# Konno-Rastan operation in a patient with dextrocardia

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#### Abstract

Konno-Rastan operation is the treatment of choice for relief of complex left ventricular outflow obstruction, especially in those with a small aortic annulus. In this report, we present a case of a ten-year-old boy with the diagnosis of recurrent diffuse subaortic stenosis and dextrocardia who underwent Konno-Rastan operation successfully.

# Konno-Rastan operation in a patient with dextrocardia

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# Abstract:

Konno-Rastan operation is the treatment of choice for relief of complex left ventricular outflow obstruction, especially in those with a small aortic annulus. When associated with dextrocardia, important aspects should be kept in mind due to the mirror-image anatomy. In this report, we present a case of a ten-year-old boy with the diagnosis of recurrent diffuse subaortic stenosis and dextrocardia who underwent Konno-Rastan operation successfully, and was asymptomatic with normal physical activity after a follow-up period of 6 months.

# Key words:

Left ventricular outflow obstruction, subaortic stenosis, konno-Rastan operation, dextrocardia

## Key Clinical Message:

konno-Rastan operation is the treatment of choice for diffuse subaortic stenosis especially in those with a small aortic annulus. When associated with dextrocardia, there will be important aspects that should be kept in mind throughout the operation due to the mirror-image anatomy.

# Introduction:

Aortoventriculoplasty (known as Konno procedure) was first described by both Konno and colleagues (1), and Rastan and Koncz (2) for the treatment of tunnel-type subaortic stenosis. It is considered the final

option for relief of complex left ventricular outflow obstruction (LVOTO) (3). Originally, it involved patch enlargement of both ventricles' outflow tracts along with the insertion of a mechanical aortic valve prosthesis (1, 2). Later, it became the procedure of choice for multilevel LVOTO, especially in those with a small aortic annulus (4, 5). Herein, we present a case of recurrent subvalvar and valvar aortic stenosis in a 10-year-old boy with associated dextrocardia who underwent Konno procedure.

## Case presentation:

A ten-year-old boy presented to our hospital with progressive exertional dyspnea and episodes of chest pain. He had undergone surgical repair of subaortic stenosis and supramitral ring 7 years ago. Transthoracic echocardiogram (TTE) revealed the following findings: situs inversus with dextrocardia, LVOTO with peak gradient (PG) of 90 mmHg along with moderate to severe aortic insufficiency (AI), moderate mitral regurgitation (MR) with mitral stenosis (MS) (PG across the mitral valve was 35 mmHg). Diagnostic cardiac catheterization was performed, and showed tunnel type LVOTO (Figure 1). Based upon these findings, the indication of Konno-Rastan operation was established. The patient was prepared appropriately and scheduled for elective surgical repair. The operation was performed via median sternotomy with great caution to avoid inadvertent rupture of any cardiac cavity due to the heavy adhesions from previous operation, and dextrocardia was kept in mind. The aorta was cannulated just below the take off of the innominate artery to gain as much length as possible on the ascending aorta. Bicaval cannulation was performed on the left side (Dextrocardia) to have a bloodless field. We placed a left ventricular vent via the left superior pulmonary vein. Aortic cross clamp was applied and the ascending aorta was opened longitudinally on the anterior aspect (Figure 2), and the cardioplegic solution was administered via the coronary ostia due to the severe AI. This incision was extended into the right coronary sinus to the right of the right coronary ostium (Dextrocardia) (Figure 3). By staying close to the right/left commissure, the conduction system is protected. The right ventricular outflow tract was opened and then cutting through the aortic annulus and ventriculoinfundibular fold into the ventricular septum (Figure 4). This incision is made between the right ostium and right/left commissure, staying closer to the right/left commissure as shown in Figure 3. The septal incision is usually about 10-15 mm in length and allows the aortic annulus to separate nicely (Figure 5). A sizer was placed to estimate the patch width and decide if the septal incision is adequate. The width of the patch will equal the additional annular circumference. The inferior aspect of the patch was sewn to the defect created in the ventricular septum (Figure 6). The superior aspect of the patch was used to augment the aortic sinus and sinotubular junction. Then, 2-0 ethibond horizontal mattress sutures were passed through the aortic annulus as per any aortic valve replacement and some of these sutures were passed through the patch in correspondence with the aortic annulus. The superior aspect of the patch was sutured to the aorticomy, and a second patch was used to augment the right ventricular outflow tract. The remainder of the operation was completed uneventfully. Postoperative TTE showed residual subaortic stenosis with PG of 40 mmHg. and there was significant improvement of MS with PG across the mitral valve of 12 mmHg. The patient was followed up for 6 months and was asymptomatic with normal physical activity.

## Discussion:

Diffuse subaortic stenosis is difficult to treat, and needs extensive resection to effectively relieve the obstruction. Anterior aortoventriculoplasty (Konno-Rastan procedure) has been shown to be the effective management of complex LVOTO especially in patients with small aortic annulus (1, 2). The most challenging step of the operation is the incision across the aortic annulus which should be performed with great caution to avoid any injury to the conduction system, the native pulmonary valve, and the right coronary ostium (3). In our patient who had dextrocardia, this issue was of great importance due to the mirror-image anatomy, which means that this incision should be to the right (instead of the left) of the right coronary ostium (6, 7). In a retrospective study, the overall 30-day mortality was 11.5%, with significant improvement in the PG across the left ventricular outflow tract (6). To the best of our knowledge this is the first and only published case of Konno-Rastan operation in a patient with dextrocardia.

# Conclusion:

The classic Konno-Rastan procedure using a mechanical valve can be performed with acceptable morbidity and mortality in this difficult group of patients. Associated dextrocardia mandates greater caution during operation due to the mirror-image anatomy.

# Figure legends:

Figure 1: Image of angiography showing the left ventricular outflow obstruction (subvalvar and valvar) and the dextrocardia.

Figure 2: A drawing showing the cannulated agrta and the site of both the agrtotomy and the right ventricular cardiomyotomy, and the right coronary artery.

Figure 3: A drawing showing the extension of the aortotomy to the right (Dextrocardia) of the right coronary artery.

Figure 4: A drawing the cutting through the aortic annulus and ventriculo-infundibular fold into the ventricular septum.

Figure 5: Intraoperative image showing the opened ventricular septum (dark line), the left ventricular cavity (dark circle), and the right ventricular cavity (dark star).

Figure 6: Intraoperative image showing the patch sewn to the opened ventricular septum. 1: right ventricular free wall, 2: right ventricular cavity, 3: the suture line between the patch and the edge of the opened ventricular septum, 4: the patch.

## **Author Contribution**

**Alwaleed Al-Dairy**: Planned and performed the work leading to the report. Wrote and reviewed successive versions and participated in their revisions.

**Bairak Salameh**: Participated in writing the report and approved the final version

Hadeel Badran: wrote and reviewed the successive versions and participated in their revisions

Hasan Hasan: wrote and reviewed the successive versions and participated in their revisions

Lujain Alkyakhii: wrote and reviewed the successive versions and participated in their revisions, and performed the drawings.

### **Author's Statement:**

Consent: Written informed consent was obtained from the patient's parents to publish this report in accordance with the journal's patient-consent policy.

#### Author's Declaration:

None of the authors listed on the manuscript are employed by a government agency that has a primary function other than research and/or education. Moreover, none of the authors are submitting this manuscript as an official representative or on behalf of the government.

## Conflict of interest:

The authors have no conflict of interest

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## Ethics statement:

The manuscript was approved by ethics committee at Damascus University.

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## Data availability statement:

The data that support the findings of this study are available from the corresponding author, [A.A], upon reasonable request.

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