C) was reported as 21.9±4.4% (range, 15%-30%).

Blood gas managements

Normothermia to mild hypothermia (34-36°C). Alpha-stat control was used by 88% of the centers, pH-stat control by 12% of the centers.

Moderate hypothermia (28-30°C). Alpha-stat control was used by 81% of the centers, pH-stat control by 19% of the centers.

Deep hypothermia (18-22°C). Alpha-stat control was used by 64% of the centers, pH-stat control by 36% of the centers.

Hypothermia technique (brain protection)

Hypothermia with circulatory arrest was the preferred perfusion technique for complex procedures on neonates in 24% of centers, while 37% of the centers reported also using deep hypothermia with isolated cerebral perfusion. Only one center used mild hypothermia without circulatory arrest, using isolated cerebral and rim perfusion techniques.

Myocardial Protection

Cardioplegia solution. Fifty-eight percent of the centers used crystalloid cardioplegia solution, 32% used blood cardioplegia solution. Ten percent used both crystalloid and blood cardioplegia solution. No center used warm blood cardioplegia.

Temperature. The infusion temperature of the cardioplegia solution was 0-5°C at 46% of the centers, 5-10°C at 24%, 10-15°C at 13%, 15-20°C at 7%, 20-28°C at 3% and others (no response: 7%).

ECMO

Procedures. The number of ECMO procedures performed was 98 in 2007, 109 in 2008, and 119 in 2009.

Pump. The centrifugal pump was used for ECMO by 58% of the centers, the roller pump by 30%, 6% of centers did not use ECMO, and from 6% there was no response.

Oxygenator. For oxygenation, 49% of the centers used the Biocube 2000 (Nipro, Osaka, Japan), 13% the Biocube 4000 (Nipro), 3% the Biocube 6000 (Nipro), 13% the SX-10 (Terumo), 2% the RX-05 (Terumo), 2% the FX-05 (Terumo) 2%, the HSO-05 (Mera, Senko Medical Instrument Mfg., Tokyo, Japan), 2% the excelung-kids (Mera), 2% the 6505R1 (Medtronic, Tokyo, Japan), 6% of centers did not use ECMO, and 6% did not respond.

Staff (perfusionists)

There were 288 pediatric perfusionists — 2 in 10% of the centers, 3 in 15%, 4 in 26%, 5 in 8%, and 6 in 13%. There were 100 certified clinical perfusionists — 1 in 25% of the centers, 2 in 25% of the centers, 3 in 12% of the centers, and none in 22% of the centers. Among the pediatric perfusionists, 49% were 20-30 years old, 33% were 31-40 years old, and 16% were 41-50 years old. Annual income was $25,000-$37,500 for 6% of the perfusionists, $37,501-$62,500 for 49%, $62,501-$87,500 for 27%, and $87,501-$112,500 for 8%.

Discussion

The surgical outcomes for congenital heart disease are influenced by not only surgical managements, but also CPB practices[6,8,9]. Several perfusion surveys have been published on different aspects[8,14]. Groom et al. regularly report pediatric perfusion surveys in North America[10,12-15]. Multicenter organizations such as the Multi-Societal Database Committee for Pediatric and Congenital Heart Disease and the International Consortium for Evidence-Based Perfusion can facilitate collaboration between healthcare providers interested in improving the quality of care delivered to congenital heart disease patients[5,6]. Although we are aware of the importance of evidenced-based "best practices" in perfusion and in initiating quality improvement in Japan[9], we have not yet established a formal community to manage a special database for pediatric perfusion. As an initial step to that end, we report here our first pediatric perfusion survey.

In over 70% of the centers, 2 perfusionists worked on each pediatric perfusion procedure, the number recommended for safety management and good clinical outcomes[16]. Moreover, more than 70% of the perfusionists were certified (20% were undergoing clinical training). The median number of CPB procedures performed per center per year in our study was 50, which is about 39% of the number performed in France (128 in 2005), and the mean number performed by each perfusionist in Japan (72% of perfusionists performed less than 50) was less than 1/3 the number performed in France (134 in 2005; range, 22-322)[17]. The difference may be due to Japan having many more congenital heart centers per population than France (1 per 200,000 vs. 1 per 400,000). Alternatively, the number of perfusionists per patient load may be greater in Japan. The low number of procedures performed by each perfusionist in Japan suggests that perfusionists may not gain enough practice to maintain their skills and that could engender a safety risk. For both patient safety and job security, it would be better to have a better balance of employment and caseload.

