Consensus Statement of Pericardial Diseases /Abridged Version

Argentine Society of Cardiology

Consensus Director
Dr. J. Horacio Casabé

Coordinators
Acute Pericarditis Chapter: Dr. Pablo Klin
Pericardial Effusion Chapter: Dr. Pablo Oberti
Cardiac Tamponade Chapter: Dr. Marcelo Trivi
Constrictive Pericarditis Chapter: Dr. Manuel Vázquez Blanco

INTRODUCTION

Although pericardial disease is very frequent found in daily clinical practice, there are no reliable data of its epidemiology in our country or published in the international literature. The Argentine Society of Cardiology entrusted a group of experts to write the first Argentine Consensus of Pericardial Diseases. In order to complete this task, we consulted the local and international literature and created four committees, each with a pericardial syndrome: 1) Acute Pericarditis, 2) Cardiac tamponade, 3) Pericardial effusions and 4) Constrictive Pericarditis. Each committee worked independently analyzing the literature, which in its great majority is level of evidence B and C. They are mainly observational descriptive studies of a not very large number of patients and expert opinions. Recently, however, well-designed therapeutic works have begun to appear, mainly for the treatment of acute and recurrent pericarditis. The three consensuses published so far were also taken into account. The present Consensus has mainly a clinical approach. To do this, we divided the disease into pericardial syndromes. Each of the syndromes is approached from an etiological, physiopathogical, clinical and therapeutic point of view, placing special emphasis on imaging diagnosis (echocardiography, computed tomography and magnetic resonance imaging) which due to their recent high development ("Multimodality images") can often provide an etiological diagnosis.

The degree of consensus reached and the levels of evidence for the final recommendations were based on SAC's Standardization and Consensus Area Regulations:

- **Class I**: Conditions for which there is evidence and/or general agreement that the treatment/procedure is beneficial, useful and effective.
- **Class II**: Conflicting evidence and/or opinion divergence about the usefulness, efficacy of the method, procedure and/or treatment.
- **Class IIa**: The weight of evidence/opinion favors usefulness / efficacy.
- **Class IIb**: Usefulness / efficacy are less well established.
- **Class III**: Evidence or general agreement that the treatment method/procedure is not useful / effective and in some cases can be harmful.

Regarding Levels of Evidence:

- *Level of Evidence A*: Strong evidence from randomized or cohort studies with adequate design to achieve statistically correct and biologically meaningful conclusions.
- *Level of evidence B*: Data from a single randomized clinical trial or large non-randomized studies.
- *Level of evidence C*: Consensus of expert opinion.

The Consensus Statement of Pericardial Diseases is published in its complete version on the website of the Argentine Society of Cardiology (http://www.sac.org.ar/area-de-consensos-y-normas/) with its corresponding complete literature. This publication summarizes the main concepts and recommendations of that document.

Abbreviations

aPTT	Activated partial thromboplastin time
CPK-MB	Creatine phosphokinase MB fraction
CAT	Computerized axial tomography
ECG	Electrocardiogram
LDH	Lactate dehydrogenase
NMR	Nuclear magnetic resonance
NSAID	Nonsteroidal anti-inflammatory drugs
PPS	Postpericardiotomy syndrome
TB	Tuberculosis
Tn	Troponin

Chapter 1
ACUTE PERICARDITIS

Coordinator
Dr. Pablo Klin

Participants
Dra. Carola Zambrano
Dr. Sergio Baratta
Dr. Jorge Lax
Dr. Máximo Santos

Contents

1.1. INTRODUCTION
Acute pericardial inflammation with or without associated effusion can occur as an isolated clinical problem or as manifestation of a systemic pathology. Acute pericardial diseases comprise a set of entities including acute pericarditis, myopericarditis and perimyocarditis, recurrent pericarditis, and incessant pericarditis.

1.2. DEFINITIONS
- *Acute pericarditis*: The diagnosis of acute pericarditis requires that at least two of the following criteria be met: typical chest pain (acute and pleuritic pain that improves in the seated position or with forward tilt), pericardial rub, suggestive electrocardiographic changes (diffuse ST-segment elevation or PR-segment depression) and presence of new pericardial effusion or worsening of a previous effusion.
- *Acute myopericarditis*: It defines the clinical presentation of acute pericarditis with myocardial necrosis marker (Tn I or T or CK-MB) elevation without focal or global left ventricular contractile function involvement assessed by Doppler echocardiography or magnetic resonance imaging (MRI).
- *Acute perimyocarditis*: It is defined as acute pericarditis with elevated myocardial necrosis markers (Tn I or T or CK-MB) and focal or global left ventricular contractile function involvement assessed by Doppler echocardiography or MRI.
- *Recurrent pericarditis*: Documented first pericarditis attack, followed by a period of at least 4-6 weeks free of symptoms followed by presentation of new pericarditis.
- Incessant pericarditis: Pericarditis lasting more than 4 to 6 weeks and less than 3 months without remission.
- *Chronic pericarditis*: Pericarditis lasting for more than 3 months.

In turn, pericarditis may be dry, fibrous or effusive regardless of its etiology.

1.3. CLINICAL PRESENTATION AND DIAGNOSIS

Table 1 describes the main clinical diagnostic elements of acute pericarditis.

Table 1. Recommendations for the diagnosis of acute pericarditis

Recommendation	Class	Level of evidence
The auscultation of a pericardial rub is highly suggestive of acute pericarditis.	I	B
The presence of progressive diffuse ST-segment elevation in the ECG is highly suggestive of acute pericarditis.	I	B
Characteristic retrosternal chest pain that increases with coughing and inspiration and sometimes improves with forward tilt is highly suggestive of acute pericarditis.	I	B
The occurrence of accelerated ESR, increased CRP, leukocytosis, increased troponins and LDH can guide the diagnosis.	I	B
The onset or increase of a previous pericardial effusion or signs of tamponade in this context is highly suggestive of acute pericarditis.	I	B
Chest X-ray may be normal or show signs of pericardial effusion or mediastinal or pulmonary disease.	I	B

ECG: Electrocardiogram. ESR: Erythrocyte sedimentation rate. CRP: C-reactive protein. LDH: Lactate dehydrogenase.

Diagnostic Sequence

Some authors have proposed a three-step protocol for the diagnostic management of pericardial diseases:

- Stage I: It includes medical history, physical examination, electrocardiogram (ECG), chest X-ray, tuberculosis (TB) assessment, serum antinuclear antibodies and thyroid hormone determinations, as well as other studies suggested after the initial evaluation.
- Stage II: It includes pericardiocentesis in patients with cardiac tamponade, suspected purulent pericarditis or large chronic pericardial effusions.
- Stage III: It includes a surgical pericardial biopsy in patients with persistent or recurrent tamponade following pericardiocentesis, and when effusion lasts more than 3 weeks after hospital admission without an etiological diagnosis.

