PATIENT EDUCATION IN

LEFT VENTRICULAR NONCOMPACTION CARDIOMYOPATHY

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Tina Aldrich

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Abstract

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Tina Aldrich

Under the Supervision of Patricia L. Bromley, Ph.D.

This paper sets forth the information needed to educate families about the genetic link of left ventricular noncompaction cardiomyopathy. The current generation will have an opportunity to take advantage of the latest technologies and medications. This paper describes the importance of echocardiogram testing of the asymptomatic patient and defines the medical terminology so the average family member can actually understand the contents. The following parts are be covered: clinical findings, physical symptoms, actions and possible treatment plans and hereditary responsibility. An appendix is included with a patient advisory question and answer pamphlet. Forms have been created to assist with patient tracking of cardiology appointments, medications, physicians, and family cardiac health history. Also included are links to additional articles to read as well as support groups to join.

This paper examines this disease and explains what steps can be done to help keep the heart from unnecessary stress and trauma. Through this increased awareness, early detection of progression and the prevention of complications, there will be a positive change in the management of left ventricular noncompaction cardiomyopathy.

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Chapter One: Introduction

Many coaches, parents and the general population have heard stories of "healthy" teenage athletes dropping dead while competing in sporting events, or simply completing a workout with their teams. This type of sudden death, while in action or at rest, can happen to children and adults alike and is often caused by cardiomyopathy. In many of these sad cases, the deceased had no known heart problems and yet succumbed to a tachyarrhythmia which resulted in sudden cardiac death. This is a condition that is prevalent in the rare disease of left ventricular noncompaction (LVNC) cardiomyopathy (Suvarna, Deshmukh, & Hajela, 2009).

Some other people are actually diagnosed with a cardiomyopathy because of poor health conditions. On July 30, 1998, Ronald A. Wyss was diagnosed with idiopathic cardiomyopathy. This diagnosis meant that the heart of this 32 year old patient was enlarged, weakened and not functioning properly, but there was no known reason for this heart condition. The patient's father, Robert H. Wyss, had died suddenly at the age of 46, but was thought to have died from a blood clot stemming from a recent neck surgery. The patient, Ronald A. Wyss, was given drug therapy to help control the symptoms and decrease the work load placed on the heart. The drugs that were prescribed were: blood thinners to control the possibility of blood clotting in the heart from stasis of the blood, beta blockers to slow the heart rate down so it did not work so hard and angiotensin-converting-enzyme inhibitors to reduce the workload of the heart. The heart had an extremely low ejection fraction and some arrhythmias were noted. After further testing, the patient was reclassified as having idiopathic dilated cardiomyopathy. The patient was given an option for a defibrillator to be placed, but refused this. In 1998 this patient was given a five to ten year life expectancy, but he suddenly died from an arrhythmia on October 16, 2004. This patient was only 38 years old.

The patient left behind two biological children, Brianna N. Wyss and Robert D. Wyss, ages seven and eight. The possibility of a genetic link to this cardiomyopathy needed to be addressed to benefit the living relatives. The autopsies and medical records of Ronald A. Wyss and Robert H. Wyss, along with the medical records of the paternal grandparents of the patient, Henry Wyss Jr. and Josephine B. Wyss, were obtained and researched. This research was then brought to a cardiologist who aided in determining the problematic heart conditions that influenced the deaths of the patient's father and both paternal grandparents. The first degree relatives were notified and the biological children started cardiac testing. The tests for the children included EKG's, Holter monitors, echocardiograms and treadmill stress tests which were administered by a pediatric cardiologist. These tests were performed every two years and all test results were normal.

The patient's only sibling, sister Carolyn Wyss Peters, was expecting a baby in July of 2005. During her childbirth she experienced heart problems and was diagnosed as having postpartum cardiomyopathy. This cardiomyopathy did not go away months after childbirth and in 2009, at the age of 40, was finally classified correctly as left ventricular noncompaction cardiomyopathy, which is a genetic cardiomyopathy. Ronald A. Wyss' autopsy and medical records were researched again and the diagnosis changed from idiopathic dilated cardiomyopathy to left ventricular noncompaction cardiomyopathy. In 2011, the patient's children went to see this new cardiologist and were both classified as having left ventricular noncompaction morphology through echocardiogram testing. The only three cousins in the Wyss family were also urged to check with this same doctor to verify heart health. In 2011, two of the cousins were diagnosed as heart healthy, while the third, Daniel R. Sterk, was diagnosed

with left ventricular noncompaction cardiomyopathy at the age of 40. This patient had experienced no prior physical symptoms.

