Differentiating Constrictive Pericarditis from Restrictive Cardiomyopathy

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Constrictive pericarditis and restrictive cardiomyopathy are 2 forms of diastolic dysfunction with similar presentation but different treatment options. Whereas constrictive pericarditis has the potential of being cured with pericardiectomy, restrictive cardiomyopathy is usually incurable. It is therefore crucial to differentiate between the 2 disorders. In the last few years, new diagnostic techniques have become available to differentiate these causes of diastolic dysfunction from each other. This review provides a complete, in-depth comparison of the 2 disorders with regard to their symptoms and clinical features, etiology, pathophysiology, hemodynamics, echocardiographic presentation, and finally the different available management options.

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Key words: Constrictive pericarditis • Restrictive cardiomyopathy • Diastolic dysfunction • M-mode color Doppler imaging • Tissue Doppler imaging • Radionuclide ventriculogram • Pericardiectomy

Diastolic dysfunction is defined in the absence of systolic dysfunction, as the requirement for elevated filling pressure to maintain cardiac output.[1] Augmented filling pressure might result in right-sided and/or left-sided congestion with symptoms of heart failure. Many processes might affect the myocardium or pericardium and lead to diastolic dysfunction (Figure 1). Among the processes that cause diastolic dysfunction by affecting the myocardium, the most common in Western nations is hypertensive cardiomyopathy.[2] The myocardial

