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CASE REPORT

One-and-a-Half Ventricle Repair with Hemi-Mustard/Bidirectional Glenn/Rastelli Procedure in a Patient with Double Outlet Right Ventricle and Atrioventricular Discordance: A Case Report

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ABSTRACT: Background Double outlet right ventricle (DORV) is a complex congenital heart disease with significant anatomical variations. Patients with atrioventricular discordance have traditionally been managed with univentricular palliation strategies. However, advances in surgical concepts and techniques have enabled biventricular or one-and-a-half ventricle repair in selected patients, even after prior palliation. Case presentation We report a case of a 32-year-old woman with DORV and atrioventricular discordance who successfully underwent conversion to a one-and-a-half ventricle circulation using a combined Hemi-Mustard/Rastelli procedure with preservation of a pre-existing bidirectional Glenn shunt, performed 18 years after initial palliation. Conclusion This case provides insight into the surgical decision-making and technical considerations for managing similarly complex congenital anomalies.

KEYWORDS: Hemi-mustard/bidirectional Glenn/Rastelli procedure; double outlet right ventricle with atrioventricular discordance; post-palliation; one-and-a-half ventricle repair

1 Case Presentation

A 32-year-old woman (height 155 cm, weight 50.2 kg) was admitted to our hospital for evaluation of progressive cyanosis and reduced exercise tolerance. At 14 years of age, she had undergone a bidirectional Glenn shunt, pulmonary artery banding, and atrial septal defect (ASD) enlargement at our center for dextrocardia, atrial situs solitus, ventricular inversion, DORV, ASD, and pulmonary stenosis (PS). Her postoperative course had been uneventful; however, she was lost to long-term follow-up. Six months before the current admission, she developed cyanosis and exercise intolerance. Transthoracic echocardiography demonstrated a patent bidirectional Glenn shunt, dilatation of the superior vena cava (SVC), a non-restrictive remote VSD, a non-restrictive ASD, and a pressure gradient of 41 mmHg across the pulmonary artery band. She was diagnosed with DORV with atrioventricular discordance (AVD) and had undergone palloative surgery 18 years earlier. Her medical history included a therapeutic abortion 6 months earlier due to severe hypoxemia during pregnancy (SpO_2 78%).

2 Preoperative Evaluation

On admission, the patient was in New York Heart Association (NYHA) functional class IV, with a resting peripheral oxygen saturation of 84%. Cardiopulmonary exercise testing indicated an intermediate risk for

heart failure and a high risk for myocardial ischemia, with poor exercise tolerance. Detailed anatomical assessment was performed using cardiac computed tomography (CT) and a patient-specific 3D-printed heart model (Figs. 1 and 2). Cardiac magnetic resonance imaging demonstrated preserved biventricular systolic function, with a left ventricular ejection fraction (LVEF) of 64.16% and a right ventricular ejection fraction (RVEF) of 64.1%, as well as relatively balanced ventricular volumes (LVEDVi 48.02 mL/m^2; RVEDVi 56.77 mL/m^2), Cardiac catheterization revealed balanced intracardiac pressures, including a SVC pressure of 11 mmHg and a main pulmonary artery (MPA) pressure of 10 mmHg, with a pulmonary-to-systemic blood flow ratio (Qp:Qs) of 0.68.

