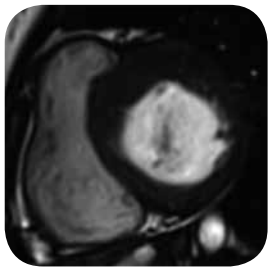

Left Ventricular Outflow Tract Obstruction

Obstrucción del tracto de salida del ventrículo izquierdo

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Key words (MeSH)

Magnetic resonance imaging
Heart
Heart ventricles

Summary

The left ventricular outflow tract (LVOT) is the anatomic structure through which the left ventricular stroke volume passes towards the aorta. The LVOT consists of three components: subvalvular component, which is delimited by the membranous and basal muscular portions of the interventricular septum; valvular component (the aortic valve); and supravalvular component. This academic review evaluates different obstructive pathologies of the LVOT, including entities located at the aortic valve level (valvular), in the ascending aorta (supravalvular), and in the proximal portion of the LVOT (subvalvular).

Palabras clave (DeCS)

Imagen por resonancia magnética
Corazón
Ventrículos cardiacos

Resumen

El tracto de salida del ventrículo izquierdo (TSVI) es la estructura anatómica a través de la cual sale el flujo sistólico del ventrículo izquierdo (VI) hacia la aorta. El TSVI está conformado por tres componentes: subvalvular, el cual es delimitado por el septo interventricular en sus porciones membranosas y muscular basal y la valva anterior de la válvula mitral; el componente valvular, corresponde a la válvula aórtica, y el supravalvular. En esta revisión académica se evaluarán las patologías obstructivas del tracto de salida del ventrículo izquierdo, incluyendo entidades localizadas en el sector valvular aórtico (valvulares), en la aorta ascendente (supravalvulares) y en el tracto de salida del ventrículo izquierdo proximal al plano valvular (subvalvulares).

Subvalvular obstruction

The subvalvular left ventricular outflow tract (LVOT) is delimited by the interventricular septum in its basal membranous and muscular portion and fibrous continuity tissue between the aortic valves and the mitral valve (anterior valve to the mitral valve). Through these structures passes the systolic flow of the left ventricle before reaching the aortic valve. Both components are highly mobile so the subvalvular LVOT area may be modified during the cardiac cycle (1).

Aortic subvalvular obstruction may be the consequence of different pathophysiological mechanisms: dynamics, such as those that can present after mitral and aortic valve surgery; mixed, as usually occurs in hypertrophic cardiomyopathy and mechanisms in which a fixed anatomical substrate prevails as in the membranes or the diaphragms.

Hypertrophic cardiomyopathy (HCM)

The accepted clinical definition for HCM is left ventricular (LV) hypertrophy associated to a non-dilated ventricular chamber, a condition that can't be explained by heart or systemic disease (2-5). HCM is caused by mutations in the genes encoding cardiac sarcomeres proteins. It is an autosomal dominant disease with variable expression and penetrance. The usual morphological phenotype is the thickening of the wall of the left ventricle. However, because of the multiplicity of genetic mutations and their transmission, the cardiac in-

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volvement may vary and rarely appears with normal wall thickness of the LV or with compromise of the right ventricle (RV) (4). Accepted diagnostic criteria for HCM consist of a maximum wall thickness of LV ≥15 mm and a ratio between the thickness of the septal wall and that of the lower wall higher than 1.5. In children, a thickness of the wall ≥ 2 standard deviations above the mean (score z ≥2) is used (6). The clinical presentation of HCM is varied and in all age groups, the majority of patients have a normal life expectancy without disability or need for mayor therapeutical procedures. When manifestations occur clinical symptoms usually come in three forms: sudden death due to malignant arrhythmias (more common in children under 35 years and athletes), heart failure with exertional dyspnea and paroxysmal or chronic atrial fibrillation (6). The HCM has different subtypes: septal, asymmetric, midventricular, apical and concentric, among others (7,8).

The mechanism causing obstruction of the outflow tract of the LV at the HCM is mixed:

1. Anatomical substrate

- Protrusion of the hypertrophic basal interventricular septum towards the exit tract (figure 1).
- Anomalies of the valvular and subvalvular mitral apparatus, including elongated mitral valves, small mitral annulus or endocardial thickening of the ventricular surface of the anterior valve (9).
- Subvalvular abnormalities include hypertrophy and variants in the papillary muscles, which can be bifid or have variation in the number of implantation, as in the case of short tendon cords or direct insertion of the papillary muscles in the mitral valves (10). The anterolateral displacement of the papillary muscles may contribute to the obstruction of the outflow tract. These alterations may produce obstruction even in patients with ventricular wall thickness very close to normal (11,12).