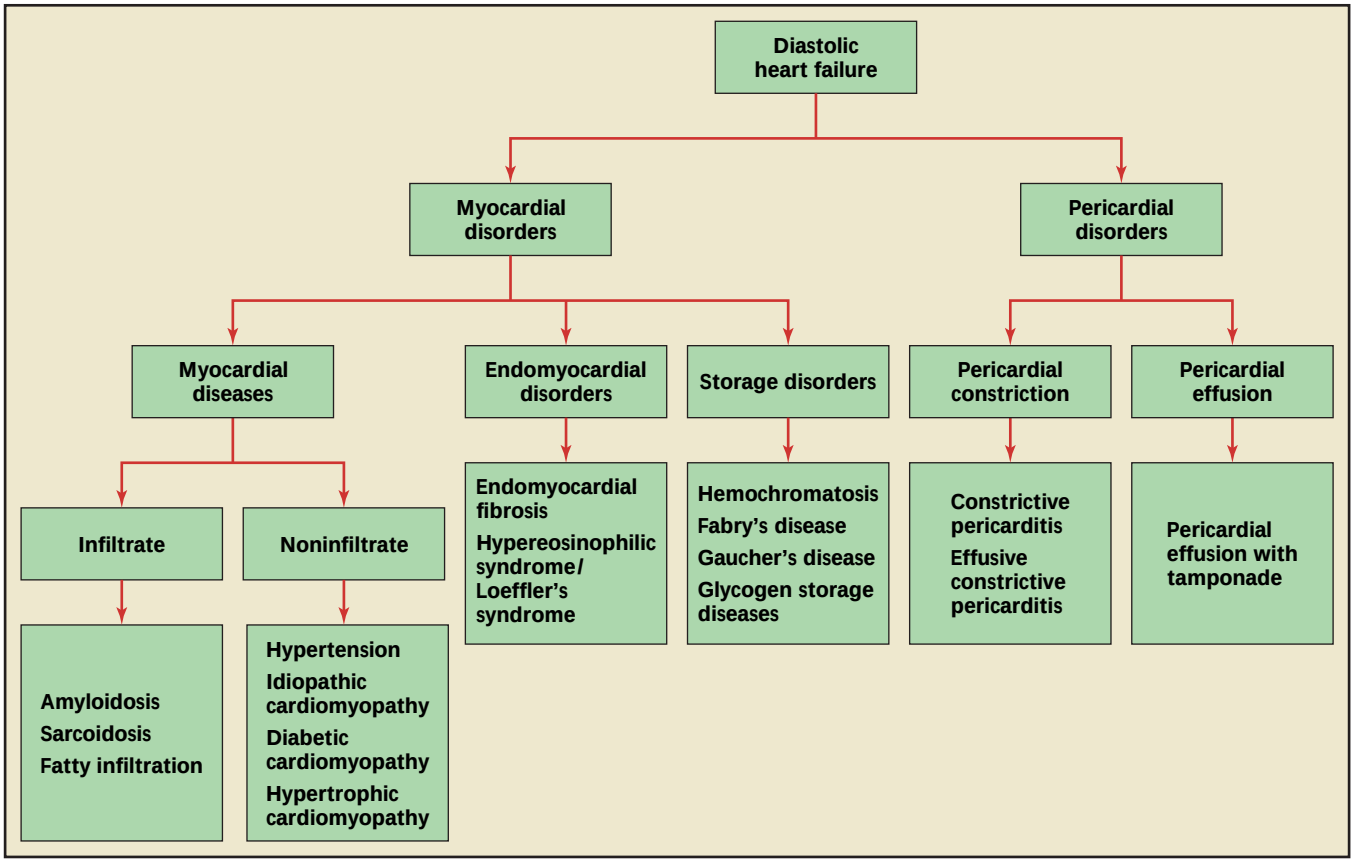


Figure 1. *Causes of diastolic dysfunction.*

conditions that lead to diastolic dysfunction exert their effect by slowing and delaying ventricular relaxation. Restrictive cardiomyopathies are among this group of disorders.[3] The other group of conditions leading to diastolic dysfunction comprises pericardial disorders, such as constrictive pericarditis (Figure 1). In constrictive pericarditis most of the ventricular filling occurs in early diastole, and further filling and expansion of the ventricle(s) is limited by the thickened pericardium. Although restrictive cardiomyopathy and constrictive pericarditis are the result of 2 different pathologic processes, they commonly manifest similarly, with diastolic right-sided heart failure, and they can be difficult to differentiate by clinical examination and simple diagnostic workup. The differentiation of these 2 disorders is crucial because constrictive pericarditis can potentially be cured by performing a pericardiectomy, whereas restrictive cardiomyopathy is usually treated symptomatically.[4] It should be emphasized that certain entities of restrictive cardiomyopathy are curable, and a correct differentiation of these from the other causes of restrictive cardiomyopathy is important. Table 1 summarizes the causes of restrictive cardiomyopathy.

Symptoms

Patients with restrictive cardiomyopathy and constrictive pericarditis present with symptoms of diastolic dysfunction. This can be in the form of either right-sided or left-sided heart failure. Usually the right-sided symptoms are noted as filling pressure and

Table 1
Causes of Restrictive Cardiomyopathy

Myocardial disease
Infiltrative
Amyloidosis
Sarcoidosis
Fatty infiltration
Non-infiltrative
Idiopathic cardiomyopathy
Diabetes mellitus
Endomyocardial disease
Endomyocardial fibrosis
Loeffler's syndrome
Storage disease
Glycogen storage disease
Fabry's disease
Gaucher's disease
Hemochromatosis

systemic venous pressure exceeding 10 to 15 mm Hg, with postprandial fullness, anorexia, flatulence, dyspepsia and abdominal swelling due to ascites, liver congestion and mesenteric venous congestion, and peripheral edema. Once filling pressure exceeds 15 to 30 mm Hg, symptoms of pulmonary venous congestion appear. As the disease progresses the symptoms of low cardiac output, such as severe fatigue, muscle wasting, and weight loss dominate.

Patients with restrictive cardiomyopathy frequently present with the initial symptom of exercise intolerance due to rapid development of pulmonary venous congestion and inability of cardiac output to increase with exercise and increased heart rate. This is due to the excessive drop of diastolic filling with increased heart rate in these patients.

Compared with constrictive pericarditis, patients with restrictive cardiomyopathy more commonly have arrhythmias because of the infiltrative nature of the disease and myocardial damage. Patients with constrictive pericarditis might develop myocardial ischemia and anginal chest pain due to compression and impingement of coronary arteries by the growing thick and fibrotic pericardium.[5] Finally, platypnea has been described in constrictive pericarditis.[6] The mechanism for this symptom is not understood in constrictive pericarditis. Table 2 summarizes the symptoms of diastolic dysfunction and low cardiac output seen in these conditions.

Table 2
Symptoms of Constrictive Pericarditis and Restrictive Cardiomyopathy

Right-sided heart failure
Peripheral edema
Ascites and liver congestion
Postprandial fullness
Flatulence
Anorexia
Left-sided heart failure
Pulmonary venous congestion
Shortness of breath
Orthopnea
Low cardiac output
Fatigue
Muscle wasting
Weight loss

Clinical Findings

The most common finding in patients with constrictive pericarditis and restrictive cardiomyopathy is elevated systemic venous pressure manifested by elevated jugular venous pressure, enlarged pulsatile liver, and peripheral edema on physical examination. Whereas patients with constrictive pericarditis do not have palpable point of maximum intensity (PMI) (due to separation of the heart from the chest wall by the pericardium), PMI is easily palpable in patients with restrictive cardiomyopathy. In constrictive cardiomyopathy an additional acoustic finding, pericardial knock, is heard after occurrence of second heart sound (S2). Pericardial knock occurs when the ventricle reaches its maximum filling limit set by the surrounding pericardium. This sound is temporarily earlier than an S3 (third heart sound) would be. In restrictive cardiomyopathy commonly a fourth heart sound (S4) is heard, produced by atrial kick in the hypertrophied myocardium, if sinus rhythm is present. Kussmaul's sign (inspiratory increase in systemic venous pressure) is described in both conditions but is more commonly seen in constrictive pericarditis; however, it might be difficult to appreciate at bedside. Pulsus paradoxus is uncommon in either condition, but it does occur, if rarely, in constrictive pericarditis. Freidrich's sign (prominent Y descent on early diastole) is seen in constrictive pericarditis. Muscle wasting happens late in both disorders once the low cardiac output stage is reached.

Etiology

Restrictive cardiomyopathy was first differentiated from the other forms of cardiomyopathy (hypertrophic and dilated cardiomyopathy) by Goodwin and colleagues in 1961.[7] Although restrictive cardiomyopathy was initially thought to be an idiopathic disorder, many diseases have since become known to cause this group of disorders.[8] Among the many causes of restrictive cardiomyopathy, amyloidosis is reported most commonly. Classification of all common causes of restrictive cardiomyopathy is summarized in Table 1.[8,9] Although restrictive cardiomyopathy carries a poor prognosis, some causes (eg, hemochromatosis) can be potentially reversible.