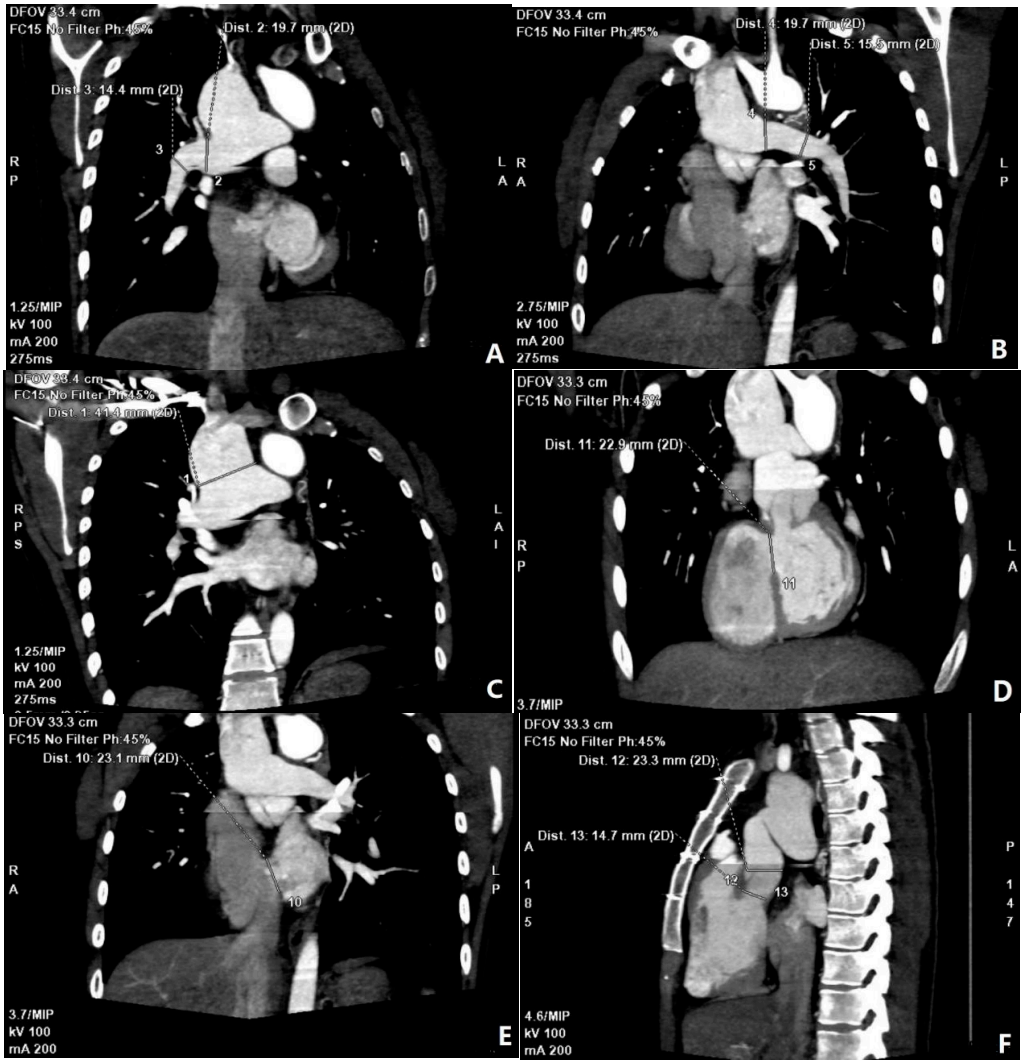

Figure 1: Preoperative contrast-enhanced CT images. (**A**) Proximal right pulmonary artery diameter: 19.7 mm, distal: 14.4 mm; (**B**) Proximal left pulmonary artery diameter: 19.7 mm, distal: 15.5 mm; (**C**) Dilated superior vena cava; diameter of the right-sided cavopulmonary anastomosis: 41.4 mm; (**D**) Diameter of the ventricular septal defect: 22.9 mm; (**E**) Diameter of the atrial septal defect: 23.1 mm; (**F**) Both the aorta and pulmonary artery originate from the right ventricle; Subpulmonary stenosis with a diameter of 14.7 mm; significant stenosis at the distal main pulmonary artery banding site with a diameter of 6.1 mm.