The use of oxygenators with an integrated arterial filter permits lower priming volumes, but only 1/4 of the Japanese centers used that setup (Table 2). We found that vacuum-assisted venous drainage was not a standard perfusion technique, probably because gaseous microemboli are associated with it and trouble-shooting is difficult[18]. Retrograde autologous priming was also not standard, probably because perfusionists want to avoid the complexity it involves at the start of the CPB. Since the Boston hematocrit trials suggested the potential advantage of a higher hematocrit level during hypothermic CPB[19-21], the trend in Japan has been to maintain hematocrit levels at over 25% during hypothermic CPB, especially during complex congenital heart surgery. Thus, most pediatric perfusionists avoid the non-transfusion CPB technique and vacuum-assisted venous drainage and retrograde autologous priming are not necessary for non-transfusion CPB techniques for young children. Hence, reducing the priming volume of the CPB circuit may not be important in the management of perfusion safety, especially in complex cases.

Concerning CPB components and circuits, the Terumo RX-05 oxygenator was popular because of its low priming volume, and the Sorin heart-lung machine was used predominantly. In General, products made in Japan were preferred, probably because of their easy maintenances and customizability.

We found that high flow management with alpha-stat blood gas control was standard during moderate to normothermic CPB. A North American survey taken in 2002, however, reported an increase in the use of pH-stat control[15], a technique that had become standard after 1993[16,22], but shifted to alpha-stat management to avoid deep hypothermic circulatory arrest in the presence of moderate to mild hypothermia. Related to that was the aggressive use of ultrafiltration methods such as prebypass, dilutional, and modified ultrafiltration, which may elevate the metabolic rate and increase collateral flow, to manage higher temperatures settings. Both high

flow management and ultrafiltration methods might help acid-base management.

Crystalloid cardioplegia was the predominant solution used for myocardial protection. In North America, 67% of centers used blood cardioplegia in 2002[15]. Future surveys will track how this changes.

In-line continuous blood gas monitoring, which is recommended by the Japanese Society of Extra-corporeal Technology, was used at over 50% of the centers and is increasing due to a keen awareness of perfusion safety management in Japan. In Australia and New Zealand, in contrast, only 5.2% of centers reported in-line blood gas monitoring routinely in 2006[9]. The technique is costly, so differences in usage may follow from differences in medical insurance systems (in-line monitoring is covered by medical insurance in Japan).

In conclusion, we have outlined the results of the first pediatric perfusion survey in Japan and compared some of the results with those of similar surveys conducted in other countries. Our survey lays the foundation for a Japanese pediatric perfusion database. Although we recognize the importance of "experience-based" practices to congenital heart surgery, we believe that data are also important and are necessary if we are to place perfusion on a sound scientific footing. We will update the survey at regular intervals so as to detail the progression of changes of pediatric perfusion practices in Japan.

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Conflict of Interest Statement

The authors declare that there is no conflict of interest.

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Dr Photiadis: Right.

Dr Gaynor: So what shunt, what operation would you do for that patient?

Dr Photiadis: Right now I think there is no data. Our data do not show it, and there are no other data, so we would still use the B-T shunt.

Dr. S. Sano *(Okayama, Japan)*: Yes, a Sano would be probably a reasonable alternative.

Dr Gaynor: That is the one finding that did come out in your studies.

Dr E. Bove *(Ann Arbor, MI, USA)*: I enjoyed your paper, and I agree with your conclusion, but I rise to support what Dr. Gaynor said, and I think that we have to be very careful how we interpret data. It is a retrospective study, and if I understood from your abstract, it was surgeon preference which was not very clear. Does that mean one surgeon did one technique, another surgeon did a different one so that each surgeon decided preoperatively what he felt would be the best shunt? Additionally, looking at postoperative risk factors and showing that the Aristotle scores are equal does not have the same validity as a prospective randomized trial to reach the conclusion as you have reached. So even though I would like to agree with your conclusion, I think we have to be very careful how we interpret these data.