1.4. ACUTE PERICARDITIS RISK STRATIFICATION (Tables 2 and 3)

Table 2. Acute pericarditis risk stratification

Recommendation	Class	Level of evidence
The following are increased risk characteristics in a patient with acute pericarditis. They are in general predictors of a non-viral, non-idiopathic etiology and of complications during follow-up:		
Presence of fever >38 °C.	I	B
Subacute outcome.	I	B
Echo-free diastolic spaces >20 mm in the echocardiogram or evidences of tamponade.	I	B
Failure of ASA or NSAID treatment.	I	B

ASA: Acetylsalicylic acid. NSAID: Nonsteroidal anti-inflammatory drugs.

Table 3. Hospitalization criteria for acute pericarditis.

Recommendation	Class	Level of evidence
Patients with acute pericarditis and with the following characteristics should be hospitalized for better control and treatment: Patients with high-risk characteristics (fever >38 °C, subacute evolution, echo-free diastolic space >20 mm or evidence of cardiac tamponade in the echocardiogram, failure with ASA or NSAID).	I	B
Evidence of myocardial involvement (myopericarditis).	I	B
Traumatic etiology or immunosuppression or concomitant anticoagulant therapy.	I	C

ASA: Acetylsalicylic acid. NSAID: Nonsteroidal anti-inflammatory drugs.

Basic etiological diagnosis (Etiological search: see Chapter 2: Pericardial effusions)

Although the clinical diagnosis of pericarditis is relatively simple, establishing the cause is often difficult. The causes of acute pericarditis are varied and the cardiologist must identify those that require specific therapies

In developed countries, the most common final diagnoses in immunocompetent hosts are idiopathic and viral pericarditis. Therefore, from a medical and practical perspective, a more accurate diagnosis is usually irrelevant for the management of most patients. However, the most important specific causes to be ruled out are tuberculous and neoplastic pericarditis and those associated with systemic (usually autoimmune) diseases, since they require specific treatments. Each of these causes has a frequency of about 5% of all unselected cases of pericarditis in developed countries.

Emerging causes should include those of iatrogenic etiology (percutaneous coronary interventions, pacemaker implantation, ablation procedures, among others). These are contemporary examples of cardiac injury syndromes, in which the etiology is determined by a combination of direct pericardial trauma, pericardial bleeding, and individual predisposition.

Radiological Characteristics

Chest X-ray, computerized axial tomography (CAT) scan and MRI findings are shown in Table 4.

Echocardiography

Echocardiography is essential for detecting pericardial effusion and evaluating concomitant structural heart disease or paracardiac disease, and CAT and MRI should be considered as additional methods to echocardiography in cases of loculated or hemorrhagic effusions, suspicion of pericardial thickening and constriction, as well as in the presence of pericardial masses, but also when echocardiographic findings are difficult to interpret or conflicting with clinical data, and when examination of the chest is necessary to evaluate possible neoplasia or TB. Presence of large pericardial effusions and/or cardiac tamponade is a strong indicator of a probable specific etiology.

Table 4. Abnormal pericardial patterns, their visualization on chest x-ray, computed tomography and magnetic resonance imaging*

Pattern	Anatomo-pathological origin	CXR	CAT	MRI	Interpretation (differential diagnosis)
Normal thickness		Lateral view between mediastinal and subepicardial fat	Thin line in front of the RA and RV, between the mediastinum and subepicardial fat +++	Signal-free lines between mediastinal and subepicardial fat ++	Absence of disease
↑Smooth thickness	Acute inflammatory process; Effusion	Thickened pericardial line in lateral view +	CAT scan values for DD +++	MRI signals for DD ++	Acute, subacute pericarditis, pericardial effusion, (DD according to liquid, semi-liquid, hemorrhagic, purulent or solid fluid)
↑Irregular thickness	Chronic inflammatory process	Irregular cardiac silhouette	+++	+++	Chronic pericarditis, pericardial fibrosis, tumor, metastasis, post surgical
↑Calcified irregular thickness	End-stage traumatic or hemorrhagic inflammatory process	↑Density	High CAT scan value +++	Weak signal ++	Pericardial calcification,, calcified tumors

* Adapted from Verhaert D, et al.
+ visible; ++ good; +++ optimal visualization
CAT: Computerized axial tomography. MRI: Magnetic resonance imaging. RA: Right atrium. RV: Right ventricle.DD: Differential diagnosis.

1.5. TREATMENT

The management of pericardial diseases in general and of acute pericarditis, in particular, has been carried out empirically over the years. Only in the last 10 years, following the publication of a series of randomized clinical trials, have the foundations of rational treatment been established on objective evidence.

Unlike other heart diseases, there are few randomized clinical trials on pericardial disease, and they include a small number of patients; consequently most of the diagnostic and therapeutic indications are based on levels of evidence B and C. Tables 5, 6 and 7 present the main recommendations for non-steroidal anti-inflammatory drugs (NSAID), colchicine and corticosteroid treatment, which constitute the main therapeutic tools.

Table 5. Anti-inflammatory treatment of acute pericarditis

Recommendation	Class	Level of evidence
Non-steroidal anti-inflammatory drugs are the first-choice drugs for the treatment of this disease.	I	B
Acetylsalicylic acid should be administered in doses of 2 to 4 grams/day.	I	B
Ibuprofen should be administered in doses of 1,000 -3,200 mg/day.	I	B
Indomethacin should be administered in doses of 75-150 mg/day.	I	B
Prolonged non-steroidal anti-inflammatory drug administration should be established until disappearance of symptoms and CRP and/or ESR normalization.	I	B

CRP: C-reactive protein. ESR: Erythrocyte sedimentation rate.

Table 6. Use of colchicine in acute pericarditis

Recommendation	Class	Level of evidence
In the absence of contraindications or specific indications, colchicine should be considered as first-line treatment associated with NSAID in acute and recurrent pericarditis.	I	A
The dose to be administered is initially 2mg/day during 1 to 2 days, and 1mg/day as maintenance dose divided into two intakes of 0.5 mg/day in patients <70 kg,. In case of intolerance (mainly diarrhea) it can be reduced to 0.5 mg/day.	I	B
Treatment duration should be at least 3-6 months.	I	B

NSAID: Non-steroidal anti-inflammatory drugs

Table 7. Recommendations for the administration of corticosteroids in acute pericarditis

Recommendation	Class	Level of evidence
Existing evidence for the use of corticosteroids in acute pericarditis is weak. They are predictors of recurrent pericarditis, probably due to involvement in the resolution of the initial viral presentation.	IIa	B
Corticosteroids should be indicated only in patients with general deterioration, recurrent pericarditis, connective tissue and autoimmune diseases or uremic pericarditis.	IIa	C
Low doses are recommended (prednisone 1-1.5 mg/kg for at least 1 month, with very gradual decrease only after obtaining stable remission with resolution of symptoms (similar to polymyalgia rheumatica).	IIa	B
If symptoms recur during the gradual decline of steroids, the last effective dose should be prescribed once more, keep it for 2-3 weeks and restart the gradual decrease again.	IIa	B
During the reduction of steroid doses (over a period of at least 3 months) the use of NSAID (ASA or ibuprofen) and colchicine (low doses, 0.5-0.6 mg/day) should be considered to improve digestive tolerance and maintained for at least 3 months.	IIa	B

NSAID: Non-steroidal anti-inflammatory drugs. ASA: Acetylsalicylic acid

Other drugs

Gastric protectors: Gastric protection with ranitidine or proton pump inhibitors (omeprazole and related drugs) should be indicated to all patients.