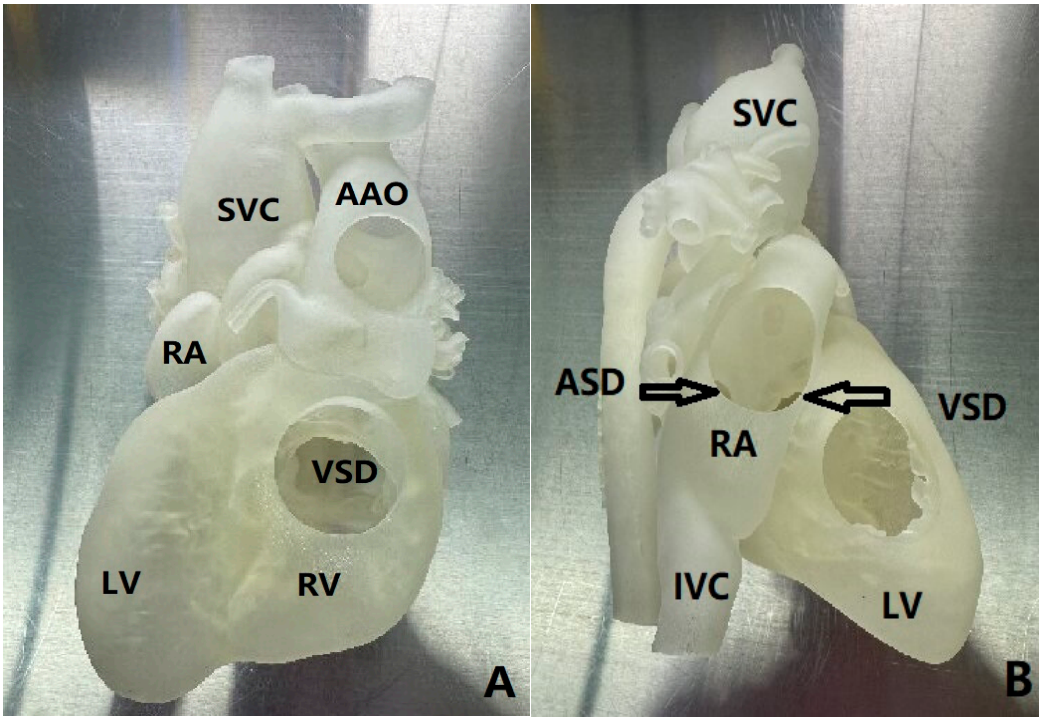

Figure 2: 3D-printed heart model. (**A**) coronal view. (**B**) sagittal view. The superior vena cava is markedly dilated with a patent anastomosis to the right pulmonary artery. The right atrium connects to the left ventricle, the left atrium connects to the right ventricle. Both the aorta and pulmonary artery arise from the right ventricle, and the ventricular septal defect is non-restrictive and remote from the great arteries. SVC: Superior vena cava; AAO: Ascending aorta; RA: Right atrium; LV: Left ventricle; RV: Right ventricle; VSD: Ventricular septal defect; ASD: Atrial septal defect; IVC: Inferior vena cava.

Following multidisciplinary team (MDT) discussion and surgical simulation using the 3D model, a one-and-a-half ventricle repair was selected. The planned strategy consisted of a combined Hemi-Mustard and Rastelli procedure, with preservation of the existing bidirectional Glenn shunt. Conversion to a 1.5-ventricle circulation was favored over biventricular repair for several reasons: (1) marked SVC dilation (>40 mm), which would complicate reanastomosis; (2) increased risk of conduction system injury associated with extensive intracardiac manipulation; (3) concern regarding excessive right-sided volume loading and potential exacerbation of tricuspid regurgitation following restoration of full venous return; and (4) institutional experience indicating advantages of 1.5-ventricle repair in terms of reduced operative duation, shorter mechanical ventilation, and fewer perioperative complications.

