

13. BÖLÜM

DOĞUŞTAN DÜZELTİLMİŞ BÜYÜK ARTERLERİN TRANSPOZİSYONU

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GİRİŞ

Büyük arterlerin levo- veya L-ilmekli transpozisyonu (L-TGA), atriyo-ventriküler ve ventrikülo-arteriyel uyumsuzluk ile karakterize nadir bir konjenital kalp hastalığı formudur. Aynı zamanda genellikle doğuştan düzeltilmiş TGA, çift diskordans veya ventriküler inversiyon olarak da adlandırılır (Şekil 1). L-TGA, ilişkili kalp kusurları olmadıkça genellikle siyanoz ortaya çıkmaz. İzole L-TGA, oksijeni azalmış sistemik venöz kan pulmoner dolaşıma ve oksijenli pulmoner venöz kan sistemik dolaşıma döndüğü için «fizyolojik olarak düzeltilmiştir». L-TGA' lı yetişkin hastalar, sistemik sağ ventrikül fonksiyonundaki ilerleyici düşüş nedeniyle kalp yetmezliği riski altındadırlar.

Embriyoloji ve Anatomi

Kardiyak döngü oluşumu - Gebeliğin üçüncü haftasında düz kalp tüpünün döngülenmesi, kalbin dört odasının doğru anatomik hizalanması için anahtar embriyolojik işlemlerden biridir. Normal olarak, ilkel kalp tüpü sağa doğru (dekstro veya D halkası) “kıvrımlar” yaparak, sağ ventrikülün (SağV) sol ventrikülün (SolV) sağına normal morfolojik konumlanmasına neden olur. Bununla birlikte, sola döngü (levo veya L döngü), ventriküllerin anormal konumlanmasına ve kalbin atriyal, ventriküler ve arteriyel segmentleri arasında anormal bağlantılara yol açar. L-TGA' nın anatomisi atriyoventriküler ve ventriküloarteriyel uyumsuzluk ile karakterizedir. Koroner arter anatomisi değişkendir.

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KAYNAKLAR

1. Dyck JD, Atallah JA. Transposition of the great arteries. In: Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult, 7th ed, Allen HD, Shaddy RE, Driscoll DJ, Feltes TF (Eds), Wolters Kluwer Health/ Lippincott Williams & Wilkins, Philadelphia 2008. p.1087.
2. Warnes CA. Transposition of the great arteries. *Circulation*. 2006;114(24):2699-2709. doi: 10.1161/CIRCULATIONAHA.105.592352.
3. Hornung TS, Calder L. Congenitally corrected transposition of the great arteries. *Heart*. 2010;96(14):1154-1161. doi: 10.1136/hrt.2008.150532.
4. Ferencz C, Rubin JD, McCarter RJ, et al. Congenital heart disease: prevalence at live-birth. The Baltimore-Washington Infant Study. *Am J Epidemiol*. 1985;121(1):31-36. doi: 10.1093/oxfordjournals.aje.a113979.
5. Bravo-Valenzuela NJ, Peixoto AB, Araujo Júnior E. Prenatal diagnosis of transposition of the great arteries: an updated review. *Ultrasonography*. 2020;39(4):331-339. doi: 10.14366/usg.20055.
6. Samánek M, Vorísková M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemia survival study. *Pediatr Cardiol*. 1999;20(6):411-417. doi: 10.1007/s002469900502.
7. Kuehl KS, Loffredo CA. Population-based study of l-transposition of the great arteries: possible associations with environmental factors. *Birth Defects Res A Clin Mol Teratol*. 2003;67(3):162-167. doi: 10.1002/bdra.10015.
8. Becker TA, Van Amber R, Moller JH, et al. Occurrence of cardiac malformations in relatives of children with transposition of the great arteries. *Am J Med Genet*. 1996;66(1):28-32. doi: 10.1002/(SICI)1096-8628(19961202)66:1<28::AID-AJMG7>3.0.CO;2-S.
9. Hornung TS, Kilner PJ, Davlouros PA, et al. Excessive right ventricular hypertrophic response in adults with the mustard procedure for transposition of the great arteries. *Am J Cardiol*. 2002;90(7):800-803. doi: 10.1016/s0002-9149(02)02619-x.
10. Kral Kollars CA, Gelehrter S, Bove EL, et al. Effects of morphologic left ventricular pressure on right ventricular geometry and tricuspid valve regurgitation in patients with congenitally corrected transposition of the great arteries. *Am J Cardiol*. 2010;105(5):735-739. doi: 10.1016/j.amjcard.2009.10.066.
11. Hornung TS, Bernard EJ, Celermajer DS, et al. Right ventricular dysfunction in congenitally corrected transposition of the great arteries. *Am J Cardiol*. 1999;84(9):1116-1119. doi: 10.1016/s0002-9149(99)00516-0.
12. Wan AW, Jevremovic A, Selamet Tierney ES, et al. Comparison of impact of prenatal versus postnatal diagnosis of congenitally corrected transposition of the great arteries. *Am J Cardiol*. 2009;104(9):1276-1279. doi: 10.1016/j.amjcard.2009.06.047.
13. Graham TP Jr, Bernard YD, Mellen BG, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol*. 2000;36(1):255-261. doi: 10.1016/s0735-1097(00)00682-3.
14. Presbitero P, Somerville J, Rabajoli F, et al. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. *Br Heart J*. 1995;74(1):57-59. doi: 10.1136/hrt.74.1.57.