Practically any cause of acute pericarditis has the potential of creating an eventual chronic picture of constrictive pericarditis. Therefore, each of the causes of acute pericarditis might also be accounted as a cause of constrictive pericarditis. In the pre-antibiotic era, the most common cause of chronic constrictive pericarditis was tuberculosis, and this might still be the case in some developing countries.[10-12] In the Western world iatrogenic causes, such as radiation exposure and cardiac surgery, have become the dominant causes of constrictive pericarditis and are now responsible for one third of all the cases in some reports.[13] Table 3 summarizes causes of constrictive pericarditis.

Diagnosis

Many varieties of constrictive pericarditis have been described on the

Table 3
Causes of Constrictive Pericarditis

Idiopathic
Trauma: penetrating or nonpenetrating, causing hemopericardium
Uremia
Iatrogenic: post-thoracotomy, post-iatrogenic tamponade (pacemaker/AICD insertion), post-PTCA
Drugs: hydralazine, procainamide, methysergide
Post-myocardial infarction: early and late (Dressler's syndrome)
Infections: viral (coxsackie, hepatitis, echo virus), bacterial (tuberculosis, pneumococcus, streptococcus, staphylococcus), fungus (histoplasmosis, coccidioidomycosis, candida)
Exotic organisms: parasitic (amebiasis, echinococcosis)
Neoplasia: breast cancer, lung cancer, lymphoma, leukemia, melanoma, mesothelioma
Vasculitis/connective tissue disorders: SLE, rheumatoid arthritis, PAN, inflammatory bowel disease
Amyloidosis
Sarcoidosis
Toxic agents (eg, asbestos)
Other causes, including radiation

AICD, automatic implantable cardioverter defibrillator; PTCA, percutaneous transluminal coronary angioplasty; SLE, systemic lupus erythematosus; PAN, polyarteritis nodosa.

basis of their chronicity, appearance, effusive component, localization, and time of presentation (Table 4). Although many forms of constrictive pericarditis have been reported, all of them have similar clinical manifestations.[14-18]

Diagnosing constrictive pericarditis and restrictive cardiomyopathy and differentiating the 2 entities can be difficult at times, especially when the obvious hallmarks of the disease, such as eggshell calcification surrounding the heart in classic constrictive pericarditis, are not present. Many diagnostic modalities have been used to differentiate these 2 disease entities.[19] Although each diagnostic procedure might contribute to the diagnosis of the specific disorder, in most cases no single diagnostic modality has enough sensitivity and specificity by itself to diagnose the disorder, and a combination of these tests is needed to correctly diagnose the cause of diastolic dysfunction.

Table 4
Varieties of Constrictive Pericarditis

Classic forms
Chronic calcific rigid shell
Subacute noncalcific
Effusive form
Localized
Occult

Electrocardiogram

Because the myocardium is commonly highly infiltrated in restrictive cardiomyopathy, an observation of atrioventricular and intraventricular conduction delay or block is not uncommon in this condition. A low-voltage electrocardiogram (ECG) is a classic finding seen in restrictive cardiomyopathy associated with amyloidosis. In long-standing cases of restrictive cardiomyopathy, atrial abnormality or hypertrophy is noted on ECG. The elevated intra-atrial pressure might also cause predisposition to atrial fibrillation.[5]

Although the above-mentioned ECG findings might also be seen in patients with constrictive pericarditis, they are much less common in this condition. When these ECG findings are present, they are most likely related to an extension of calcification into the myocardium.[20] Ischemic changes with ST-T changes might be noted on the ECG of patients with constrictive pericarditis, owing to calcification processes involving pericardial coronary arteries externally.

Chest X-Ray

Although a calcified pericardium on chest roentgenogram is a classic finding in cases of constrictive pericarditis, this finding might not be present despite hemodynamically severe constrictive pericarditis. Calcified pericardium is noted on chest roentgenogram in only 20% to 30% of the patients with constrictive pericarditis.[21] In addition, a calcified pericardium is not necessarily synonymous with constrictive pericarditis, because calcification might be present without hemodynamic effect on the heart function. Calcification might also occur in an aneurysmal segment of a previously infarcted left ventricle and be misinterpreted as pericardial calcification.

The cardiac silhouette might be small, normal, or enlarged in both constrictive pericarditis and restrictive cardiomyopathy. Prominence of the right superior mediastinum is sometimes noted as a result of superior vena cava and right atrium engorgement in either condition.[22]

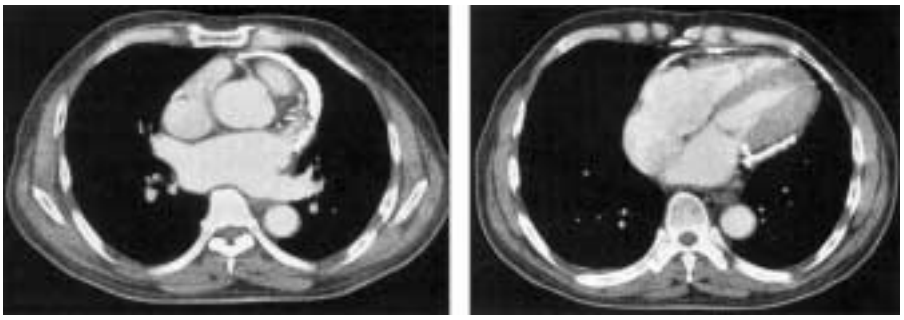


Figure 2. *Cine computed tomography scans at the base of the heart (left) and at midventricular level (right) of a patient with constrictive pericarditis. There is heavy calcification of the pericardium extending into the posterior atrioventricular grove. Adapted with permission from Braunwald.*[4]