Anticoagulants: During hospitalization, if the patient requires the use of anticoagulants, the use of heparin is recommended under strict control of activated partial thromboplastin time (aPTT). Anticoagulants can also be prescribed for other indications (e.g., atrial fibrillation, confirmed or suspected pulmonary embolism, previous prosthetic valve implantation, etc.).

Antiarrhythmics: The association of acute pericarditis with atrial fibrillation, atrial flutter, and other supraventricular arrhythmias may require the use of antiarrhythmic drugs. Among these drugs, the most frequently indicated are Class I C (flecainide, propafenone) and Class III (amiodarone) agents. There is frequent need to prescribe beta-blockers for the control of ventricular response, although hypovolemia, hyperthyroidism, cardiac tamponade and the consequences of systemic inflammatory activity observed in patients with acute pericarditis should be ruled out in patients with sinus rhythm.

Other immunosuppressors: Azathioprine (75/100 mg/day) or cyclophosphamide has been proposed in patients who present with recurrence and do not respond adequately to conventional treatment (including steroids). In these cases, the experience in the use of these drugs is scarce. (Class IIa, level of evidence C indication).

Drugs for the prevention of osteoporosis: The addition of calcium and vitamin D (1,500 mg/day and 800 IU/day, respectively) or some form of activated vitamin D (such as alpha-calcidiol 1 μg/day or calcitriol 0.5 μg/day) should be recommended to all patients receiving systemic steroids to restore normal calcium balance. The use of bisphosphonates is also recommended for the prevention of bone resorption in all men and postmenopausal women in whom systemic steroid therapy is initiated at prednisone doses >5 mg/day or equivalent drugs.

Restriction of physical activity

Rest is part of the treatment, and should be maintained until the patient is asymptomatic. It is recommended for a period of 4-6 weeks, together with serial echocardiographic monitoring.

Regarding the resumption of sports activity, it should be considered after a period of 6 months, only if the patient is asymptomatic, with normalization of ECG, inflammation markers and ventricular function parameters.

It is advisable to perform Holter monitoring and a stress test to assess functional capacity (conventional exercise stress test or stress echo) prior to resumption of sports activity.

1.6. ACUTE REFRACTORY AND RECURRENT PERICARDITIS

Pericardiocentesis has not shown therapeutic benefit and has been exclusively used in patients with relapsed pericarditis, very symptomatic and refractory to treatment or with tamponade, suspected purulent or neoplastic pericarditis.

The cases that recur after the decrease of steroid doses (very frequent) should not be considered as refractory; this definition should only apply to those cases requiring long-term, unacceptably high doses of steroids for their control (e.g. prednisone >25 mg/day). In these cases (<5% of recurrence cases) a number of immunosuppressive drugs (azathioprine, cyclophosphamide, methotrexate, hydroxychloroquine, intravenous immunoglobulins) have been used, azathioprine being the drug of choice at doses of 2-3 mg/kg/day. There is no solid evidence to recommend their use. If indicated, therapy should be personalized in each case, under strict follow-up of experienced physicians in their use and with informed consent.

In the more difficult cases, the association of three drugs (a NSAID, colchicine and low doses of prednisone) could be considered, but the evidence for this association is very scarce.

Although a systematic review of cases published between 1966 and 2006, including a total of 230 patients with recurrent idiopathic pericarditis followed-up for approximately 5 years, showed that the prognosis is generally excellent and that complications are rare, long-term prognosis of idiopathic recurrent pericarditis has not been clearly established and the possibility of progression to constrictive pericarditis should be controlled.

Role of pericardiectomy, pericardial window and other interventional techniques in patients with acute pericarditis

The European guidelines (2015) considered pericardiectomy for the treatment of frequent and highly symptomatic recurrences, refractory to medical treatment with a Class IIa recommendation. Other reported indications include recurrences with cardiac tamponade and evidence of severe toxicity due to steroid use.

Pericardiectomy is generally considered a therapeutic option of dubious efficacy in recurrent idiopathic pericarditis and should be considered only in exceptional cases. It is indicated only for frequent and highly symptomatic recurrence, resistant to maximum medical treatment (Class IIa, level of evidence B indication).

Prior to pericardiectomy, the patient should be steroid-free for a period of several weeks. Post-pericardiectomy recurrence cases have been described, probably related to incomplete pericardial resection. There have been reported cases in which, for reasons not completely clarified, the removal of the pericardium ended the

syndrome, but in other cases it continued its progression or relapsed after the intervention following an apparent clinical control.

Indications of pericardiectomy in cases of recurrent pericarditis are currently based on expert opinion rather than on its proven benefits. On the other hand, the benefits of pericardiectomy are well established in constrictive pericarditis.

Transient constriction has been reported in up to 9% of patients with acute idiopathic pericarditis, where the characteristics of constriction were noted in the subacute stage of pericarditis, once the effusion had disappeared or was minimal. In patients developing "acute" pericardial constriction following an acute pericarditis episode, coursing with hemodynamic stability, an initial conservative strategy (anti-inflammatory drugs, colchicine, with or without associated steroids) may be applied during 2-3 months, prior to considering pericardiectomy.

Acute pericarditis in the postoperative period of cardiovascular surgery. Postpericardiotomy syndrome.

Postpericardiotomy syndrome (PPS) is a potential complication of cardiovascular surgery that can develop within the first 6 months, mainly around the first postoperative month, whose incidence reaches up to 20-40% of cases. Its etiopathogenesis is not fully understood, but an autoimmune mechanism is postulated. Conventional treatment includes steroids. However, a controlled clinical trial evaluating the response to colchicine administration (COPPS) has been published. This study included 360 patients coursing the third day of cardiovascular surgery and who were randomized to placebo or colchicine. The primary endpoint was to develop PPS at 12 months. The secondary endpoints were combined rates of cardiac tamponade, constrictive pericarditis, recurrence, and hospitalizations for pericardial pathology. Colchicine significantly reduced the incidence of PPS (8.9% vs. 21.1%, p=0.002 and NNT=8). It also reduced the secondary endpoint (0.6% vs. 5.0%, p=0.024). The rate of adverse effects (mainly related to digestive intolerance) was similar in both groups (p=ns). The authors concluded that colchicine was safe and effective in preventing PPS and its related complications and could reduce by 50% the risk of developing PPS after cardiovascular surgery.