15. Keane JF, Fyler DC. "Corrected" transposition of the great arteries. In: Nadas' Pediatric Cardiology, 2nd ed, Keane JF, Lock JE, Fyler DC (Eds), Saunders, Philadelphia 2006. p.791.
16. Prieto LR, Hordof AJ, Secic M, et al. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. *Circulation*. 1998;98(10):997-1005. doi: 10.1161/01.cir.98.10.997.
17. Beauchesne LM, Warnes CA, Connolly HM, et al. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. *J Am Coll Cardiol*. 2002;40(2):285-290. doi: 10.1016/s0735-1097(02)01952-6.
18. Murtuza B, Barron DJ, Stumper O, et al. Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience. *J Thorac Cardiovasc Surg*. 2011;142(6):1348-1357.e1. doi: 10.1016/j.jtcvs.2011.08.016.
19. Friedberg MK, Silverman NH, Moon-Grady AJ, et al. Prenatal detection of congenital heart disease. *J Pediatr*. 2009;155(1):26-31. doi: 10.1016/j.jpeds.2009.01.050.
20. Mongeon FP, Connolly HM, Dearani JA, et al. Congenitally corrected transposition of the great arteries ventricular function at the time of systemic atrioventricular valve replacement predicts long-term ventricular function. *J Am Coll Cardiol*. 2011;57(20):2008-2017. doi: 10.1016/j.jacc.2010.11.021.
21. Winlaw DS, McQuirk SP, Balmer C, et al. Intention-to-treat analysis of pulmonary artery banding in conditions with a morphological right ventricle in the systemic circulation with a view to anatomic biventricular repair. *Circulation*. 2005;111(4):405-411. doi: 10.1161/01.CIR.0000153355.92687.FA.
22. Helvind MH, McCarthy JF, Imamura M, et al. Ventriculo-arterial discordance: switching the morphologically left ventricle into the systemic circulation after 3 months of age. *Eur J Cardiothorac Surg*. 1998;14(2):173-178. doi: 10.1016/s1010-7940(98)00172-9.
23. Quinn DW, McQuirk SP, Metha C, et al. The morphologic left ventricle that requires training by means of pulmonary artery banding before the double-switch procedure for congenitally corrected transposition of the great arteries is at risk of late dysfunction. *J Thorac Cardiovasc Surg*. 2008;135(5):1137-1144. doi: 10.1016/j.jtcvs.2008.02.017.
24. Myers PO, del Nido PJ, Geva T, et al. Impact of age and duration of banding on left ventricular preparation before anatomic repair for congenitally corrected transposition of the great arteries. *Ann Thorac Surg*. 2013;96(2):603-610. doi: 10.1016/j.athoracsur.2013.03.096.
25. Ma K, Gao H, Hua Z, et al. Palliative pulmonary artery banding versus anatomic correction for congenitally corrected transposition of the great arteries with regressed morphologic left ventricle: long-term results from a single center. *J Thorac Cardiovasc Surg*. 2014;148(4):1566-1571. doi: 10.1016/j.jtcvs.2013.12.044.
26. Devaney EJ, Charpie JR, Ohye RG, et al. Combined arterial switch and Senning operation for congenitally corrected transposition of the great arteries: patient selection and intermediate results. *J Thorac Cardiovasc Surg*. 2003;125(3):500-507. doi: 10.1067/mtc.2003.158.