2. Proposed dynamic mechanisms

- Protrusion of the hypertrophic septum generates acceleration of flow in the LVOT and reduction of lateral pressure, triggering a venturi effect that "attracts" the anterior valve of the mitral valve and causes it to contact the interventricular septum accentuating the obstructive effect. This phenomenon is known as systolic anterior movement (SAM) of the anterior valve of the mitral valve (Figure 1b and d). When SAM is present, it produces flow of mitral insufficiency (13-16).
- The second mechanism suggests an abnormal ventricular morphology with an unusual relationship of the output and input tracts of the LV. A decrease in the aortomitral angle of projection projects the valvular and subvalvular mitral apparatus to the LVOT exposing the valve of the mitral valve to the outflow of the LV and favoring its displacement (13).
- Recently it has been proposed that the aortoseptal angulation in the projection of three chambers is important in the obstruction of the LVOT and can be used as a predictor of inducible obstruction in symptomatic patients who do not have a significant gradient of the resting tract at rest (13,17). The aortoseptal angle is defined as that formed between a line drawn along the border of the left and right interventricular septum and a line along the longitudinal axis of the aortic root. A value of 180 ° represents a straight line from the septum to the aorta and lower values indicate an increase in angulation. In the study conducted by Critoph et al., patients with HCM had a lower aortoseptal angle than controls (113 ± 12 vs. 126 ± 6, respectively) (13,18,19).

Obstruction of the left ventricular outflow tract (LVOTO) caused by SAM is found in one-third of patients with HCM at rest (13, 15) and occurs in up to two thirds of symptomatic patients which have no obstruction at rest but do so during manoeuvres that reduce preload and afterload or increase contractility (20). LVOTO in patients with HCM is considered clinically significant when a gradient greater than 30 mm Hg at rest or greater than 50 mm Hg is encountered during manoeuvres by echocardiography, such as exercise, Valsalva or pharmacological stress (21). Surgical intervention is recommended in patients who have not responded to pharmacological management. Septal ablation with alcohol is recommended in patients who are not candidates for surgery and older patients regardless of whether or not they are candidates for surgical management (6).

Transthoracic (TT) or transesophageal (TE) echocardiography with Doppler is usually the first test performed when there is suspicion of LVOTO or HCM, for the identification and classification of the severity of subvalvular stenosis according to the pressure gradient. Cardiac MRI may provide important additional information to define in detail the anatomy of the LVOT, including the degree of hypertrophy and the precise location of the thickening of the myocardium, evaluation of the valvular and subvalvular mitral apparatus and presence of myocardial fibrosis that is manifested in late enhancement images in inversion sequences (22). Cardiac MRI can also be used to measure the area of the hole of LVOTO, which is determined by transplanar flow planimetry using phase contrast sequences (figure 1f), which has demonstrated to be a reliable parameter for characterizing patients with obstructive HCM. Contrary to routinely used parameters on echocardiography, MRI is free of interobserver variability, conditions of image variability and is a non-invasive method (22-24). Measurement of the tract area by MRI is useful to define the degree of obstruction. An outflow tract <2.7 cm^2 during systole, with cardiac MR, has a sensitivity and specificity of 100% to differentiate obstructive and non-obstructive form of HCM (25,26). The main role of MRI in patients with HCM is to clarify the diagnosis and phenotype (6,27-31). In patients in whom echocardiography is inconclusive usually through poor acoustic window or when hypertrophy is located in regions that are not easily visualized by this method, cardiac MRI should be performed (6,32). Late enhancement sequences allow the identification of areas of myocardial fibrosis in patients with HCM. Patients with evidence of myocardial fibrosis and CMR have been associated with risk markers for sudden death, malignant ventricular arrhythmias and electrocardiographic disorders (33-38).