Chapter 2
PERICARDIAL EFFUSIONS

Coordinator

Dr. Pablo Oberti

Participants

Dr. Pablo Desmery
Dra. Amalia Elizari
Dr. Eduardo Guevara
Dr. Ricardo Obregón

Contents

2.1. DEFINITION

The presence of pericardial effusion is defined when the amount of pericardial fluid present in the pericardial space exceeds the amount considered normal (approximately 50 ml). Pericardial effusions are commonly diagnosed in clinical practice either as an incidental finding or as manifestation of a cardiac or systemic disorder. Pericardial effusions may be, according to the rate of development, of slow fluid accumulation or of sudden progression rapidly evolving to cardiac tamponade. In general, the prognosis depends on the cause and size of the effusion, although small effusions are not always benign.

There is scarce epidemiological information in our setting. Relevant data from a recent Italian study reported an incidence of 3% effusions in 6 years of experience in patients referred to echocardiography labs and a prevalence of 9%.

2.2. PHYSIOPATHOLOGY

The normal pericardial sac contains approximately 10-50 ml of pericardial fluid that acts as a lubricant between the pericardial layers.

The presence of fluid in the pericardium raises intrapericardial pressure, but the magnitude of this elevation depends not only on the absolute amount of the fluid but also on the velocity with which it has accumulated and the physical characteristics of the pericardium.

Any pathological condition may cause an inflammatory process with the possible increase in the production of pericardial fluid (exudate). An alternative mechanism of pericardial fluid accumulation may be lower reabsorption due to an increase in systemic venous pressure, usually as a result of congestive heart failure, nephrotic syndrome, cirrhosis, or pulmonary hypertension (transudate). Generally, pericardial fluid accumulates in the posterior portion of the left ventricle, following gravitational forces. When the effusion is moderate to severe, its distribution is usually circumferential. The presence of isolated pericardial fluid in the anterior portion, without prior surgical or pericarditis history, should be more likely considered as pericardial fat rather than pericardial fluid. (7) The aid of CAT scan or MRI is often useful for differentiation.

In case of pericardial effusion, the first step is to assess its size and hemodynamic repercussion, as well as its possible association with concomitant diseases.

The currently available guidelines for the use of echocardiography have recommended echocardiography as the first diagnostic tool for this evaluation.

Pericardial effusions can be classified according to their type of onset as acute, subacute or chronic (with more than 3-month progression), by their distribution as circumferential or loculated, by their hemodynamic impact as none, cardiac tamponade or effusive-constrictive, by their composition as exudate, transudate, hemorrhagic, chylopericardium or pyopericardium, with air or gas derived from a bacterial infection, and especially by their size as mild, moderate or severe. This evaluation, which can be derived from a semiquantitative echocardiographic analysis or a CAT scan or MRI has also proved to be useful to estimate the risk of specific etiologies and complications during follow-up,

The sudden increase of 100-200 ml in pericardial volume, as in hemopericardium can raise pericardial pressure up to 20-30 mmHg with acute tamponade (acute or surgical tamponade). In contrast, slow pericardial fluid accumulation may allow pericardial distension to accumulate up to 1-2 liters of pericardial fluid without the development of tamponade until advanced stages, often due to intercurrent events (chronic or medical tamponade).

2.3. ETIOLOGY (Table 1)

In the case of a patient presenting with pericardial effusion, the first challenge is to identify its etiology. A wide variety of etiological agents may be responsible for pericardial effusions, since all known agents of pericardial disease may be causative agents.

Common causes include infections (viral, bacterial, TB), cancer, heart failure, connective tissue diseases, pericardial injury (post-infarction effusion, postpericardiotomy syndromes, post-traumatic pericarditis, etc.), metabolic causes (renal failure, hypothyroidism, hypoalbuminemia), myopericarditis disease and acute aortic syndromes secondary to the use of certain drugs (e.g. minoxidil).

Table 1. Etiology of pericardial effusions

Infective
-Viral (echovirus, coxsackievirus, influenza, CMV, EBV, HIV, others)
-Bacterial (TB, cocci, others)
-Fungal (histoplasmosis) and parasites (toxoplasma), rare
Non-infective
Hemodynamic (CHF, pulmonary hypertension and hypoalbuminemia)
Metabolic (uremia, myxedema)
Systemic inflammatory diseases (SLE, Sjögren, RA, others)
Pericardial injury syndromes (post-AMI, post-pericardiectomy, others)
Metastatic tumors (lung, breast, lymphomas, melanomas, others)
Trauma (penetrating and non-penetrating thoracic injury, iatrogenic, others)
Mediastinal radiation (recent or remote)
Primary tumors (mesothelioma)
Drugs and toxins (immunosuppressors, phenytoin, isoniazid, others)

CMV: Cytomegalovirus. EBV: Epstein-Barr virus. HIV: Human immunodeficiency virus. TB: Tuberculosis. CHF: Congestive heart failure. SLE: Systemic lupus erythematosus. RA: Rheumatoid arthritis. AMI: Acute myocardial infarction.

2.4. CLINICAL PRESENTATION

The clinical presentation of pericardial effusion mainly depends on the speed of installation and etiology of the effusion, often with symptoms that may be related to the causal disease. The rate of fluid accumulation is critical for the clinical presentation. If this is fast, such as after trauma or iatrogenic perforation, the evolution is dramatic and only small amounts of blood are responsible for the rapid increase of intrapericardial pressure with development of cardiac tamponade in minutes or hours. Conversely, a slow accumulation of pericardial fluid allows the development of a large effusion in days or weeks before causing a significant increase in intrapericardial pressure with development of signs and symptoms. (

Pericardial effusion may be asymptomatic or give nonspecific symptoms which may include exercise dyspnea with progression to orthopnea, chest pain and/or feeling of fullness. Occasionally, the symptoms can be provoked by local compression and include nausea, dysphagia, hoarseness and hiccup due to phrenic nerve irritation. Other symptoms such as fever, cough, weakness, fatigue, anorexia, chills, and palpitations can also occur, reflecting the compressive character of the pericardial fluid on adjacent anatomical structures or be related to the causal disease

Many patients with a large (more than 20 mm by echocardiography), chronic (more than 3 months), idiopathic pericardial effusion may be asymptomatic and remain clinically stable for many years. However, this condition may lead to the development of cardiac tamponade in up to 29% of patients. The trigger for tamponade is unknown, but hypovolemia, paroxysmal tachyarrhythmias or recurrent acute pericarditis may precipitate it.

