

# Ascending Aorta and Aortic Root Replacement With the Bentall Procedure in a Patient With Dextrocardia and Situs Inversus Totalis

Özlem Balcıoğlu<sup>1</sup>, Barçın Özçem<sup>1</sup>, Aşkın Ali Korkmaz<sup>2</sup>

<sup>1</sup>Department of Cardiovascular Surgery, Near East University Faculty of Medicine, Nicosia, Cyprus

<sup>2</sup>Department of Cardiovascular Surgery, Memorial Hospital, İstanbul, Turkey

## ABSTRACT

We report a case of situs inversus totalis with severe aortic regurgitation, ascending aorta aneurysm and aortic root dilatation in an adult patient. This is the first report in the literature concerning a Bentall operation which was performed successfully in situs inversus totalis patient. The post-operative period of our patient was uneventful and he was discharged from the hospital after 10 days.

**Keywords:** Situs inversus totalis, dextrocardia, Bentall procedure, ascending aort aneurysm

## INTRODUCTION

Situs inversus totalis is an autosomal recessive congenital anomaly characterized by the opposite positioning of the thoracic and abdominal organs in a reverse mirror image. Dextrocardia is a component of situs inversus totalis. In this particular case, aortic valve insufficiency, aortic root and ascending aorta dilatation were also co-existent. We report a successful Bentall operation that can be a reference for the treatment plans of similar cases in the future

## CASE PRESENTATION

A 57-year-old male patient with known dextrocardia was admitted to the cardiology department with shortness of breath and chest pain complaints. In spite of his suspicious symptoms such as chronic sinusitis, chronic rhinitis and red eye that were consistent with primary ciliary dyskinesia syndrome; he was fertile and did not suffer from any lower respiratory tract diseases. Physical examination revealed the presence of a sinus rhythm and a diastolic murmur becoming more prominent at the left parasternal border.

Echocardiography showed dextrocardia with left ventricular (LV) dilatation, a slightly depressed LV ejection fraction (45%–50%) and severe aortic regurgitation. Sinus valsalva, sinotubular junction and ascending aorta measurements were recorded as 52 mm, 49 mm and 47 mm, respectively (Figure 1). After, contrast enhanced computerized tomography (CT), the patient was diagnosed with “Situs Inversus Totalis” (SIT) which is the presence of reversed mirror images of all thoracoabdominal organs (Figures 2 and 3).

Surgery was performed under general anesthesia using standard median sternotomy. After the pericardiectomy, dextrocardia and right-sided aortic arch were observed. The brachiocephalic trunk and both vena cava were located on the left side of the patient (Figure 4). No venous return abnormalities were detected. Despite the dilatation of the proximal ascending aorta, there was a safety cross clamping area just below the brachiocephalic trunk. Cardiopulmonary bypass was established by the chief surgeon standing on the right side of the patient by using the left sided right atrial appendage and the distal ascending aorta for cannulation. After the aortic cross clamp, both anterograde and retrograde cardioplegia were given and subsequently, aortotomy was performed. The aortic wall was extremely thinned, the valve ring

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**ORCID iDs of the authors:** Ö.B. 0000-0002-8935-1477; B.Ö. 0000-0003-1459-7939; A.A.K. 0000-0002-2263-785X.



**Address for Correspondence:** Özlem Balcıoğlu

**E-mail:** ozlem.balcioglu@neu.edu.tr

**ORCID ID:** orcid.org/0000-0002-8935-1477

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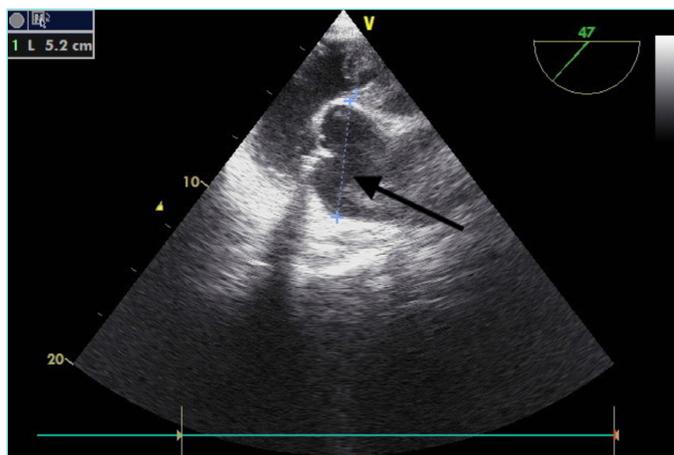
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was dilated and the cusps were highly degenerated. For complete myocardial protection, selective cardioplegia was also applied.

Severe aortic regurgitation, ascending aorta aneurysm and dilatation of the aortic anulus led us to consider that valve sparing surgery would not be beneficial; therefore, the team decided to replace the root and the ascending aorta completely by performing the Bentall procedure. Both coronary buttons were prepared as usual and the aortic valve was excised. A 25 mm mechanical aortic valve (SJM™ Masters Series Mechanical Heart Valve SJM) was sutured to the 30 mm dacron graft



**Figure 1.** Preoperative aortic root measurements by transthoracic echocardiography and sinus valsalva was measured at 5.2 cm and is shown with an arrow.



**Figure 2.** Preoperative CT scans showed reverse mirror images of all thoracoabdominal organs.

CT: Computed tomography

and this was implanted with a continuous suturing technique, by using a 4/0 propylene suture. After the re-connection of the coronary buttons, the tubular graft length was trimmed and distal anastomosis was completed. The patient was weaned from the cardiopulmonary bypass with normal sinus rhythm. No complications were observed in the early postoperative period and no inotropic support was required. The patient was extubated at the 6<sup>th</sup> hour of the post-operative period and was discharged from the hospital after 10 days.

