# PULMONARY INFUNDIBULAR STENOSIS SECONDARY TO A MEDIASTINAL HOGDKIN LYMPHOMA TUMOUR INVASION IN A YOUNG ADULT MALE: What a rare occurrence!


## Abstract

**Introduction:** Cardiac involvement is a rare unusual localization of Hodgkin lymphoma, as it's known that primary lymphomas of the heart are rare. They develop primarily in the right atrium, and extend to the pericardium in about one third of the cases. Pulmonary infundibular stenosis secondary to mediastinal mass Hodgkin lymphoma invasion is rare but not impossible especially in relapse cases.

**Case report**: A rare case of a young adult patient diagnosed with Hodgkin Lymphoma under chemotherapy presents a relapse of his pathology associated with heart failure; predominantly, right heart failure due to extension of the mediastinal mass tumor to the right cavities of the heart causing pulmonary infundibular stenosis which is a rare occurrence. He was put under an intense chemotherapy and symptomatic heart failure treatment with close follow-up after multi-disciplinary discussions including cardiologist, onco-hematologists and cardiothoracic surgeons. Three months later, a significant reduction of the mass was observered.

**Conclusion :** Pulmonary infundibular stenosis secondary to mediastinal Hodgkin lymphoma invasion is a rare entity of secondary tumors of the heart. Multimodal imagery is the key tool for diagnosis. There prognosis depend on multiple factors but generally good in younger adult patients compared to aged individuals.



**Keywords:** Intracardiac mass, infundibulum pulmonary stenosis, Hodgkin Lymphoma, CT scan, Transthoracic echocardiography**.**


## Introduction

Hodgkin's lymphoma is defined by a malignant proliferation of Reed-Sternberg or Hodgkin cells that are clonally related B-cell-derived malignant cells. This disease is characterized by a good outcome (cure rate more than 80%) [1,2]. Initial thoracic involvement is usual and the more frequent localization is the mediastinum, following by the lung parenchyma and the pleura [3]. Cardiac involvement is a rare in Hodgkin lymphoma [4], as it's known that primary lymphomas of the heart are rare. They are linked to extra-nodal lymphomas since the heart does not normally contain lymphoid tissue. They develop primarily in the right atrium, and extend to the pericardium in about one third case [5]. The most common histological types are diffuse large B lymphoma cells, follicular B lymphoma, and Burkitt lymphoma [6,7]. We hereby report an extremely rare case of a pulmonary infundibular stenosis secondary to a relapse Hodgkin lymphoma responsible for right heart failure in a young male adult patient who interrupted his initial chemotherapy treatment without a justified medical reason. Patient's chemotherapy treatment was reinitiated with a net clinical outcome and

regression of the intra cardiac mass.

## Case Report

A 27year old young adult male patient with no cardiovascular disease history was followed up at the hematology cancer centre since 2021 for Hodgkin lymphoma under chemotherapy with a good evolution. Later, in 2023, patient was concomittenly diagnosed with pulmonary tuberculosis treated by therapy protocol (RHZE quadritherapy for 2 months and RH bitherapy for 6 months), declared cured. During this period of tuberculosis treatment, patient was lost of sight which interrupted his chemotherapy treatment for almost 8 consecutive months.

The young patient was currently re-admitted initially for a large mediastinal mass invading the right heart side obstructing the right ventricular outflow confirming lymphoma relapses.

Clinical exam findings showed a conscious patient, cachectic with a weight lost of 10kg three months prior to hospitalization. He presented a stage III dyspnea according to the NYHA scale, with edema of the right upper limb and asymetric edema of the lower limbs rising up to the ankles. He was afebrile at 37.5°C, blood pressure (BP) of 140/90mmHg, heart rate of 124 beats per minute (bpm) and a saturation level in open air of 97%. His supraclavicular and right axillary ganglions were palpated with the presence of filling supraclavicular hollows responsible for superior cava syndrome. The later caused cyanosis of the upper cava territory during efforts like coughing or exercise. Heart auscultation found heart systolic murmurs at the pulmonary focus.