Left ventricular noncompaction cardiomyopathy is a rare genetic heart disease that actually begins in the womb. This means that all of these patients, including the "healthy" athletes, were actually born with this disease. It is important to understand family health histories and be proactive in testing through echocardiograms and in verifying heart health with more than one cardiologist. The earlier this condition is found, the more tools can be used to alleviate some of the strain that is placed on the heart. In today's world of medical imaging and advances, all of these patients could have had an opportunity to be diagnosed while young and asymptomatic. The idea is for the patient to live a healthy lifestyle and then to treat a patient immediately if they do indeed become symptomatic. Early diagnosis would have taken off years of undue stress placed on the heart of these patients and made the prognosis for the disease better.

Left ventricular noncompaction cardiomyopathy, also called spongiform cardiomyopathy, is a rare congenital cardiomyopathy that affects both children and adults. It results from the failure of myocardial development during embryogenesis (Suvarna et al., 2009). This is a condition that develops in the womb, affects the left ventricle and can manifest itself as an enlarged and weakened heart. Because the left ventricle is affected, the common name for this disease is left ventricular noncompaction cardiomyopathy (Paterick, Gerber, Pradhan, Lindor & Tajik, 2010). It is possible to be diagnosed with this condition at any age through the use of an echocardiogram, yet not to ever suffer from any of the symptoms associated with heart disease. Likewise it is possible to suffer from severe heart failure, which even though the condition is present from birth, may only manifest itself later in life.

Most people are diagnosed with LVNC cardiomyopathy because they are already symptomatic. The physical symptoms may include breathlessness, fatigue, swelling of the ankles, racing heartbeat, limited physical capacity and exercise intolerance. The clinical findings may include depression of systolic and diastolic function, poor pumping performance of the heart and an increased ratio of noncompacted wall to compacted wall of both heart ventricles. Once the patients are diagnosed, the management of LVNC includes treatment of heart failure, arrhythmia, and thromboembolism. Some patients may need an implanted internal defibrillator, and others may need cardiac transplantation (Hurst, 2002). There is also the possibility of prescriptions or restrictions on physical activity (Paterick et al., 2010).

Left ventricular noncompaction cardiomyopathy was given its most recent name in 1990 and the World Health Organization has it listed as an unclassified cardiomyopathy. It was classified as a genetic cardiomyopathy by the American Heart Association in 2006 (Paterick, Umland, Jan, Ammar, Kramer, Khandheia, Seward & Tajik, 2012). Due to LVNC cardiomyopathy being a relatively new disease, its impact on life expectancy is not very well understood (Espinola-Zavaleta, Soto, Castellanos, Játiva-Chávez & Keirns, 2007). Most of the clinical studies on this disease deal with patients who are symptomatic and so are suffering from more severe forms of LVNC cardiomyopathy than might be found typically in the population.

Because LVNC cardiomyopathy is a genetic disease, immediate family members are being tested as a precaution. This testing is turning up more supposedly healthy people with LVNC morphology/cardiomyopathy who are asymptomatic. The long-term prognosis for these people is currently unknown. The early detection of asymptomatic LVNC cardiomyopathy places families in charge of being honest with and interested in their health histories and

supportive of echocardiogram testing. A critical part of patient education is to have regular echocardiograms to monitor for the first signs of any clinical findings before physical symptoms are noticed. This would allow the patient to be treated and monitored before the physical changes cause undue harm to the heart itself.

This research paper focuses on early detection for LVNC cardiomyopathy and the education that can be provided to families and patients so diagnosed. Increasing the number of asymptomatic patients who can be monitored will result in better life expectancy and general wellness for those with the disease of LVNC cardiomyopathy.

