

Chapter 5

OVERVIEW AND CURRENT SURGICAL APPROACHES FOR ATRIAL SEPTAL DEFECT

Mehmet Erin TÜYSÜZ¹

Introduction

The first successful surgical atrial septal defect (ASD) closure was performed using a pump oxygenator by John Gibbon on May 6, 1953. Initially, ASD used to be closed by performing a highly risky technique in which the defect is sewn with sutures placed from the outside of heart under deep hypothermic circulatory arrest. As the surgical experience increases, the surgical closure of ASD has been performed successfully (1).

Incidence and Etiology

ASD is the third most common congenital heart disease. It is encountered in 56 of 100000 live births (2). 30% out of adult congenital heart defects is ASD (3). The most common type is ostium secundum septal defect and it is seen the rate of 65-70%. Many ASDs are sporadic and the cause of it is not certain. Autosomal dominant inheritance has been reported in familial type (4,5). The mutation in the 14q12 gene, which plays a major role in cardiac septation, is considered to be responsible for the pathogenesis (6). The mutation in NKX2-5 gene is also the responsible factor for the pathogenesis of ASD with atrioventricular block (7,8,9). Besides, the risk of ASD is higher in the family group, particularly in the twins (10). There is a relationship between ASD and genetic disorders such as the syndromes of Holt-Oram, Ellis van Creveld, Noonan, Down, Budd-Chiari and Jarcho-Levine (11-18). Holt-Oram syndrome in which there is a NKX2-5 mutation presents in 66% out of ASD patients. ASD is also encountered in patients with trisomy 21 syndrome (19). In pregnancy, various factors such as the exposure of some chemical substances, fetal alcohol syndrome (20), the smoking in the first trimester (21,22) and the usage of some antidepressant drugs (23-25) increase the risk of ASD. Furthermore, it is reported that there is an increased ASD risk in women without diabetes who eat foods with high glycemic index (26,27) and in women with pregnancy over the age of 35 (28,29).

Embryologic and Anatomic Features

The atrial septum originates from the fossa ovalis which consists of two anatomical structures as the part of muscular and valvular. The muscular part of fossa ovalis

¹Mersin City Training and Research Hospital, Department of Cardiovascular Surgery, Mersin, Turkey

ASD closure with the help of 3D printing technology

When the echocardiography and computed tomography which are conventional imaging methods are inadequate in some conditions, 3D printing technology may be useful in the preoperative evaluation (97). In this method, the heart rate of patients being in the procedure is decreased by beta blocker and the images of patients are obtained by multi-slice computed tomography. The simulation of cardiac structures and ASD is achieved by 3D printer and 3D model is printed. This method is feasible in both surgical and transcatheter closure for ASD.

ASD Closure Surgery in the Pregnancy

In generally pregnancy, during cardiac surgery with cardiopulmonary bypass, while there is no an increase in maternal mortality rate, fetus death due to hypotension and non-pulsatile blood flow is 10-15% (98,99).

The pulmonary and systemic vascular resistances of patient with pregnancy and ASD decrease in the first trimester (100). In patient with pregnancy and ASD, although the pregnancy is well tolerated, there is a high risk of mortality, especially one month after giving birth, in pregnant patients with pulmonary hypertension (101). There are different treatment opinions in this patient group. The percutaneous ASD closure has been applied in the past, however, this procedure is not appropriate method in ASD with large defect (102). The second option is the therapeutic termination of pregnancy which is not accepted by most of patients, and then the surgical closure for ASD. The other option is that the pulmonary hypertension is kept under control by medical approaches until the birth and then the surgical closure for ASD is performed.