The ECG noted a sinus tachycardia at 124 bpm, with an incomplete right bundle branch bloc (RBB) without secondary repolarization disorders (Fig 1).

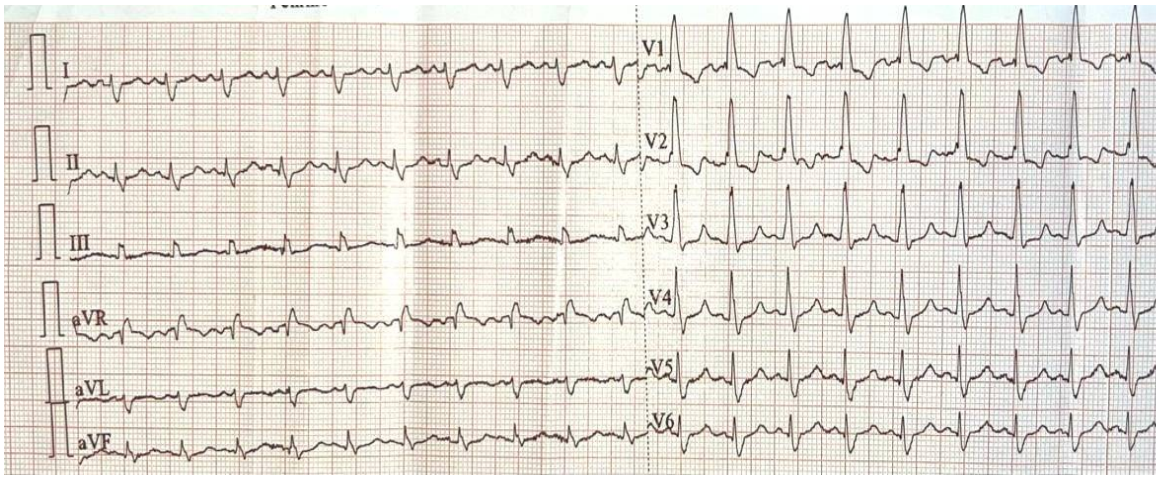



**Figure 1** : 12 leads electrocardiogram (ECG) : Sinus Tachycardia associated with an incomplete right bundle branch bloc (RBB).

A transthoracic echocardiography was performed to explore the involvement of the heart cavities ; it showed a dilated right heart side cavity (Basal diameter of the right ventricle : 53mm) without hypertrophy of the walls of the right ventricule (RV) and longitudinal systolic dysfunction of the RV (S'VD :6m/s TAPSE :12mm) [Figure 2].

