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Three-dimensional-printed heart model can determine univentricular repair strategy in borderline double-outlet right ventricle

Yuzo Katayama[1*], Sho Isobe[1], Tsukasa Ozawa[1] and Takeshiro Fujii[1]

Abstract

Background As *double-outlet right ventricle* has a wide pathophysiology spectrum, its comprehensive treatment strategy is determined based on relevant factors, such as the location and size of the ventricular septal defect, ventricular volume, and relationship of the great arteries. However, for borderline *double-outlet right ventricle* cases, it is occasionally difficult to decide the treatment strategy preoperatively. Recently, advances in 3D printing technology based on computed tomography have enabled the creation of 3D heart models of congenital heart disease that can precisely reproduce the anatomical structure of each patient even for complex anomalies. Herein, we describe a young patient in whom univentricular repair could be decided after confirming the 3D heart model and intracardiac structure under direct vision.

Case presentation We describe a 3-year-old girl who was diagnosed with *double-outlet right ventricle* and severe pulmonary valve stenosis at birth and who underwent a left modified Blalock–Taussig shunt at 2 years of age. Preoperative examination revealed a borderline condition for biventricular repair characterized by a small left ventricle volume and side-by-side relationship of the great artery. After a preoperative discussion using a 3D heart model, we concurred that an intraoperative assessment would be made as to whether biventricular repair was possible or not. After confirming the intracardiac structure under direct vision, we assessed that intraventricular rerouting was not possible owing to the high risk of subvalvular aortic stenosis as there was no tissue that could be incised between the right ventricular free wall and the primary interventricular foramen, as indicated in the 3D heart model. Thus, atrial septostomy and Glenn anastomosis were performed.

Conclusions We report a 3-year-old girl with a borderline *double-outlet right ventricle* in whom a univentricular repair strategy could be decided after confirming the 3D heart model and intracardiac structure under direct vision. A 3D-printed heart model can be useful in patients whose repair strategy is difficult to judge for the borderline double-outlet right ventricle.

Keywords Double-outlet right ventricle, 3D printed heart model, Univentricular repair

Background

As *double-outlet right ventricle* (DORV) has a wide pathophysiology spectrum, a comprehensive treatment strategy is determined based on important factors, such as the location and size of the ventricular septal defect, ventricular volume, and the relationship of the great arteries. However, for borderline DORV cases, it is occasionally difficult to decide the treatment strategy preoperatively.

*Correspondence:
Yuzo Katayama
yuuzou.katayama@med.toho-u.ac.jp
[1] Division of Cardiovascular Surgery, Department of Surgery, Toho University Omori Medical Center, 6-11 Omori-Nishi Ota-Ku, Tokyo 143-8541, Japan

Recently, advances in 3D printing technology based on computed tomography (CT) have enabled the creation of 3D heart models of congenital heart disease that can precisely reproduce the anatomical structure of each patient even for complex anomalies.

Herein, we describe a young patient in whom a univentricular repair (UVR) strategy could be decided after confirming the 3D heart model and intracardiac structure under direct vision.

Case presentation

The patient was a 3-year-old girl who was diagnosed with DORV and severe pulmonary valve stenosis at birth and who was followed up with home oxygen therapy with a view of biventricular repair (BVR). At 2 years of age, she underwent urgent left modified Blalock–Taussig shunt owing to the development of severe hypoxia.

Echocardiography revealed severe overriding of the aorta and subaortic ventricular septal defect (Fig. 1a). Catheterization and 3D-CT revealed a side-by-side relationship between the ascending aorta and the pulmonary trunk (Fig. 1b). As there was a concern for the development of left ventricular outflow stenosis (LVOTS) after intraventricular rerouting, we used the "CARDIO Simulator® (cross Medical, Inc., Japan)" as a 3D heart model, and we had a preoperative discussion on whether intraventricular rerouting is possible. The 3D heart model showed that the primary interventricular foramen (PIF) is of the subaortic type and is small, and therefore requires enlargement for intraventricular rerouting. However, it appeared that achieving enough enlargement of the PIF while avoiding LVOTS was not possible because the anterior free wall of the right ventricle was close to the PIF and that the route from the left ventricle to the aorta via the PIF was sharply curved.