References

1. Constantine M, Carl B. Pediatric Cardiac Surgery. 4th ed. Chicago: Wiley-Blackwell: 879-82.
2. Hoffman J, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002; 39: 1890-900.
3. Kotowycz M, Therrien J, Ionescu-Ittu R, et al. Long-term outcomes after surgical versus transcatheter closure of atrial septal defects in adults. *JACC Cardiovasc Interv.* 2013; 6: 497-503.
4. Caputo S, Capozzi G, Russo M, et al. Familial recurrence of congenital heart disease in patients with ostium secundum atrial septal defect. *Eur Heart J.* 2005; 26: 2179-84.
5. Chen Y, Han Z, Yan W, et al. A novel mutation in *GATA4* gene associated with dominant inherited familial atrial septal defect. *J Thorac Cardiovasc Surg.* 2010; 140: 684-87.
6. Ching Y, Ghosh T, Cross S, et al. Mutation in myosin heavy chain 6 causes atrial septal defect. *Nat Genet.* 2005; 37: 423-28.
7. Bjørnstad P, Leren T. Familial atrial septal defect in the oval fossa with progressive prolongation of the atrioventricular conduction caused by mutations in the *NKX2.5* gene. *Cardiol Young.* 2009;19: 40-44.
8. Rifai L, Maazouzi W, Sefiani A. Novel point mutation in the *NKX2-5* gene in a Moroccan family with atrioventricular conduction disturbance and an atrial septal defect in the oval fossa. *Cardiol Young.* 2007; 17: 107-09.
9. Gelernter-Yaniv L, Lorber A. The familial form of atrial septal defect. *Acta Paediatr.* 2007; 96: 726-30.
10. Caputo S, Capozzi G, Russo M, et al. Familial recurrence of congenital heart disease in patients with ostium secundum atrial septal defect. *Eur Heart J.* 2005; 26: 2179-84.

11. Strauss R, Ferguson A, Rittey C, Cork M. Microcephaly-lymphoedema-chorioretinal-dysplasia syndrome with atrial septal defect. *Pediatr Dermatol.* 2005; 22: 373-74.
12. Yoshihara K, Ozawa T, Sakuragawa H, et al. Noonan syndrome associated with atrial septal defect, pulmonary stenosis, and completely unroofed coronary sinus without LSVC: a case report. *Kyobu Geka.* 1999; 52: 134-37.
13. Cunningham E, Elliott D, Miller N, et al. Familial Axenfeld-Rieger anomaly, atrial septal defect, and sensorineural hearing loss: a possible new genetic syndrome. *Arch Ophthalmol.* 1998; 116: 78-82.
14. Diegeler A, Van Son J, Mohr F. Budd-Chiari syndrome as late complication of secundum atrial septal defect closure. *Eur J Cardiothorac Surg.* 1997; 12: 501-03.
15. Shimizu K, Arai H, Sakamoto T, et al. Jarcho-Levin syndrome associated with atrial septal defect and partial anomalous pulmonary venous return: a case report. *J Card Surg.* 1997; 12: 198-200.
16. Aynaci F, Ozdemir M, İşik Y. Atrial septal defect in Hallermann Streiff syndrome. *Genet Couns.* 1997; 8: 145-46.
17. Holt M, Oram S. Familial heart disease with skeletal malformations. *Br Heart J.* 1960; 22: 236-42.
18. Ganie M, Laway B, Ahmed S, et al. Mayer-Rokitansky-Kuster-Hauser syndrome associated with atrial septal defect, partial anomalous pulmonary venous connection and unilateral kidney—an unusual triad of anomalies. *J Pediatr Endocrinol Metab.* 2010; 23: 1087-91.