**B**

**A**

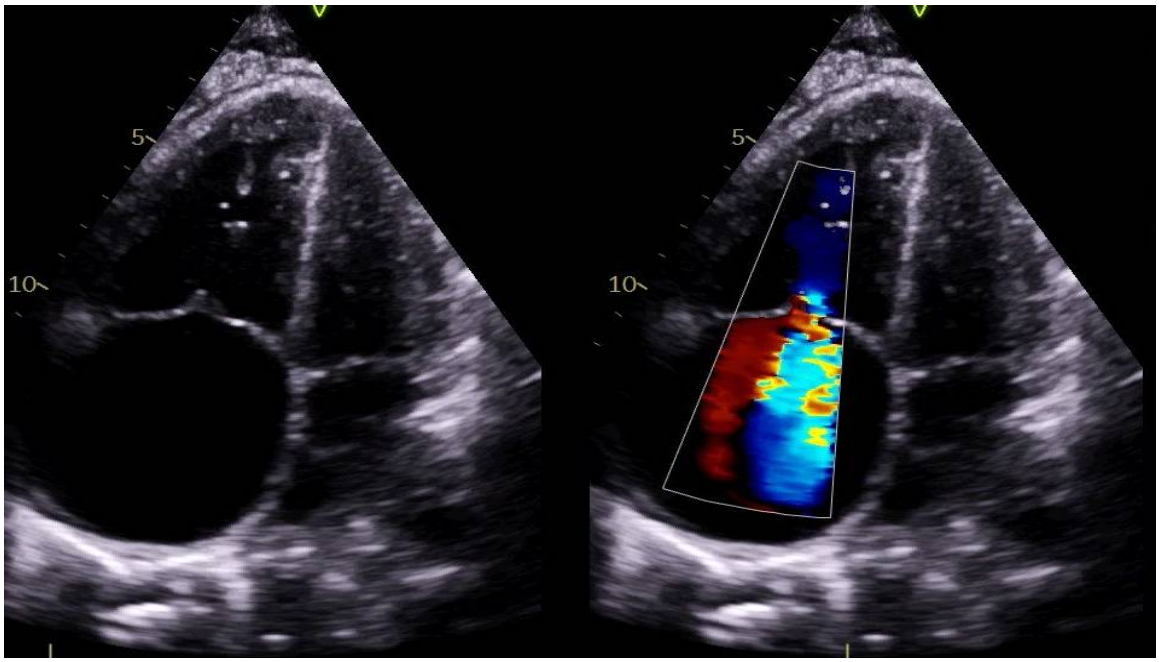



**Figure 2** : Transthoracic echocardiography (TTE) ; Apical 4-chambers view : **A**-showing a dilated right heart side cavity (Basal diameter of the right ventricle : 53mm) without hypertrophy of the walls of the right ventricule (RV) and longitudinal systolic dysfunction of the RV. **B**- significant tricuspid regurgitation.

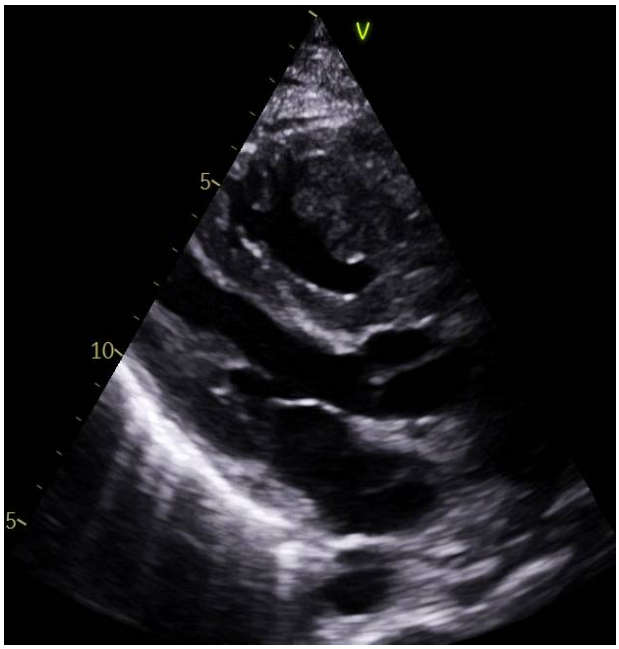
We noted the presence of an extracardiac mass invading the RV at the level of the proximal and distal pulmonary infundibulum site extending to ejection way of the RV and trunk of the pulmonary artery (PA) causing a pulmonary stenosis with a maximum gradient of 115mmHg, significant tricuspid regurgitation estimating the RV systolic pressure at 98mmHg (Figure 3 and 4).