Physical examination in patients with pericardial effusion may be normal. In some cases, due to the increase in intrapericardial pressure and the presence of a large volume of fluid, there is distension of the neck veins with an increase in jugular venous pressure and decrease of cardiac sounds during auscultation. Pericardial friction rub is not commonly found; however there is no precise correlation between this finding and the presence or size of the effusion.

2.5. DIAGNOSIS

The diagnostic algorithm should be guided by epidemiology and clinical presentation to avoid a series of long blinded studies. Once its presence and severity is diagnosed, the most complex task consists in the evaluation of etiology.

In developing countries there is high frequency of pericardial effusions related to tuberculosis and/or human immunodeficiency virus. The non-idiopathic and non-viral etiologies are associated with increased risk of severe effusion and cardiac tamponade, requiring mandatory pericardiocentesis.

Pericardial biopsy by pericardioscopy has shown to be useful in the diagnosis of neoplastic effusion.

a. Electrocardiogram

Pericardial effusion may cause non-specific electrocardiographic changes as reduction in the QRS voltage (defined as the sum of R+S waves in L1+L2+L3 <15 mm) and diffuse flattening of the T waves.

In case of severe pericardial effusion and cardiac tamponade, there is evidence of electrical alternans (change in the shape and voltage of the QRS complex in alternate beats) due to the oscillating movement of the heart in a cavity with a great volume of fluid.

b. Chest X-ray

Chest X-ray can show the presence of an enlarged cardiac silhouette with clear lung fields. This finding, which may be unexpected in the routine exam of an asymptomatic patient, should pose the doubt of pericardial effusion. An enlarged cardiac silhouette generally appears when more than 200-250 ml of fluid accumulates in the pericardial sac. Therefore, a normal or slightly abnormal chest X-ray does not rule out the presence of a low volume pericardial effusion. In some cases, the cardiac silhouette adopts a globular bottle-like shape, effacing the contour along the left cardiac border, and concealing the hilar vessels.

Some studies have shown that the enlarged cardiac silhouette in the chest X-ray has moderate sensitivity (70%) but low specificity (41%) for the diagnosis of pericardial effusion.

On the other hand, radioscopy enables observing the absence of pulsations in the cardiac silhouette or absence of changes in its size and configuration associated with inspiration.

c. Echocardiography (Table 2)

Echocardiography is the most frequently used diagnostic method, as it allows a fast and easily accessible semiquantitative assessment of effusion size and its hemodynamic effect. Table 2 summarizes the recommendations for echocardiography in pericardial effusion.

Table 2. Recommendations for the use of echocardiography in the management of pericardial effusion

Recommendation	Class	Level of evidence
Echocardiography is recommended to establish the diagnosis and severity of pericardial effusion, its hemodynamic impact, and quantify its severity to define possible drainage.	I	B
Echocardiography is recommended to guide pericardiocentesis.	I	B
Echocardiography is recommended to follow-up the evolution of pericardial effusion and to diagnose and assess the progression of cardiac tamponade.	IIa	B
Echocardiography is recommended for the initial diagnosis of acute pericarditis.	IIb	C

d. Computerized axial tomography and magnetic resonance imaging (Table 3)

Although echocardiography is the main and first method used for the diagnosis of pericardial effusion, other approaches and methods can complement its assessment. Together with echocardiography, cardiac CAT scan and MRI have shown to be very useful in the search of the cause for effusion. Magnetic resonance imaging can also provide a combined morphological and functional assessment. The pericardium can be adequately evaluated with both modalities due to the natural contrast presented by the pericardial layers separated with pericardial fluid and the contiguous adipose tissue in the mediastinal and subepicardial spaces. Computerized axial tomography density measurements and MRI signal analysis allow better characterization of pericardial fluid than echocardiography. Magnetic resonance imaging is superior to CAT scan in differentiating thickened pericardium fluid, especially in exudative effusions with high protein content. Conversely, CAT scan can detect even minimal amounts of pericardial calcium, while MRI may err in its detection. Computerized axial tomography usually requires less time than echocardiography and MRI, but demands the use of intravenous contrast and ionizing radiation. Table 3 summarizes the recommendations for the use of MRI and CAT scan in patients with pericardial effusion.

Table 3. Recommendations for the use of CAT scan or MRI in the diagnosis of pericardial effusion

Recommendation	Class	Level of evidence
Computed tomography and/or magnetic resonance imaging are recommended in case of suspected neoplastic pericarditis to document pericardial tumor thickening or suspicious images in the neighboring structures.	I	B
Computed tomography and/or magnetic resonance imaging are recommended to document localized pericardial effusion or localized cardiac tamponade, especially in the postoperative period of cardiac surgery.	I	B
CAT and MRI are alternative techniques to assess pericardial effusion, mainly to determine its location, pericardial thickening, presence of calcium, masses and associated thoracic involvement.	IIa	B
Computed tomography and/or magnetic resonance imaging are recommended to evaluate chronic pericardial effusions (>3 months).	IIa	B
Computed tomography and/or magnetic resonance imaging are recommended to establish the nature of the pericardial fluid through tissue characterization, attenuation (CAT scan) or signal intensity (MRI).	IIa	B
Computed tomography and/or magnetic resonance imaging are recommended as initial diagnostic method and quantification of pericardial effusion in common situations.	III	B

CAT: Computerized axial tomography. MRI: Magnetic resonance imaging.

Analysis of pericardial fluid (Table 4)

The analysis of pericardial fluid can establish the diagnosis of infective or neoplastic effusion. The differentiation between a tuberculous or neoplastic origin can be determined with the presence of low adenosine deaminase and high carcinoembryonic antigen levels. The identification of the tuberculous cause of effusion is especially important due to the high mortality of the disease if not adequately treated, together with an elevated risk of progression to constrictive pericarditis (30-50% of cases). In case of suspected bacterial infection, at least three samples of pericardial fluid should be taken for aerobic and anaerobic germ cultures, as well as for blood cultures.

The effusion may appear as transudate (hydropericardium), exudate, pyopericardium, chylopericardium or hemopericardium. The analysis of pericardial fluid will help to diagnose the type of effusion: density (>1.015), protein level (>3.0 g/dl), pericardial fluid to plasmatic protein ratio >0.5, LDH >200 mg/dl, plasma/pericardial fluid ratio >0.6 and glucose can separate exudates from transudates. Purulent effusions with positive cultures have a significantly lower amount of glucose than non-infective effusions, as well as a higher white cell count compared with mixedema.

The diagnosis of autoimmune effusions is established by: 1. Increased number of lymphocytes and mononuclear cells (>5,000/mm3) or the presence of antisarcolemmal antibodies; 2) Signs of myocarditis in myocardial biopsy; 3) Exclusion of viral infection; 4) Exclusion of TB and other bacterial infections; 5) Absence of neoplastic infiltration in the pericardial fluid; and 6) Exclusion of systemic and metabolic diseases and uremia.

