FDC BEAT

Newsletter of the Familial Dilated Cardiomyopathy Project at Oregon Health & Science University
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Diagnostic Tests: Electrocardiograms, Echocardiograms, & Cardiac Catheterization

Echocardiograms and **electrocardiograms** are the two types of tests the FDC Research Project recommends first-degree relatives (siblings, parents, and children) of a person with idiopathic or familial dilated cardiomyopathy have performed every 3-5 years. Both of these tests are non-invasive and informative for diagnosing early signs of FDC. In this issue of the FDCBeat, we would like to explain how the results of these tests help your health care provider determine if your heart is functioning properly, and if FDC may be in your family.

Electrocardiograms and echocardiograms are important tests for detecting early signs of cardiomyopathy because they assess the electrical system and muscle function of your heart respectively. The heart has four main chambers: a right atrium and ventricle, and a left atrium and ventricle. The heart is responsible for supplying oxygenated blood from the lungs to your body and receiving deoxygenated blood from the body for the lungs to reoxygenate. This process is called the cardiac cycle which occurs as follows: Deoxygenated blood from your body flows into the right atrium, is then pumped into the right ventricle, and out to the lungs. Once oxygenated, blood is pumped into the left atrium and finally to the left ventricle to be pumped out to the body. Blood is pumped out of the heart to the lungs and body during contraction of the heart chambers (**systole**), and flows into the heart during chamber relaxation (**diastole**).

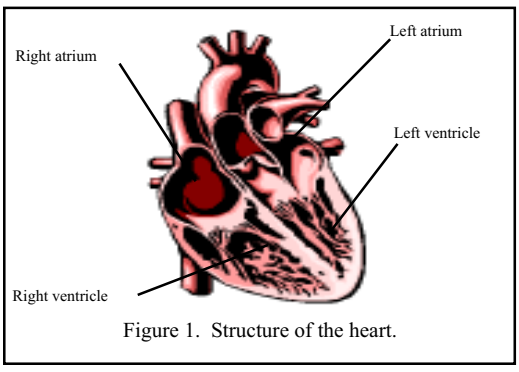

Figure 1. Structure of the heart.

The pumping of the heart muscles is controlled by clusters of heart cells that transmit electrical impulses through the heart. The **sinoatrial node**, the pacemaker of your heart, generates electrical signals that cause the atria to contract. The electrical impulse is then received by the **atrioventricular node**, and then travels to the **AV Bundle of His**, which branches out over the ventricles (known as the **left and right bundle branches**), causing the ventricles to contract. The contraction of the ventricles occurs .1 to .2 seconds after the atria contract. For more detailed information about how the heart functions, see our article in the June 2001 issue of the FDCBeat.

Electrocardiogram. The purpose of an **electrocardiogram** (EKG or ECG) is to assess the electrical activity of the heart. A 12-lead EKG is performed using 12 electrodes. A trained technician, nurse, or doctor will place electrode discs (usually sticky patches) on each leg, upper arm, and the other 8 around the left side of your chest. The electrodes are then clipped onto the discs to record the signals as your heart contracts and relaxes. The electroactivity is displayed as a wavelength on a moving strip of paper. On page 2 is an example of what one cardiac cycle or heartbeat looks like on an EKG.

The small P peak corresponds to the contraction of the upper heart chambers (right and left atria), called the P wave. This is followed by the QRS complex which tracks the electrical

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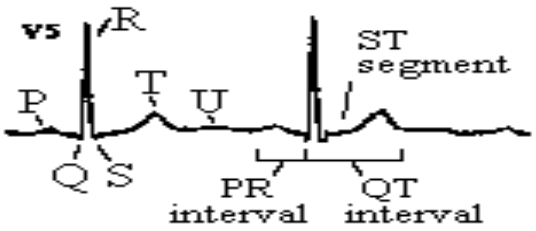
impulses through the lower heart chambers when they contract (right and left ventricles). The third peak,T wave, is the relaxation period of the lower chambers.

EKG reports will include a measurement of the heart's rate and rhythm. Rhythm abnormalites are determined by measuring the length of time it takes electrical signals to travel to different parts of the heart, illustrated by the distance between peaks, such as the PR interval (beginning of the P wave to the start of the QRS complex) and QRS intervals. Other intervals may also be calculated and recorded on an EKG report.



A health care provider evaluating an EKG report would look at all 12 leads to check for normal appearance and size of the P wave, QRS complex, and T wave. Abnormally long intervals may mean that the heart's electrical signals that control chamber contraction are delayed. This is known as **heart block,** such as an **AV block** (delayed communication between the atria and ventricles, shown as a long PR interval), and a **Bundle branch block** (delayed contraction of the left and/or right ventricle, seen as a wide QRS complex). These **conduction system abnormalities**, as well as others, may be accompanied by symptoms of dizziness, fainting, loss of conciousness, or no symptoms.

EKG abnormalities may be early signs of FDC/IDC. Sometimes, computer generated EKG reports diagnose potential problems; however, a health care provider should review the printout taking into account your symptoms and health history to confirm a diagnosis and determine appropriate follow-up.

Echocardiogram. The purpose of an **echocardiogram** (echo) is to take a picture of the heart while it is pumping to determine the size of each chamber, thickness of the ventricle walls, pumping function, function and structure of the valves, and the blood flow through your heart.