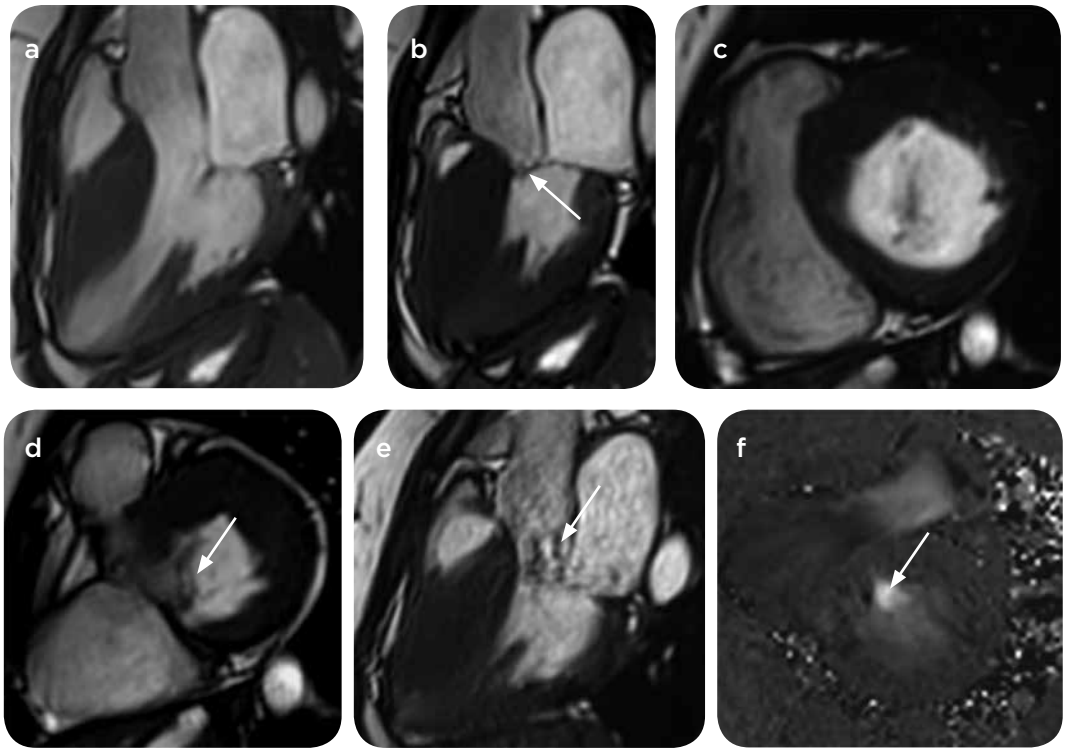

Figure 1. 35-year-old patient with predominantly anteroseptal basal HCM and obstruction of the LVOT due to hypertrophy of the basal septum and SAM. CMR, SSFP (steadystate Free precession) sequences in different phases of the cardiac cycle, long axis projection of three chambers (a and b) and short axis in the Mitral valve plane (c and d) where systolic blood pressure movement (SAM) is observed both of the anterior valve as well as the posterior leaflet (arrows in b and d) which determines the contact of the same with the interventricular septum. Note the normal location of the leaflets (central plane) in the others phases of the cardiac cycle (a and c). Immediately after SAM note the acceleration of flow in the LVOT (arrow on e). Phase contrast sequence in a plane perpendicular to the tract to determine peak speed and area of the outflow tract in systole (arrow on f).

LVOTO by membranes, diaphragms and subvalvular infundibulums

They may have different morphologies; the most frequent form is a fibroelastic tissue membrane or spine that extends from the endocardial surface of the basal septum and protrudes to the outflow tract, being able to extend to the base of the anterior valve of the Mitral valve (Figure 2). Indications for surgical treatment of patients with membranes, diaphragms or infundibuli of the ventricular outflow tract depend on the clinical manifestations, the gradient pressure and morphology of the lesions (1,12).

LVOTO in patients after closure of ventricular septal defect (VSD)

La obstrucción subvalvular también se ha informado luego del cieSubvalvular obstruction has also been reported after surgical closure of ventricular septal defects and is believed to be due to proliferation of fibrotic tissue at turbulent flow sites (39). The membranes located immediately adjacent to the aortic valve or extending to the anterior Mitral valve are more likely to lead to progressive obstruction and aortic valve injury with insufficiency (Figure 3) (1,40).

LVOTO after Mitral valve surgery

In patients undergoing Mitral valve replacement, outflow tract obstruction is one of the postoperative complications (22). Obstruction occurs by a high profile protrusion of the prosthetic valve to the outflow tract or by an abnormal position of the prosthesis at the subvalvular level. If the prosthesis is not properly oriented, one of the ends can obstruct the LVOT (figure 4) (41).

Sigmoid septum

The interventricular sigmoid septum involves the protrusion of segments of the septum to the outflow tract of the LV and appears, usually, in elderly patients with a history of hypertension with low-grade concentric hypertrophy of the LV myocardium (thickness of the myocardium in diastole usually between 12 to 16 mm) (figure 5). The decrease in the aortoseptal angle Is typical of patients with sigmoid septum. Recently It has been proposed that the sigmoid configuration of the septum may lead to dynamic obstruction of the LVOT even without ventricular hypertrophy. In some cases the sigmoid septum may cause stenosis of the LVOT with dynamic obstruction and an increase in the pressure of the LV, especially if there is hypertrophy, physical exercise, general anaesthesia, acute myocardial infarction (AMI), dehydration or aggressive management of hypertension (42,43).

Valve obstruction

Aortic valve stenosis

Aortic stenosis is a common disease that usually affects older patients. There are two important factors that make it one of the most common valvulopathies: a. approximately 1-2 % of the population is born with a bicuspid aortic valve, which is prone to stenosis and b. aortic stenosis develops with age.

Calcified aortic stenosis was considered a degenerative lesion; However, it shares many characteristics with coronary artery

disease. Both are more common in men, older people, patients with hypercholesterolemia and both are derived from a chronic inflammatory process. Aortic stenosis differs from sclerosis due to the degree of valve involvement. In aortic sclerosis, the valves are abnormally thickened and the obstruction of the outflow tract is minimal. In contrast, in aortic stenosis the functional area of the leaflets has diminished sufficiently to cause measurable flow obstruction (44).