Computed Tomography and Magnetic Resonance Imaging

Findings on chest images from computed tomography (CT) might not only be helpful in diagnosing constrictive pericarditis but also have some prognostic value in the screening of patients for surgery (Figure 2). CT has a higher sensitivity for detecting pericardial calcification than chest roentgenogram, although this increase in sensitivity is only on the order of approximately 10% to 15%.[21] In addition, with CT one can identify dilation of the venae cavae and atrial and right ventricular deformity.[23,24]

Myocardial fibrosis or atrophy of the myocardium is noted on the CT as nonvisualization of the posterolateral wall, and it indicates a poor prognosis after pericardiectomy.[25]

CT imaging can also be useful in the diagnosis of certain types of restrictive cardiomyopathy. For instance, iron deposition in myocardium seen in hemochromatosis can cause significantly bright myocardium and raise the suspicion of this disorder.

Magnetic resonance imaging (MRI) can also detect the same findings as mentioned above with CT (Figure 3). Although the experience with MRI in these disorders is not as extensive as with CT, it seems that MRI is more sensitive than CT for delineating pericardial thickening and any associated myocardial atrophy or fibrosis.[26,27]

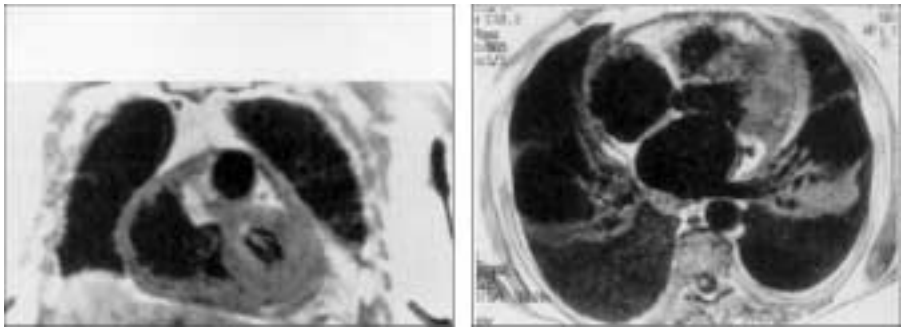
Echocardiogram

M-mode echocardiography might be helpful in measuring pericardial thickness. There are 2 patterns that might be visualized: either 2 parallel lines representing the visceral and parietal pericardium or multiple dense echoes with their width as the full thickness of the pericardium.[28] M-mode might also demonstrate the "septal bounce" (abrupt posterior motion of the interventricular septum during early diastole), a common finding in constrictive pericarditis.

Two-dimensional echocardiography might show a dense, immobile, and thick pericardium in constrictive pericarditis, but its resolution and sensitivity are not as good as CT/MRI for clearly distinguishing pericardium from the surrounding tissue. It might also show "septal bounce" in constrictive pericarditis.[29] Furthermore, 2-dimensional echocardiography can reveal dilated inferior vena cava, hepatic veins, and right/left atrial enlargement in both constrictive pericarditis and restrictive cardiomyopathy. The diagnostic yield of these findings might be low because they can also be seen in other conditions, such as in patients with right heart failure, pacemaker, and/or bundle branch block.[30] In cardiac amyloidosis, a common form of restrictive cardiomyopathy, the myocardium has a ground-glass appearance on two-dimensional echocardiography when the harmonic function is not in use.[31]

Spectral Doppler echocardiography has been found to be a useful tool for diagnosing constrictive pericarditis and differentiating it from restrictive cardiomyopathy.[32,33] Although both conditions can show any of the 3 phases of restriction on conventional Doppler measurement of the left ventricular inflow through the mitral valve, respiratory changes of mitral valve inflow are quite different in these 2 conditions.[34] Because the myocardium is isolated from the intrathoracic respiratory pressure

Figure 3. *Magnetic resonance imaging in coronal (left) and transverse (right) planes of a patient with constrictive pericarditis. There is significant thickening of the pericardium, which extends over the pulmonary artery on the coronal image. Adapted with permission from Braunwald.*[4]



changes in constrictive pericarditis, there are significant flow changes over the mitral and tricuspid valves during inspiration and expiration. During inspiration, the pulmonary capillary pressure drops, whereas the intracardiac pressure is not affected; therefore, the inflow to the left ventricle over the mitral valve is reduced in constrictive pericarditis. This can be visualized by Doppler echocardiography over the mitral valve with simultaneous graphic recording of the phases of respiration. On the other hand, the flow over the tricuspid valve increases during inspiration, owing to increased venous return to the heart. The interventricular septum will also show a leftward shift during early diastole under inspiration, owing to the above-mentioned changes and ventricular interdependence resulting from the encasement of the heart in the rigid pericardium shell. Although these techniques have been found to be reasonably sensitive and specific (as high as 85%–90% in expert hands), they are cumbersome to perform because simultaneous respiratory recording has to be performed. In addition, irregular breathing, irregular cardiac rhythm, and short diastolic periods resulting from rapid heart rate cause difficulty in interpretation.[19]