An echo is performed using ultrasound: a transducer that sends and receives sound waves is placed on your chest near your heart and a visual image is created and displayed on a monitor.

An echo is performed by a highly trained technician and video taped. A cardiologist that specializes in interpreting echocardiograms reviews the images and makes a conclusive summary in a written report.

Echo reports usually include abbreviations such as **LVEDD** (left ventricular end dimension at diastole) or **LVIDd** (left ventricular internal dimension at diastole). Both of these abbreviations correspond to the size of the left ventricle chamber at relaxation. The abbreviation **LVEDS** (left ventricular end dimension at systole) or **LVIDs** (left ventricular internal dimension at systole) is the size of the left ventricle when it is contracting. Here at the FDC Research Project, we compare the LVIDd with a chart that shows the percentage of people the same height and sex that would have a heart size as large as yours. If the LVIDd measurement is above the 95th percentile, meaning 5% or less have a heart size larger, the left ventricle is considered dilated.

Other abbreviations include **LVPW** and **LVSW**; these are measurements of the left ventricular outer and inner wall thicknesses (posterior and septal). A thickened left ventricular wall can be associated with another type of cardiomyopathy called **hypertrophic** cardiomyopathy. Many of the symptoms that occur with IDC or FDC also occur with hypertrophic cardiomyopathy. In the February 2002 issue of the FDCBeat, the front-page article discusses hypertrophic and other types of cardiomyopathy.

During an echo the **ejection fraction** is also estimated. An ejection fraction is the percentage of blood pumped out of the left ventricle to the body during the contraction. A normal ejection fraction is above 55%. Below 50% is abnormally low and is referred to as **left ventricular systolic dysfunction**. This means

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Peripartum Cardiomyopathy and FDC

Peripartum (meaning around birth) **cardiomyopathy** (**PPCM**) is a form of dilated cardiomyopathy that affects women and develops during or after pregnancy. PPCM occurs with approximately 1 in every 3000-4000 births. A diagnosis of PPCM is made when a woman with no prior history of heart problems develops unexplained heart failure, usually in the last month of pregnancy or within 5 months after delivery. An echocardiogram will reveal a low ejection fraction or left ventricular systolic dysfunction (see previous article for a detailed discussion of these echo terms). PPCM may easily go unrecognized, as some symptoms of heart failure - shortness of breath, fatigue, leg/ankle swelling - are common in the later months of normal pregnancy. The severity of PPCM, like FDC, is very variable, with cardiomyopathy resolving in about half of women, while the other half experience continued heart failure that may be life threatening or require transplantation.

The cause of PPCM is unknown. There are reports of familial PPCM (PPCM in more than one family member) in the medical literature. In addition, amongst research families we have noted both FDC (unexplained cardiomyopathy in men and non-pregnant women) and PPCM within the same family. This leads us to believe that at least some percentage of PPCM may actually be an expression of FDC occurring in pregnancy, and in these cases may have an underlying genetic cause.

Based on this thinking, the recommendations given to family members of individuals with FDC or idiopathic dilated cardiomyopathy (IDC) may also apply to women with PPCM. Women with a diagnosis of PPCM should be asked about their family history of heart problems, and echocardiogram and EKG screening of first-degree relatives (parents, siblings, children) should be considered.

We hope to learn more about the possible association of PPCM and FDC as we continue our research and enroll new families. If you or anyone in your family has been diagnosed with PPCM, please let us know!

New Publication:

A novel lamin A/C mutation in a family with dilated cardiomyopathy prominent conduction system disease and need for permanent pacemaker implantation. 2002. RE Hershberger, EL Hanson, PM Jakobs, H Keegan, K Coates, S Bouman, M Litt. *Am Heart J* 144(6):1081-1086.

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FDC BEAT Newsletter

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that your heart is not pumping enough blood out of the left ventricle to the body during contraction.

Cardiac Catheterization. There are many other types of cardiovascular testing that a health care provider can request. **Cardiac catheterization/Angiogram** (CC) is sometimes performed after EKG, echo, and other cardiac tests, to further assess and diagnose the cause of cardiomyopathy. With CC, a tube (catheter) is inserted usually in an artery in the groin and guided through to the coronary arteries, the blood vessels that supply blood to the heart itself. A dye is then injected through the catheter and X-rays are taken to follow the flow of the dye through these arteries. An angiogram provides an accurate picture of the coronary arteries, and will detect blockages in the arteries (**coronary artery disease, or CAD**). If a person is younger or does not have risk factors for coronary artery disease (family history, high cholesterol, diabetes, high blood pressure, smoking. etc.), CC may not be done.

Screening & The FDC Project. Screening is an important step for family members because many of the EKG and echo abnormalities can occur without symptoms. EKG abnormalities or early signs of left ventricular enlargement or systolic dysfunction may benefit from early treatment.

A diagnosis of FDC is made by our research project when two or more people within a family have **chamber enlargement (an LVIDd above the 95th percentile), systolic dysfunction and other possible causes of cardiomyopathy (such as CAD or hypertrophic cardiomyopathy) are excluded**. Since we are unable to visit and screen every family that is involved in our project, we request medical records. When reviewing records we look for EKG, echo, and CC reports. These tests, as well as others help your health care provider determine a cause for your cardiomyopathy. As always if you or any of your family members have had any of these tests performed, please let us know! You can contact us at our toll free number: 1-877-800-3430 or via our website: www.fdc.to .

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