The bivalve aorta (Figure 6) occurs by abnormal valvulogenesis with the formation of a small cusp and a larger cusp, usually by congenital fusion of one of the valvular commissures. This disposition makes the valve more susceptible to trauma and finally leads to calcification and fibrosis. By the time the obstruction of the outflow tract causes significant symptoms, the valve is a rigid, calcified mass that makes it difficult to determine the etiology of process. This type of valve configuration is usually not obstructive early. Valvular stenosis develops between the fourth and sixth decade of life and represents > 50 % of cases of aortic stenosis disease in people under 70 years of age. The presence of a bivalve aortic valve is associated with an increase in the incidence of complications such as stenosis, insufficiency, endocarditis and aneurysmal dilatation of the aorta (1).

In developed countries, aortic stenosis is usually related to risk factors similar to those which cause atherosclerosis. Another major cause of aortic stenosis is rheumatic disease, which generates inflammatory adhesions of the valvular cusps that lead to fusion of the commissures and, consequently, to valve stenosis and/or regurgitation (45,46). As the compromise is greater, ventricular dysfunction occurs. Initially, the patients are asymptomatic and a systolic breath can be found in an incidental manner. As the disease progresses, the symptoms it produces are angina, dyspnea, syncope and, finally, heart failure. Once the symptoms appear, survival without surgical treatment is 2 to 3 years (45).

Doppler ultrasound can be performed in most patients, but the severity of the stenosis may be underestimated if the quality of the image is poor and is affected by technical factors. CT is useful for the quantification of valvular calcification (severe: > 1000 Agatston units) and in patients to whom they will practice transcatheter replacement, to undergo planimetry and take measurements of the valvular ring area, valve leaflet length and the distance from the ring to the coronary ostium; It is also sought to determine the dimensions of the aortic root, the severity of the vascular disease and the state of the coronary arteries (Figure 7) (47).

MRI allows a better characterization of the myocardium in patients with aortic stenosis and the detection of fibrosis with late enhancement sequences. A pattern of patched and subendocardial enhancement has been described, which predominates in basal segments. It has been shown that the late enhancement, as an indicator of myocardial fibrosis, is a factor of independent risk of mortality in patients with moderate or severe aortic stenosis, and is associated with a worse prognosis after valvular replacement (45, 48-50).

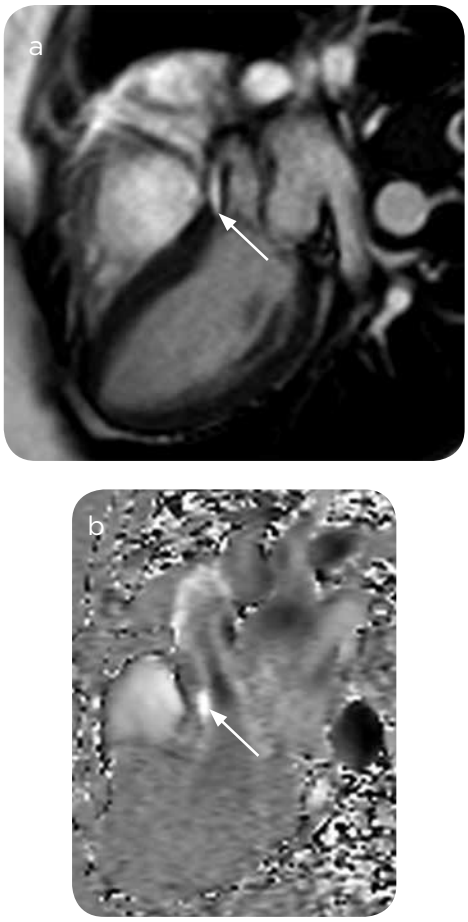

Figure 2. 25-year-old female patient with a membrane in the LVOT. Cardiac MRI, SSFP cinema sequences, long axis projection of three chambers (a) and phase contrast sequence in the plane of the outflow tract of the left ventricle (b), where the membrane is seen extending from the basal septum to the base of the anterior valve of the Mitral valve (arrow in a) which causes acceleration of the flow (arrow in b), in systole in the outflow tract.

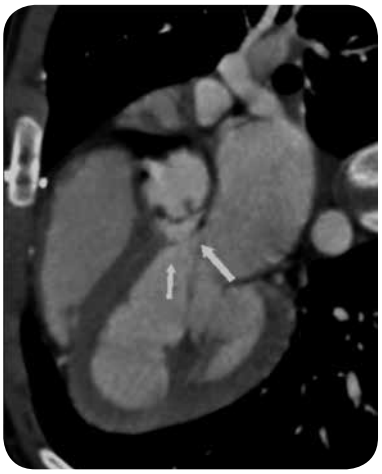

Figure 3. 21-year-old patient with multiple cardiovascular malformations, including LVOT membrane, partial pulmonary venous drainage abnormality and abnormal papillary muscles of the left ventricle with septal insertions. Correction of CIV. Cardio CT with reconstruction in long axis three chambers where an oblique membrane is observed in the LVOT (arrows).