Statement of the Problem

The purpose of this project was to prepare materials which will be useful in educating and treating LVNC cardiomyopathy patients.

Definition of Terms

- Trabecula is a fibrous strand of connective tissue that supports the heart muscle in place. The characteristic appearance of numerous, excessively prominent trabeculae and deep intertrabecular recesses observed in one or more ventricular wall segment is characteristic in left ventricular noncompaction cardiomyopathy (Suvarna et al., 2009)
- Left Ventricular Noncompaction (LVNC) Cardiomyopathy. During heart development, the majority of the heart muscle is a sponge-like meshwork of interwoven myocardial fibers. As normal development progresses, these trabeculated structures undergo significant compaction that transforms them from spongy to solid. Noncompaction cardiomyopathy results when there is failure of compaction during this process. This disease is characterized anatomically by deep trabeculations in the ventricular wall, which define recesses communicating with the main ventricular chamber. Major clinical

correlates include systolic and diastolic dysfunction, associated at times with systemic embolic events (Espinola-Zavaleta et al., 2007).

- Stasis is the state in which the normal flow of a body liquid stops, for example the flow of blood through vessels (Taber's Medical Dictionary Online, 2000-2012).
- Morphology refers to the form and structure of the internal parts like organs and bones (Taber's Medical Dictionary Online, 2000-2012).
- Tachyarrhythmia is any cardiac rhythm disturbance in which the heart rate exceeds 100 beats per minute (Taber's Medical Dictionary Online, 2000-2012).
- Myocardial refers to the myocardium, the middle of the three layers forming the wall of the heart (Taber's Medical Dictionary Online, 2000-2012).
- Embryogenesis is the process by which the embryo is formed and develops, until it develops into a fetus (Taber's Medical Dictionary Online, 2000-2012).
- Echocardiogram is a visual record made by echocardiography which is the use of ultrasound to examine and measure the structure and functioning of the heart and to diagnose abnormalities and disease (Merriam-Webster, Incorporated, 2012)
- Thromboembolism is the blocking of a blood vessel by a particle that has broken away from a blood clot at its site of formation (Suvarna et al., 2009)

Delimitations of Research

Left ventricular noncompaction research is limited because of the controversy over whether it is an actual disease entity. There is extensive prognosis overlap with dilated cardiomyopathy, apical hypertrophy, and hypertrophic cardiomyopathy (Paterick et al., 2010). It takes an extremely knowledgeable cardiologist to know exactly what to look for in the echocardiogram readings to diagnose correctly. This paper covers the importance of

echocardiogram diagnosis as well as addressing possible clinical findings and physical symptoms. It focuses on working together with physicians and families to have an accurate treatment plan. This paper does not address the technical results of echocardiograms or the possible complications in some of the treatment plans.

Method of Approach

An extensive review of the disease LVNC cardiomyopathy was conducted. A review of literature relating to research studies, illustrations of echocardiograms, postmortem examinations, magnetic resonance imaging, video clips, and actual cases of symptomatic and asymptomatic noncompaction patients was studied. Actual echocardiograms performed on two patients were witnessed and e-mail and personal conversations with doctors specializing in this type of cardiomyopathy were conducted. The findings were summarized and recommendations made.

Chapter Two: Review of Related Literature

Echocardiogram Diagnosis of Asymptomatic Patients

Left ventricular noncompaction cardiomyopathy has distinct structures and features that present themselves during the end stage of the myocardial growth process in the womb. During the embryonic growth between five and eight weeks, intertrabecular recesses form such that they are in communication with the left ventricular endocardium. As normal cardiac growth and development progresses, myocardial compaction occurs and the intertrabecular recesses form into capillary beds. Between 12 and 18 weeks in the womb, the process of compaction of the trabecular supporting connective tissue occurs. This compaction adds to the proportion and thickness of the compact myocardium (Paterick et al., 2012).

It is hypothesized that the early ending of the compaction of the trabecular meshwork gives rise to LVNC in utero. The result is a spongy meshwork of bilayered myocardium that has prominent trabeculae and deep intertrabecular recesses that communicate with the left ventricular cavity less effectively than a compacted myocardial would. The gestational age at which the myocardium ceases to compact may determine the severity and extent of the LVNC (Paterick et al., 2012).