PA

Intra-cardiac mass

B

A



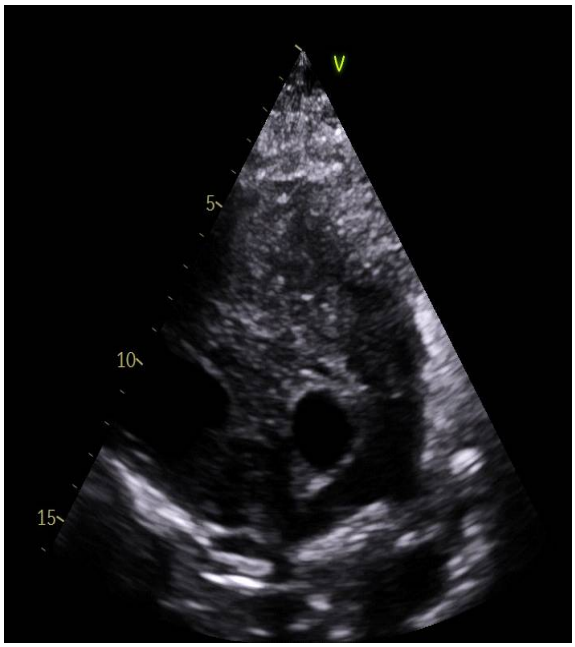




**Figure 3 :** Transthoracic echocardiography : **A**- Parasternal long axis view showing an intracardiac mass occupying the proximal and distal pulmonary infundibulum RV compressing the left ventricle **B**- Parasternal short axis view showing an extension extracardiac portion of the mass with obstruction of the pulmonary trunk.

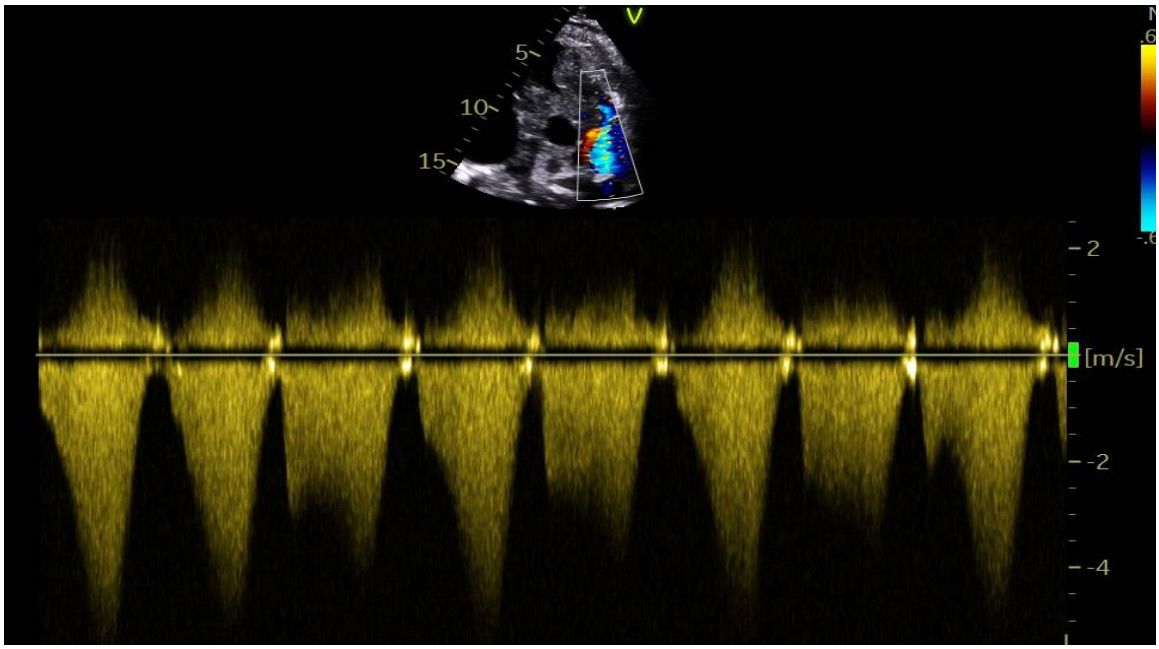



**Figure 4 :** Transthoracic echocardiography :C-pulse doppler wave affirming a pulmonary stenosis with a maximum gradient of 115mmHg and Vmax 4m/s

The pulmonary artery branches were free without a visible thrombus. A dilated non compliance inferior vena cava with (Fig 5) no pericardial effusion but a bilateral pleural effusion more marked on the left side was noted in the subcoastal and 4 chambers apical windows view.