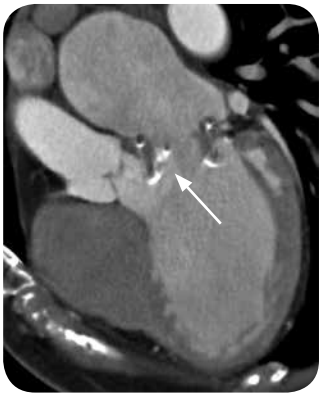

Figure 4. 65-year-old patient with Mitral bioprosthesis partially obstructing the outflow tract of the left ventricle. Study of cardio CT with long axis of three chambers multi-planar reconstruction. Protrusion of one of the pillars of the bioprosthesis ring towards the outflow tract of the left ventricle (arrow) with alteration of aortic angle, which favors this phenomenon.

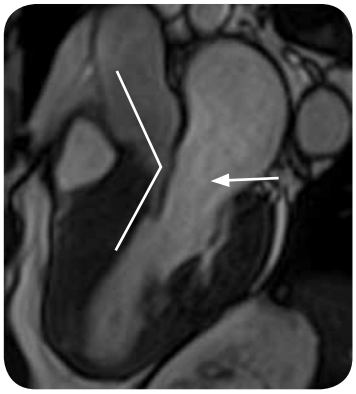

Figure 5. Aortoseptal angle in patient with sigmoid septum. CMRI, SSFP (steady-state free precession) long axis projection of three chambers cinema sequence. Basal septum protrusion (arrow) to the outflow tract in a patient with chronic hypertension, with aortoseptal angle and discreet concentric hypertrophy of the myocardium.

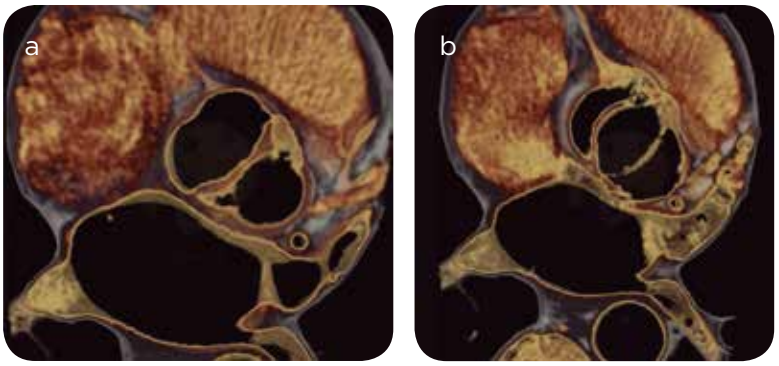

Figure 6. Cardiac CT in a patient with bivalve aorta without stenosis. 3D reconstruction in systole a) and diastole b).

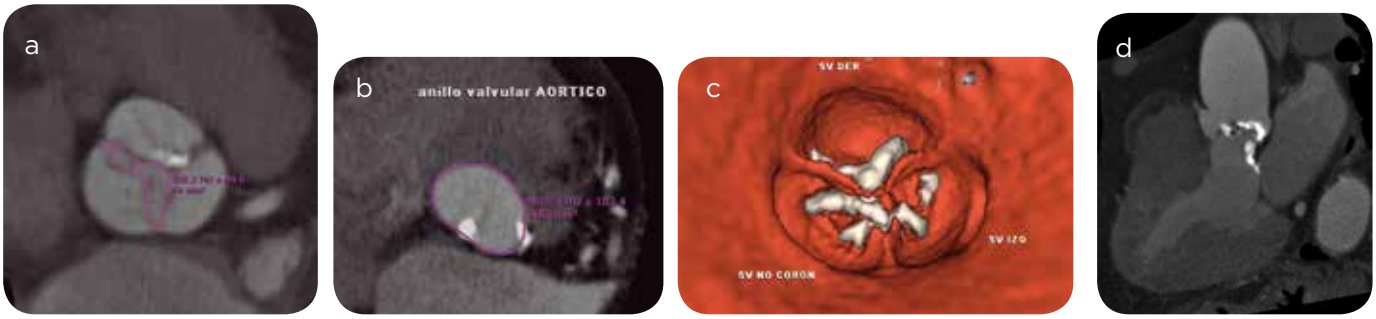

Figure 7. 78-year-old patient with severe aortic valve stenosis. Patient in protocol for TAVI planning, cardio-triggered CT, a) orthogonal multiplanar reconstructions of the aortic valve in systole and b) of the valve ring, c) three-dimensional reconstruction with superior sight of the aortic valve in diastole, d) multi-planar reconstruction long axis of three chambers. Anatomically trivalent aortic valve patient, but with complete fusion of the right and left valves, which causes the valve to behave physiologically as a bivalve with semilunar opening in systole (a). A valvular area in systole is identified by planimetry of 0.9 cm2 that correlates with severe stenosis. The extent of aortic valve calcification is identified through the outflow tract to the base of the anterior valve of the Mitral valve, a finding that must be reported since it significantly increases the risk of perforation in TAVI procedures (d).

