



BÖLÜM | 25

Primer ve Sekonder Kardiyomiyopatiler ve Acil Durumlar

Yusuf DEMİR¹

Giriş

Kardiyomiyopatinin tanımı ve sınıflandırılması zaman içinde değişiklik göstermekle birlikte temel olarak koroner arter hastalığı, hipertansiyon, kalp kapak hastalığı, konjenital kalp hastalığının eşlik etmediği, kalp boyutunda ve duvar kalınlıklarında kalınlaşma, incelme veya kalbin fonksiyonel kontraktıl işlev bozukluğu ile karakterize geniş bir yelpazeye sahip kalp kası hastalıkları grubudur. 2006 yılında Amerikan Kalp Cemiyeti (AHA) tanımına göre; Kardiyomiyopatiler, genellikle uygun olmayan ventriküler hipertrofi veya kalp boşluklarında dilatasyon gösteren ve çoğu zaman genetik temele dayanan, mekanik ve/veya elektriksel işlev bozukluğu ile ilişkili olan, miyokardın heterojen bir hastalık grubudur (1). Yine 2008 yılında Avrupa Kardiyoloji Derneği (ESC) kardiyomiyopatiyi; kalp kasının yapısal ve fonksiyonel olarak anormal olduğu kalp kası hastalığı olarak tanımlamıştır (2).

Sınıflama

Klinik uygulamada etiyoloji ve patofizyoloji net olmadığından kardiyomiyopatilerin sınıflandırılmasında fikir birliği hala tam olarak sağlanamamıştır (3). Kardiyomiyopatileri 2006 yılında Amerikan kalp cemiyeti Primer kardiyomiyopatiler ve sekonder kardiyomiyopatiler olarak iki ana kategoride incelemektedir (Tablo 1). Primer kardiyomiyopatiler; sadece veya daha çok kalp kasını ilgilendiren ve daha az görülen hastalıklar (genetik, genetik dışı, kazanılmış), daha sık olan sekonder kardiyomiyopatiler ise; sistemik hastalığın bir parçası olarak ortaya çıkan patolojik miyokard tutulumunu kapsayan kardiyomiyopatiler olarak tanımlanmıştır (1).

¹ Uzm. Dr., Menemen Devlet Hastanesi, yusufdemir2502@gmail.com

ları olan bir hastalıktır. Ekg de özellikle göğüs derivasyonlarında ST segment elevasyonu görülür.

Özellikle ST elevasyonu varlığında takotsuba kardiyomiyopatisi ve akut koroner sendrom ayrımı yapılması mümkün değildir. Bu nedenle bu hastalara koroner anjiyografi gerekmektedir. Ve bu hastalara ayırım yapılamadığından ST elevasyonlu MI protokolü uygulanmalıdır. Hastaların koroner anjiyografileri normal veya ciddi olmayan koroner arter darlığı olarak görülür. Ventrikülografide apikal balonlaşma görülür.

Kaynaklar

1. Maron BJ, Towbin JA, Thiene G, Antzelevitch C, Corrado D, Arnett D, ve ark. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation*. 2006;113(14):1807-16.
2. Elliott P, Andersson B, Arbustini E, Bilinska Z, Cecchi F, Charron P, ve ark. Classification of the cardiomyopathies: a position statement from the European Society Of Cardiology Working Group on Myocardial and Pericardial Diseases. *Eur Heart J*. 2008;29(2):270-6.
3. Sisakian H. Cardiomyopathies: Evolution of pathogenesis concepts and potential for new therapies. *World J Cardiol*. 2014;6(6):478-94.
4. Maron BJ, Hypertrophic cardiomyopathy: A systematic review. *JAMA*. 2002; 287 : 1308.
5. Teare D, Asymmetrical hypertrophy of the heart in young adults. *Br Heart J*. 1958; 20: 1- 18
6. Kamil Adalet. Klinik Kardiyoloji. II. Baskı. Kısım IX, miyokardit ve kardiyomiyopatiler. 721-749
7. Braunwald Kalp Hastalıkları. IX. Baskı. Kısım XXIV, Bölüm 69. Hipertrofik kardiyomiyopati. 1582-1595
8. Brian P Griffin. IV. Baskı. Kardiyovasküler Hastalıklar. Kısım II, bölüm 10. Hipertrofik Kardiyomiyopati. 160-176
9. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy
10. Spirito P, Seidman CE, Mc Kenna WJ, et al. The management of hypertrophic cardiomyopathy. *N Engl J Med*. 1997; 336(11): 775-85
11. Maron BJ. The 2009 international hypertrophic cardiomyopathy summit. *Am J Cardiol*. 2010;105(8):1164-8
12. Hess MO, Mc Kenna W, Schultheiss with co-authors Hullin R, Kühl U, Pauschinger M, Noutsias M, Sen- Chowdhry S. Myocardial disease. In Camm JA, Lüscher FT, Serruys WP eds. *The ESC textbook of cardiovascular medicine*. 1st ed.:Blackwell publishing. 2006, p.453-515.
13. Maron BJ, Nishimura RA, Danielson GK. Pitfalls in clinical recognition and a novel operative approach for hypertrophic cardiomyopathy with severe outflow obstruction due to anomalous papillary muscle. *Circulation*. 1998 ;98(23):2505-8.
14. Levine RA, Vlahakes GJ, Lefebvre X, et al. Papillary muscle displacement causes systolic anterior motion of the mitral valve. Experimental validation and insights into the mechanism of subaortic obstruction. *Circulation*. 1995;91(4):1189-95.
15. Sherrid MV, Gunsburg DZ, Moldenhauer S, et al. Systolic anterior motion begins at low left ventricular outflow tract velocity in obstructive hypertrophic cardiomyopathy. *J Am Coll*

