Neonatal repair of atypical double outlet right ventricle

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Abstract

Primary repair was carried out in a neonate with an atypical form of double outlet right ventricle; with a non-committed ventricular septal defect and lack of the outlet septum between the semilunar valves. The aortic arch was right-sided. The procedure required a right ventricular incision. Intraventricular rerouting could be achieved concomitantly with the arterial switch maneuver. Retrospectively, several strategies were contemplated to seek whether any other approach could have been superior to our present choice.

Case report

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Key words:

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Abstract

Primary repair was carried out in a neonate with an atypical form of double outlet right ventricle; with a non-committed ventricular septal defect and lack of the outlet septum between the semilunar valves. The aortic arch was right-sided. The procedure required a right ventricular incision. Intraventricular rerouting could be achieved concomitantly with the arterial switch maneuver. Retrospectively, several strategies were contemplated to seek whether any other approach could have been superior to our present choice.

Ethical Standards

The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human medical regulations and with the Helsinki Declaration of 1975, as revised in 2008. An individual ethical approval for this study was waived according to the policy (case report in an anonymous style) of the Nara Medical University review board.

Introduction

A newborn posed us multiple issues when repairing an atypical form of double outlet right ventricle (DORV). Primary repair turned out to be successful. We retrospectively contemplated what could have been done differently.

Case Report

A fetus was diagnosed as having DORV with subpulmonary ventricular septal defect (VSD). The baby was born by the Caesarean section (gestational age 38 weeks and 1day, body weight 3.4 kg, female). Her oxygen saturation was 85%. The arterial duct was maintained on Prostaglandin. Balloon atrio-septostomy was not carried out. Left and right ventriculography exclusively enhanced streaming to the pulmonary trunk and the aorta, respectively. On computed tomography (CT), the aortic valve was right-anterior to the pulmonary valve. The right ventricular (RV) outflow tract was widely patent to either of the semilunar valves. The aortic arch was right-sided. We judged that neonatal primary repair was sensible switching the great arteries.

Surgery was carried out at 17 days old. The architecture of the RV could not be visualized via the tricuspid valve, but identified through a RV incision. The anterior limb of the trabeculo-septo-marginalis sit between the pulmonary valve and the rim of the interventricular communication (typical for DORV with a non-committed VSD). The outlet septum was entirely lacking; the aortic and the pulmonary valves possessed equivalent diameters and fibrous continuity without offsetting (a feature of DORV with a doubly-committed VSD) [1].

The VSD was enlarged antero-superiorly by the wedge resection of the septal muscle. Interventricular rerouting was achieved, from the left ventricle (LV) to the morphologically pulmonary valve, using an oval polytetrafluoroethylene patch [Figure 1]. Particular attention was paid when putting stitches to the fibrous tissues between the semilunar valves; no pledgets were placed so as not to interfere motion of the leaflets. The arterial switch was completed in a standard way with the Lecompte maneuver. Placing the arterial cannula high and mobilizing the ascending aorta extensively, right aortic arch did not matter the arterial switch.

Postoperative CT showed excellent results a month after repair. Transthoracic echocardiography demonstrated widely patent coronary arteries. Flow was not accelerated across the intraventricular tunnel or the pathway from the RV to the neo-pulmonary trunk. No residual shunts. No tricuspid valvar impediments thus far.

Comment

The initial diagnosis was given as “false Taussig Bing” malformation on fetal echo examination as a shorthand; we had been aware that the term is often confusing [2]. The pattern of blood streaming on ventriculography also deceived us. We should have been super-cautious about the intracardiac morphology. Retrospectively, the 3-dimensional CT had been informative enough to recognize the variation of DORV. [Figure 2] At the preoperative stage, right aortic arch gathered more attention of the surgeons; whether any technical pitfalls

were hidden. The arterial switch is relatively rare in such a circumstance [3]. Fortunately, it did not turn out to be an issue.

Having admitted that the preoperative diagnosis could have been more precise, what would have been the alternative strategy? Initial banding of the pulmonary trunk would have been less invasive and provided broad strategical choices in the future. An immediate downside of the palliation would be considerable arterial desaturation persisting because of transposition physiology.

This streaming issue would be much less when banding is carried out concomitantly with the arterial switch maneuver. Oxygenated blood would mainly flow to the neo-aorta, providing a pinker circulation. When the patient became older beyond early infancy, intraventricular rerouting could be less demanding. A RV incision is dispensable at the neonatal stage.

The Réparation à l'Etage Ventriculaire (REV) procedure is an alternative, rerouting the LV to both semilunar valves[4]. The outlet septum was missing; therefore, nothing to resect. VSD enlargement is anatomically limited; better not to penetrate the ventriculo-infundibular fold. A downside of the REV procedure would be pulmonary regurgitation in the longer terms. Pulmonary valve replacement will not be straightforward because of the geometry around.