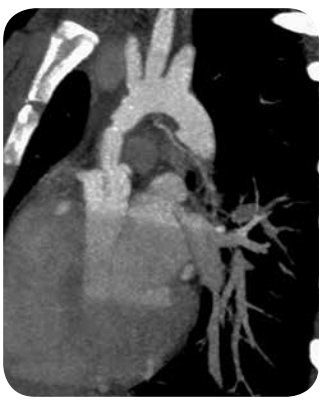

Figure 8. 2 year old patient with Williams-Beuren syndrome with supravalvular aortic stenosis. study of triggered cardio CT, sagittal reconstruction of the aorta and its root where aortic ascending stenosis from the sinotubular junction to the origin of the brachiocephalic trunk with decreased concentric vascular lumen is identified.

The severity of aortic valve stenosis is classified according to the following parameters (48):

	Low	Moderate	Severe
Valve area (cm^2) (mts / sec) Estimated gradient (mm Hg)	1,6-2,0 2,0-2,9 20-35	1,0-2,5 3,0-3,9 36-63	< 1,0 > 4,0 > 64

Supravalvular obstruction

Aortic supravalvular lesions are the rarest cause of LVOTO. They are characterized by a diffuse or focal stenosis that initiates at the sinotubular junction and occasionally extends throughout the ascending aorta. It rarely involves the aortic arch.

Aortic supravalvular stenosis is often associated with Williams-Beuren syndrome (Figure 8). This is an autosomal condition which occurs in 1 in 20.000 live births and in which 71 % of the time there is supravalvular aortic stenosis; It is also accompanied by mental retardation, stenosis of the pulmonary arteries, prolapse and up to 50 % of patients present aortic valve abnormalities, mainly aorta bivalve. This syndrome can be accompanied by alteration in the perfusion secondary to some degree of aortic valve adhesion in the sinotubular junction that restricts the diastolic filling of the coronary arteries (1).

Stenosis of the sinotubular region can be surgically extended when the patient has symptoms such as angina, dyspnea and syncope or when there is an average pressure gradient greater than 50 mm Hg (1).

Conclusion

Currently, cardiac magnetic resonance plays a leading role in the evaluation of patients with obstruction of the left ventricle, especially in the assessment of hypertrophic cardiomyopathies and in patients where echocardiography is unfinished or technically limited. The detection of myocardial fibrosis using late enhancement allows detecting those patients who are at increased risk of sudden death, malignant arrhythmias and electrocardiographic disorders.

