Türk Göğüs Kalp Damar Cerrahisi Dergisi 2017;25(3):450-454
http://dx.doi.org/doi: 10.5606/tgkdc.dergisi.2017.14312

Case Report / Olgu Sunumu

Totally endoscopic robotic atrial septal defect closure in an adult with Down syndrome

Down sendromlu bir erişkinde atriyal septal defektin total endoskopik robotik kapatılması

Burak Onan, Ersin Kadiroğulları, Ünal Aydın, Yeşim Güner, Korhan Erkanlı

Department of Cardiovascular Surgery, İstanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Turkey

ABSTRACT

Cardiac anomalies in Down syndrome often present with atrioventricular canal defects, ventricular septal defects, and atrial septal defects. Although complex cardiac pathologies are mostly seen during infancy, milder cardiac defects such as atrial septal defects can be seen in adulthood. Herein, we report a 25-year-old male case of Down syndrome who presented with a secundum-type atrial septal defect and rim deficiency and treated with a minimally invasive robotic surgery for the atrial septal defect closure to minimize the surgical risks.

Keywords: Atrial septal defect; Down syndrome; robotic surgery.

ÖZ

Down sendromunda kardiyak anomaliler genellikle atriyoventriküler kanal defektleri, ventriküler septal defektler ve atriyal septal defektler ile birlikte seyreder. Kompleks kardiyak patolojiler sıklıkla yenidoğan döneminde görülmekle birlikte, atriyal septal defektler gibi daha hafif kardiyak defektler erişkinlikte görülebilmektedir. Bu yazıda, rim yetersizliği ve sekundum tip atriyal septal defekti olan ve atriyal septal defektin kapatılmasında cerrahi riskleri en aza indirgemek amacıyla minimal invaziv robotik cerrahi ile tedavi edilen 25 yaşında Down sendromlu bir erkek olgu sunuldu.

Anahtar sözcükler: Atriyal septal defekt; Down sendromu; robotik cerrahi.

Down syndrome is a common chromosomal disorder which is seen in 1:1000 of the births.[1-3] Several studies have shown that the probability of concomitant emergence of congenital disorders with Down syndrome as high as 44%.[1] Although complex forms of cardiovascular anomalies, such as atrioventricular canal defects and ventricular septal defects (VSDs) are diagnosed in the early postnatal period, atrial septal defects (ASDs) may present with an incidence of about 16.7% in adulthood.[2,3] Early ASD closure prevents the potential risks of right ventricular dilatation and pulmonary dysfunction.[2,3] Alternative to traditional sternotomy and thoracotomy incisions, totally endoscopic robotic ASD closure surgery has proved its safety in adults.[4-7] Robotic surgery can be a preferred approach to minimize the risks of conventional sternotomy and thoracotomy incisions.

Herein, we report a 25-year-old male case of Down syndrome who presented with a secundum-type ASD and rim deficiency and treated with a minimally invasive robotic surgery for the closure.

CASE REPORT

A 25-year-old male patient with Down syndrome presented to our clinic with exertional dyspnea and fatigue. The patient was mentally competent and physically active on his daily life. On admission, vital signs were stable. Physical examination revealed hypertelorism, depressed nasal bridge, protrusion of the tongue, and a broad and short neck. Cardiac auscultation showed a 1 to 2/6 grade diastolic murmur over the left parasternal area. Biochemistry test results were also normal. Electrocardiography revealed a right axis deviation, suggesting right ventricular dilatation. Chest X-ray showed a normal cardiothoracic ratio with

Available online at
www.tgkdc.dergisi.org
doi: 10.5606/tgkdc.dergisi.2017.14312
QR (Quick Response) Code