- Cardiol. 2000; 36: 1344-54. 47
15. Grigg LE, Wigle ED, Williams WG, Daniel LB, Rakowski H. Transesophageal Doppler echocardiography in obstructive hypertrophic cardiomyopathy. Clarification of pathophysiology and importance in intraoperative decision making. *J Am Coll Cardiol* 1992;20:42-52.
 16. Maron BJ, McKenna WJ, Danielson GK et al. American College of Cardiology/European Society of Cardiology Clinical Expert Consensus Document on Hypertrophic Cardiomyopathy. A Report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines and Policy Conferences. *J Am Coll Cardiol.* 2003;42:1587-713.
 17. B. J. Gersh et al., "2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: A report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines Developed in Collaboration with the American Ass," *J. Am. Coll. Cardiol.*, vol. 58, no. 25, pp. e212–e260, 2011, doi: 10.1016/j.jacc.2011.06.011.
 18. B. Moore, C. Semsarian, K. H. Chan, and R. W. Sy, "Sudden Cardiac Death and Ventricular Arrhythmias in Hypertrophic Cardiomyopathy," *Hear. Lung Circ.*, vol. 28, no. 1, pp. 146–154, 2019, doi: 10.1016/j.hlc.2018.07.019.
 19. K. R. Kumar, S. N. Mandleywala, and M. S. Link, "Atrial and Ventricular Arrhythmias in Hypertrophic Cardiomyopathy," *Card. Electrophysiol. Clin.*, vol. 7, no. 2, pp. 173–186, 2015, doi: 10.1016/j.ccep.2015.03.002.
 20. J. L. Zamorano et al., "2014 ESC guidelines on diagnosis and management of hypertrophic cardiomyopathy: The task force for the diagnosis and management of hypertrophic cardiomyopathy of the European Society of Cardiology (ESC)," *Eur. Heart J.*, vol. 35, no. 39, pp. 2733–2779, 2014, doi: 10.1093/eurheartj/ehu284.
 21. Sakamoto T. Apical hypertrophic cardiomyopathy (apical hypertrophy): An overview. *J Cardiol.* 2001;37(Suppl 1):161-20
 22. Maron BJ. Hypertrophic cardiomyopathy: A systematic review. *JAMA.* 2002;287:1308-20.
 23. Maron BJ, McKenna WJ, Danielson GK et al. American College of Cardiology/European Society of Cardiology Clinical Expert Consensus Document on Hypertrophic Cardiomyopathy. A Report of the American College of Cardiology Task Force on Clinical Expert Consensus Documents and the European Society of Cardiology Committee for Practice Guidelines and Policy Conferences. *J Am Coll Cardiol.* 2003;42:1587-713.
 24. Pasternac A, Noble J, Streulens Y, et al. Pathophysiology of chest pain in patients with cardiomyopathies and normal coronary arteries. *Circulation.* 1982;65:778-89.
 25. Petersen SE, Jerosch-Herold M, Hudsmith LE, et al. Evidence for microvascular dysfunction in hypertrophic cardiomyopathy: new insights from multiparametric magnetic resonance imaging. *Circulation.* 2007;115:2418- 25.
 26. Choudhury L, Mahrholdt H, Wagner A, et al. Myocardial scarring in asymptomatic or mildly symptomatic patients with hypertrophic cardiomyopathy. *J Am Coll Cardiol.* 2002;40: 2156-64.
 27. Authors/Task Force members, Elliot PM, Anastasakis A, Borger MA, Borriggreffe M, Cecchi F, Charron P, Hagege AA, Lafont A, Limongelli G, Mahrholdt H, McKenna WJ, Mogensen J, Nihoyannopoulos P, Nistri S, Pieper PG, Pieske B, Rapezzi C, Rutten FH, Tillmanns C, Watkins H. 2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy: The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC). *Eur Heart J.* 2014;35(39):2733-79
 28. Rosenkranz S, Flesch M, Amann K, et al. Alterations of β adrenergic signaling and cardiac hypertrophy in transgenic mice overexpressing TGF- β (1). *Am J Physiol Heart Circ Physiol.* 2002;283:1253–1262.
 29. Debonnaire, P., Katsanos, S., Joyce, E., Van den Brink, O. V., Atsma, D. E., Schalij, M. J., ... &