Dr Photiadis: You are absolutely right, but again, randomisation may not give you the whole answer. If we have a patient, say aortic atresia, mitral atresia, who is stable preoperatively and whom you randomly assign to the Sano shunt, and then you have another patient with aortic atresia and mitral atresia, same anatomic type, was not prenatally diagnosed, decompensation without closure of the duct and cannot be stabilised, if you look on just the anatomic type, they are the same, but with respect to preoperative risk in fact they are not. By means of the Aristotle score these differences can be detected, with one patient being assigned to the low-risk and the other one to the high-risk group.

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EDITORIAL COMMENT

Re: Does the shunt type determine mid-term outcome after Norwood operation?

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There is always a debate as to which shunt is better in a Norwood procedure [1–4]. Some say the modified Blalock–Taussig (MBT) shunt is better than the right ventricle-pulmonary artery (RV-PA) shunt, while others say the RV-PA shunt is better than the MBT shunt. Photiadis *et al.* [1] compared their experience of MBT shunts with that of RV-PA shunts in 109 patients. They concluded that there was no significant difference in the survival rates between the two shunt types. However, the incidence of shunt-related interventions was significantly increased with the RV-PA (Sano) shunt group. Ohye *et al.* [2] reported the results of the first multicentre, randomized trial of the Norwood procedure with comparison of an RV-PA shunt and MBT shunt. They summarized that the 12-month transplantation-free survival was higher with the use of an RV-PA shunt than with the use of an MBT shunt. However, the RV-PA shunt was associated with a higher rate of unintended cardiovascular interventions and complications during the first 12 months after randomization. Their conclusion was that there was no significant difference between the two groups with respect to transplantation-free survival beyond 12 months.

The Norwood procedure using an MBT shunt creates pulmonary atresia from aortic atresia, while the idea of an RV-PA shunt is to create tetralogy of Fallot from aortic atresia. Postoperative management after the implantation of an MBT shunt in patients with pulmonary atresia was not easy because it is important to keep a balance between systemic and pulmonary circuits. Babies with an MBT shunt sometimes collapse suddenly in the ward or at home. However, almost no babies experience a sudden collapse in tetralogy of Fallot. While many surgeons and many centres report almost no or minimal mortality in patients with MBT shunt, and that it is easy to keep a balance between systemic and pulmonary circuits, the discharge mortalities in neonates after MBT shunts were 7.2 and 10.6%; in the Society of Thoracic Surgeons database [5] and in the European Society for Cardio-Thoracic Surgery database (http://www.eactscongenitaldb.org/index.php?LANG=en&level=1&struct=14), respectively. These data clearly show that the MBT shunt has a high mortality, and this is the reality of the situation.

Creating a tetralogy of Fallot instead of pulmonary atresia was the simple idea behind the RV-PA shunt when I started using the RV-PA shunt to treat the hypoplastic left heart syndrome. This was because postoperative management after the implantation of a MBT shunt was not easy.

We all worry about the adverse effects on RV function after a ventriculotomy; however, there are many papers that have demonstrated no adverse effects in the use of a RV-PA shunt [3, 6]. The site and size of the ventriculotomy as well as the size of the shunt are important. Initially, we created an RV hole by

using a knife and a pair of scissors and this was changed to a coronary puncher to create uniform and minimally sized holes. Furthermore, pulmonary blood flow is controlled using a clip on the graft with oxygen saturation staying at around 80–85%; on room air. These changes may affect long-term RV function. The authors and others also alluded to a higher rate of unintended cardiovascular interventions and complications with the use of the RV-PA shunt. A ring-enforced polytetrafluoroethylene (PTFE) graft is the option to avoid proximal graft stenosis [7]. Recently, we have begun to use ring-enforced PTFE grafts frequently instead of a ringless graft. By adopting these modifications, we may be able to decrease the mortality and morbidity after a RV-PA shunt.