Tissue Doppler echocardiography and color M-mode echocardiography were found to be both sensitive and specific diagnostic tools in the differential diagnosis of constrictive pericarditis from restrictive cardiomyopathy in the late 1990s.[35] Longitudinal axis expansion of the mitral valve ring can be measured by tissue Doppler echocardiography. In healthy normal subjects, a peak early velocity longitudinal expansion (peak Ea) of more than 10 cm/s is observed. In restrictive cardiomyopathy Ea is reduced, whereas it is normal or elevated in constrictive pericarditis owing to normal relaxation of the myocardium in the early phase of diastole in this condition and because most of the filling occurs early in diastole in constrictive pericarditis. Rajagopalan and colleagues[36] found in a limited study that the sensitivity and specificity of tissue Doppler echocardiography reach 89% and 100%, respectively, when a peak Ea velocity of at least 8.0 cm/s is used for the differentiation of constrictive pericarditis from restrictive cardiomyopathy. In the same study,[36] the investigators found sensitivity and specificity of 74% and 91%, respectively, for color M-mode in the diagnosis of constriction when a slope of 100 cm/s or more for the first aliasing contour was used in color M-mode propagation. In a recent study by Ha and colleagues,[37] an Ea cut-off value of 8 cm/s during tissue Doppler imaging resulted in very high sensitivity and specificity (95% and 96%, respectively) for the diagnosis of constrictive pericarditis.

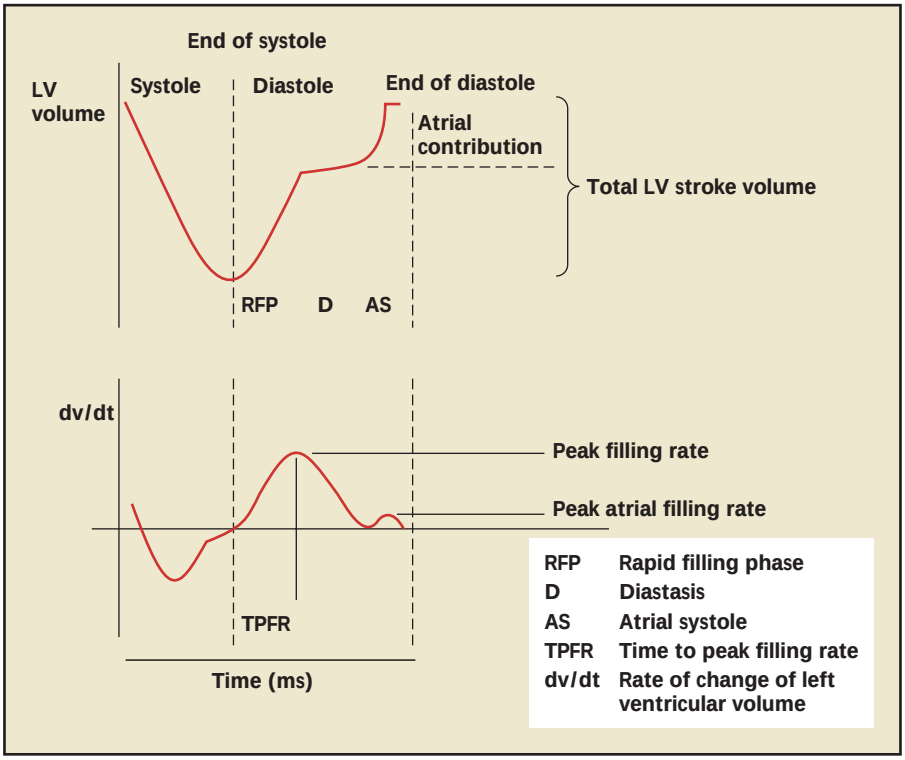
Radionuclide Ventriculography

Use of the first-pass time of the radionuclide tracer during diastole has been proved to be helpful in the differentiation of constrictive pericarditis from restrictive cardiomyopathy. In constrictive pericarditis most of the flow/filling occurs in early diastole, with minimal atrial filling contribution to the total ventricular diastolic filling. Conversely, the filling/flow is sluggish in early diastole in restrictive cardiomyopathy, and the atrial filling contribution is enhanced.[38] Figure 4 shows an example of radionuclide ventriculography in a healthy subject.

Cardiac Catheterization

Difficulty in differentiating constrictive pericarditis from restrictive

Figure 4. *Radionuclide ventriculography in a healthy subject. Most of the diastolic filling is occurring in early diastole, and there is minimum contribution of atrial contraction.*



cardiomyopathy was reported in the literature once pressure tracing was widely used as a diagnostic tool.[19] Some investigators argued that the diagnostic dilemma is almost created by similar hemodynamic tracings in these 2 disorders rather than by similar clinical presentations.[19]

The dip-plateau, or square root-like waveform in the right ventricular or left ventricular pressure tracings is the classic hallmark of both constrictive pericarditis and restrictive cardiomyopathy. Still, there are several hemodynamic clues that can be helpful to differentiate constrictive pericarditis from restrictive cardiomyopathy. Equilibration of the diastolic pressures in all chambers of the heart with a discrepancy of less than 5 mm Hg is usually indicative of constrictive pericarditis rather than restrictive cardiomyopathy, whereas in restrictive cardiomyopathy the left ventricular diastolic pressure is usually higher than the right ventricular diastolic pressure. The ventricular interdependence of constrictive pericarditis can also be noted on the hemodynamic tracings by disconcordant variation of the right and left ventricular peak systolic pressure levels with respiration.[34] In addition, right ventricular systolic pressure is commonly higher in restrictive cardiomyopathy owing to a higher pulmonary systolic pressure. This will lead to a right ventricular end diastolic pressure (RVEDP) over right ventricular systolic pressure (RVSP) ratio of less than one third (RVEDP/RVSP < 1/3) in restrictive cardiomyopathy, whereas the RVEDP/RVSP ratio is often greater than one third in constrictive pericarditis.[39] Although presence of pulsus paradoxus is rather indicative of tamponade, it has also been noted rarely in constrictive pericarditis and even less frequently in restrictive cardiomyopathy.[40] These differences can be seen in Figures 5 and 6.