References

1. Aboulhosn J, Child JS. Left ventricular outflow obstruction: subaortic stenosis, bicuspid aortic valve, supravalvar aortic stenosis, and coarctation of the aorta. Circulation. 2006;114:2412-22.
2. Fattal J, Henry MA, Ou S, Bradette S, Papas K, Marcotte F, et al. Magnetic resonance imaging of hypertrophic cardiomyopathy: beyondleft ventricular wall thickness. Can Assoc Radiol J. 2015;66:71-8.
3. Stojanovska J, Garg A, Patel S, Melville DM, Kazerooni EA, Mueller GC. Congenital and hereditary causes of sudden cardiac death in young adults:diagnosis, differential diagnosis, and risk stratification. Radiographics. 2013;33:1977-2001.
4. Wigle ED. Cardiomyopathy: The diagnosis of hypertrophic cardiomyopathy. Heart. 2001;86:709-14.
5. Karamitsos TD, Francis JM, Neubauer S. The current and emerging role of cardiovascular magnetic resonance in the diagnosis of nonischemic cardiomyopathies. Prog Cardiovasc Dis. 2011;54:253-65.
6. Gersh BJ, Maron BJ, Bonow RO, Dearani JA, Fifer MA, Link MS, et al. 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 Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. J Am Coll Cardiol. 2011;58:e212-60.
7. Radermecker M, Canivet JL, Lancellotti P, Limet R. The usual causes of left ventricular outflow tract obstruction below the aortic valve in normal ventriculoarterial connection: review of the physiopathology and surgical implications. Acta Chir Belg. 2005;105:475-81.
8. Noureldin RA, Liu S, Nacif MS, Judge DP, Halushka MK, Abraham TP, et al. The diagnosis of hypertrophic cardiomyopathy by cardiovascular magnetic resonance. J Cardiovasc Magn Reson. 2012;14:17.
9. Rollán MJ, San Román JA, Muñoz C, Cobos MA, Bratos JL. [Congenital anomalies of the mitral valve in the adult: presentation of 3 cases]. Rev Esp Cardiol. 1998;51:912-4.
10. Oosthoek PW, Wenink AC, Wisse LJ, Gittenberger-de Groot AC. Development of the papillary muscles of the mitral valve: morphogenetic background of parachute-like asymmetric mitral valves and other mitral valve anomalies. J Thorac Cardiovasc Surg. 1998;116:36-46.
11. Desai MY, Ommen SR, McKenna WJ, Lever HM, Elliott PM. Imaging phenotype versus genotype in hypertrophic cardiomyopathy. Circ Cardiovasc Imaging. 2011;4:156-68.
12. Bogaert J, Olivotto I. MR Imaging in hypertrophic cardiomyopathy: From magnet to bedside. Radiology. 2014;273:329-48.
13. Critoph CH, Pantazis A, Tome Esteban MT, Salazar-Mendiguchía J, Pagourelias ED, Moon JC, Elliott PM. The influence of aortoseptal angulation on provocable left ventricular outflow tract obstruction in hypertrophic cardiomyopathy. Open Heart. 2014;1:e000176.
14. Maron BJ, Gottdiener JS, Roberts WC, Henry WL, Savage DD, Epstein SE. Left ventricular outflow tract obstruction due to systolic anterior motion of the anterior mitral leaflet in patients with concentric left ventricular hypertrophy. Circulation. 1978;57:527-33.
15. Maron BJ, Harding AM, Spirito P, Roberts WC, Waller BF. Systolic anteriormotion of the posterior mitral leaflet: a previously unrecognized cause of dynamic subaortic obstruction in patients with hypertrophic cardiomyopathy. Circulation. 198368:282-93.
16. Shah PM, Gramiak R, Kramer DH. Ultrasound localization of left ventricular outflow obstruction in hypertrophic obstructive cardiomyopathy. Circulation. 1969;40:3-11.
17. Kwon DH, Smedira NG, Popovic ZB, Lytle BW, Setser RM, Thamilarasan M, et al. Steep left ventricle to aortic rootangle and hypertrophic obstructive cardiomyopathy: study of a novel associationusing three-dimensional multimodality imaging. Heart. 2009;95:1784-91.
18. Barkhordarian R, Wen-Hong D, Li W, et al. Geometry of the left ventricular outflow tract in fixed subaortic stenosis and intact ventricular septum: an echocardiographic study in children and adults. J Thorac Cardiovasc Surg. 2007;133:196-203.
19. Fowles RE, Martin RP, Popp RL. Apparent asymmetric septal hypertrophy due to angled interventricular septum. Am J Cardiol. 1980;46:386-92.
20. Shah JS, Esteban MT, Thaman R, Sharma R, Mist B, Pantazis A, et al. Prevalence of exercise-induced left ventricular outflow tract obstruction in symptomatic patients with non-obstructive hypertrophic cardiomyopathy. Heart. 2008;94:1288-94.
21. Fowles RE, Martin RP, Popp RL. Apparent asymmetric septal hypertrophy due to angled interventricular septum. Am J Cardiol. 1980;46:386-92.
22. Aboulhosn J, Child JS. Left ventricular outflow obstruction: subaortic stenosis, bicuspid aortic valve, supravalvar aortic stenosis, and coarctation of the aorta. Circulation. 2006;114:2412-22.
23. Dulce MC, Mostbeck GH, Higgins CB, et al. Magnetic resonance tomography (MRT) in the evaluation of heart disease: quantitative determination of aortic regurgitation volume. Rontgenpraxis. 1994;47:65- 69.
24. Allison JD, Flickinger FW, Wright JC, et al. Measurement of left ventricularmass in hypertrophic cardiomyopathy using MRI: comparison with echocardiography. Magn Reson Imaging. 1993;11:329-34.
25. Bogaert J, Olivotto I. MR Imaging in Hypertrophic Cardiomyopathy: From Magnet to Bedside. Radiology. 2014;273:329-48.
26. Schulz-Menger J, Abdel-Aty H, Busjahn A, et al. Left ventricular outflow tract planimetry by cardiovascular magnetic resonance differentiates obstructive from non-obstructive hypertrophic cardiomyopathy. J Cardiovasc Magn Reson. 2006;8:741-6.
27. Maron MS, Olivotto I, Zenovich AG, et al. Hypertrophic cardiomyopathyis predominantly a disease of left ventricular outflow tract obstruction. Circulation. 2006;114:2232-9.
28. Maron BJ. Hypertrophic cardiomyopathy: a systematic review. JAMA. 2002;287:1308-20.
29. Maron BJ, McKenna WJ, Danielson GK, et al. American College of Cardiology/European Society of Cardiology clinical expert consensusdocument on hypertrophic cardiomyopathy. J Am Coll Cardiol. 2003;42:1687-713.
30. Maron MS, Maron BJ, Harrigan C, et al. Hypertrophic cardiomyopathy phenotype revisited after 50 years with cardiovascular magnetic resonance. J Am Coll Cardiol. 2009;54:220-8.
31. Maron BJ, Gottdiener JS, Epstein SE. Patterns and significance of distribution of left ventricular hypertrophy in hypertrophic cardiomyopathy: a wide angle, two dimensional echocardiographic study of 125 patients. Am J Cardiol. 1981;48:418-28.
32. Maron BJ, Maron MS, Wigle ED, et al. The 50-year history, controversy, and clinical implications of left ventricular outflow tract obstruction in hypertrophic cardiomyopathy: from idiopathic hypertrophic subaortic stenosis to hypertrophic cardiomyopathy. J Am Coll Cardiol. 2009;54:191-200.
33. Maron BJ, Sherrid MV, Haas TS, et al. Novel hypertrophic cardiomyopathy phenotype: segmental hypertrophy isolated to the posterobasal left ventricular free wall. Am J Cardiol. 2010;106:750-2.