19. Freeman S, Bean L, Allen E, et al. Ethnicity, sex, and the incidence of congenital heart defects: a report from the national Down syndrome project. *Genet Med.* 2008; 10: 173-80.
20. Burd L, Deal E, Rios R, et al. Congenital heart defects and fetal alcohol spectrum disorders. *Congenit Heart Dis.* 2007; 2: 250-55.
21. Alverson C, Strickland M, Gilboa S, et al. Maternal smoking and congenital heart defects in the Baltimore-Washington Infant Study. *Pediatrics.* 2011; 127: 647-53.
22. Lee L, Lupo P. Maternal smoking during pregnancy and the risk of congenital heart defects in offspring: a systematic review and metaanalysis. *Pediatr Cardiol.* 2013; 34: 398-407.
23. Louik C, Lin A, Werler M, et al. First-trimester use of selective serotonin-reuptake inhibitors and the risk of birth defects. *N Engl J Med.* 2007; 356: 2675-83.
24. Polen K, Rasmussen S, Riehle-Colarusso T, et al. Association between reported venlafaxine use in early pregnancy and birth defects, national birth defects prevention study, 1997-2007. *Birth Defects Res A Clin Mol Teratol.* 2013 Jan; 97(1):28-35.
25. Bakker M, Kerstjens-Frederikse W, Buys C, et al. First-trimester use of paroxetine and congenital heart defects: a population-based case-control study. *Birth Defects Res A Clin Mol Teratol.* 2010; 88: 94-100.
26. Correa A, Gilboa S, Besser L, et al. Diabetes mellitus and birth defects. *Am J Obstet Gynecol.* 2008; 199: 237 e231-39.
27. Parker S, Werler M, Shaw G, et al. Dietary glycemic index and the risk of birth defects. *Am J Epidemiol.* 2012; 176: 1110-20.
28. Miller A, Riehle-Colarusso T, Siffel C, et al. Maternal age and prevalence of isolated congenital heart defects in an urban area of the United States. *Am J Med Genet A.* 2011; 155A: 2137-45.
29. Reefhuis J, Honein M, Schieve L, et al. Assisted reproductive technology and major structural birth defects in the United States. *Hum Reprod.* 2009; 24: 360-66.
30. Van Praagh S, Carrera M, Sanders S, et al. Sinus venosus defects: unroofing of the right pulmonary veins-anatomic and echocardiographic findings and surgical treatment. *Am Heart J.* 1994; 128: 365-79.
31. Blom N, Gittenberger-de Groot A, Jongeneel T, et al. Normal development of the pulmonary veins in human embryos and formulation of a morphogenetic concept for sinus venosus defects. *Am J Cardiol.* 2001; 87: 305-09.
32. Banka P, Bacha E, Powell A, et al. Outcomes of inferior sinus venosus defect repair. *J Thorac Cardiovasc Surg.* 2011; 142:517-22.
33. Chin A, Murphy J. Identification of coronary sinus septal defect (unroofed coronary sinus) by color Doppler echocardiography. *Am Heart J.* 1992; 124: 1655-57.
34. Fuse S, Tomita H, Hatakeyama K, et al. Effect of size of a secundum atrial septal defect on shunt volume. *Am J Cardiol.* 2001; 88: 1447-50.