All available data show no difference in mortality between the two different shunts. Some surgeons prefer to do an MBT shunt and some prefer to do an RV-PA shunt. Whichever shunt is used, more efforts should be made to improve mortality and morbidity.

Conflict of interest: none declared.

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Extracorporeal membrane oxygenation following pediatric cardiac surgery: development and outcomes from a single-center experience

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Abstract

Extracorporeal membrane oxygenation (ECMO) has emerged as an effective mechanical support following cardiac surgery with respiratory and cardiac failure. However, there are no clear indications for ECMO use after pediatric cardiac surgery. We retrospectively reviewed medical records of 76 pediatric patients [mean age, 10.8 months (0–86); mean weight, 5.16 kg (1.16–16.5)] with congenital heart disease who received ECMO following cardiac surgery between January 1997 and October 2010. Forty-five patients were treated with an aggressive ECMO approach (aggressive ECMO group, April 2005–October 2010) and 31 with a delayed ECMO approach (delayed ECMO group, January 1997–March 2005). Demographics, diagnosis, operative variables, ECMO indication, and duration of survivors and non-survivors were compared. Thirty-four patients (75.5%) were successfully weaned from ECMO in the aggressive ECMO group and 26 (57.7%) were discharged. Conversely, eight patients (25.8%) were successfully weaned from ECMO in the delayed ECMO group and two (6.5%) were discharged. Forty-five patients with shunted single ventricle physiology (aggressive: 29 patients, delayed: 16 patients) received ECMO, but only 15 (33.3%) survived and were discharged. The survival rate of the aggressive ECMO group was significantly better when compared with the delayed ECMO group ($p<0.01$). Also, ECMO duration was significantly shorter among the aggressive ECMO group survivors (96.5 ± 62.9 h, $p<0.01$). Thus, the aggressive ECMO approach is a superior strategy compared to the delayed ECMO approach in pediatric cardiac patients. The aggressive ECMO approach improved our outcomes of neonatal and pediatric ECMO.

Keywords

extracorporeal membrane oxygenation; congenital heart disease; cardiac surgery; pediatric; hypoplastic left heart syndrome

Introduction

Extracorporeal membrane oxygenation (ECMO) has emerged as an effective mechanical support following cardiac surgery with respiratory and cardiac failure. In 1976, Bartlett et al.[1] reported the successful use of ECMO for a neonatal patient with respiratory failure and, since then, ECMO has been used effectively for a variety of indications, including preoperative hemodynamic support, low cardiac output after cardiopulmonary bypass (CPB), sudden cardiac arrest, and as a bridge to heart transplantation[2]. The Extracorporeal Life Support Organization (ELSO) Registry reports that the rates of survival off ECMO and survival to discharge among neonatal cardiac patients are 59% and 39%, respectively; in pediatric patients, these values are 62% and 46%, respectively. The rates of survival off ECMO and to discharge among neonates subject to extracorporeal cardiopulmonary resuscitation are 63% and 37% and, in pediatric patients, these rates are 52% and 38%, respectively[3]. Outcomes of ECMO have been developing and still have been keeping the improvement scope for better outcomes, especially in patients with congenital heart disease.

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In our institute, we did not have any criteria for ECMO introduction until March 2005, but we changed our strategy of ECMO therapy and introduced an aggressive approach toward indications for ECMO from April 2005 wherein we did not hesitate to introduce ECMO. Before we had changed our strategy of ECMO therapy in April 2005, we had felt the negative image of ECMO because of our poor results of survival off ECMO in patients with congenital heart disease.

The purpose of this study was to evaluate the effects of this aggressive approach of ECMO initiation since we changed our strategy compared with the previously followed delayed approach for introducing ECMO after pediatric cardiac surgery, by reviewing our single center experiences.

Materials and methods

We retrospectively reviewed medical records of 76 pediatric patients with congenital heart disease who received ECMO following cardiac surgery between January 1997 and October 2010 at Okayama University Hospital, Japan.