These distinct morphological features lend well to echocardiography for the diagnostic tool of choice. The echocardiogram reveals the features of the bilayered myocardium, the prominent ventricular trabeculations and the deep intertrabecular recesses. Widespread use of and advances in the technology of echocardiography and cardiac magnetic resonance imaging are increasing awareness of LVNC, and the cardiac magnetic resonance imaging is improving the ability to stage the severity of the disease (Paterick et al., 2012).

Clinical Findings

The use of the echocardiograms is valuable for two separate reasons. First, it can give an asymptomatic patient peace of mind. This means that the noncompacted myocardium, or spongy meshwork, can be traced to a small degree, but that the actual ratio of noncompacted to compacted myocardium is not at the stage that treatment is needed. Secondly, there is no crystal clear answer as to whether the asymptomatic patient will ever have an actual onset of LVNC cardiomyopathy. This disease entity requires close surveillance through echocardiograms so that if changes in the noncompacted to compacted myocardium ratio reach a clinically established number, treatment could begin immediately.

The change in the ratio of noncompacted to compacted myocardium is the basic sign for diagnosing that the asymptomatic patient now should be categorized and monitored for LVNC morphology/cardiomyopathy. There is currently no intervention available to prevent the ratio from increasing. Some of the clinical findings that can be measured are poor pumping performance, or ejection fraction, of the heart, the signs of thromboembolic events, ventricular arrhythmias, and the depression of systolic and diastolic function.

Ejection fraction is a measurement of the percentage of blood leaving the heart each time it contracts. During each heartbeat cycle, the heart contracts and relaxes. When the heart contracts it ejects blood from the two pumping chambers or ventricles. When the heart relaxes, the ventricles refill with blood. No matter how forceful the contraction, it does not empty all of the blood out of a ventricle. Because the left ventricle is the heart's main pumping chamber, ejection fraction is usually measured only in the left ventricle. A normal left ventricle ejection fraction is generally greater than 55%. The ejection fraction may decrease if the heart muscle is weak due to LVNC cardiomyopathy or even long-standing uncontrolled high blood pressure

(Grogan, 2010). One problem with a decreased ejection fraction can be the excessive amount of blood stasis in the ventricle where the heart function is depressed and does not move much which can cause the blood to clot. Parts of this clot can then break off and could cause blockage of a blood vessel which could result in a stroke or peripheral embolization and is categorized as a thromboembolic event.

Another clinical finding that could be present is a ventricular arrhythmia. These arrhythmias include ventricular tachycardia and ventricular fibrillation. Both are life threatening arrhythmias most commonly associated with heart attacks. When an abnormal heart rhythm comes from the lower chambers of your heart, the ventricle, it is called a ventricular arrhythmia. Rhythms of this type include ventricular tachycardia and ventricular fibrillation. Both are life threatening arrhythmias most commonly associated with heart attacks or scarring of the heart muscle from a previous heart attack.

Ventricular tachycardia is a fast heart rhythm that occurs in one of the ventricles of your heart. It is like an electrical short circuit that races in a circle. In a ventricular tachycardia, the heart beat races around the circuit at rates from 150 to 250 beats per minute. As the heart beats faster, it pumps less blood because there is not enough time for the heart to fill with blood between beats. If this fast heartbeat continues, the brain and body may not receive enough blood and oxygen.

Another type of ventricular arrhythmia is ventricular fibrillation. Ventricular fibrillation originates from many different locations in the ventricles, each one trying to signal the heart to beat. In this case, the heart beats much faster than normal, sometimes over 300 beats per minute. The lower chambers quiver instead of contract, and very little, if any, blood is pumped from the

heart to the rest of the body. If a heart is in ventricular fibrillation, the person can become unconscious very quickly. The person might not remember anything that happened just before or during the episode.

If these irregular, fast heart rhythms continue for a length of time, the body will not get enough oxygen-carrying blood. Without oxygen, the brain and body tissues cannot function normally and die.