## DISCUSSION

SIT is an autosomal recessive congenital anomaly characterized by opposite positioning of the thoracic and abdominal organs as mirror images. Dextrocardia with SIT is a very rare condition occurring in approximately 1–2 per 10,000 in the general population.<sup>1</sup> Apart from the small percentage of SIT patients that have other congenital heart anomalies, such as transposition of the great vessels or heart defects, most affected adult patients live a normal lifespan and diagnosis takes place by chance when an X-ray or CT scan is performed for another reason.<sup>2</sup>



**Figure 3.** Preoperative CT scans showed reverse mirror images of all thoracoabdominal organs.

CT: Computed tomography

Reverse positioning of the internal organs takes place during embryological development. Although the genes that regulate the anatomical positioning of the organs are known, the specific genetic cause of dextrocardia with SIT is still unidentified. Some patients have association with primary ciliary dyskinesia called Kartagener syndrome and are diagnosed in the early childhood period. For the genetic research of this case, blood samples of the patient and family were collected and further investigations were performed.

In our opinion, the pre-operative evaluation, the imaging techniques that were used and the surgical plan were the key points in the success of this case. CT imaging presented the anatomical abnormalities sufficiently and it was extremely helpful for planning the cannulation strategy, setting-up the operative tools and positioning the team during the surgery. However, conventional angiography was also applied in order to eliminate the presence of concurrent coronary artery disease and venous perfusion defects. No coronary artery lesions or venous perfusion defects were found. As neither a left atrial approach nor coronary bypass surgery were required, in contrast with some articles in the literature, the surgical team decided to implement the conventional positioning where the chief surgeon stands on the right side of the patient.<sup>3</sup> The surgery was successfully completed with no difficulties or complications encountered.

Ascending aortic aneurysm is a life-threatening disease related with rupture and aortic dissection risk. As is well-known, a well-planned elective surgery, applied by experienced centers, is the curative treatment. The Bentall procedure is still the most helpful technique,

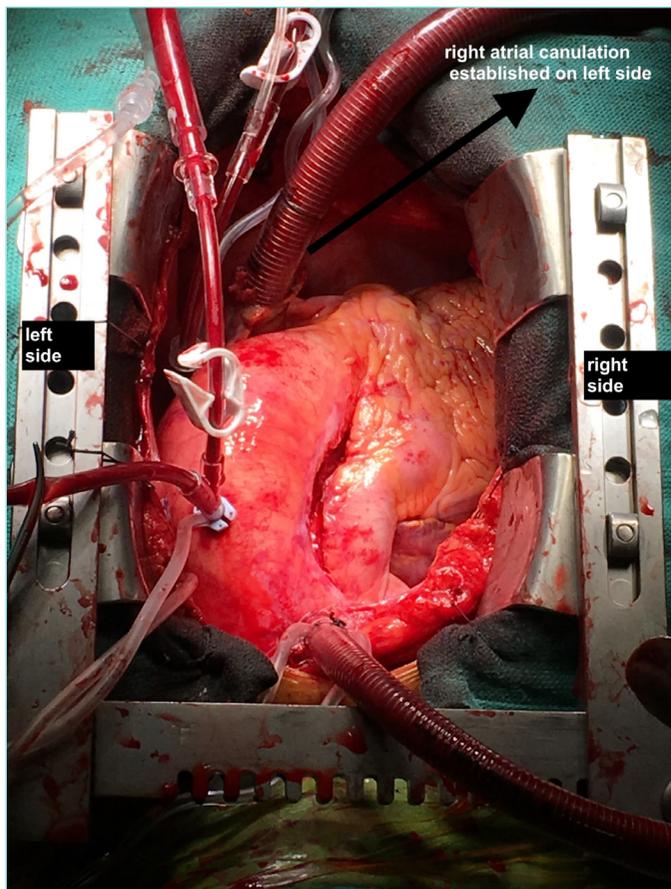


Figure 4. Preoperative view.

especially for ascending aortic aneurysms that include aortic root dilation and aortic valve regurgitation and are inappropriate for valve sparing surgery.<sup>4</sup>

In conclusion, the presence of SIT with dextrocardia is an extremely rare condition. In this particular case, aortic valve insufficiency, aortic root and ascending aorta dilatation were also co-existent with SIT with dextrocardia. In the literature, no cases which involved the use of the Bentall procedure were reported. In our opinion, the Bentall procedure can be safely applied to such cases without any requirement to change the positions of the surgical team in the surgery. This particular case can be a reference for the treatment plans of similar cases which may be encountered in the future.

## MAIN POINTS

- The Bentall Operation is still a life-saving procedure in aortic root dilatation and severe aortic regurgitation if aortic valve sparing techniques are not suitable.
- Complete diagnosis and well-planned operation techniques carried out by experienced centers is the key point of success in such cases.
- A small percentage of SIT patients have additionally heart defects which may lead to diagnosis in early childhood.
- Chronic sinusitis, chronic rhinitis, red eyes and infertility may be associated with primary ciliary dyskinesia in SIT patients and these patients should undergo genetic investigation for Kartagener syndrome.

## ETHICS

**Informed Consent:** Informed consent was obtained from the patient before the surgery.

**Peer-review:** Externally peer-reviewed.

## Authorship Contributions

Concept: Ö.B., Design: Ö.B., Supervision: A.A.K., Data Collection and/or Interpretation: B.Ö., Literature Search: B.Ö., A.A.K., Writing: Ö.B., Critical Review: A.A.K.

## DISCLOSURES

**Conflict of Interest:** No conflict of interest was declared by the authors.

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