Received: January 07, 2017 *Accepted:* March 14, 2017

Correspondence: Burak Onan, MD. İstanbul Mehmet Akif Ersoy Göğüs ve Kalp ve Damar Cerrahisi Eğitim ve Araştırma Hastanesi, Kalp ve Damar Cerrahisi Kliniği, 34303, Küçükçekmece, İstanbul, Turkey.
Tel: +90 553 - 622 38 78 e-mail: burakonan@hotmail.com

an increased pulmonary vascularity. Transthoracic echocardiography examination revealed left-to-right shunting, enlarged right cardiac chambers, and a mild tricuspid regurgitation (Figure 1). The size of the septal defect was 3x2 cm in diameter. It showed ejection fraction of 65%, pulmonary artery pressure of 35 mmHg, and Qp/Qs ratio of 2.0. Transesophageal echocardiography confirmed a secundum-type ASD with an aortic rim of 2 mm, a superior vena cava rim of 17 mm, and an inferior vena cava rim of 2 mm, excluding the possibility of transcatheter closure. The patient underwent a totally endoscopic robotic surgery to minimize the risks of traditional sternotomy/thoracotomy incisions and to offer a better recovery period.

The da Vinci SI robotic surgery system (Intuitive Surgical Inc., Sunnyvale, CA, USA) was used. The patient was intubated for single-lung ventilation under general anesthesia. After systemic heparinization, the right internal jugular vein and the right femoral vessels were cannulated for cardiopulmonary bypass (CPB). A service port of 2 cm was opened through the fourth intercostal space in the anterior axillary line (Figure 2). A 30-degree endoscope was, then, inserted into the pleural space through the fourth intercostal space anteriorly. Two additional instrument ports in the third and fifth intercostal spaces were used. Atrial retractor was introduced through the fifth intercostal space anteriorly. The surgical field was flooded with carbon dioxide.

An autologous glutaraldehyde-treated pericardial patch was used for the closure. After delivery of antegrade blood cardioplegia at moderate hypothermia, cardiac arrest was established. Both venae cavae were occluded using atraumatic vascular bulldog clamps. The right atriotomy incision was done obliquely. The secundum-type ASD was explored (Figure 3). There was no associated anomaly of systemic or pulmonary venous return. The defect was closed using a glutaraldehyde-treated autologous pericardial patch of 3x2 cm and 5/0 polytetrafluoroethylene suture. The pericardial patch was used in this large defect with an inferior rim deficiency. Following de-airing and closure of the atrium, the patient was uneventfully weaned from CPB. The total CPB and aortic clamp times were 62 min and 35 min, respectively. Surgery took about three hours with a ventilation time of four hours and length of intensive care unit stay of about 16 hours. During surgery, no blood product was needed.

The postoperative period was uneventful and the patient was discharged in the third postoperative day. He is still followed without any complication at three months.

