

Ask the Clinical Instructor

A Q&A column for those new to the cath lab

Questions are answered by:
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Amyloidosis (am'i - loy- doh'sis) is a disease characterized by extracellular accumulation of amyloid in various organs of the body.¹ The cause of this disease is the deposit of amyloid, a type of protein, that when abnormally present in the body, can attach itself within the tissues and organs of the body. If these deposits replace or surround muscle sections, it can make it hard for that muscle or structure to work properly.

This is a rather rare disease that is sometimes also called an infiltrative disease. There are many types of amyloidosis and cardiac amyloidosis is one of them. Cardiac amyloidosis is more common in men than in women and the disease is rare in people under age 40.

This disease can also be known as the "stiff heart" syndrome or secondary cardiac amyloidosis. The primary concern is that when these deposits take the place of normal heart tissue, the heart becomes "stiff." It is the most typical type of restrictive cardiomyopathy. Other types of restrictive/infiltrative diseases are sarcoidosis (a granuloma disease), hemochromatosis (high deposits of iron in the muscles), fibrosis from radiation, and various tumors and infiltrations of the heart.

The cardiac variation of this disease can present with 4 specific syndromes: restrictive cardiomyopathy, systolic heart failure (cardiomyopathy), orthostatic hypotension (very poor prognosis) and a conduction pathway disease.

The patient who comes to the lab can have many complaints, but some common ones usually occur, which can include palpitations, swelling of the legs and ankles, a history of

excessive urination at night, fatigue, shortness of breath at night or trouble breathing while lying down (orthopnea). Because of the effects of the amyloids on the heart muscle, these patients can have congestive heart failure due to the cardiomyopathy.

Unfortunately, the only definitive diagnosis of cardiac amyloidosis is a cardiac biopsy. If you don't perform those procedures in your lab right now, chances are you never will, since it is usually reserved for transplant and research facilities. With a cardiac biopsy, a special catheter is placed within the heart chambers, and a "bite" is taken out of the tissue (Figure 1). This tissue is then sent to the pathology lab for analysis. Without the use of a biopsy, diagnosis is made by history, signs and symptoms, and by ruling out other diseases. Sometimes a measurement of circulating serum proteins can be helpful in the confirmation of the disease³ or proteins in the urine can provide clues to the cardiologist.

Now you know that these patients suffer from cardiomyopathy because their heart muscle is not allowed to function properly. When these patients receive a cardiac catheterization, we might see some specific things. If the heart muscle is "stiff," we will see an elevated left ventricular end-diastolic (ED) pressure. If the myocardium is diffusely fibrotic or infiltrated with the amyloid, a very high end-diastolic stretching force may be required to produce even a normal end-diastolic sarcomere length.² Remember that sarcomeres are the components in the heart muscle that contract and relax. There may need to be more pressure placed on them to relax in order to allow the heart to fill with blood and also

"Recently, we had a patient in the lab for a cath, and the doctor stated that he suspected the cause of the heart failure to be amyloidosis. What? I've never heard of that."

— Email from anonymous to tginapp@rcisreview.com



Figure 1. The catheter will be inserted into a vein or an artery, under flouroscopy, depending upon from where the sample needs to be obtained. A procedural note: assure that the jaws, or bioptomes, are CLOSED during insertion. Once the sample is obtained, the bioptomes should also remain closed during catheter removal. Image courtesy of Cook Medical.

get a proper contraction to expel the blood from the ventricle. This "forced filling" pressure of the ventricle results in higher end-diastolic pressures.

On the left ventriculogram, you will likely see the classic cardiomyopathy silhouette (large left ventricle, poor ejection fraction), as well as a large atrium. The large atrium is also part of the cardiomyopathy process due to the inability to get all the necessary flow into the left ventricle. This leaves the atrium "holding the ball." Over a period of time, the atrium will expand/enlarge to accommodate this flow imbalance.

With today's echocardiogram technology, there is seldom a need to perform a right heart catheterization (RHC) on these patients. If a RHC were performed on the symptomatic patients, there would likely be the classic 'dip and plateau' square root sign. This is indicative of the impairment of chamber filling due to the restrictive pathology (Figure 2).

There may also be times where these patients come in with what appears to be an acute coronary syndrome (chest pain, dyspnea, electrocardiogram changes, etc.), but after angiography, the coronary arteries appear normal. If the amyloids are deposited in the intramural (in the muscle wall) arteries, particularly in the media and adventitia layers, there can be impairments in coronary perfusion.³ This lack of perfusion can create

chest pain due to the lack of proper oxygenation to the muscle. However, the main arteries will appear 'normal' during angiograms.

These patients have a poor prognosis after symptoms appear and once they are diagnosed. Cardiac involvement generally denotes a high mortality rate, regardless of the method of treatment. The median survival rate from the onset of congestive heart failure is only 6 months.⁴ Syncope indicates a poor prognosis as well, and is often a precursor of sudden cardiac death.⁵ In the presentation of cardiomyopathy, these patients may only have a life expectancy of a few years. Heart transplant is generally not an option because amyloidosis will also affect other organs at the same time, excluding the individual from being a transplant patient.

Treatments are generally based upon managing the symptoms without interfering with cardiac output. During left ventricular analysis, a left ventricular ED pressure of 25 or more is not uncommon. However, if the increase in ED pressure is treated, a reduction in the sarcomere stretch can occur, and this will result in an equal decrease in cardiac output. Pacemakers and ICDs may be used in cases where cardiac amyloidosis is thought to be in early stages.