3 Surgical Technique

The operation was performed via median sternotomy under standard cardiopulmonary bypass. The key operative steps were as follows:

(1) **Intra-atrial Baffle Construction**

A right atriotomy was made parallel to the atrioventricular groove. The ASD was enlarged toward the inferior vena cava (IVC), and the coronary sinus was unroofed. A pericardial patch was used to construct an intra-atrial baffle directing IVC flow across the ASD to the tricuspid valve, which served as the inlet to the right ventricle, separating systemic and pulmonary venous return.

(2) **VSD Enlargement and Left Ventricle-to-Aorta Intraventricular Tunnel**

Through an infundibular incision in the right ventricle, the obstructing conal muscle was resected to optimize the left ventricular outflow tract (LVOT). The tricuspid valve chordae attached to the resected muscle were carefully detached and preserved. A second pericardial patch was used to create an intraventricular tunnel from the VSD to the aortic valve, forming a smooth, non-obstructive left ventricle-to-aorta pathway. The detached chordae were subsequently reattached.

(3) **Glenn shunt preservation and Right Ventricular Outflow Tract Reconstruction**

The MPA was transected at its root, and the proximal stump was oversewn while preserving the bidirectional Glenn shunt. Right ventricular outflow tract (RVOT) reconstruction was completed using a 17 mm valved bovine jugular vein conduit connecting the morphologic right ventricle to the distal pulmonary artery.

The cardiopulmonary bypass time was 445 min, and the aortic cross-clamp time was 220 min. The heart resumed spontaneous beating, and the electrocardiogram showed sinus rhythm. Intraoperative transesophageal echocardiography confirmed satisfactory hemodynamics, including pulmonary venous inflow velocity of 0.6 m/s, IVC baffle flow velocity of 1.3 m/s, LVOT velocity of 0.8 m/s, right ventricular outflow tract (RVOT) conduit velocity of 1.0 m/s, and pulmonary conduit velocity of 1.8 m/s, with only mild valvular regurgitation.

4 Postoperative Course and Follow-Up

The patient was extubated on postoperative day 3 and transferred from the intensive care unit on day 7 without rhythm-related and other major complications. She was discharged after 3 weeks due to excessive postoperative chest drainage. At the 2-month follow-up, she had improved to NYHA functional class II, with oxygen saturation of 98%. Echocardiography demonstrated a patent Glenn shunt (velocity 0.47 m/s), IVC baffle velocity of 1.66 m/s, pulmonary conduit velocity of 1.8 m/s, mild valvular regurgitation, and LVEF of 68.1%. Follow-up cardiac CT confirmed excellent anatomical repair without residual obstruction or shunting (Fig. 3).

5 Discussion

DORV with AVD is a rare and highly complex congenital anomaly, accounting for approximately 10% of all DORV cases [1,2]. As in congenitally corrected transposition of the great arteries (ccTGA), definitive surgical intervention is required. Initial palliation, such as systemic-to-pulmonary shunting or pulmonary artery banding, is often performed early in life (<6 months) to improve oxygenation and promote pulmonary vascular development [3].

In this patient, initial palliation at 14 years of age was chosen due to unfavorable factors for primary biventricular repair, including advanced age, elevated pulmonary vascular resistance, a remote VSD, and a restrictive ASD. This strategy aimed to stabilize circulation, reduce ventricular volume load, and preserve pulmonary vascular integrity.

Eighteen years later, reassessment revealed favorable anatomy and physiology, including a patent Glenn shunt, adequate pulmonary artery development (McGoon ratio 1.78), non-restrictive intracardiac shunts, balanced ventricular volumes, preserved systolic function, and normal intracardiac pressures. Although Fontan completion was a conventional option, its known long-term complications, including heart failure, arrhythmias, and protein-losing enteropathy, prompted consideration of alternatives [4]. Advances in surgical techniques and 3D modeling have enabled conversion from univentricular to biventricular or

1.5-ventricle circulation in selected patients [5]. Given the patient's age, desire for future pregnancy, and long-term quality-of-life considerations, a one-and-a-half ventricle repair was selected.