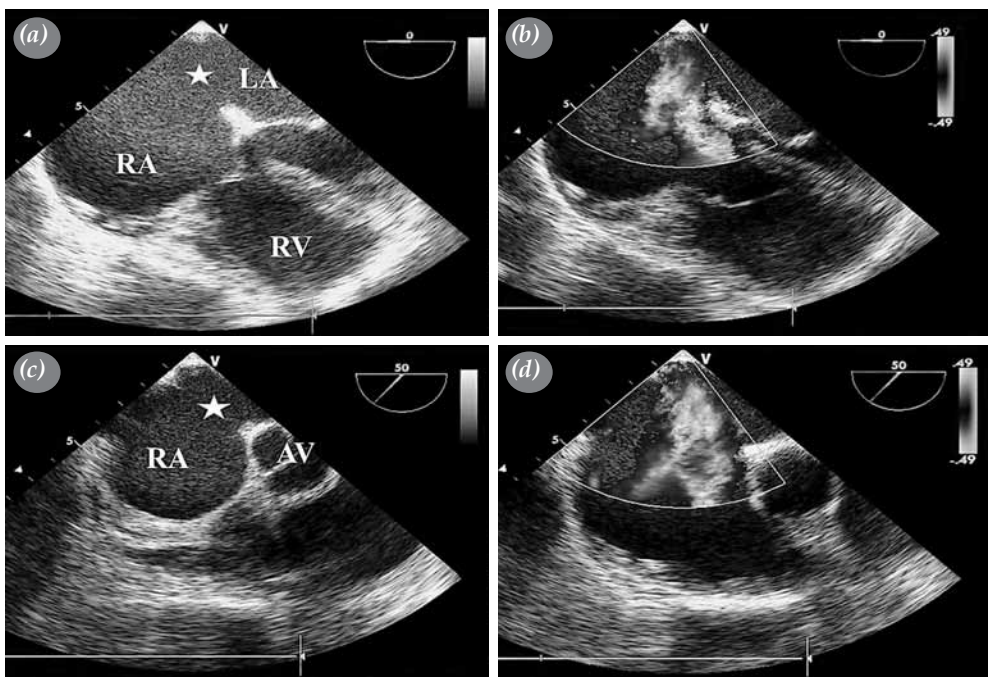

Figure 1. Transesophageal echocardiography showing a secundum-type atrial sepal defect (white star) with left-to-right interatrial shunting.

AV: Aortic valve; LA: Left atrium; RA: Right atrium; RV: Right ventricle.

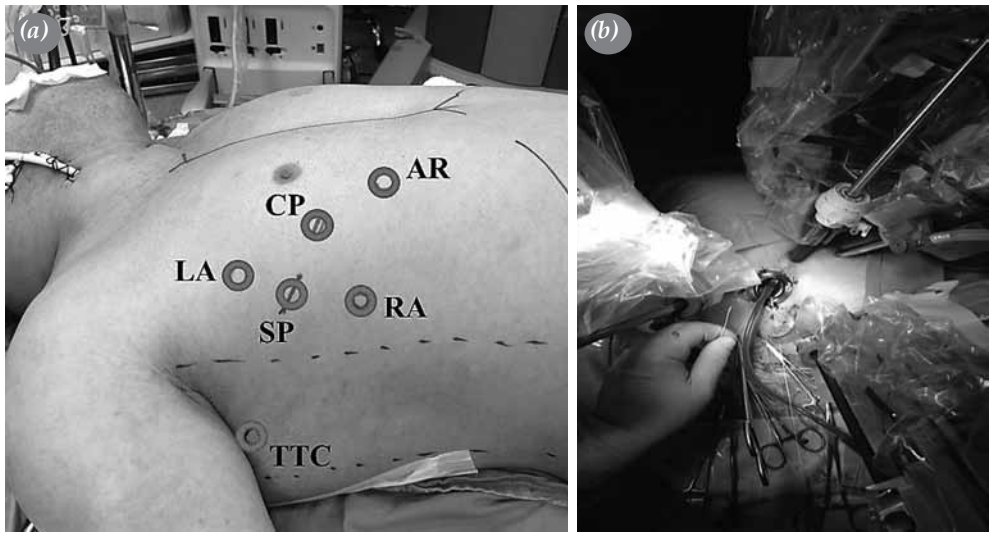

Figure 2. Operative view.
AR: Atrial retractor; CP: Camera port; LA: Left arm; RA: Right arm; SP: Service port; TTC: Transthoracic clamp.

DISCUSSION

Congenital anomalies of the heart are common in Down syndrome with ASDs and VSDs, accounting for the most common defects.[2,3] Previously, Nisli et al.,[3] evaluated the incidence and types of coronary heart disease patterns in the Turkish children with Down syndrome. The authors reported that, in 421 patients with Down syndrome, 77.6% had a single cardiac lesion, while the remaining 22.4% had multiple defects. The most common single defect was an atrioventricular septal defect in 34.2% of patients, followed by secundum-type ASDs in 16.7% and VSDs in 16.5%.

Atrioventricular canal defects or VSDs with pulmonary hypertension should be surgically corrected in the first six months of life. However, some milder forms of cardiac defects such as ASDs can be diagnosed in adulthood. In these cases, surgical or percutaneous repair is needed, after a detailed examination of organ systems to reduce morbidity and mortality.