**B**

**A**

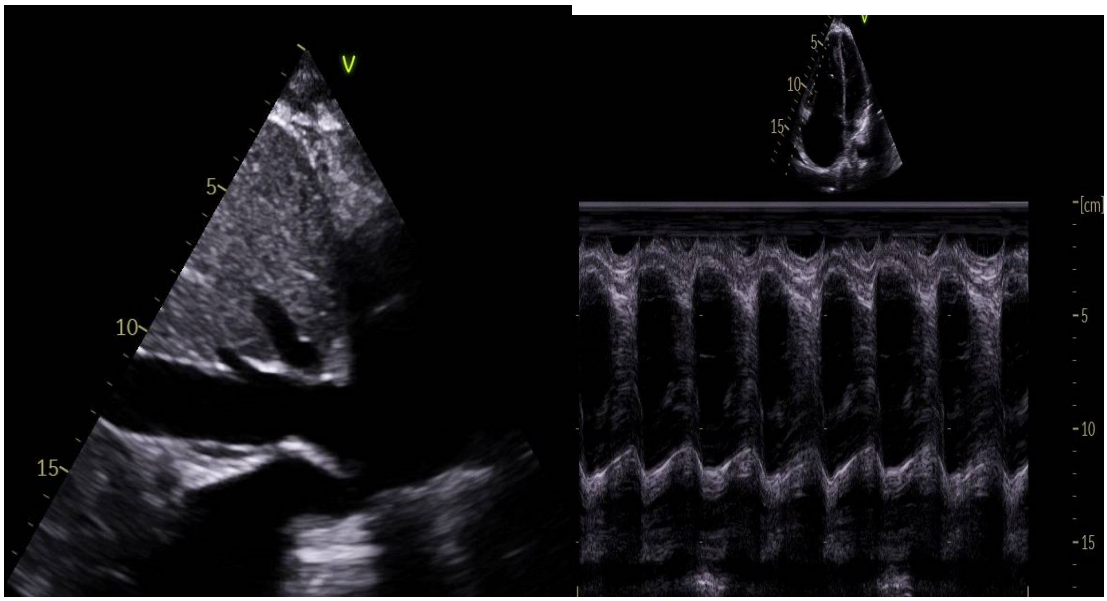



**Figure 5** : TTE ; **A**- dilated inferior vena cava at 22mm and **B**- longitudinal systloic dysfunction with a tricuspid annular plan exercusion value of 12mm on time mode (TM).

Furthermore, the left ventricule was of normal size and function with an ejection fraction estimated at 55%, compressed by the dilated RV. There was no significant changes of the left ejection fraction values since his last control. No valve disease or other congenital heat disease were spotted.

A full body computed tomography scan (body CT Scan) was performed ; it showed a locally advanced mediastinal lesion process with supra and infra diaphragmatic lymph node involvement, as well as broncho-pulmonary, cardiac and vascular lymph nodes (pulmonary trunk and superior vena cava invaded +++) ; possibly related to a lymphoma relapse with pericardial effusion and moderate bilateral pleural effusion (Fig 6).