There are some symptoms of these arrhythmias. If a person is experiencing a fast ventricular arrhythmia, they may feel as though their heart is skipping beats or fluttering. If the rhythm is very fast, they may experience fainting spells, blackouts, temporary blind spots, or dizziness. Eventually, the person may become unconscious and their heart might stop. The diagnosis of ventricular arrhythmias, which are often unpredictable, can be challenging. Arrhythmias can be diagnosed through Holter monitors, electrocardiograms (EKG's) and treadmill stress tests.

Both ventricular tachycardia and ventricular fibrillation are life-threatening heart rhythms. In emergencies, paramedics or medical personnel can treat these arrhythmias with an automated external defibrillator (AED). Long-term treatment options vary according to the type of arrhythmia and individual patient circumstances ("Ventricular arrhythmias -," 2012).

Physical Symptoms

The most common symptoms of LVNC cardiomyopathy are the symptoms of heart failure. Some symptoms of heart failure include the following: difficulty breathing, shortness of breath, chest pain, lightheadedness, limited physical capacity, exercise intolerance, swelling of

the lower extremities or edema, fatigue and distortion of the jugular veins. A chest x-ray usually shows congestion in the lungs from excess fluid buildup. Heart failure is a chronic condition that does not indicate LVNC cardiomyopathy unless accompanied by other symptoms. The additional symptoms that may indicate LVNC cardiomyopathy include heart rhythm disorders called arrhythmias, a very fast heart beat or tachycardia and an incidence of blood clots or embolisms. A person can experience these symptoms from birth or not have any symptoms until later in life, even though LVNC cardiomyopathy is a congenital defect (King, 2011).

Actions and Possible Treatment Plans

Asymptomatic patients should follow a healthy lifestyle. They should minimize alcohol consumption, avoid recreational drugs, eliminate smoking and tobacco products, maintain a healthy weight, have a healthy nutritional plan including appropriate vitamins, fats and oils, obtain an appropriate amount of sleep and a consistent sleep pattern, exercise daily and create a low stress lifestyle. It is also important to have regular physicals and echocardiograms. The patient should live an emotionally healthy and active lifestyle and should not dwell on the fact that there could be heart complications in the future. By having the patient stay on top of their echocardiograms and have them be honest with how their body feels, they should be confident about their healthy lifestyle and enjoy a normal life. If the asymptomatic patient has a change in his or her heart health, the basic healthy lifestyle should continue to be followed but other treatment plans may be added.

There are several possible treatment plans for LVNC cardiomyopathy. Drug therapy may include beta blockers, blood thinners, and angiotensin-converting-enzyme (ACE) inhibitors. The patient may need to have an automatic internal cardioverter defibrillator inserted; some patients may be candidates for a heart transplant. Each actual treatment plan is designed carefully and

uniquely for each individual patient tailored to their specific clinical results and physical symptoms. The benefit for each of the treatments must outweigh the risks.

One type of medication that may be prescribed to help the heart perform more effectively is a beta-blocker. One example of this type of medicine is a drug named carvedilol or Coreg. The weakened heart is wearing itself out, trying to make up for its weak pumping action by pumping more times per minute. Beta-blockers slow the heart rate, which lessens this wearing-out process. Coreg also blocks alpha receptors in artery walls. This relaxes or expands the arteries, lowering the resistance against which the heart pumps, which is the blood pressure. Beta-blockers can also reduce some heart arrhythmias.

Another type of medication that may be used is an antiplatelet or blood thinning medication. An example of this type of medication is Warfarin. Another option is baby aspirin (Hitti, 2007). These medications would help prevent the blood from clotting in the ventricles. The blood that is not being pumped back out of the heart due to a low ejection fraction and weakened heart pools in the ventricle. With the addition of the blood thinning medication, the blood should not clot because the blood is now thinner. This would eliminate or drastically decrease the chance of a piece of a blood clot breaking off and clogging an artery, causing a stroke or peripheral embolization. The addition of a blood thinner helps decrease the likelihood of these thromboembolic events.

Angiotensin-converting-enzyme inhibitors are another category of drugs that can help with heart failure. They are vasodilators; that means they dilate or widen the blood vessels to improve blood flow, which helps to decrease the amount of work the heart has to do. They also block some of the harmful substances in the blood. One of these, angiotensin, is produced as a result of heart failure. Angiotensin is one of the most powerful vasoconstrictor in the body.