27. Poirier NC, Yu JH, Brizard CP, et al. Long-term results of left ventricular reconditioning and anatomic correction for systemic right ventricular dysfunction after atrial switch procedures. *J Thorac Cardiovasc Surg.* 2004;127(4):975-981. doi: 10.1016/j.jtcvs.2003.10.024.
28. Ly M, Belli E, Leobon B, et al. Results of the double switch operation for congenitally corrected transposition of the great arteries. *Eur J Cardiothorac Surg.* 2009;35(5):879-883; discussion 883-884. doi: 10.1016/j.ejcts.2009.01.051.
29. Mattes A, Haun C, Blaschczok HC, et al. Functional outcome of anatomic correction of corrected transposition of the great arteries. *Eur J Cardiothorac Surg.* 2011;40(5):1227-1234. doi: 10.1016/j.ejcts.2011.01.077.
30. Langley SM, Winlaw DS, Stumper O, et al. Midterm results after restoration of the morphologically left ventricle to the systemic circulation in patients with congenitally corrected transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2003;125(6):1229-1241. doi: 10.1016/s0022-5223(02)73246-7.
31. Bautista-Hernandez V, Marx GR, Gauvreau K, et al. Determinants of left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries. *Ann Thorac Surg.* 2006;82(6):2059-2065; discussion 2065-2066. doi: 10.1016/j.athoracsur.2006.06.045.
32. Hraska V, Duncan BW, Mayer JE Jr, et al. Long-term outcome of surgically treated patients with corrected transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2005;129(1):182-191. doi: 10.1016/j.jtcvs.2004.02.046.
33. Rutledge JM, Nihill MR, Fraser CD, et al. Outcome of 121 patients with congenitally corrected transposition of the great arteries. *Pediatr Cardiol.* 2002;23(2):137-145. doi: 10.1007/s00246-001-0037-8.
34. Hraska V, Vergnat M, Zartner P, et al. Promising Outcome of Anatomic Correction of Corrected Transposition of the Great Arteries. *Ann Thorac Surg.* 2017;104(2):650-656. doi: 10.1016/j.athoracsur.2017.04.050.
35. Hiramatsu T, Matsumura G, Konuma T, et al. Long-term prognosis of double-switch operation for congenitally corrected transposition of the great arteries. *Eur J Cardiothorac Surg.* 2012;42(6):1004-1008. doi: 10.1093/ejcts/ezs118. Erratum in: *Eur J Cardiothorac Surg.* 2013;43(3):663.
36. Malhotra SP, Reddy VM, Qiu M, et al. The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: rationale and midterm results. *J Thorac Cardiovasc Surg.* 2011;141(1):162-170. doi: 10.1016/j.jtcvs.2010.08.063.
37. Bautista-Hernandez V, Myers PO, Cecchin F, et al. Late left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2014;148(1):254-258. doi: 10.1016/j.jtcvs.2013.08.047.
38. Mainwaring RD, Patrick WL, Ibrahimiyeh AN, et al. An Analysis of Left Ventricular Retraining in Patients With Dextro- and Levo-Transposition of the Great Arteries. *Ann Thorac Surg.* 2018;105(3):823-829. doi: 10.1016/j.athoracsur.2017.11.047.
39. Schwartz ML, Gauvreau K, del Nido P, et al. Long-term predictors of aortic root dilation and aortic regurgitation after arterial switch operation. *Circulation.* 2004;110(11 Suppl 1):II128-II132. doi: 10.1161/01.CIR.0000138392.68841.d3.

40. Brizard CP, Lee A, Zannino D, et al. Long-term results of anatomic correction for congenitally corrected transposition of the great arteries: A 19-year experience. *J Thorac Cardiovasc Surg.* 2017;154(1):256-265. doi: 10.1016/j.jtcvs.2017.03.072.
41. Cotts T, Malviya S, Goldberg C. Quality of life and perceived health status in adults with congenitally corrected transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2012;143(4):885-890. doi: 10.1016/j.jtcvs.2011.08.031.
42. Fredriksen PM, Chen A, Veldtman G, et al. Exercise capacity in adult patients with congenitally corrected transposition of the great arteries. *Heart.* 2001;85(2):191-195. doi: 10.1136/heart.85.2.191.
43. Gaies MG, Watnick CS, Gurney JG, et al. Health-related quality of life in patients with congenitally corrected transposition of the great arteries. *J Thorac Cardiovasc Surg.* 2011;142(1):136-141. doi: 10.1016/j.jtcvs.2010.11.035.
44. Van Hare GF, Ackerman MJ, Evangelista JA, et al. Eligibility and Disqualification Recommendations for Competitive Athletes With Cardiovascular Abnormalities: Task Force 4: Congenital Heart Disease: A Scientific Statement From the American Heart Association and American College of Cardiology. *Circulation.* 2015;132(22):e281-e291. doi: 10.1161/CIR.0000000000000240.
45. Connolly HM, Grogan M, Warnes CA. Pregnancy among women with congenitally corrected transposition of great arteries. *J Am Coll Cardiol.* 1999;33(6):1692-1695. doi: 10.1016/s0735-1097(99)00046-7.
46. Therrien J, Barnes I, Somerville J. Outcome of pregnancy in patients with congenitally corrected transposition of the great arteries. *Am J Cardiol.* 1999;84(7):820-824. doi: 10.1016/s0002-9149(99)00444-0.