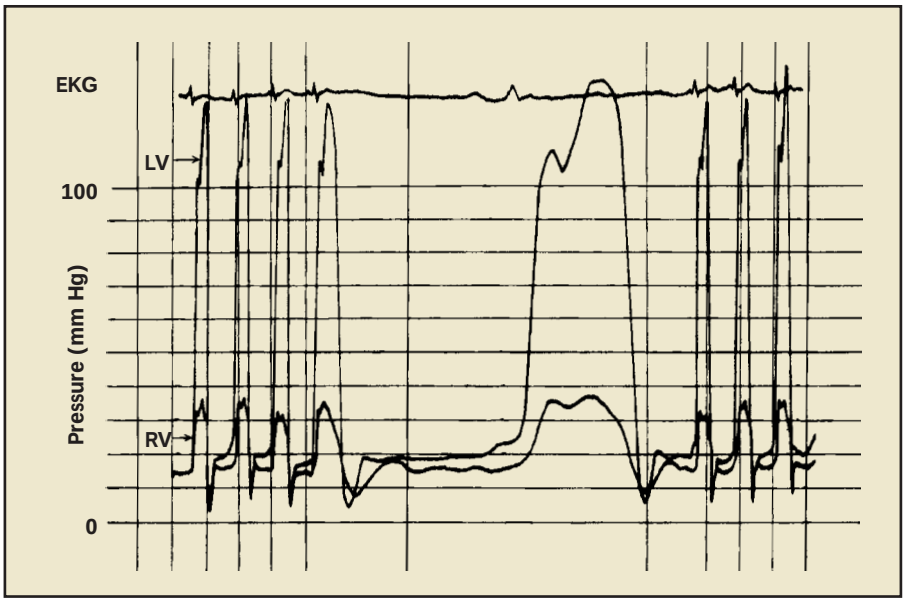


Figure 5. *Simultaneous left ventricular and right ventricular pressure recordings in a patient with restrictive cardiomyopathy, illustrating the early dip and plateau and near equilibration of diastolic pressures. Reproduced with permission from Benotti et al.*[46]

Endomyocardial Biopsy

Endomyocardial biopsy is essential in the final diagnosis of restrictive cardiomyopathy. Biopsy is also essential if thoracotomy is considered for diagnosis of constrictive pericarditis. Endomyocardial biopsy has high specificity in certain types of restrictive cardiomyopathy, such as amyloidosis, hemochromatosis,

Figure 6. *Simultaneous right ventricular and left ventricular pressure tracings in a patient with constrictive pericarditis. Reproduced with permission from Myers and Spodick.*[47]

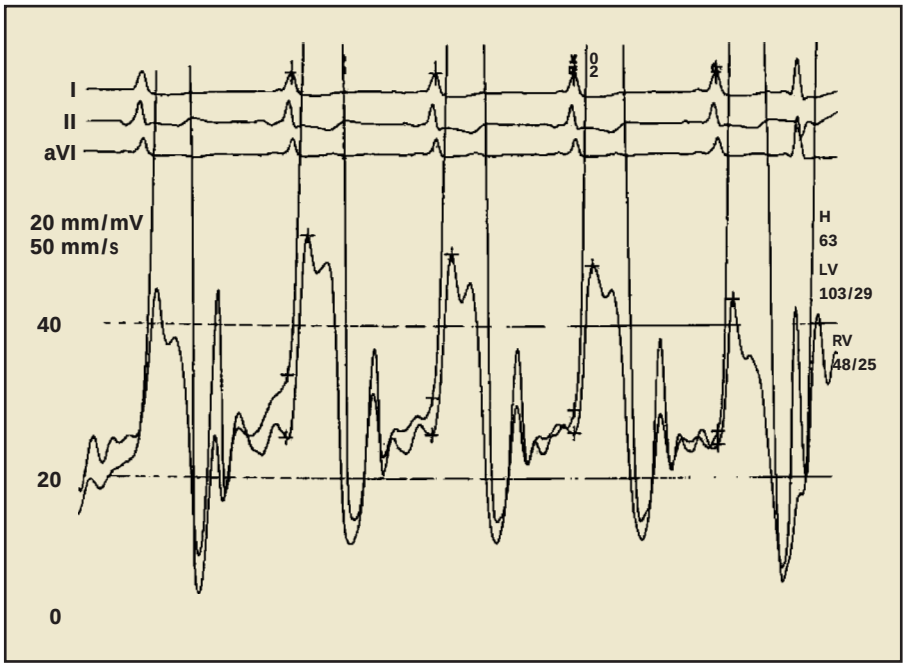


Table 5
Differentiation Between Restrictive Cardiomyopathy and Constrictive Pericarditis