Angiotensin-converting-enzyme inhibitors are critical in the treatment of heart failure when systolic dysfunction is present and may also be prescribed for the treatment of diastolic dysfunction. They are also used to control high blood pressure and prevent ongoing heart damage after a heart attack. An example of an ACE inhibitor is a drug called Lotensin.

If detected in the earlier stages, cardiomyopathy may be controlled with long-term drug therapy. Occasionally antiarrhythmia medications or surgery are used in an effort to control or destroy the portion of heart tissue that causes the abnormal rhythms. These treatments may be insufficient and may need to be augmented or replaced in treating the heart when dangerous arrhythmias must be regulated ("Ventricular arrhythmias -," 2012). At this time the placement of an Automatic Implantable Cardioverter Defibrillator (AICD) may be added ("Children's cardiomyopathy foundation," 2012). The procedure involves implanting a small mechanical device under the skin of the chest with wire leads threaded through veins into the heart. It is considered minor surgery, requiring a short hospital stay. Once a defibrillator is inserted, it requires careful monitoring to determine when it needs to be replaced and to ensure that the electrical settings are correct. The decision to implant a defibrillator depends on the specific heart problem. An automatic internal cardioverter defibrillator is used to detect and treat very fast, lethal heart rhythms. Often referred to as an "emergency room in the chest," an AICD will send a small electrical shock to the heart to restore a normal rhythm if a dangerously high heart rate occurs. It may benefit higher risk patients who have experienced serious episodes of fainting, been resuscitated from cardiac arrest, have experienced life threatening arrhythmias or are susceptible to sudden death ("Children's cardiomyopathy foundation," 2012).

If all other methods of trying to control the heart function are not effective the patient may be a candidate for a heart transplant. Heart transplantation is the procedure by which the failing heart is replaced with another heart from a suitable donor. It is generally reserved for patients with end-stage congestive heart failure who are estimated to have less than 1 year to live without the transplant and who are not candidates for or have not been helped by conventional medical therapy (Tigen, Karaahmet, Kahveci, Mutlu & Basaran, 2008). In addition, most candidates are excluded from other surgical options because of the poor condition of the heart. Attempts are made to stabilize the cardiac condition while the evaluation process is undertaken (Botta & Mancini, 2012).

Hereditary Responsibility

Family screening is often not carried out because the focus is on attending to the patient with LVNC cardiomyopathy or grieving the death of a loved one who has succumbed to this disease. Identifying who else may be affected needs to be addressed. In addition to assessing the risk to relatives and siblings, examining the ramifications for family planning is important.

Since LVNC cardiomyopathy can be inherited and present without any signs or symptoms, it is recommended that all first-degree relatives of a patient, such as parents, siblings, and children, be screened. It is also advisable to screen grandparents, aunts, uncles, and cousins. This is especially the case if there is a family history of sudden infant death or sudden cardiac arrest. Even if there is no evidence of the disease, it is advisable to screen more than once with at least a couple of different cardiologists who specialize in cardiomyopathy. For children with an affected family member, parent or sibling, but without symptoms, an echocardiogram and EKG

should be regularly scheduled every 1 to 3 years prior to age 12 and then more frequently from age 12 to 21.

If by early adulthood there is no evidence of LVNC cardiomyopathy, it is unlikely that the condition will develop. However, those with a family history of cardiomyopathy may be advised to continue screening every 5 years throughout life even after the age of 21. These are general screening guidelines that may need to be adjusted on an individual basis by the evaluating cardiologist. Factors that typically influence the frequency of screening include: 1) type of cardiomyopathy diagnosed, 2) family history indicating the likelihood of familial cardiomyopathy and 3) clinical presentation profile of the affected family member ("Children's cardiomyopathy foundation," 2012).

It is not only important to share information and encourage testing of relatives, but also to look back at medical records and autopsies of relatives who have had sudden death. This research can help provide a more accurate family cardiac history and aid in the present for correct diagnosis and treatment plans of the living relatives.