Surgical treatment is an alternative to percutaneous techniques for closure of large ASDs and rim deficiencies.[4-7] Surgical ASD closure is traditionally performed using sternotomy or thoracotomy incisions. Majdalany et al.[2] reported that adult patients with Down syndrome could undergo cardiac surgery with a low risk of mortality and acceptable morbidity. As an alternative approach, robotic surgery can be an alternative option with a negligible operative risk, low morbidity, and early return to daily activities in Down syndrome.[4-7]

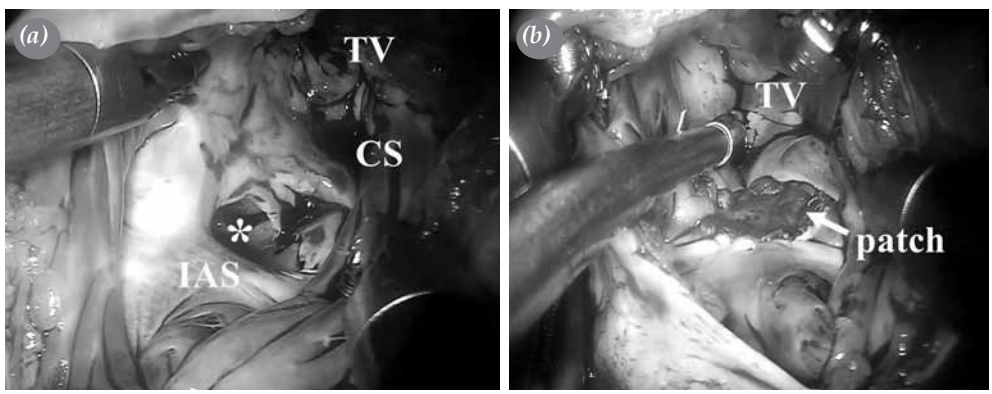

Figure 3. Endoscopic view.
CS: Coronary sinus; IAS: Interatrial septum; TV: Tricuspid valve; * Atrial septal defect.

Robotic surgery is technically feasible for primum- or secundum-type defects and sinus venous defects with partial anomalous venous return of the right lung.[4-7] The main advantages of robotic surgery include less pain, shortened hospital stay, early return to daily life and work, and almost no transfusion requirements.[4-10] In addition, morbidity and mortality rates are similar to traditional incisions, despite some technical challenges.[4,5] However, there are some disadvantages such as limited availability of robotic systems, high costs, and necessity of a learning curve.[4-10] Although the operative times of robotic operations are relatively longer than conventional procedures, CPB and aortic clamping times can be reduced to acceptable levels with increasing experience. Previously, Bonaros et al.[5] showed that longer operation times had no adverse impact on intraoperative and postoperative outcomes. Our experience also confirms that operation time of robotic surgery is comparable with conventional sternotomy and thoracotomy procedures.

The most common postoperative complications in Down syndrome are related to pulmonary dysfunction.[2] Although pulmonary abnormalities are seen more frequently in infants and children, it is a rare complication in the adults with Down syndrome.[2] However, the weaning period, hospital stay, and recovery period can be longer due to pulmonary dysfunction causing obstructive and restrictive ventilation patterns.[2,3] In the literature, previous studies have shown that robotic approach can be an option for patients seeking a safe, minimally invasive ASD repair with an excellent long-term result.[4,5] Therefore, in our case, we changed our surgical strategy from median sternotomy or thoracotomy approach to a totally endoscopic robotic surgery for the closure to minimize postoperative complications.

Another potential morbidity in Down syndrome is postoperative atrial arrhythmia.[2] It has been reported that atriotomy incisions and related scars can be substrates for atrial arrhythmias following congenital heart surgery in adults.[8] Yang et al.[8] also reported that most postoperative macro-reentry atrial tachycardia were atriotomy-related. The authors concluded that the incidence of postoperative atrial arrhythmias can be reduced, if the directions of atriotomy lines are modified, which means less manipulation of the atrial wall. In our case, there was no postoperative supraventricular arrhythmia such as atrial ectopic beats, fibrillation, or tachycardia. This can be related to a less inflammatory response, which is related to the lack of sternotomy and mediastinal dissection. No additional incision on the right atrial wall is made for cannulation, as peripheral cannulation is used.