	Constrictive Pericarditis	Restrictive Cardiomyopathy
History	Pericarditis, radiation, cardiac surgery or trauma, uremia or other causes of pericarditis	Family medical history of hemochromatosis or other causes of restrictive cardiomyopathy
Physical examination	Pericardial knock, apical impulse not palpable, pulsus paradoxus, Kussmaul's sign	S3 (late in disease) and S4 (early in disease) frequently present, regurgitant murmurs common
Electrocardiogram	P wave abnormalities	Low voltage in amyloidosis, T wave abnormalities, conduction disturbances
Chest x-ray	Pericardial calcification in 30%	Pericardial calcification in <30%
2D-echocardiogram	Normal wall thickness, pericardial thickening might be seen, "septal bounce"	Small LV cavity with LVH, thickened cardiac valves and granular sparking texture (in amyloidosis)
Doppler inflow patterns		
Mitral valve	Reduced flow during inspiration and opposite during expiration (more than 25%), prolonged IVRT and short DT during inspiration	No significant respiratory variation, E/A > 2, short DT (<150 ms), short IVRT (<60 ms)
Tricuspid valve	Increased flow during inspiration and opposite changes during expiration (E wave changes typically more than 40%)	Similar to the finding over mitral inflow with inspiration, further shortening of DT with an increased E/A (E/A > 2)
Pulmonary vein	S/D = 1, similar respiratory flow changes as with mitral valve	Blunted S/D ratio (0.5) with small S and large D, prominent AR, no significant respiratory variation of D wave
Hepatic vein	S greater than D wave with small AR	Similar finding as with pulmonary vein flow, with blunted S/D ratio, reversal of S and D size (D > S), with increased reversal prominence during respiration and intact AR
Color M-mode	Rapid flow propagation (>100 cm/s)	Slow flow propagation
Tissue Doppler over mitral annular motion	Rapid early filling (>8 cm/s)	Low velocity early filling (<8 cm/s)
Hemodynamics	Dip and plateau/square root sign Equal diastolic pressures (RVEDP = LVEDP) Ventricular interdependence with ↑ RV systolic pressure by inspiration	Dip and plateau/square root sign LVEDP > RVEDP by at least 5 mm Hg RVSP > 50 mm Hg RVEDP < one third of RVSP
CT/MRI	Pericardium commonly thickened and calcification might be noted	Usually normal-looking pericardium
Endomyocardial biopsy	Might be normal or show nonspecific changes (myocardial hypertrophy or fibrosis)	Might reveal specific cause of restrictive pericarditis

LV, left ventricular; LVH, LV hypertrophy; IVRT, isovolumic relaxation time; DT, deceleration time; E/A ratio, early to late diastolic filling ratio; S/D, systolic/diastolic ratio; AR, atrial reversal wave; RVEDP, right ventricular end diastolic pressure; LVEDP, LV end diastolic pressure; RVSP, RV systolic pressure; CT, computed tomography; MRI, magnetic resonance imaging.

and eosinophilic cardiomyopathy, whereas its specificity is low in other types of restrictive cardiomyopathy, such as idiopathic restrictive cardiomyopathy. The nonspecific histologic changes seen in idiopathic restrictive cardiomyopathy might also be seen in constrictive pericarditis. Table 5 summarizes the differentiation between restrictive cardiomyopathy and constrictive pericarditis.

Treatment

Both restrictive cardiomyopathy and constrictive pericarditis have progressive natures if they are left untreated. Treatment of restrictive cardiomyopathy depends on the histologic type and can range from chemotherapy for amyloidosis to phlebotomy and iron chelating agents for hemochromatosis. On the other hand, treatment for constrictive pericarditis is surgical, with pericardiectomy.

Although pericardiectomy is curative in these patients, it carries a high perioperative mortality risk.[41,42] The average operative mortality has been reported as 5.6% to 19%.[41-44]

Prognosis

The prognosis of restrictive cardiomyopathy is highly variable, depending on its type. Whereas certain types, such as hemochromatosis, can be reversed if diagnosed and treated early in the disease course, other types, such as amyloidosis, might run a malignant course.

Prognosis of patients undergoing pericardiectomy for constrictive pericarditis depends on several factors. Most importantly, prognosis depends on how well the surgical procedure is performed. Certain groups of patients will still do poorly despite a successful complete pericardiectomy. Patients with constrictive pericarditis with unfavorable outcome despite pericardiectomy are those with severe preoperative functional disability (New York Heart Association Class III/IV), preoperative renal insufficiency, presence of extensive nonresectable calcification, incomplete pericardial resection, history of radiation pericarditis, and concomitant restrictive cardiomyopathy.[41-45] Table 6 summarizes the prognostic factors involved in the outcome of pericardiectomy for constrictive pericarditis.