34. Rickers C, Wilke NM, Jerosch-Herold M, et al. Utility of cardiac magnetic resonance imaging in the diagnosis of hypertrophic cardiomyopathy. Circulation. 2005;112:855- 61.
35. Maron MS, Lesser JR, Maron BJ. Management implications of massiveleft ventricular hypertrophy in hypertrophic cardiomyopathy significantly underestimated by echocardiography but identified by cardiovascularmagnetic resonance. Am J Cardiol. 2010;105:1842-3.
36. Adabag AS, Maron BJ, Appelbaum E, et al. Occurrence and frequency of arrhythmias in hypertrophic cardiomyopathy in relation to delayed enhancement on cardiovascular magnetic resonance. J Am Coll Cardiol. 2008;51:1369-74.
37. Rubinshtein R, Glockner JF, Ommen SR, et al. Characteristics and clinical significance of late gadolinium enhancement by contrastenhanced magnetic resonance imaging in patients with hypertrophic cardiomyopathy. Circ Heart Fail. 2010;3:51-8.
38. O'Hanlon R, Grasso A, Roughton M, et al. Prognostic significance of myocardial fibrosis in hypertrophic cardiomyopathy. J Am Coll Cardiol. 2010;56:867-74.
39. Cilliers AM, Gewillig M. Rheology of discrete subaortic stenosis. Heart. 2002;88:335-6.
40. Bruder O, Wagner A, Jensen CJ, et al. Myocardial scar visualized bycardiovascular magnetic resonance imaging predicts major adverse events in patients with hypertrophic cardiomyopathy. J Am Coll Cardiol. 2010;56:875- 87.
41. Cilliers AM, Gewillig M. Rheology of discrete subaortic stenosis. Heart. 2002;88:335-6.
42. Guler N, Ozkara C, Akyol A. Left ventricular outflow tract obstruction after bioprosthetic mitral valve replacement with posterior mitral leaflet preservation. Tex Heart Inst J. 2006;33:399-401.
43. Polyakova T, Zvereva L, et al. Longitudinal 2D strain imaging in patients with sigmoid shaped interventricular. Scientific exhibit in ECR 2013.
44. Gentille-Lorente D, Salvadó-Usach T. [Sigmoid septum: A variant of the ventricular hypertrophy or of the hypertrophic cardiomyopathy?]. Arch Cardiol Mex. 2016;86:110-22.
45. Carabello BA. Clinical practice. Aortic stenosis. N Engl J Med. 2002;346:677-82.
46. Bennett CJ, Maleszewski JJ, Araoz PA. CT and MR imaging of the aortic valve: radiologic-pathologic correlation. Radiographics. 2012;32:1399-420.
47. Maganti K, Rigolin VH, Sarano ME, Bonow RO. Valvular heart disease: diagnosis and management. Mayo Clin Proc. 2010;85:483-500.
48. Nishimura RA, Otto CM, Bonow RO, Carabello BA, Erwin JP 3rd, Guyton RA, et al. AHA/ACC guideline for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. J Am Coll Cardiol. 2014;63:e57-185.
49. Dweck MR, Joshi S, Murigu T, Alpendurada F, Jabbour A, Melina G, et al. Midwall fibrosis is an independent predictor of mortality in patients with aortic stenosis. J Am Coll Cardiol. 2011;58:1271-9.
50. Azevedo CF, Nigri M, Higuchi ML, Pomerantzeff PM, Spina GS, Sampaio RO, et al. Prognostic significance of myocardialfibrosis quantification by histopathology and magnetic resonance imaging in patients with severe aortic valve disease. J Am Coll Cardiol. 2010;56:278-87.

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