35. Walker R, Moran A, Gauvreau K, et al. Evidence of adverse ventricular interdependence in patients with atrial septal defects. *Am J Cardiol.* 2004; 93: 1374–77.
36. Masutani S, Senzaki H. Left ventricular function in adult patients with atrial septal defect: implication for development of heart failure after transcatheter closure. *J Card Fail.* 2011; 17: 957–63.
37. Hanslik A, Pospisil U, Salzer-Muhar U, et al. Predictors of spontaneous closure of isolated secundum atrial septal defect in children: a longitudinal study. *Pediatrics.* 2006; 1560–65.
38. Hager A, Eicken A, Lange R, et al. Need for closure of secundum atrial septal defect in infancy. *J Thorac Cardiovasc Surg.* 2005; 129: 1353–57.
39. Hanslik A, Pospisil U, Salzer-Muhar U, et al. Predictors of spontaneous closure of isolated secundum atrial septal defect in children: a longitudinal study. *Pediatrics.* 2006; 1560–65.
40. Helgason H, Jonsdottir G. Spontaneous closure of atrial septal defects. *Pediatr Cardiol.* 1999; 20: 195–99.
41. McMahon C, Feltes T, Fraley J, et al. Natural history of growth of secundum atrial septal defects and implications for transcatheter closure. *Heart.* 2002; 87: 256–59.
42. Andrews R, Tulloh R, Magee A, et al. Atrial septal defect with failure to thrive in infancy: hidden pulmonary vascular disease? *Pediatr Cardiol.* 2002; 23: 528–30.
43. Lammers A, Hager A, Eicken A, et al. Need for closure of secundum atrial septal defect in infancy. *Thorac Cardiovasc Surg.* 2005; 129: 1353–57.
44. Goetschmann S, Dibernardo S, Steinmann H, et al. Frequency of severe pulmonary hypertension complicating “isolated” atrial septal defect in infancy. *Am J Cardiol.* 2008; 102: 340–42.
45. Rhodes J, Patel H, Hijazi Z. Effect of transcatheter closure of atrial septal defect on the cardiopulmonary response to exercise. *Am J Cardiol.* 2002; 90: 803–06.
46. Lee Y, Jeng M, Tsao P, et al. Pulmonary function changes in children after transcatheter closure of atrial septal defect. *Pediatr Pulmonol.* 2009; 44: 1025–32.
47. Van De Bruaene A, Buys R, Vanhees L, et al. Cardiopulmonary exercise testing and SF-36 in patients with atrial septal defect type secundum. *J Cardiopulm Rehabil Prev.* 2011; 31: 308–15.
48. Berger F, Vogel M, Kramer A, et al. Incidence of atrial flutter/ fibrillation in adults with atrial septal defect before and after surgery. *Ann Thorac Surg.* 1999; 68: 75–78.
49. Nouira S, Kamoun I, Ouragini H, et al. Clinical and genetic investigation of atrial septal defect with atrioventricular conduction defect in a large consanguineous Tunisian family. *Arch Med Res.* 2008; 39:429–33.
50. Khoury G, Hawes C. Atrial septal defect associated with pulmonary hypertension in children living at high altitude. *J Pediatr.* 1967; 70: 432–35.
51. Humenberger M, Rosenhek R, Gabriel H, et al. Benefit of atrial septal defect closure in adults: impact of age. *Eur Heart J.* 2011; 553–60.
52. Yalonetsky S, Lorber A. Comparative changes of pulmonary artery pressure values and tricuspid valve regurgitation following transcatheter atrial septal defect closure in adults and the elderly. *Congenit Heart Dis.* 2009; 4: 17–20.
53. Steele P, Fuster V, Cohen M, et al. Isolated atrial septal defect with pulmonary vascular obstructive disease—long-term follow-up and prediction of outcome after surgical correction. *Circulation.* 1987; 76: 1037–4.
54. Sachweh J, Daebritz S, Hermanns B, et al. Hypertensive pulmonary vascular disease in adults with secundum or sinus venosus atrial septal defect. *Ann Thorac Surg.* 2006; 81: 207–13.
55. Campbell M. Natural history of atrial septal defect. *Br Heart J.* 1970; 820–26.
56. Murphy J, Gersh B, McGoan M, et al. Long-term outcome after surgical repair of isolated atrial septal defect. Follow-up at 27 to 32 years. *N Engl J Med.* 1990; 323: 1645–50.
57. Martín-Reyes R, Lopez-Fernandez T, Moreno-Yanguela M, et al: Role of real-time three-dimensional transesophageal echocardiography for guiding transcatheter patent foramen ovale closure. *Eur J Echocardiogr.* 2009; 10: 148–150.
58. PerK G, Lang R, Garcia-Fernandez M, et al: Use of real time three-dimensional transesophageal echocardiography in intracardiac catheter based interventions. *J Am Soc Echocardiogr.* 2009; 22: 865–882.
59. Van den Bosch A, Ten Harkel D, McGhie J, et al. Characterization of atrial septal defect assessed by real-time 3-dimensional echocardiography. *J Am Soc Echocardiogr.* 2006; 19: 815–21.