Table 6
Poor Prognostic Factors for Outcome of Pericardiectomy in Patients with Constrictive Pericarditis

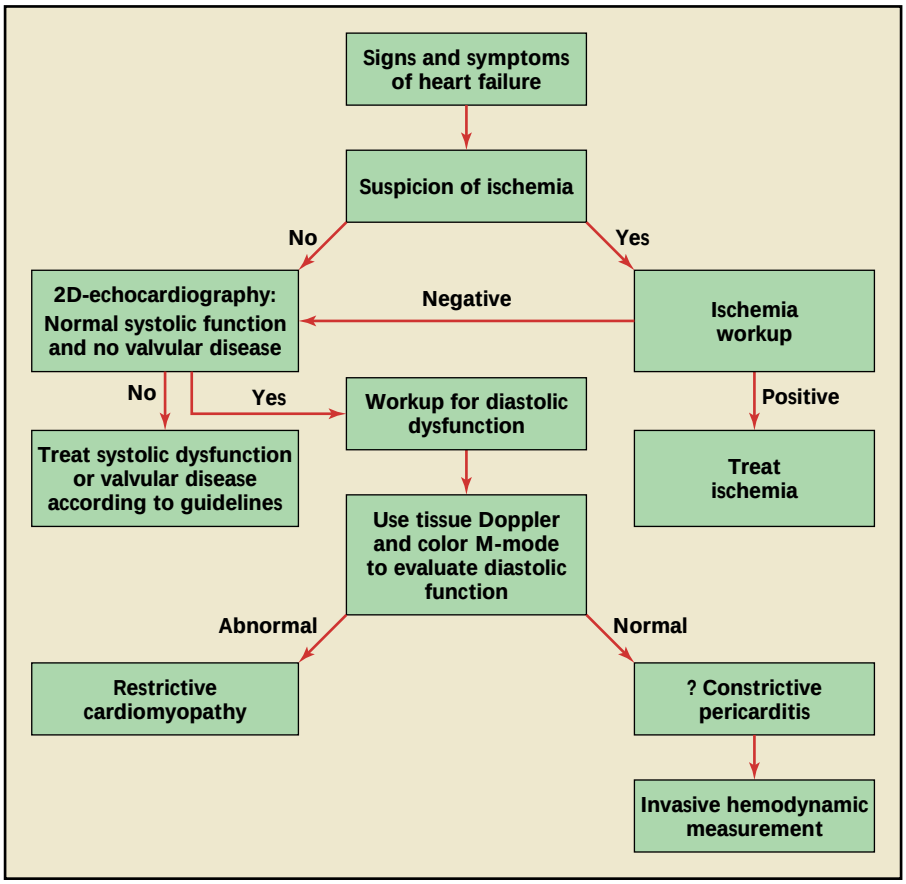
- Previous radiation
- New York Heart Association Class III/IV
- Age > 55 years
- Ascites
- Other organ failure/insufficiency (particularly renal failure)
- Extensive nonresectable calcification and/or incomplete pericardial resection
- Presence of restrictive cardiomyopathy

Discussion

Constrictive pericarditis and restrictive cardiomyopathy are 2 completely different disease entities that require different treatment approaches. Although these disease entities involve different parts of the heart, their clinical manifestations can be similar. The difficulty in differentiating constrictive pericarditis from restrictive cardiomyopathy seems to have been brought to clinicians' attention once their similar hemodynamic tracings were noted. In recent years, many investigators have reported new modalities for diagnosing these disorders and differentiating them from each other. Efforts have been made to avoid more invasive techniques for the diagnosis and also to avoid thoracotomy unless necessary for therapeutic purposes.

In the 1980s, Doppler echocardiography with simultaneous recording of respiratory changes of flow in the pulmonary vein and hepatic vein and over the tricuspid valve and mitral valve became popular as a diagnostic tool, with reasonable sensitivity and specificity in this setting. The difficulty of performing simultaneous respiratory recording and getting

Figure 7. *Suggested workup plan for patients with diastolic heart failure.*



a good signal from pulmonary or hepatic vein sites has demonstrated this method to be cumbersome at times. In the 1990s, tissue Doppler echocardiography and color M-mode were noted to be reliable tests with high sensitivity and specificity for diagnosing and differentiating these conditions. The noninvasive, low-cost, and comfortable nature of this method has made it a favorable tool for evaluation of patients with a suspicion of these disease entities. It still seems that a combination of complementary diagnostic modalities is needed to establish the final correct diagnosis. With the new advances made in tissue Doppler echocardiography, color M-mode, and radionuclide ventriculography, differentiation between constrictive pericarditis and restrictive cardiomyopathy is made with more certainty and less need for more invasive and cumbersome tests. Figure 7 shows a suggested workup plan for patients who present with signs and symptoms of diastolic heart failure.

Conclusion

Constrictive pericarditis and restrictive cardiomyopathy are 2 forms of diastolic heart failure that might have similar clinical presentations. Although in past the differentiation of these entities was difficult and required more invasive approaches, such as endomyocardial biopsy or even thoracotomy at times, the task has been made easier with the new advances made in the world of echocardiography. ■

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Main Points

- Constrictive pericarditis and restrictive cardiomyopathy are 2 forms of diastolic dysfunction with similar clinical presentations but differing treatment options. Whereas constrictive pericarditis has a curative potential with pericardiectomy, the prognosis of restrictive cardiomyopathy is highly variable.
- Many diagnostic modalities have been used to differentiate between these 2 disease entities; a combination of these tests is needed to correctly diagnose the cause of diastolic dysfunction.
- With advances in tissue Doppler echocardiography and color M-mode echocardiography, differentiation can be made with more certainty and less need for more invasive and cumbersome tests.
- Endomyocardial biopsy is essential in the final diagnosis of restrictive cardiomyopathy and is essential if thoracotomy is considered for diagnosis of constrictive pericarditis.
- The prognosis of restrictive cardiomyopathy is highly variable, depending on its type: certain types can be reversed if diagnosed and treated early, whereas others might run a malignant course; the prognosis of patients undergoing pericardiectomy for constrictive pericarditis depends on several factors, most importantly on how well the surgical procedure is performed.

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