In robotic procedures, preservation of the sternum and its stability is an advantage in the postoperative period.[9,10] However, in patients with mental retardation, the weaning from ventilation in the intensive care unit and postoperative rehabilitation in the ward can be challenging due to limited cooperation and unawareness of medical treatment and interventions. In particular, the sternum can be at risk of dehiscence following surgery due to increased pulmonary secretions, pain, and uncontrolled mobility in the bed and movements. In addition, superficial or deep wound infections, sternal osteomyelitis, and mediastinitis can be regarded as the other potential risks following sternal dehiscence.[4-10] Therefore, totally endoscopic robotic approach may be beneficial to avoid sternal and mediastinal complications in patients with Down syndrome.

In the presented case, we performed an ASD closure using the Da Vinci SI surgical system. To the best of our knowledge, this is the first case of robotic ASD closure in Down syndrome. In our case with Down syndrome, we used robotic approach to minimize postoperative complications.

In conclusion, robotic surgery is a feasible technique for the closure of atrial septal defects in adults with Down syndrome. Although percutaneous closure is the initial procedure of choice for atrial septal defect closure, robotic surgery can be preferred as an alternative approach, rather than traditional surgery techniques, in patients who are not eligible candidates for percutaneous treatment.

Declaration of conflicting interests

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding

The authors received no financial support for the research and/or authorship of this article.

REFERENCES

1. Freeman SB, Taft LF, Dooley KJ, Allran K, Sherman SL, Hassold TJ, et al. Population-based study of congenital heart defects in Down syndrome. Am J Med Genet 1998;80:213-7.
2. Majdalany DS, Burkhart HM, Connolly HM, Abel MD, Dearani JA, Warnes CA, et al. Adults with Down syndrome: safety and long-term outcome of cardiac operation. Congenit Heart Dis 2010;5:38-43.
3. Nisli K, Oner N, Candan S, Kayserili H, Tansel T, Tireli E, et al. Congenital heart disease in children with Down's syndrome: Turkish experience of 13 years. Acta Cardiol 2008;63:585-9.

4. Xiao C, Gao C, Yang M, Wang G, Wu Y, Wang J, et al. Totally robotic atrial septal defect closure: 7-year single-institution experience and follow-up. Interact Cardiovasc Thorac Surg 2014;19:933-7.
5. Bonaros N, Schachner T, Oehlinger A, Ruetzler E, Kolbitsch C, Dichtl W, et al. Robotically assisted totally endoscopic atrial septal defect repair: insights from operative times, learning curves, and clinical outcome. Ann Thorac Surg 2006;82:687-93.
6. Onan B, Aydın Ü, Kahraman Z, Bakır İ. Robotic surgery for atrial septal defect closure in a case of Kabuki syndrome. Turk Kardiyol Dern Ars 2017;45:77-81.
7. Onan B, Aydin U, Basgoze S, Bakir I. Totally endoscopic robotic repair of coronary sinus atrial septal defect. Interact Cardiovasc Thorac Surg 2016;23:662-4.
8. Yang G, Du X, Ni B, Chen H, Qi R, Cai C, et al. Prevention of postsurgical atrial tachycardia with a modified right atrial free wall incision. Heart Rhythm 2015;12:1611-8.
9. Sagbas E, Sanisoglu I, Guden M, Caynak B, Akpinar B. Robotically enhanced totally endoscopic coronary artery bypass surgery in three cases. Turk Gogus Kalp Dama 2008;16:254-6.
10. Kocyigit M, Akpek EA, Tetik O, Senay S, Alhan C. Our anesthesia experiences at robotic-assisted and robotic cardiac surgery. Turk Gogus Kalp Dama 2013;21:972-7.