Two groups were compared: those who were treated with the aggressive ECMO approach (45 patients from April 2005 to October 2010) and those who were treated with the delayed ECMO approach (31 patients from January 1997 to March 2005). The aggressive ECMO approach was defined as commencement of ECMO as early as possible before end-organ dysfunction or complete circulatory collapse, and without hesitation for the introduction of ECMO. The outcomes were categorized as follows: survived off ECMO (successful weaning from ECMO support), survival to hospital discharge, and survival during outpatient follow-up. Demographics, diagnosis, operative variables, and ECMO indication and duration of survivors and non-survivors were compared. We compared the lactate levels of both groups before and 2 hours after the introduction of ECMO.

We defined the indications for aggressive ECMO as follows: mean arterial blood pressure less than 35 mmHg, anuria, high lactate level (greater than 5.0 mmol/L) and acidosis with a pH less than 7.3, hypoxia with arterial oxygen saturation less than 60% with 100% FiO_2 (fraction of inspired oxygen) on mechanical ventilatory support. ECMO was established via a median sternotomy and cannulation of the ascending aorta or inominate artery for return, and drainage via a 3-mm polytetrafluoroethylene graft to the right atrium (in cases after a Norwood stage 1 procedure). For patients with a right ventricle (RV) to pulmonary artery (PA) shunt after a Norwood stage 1 procedure in hypoplastic left heart syndrome, we opened the RV-PA shunt and controlled the pulmonary blood flow by the degree of clipping the shunt.

Table 1. The demographic data of aggressive and delayed ECMO groups

Aggressive ECMO(n=45)			
Biventricle (n=16)		Single ventricle (n=29)	
Critical AS	4	HLHS	9
DORV	3	HLHS variant	7
TOF	3	SA/SV	5
PA/VSD	2	TA	3
TAPVC	2	DILV	2
CoA/VSD	1	PA/IVS	1
BEG syndrome	1	IAA	1
		Congenital MS	1
Delayed ECMO (n=31)			
Biventricle (n=15)		Single ventricle (n=16)	
DORV	3	HLHS	5
PA/VSD	3	HLHS variant	3
AVSD	2	PA/IVS	3
TOF	2	DILV	2
TAPVC	1	SA/SV	2
TGA	1	TA	1
Critical AS	1		
PTA	1		
Myocarditis	1		

AS, aortic stenosis; DORV, double-outlet right ventricle; TOF, tetralogy of Fallot; PA, pulmonary atresia; VSD, ventricular septal defect; TAPVC, total anomalous pulmonary venous connection; CoA, coarctation of the aorta; BWG, Bland-White-Garland; HLHS, hypoplastic left heart syndrome; SA, single atrium; SV, single ventricle; TA, tricuspid atresia; DILV, double-inlet left ventricle; PA, pulmonary atresia; IVS, intact ventricular septum; IAA, interrupted aortic arch; MS, mitral stenosis; AVSD, atrioventricular septal defect; TGA, transposition of the great arteries; PTA, persistent truncus arteriosus

We used a hollow-fiber membrane oxygenator (Biocube®; Nipro, Osaka, Japan) and a centrifugal pump (Gyro®; Kyocera, Kyoto, Japan) with a 6-mm heparin-coated tube (Biomate®; Toyobo, Osaka, Japan) for the ECMO device. The initial ECMO flow rate was set at 150–180 ml/kg/min. Anticoagulation was accomplished by drip infusion of sodium heparin to maintain an activated clotting time of 150–200 seconds. The hemoglobin concentration was maintained above 10 g/dL by transfusion of packed red blood cells during full support. We maintained the bladder temperature at 35°C and applied minimal ventilatory support during ECMO. The intrathoracic cavity was irrigated with sterile, warmed, normal saline every 3 days.

We tried to wean the patient off ECMO when the lactate level was less than 2.0 mmol/L, the urine output had increased to equal or greater than 1.0 ml/kg/h for as long as 24 h, and the arterial pulse pressure had increased to equal or greater than 10 mmHg. We started inotropic agents for 6 h and gradually reduced the ECMO flow rate