An intraventricular tunnel becomes long and tortuous in DORV with a non-committed VSD [5,6] when biventricularly repaired. Even for the REV procedure, the baffle design is compromised because of the presence of the tricuspid valve and its tension apparatus. As described by Belli et al. [7], the intraventricular tunnel becomes straighter and shorter by switching the great arteries. Still, the tricuspid valve interferes in accommodating a baffle. The septal leaflet would most likely adheres to the baffle, eventually causing tricuspid regurgitation. Thus, the Fontan type procedure remains as a definitive correction of the cyanotic circulation. Using the ventricles as a solitary chamber, the ventricular outlet is not compromised with a baffle. The tricuspid valve remains intact. Downside of this strategy is, of course, the unique circulation itself. It causes certain impediments in the longer terms. Heart failure and Fontan-associated liver disease are among current topics of concern. Biventricular physiology, if achievable, is preferred in this respect.

It is uncommon to repair of DORV of a non-committed VSD type during a neonatal period. We have come across only one case reported in literature [8]. Although the doubly-committed VSD type has a morphological spectrum [1], the combination of DORV with a non-committed VSD type and lack of the outlet septum has not been described in a clinical series to the best of our knowledge. Sequelae could grow in due course. Obstruction across the LV outflow tract and tricuspid regurgitation would be related to the intraventricular baffle. Neonatal ventricular incision could consequently cause ventricular dysfunction or ventricular arrhythmia. Function of the semilunar valves might deteriorate eventually, despite the baffle was meticulously fixed there.

After all, what could we have done better? No definitive answer. As far as the results in the immediate postoperative term is concerned, our neonatal repair was not unreasonable. Should we take a similar approach in an identical case? Yes, although a rare form of DORV is to be preoperatively recognized next time.

Financial Disclosure

The authors have no financial relationship to disclose relevant to this article.

Conflict of Interest

The authors have no conflicts of interest to disclose.

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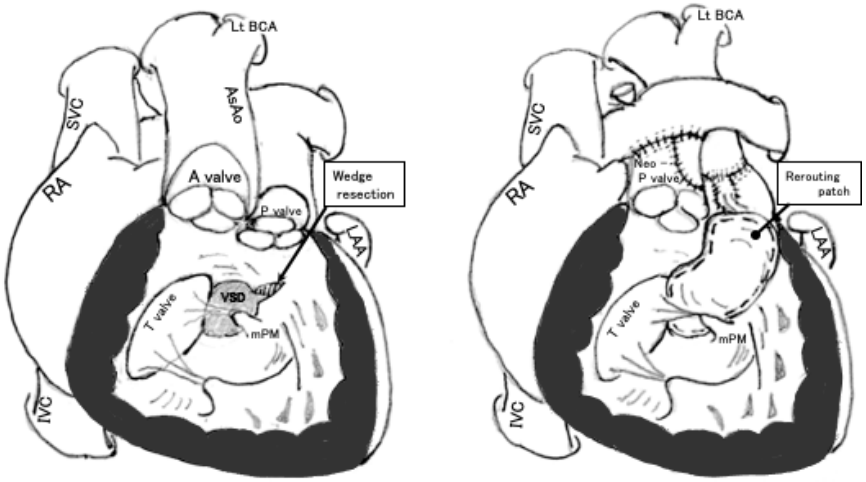
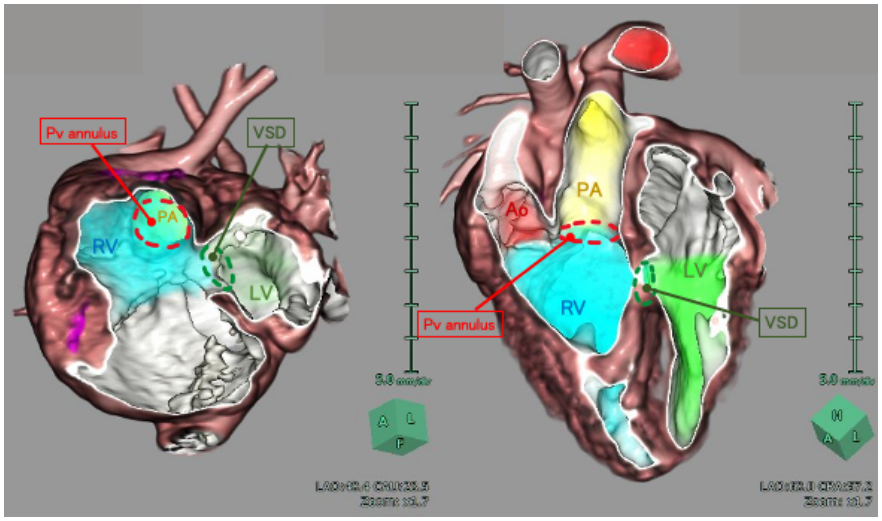
Figure legends

Figure 1: Schema of the procedure



SVC, superior vena cava; IVC, inferior vena cava; RA, right atrium; T valve, tricuspid valve; Avalve, aortic valve; P valve, pulmonary valve; Lt BCA, left brachiocephalic artery; AsAo, ascending aorta; VSD, ventricular septal defect; mPM, medial papillary muscle; LAA, left atrial appendage

Figure 2: Preoperative 3-dimensional computed tomography (images retrospectively reconstructed after repair)



Pv annulus, pulmonary valve annulus; VSD, ventricular septal defect; RV, right ventricle; LV, left ventricle; PA, pulmonary artery; Ao, aorta