The 3D heart model also showed the presence of a long lesion of right ventricular outflow tract stenosis in addition to the pulmonary valve stenosis (Supplementary video). For BVR, it was necessary to perform transannular repair with a long incision of the right ventricle. These findings from the 3D heart model suggested that BVR should be avoided.

The operation was planned so that we could make an intraoperative assessment under direct vision of whether BVR was possible or not. After the establishment of cardiopulmonary bypass and cardiac arrest, right atrial incision was performed and the heart was observed. As the intracardiac lesion findings were very similar to those of the 3D heart model (Fig. 2), enlargement of the atrial septal defect and right atrial closure were performed. Glenn anastomosis was additionally performed after declamping of the aorta. The patient also underwent completion of the Fontan procedure.

Discussion

As DORV presents with anatomical and hemodynamic variations, surgical management varies and it is necessary to choose the optimal strategy based on individual cases. BVR is recommended in cases wherein both the right and left ventricles are balanced and well-functioning, as Fontan circulation may not be safe and stable in the long term. However, in cases of DORV involving a complex anatomy, it is difficult to select the optimal surgical strategy.

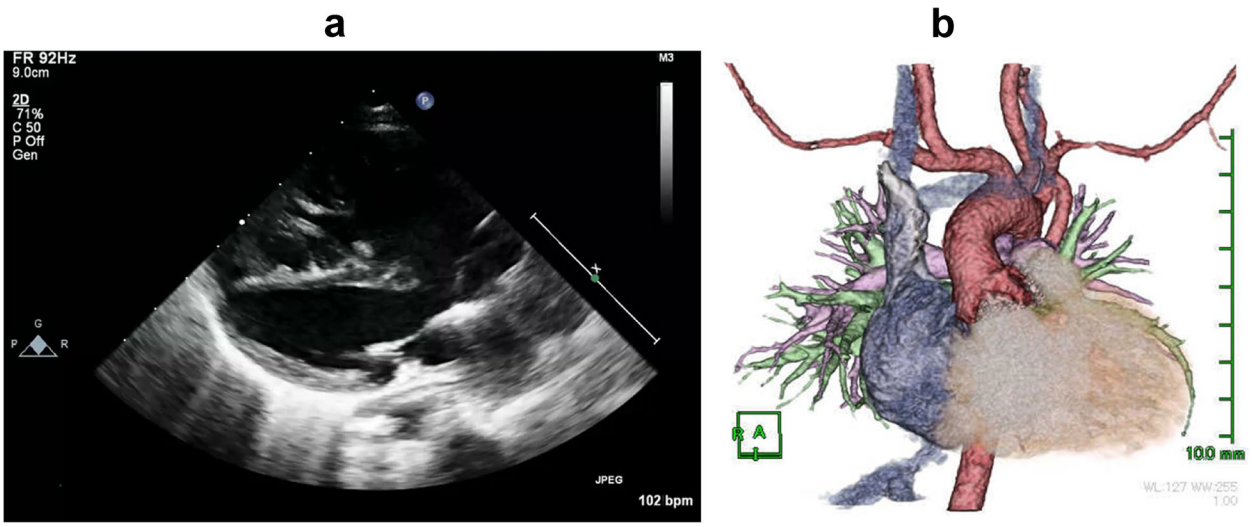

Fig. 1 **a** Transthoracic echocardiography revealed severe overriding of the aorta and subaortic ventricular septal defect. **b** 3D-CT revealed a side-by-side relationship between the ascending aorta and the pulmonary trunk