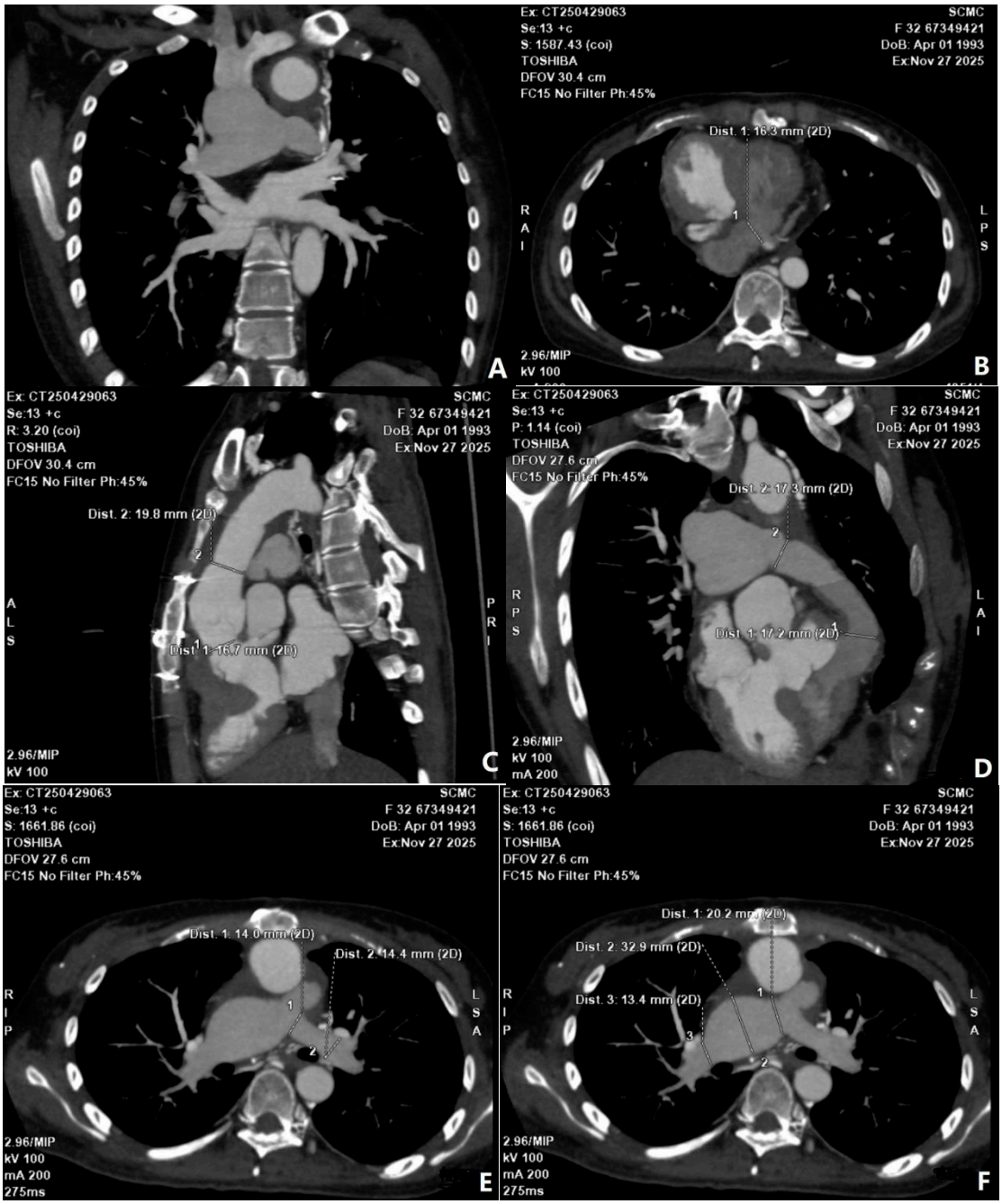

Figure 3: Cardiac CT images at 2-month postoperative follow-up. (**A**) PV draining into the right atrium without stenosis, cavopulmonary anastomosis 38.5 mm; (**B**) Inferior vena cava-left atrial junction 16.3 mm; (**C**) LVOT 16.7 mm, AAO 19.8 mm; (**D**) RVOT conduit patent, proximal 17.2 mm, distal 17.3 mm; (**E**) LPA proximal 14.0 mm, distal 14.4 mm; (**F**) RPA proximal 20.2 mm, distal 13.4 mm.

The combined Hemi-Mustard/Bidirectional Glenn/Rastelli procedure, initially described by the Baylor group for complex ccTGA [6], offers a flexible solution for patients with dextrocardia, ventricular

dysfunction, tricuspid regurgitation, or pulmonary stenosis [7–9]. Compared with Fontan completion or conventional biventricular repair, the 1.5-ventricle approach (Hemi-Mustard/Glenn/Rastelli) offers several advantages: it reduces preload on the systemic right ventricle, shields the tricuspid valve from systemic pressures, is technically less demanding than a complete atrial switch, and permits use of a smaller right ventricle-to-pulmonary artery conduit with potentially improved durability [10]. Consistent with these benefits, our institutional experience with 90 ccTGA patients demonstrated that 1.5-ventricle repair was associated with superior postoperative survival, lower complication rates, and higher freedom from reoperation compared with both double-switch and Fontan procedures [11,12].

In this case, preserving the established Glenn shunt avoided a technically challenging takedown, limited systemic venous return to the systemic ventricle, reduced stress on the systemic tricuspid valve, simplified atrial baffling, and allowed use of a smaller subpulmonary conduit, potentially reducing the need for future replacement.

6 Conclusion

One-and-a-half ventricle repair using a combined Hemi-Mustard/Bidirectional Glenn/Rastelli approach is a viable and effective option for selected patients with complex congenital heart disease. This case demonstrates that late conversion following prior univentricular palliation is feasible in DORV with AVD, offering an alternative strategy with potential long-term benefits. Further studies with larger cohorts and extended follow-up are required to confirm durability and quality-of-life outcomes.