Chapter Three: Conclusions and Recommendations

In summary, this paper focuses on patient education in LVNC cardiomyopathy. This paper describes the importance of echocardiogram testing of the asymptomatic patient and defines the medical terminology so the average family member can actually understand the contents. The following parts were covered: clinical findings, physical symptoms, actions and possible treatment plans and hereditary responsibility.

The existing literature on the topic leads to the following conclusions: through this increased awareness, early detection progression and the prevention of complications, there may be a positive change in the management of left ventricular noncompaction cardiomyopathy. This awareness is critical to the detection and management of LVNC. Patients need to know that they can live a long and healthy life with LVNC morphology/cardiomyopathy. The patients should be proactive in treatments and appointments to ensure the latest medical options are being used in the patient's treatment plan. Obtaining a healthy lifestyle is extremely beneficial in the management of the disease.

Based on these conclusions, it is recommended that family members maintain open lines of communication regarding the status of the LVNC cardiomyopathy disease. Patient educators should encourage family members to be tested and work together as a unit to aid the family in the diagnosis and treatment of this heart condition. By utilizing the newest technologies in heart imaging, early detection can reveal the presence of the disease in asymptomatic patients. This early detection, proper treatment and healthy lifestyle will likely increase the number of asymptomatic patients who can be monitored and should result in better life expectancy and general wellness in living with the disease of LVNC cardiomyopathy.

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Appendix

Educational Readings and Support Groups

Informational readings are available to help the patient understand the disease of left ventricular noncompaction cardiomyopathy. The patient and family are encouraged to continue to research the disease as well as communicate with other people that are impacted with LVNC morphology/cardiology.

Education Readings and Support Groups for LVNC Cardiomyopathy

Articles

- The American Heart Association - http://www.heart.org/HEARTORG/
- The Cardiomyopathy Association - Left Ventricular Noncompaction http://www.cardiomyopathy.org/index.php?id=274
- American Society of Echocardiography - LVNC - A 25-year Odyssey http://www.asecho.org/i4a/pages/index.cfm?pageID=4396

Support Networks

- Contact your health provider to help find support groups in your area
- On-line chat groups for cardiomyopathy
- Hospice to help the patient and family deal with a lifelong disease

Patient Advisory Pamphlet for LVNC Cardiomyopathy

What is Cardiomyopathy?

Cardiomyopathy is a disease of the heart muscle.

What is LVNC Cardiomyopathy?

Left ventricular noncompaction is a rare and primarily genetic cardiomyopathy affecting the left ventricle of the heart. During growth and development in the womb, the majority of the heart muscle is a sponge-like meshwork of fibers. As normal development progresses, these fibers undergo significant compaction that transforms them from spongy to solid. Noncompaction cardiomyopathy results when there is failure of this process of compaction.

What are the symptoms?

The symptoms caused by this condition are not specific and are mainly determined by how much the heart function is affected. Individuals who have the described structural features but normal performance of the heart may be entirely free of symptoms.

Common symptoms associated with a reduced pumping performance of the heart include:

- Breathlessness
- Fatigue
- Swelling of the lower extremities
- Limited physical capacity and exercise intolerance

How is it diagnosed?

Echocardiogram - An ultrasound of your heart that allows your doctor to see each section of your heart, the thickness of the heart muscle, the major blood vessels connected to your heart, your heart valves, and the thin sack around your heart known as the pericardium. This is perhaps the most valuable, painless test to determine the extent of heart disease.

Electrocardiogram (EKG) - This test shows your doctor how your heart is beating and often whether there has been any damage or changes to your heart muscle. Electrical wires are placed on your chest, arms, and legs to record the electrical activity of your heart. This procedure is also painless.

Holter Monitors - By wearing a small portable electrocardiogram, your heart rhythm can be recorded over a 24 hour period. It is a totally painless and safe test.

Cardiac Magnetic Resonance Imaging - Cardiac MRI creates both still and moving pictures of your heart and major blood vessels. Doctors use cardiac MRI to get pictures of the beating heart and to look at its structure and function. These pictures can help them decide the best way to treat people who have heart problems. This is a safe and pain free test.

How is it treated?