60. Rosenzweig B, Nayar A, Varkey M, et al. Echo contrast-enhanced diagnosis of atrial septal defect. *J Am Soc Echocardiogr.* 2001; 14: 155–57.
61. Valverde I, Simpson J, Schaeffter T, et al. 4D phase-contrast flow cardiovascular magnetic resonance: comprehensive quantification and visualization of flow dynamics in atrial septal defect and partial anomalous pulmonary venous return. *Pediatr Cardiol.* 2010; 31: 1244–48.
62. Teo K, Disney P, Dundon B, et al. Assessment of atrial septal defects in adults comparing cardiovascular magnetic resonance with transoesophageal echocardiography. *J Cardiovasc Magn Reson.* 2010; 12: 44.
63. Banka P, Bacha E, Powell A, et al. Outcomes of inferior sinus venosus defect repair. *J Thorac Cardiovasc Surg.* 2011;142:517–22.
64. Stout K, Daniels C, Aboulhosn J, et al. 2018 AHA/ACC guideline for the management of adults congenital heart disease: A report of the American College of Cardiology/ American Heart Association task force on clinical practice guidelines. *J Am Coll Cardiol.* 2018. pii: S0735-1097(18)36846-3.
65. Black M, Freedom R. Minimally invasive repair of atrial septal defects. *Ann Thorac Surg.* 1998; 65: 765-7.
66. Cremer J, Boning A, Anssar M, et al. Different approaches for minimally invasive closure of atrial septal defects. *Ann Thorac Surg.* 1999; 67: 1648-52.
67. Grinda J, Folliguet T, Dervanian P, et al. Right anterolateral thoracotomy for repair of atrial septal defect. *Ann Thorac Surg.* 1996; 62: 175-8.
68. Massetti M, Babatasi G, Rossi A, et al. Operation for atrial septal defect through a right anterolateral thoracotomy: current outcome. *Ann Thorac Surg.* 1996; 62: 1100-3.
69. Yoshimura N, Yamaguchi M, Oshima Y, et al. Repair of atrial septal defect through a right posterolateral thoracotomy: a cosmetic approach for female patients. *Ann Thorac Surg.* 2001; 72: 2103-5.
70. Min H, Yang J, Jun T, et al. Closure of atrial septal defects through a video-assisted mini-thoracotomy. *Korean J Thorac Cardiovasc Surg.* 2008; 41: 568-72.
71. Bonaros N, Schachner T, Oehlinger A, 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.
72. Ma Z, Dong M, Yin Q, et al. Totally thoracoscopic repair of atrial septal defect without robotic assistance: a single-center experience. *J Thorac Cardiovasc Surg.* 2011;141:1380-3.
73. Kim J, Jung S, Kim G, et al. Surgical outcomes of congenital atrial septal defect using da Vinci TM Surgical Robot System. *Korean J Thorac Cardiovasc Surg.* 2013; 46: 93-7.
74. Black M, Freedom R. Minimally invasive repair of atrial septal defects. *Ann Thorac Surg.* 1998; 65: 765 – 767.
75. Ewert P, Berger F, Daehnert I, et al. Transcatheter repair of atrial septal defect without fluoroscopy: Feasibility of new method. *Circulation.* 2000; 101: 847 – 849.
76. Ishikawa N, Watanabe G, Tomita S, et al. Robot-assisted minimally invasive direct coronary artery bypass grafting: ThoraCAB. *Circ J.* 2014; 78: 399 – 402.
77. Jeevan N, Hamad F, Bob K, et al. Minimally invasive endoscopic repair of atrial septal defects via right minithoracotomy, *Multimedia Manual of Cardio-Thoracic Surgery.* 2016 Feb 1; pii: mmv042.
78. Ak K, Aybek T, Wimmer-Greinecker G, et al. Evolution of surgical techniques for atrial septal defect repair in adults: a 10-year single-institution experience. *J Thorac Cardiovasc Surg.* 2007;134:757–764.
79. Grossi E, Loulmet D, Schwartz C, et al. Evolution of operative techniques and perfusion strategies for minimally invasive mitral valve repair. *J Thorac Cardiovasc Surg.* 2012;143: 68–70.
80. Chan E, Lumbao D, Iribarne A, et al. Evolution of cannulation techniques for minimally invasive cardiac surgery: a 10-year journey. *Innovations.* 2012; 7: 9–14.
81. Sabik J, Lytle B, McCarthy P, et al. Axillary artery: an alternative site of arterial cannulation for patients with extensive aortic and peripheral vascular disease. *J Thorac Cardiovasc Surg.* 1995;109:885–90.
82. Schachner T, Nagiller J, Zimmer A, et al. Technical problems and complications of axillary artery cannulation. *Eur J Cardiothorac Surg.* 2005; 27: 634–7.
83. Hendrickson S, Glower D. A method for perfusion of the leg during cardiopulmonary bypass via femoral cannulation. *Ann Thorac Surg.* 1998; 65(6):1807–8.
84. Jackson K, Timpa J, McIlwain R, et al. Side-arm grafts for femoral extracorporeal membrane oxygenation cannulation. *Ann Thorac Surg.* 2012; 94: 111–2.