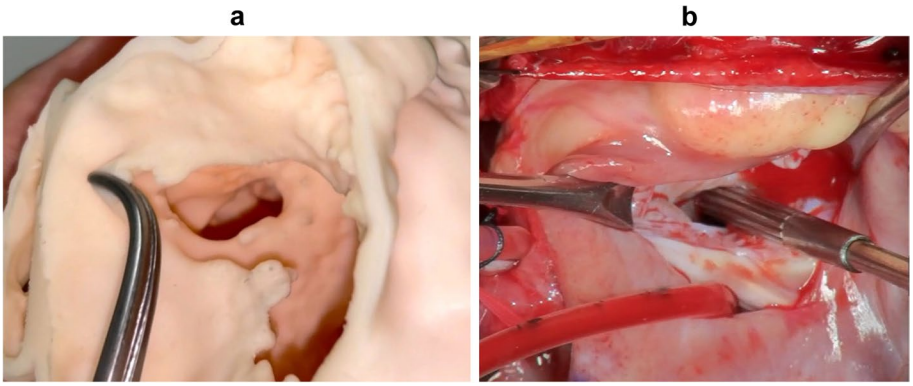

Fig. 2 Primary interventricular foramen in **a** 3D printed heart model and **b** intraoperative findings

Bradley et al. [1] reported that BVR, particularly Rastelli-type reconstruction, is associated with higher late mortality and reintervention than Fontan repair. On the other hand, Oladunjoye et al. [2] reported that BVR for DORV with complex anatomy resulted in acceptable midterm outcomes with low mortality. However, freedom from surgical reintervention for LVOTS was low after BVR.

Thus, in patients having DORV with a complex anatomy, it is required to thoroughly understand the precise intracardiac structures and carefully consider an operative plan with preferable consequences and fewer complications. A 3D-printed heart model precisely replicates the anatomical structure of congenital heart disease and enables an easier preoperative evaluation. Shiraishi et al. [3] created a flexible stereolithographic biomodel using rubber-like urethane which enabled preoperative simulation of individual surgical operations. Furthermore, they developed superflexible polyurethane heart models similar to our model in the present case which have realistic flexibility that enables retraction, incision, and suturing for preoperative simulation. Hoashi et al. [4] created 3D-printed heart models for preoperative surgical simulation in 20 patients including those with DORV, and reported that 3D-printed heart models are useful for understanding the relationship between intraventricular communications and great vessels and for simulations to create intracardiac pathways. Zhao et al. [5] reported that surgical planning based on 3D printing could decrease bypass and cross-clamp times during surgery.

In the present case, the use of a 3D-printed heart model facilitated the decision-making of a surgical plan and the performance of UVR that avoids complications after BVR. A 3D-printed heart model can be a useful alternative in patients whose repair strategy (i.e., BVR or UVR) is difficult to judge similar to our case.

Conclusions

We report a 3-year-old girl with a borderline *double-outlet right ventricle* in whom a univentricular repair strategy could be decided after confirming the 3D heart model and intracardiac structure under direct vision. A 3D-printed heart model can be useful in patients whose repair strategy is difficult to judge for the borderline double-outlet right ventricle.

Abbreviations

DORV	Double-outlet right ventricle
UVR	Univentricular repair
BVR	Biventricular repair
LVOTS	Left ventricular outflow stenosis
PIF	Primary interventricular foramen

Supplementary Information

The online version contains supplementary material available at https://doi.org/10.1186/s43054-023-00229-z.

Additional file 1: Supplementary video. A preoperative simulation was performed using a 3D printed heart model based on multislice computed tomography created by CARDIO Simulator® (cross Medical, Inc., Japan).

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Authors' contributions

YK drafted the original manuscript. SI, TO, and TF substantially contributed to the revisions of the manuscript drafts. All authors have approved the submitted version of the manuscript and agree to be accountable for any part of the work. All the authors have read and approved the final manuscript.

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Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

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