Aortic Valve-Sparing Operation after Correction of Heart Displacement due to Pectus Excavatum Using Nuss Procedure in a Marfan Syndrome Patient

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Cardiovascular surgery in the setting of chest wall deformities is a clinical challenge. Pectus excavatum, for example, can cause heart displacement to the left thoracic cavity, following the poor operative field. This report highlights a case in which a successful aortic valve-sparing operation via conventional median sternotomy after correction of the heart displacement due to pectus excavatum using Nuss procedure in Marfan syndrome. This technique can be one surgical option in Marfan syndrome patients with pectus excavatum and thoracic aortic aneurysm under close follow up.

Keywords: aortic valve-sparing operation, heart displacement, pectus excavatum, Nuss procedure, Marfan syndrome

Introduction

Cardiovascular surgery in the setting of chest wall deformities such as pectus excavatum is the clinical challenge. The deformities make conventional median sternotomy technically more difficult with the possibility of injury to structures including heart, aorta and others. Moreover, pectus excavatum can cause heart displacement to left thoracic cavity, following the poor operative field.

Simultaneous repair of pectus excavatum and cardiovascular diseases has been reported in the literature.1) However, it may be associated with increased bleeding due to intraoperative heparinization and prolonged operative time.2) Nuss procedure, which is the minimally invasive procedure has been popularized,3) and simultaneous repair using this procedure has been also reported.4) Herein, we present a case of aortic valve-sparing operation after correction of heart displacement to left thoracic cavity due to pectus excavatum using Nuss procedure about 18 months previously.

Case Report

A 20-year-old man with Marfan syndrome was admitted due to an operation for annuloaortic ectasia (AAE). His mother and older sister with Marfan syndrome underwent the Bentall procedure and an aortic valve-sparing operation, respectively. He had been followed up by pediatricians since childhood. Serial transthoracic echocardiograph showed a gradual dilatation of sinus of Valsalva with a maximum diameter of up to 45 mm and mild aortic valve regurgitation. The pulmonary function test was within the normal range.

A problem was that heart was displaced into the left thoracic cavity due to pectus excavatum shown by computed tomography (CT) (Fig. 1A). It was the disadvantage for an operation through the conventional median sternotomy approach. That was why foreseeing the coming
operation for AAE, about 18 months previously; he underwent surgical correction for pectus excavatum using Nuss procedure by pectus bar (Walter Lorenz Surgical, Inc. Jacksonville, FL, US).

In addition to the surgical correction, medical interventions including oral angiotensin receptor blocker and β blocker were also initiated to prevent the progressive dilatation of aorta.

After the chest wall deformity and heart displacement were corrected (Fig. 1B), removal of the pectus bar was done prior to an operation for AAE during this admission. We selected the conventional median sternotomy approach to expose adequate position of the heart. The lower portion of sternum was adhered strongly due to the pectus bar, but we could dissect the sternum from surrounding tissue. Pericardiotomy was performed in a routine fashion easily. The heart itself was rotated slightly to the left, and the superior vena cava and aorta were also identified to be slightly to the left side, compared with usual cases (Fig. 2). Aortic valve-sparing operation using J Graft SHIELD NEO 30MM (Japan Lifeline Co., Ltd., Tokyo, Japan) by reimplantation technique was performed in a standard fashion safely and successfully through that approach. Additionally, neo-aortic sinuses could be created by plicating the tubular graft using 5-0 polypropylene above each commissure. Although atelectasis of the right lung occurred postoperatively due to possibly chest wall instability, he was discharged in a good condition with a stable chest wall appearance.

**Discussion**

In the setting of chest wall deformities, it is difficult to obtain an adequate operative field through conventional
median sternotomy, because of severe, posterior concavity of the sternum or heart displacement towards the left thoracic cavity.\textsuperscript{4)}

The deeper the sternal depression and the larger the CT depression index, the more pronounced is the cardiac rotation.\textsuperscript{5)}

The right ventricle is the most vulnerable chamber to be rotated or compressed in chest wall deformities such as pectus excavatum. The depressed sternum results in not only compression of the right ventricle, but also myocardial ischemia leading to the electrocardiogram changes such as partial right bundle-branch block.\textsuperscript{5)}

Nuss D et al. introduced the minimal invasive procedure using the convex bar to repair pectus excavatum.\textsuperscript{3)} Because its technique is safe and successful in managing pectus excavatum, it has become widely accepted.

In reports about patients with both pectus excavatum and cardiovascular disease, one describes simultaneous repair,\textsuperscript{1,2,4)} and another, a staged repair.\textsuperscript{2)}

In our report, taking into consideration the upcoming operation for dilatation of the aorta and heart displacement to left thoracic cavity, we decided on the staged, aortic valve-sparing operation, and gave priority to the correction of the pectus excavatum by using the Nuss procedure to keep the sternum intact.

To make the thoracic cage morphometrically steady after this procedure, it takes a longer follow-up period before removal of the pectus bar. In the present case, the dilatation of aorta progressed earlier than expected, which increased the risk of rupture. Therefore, the bar had to be removed prematurely. It was, however, enough to correct the heart displacement, but it was not sufficient for steadying the sternum.

Needless to say, careful attention must be paid first and foremost to progressive dilatation of the aorta by imaging modalities. And the position of pectus bar to prevent the life-threatening complications is also important.\textsuperscript{6)}

Additionally, medical interventions are mandatory during the follow-up period.

**Disclosure Statement**

Fukunaga N and co-authors have no conflict of interest.

**References**