Table 4. Recommendations for the analysis of pericardial fluid in the diagnosis of pericardial effusion

Recommendation	Class	Level of evidence
The pericardial fluid should be analyzed in the following situations: Suspicion of purulent, tuberculous or neoplastic pericarditis.	I	B
In all patients requiring pericardial drainage as in tamponade treatment.	I	B
In patients with moderate to severe amount of fluid without confirmed etiology, unresponsive to anti-inflammatory treatment.	I	B
Samples should be sent to the microbiology laboratory for Gram staining, bacterial and mycological cultures, acid-alcohol-resistant bacilli staining and to the pathology laboratory for cytology, in addition to biochemical parameter measurements and special determinations (interferon, ADA).	I	A
The biochemical analysis of the fluid should be performed to obtain protein, LDH, glucose and cellular count values.	IIa	C
The suspicion of neoplasia demands not only a cytological analysis, but also tumor marker assessments, as carcinoembryonic antigen (CEA) and serum cytokeratin-19 fragments (CYFRA 21-1).	IIa	C
The use of other markers as alpha-fetoprotein and carbohydrate antigens (CA) applied according to patient characteristics (CA 125, CA 15-3, CE 19-9) are difficult to interpret, as the cut-off values are not determined and their clinical usefulness is uncertain.	IIb	C
Routine use of virological studies in pericardial fluid is not recommended and is not used in clinical practice, except in special situations as immunocompromised patients in whom CRP, immunofluorescence studies or viral cultures may be necessary.	IIb	B
A pericardial biopsy should be obtained in case of surgical access.	IIb	B
The use of pericardioscopy allows the identification of injured areas, increasing the diagnostic yield with a minimally invasive procedure.	IIb	B
A blind pericardial biopsy should be performed according to the clinical context, as the increase of diagnostic accuracy is not higher than 10%.	IIb	B

ADA: Adenosine deaminase. LDH: Lactic dehydrogenase. CRP: C-reactive protein.

2.6. TREATMENT (Table 5)

Whenever possible, treatment of pericardial effusion should be oriented to its etiology. In 60% of cases, the effusion is associated with known causes and the treatment should be oriented to the underlying cause. When the pericardial effusion id associated with pericarditis, management should focus on pericarditis treatment.

Table 5. Indications for the treatment of pericardial effusion

Recommendation	Class	Level of evidence
All patients with known etiology should receive medical treatment for the underlying disease.	IIa	B
Patients with pericardial effusion of inflammatory or idiopathic etiology should receive aspirin or NSAID (ibuprofen) and assess their response.	IIa	B

NSAID: Nonsteroidal anti-inflammatory drugs

2.6.1. Pericardial drainage procedures (Table 6)

Pericardial drainage procedures can be performed for diagnostic or therapeutic purposes (e.g. in patients with cardiac tamponade).

Various invasive procedures, ranging from simple needle pericardiocentesis to open surgery evacuation, are useful for pericardial drainage. The selection of a particular procedure depends mostly on the etiology of the pericardial effusion. In patients with viral or idiopathic pericardial effusions, a simple pericardiocentesis is usually enough for its evacuation.

The general consideration is that pericardial drainage procedures are not routinely justified in patients without hemodynamic involvement.

Table 6. Indications for the use of pericardiocentesis

Recommendation	Class	Level of evidence
Pericardiocentesis is indicated in case of suspected purulent or tuberculous pericardial effusion.	I	B
Pericardiocentesis is indicated for diagnostic purposes in pericardial effusions >20 mm by echocardiography and in patients with minor effusions.	IIa	B
Pericardiocentesis is indicated in patients with massive chronic pericardial effusion (echo-free spaces in anterior and posterior pericardial sacs >20 mm).	IIa	B
Pericardiocentesis is indicated in patients with neoplastic pericardial effusion.	IIa	B
Pericardiocentesis should be avoided in pericardial effusion secondary to acute aortic syndrome.	III	B
Pericardiocentesis should be avoided in mild or moderate pericardial effusion without hemodynamic involvement.	III	C

2.7 PERICARDIAL CYSTS AND CONGENITAL PERICARDIAL ABNORMALITIES

1. Pericardial cysts.
2. Pericardial diverticula
3. Congenital absence of the pericardium.

Pericardial cysts and pericardial diverticula are, mainly, congenital malformations produced by an embryological alteration of the celomic cavity. They are benign, rare anomalies (7% of mediastinal tumors and 20% of cardiac and pericardial tumors) with an incidence of 1 per 100,000. The clinical characteristics and the diagnostic and recommended therapeutic methods for these rare diseases are detailed on the website version of this document

Chapter 3
CARDIAC TAMPONADE

Coordinator
Dr Marcelo Trivi

Participants
Dr. Claudio Higa,
Dr Martín Lombardero
Dr Guillermo Vaccarino
Dr Mariano Benzadón.

Contents

3.1. INTRODUCTION
Definition: Cardiac tamponade is a clinical condition in which the increase in pericardial fluid raises pressure in the heart chambers above central venous pressure, reducing venous return and anterograde flow.

3.2. ETIOLOGY
The causes of cardiac tamponade are mostly the same as those for pericardial effusion and constrictive pericarditis. However, most frequently cardiac tamponade presents associated with any of these conditions. Generally, three types of tamponade can be identified: serous, serohematic and hemopericardium. The main causes are:
- Serous or serohematic: viral, autoimmune or metabolic.
- Hematic or serohematic: cardiac rupture, aortic dissection, iatrogenic or neoplastic perforation.

This differentiation according to whether the type of effusion is blood or another fluid has prognostic and therapeutic implications. Hemopericardium is more severe than serous or serohematic effusion and, except in neoplasms, frequently requires emergency surgery. With the increasing number of procedures, iatrogenic tamponade is more frequent in interventional centers. Specially, radiofrequency ablation and endocavitary electrode implantation are expanding as frequent causes of cardiac tamponade.

3.3. PHYSIOPATHOLOGY
The pericardium has the ability of adapting to physiological changes in the volume of fluid occupying the space between its serous and visceral layers, but if the amount of fluid exceeds its stretching limit, it results in compression of cardiac chambers (“last drop phenomenon”), first the right and then the left chambers. This generates diastolic failure of heart filling which is exacerbated during inspiration, since the reduction in intrathoracic pressure raises right chamber filling (increasing right murmurs) building up the amount of blood in the pulmonary bed. The interventricular septum bends to the left decreasing left ventricular ejection and anterograde flow. This reverts during expiration: intrathoracic pressure increases, the right heart filling is reduced and the left heart filling increases. This is the explanation of the wrongly termed paradoxical pulse, as it is the escalation of a normal physiological phenomenon. Classically measured with a sphygmomanometer, it is currently more difficult to assess with electronic devices, though it can be easily observed in the pulse oximetry tracing in intensive care unit monitors. It also explains the marked respiratory variations seen with cardiac Doppler both in atrioventricular valve flow and aortic flow.