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Author Contributions: Kai Luo, Hao Zhangand and Xiaomin He were the major contributors in writing and revising the manuscript. Jinghao Zheng and Yanjun Pan identified the worthiness of the case and drafted the manuscript. Kai Luo and Zhongqun Zhu collected the patient information and did follow-up work. Hao Zhang provided funding support for this work. All authors reviewed and approved the final version of the manuscript.

Availability of Data and Materials: Not applicable.

Ethics Approval: This case has been filled by The Ethics Committee of Shanghai Children's Medical Center and confirmed that formal approval was not required as it describes a single anonymized patient (proof of exemption available upon request). The study was conducted in accordance with the principles of the Declaration of Helsinki. Written informed consent for publication of clinical details and images was obtained from the patient.

Conflicts of Interest: The authors declare no conflicts of interest.

Abbreviations

DORV	double-outlet right ventricle
AVD	atrioventricular discordance
ASD	atrial septal defect
PS	pulmonary valve stenosis
SVC	superior vena cava
VSD	ventricular septal defect
NYHA	New York Heart Association
CT	computed tomography

MRI	magnetic resonance imaging
LVEF	left ventricular ejection fraction
LVEDVi	left ventricular end-diastolic volume index
RVEF	right ventricular ejection fraction
RVEDVi	right ventricular end-diastolic volume index
IVC	inferior vena cava
AAO	ascending aorta
RA	right atrium
LV	left ventricle
RV	right ventricle
MPA	main pulmonary artery
RVOT	right ventricular outflow tract
LVOT	left ventricular outflow tract
ccTGA	congenitally corrected transposition of the great arteries

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