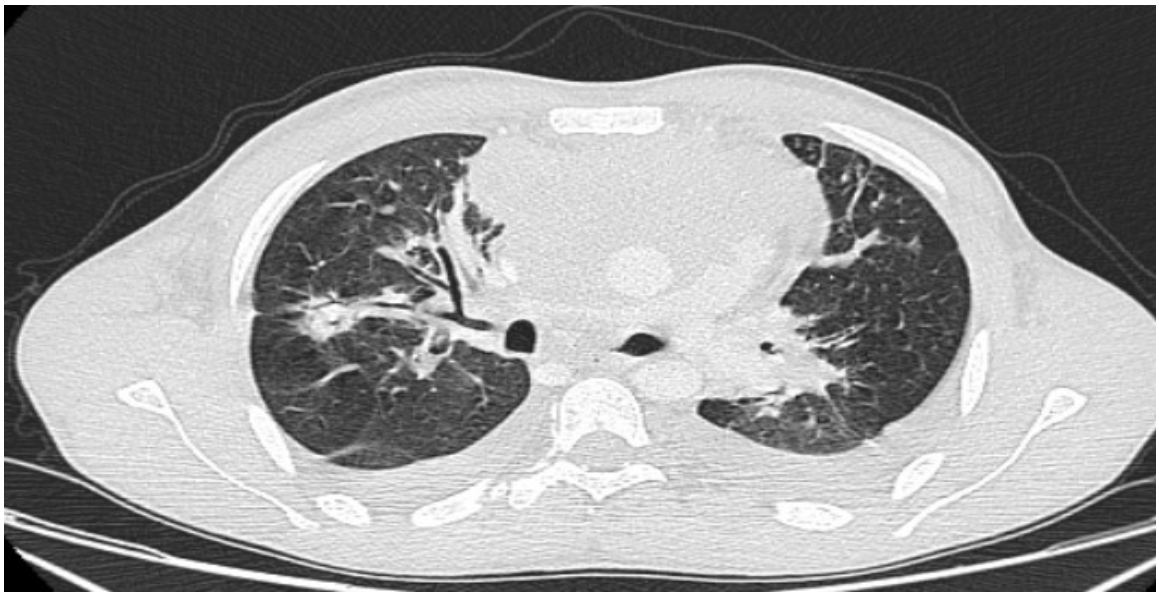


**Figure 6:** CT scan at the thoracic level without injection: showing a locally advanced mediastinal lesion process with lymph node invasions (red arrows), enlarged heart with pericardial effusion evoking a relapse Hodgkin Lymphoma.

Blood workout findings showed a mild anemia with a hemoglobin level of 11.9g/dl, white blood cells count at 6550/$mm^3$ and normal platelets level at 255000/$mm^3$ . Blood electrolytes (Na+, K+, creatinine, albumin, liver enzymes such as ALAT and ASAT) were normal except a slight increase in C-reative protein of 59ng/l.

Patient was given symptomatic treatment based on loop duiretics (furosemide 40mg tablet x 3 a day with potassium supplmentation) for 48hrs then reduction of the dose to 4àmg daily with a minerocorticoide (Spironolactone 50mg, half tablet a day). The clinical evolution was marked with the regression and dispiration of the limb edema but persistent of that of the upper limbs which was secondary to the mediastinal mass. A decision was taking after pluridisplinary work out with the hematology, oncology and cardiology team to re-initiate chemotherapy with regular fellow-up.

We prescribed a low salt diet intake, optimal fluid intake to avoid congestion. Patient was placed under chemotherapy protocol R-DHAOX or DHAC or DHAP which include Rituximab 375mg/$m^2$, Dexamethasone 40mg, A racytine 2g/$m^2$ (twice daily), oxalipatine 130mg/$m^2$. The first week of chemotherapy was without incidence. Patient transthoracic echocardiogaphy follow-up showed reduced intrac cardiac mass three months prior to intense chemotherapy.

**Discussion**

Hodgkin's disease was first described by Thomas Hodgkin in 1832. It was recognized as a

non-infectious pathology in 1940 [8]. This entity is characterized by the presence of Reed-Sternberg cells, implemented evidence in 1898 by Sternberg and 1902 by Reed, associated with an extensive immune response (T lymphocyte CD4+ Th 2), within tumor proliferation. Molecular studies carried out on isolated Reed-Sternberg tumor cells by microdissection made it possible to designate the origin B centro-follicular clonal [2, 9].