85. Rosu C, Bouchard D, Pellerin M, et al. Pre-operative vascular imaging for predicting intraoperative modification of peripheral arterial cannulation during minimally invasive mitral valve surgery. *Innovations*. 2015; 10: 39–43.
86. Muhs B, Galloway A, Lombino M, et al. Arterial injuries from femoral artery cannulation with port access cardiac surgery. *Vasc Endovascular Surg*. 2005; 39: 153–8.
87. Gates J, Bichell D, Rizzo R, et al. Thigh ischemia complicating femoral vessel cannulation for cardiopulmonary bypass. *Ann Thorac Surg*. 1996; 61: 730–3.
88. Bisdas T, Beutel G, Warnecke G, et al. Vascular complications in patients undergoing femoral cannulation for extracorporeal membrane oxygenation support. *Ann Thorac Surg*. 2011; 92: 626–31.
89. Peng Z, MD, Yong S, MD, Qian Y, et al. Intraoperative device closure of atrial septal defects with minimal transthoracic invasion. *Tex Heart Inst J*. 2013; 40(3):256-60.
90. Wei X, Yi W, Xu X, et al. Transthoracic occlusion for secundum atrial septal defects unsuitable for transcatheter occlusion approach. *J Thorac Cardiovasc Surg*. 2011;142(1):113-9.
91. Sabin J, Aaron G, Ivan I, et al. Minimally invasive periareolar approach to unroofed coronary sinus atrial septal defect repair. *Ann Thorac Surg*. 2016; 102:223–5.
92. Torracca L, Ismeno G, Alfieri O. Total endoscopic computer-enhanced atrial septal defect repair in six patients. *Ann Thorac Surg*. 2001; 72: 1354 – 1357.
93. Norihiko I, Go W, Tatsuya T, et al. Two-port robotic cardiac surgery (TROCS) for atrial septal defect (ASD) using cross-arm technique. *Circ J*. 2015; 79: 2271 – 2273.
94. Hekmat K, Mehlhorn U, Rainer de V. Surgical repair of a large residual atrial septal defect after transcatheter repair. *Ann Thorac Surg*. 1997; 63: 1456 – 1458.
95. Bohm J, Bittigau K, Kohler F, et al. Surgical removal of atrial septal defect occlusion system-devices. *Eur J Cardiothorac Surg*. 1997; 12: 869 – 872.
96. Komar M, Przewłocki T, Olszowska M, et al. Conduction abnormality and arrhythmia after trans-catheter closure of atrial septal defect. *Circ J*. 2014; 78: 2415 – 2421.
97. Zhongmin W, Yuhao L, Yu X, et al. Three dimensional printing-guided percutaneous transcatheter closure of secundum atrial septal defect with rim deficiency: First-in-human series. *Cardiol J*. 2016;23:599-603.
98. Strickland R, Oliver W, Chantigian R, et al. Anesthesia, cardiopulmonary bypass, and the pregnant patient. *Mayo Clin Proc* .1991; 66: 411–429.
99. Pomini F, Mercogliano D, Cavalletti C, et al. Cardiopulmonary bypass in pregnancy. *Ann Thorac Surg*. 1996; 61: 259–268.
100. Abbas A, Lester S, Connolly H. Pregnancy and the cardiovascular system. *Int J Cardiol*. 2005; 98: 179–189.
101. Bedard E, Dimopoulos K, Gatzoulis M. Has there been any progress made on pregnancy outcomes among women with pulmonary arterial hypertension? *Eur Heart J*. 2009; 30: 256–265.
102. Soydemir D, Johnston T, Clarke B. Percutaneous closure of an atrial septal defect during pregnancy using an Amplatzer occlusion device. *J Obstet Gynaecol* 2005; 25: 715–716.