Healthy Lifestyle

- Minimize alcohol consumption
- Avoid recreational drugs
- Eliminate smoking and tobacco products
- Maintain a healthy weight
- Obtain appropriate sleep and sleep pattern

Drug Therapy

- Beta Blockers to relax the heart muscle. This helps lower blood pressure and heart rate so that the heart does not have to work as hard.
- Angiotensin-converting-enzyme (ACE) inhibitors to relax blood vessels and lower blood pressure. This helps the heart to pump more blood out to the body.
- Blood Thinners to help keep the blood from clotting and prevent artery blockages and strokes.

Procedures

- Automatic Implantable Cardioverter Defibrillator (AICD) may be surgically inserted to treat abnormal heart rhythms.
- Heart transplantation is a procedure by which the failing heart is replaced with another heart from a suitable donor. This is reserved for patients who have not been helped by conventional therapy.

Is there a cure?

There is currently no medication to treat or cure LVNC cardiomyopathy. Medication therapy treats the symptoms of the disease.

Is this disease genetic?

Yes, this disease is considered primarily a genetic cardiomyopathy. It is recommended that all first-degree relatives of a patient such as parents, siblings, and children be screened by echocardiogram testing. It is also advisable to screen grandparents, aunts, uncles, and cousins.

Will my children inherit this disease from me?

There is no way to know at this point whether this disease will travel to your children. It is advisable to have your children's physicians be aware of this genetic cardiomyopathy and schedule regular echocardiograms to monitor for any signs of LVNC morphology/cardiomyopathy.

What is the Prognosis?

The prognosis for people affected by this condition is difficult to determine and must be individualized. In general prognosis is associated with the degree of cardiac impairment. In severe cases it seems to be no different than the prognosis for any other heart failure. But increased awareness, the early detection of progression and the prevention of complications can really make the difference in improving prognosis.

Knowledge is power!

Patient Cardiologist Appointments

The patient should list cardiologist appointments and any referring doctor in the chart provided. This is a chart that should be copied off and updated. The patient should fill in each visit to help remember what has been done. This aids in planning for further tests and dates. It will help the patient be more in control of the disease and accountable for maintaining their own body.

Patient Cardiologist Visit and Follow-Up

Date:
Physician:
Referring Physician:
Reason for Visit:
Comments:

Problems since last visit:

Echocardiogram:

Other Procedures:

Next appointment date:
Next appointment plan:

Medication Changes:

Weight:
Blood Pressure:
Cholesterol:
Alcohol Use:
Tobacco Use:

Patient Current Medication Information

The patient should list any current or newly-prescribed medications. This listing will help maintain accurate records of dosages and reasons for taking a medication. In the comment section the patient should log any reactions, positive or negative. The patient is advised to make sure pharmacies have a complete listing of the patient's medications.

Patient Current Medication Information

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

Start Date:

Patient Medication History

The patient should list any medications that have been prescribed but usage has been discontinued. This listing will help the patient maintain accurate records of dosages and reasons for taking a medication. In the comment section the patient should log any reactions, positive or negative. Pharmacies and doctors should be notified when a medication has been discontinued.

Patient Medication History

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Medication name:
Medication purpose:
Dosage:

Prescribing doctor:
Comments:

End Date:

Patient Physician History

The patient should list any physicians that are seen, and their location and contact information. This list should include primary doctors, cardiologists, and doctors seen for second opinions. It should include any changes of doctors due to insurance changes. This listing will help maintain accurate record locations to share appropriate health history with other providers.

Patient Physician Contact Information

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Physician name:
Specialty:
Purpose of Visit:
Location:
Contact Information:

Patient Family Cardiology Health History

This chart provides the opportunity for the patient to talk to family members and be open about their cardiac health histories. Everything should be listed in one place to refer back to and share with doctors.

Patient Family Cardiology Health History

Relation	Alive Deceased	Heart Disease	High Blood Pressure	High Cholesterol	Arrhythmias	Sudden Death
Mother						
Father						
Siblings:						
Paternal Grandfather						
Paternal Grandmother						
Maternal Grandfather						
Maternal Grandmother						
Paternal Aunt						
Paternal Uncle						
Maternal Aunt						
Maternal Uncle						
Other						