The epidemiology of Hodgkin lymphoma (HL) is very unique. There is a very variable incidence in function of the geographical areas studied, which implies an influence of lifestyles and socio-economic development. In France, the incidence is 3.2 per 100,000 inhabitants, according to data from 2018 register [10]. Cardiac lymphomas are very rare ; the Institute of Pathology of the Armed Forces of the United States of America reported 38 cases between 1945 and 1994 [11]. Other studies collected a total of 56 cases [12]. From a macroscopic point of view, these are polylobed masses yellowish in color, infiltrating the heart walls. Microscopic examination reveals proliferation B lymphocytes [12]. Currently
The frequency of lymphoma is increasing due to more large proportion of immunocompromised patients (AIDS). On the other hand, in transplants patients, and particularly in heart and lung transplant recipients, we can observe cardiac lymphomas in significantly higher incidence than in kidney and liver transplant recipients [13]. Primitive cardiac lymphomas are rare but cardiac infiltration of extracardiac lymphomas is described in 30% of cases in the literature [14]. Extracardiac lymphomas infiltrating the right heart side responsable for pulmonary infundibular stenosis are extremely rare compared to the forms in the literature[4,5], making our case unique.
The clinical presentation of cardiac lymphomas are variable, but considering the fact that the right heart side being oftenly infiltrated, patients can be totally asymptomatic or present symptoms like : dyspnea, orthopnea, palpation, chest pains or syncopes due to systemic embolism. [5,14]. In our case, patient presented signs and symptoms of predominant right heart side failure with a systolic dysfuction of the right.

Transthoracic echocardiography (TTE) is considered the first-line imaging modality for the assessment of cardiac masses due to its portability, lack of radiation, and wide availability. It remains the most often used modality to characterize masses and their hemodynamic consequences, as many masses are found incidentally with echocardiography; sensitivity 55–93% [16]. Transesophageal echocardiography (TEE) is used to complement TTE with
improved detection of cardiac masses compared to TTE alone, particularly in the evaluation of posterior structures and small lesions [16]. Cardiac CT scan and cardiac MRI have more specific indications concerning cardiac tumor pathologies ; size, localization, tissue characterization and their extension [17]. In our case TTE and thoracic CT scan was performed as TTE was precise in diagnostic of the mass extension to the right heart side ejection track while the CT scan studied the anatomic relationship of the mediastinal lymphoma with adjacent cardiac structures. 18-FDG PET (18-fluorodesoxyglucose positron emision tomography) has become the examination of choice in staging and therapeutic evaluation, and constitutes a decision-making examination in personalized

PET-guided treatment strategies [10].

Cardiac lymphoma requires treatment medical with the combination of doxorubicin, cyclophosphamide, vincristine and prednisolone [5]. The most accessible malignant tumors'
surgical resection are those located in the right or left atrium. Some techniques like auto transplantation has been described [18, 19]. Since in our case the patient presented relapse situation of his mediastinal lymphoma; an intensify chemotherapy was opted similar to codified treatment of relapse lymphomas [5]. Surgery was dangerous in our patient who had a systolic right ventricular dysfunction whilst chemotherapy sections were not completed. A close follow up was carried out in order to evaluate chemotherapy efficacy.

As for secondary tumors of the heart, they represent often a terminal stage of the disease. The surgical treatment aims to treat obstruction of large vessels and the heart cavity by the tumor in young patients [4]. However, Hodgkin lymphoma is a curable cancer in approximately 90% of patients. Successive protocols have been developed to maintain this high level of curability, while reducing the risks of long-term toxicity of the treatments. In relapse situations, treatments may include immunotherapy [5]. Its prognosis is favorable in young subjects compared to adults more than 60 years of age [20]. Our patient is followed up regularly and evaluated with multimodal imagery.

## Conclusion

Pulmonary infundibular stenosis secondary to mediastinal Hodgkin lymphoma invasion is a rare entity of secondary tumors of the heart. Multimodal imagery is the key tool for diagnosis and exploration of tumor extension with adjacent anatomical organs. Secondary localization of heart lymphomas are frequent compared to primary heart lymphomas. Their prognosis depends on multiple factors but generally good in younger adult patients compared to older patients.

## Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

## Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

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