A recent case study we are familiar with involved a 49-year-old male who

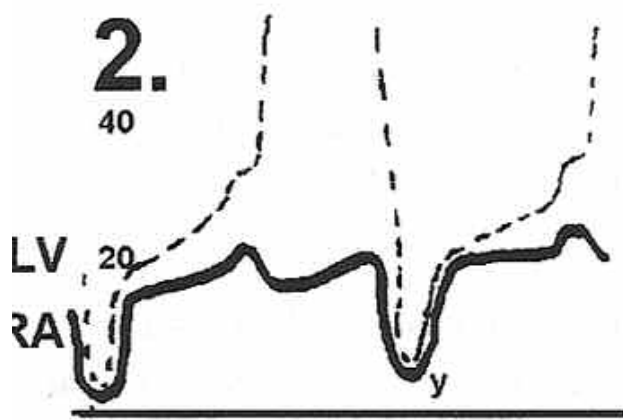


Figure 2. Reprinted with permission from Wes Todd's *Cardiovascular Self-Assessment*.

This waveform is indicative of a restrictive pathology. There are elevated venous pressures, but the left ventricle (LV) and right ventricle (RV) are not equal, as they would be in constrictive diseases. Here, a "stiff" LV fills rapidly in diastole, then the pressure plateaus. This square root sign $\sqrt{\quad}$ pattern is also described as a "dip and plateau pattern."

presented with chest tightness upon exertion, progressive dyspnea and orthopnea. Echo indicated an ejection fraction of approximately 30%, compared to normal left ventricular function a year earlier. Bi-atrial enlargement was also present and there was a positive history of early aged sudden cardiac death in the family. Left heart cardiac catheterization was performed, which yielded normal coronary arteries. The left ventriculogram revealed obvious global myopathy with regurgitation into a very large atrium. (Angiograms can be seen at <http://www.rcisreview.com/AskTheInstructor/March2008.htm>). With this history and normal coronaries, an infiltrative disease must be ruled out. At the time of this article, a definitive diagnosis is not available.

When you have a 'negative' case after the patient had presented with acute coronary signs and symptoms, but cardiomyopathy is present, infiltrative disorders, such as amyloidosis, should be considered as a possible pathology.

Next month, we will answer a question about procedures to evaluate aortic and mitral valves. ■

References

1. *Stedman's Medical Dictionary for the Health Professions and Nursing, 5th edition.* Philadelphia: Lippincott Williams & Wilkins, 2004.
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3. Libby P, Bonow RO, Mann DL, Zipes DP. *Braunwald's Heart Disease, 8th edition.* St. Louis: Saunders, An Imprint of Elsevier, 2007.
4. Grogan M, Gertz MA, Kyle RA, et al. Five or more years of survival in patients with primary systemic amyloidosis and biopsy-proven cardiac involvement. *Am J Cardiol* 2000; 85:664-665, A11.
5. Chamarthi B, Dubrey SW, Cha K, et al. Features and prognosis of exertional syncope in light-chain associated AL cardiac amyloidosis. *Am J Cardiol* 1997;80:1242-1245.

"The only foolish question is one left unasked."

New Staff: Your suggested topics and questions are needed!

(You are welcome to remain anonymous.)

Email us at: tginapp@rcisreview.com

What Do You Think?

Multiple new and ongoing questions from readers. Your responses are welcome!

Answer or pose a question at cathlabdigest@aol.com.

New Questions

Cross-Training Duties

In a cross-trained lab, do RTs and CVTs document equipment only and the RN separately handles the moderate sedation and patient care documentation?

Thank you,
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Staffing with Case Fluctuation

I am doing some research into how to handle staffing when the census/# of cases fluctuates in the cath lab. Any suggestions about how to keep employees' schedules relatively stable when the case load bounces around so much?

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Manual Pressure

I work in the cath lab. My main job is to hold manual pressure on all post cath patients. I am trying to find out how many cath labs use manual pressure compared to closure devices. Do any other cath labs have one person that is responsible for holding pressure? Thank you!

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Cross Training with Recovery Unit

Due to some staffing issues in our cath lab and recovery units, our director of nursing wants cross-training between the areas. We are a 2-room cath lab that also does electrophysiology studies as well as pacemaker and ICD implants. In the lab, we have 3 full-time (FT) RNs, 1 part-time (PT) RN, 3 FT RTs and 1 PT RT. The 12-bed recovery area has 5 FT RNs and 3 PT RNs. Does anyone cross-train with their recovery area?

Our cath lab RNs do cover recovery for lunches and breaks. I am concerned about the constant changes within the lab and keeping the recovery staff up to date. Thank you!

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Normal Caths & Standard Ambulation Times

How does your institution handle the following items?

1) Do you track normal cardiac cath numbers? If so, how? If yes, what is the criteria in place that defines a normal cardiac cath (i.e., no blockages greater than 15% in any major artery greater than 2mm diameter)?

2) What are the standard ambulation times after a diagnostic cath using 5 or 6 Fr sheaths, when the patient has not received heparin. How long do you keep patients on bedrest after hemostasis is achieved? (Manual holds only, no devices used for closure.) For those responding to this question, do you know of any studies or articles/research that supports this ambulation time?

Thank you!
Annie Ruppert
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Data on Pre/Post Beds per CCL

I am looking for data or research that supports the number of pre/post beds per cath lab. Are there any guidelines on recommendations ratios?

Thanks for your help.
Kind regards,
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