In addition, as the pericardium surrounds all the cardiac chambers without compartments, the increase in pericardial pressure is uniform in all the cardiac chambers. This explains the equalization of pressures during right cardiac catheterization: mean right atrial pressure is equal to pulmonary capillary pressure, which represents left atrial pressure, and equal to the end-diastolic pressure of both ventricles.

3.4. CLINICAL PRESENTATION
It depends on the velocity of fluid accumulation in the pericardial cavity. It can be acute or subacute, of low pressure or occult and regional.

Acute: It presents within minutes (e.g. in trauma, rupture due to infarction and aortic dissection), it is similar to cardiogenic shock (cyanosis, vasoconstriction, oliguria, symptoms of cerebral low blood flow) and requires urgent reduction of intrapericardial pressure.

Subacute: It is produced in the term of days or weeks (as in neoplasms, uremia and idiopathic cardiac tamponade), both due to low cardiac output as venous return involvement, manifested by dyspnea, chest pain, edema resulting from a compensating mechanism, asthenia and hypotension. The patient becomes thirsty and increases his fluid intake to raise venous pressure and overcome the pericardial pressure. Therefore, diuretics must be administered with caution as they decrease venous pressure and may trigger or worsen cardiac tamponade. In this case, the treatment consists in the removal of the pericardial fluid and use of diuretics should not be attempted.

Cardiac tamponade with low intrapericardial pressure: Its etiology is similar to the rest of tamponades. It presents in conditions of severe hypovolemia (hemorrhagic trauma, hemodialysis) where pericardial and diastolic pressures are only 6 to 12 mmHg. It is detected echocardiographically by collapse of both right chambers and respiratory changes in the transmitral and tricuspid flow. Occasionally, low cardiac output and paradoxical pulse symptoms are clinically found.

Regional cardiac tamponade: It is produced by hematomas or eccentric effusions, where only some chambers are selectively compressed. It may be triggered secondary to pericardiotomy or infarction and is generally asymptomatic. It can be considered as a veritable diagnostic challenge as it is detected by high clinical suspicion, needing additional echocardiographic studies (subcostal or transesophageal) and other imaging techniques (computed tomography).

3.5. DIAGNOSIS

Table 1 describes the recommendations of the different diagnostic methods used for cardiac tamponade.

Table 1. Diagnosis of cardiac tamponade

Recommendation	Class	Level of evidence
Front-view chest X-ray.	I	C
Transthoracic echocardiogram.	I	B
Transesophageal echocardiogram in case of non-diagnostic transthoracic echocardiography.	I	B
Transesophageal echocardiogram in case of suspected aortic dissection or cardiac rupture.	I	B
Transesophageal echocardiogram in the postoperative period of cardiac surgery.	I	B
Transesophageal echocardiogram in ventilated patients.	IIa	C
Thorax CAT scan due to diagnostic doubts in stable patient.	IIa	C
Thorax CAT scan or MRI to establish etiology.	IIb	C
Thorax CAT scan with contrast or MRI in decompensated patients.	III	C

CAT: Computerized axial tomography. MRI: Magnetic resonance imaging

Pericardiocentesis technique

See technique description in Chapter 2 of the complete version on the website.

Table 2. Recommendations for pericardiocentesis in cardiac tamponade

Recommendation	Class	Level of evidence
Hypotensive or in shock patient.	I	B
Remote postoperative period of cardiac surgery (Dressler syndrome).	IIa	B
Remote postoperative period of cardiac surgery with tabicated effusion.	IIb	C
Aortic dissection or cardiac rupture without shock (surgery is the recommended treatment).	III	C
Immediate postoperative period of cardiac surgery (reoperation is the recommended treatment.	III	C

3.6 POSTOPERATIVE CARDIAC TAMPONADE

Pericardial effusion is a common finding in the postoperative period of cardiac surgery, approximately affecting half of operated patients. Most of these effusions will course without hemodynamic involvement and only 2% of cases need an invasive procedure to remove the content in the pericardial cavity. Postoperative cardiac tamponade is an entity difficult to diagnose, which if not quickly detected may be potentially fatal. In fact, cardiac tamponade is one of the main causes of cardiorespiratory arrest in the immediate postoperative period of cardiac surgery.

The clinical characteristics and recommended conducts for this clinical condition are detailed in the online version of this document.

Chapter 4
CONSTRICTIVE PERICARDITIS

Coordinator:
Dr. Manuel Vázquez Blanco

Participants:
Dr. Gustavo Avegliano
Dr. Federico Cintora
Dr. Alberto Domenech
Dr. Miguel Rubio
Dr. Ariel K. Saad

Contents

4.1. DEFINITION

The normal pericardium is a thin fibroelastic membrane with two layers, a visceral and a parietal layer, separated by a small amount of fluid, which enclose the heart and facilitate its movement. Different diseases can modify these characteristics and, through a single or recurrent inflammatory process, cause loss of its elasticity with or without increased thickness, fibrosis, calcification and eventually fluid accumulation. These changes end by hampering myocardial expansion and restricting chamber filling, jeopardizing the adaptation of the heart to the physiological demands.

4.2. PHYSIOPATHOLOGY

In constrictive pericarditis, the stiff pericardium, thickened or not, limits myocardial distension, obstructing right ventricular filling. Diastolic pressure increases reducing venous return. At the same time, pressure in the pulmonary veins decreases and this also reduces left heart filling. Since the myocardium is not altered, the early filling phase is fast due to the existing obstacle and next, the thickened, rigid myocardium limits subsequent expansion. This gives rise to a typical curve of rapid increase and decrease and then a plateau in the hemodynamic tracing (square root signal or dip and plateau curve). At the same time, as the limitation involves both chambers, diastolic pressures tend to become equal.

The preservation of diastolic function allows differentiating constrictive pericarditis from cardiomyopathies which progress with restricted filling. In pericarditis, ventricular function tends to remain stable until advanced stages of the disease, in which the fibrous process can invade and hinder ventricular function.

As a consequence of restricted filling, cardiac output decreases and venous pressure increases, giving rise to the signs and symptoms characterizing this disease.

4.3. ETIOLOGY

Pericarditis is a relatively frequent disease triggered by multiple causes, but its progress to constriction is relatively rare. However, we should remember that any process affecting the pericardium may cause its fibrosis, thickening and calcification, thus generating restriction to filling.