- Marsan, N. A. (2015). QRS fragmentation and QTc duration relate to malignant ventricular tachyarrhythmias and sudden cardiac death in patients with hypertrophic cardiomyopathy. *Journal of cardiovascular electrophysiology*, 26(5), 547-555
30. McKenna WJ, Cocco F, Elliot PM. Genese and disease expression in hypertrophic cardiomyopathy, *Lancet*, 1998;10;1162-3
 31. Thiene G, Nava A, Corrado D, Rossi L, Pennelli N. Right ventricular cardiomyopathy and sudden death in young people. *N Engl J Med* 1988;318:129-33
 32. Maron BJ, Towbin JA, Thiene G, et al. Contemporary definitions and classification of the cardiomyopathies: an American Heart Association Scientific Statement from the Council on Clinical Cardiology, Heart Failure and Transplantation Committee; Quality of Care and Outcomes Research and Functional Genomics and Translational Biology Interdisciplinary Working Groups; and Council on Epidemiology and Prevention. *Circulation*. 2006;113(14):1807-16.
 33. Ulus T, Okyay K, Kabul HK, Özcan EE, Özeke Ö, Altay H, et al. Turkish Society of Cardiology consensus paper on management of arrhythmia-induced cardiomyopathy. *Anatol j Cardiol*. 2019;21(2):98-106.
 34. gopinathnair R, Etheridge Sp, marchlin- ski FE, Spinale Fg, Lakkireddy D, Olshan- sky B. Arrhythmia-induced cardiomyopathies: mechanisms, recognition, and management. *j Am Coll Cardiol*. 2015;66(15):1714- 28.
 35. Baman TS, Lange DC, ilg Kj, gupta SK, Liu TY, Alguire C, et al. Relationship between burden of premature ventricular complexes and left ventricular function. *Heart Rhythm*. 2010;7(7):865-9.
 36. Moore jp, patel pA, Shannon Km, Albers EL, Salerno jC, Stein mA, et al. predictors of myocardial recovery in pediatric tachycardia-induced cardiomyopathy. *Heart Rhythm*. 2014; 11(7):1163-9.
 37. Hasdemir C, Ulucan C, Yavuzgil O, Yuksel A, Kartal Y, Simsek E, et al. Tachycardia-induced cardiomyopathy in patients with idiopathic ventricular arrhythmias: the incidence, clinical and electrophysiologic characteristics, and the predictors. *j Cardiovasc Electrophysiol*. 2011;22:663-8.
 38. ponikowski p, Voors AA, Anker SD, Bueno H, Cleland jg, Coats Aj, et al.; Authors/Task Force members; Document Reviewers. 2016 ESC guidelines for the diagnosis and treatment of acute and chronic heart failure: The Task Force for the diagnosis and treatment of acute and chronic heart failure of the European Society of Cardiology (ESC). Developed with the special contribution of the Heart Failure Association (HFA) of the ESC. *Eur j Heart Fail*. 2016;18:891-975.
 39. Sliwa K, Hilfiker-Kleiner D, Petrie mc, mebazaa A, Pieske B, Buchmann E, et al. current state of knowledge on aetiology, diagnosis, management, and therapy of peripartum cardiomyopathy: a position statement from the Heart Failure Association of the European Society of cardiology Working group on peripartum cardiomyopathy. *Eur j Heart Fail*. 2010;12:767-78.
 40. Elliott P, Andersson B, Arbustini E, Bilinska Z, Cecchi F, Charron P, et al. Classification of the cardiomyopathies: a position statement from the European Society of Cardiology Working Group on Myocardial and Pericardial Diseases. *Eur Heart J*. 2008;29 (2):270-6.
 41. Towbin JA, Lorts A, Jefferies JL. Left Ventricular Non-compaction Cardiomyopathy. *Lancet*. 2015;386(9995):813-25.
 42. Sato H. Tako-tsubo-like left ventricular dysfunction due to multivessel coronary spasm. In: K Kodama, K, Haze M Hori, eds. *Clinical Aspect of Myocardial Injury: From Ischemia to Heart Failure*. Tokyo: Kagakuhyoronsha publishing Co; 1990. p56-64.