The most common causes are:

- Idiopathic or viral: 42-49%
- Post cardiac surgery: 11-37%
- Post radiation therapy: 9-31% (breast cancer, lymphomas, lung cancer)
- Connective tissue diseases: 3-7% (lupus, rheumatoid arthritis, sclerodermia, mixed disease)
- Post infective disease: 3-6% (tuberculosis, other germs)
- Others: 1-10% (drugs, neoplasms, thoracic trauma, chronic renal failure, post-infarction, asbestosis).

It is estimated that approximately 9% of patients suffering from acute pericarditis will develop in time pericardial constriction, although the true incidence depends mainly on the cause. Thus, in a prospective study including 500 consecutive patients, the risk of developing this complication was <2% after a mean follow-up of 6 years, with wide variations depending on the etiology. When the cause was viral or idiopathic, risk was 0.5% and 8.3% when it depended on another etiology. In this last group, the incidence of constriction per 1000 patients/year was 4.4 cases in patients suffering from a connective tissue disease, 6.3 cases in those with a neoplastic disease, 31.6 cases for those with tuberculosis etiology and 52.7 cases for those suffering purulent pericarditis.

4.4. CLINICAL PRESENTATION

The filling restriction of cardiac chambers during the course of pericarditis may adopt different clinical presentations:

- Chronic constrictive
- Transient constrictive
- Effusive constrictive
- Occult constrictive
- Localized constrictive

4.5. DIAGNOSIS (Table 1)

Clinical manifestations

Symptoms

1. Edemas: These may be localized or generalized, constituting in some cases an edematous ascites syndrome.
2. Variable degree dyspnea.
3. Fatigability, asthenia, anorexia, weakness.

Signs

Jugular distension: It depends on the increase of venous pressure as a result of restricted chamber filling. This sign can be absent when there is volume depletion and usually manifests once this is restored.

In some patients, a rise in jugular venous pressure can be observed on inspiration. This abnormal venous pressure behavior is called paradoxical venous pulse or Kussmaul sign, and can be found in up to 20% of cases.

Paradoxical arterial pulse behavior: The arterial pulse normally decreases its amplitude during inspiration as a consequence of the reduction in intrathoracic pressure. This reduction is minimal and passes unnoticed in normal persons, but in patients with constrictive pericarditis and/or pericardial effusion, the reduction is magnified and when it overcomes 10 mmHg it becomes perceptible during palpation. This manifestation can be observed in up to 20% of cases.

Hepatojugular reflux: It is produced as a result of the inability of right heart chambers to adapt to the increase in venous return.

Cardiac sounds: The intensity of the first and second heart sound may be decreased. In 50% of cases, a high frequency sound appears, occurring slightly earlier than the third heart sound. This sound, which depends on the vibrations originated in the thickened and rigid pericardium that tries to be distended during the fast ventricular filling phase, has a dry, high-frequency pitch and is called pericardial knock.

Occasionally, a pericardial rub can be auscultated.

More advanced cases may present with hydrothorax, ascites, generalized edemas, asthenia, anorexia, adynamia, hypotension, jaundice and cachexia. There is frequent hepotomegaly, evidenced in >70% of cases and its presence may erroneously suggest other diagnoses.

External recordings and imaging techniques are necessary to formulate a definitive diagnosis.

Table 1 details the recommendations for different complementary studies.

Table 1. Diagnosis of constrictive pericarditis

Recommendation	Class	Level of evidence
Use of ECG as a complementary method is recommended when constrictive pericarditis is suspected.	I	C
Use of chest X-ray as complementary method is recommended when constrictive pericarditis is suspected.	I	C
Use of cardiac Doppler echocardiography as complementary diagnostic method is recommended when constrictive pericarditis is suspected.	I	B
Use of CAT scan as complementary diagnostic method is recommended when constrictive pericarditis is suspected.	I	B
CAT scan is recommended as complementary diagnostic and prognostic method to guide treatment in patients with diagnosis of constrictive pericarditis.	IIa	B
MRI is recommended as complementary diagnostic method when constrictive pericarditis is suspected and Doppler echocardiography and CT findings are inconclusive.	IIa	B
A hemodynamic (right catheterization) study is recommended as complementary diagnostic method to differentiate constrictive pericarditis from restrictive cardiomyopathy when complementary study results are doubtful.	I	B
A preoperative hemodynamic study is recommended as complementary diagnostic method when there is associated cardiovascular disease.	I	B
Coronary angiography is recommended prior to pericardiectomy in men above 35 years and women above 45 years of age without history of coronary heart disease or associated risk factors.	IIa	B
Myocardial or pericardial biopsy is recommended in highly selected cases to differentiate constrictive pericarditis from constrictive cardiomyopathy.	IIa	B
Pericardiocentesis is recommended in case of constrictive symptoms in the presence of at least moderate pericardial effusion, as a diagnostic and eventually therapeutic method of effusive-constrictive pericarditis.	I	B
Brain natriuretic peptide dosage as complementary diagnostic method is of limited usefulness when constrictive pericarditis is suspected.	IIb	B

ECG: Electrocardiogram. CAT: Computerized axial tomography. MRI: Magnetic resonance imaging

4.6. DIFFERENTIAL DIAGNOSIS

Signs and symptoms characterizing constrictive pericarditis are common in a series of processes, among them:

- Cardiomyopathies coursing with restriction
- Pericardial tamponade
- Edematous-ascites syndromes
- Liver cirrhosis
- Nephrotic syndrome
- Heart failure
- Ovarian cancer

4.7. TREATMENT (Table 2)

In most cases, surgery is the only treatment able to definitively resolve pericardial constriction. However, on some occasions, patients may spontaneously evolve favorably or with medical treatment. The rates of 30-day mortality with surgical treatment range between 4% and 10%, depending essentially on the etiology, the occurrence of comorbidities and the degree of functional repercussion.

Table 2. Treatment of Constrictive Pericarditis

Recommendation	Class	Level of evidence
Discretional use of diuretics is recommended to decrease venous pressure, edemas and relieve symptoms.	I	C
It is recommended to initiate or continue medical treatment (diuretics, antiinflammatory agents, corticosteroids, immunosuppresosrs) for at least 3 months before indicating surgical treatment, provided the clinical condition of the patient allows it, in order to assess and treat transient constrictive pericarditis.	I	B
Medical treatment is recommended in oligosymptomatic patients or with mild symptoms and high surgical risk.	I	C
Surgical treatment with total pericardiectomy is recommended in patients with constrictive pericarditis who remain symptomatic despite medical treatment.	I	B
Complete pericardiectomy (anterior portion between both phrenic nerves and diaphragmatic pericardium) is recommended to reduce the risk of symptom recurrence.	I	B
Analysis and assessment of prognostic risk factors is recommended when indicating surgical treatment, especially in patients with unfavorable prognosis (post-radiation constrictive pericarditis, end-stage chronic renal failure, myocardial dysfunction, end-stage heart failure and end-stage liver disease.	I	C